Endoscopic Treatment of Arachnoid Cysts

16

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Arachnoid cysts are a congenital malformation, secondary to splitting or duplication of the arachnoid that becomes filled with CSF [1]. They typically arise within the arachnoidal cisterns, in most cases in the middle fossa at the sylvian fissure (30–50 %). Ten percent arise on the hemisphere convexity, 10 % in the suprasellar cistern, 10 % in the quadrigeminal cistern, 10 % in the cerebellopontine angle, and 10 % in the midline of the posterior fossa [1]. Midline cysts (suprasellar, interhemispheric, quadrigeminal, and posterior fossa) are usually associated with hydrocephalus and present earlier in life, usually during the first 10 years.

Symptomatic cysts typically present with local mass effect on neural tissue, obstruction of CSF flow, and macrocephaly in younger children. Most frequent symptoms are headache, head bobbing, focal neurologic deficits, seizures, and psychomotor retardation. An increased risk of posttraumatic extradural and subdural hematomas/hygromas is associated to the presence of the cyst. In symptomatic cysts, indication for treatment is clear. In infants, a pathologic increase (symmetric and asymmetric) in head circumference is a surgical indication. However, considerable debate continues to exist in the management of asymptomatic, incidentally discovered cysts. Because of the concern for impairment of brain development, in the pediatric population, treatment should always be recommended at the time of discovery unless the cyst is of a small size with minimal distortion of surrounding tissues and has been discovered incidentally [2, 3]. Cysts that distort surrounding neural tissues may be responsible for alteration in cerebral blood flow, thus explaining the atrophy often observed in the adjacent neural tissue.

Multiple surgical strategies have been developed in the management of symptomatic arachnoid cysts. These include endoscopic resection of the cyst wall with establishment of communication between the cyst and the contiguous CSF pathway (ventricles or basal cisterns), craniotomy with microsurgical fenestration, and shunting of the cyst in the peritoneum. The advantage of neuroendoscopy is that it avoids major surgical procedures, such as craniotomy with the risk of some major complications, and it avoids shunt dependence with all of the shunt-related problems [4, 5]. The choice between shunt and neuroendoscopy must be made on the basis of neuroradiological imaging to detect an area of contiguity between the cyst wall and the ventricular ependyma or subarachnoid spaces. This should be opened under endoscopic control to allow continuous drainage of the cyst. To avoid

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reclosure of the stoma, large openings, at least 10–15 mm in diameter with removal of cyst wall fragments, should be performed [5]. The success rate (control of symptoms and cyst size without other surgical procedures) amounts for 71–81 % in endoscopic series [5–9]. The treatment is usually easier when the cyst obstructs the CSF pathway and hydrocephalus is associated, due to the possibility of working in larger spaces.

Surgical technique and results vary according to the location of the cysts. Best results have been achieved with suprasellar cysts. On the contrary, in case of arachnoid cysts of the middle fossa, indication to endoscopy is controversial. The arterial vessels of the Sylvian fissure may course within the medial wall of the cyst in proximity of the sites where fenestrations should be made. Therefore, some authors prefer microsurgery to endoscopic surgery for the arachnoid cysts in this location.

For infants with large hemispheric arachnoid cysts, the success rate of endoscopic fenestration appears to be less favorable than in older children [10]; however, in young children also, shunting is burdened by high rate of failure, and therefore, a neuroendoscopic approach should be advocated if possible. The possible need of a secondary cyst-peritoneal shunting in case of neuroendoscopy failure is always discussed prior to the intervention [10].

16.1 Middle Fossa Cyst

The middle fossa is the most common location for arachnoid cysts. These cysts may be responsible of a wide range of signs or symptoms, which include headache, focal neurological deficits, macrocrania, and hydrocephalus. They may also present with "functional" symptoms such as seizures and developmental delay [12].

16.1.1 Indication for Surgery

Indication for surgery in case of middle fossa cyst is highly debated. In the presence of symptoms and signs related to the cyst (headache, intracranial hypertension, macrocrania, hydrocephalus, focal neurological deficits), indication for surgery is clear. In case of "functional" symptoms, such as epilepsy and neurodevelopmental delay, indication for surgery is less clear. Some authors [12] suggest efficacy in postoperative seizure control and improvement in development, although direct clinical correlation of these symptoms to the cysts may be tenuous. In the series of Di Rocco et al. [13], patients with seizures were treated only if they also presented headache and/or macrocrania and/or focal neurology and/or progressive increase in size of the cyst. They observed good outcome of seizures in these patients. On the contrary, patients with developmental delays did not improve. In our opinion, patients with functional symptoms and mass effect on neuroradiological imaging are candidates for surgery, especially if they are young babies, with the aim to allow better brain development.

The debate of indication for surgery in asymptomatic young children with very large cyst with mass effect is still open. In our opinion, the younger the patient, the stronger is the indication for surgery.

In the recent paper of Choi et al. [15], the reasons for surgery (endoscopic or microsurgical fenestration) were categorized into three groups. In one group, surgery was necessary because the symptoms were related to the cyst (hydrocephalus, intracranial hypertension). In the second group, the symptoms were ambiguous, thought to be correlated with the cyst (headache, dizziness, large head, abnormality of skull, seizures, strabismus, or developmental delay). The patients in the third group presented minimal or no clinical symptoms, but underwent surgery with the expectation of improvement of abnormal findings in radiological or other examinations (huge cyst or enlarging cyst on follow-up image, abnormality in neuropsychologic tests, severe perfusion defect of adjacent brain parenchyma in single-photon emission computed tomography – SPECT). Interestingly, while the improvement rate in the first group was satisfactory, in the second group (the cases that showed ambiguous correlations between cysts and symptoms), it was

very low. The improvement rate in this category was only 11 %, while the complication rate was as high as 43 %. In the third group, the radiological appearance of the cyst improved in 50 % of patient, and abnormality of neuropsychological tests improved in 35 %, while in no patients, there was improvement in SPECT evaluation. Therefore, they suggest surgical interventions only for those patients with symptoms indisputably related to the cyst.

Risk of traumatic hemorrhage after head injury is well known in patients with middle fossa cyst. However, surgery appears not to reduce the risk of traumatic hemorrhage. Surgery should not be proposed to prevent traumatic hemorrhage.

16.1.2 Selection of Candidates to Endoscopic Procedure Versus Alternative Treatments

Careful evaluation of preoperative imaging is crucial for considering endoscopy in case of intracranial cysts. Best evaluation is by MRI, especially with the modern CISS or DRIVE sequences that offer extraordinary contrast between CSF and brain parenchyma. The Prerequisite for safe endoscopic fenestration is the presence of enough space to safely reach the site of fenestration and the presence of a thin, not vascularized membrane to be fenestrated. In case of middle fossa cyst, microsurgical fenestration is also a straightforward procedure, with low operative risk and no need to cross the brain parenchyma. The choice between microsurgical and endoscopic fenestration should be well pondered: a minimal invasive procedure should be chosen only if it offers the same safety and the same success rate. Middle fossa cysts are usually classified in accordance with the Galassi classification [14]: type I cysts are small, spindle shaped, and limited to anterior middle cranial fossa; type II cysts show superior extension along sylvian fissure with displacement of the temporal lobe; type III cysts are large and fill the whole middle cranial fossa, with displacement of not only the temporal lobe but also the frontal and parietal lobes. Type I cysts are usually asymptomatic and only rarely treatment is necessary. In case of indication for surgery, endoscopic approach is not the ideal treatment, because the space for endoscopic maneuvers is limited. Moreover, it is necessary to cross the brain parenchyma to reach the cyst. Microsurgical fenestration is the best option in this kind of cysts. Type II and III cysts are very large with enough space for the endoscope to reach the target. The choice between endoscopic and microsurgical fenestration should be based on the appearance of the chiasmatic and interpeduncular cisterns on MRI. In case of large cisterns with thin membranes and enough space between carotid artery and optic nerve and/or tentorial notch and third cranial nerve, endoscopic surgery should be considered a valid option; otherwise, microsurgical fenestration should be preferred. Microsurgical techniques allow safer dissection of deep, thick membranes near vital structures, such as carotid artery and its branches, optic nerve, oculomotor nerves, and brain stem. In every case, the surgeon following endoscopic inspection of the deep membranes of the cyst should be ready to convert his procedure into microsurgery. This should be well explained to the patients and/or their parents in the informed consent. Several factors should be considered before performing endoscopic fenestration. Once within the cyst, the arterial vessels of the Sylvian fissure can be seen coursing the medial wall. They can be followed down to the basal cisterns, where the fenestrations can be made. The membrane of these cysts may be rich in collagen and can be difficult to penetrate, so that scissors or sharp instruments can be used to cut an opening. Often, the stoma should be enlarged with sharp instruments with some risks for the vessels of the Sylvian fissure. Open surgical procedures permit bimanual manipulation and the use of regulated suction to manipulate such thick and vascularized arachnoid membranes. Bleeding can be more easily controlled with traditional methods such as bipolar cautery and topical application of hemostatic agents (Fig. 16.1a–f).

Shunting, as the first surgical procedure, should be avoided in middle fossa cysts, because of the well-known risk common to all shunts (infection, mechanical dysfunction) and the

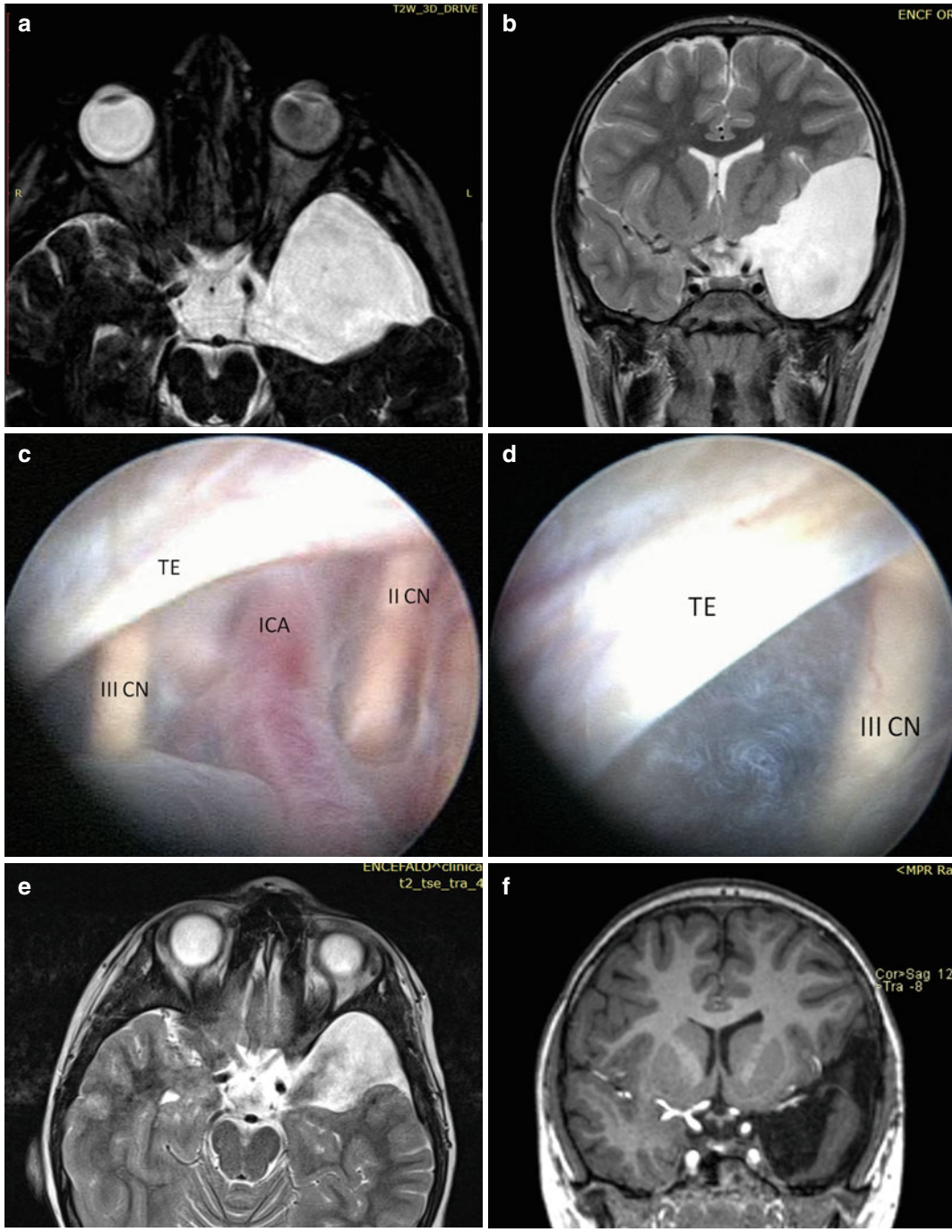


Fig. 16.1 (**a**) Arachnoid cyst of the middle fossa. Note the inner layer bulging medially behind the carotid artery. (**b**) Same patient as shown in (**a**), coronal view, where a large area of possible fenestration is easily identified. (**c**) Endoscopic view, where nerves and arteries are easily identified through the translucent layer of the cyst and delimitate areas of fenestration. (**d**) A safe, posterior window of fenestration is identified between the free edge of the tentorium and the third cranial nerve. (**e**, **f**) Postoperative MRI showing good flow artifact behind the carotid artery on axial T2 turbo spin echo sequences (**e**) and reduction of the cyst volume on coronal T1 (**f**)

higher probability of overdrainage, with slit cyst syndrome and secondary Chiari malformation that can be challenging to treat.

16.1.3 Authors' Preferred Surgical Technique

We use a surgical technique similar to those described by Di Rocco et al. [13] and Spacca et al. [12]. The aim of the procedure is to establish communication between the cyst and interpeduncular or carotid cistern (cyst-cisternostomy). The site of fenestration is between the optic nerve and the carotid artery, between the carotid artery and the oculomotor nerve, and between the third nerve and the free edge of tentorium. Electromagnetic neuronavigation is a very useful tool in order to choose the optimal entry point and to plan the best trajectory. It also provides real-time control of the endoscope position. The patient is positioned supine with the head tilted contralaterally. A small incision is performed over the temporal muscle behind the hairline. A burr hole is drilled directly above the cyst, avoiding residual cerebral mantle. The dura mater is opened with a knife to favor dural closure at the end of the procedure. A 30° free-hand rigid scope (Storz ®, Tuttlinger, Germany) is inserted within the cyst. The landmarks for the orientation are the free edge of the tentorium, the sylvian fissure with the arterial trunks, and the cranial nerves. The site of fenestration is decided on the basis of the aspect of the deep membrane (thickness and transparency). Usually the safest site is between the tentorial edge and the third cranial nerve. It allows to reach the interpeduncular cistern with good visualization of the basilar artery. Whenever possible, several holes within the deep membranes are performed. Based on the experience of Karabagli, multiple perforations are associated with decreased risk of cyst regrowth [19]. The best way to fenestrate the membranes is using Tulium LASER coagulation and forceps. Scissors should be used with great caution, because of the risk of arterial bleeding from small vessels. The stoma is also enlarged using the double-balloon catheter (neuro-balloon catheter from Integra Neurosciences®, Antipolis, France). Care is taken to open all layers. The cyst-cistern communication is considered satisfactory when it is possible to directly view the basilar artery and cerebrospinal fluid (CSF) pulsation through the fenestration. At the end of the procedure, the endoscope is removed. The dura mater is closed. The muscular fascia and the superficial layers are sutured in a standard fashion.

16.1.4 Follow-Up

CT scan or MRI is obtained until 24 h following operation to exclude major complication, in particular subdural effusion. MRI is planned in the first few weeks to confirm the presence of a flow artifact through the fenestration.

After hospital discharge, the patients are regularly controlled (clinical examination and brain MRI) 3 and 6 months and 1, 2, and 3 years after surgery.

Postoperative radiological appearance is very variable: in most successful cases, the cysts remain unchanged, or with minimal reduction of mass effect on surrounding tissue. The flow of CSF through the cyst- cisternostomies is confirmed by signal voids around ICA or tentorial edge on postoperative T2-weighted coronal MR imaging studies (*flow artifact*). There is no concordance between clinical and radiological outcome: patients who show radiological improvement not always demonstrate a corresponding improvement of clinical symptoms, especially if the reason for surgery was not clearly related to the cyst. Conversely, the patients who show clinical improvement, not always have radiological improvement [15].

Disappearance of the cyst following fenestration is very rare. In Choi's series [15], the cyst disappeared in 4/39 cases (10 %) and significantly decreased in 16/39. Sometimes the decrease of the cyst may be associated with the increase in size of the ventricles (especially at the level of the temporal horn) and presence of subdural fluid collection.

16.1.5 Complications

The main complication of the endoscopic series is subdural hygroma, with an incidence as high as 40 % of cases [4, 13, 15, 17]. This complication also occurs in microsurgical series and often requires surgical treatment with subduro-peritoneal shunt.

Leakage of CSF is another important complication, which has been reported to occur in 3.9–6 % of patients both in endoscopic and microsurgical series [16, 18]. Often, patients with CSF leaks are younger than 1 year of age and need a CSF diversion procedure.

Other surgical-related complications are subcutaneous collection, CSF infection, and third cranial nerve palsy.

16.1.6 Results

Good clinical outcome with complete recovery or significant improvement has been achieved in more than 90 % of cases in several series [12, 13, 19]. Relief of headache and other neurological deficits was obtained in all series but that of Choi et al. [15], in which headache was not always related to the presence of the cyst. These data are similar to those achieved by Levy et al. [18] following microsurgical fenestration. In the series of Choi et al. [15], the results are not so satisfactory, especially in infants, where the rate of surgical failure (need for additional operations such as a shunt or a second fenestration procedure) was 50 % (3/6). These results were also noted by others, so that some authors consider cyst-peritoneal shunt more effective in the infant group. In our opinion, the advantage of being shunt-free overcomes the risk of second surgery in the infant population, in which also the shunt-related problems (high rate of shunt revision, lifelong shunt dependency, overdrainage, and even brain herniation) are more frequent.

A second line of repeat endoscopy, craniotomy, or shunt may be considered in case of failures. In recurrent middle fossa cysts, we rarely perform repeat endoscopy and we prefer to offer craniotomy and microsurgical fenestration in case of absent flow artifact on neuroimaging (with the aim to achieve larger fenestrations of the deep membranes). If the fenestration appears to be patent on neuroimaging but symptoms are still present, cyst-peritoneal shunt is the preferred option, especially in younger children.

In conclusion, recent reports indicate that increasing number of authors prefer endoscopic approach as the first-line management of middle fossa arachnoid cysts [12, 13, 19–21]. However, larger randomized series are needed to identify the real advantages in managing middle fossa cysts by endoscopy, rather than alternative techniques (Table 16.1).

16.2 Suprasellar Cysts

Endoscopic surgery has radically changed the management of deep-seated arachnoid cyst. Patients with arachnoid cysts in the suprasellar region, especially if associated with hydrocephalus,

Table 16.1 Review of case series for pure neuroendoscopic treatment of middle fossa cysts

Authors	Cohort size	Results	Complications
Elhammady et al. [20]	6	Six successful	Subdural hygroma ($n=3$)
Di Rocco et al. [13]	17	Two recurrences	Venous bleeding ($n=1$)
Spacca et al. [12]	40	Satisfactory outcome reported in 92.5 % cases; cyst reduction in 72.5 % of cases; 10 % requiring second procedure	Subdural hygroma ($n=4$)
El-Ghandour [22]	32	Clinical improvement reported in 87.5 % cases; reduction in cyst size in 71.9 % cases; three cases of recurrence	Subdural hygroma ($n=2$), CN III palsy ($n=1$)
Karabagli and Etus [19]	20	Eighteen successful cases (in three endoscopy was repeated); two failures (CP shunt)	Asymptomatic subdural hygroma ($n=1$)

From Choudhri et al. [10], modified

are ideal candidates to endoscopic surgery, which allows wide fenestration of the cyst, both in the ventricular system and in the cisternal spaces [8], avoiding major surgical procedures, such as craniotomy and transcallosal approach. For this kind of cysts, endoscopic surgery is the treatment of choice, considering repeat endoscopy in case of failures and reserving shunting only to refractory cases. Typically, the suprasellar cysts elevate the floor of the third ventricle, often appearing just under the body of the lateral ventricle. Hydrocephalus due to obstruction of the CSF pathways either at the foramen of Monro or at the level of the cerebral aqueduct is often associated. Miyajima et al. [23] identified two different types of suprasellar arachnoid cyst, according to the position of the basilar artery. In the first type, the basilar artery is inside the cyst. They speculated that these cysts arise from invagination of the membrane of Liliequist. In the second type, the basilar artery is pushed posteriorly by the cyst. Their interpretation was that these cysts arise from cystic dilatation of the interpeduncular cistern.

16.2.1 Indication for Surgery

Suprasellar cysts are rare. They represent 8–15 % of all intracranial cysts. Treatment is indicated in case of symptomatic cysts or in case of coexisting hydrocephalus. The majority of cysts become symptomatic in early childhood, usually presenting with hydrocephalus and symptoms and signs of increased intracranial hypertension. Other frequent symptoms are visual field defects/impaired visual acuity (about 30 % of cases) and endocrinological disorders (about 60 % of cases) secondary to compression of the chiasm and hypothalamic-pituitary axis. Head bobbing in children is a rare feature, but quite typical of suprasellar cyst. Recurrent seizures are also indication for surgery.

Incidentally discovered cysts are usually not candidates for surgery, even if demonstration of cyst growth or the presence of neural compression, especially in children, should be an indication for consideration of surgical treatment to allow the potentially normal development and function of the adjacent brain [11].

16.2.2 Selection of Candidates to Endoscopic Procedure Versus Alternative Treatments

The location of the suprasellar may favor a different surgical treatment compared with cysts in other locations. The cysts are always in close relationship with the third ventricle; so, in case of sufficient ventricular dilatation, endoscopic treatment is the treatment of choice [24]. Usually, the cyst can be approached from the ventricles through a standard precoronal burr hole. Shunt placement is no longer suggested because of the need for foreign body implantation and the concomitant risk of shunt infection, failure, or lifelong shunt dependence. It should be reserved only to refractory cases.

Open surgical approaches are associated with a relatively higher surgical morbidity compared with endoscopic procedures and have success rate that does not exceed 70 % [11]. Microsurgery, through a subfrontal or pterional approach, should be reserved to those few cases not associated with ventricular dilatation. In the very rare cases in which the cyst expands from the suprasellar region to reach laterally the temporal fossa, the cyst can be approached with endoscope from a temporal burr hole, even if hydrocephalus is absent.

Different techniques have been advocated for endoscopic fenestration of suprasellar cysts. Some have advocated fenestration only of the apical membrane, usually at the level of the Monro foramen, between the ventricle and the cyst (ventriculocystostomy). Others suggested to also perform concurrent basilar fenestration toward prepontine cistern (cyst-cisternostomy), realizing a ventriculocystocisternostomy (VCC). Decq et al. demonstrated by MR-imaged CSF flow dynamics the importance of fenestrating suprasellar cysts both in the ventricles and in the basal cisterns, to prevent secondary closure of the opening and recurrence of symptoms [8]. The passage of CSF pulse waves in the cyst's cavity through sufficiently large windows in a "bipolar" fashion lowers the risk of re-formation of the cyst's membrane [8, 11]. Multiple fenestrations into all arachnoid cisterns that are accessible

should lead to a lower rate of recurrence at long-term follow-up than do less aggressive methods, such as single fenestration [11]. Also the more recent paper from Maher and Goumnerova [24], which summarized their experience with 44 published cases, concluded that endoscopic ventriculocystocisternostomy is more effective than ventriculocystostomy.

16.2.3 Authors' Preferred Surgical Technique

Our surgical technique is similar to that described by Kirollos et al. [11]. Under general anesthesia, a frontal burr hole is drilled 3–4 cm from the midline (usually on the right side, or on the larger side, in case of asymmetrical ventricular dilatation) and on the coronal suture. The ideal position of the entry point and the best trajectory is selected on the basis of preoperative MR imaging. Neuronavigation is useful but usually not mandatory in standard cases, unless the ventricular system is small. The lateral ventricle is tapped, and the endoscope is directed toward the foramen of Monro, where the dome of the cyst is usually protruding into the third ventricle and comes into view. A fenestration is made between the cyst and the ventricle with various techniques, and we prefer to use Tulium LASER coagulation and scissors. Wide fenestration, at least 10 mm in diameter, is achieved, with coagulation of the apical portion of the cyst and removal of the cyst wall if possible. The cyst is then entered with the endoscope to visualize the basal wall of the third ventricle and the position of the basilar artery (inside the cyst, or outside the cyst, pushed toward the brain stem). It is usually possible to observe all the anatomical structures around the interpeduncular cistern through the thin inner layer of the arachnoid cyst, such as the basilar artery, internal carotid artery, posterior communicating arteries, pituitary stalk, optic chiasm, and third cranial nerves. Several openings into the basal prepontine cistern should be created, usually between basilar artery and third cranial nerves from both sides. The openings can be done using a probe without the application of any current and enlarged with balloon catheters. The endoscope is then advanced through the fenestration to visualize the neurovascular structures in the basal cisterns and to ensure the creation of adequate communication between the cyst and the subarachnoid space.

In the rare cases in which suprasellar cyst expand laterally toward the temporal fossa, the cyst can be approached directly through a temporal burr hole, like middle fossa cyst. Once inside the cyst with the endoscope, a cyst-cisternostomy can be performed in standard fashion, trying to make multiple perforations, on both sides of the basilar artery (Figs. 16.2a, b and 16.3a–j).

16.2.4 Follow-Up

The follow-up is similar to middle fossa cyst: a neuroradiological investigation (preferably MRI) is obtained until 24 h following operation. The presence of flow artifact through the fenestration at MRI should be addressed before discharge from the hospital. After hospital discharge, the patients are regularly controlled 3 and 6 months and 1, 2, and 3 years after surgery.

The reduction in arachnoid cyst size following endoscopy, also for cysts in this location, is variable. The indications for further intervention depend on the persistence of the patient's symptoms and not upon the appearance of the cyst on postoperative imaging [11].

Endocrinological disorders usually persist following treatment despite the satisfactory decrease in volume of the cyst [11].

16.2.5 Complications and Results

Reported complication rate is very low in this kind of surgery [10], with few addressed cases of ventriculitis and subdural hygromas. The success rate is high. Maher and Goumnerova [24] in their review calculated a success rate (no need for subsequent surgical procedures) of 86 % when only ventriculocystostomy (VC) was performed, and that increased to 92 % when ventriculocystocisternostomy (VCC) was performed as first procedure.

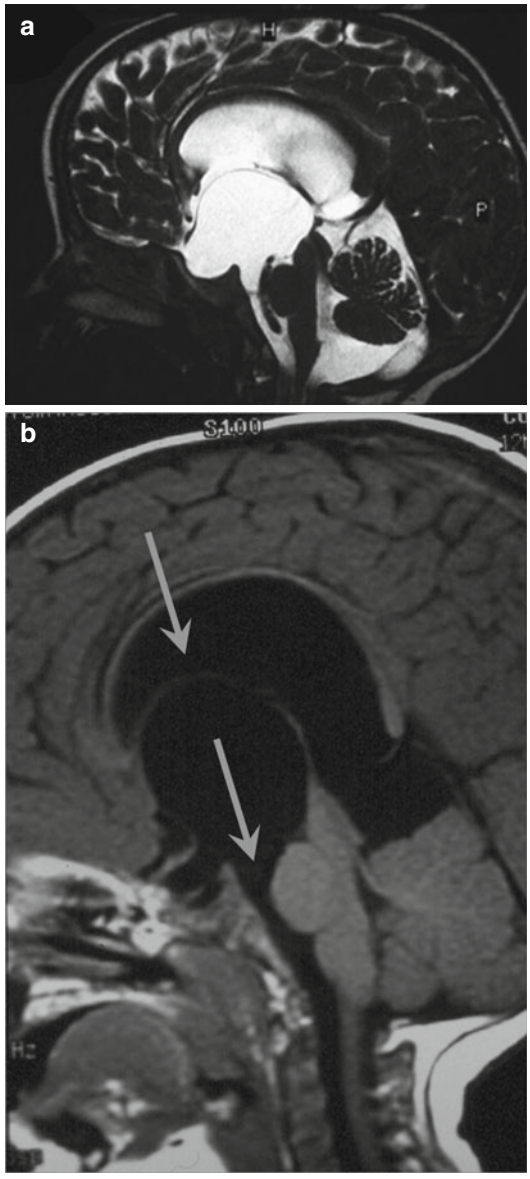


Fig. 16.2 (**a**) Type 1 suprasellar cyst, bulging into the third ventricle and occluding both foramina of Monro and the inlet of the aqueduct. (**b**) Surgical trajectory (*arrows*) for ventriculocystostomy and cyst-cisternostomy through a coronal burr hole

They analyzed separately the success rate of VC or VCC, when they were performed following a prior shunt treatment. VC alone was successful in 64 % of cases, while VCC was successful in 88 %.

Recently, Rizk et al. [25] reported a long-term clinical follow-up of six patients treated with endoscopic techniques, with an average of 46.5 months. Apart from one patient who developed ventriculitis that was successfully treated with antibiotics and temporary external drainage, no patients suffered recurrence of symptoms, abnormal imaging findings, or any sequelae of the surgical intervention.

In conclusion, endoscopic treatment of suprasellar cysts is safe and effective and should be considered in each case with compatible anatomy, in patients already treated with shunts and also in case of failure of the first endoscopic procedure. In these cases, larger fenestrations, with removal of cyst wall, especially at the level of the basal membrane, should be done by experienced neuroendoscopists.

16.3 Interhemispheric Cysts

Interhemispheric arachnoid cysts are congenital malformations usually associated with complete or partial agenesis of corpus callosum. They should be distinguished from other types of cystic lesions that can occur in the interhemispheric fissure and are part of more complex brain malformation, such as holoprosencephaly, diencephalic cyst with upward extension of the third ventricle, and porencephalic cyst. These cystic malformations are in communication with the ventricular system, do not need specific treatment, and have a very bad cognitive prognosis [26, 27].

Sometimes they can be confused with convexity cysts with interhemispheric extension (Fig. 16.4a). Often interhemispheric cysts are associated with ventricular enlargement. Various mechanisms are implicated: downward displacement of the foramina of Monro, obstruction of the aqueduct, or impairment of the resorption mechanism over the convexity.

Generally, interhemispheric cysts are diagnosed in the first year of life. Head enlargement, seizures, and psychomotor retardation are the most frequent presenting symptoms. These symptoms, however, may also result from the associated malformations (agenesis of the corpus callosum, gyral abnormality, and neuronal

heterotopia). The cysts can be diagnosed antenatally, during routine ultrasound evaluation. They can remain silent for many years (Fig. 16.4b, c).

16.3.1 Indication for Surgery

As for cysts in other locations, surgery is indicated in case of symptomatic cysts. For patients with progressive signs of increased intracranial pressure or progressive cyst or ventricle enlargement, surgery is indisputably indicated. The indication for surgery is less established in patients with epilepsy and psychomotor retardation that can be attributed to the associated brain malformation. Usually, in young children with large cyst, associated to ventricular dilatation, surgery should be considered also in the absence of specific symptoms. Asymptomatic patients, for whom a conservative treatment is chosen, require

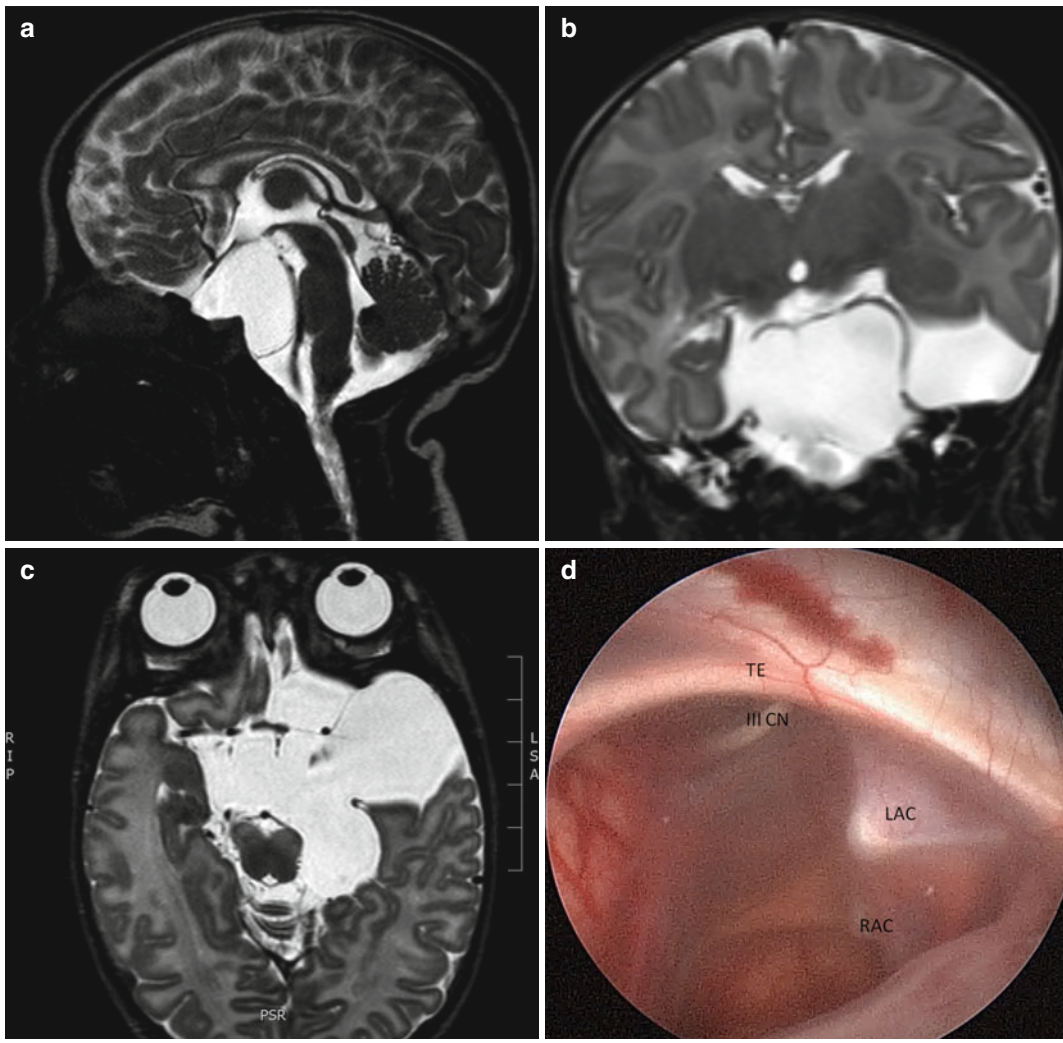
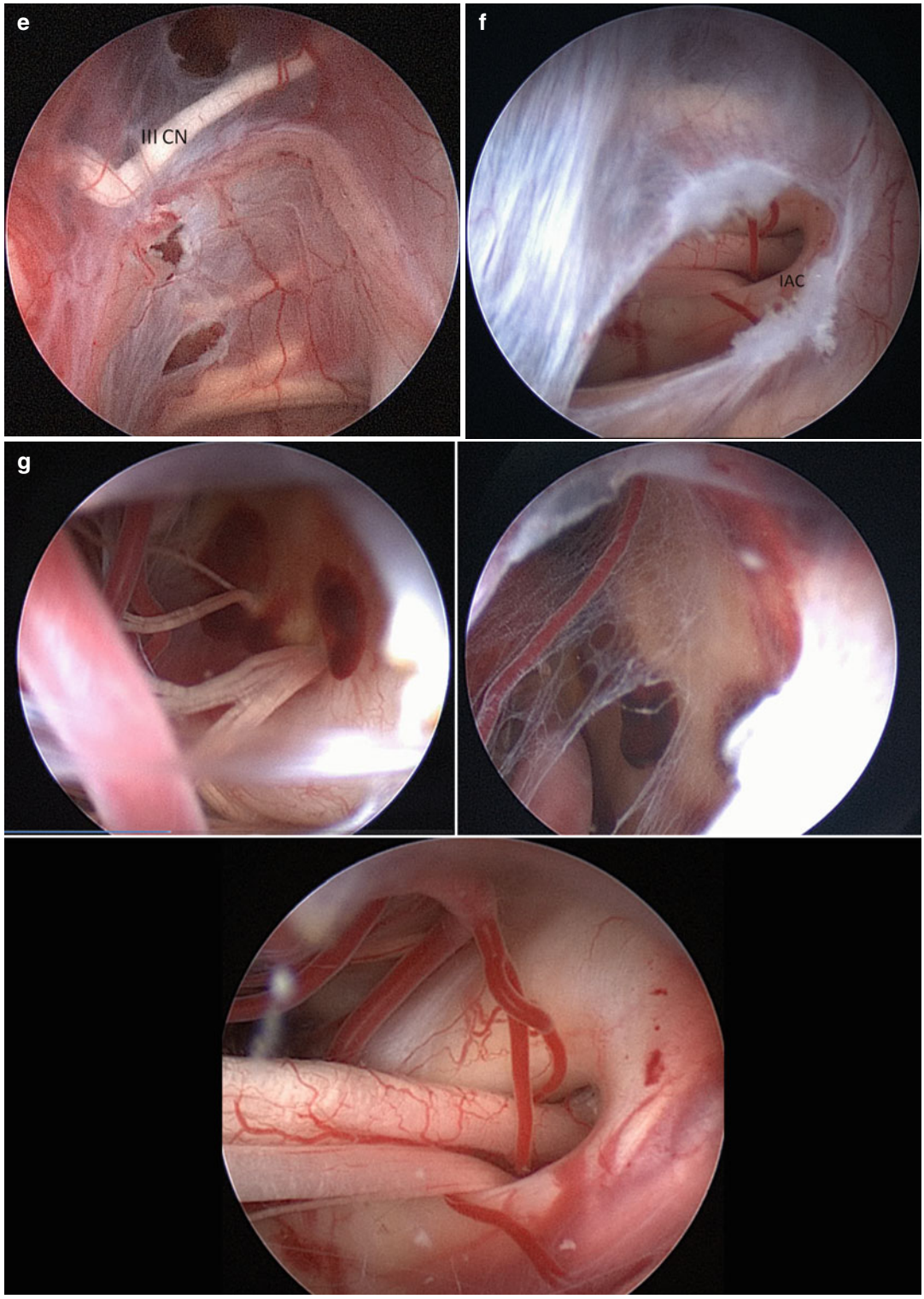


Fig. 16.3 Type 2 suprasellar cyst with lateral extension as seen on T2 sagittal (**a**), T2 coronal (**b**), and T2 axial (**c**) magnetic resonance. Endoscopic view of the skull base area chosen for fenestration (**d**). Following fenestrations, the stretching of the cranial nerves has decreased (**e**). Endoscopic views of the posterior fossa cisterns (**f**, **g**) through the fenestrations. Postoperative MRI showing significant decrease of the cyst size on sagittal (**h**), coronal (*arrow* indicate flow artifact through the stomy) (**i**), and axial (**j**) T2-weighted images



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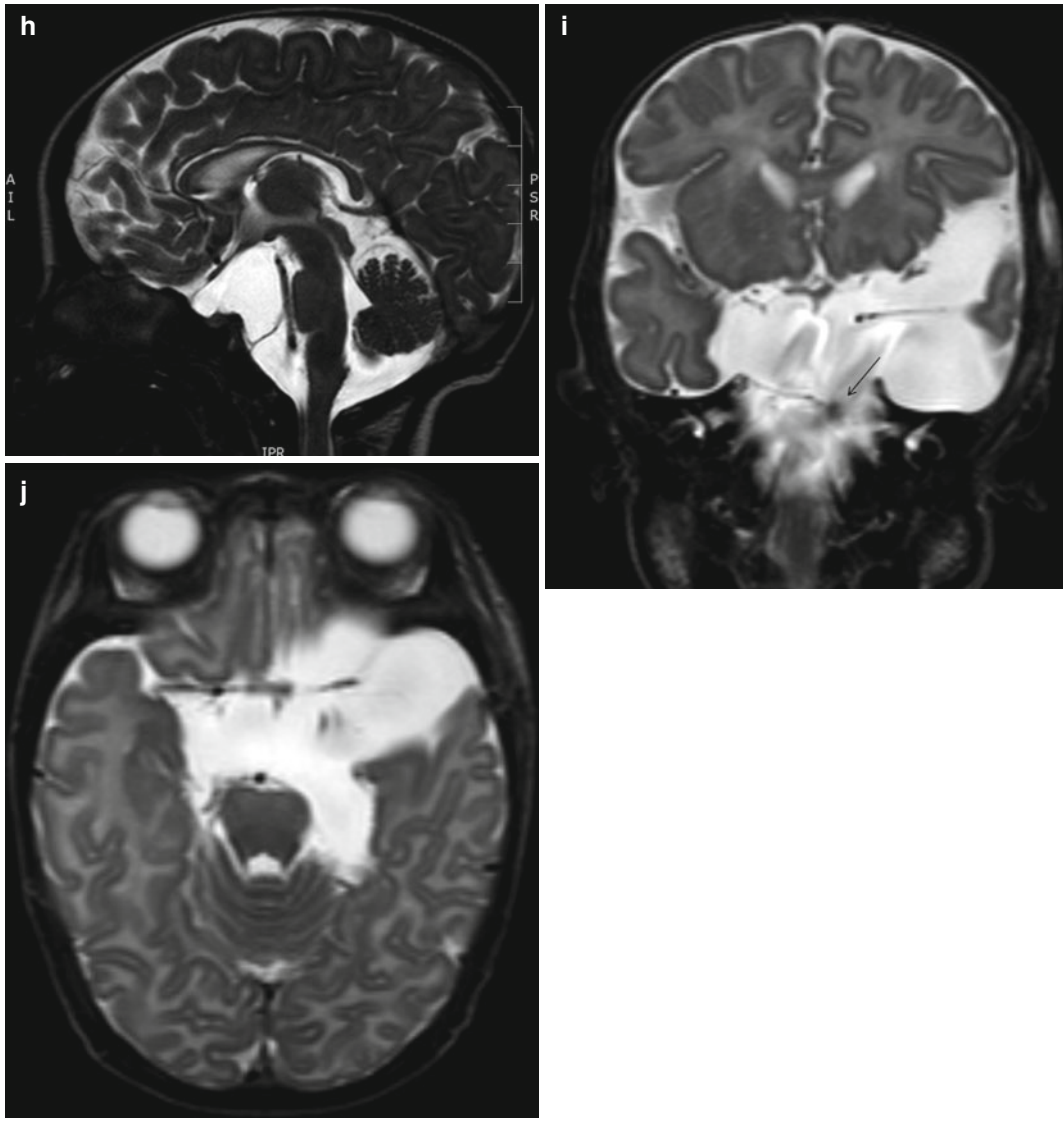


Fig. 16.3 (continued)

a close psychometric and neuroradiological follow-up, especially in the first year of life.

16.3.2 Selection of Candidates to Endoscopic Procedure Versus Alternative Treatments

According with Mori classification [26], Mori distinguished two types of arachnoid cyst in the interhemispheric fissure: unilateral parasagittal cysts and midline cysts. In the first case, the cysts have unilateral extension, are usually discovered in older children, and are not associated with corpus callosum agenesis. Usually, there is no contiguity between this kind of cysts and the third ventricle or other part of the ventricular system. Moreover, these cysts are not associated with hydrocephalus. The best option for these cysts, which usually reach the convexity, is microsurgery with excision of the lining membranes. It is a straightforward procedure, similar to removal

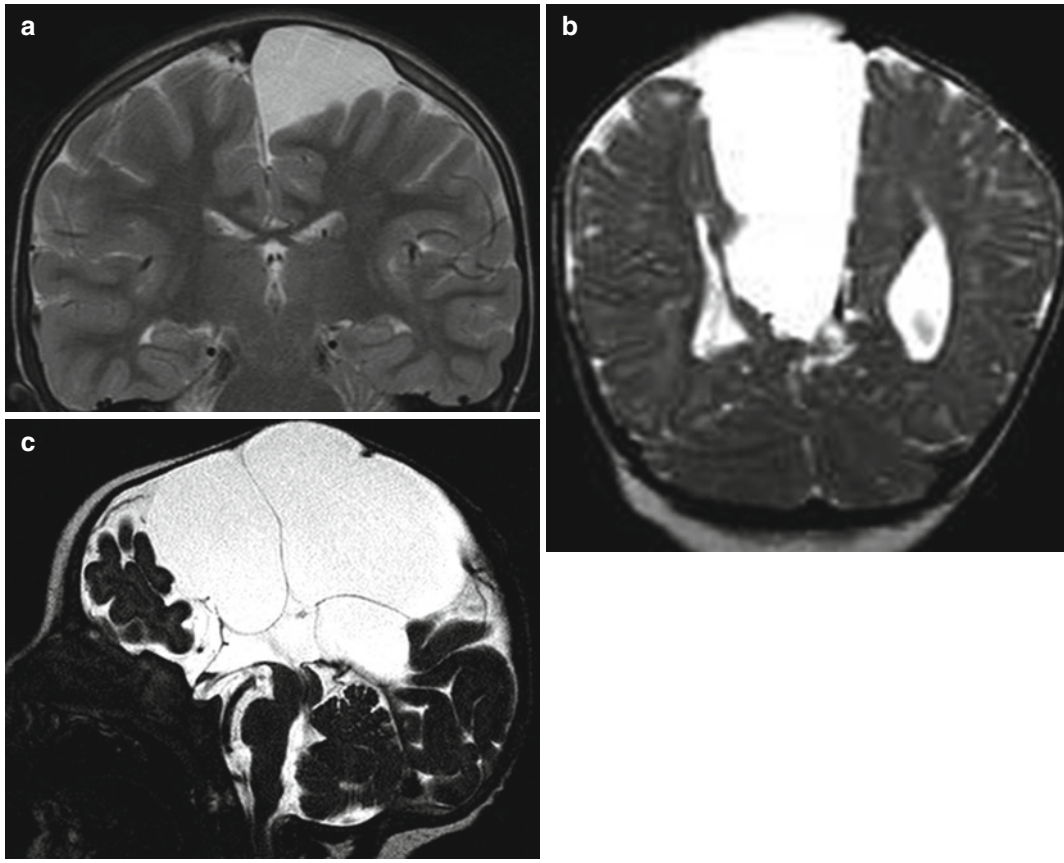


Fig. 16.4 (**a**) Convexity cyst with parasagittal expansion. Interhemisperic cyst associated with corpus callosum agenesis on coronal view (**b**) and sagittal view (**c**)

of arachnoid cysts over the convexity. On the contrary, midline cysts are more complex cyst often multiloculated, discovered in neonates and infants, and associated with agenesis of corpus callosum and ventricular dilatation. They usually expand inside the ventricular system, from which are separated by thin membranes. Due to the absence of the corpus callosum, the most frequent site of contiguity between the cyst and the ventricles is at the level of the roof of the third ventricle. The third ventricle, in fact, is often the final target of fenestration. In midline cyst, endoscopy is usually the best choice, because it allows simplification of multiloculated cysts, fenestration of the cyst with the ventricular system and the subarachnoid space of the interhemispheric/quadrigeminal cistern. In selected cases, third ventriculostomy can also be performed, to treat the associated hydrocephalus and to reduce the chance of stoma reclosure as discussed for suprasellar cysts. Craniotomy with cyst membrane excision, and creation of communication with the subarachnoid cisternal spaces and/or the ventricular system, is a major surgical procedure, with higher rate of complications, especially in neonates. Experience with open surgery is limited [26, 28], with not negligible rate of failure and secondary shunting. Shunting of the cyst was the best option prior to development of endoscopy. It is associated with low operative risk but also by all the well-known complications of the shunts in the long period, such as occlusions, inadequate drainage, and infection [28]. Moreover, in multiloculated cyst and in case of associated hydrocephalus, single cyst catheter may not be sufficient [29].

16.3.3 Authors' Preferred Surgical Technique

The goal of endoscopic surgery is to create communication between the cyst and the ventricular system (cyst-ventriculostomy) and/or the cyst and the cisternal space (cyst-cisternostomy). Surgical planning begins with careful evaluation of preoperative neuroradiological investigations (in particular MRI with DRIVE and CSF flow study) in order to detect the thinner point of the cyst walls where the stoma should be created and the multiple membranes, in case of multiconcamerated cysts. Neuronavigation is mandatory, because of the very distorted anatomy, to place the burr hole and to guide the endoscope through the target. Major vessels of the basal cisterns, the pericallosal arteries, the free edge of the tentorium, and the choroid plexus may be useful orientating landmarks.

Usually the most simple option is to penetrate first in the cyst, through a paramedian burr hole (drilled in the frontal, parietal, or occipital region, according to the location of the cyst), and hence, after recognition of the membranes to be perforated, to perform endoscopic fenestration in standard fashion. We prefer to use Tulium LASER coagulation and removal of large fragments of membranes. Also, the wall of the ventricle is usually a thin membrane. After entering the ventricle, the ependyma is easily recognized. In most cases of frontal and parietal interhemispheric cysts, associated to corpus callosum agenesis, the final target is the third ventricle. If the third ventricle is large enough, it is often possible to add a third ventriculostomy, in standard fashion, so to realize a ventriculocystocisternostomy. In case of posteriorly located interhemispheric cyst, the third ventricle is difficult to reach, and fenestration can be performed toward the interhemispheric fissure or lamina quadrigeminal cistern (cyst-cisternostomy) and toward an enlarged occipital horn of the lateral ventricle. Also in these cases, navigation is mandatory (Fig. 16.5a–e).

Postoperative imaging is essential to assess the patency of the stomas and the patency of the subarachnoid spaces in which the CSF is diverted after the endoscopic procedure.

They are also important to detect possible complications related to the rapid decrease of the cyst volume, such as subdural hygromas.

16.3.4 Complications and Results

The endoscopic experience in the treatment of interhemispheric cysts is limited. Our group reported a small series of seven patients [30] affected by interhemispheric cysts associated with corpus callosum agenesis. Complete success (control of hydrocephalus and of the cyst size with no further surgical procedures) was achieved in five cases (71 %). Partial successes were achieved in the remaining two cases: one patient was endoscopically reoperated with success 1 year later because of closure of the stoma. The remaining patient required implantation of a lumboperitoneal shunt for persistence of subcutaneous CSF collection over the burr hole. The lumboperitoneal shunt was successfully removed without recurrence of the pseudomeningocele 8 months after insertion.

Subdural collection developed in three patients; in only one case, it was managed with insertion of subduro-peritoneal shunt. This was uneventfully removed 4 months later. No patients were shunt dependent at the end of follow-up. Neurodevelopmental evaluation performed in six patients showed normal intelligence (IQT>80) in three patients, mild developmental delay (IQT range 50–80) in two patients, and persistence of severe developmental delay (IQT<50) in one patient, who had undergone surgery when he was 12 years old. Similar satisfactory results were achieved by Giannetti et al. [31], who reported another small series of five patients. They avoided shunt in all very cases and reordered two complications: one subdural hygroma (treated with subduro-peritoneal shunt) and one CSF fistula (treated with lumbar puncture).

16.4 Quadrigeminal Cysts

Arachnoid cysts originated in the quadrigeminal cistern represent only the 10 % of all intracranial cysts. They are not homogeneous but have different extension toward surrounding regions,

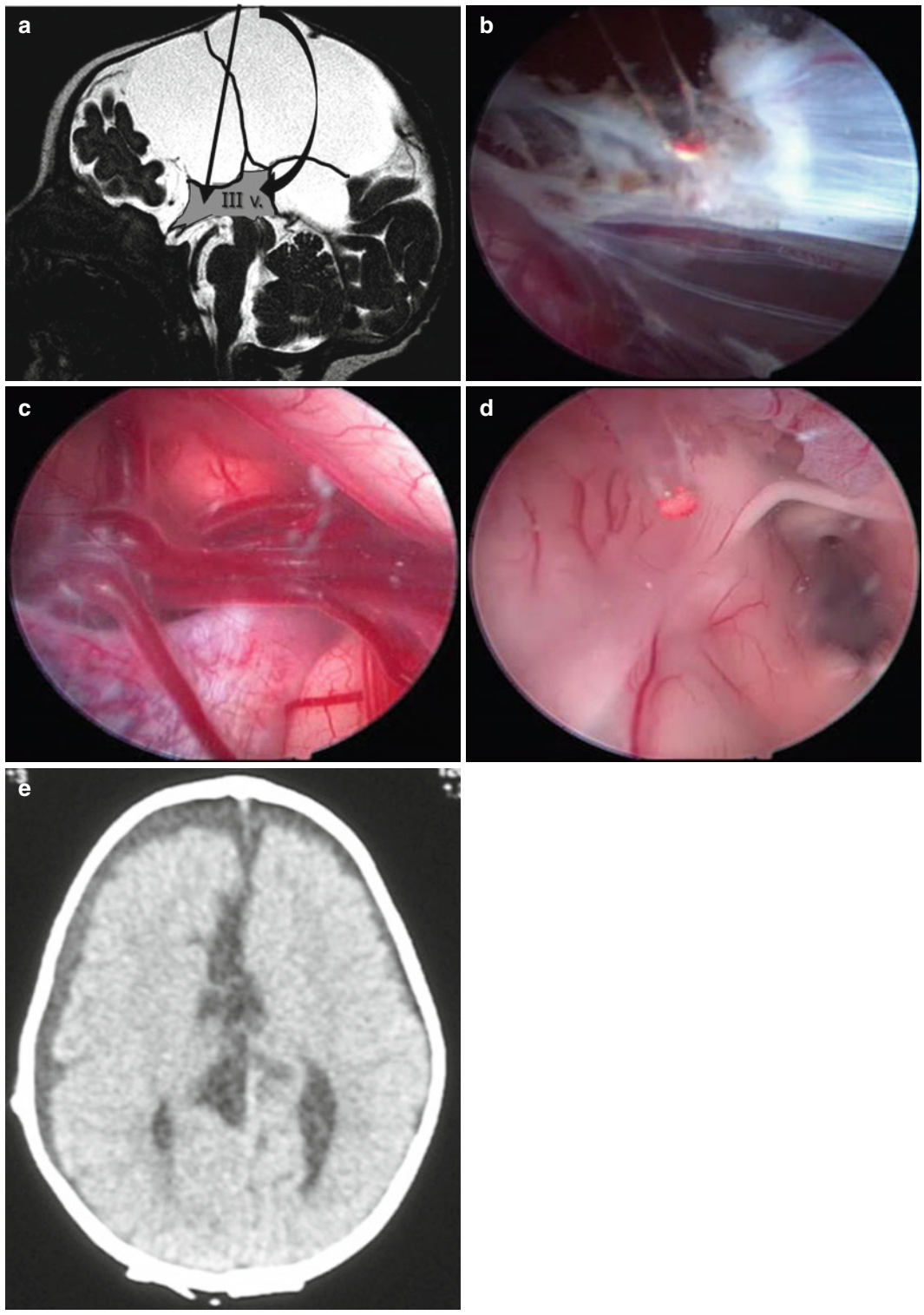
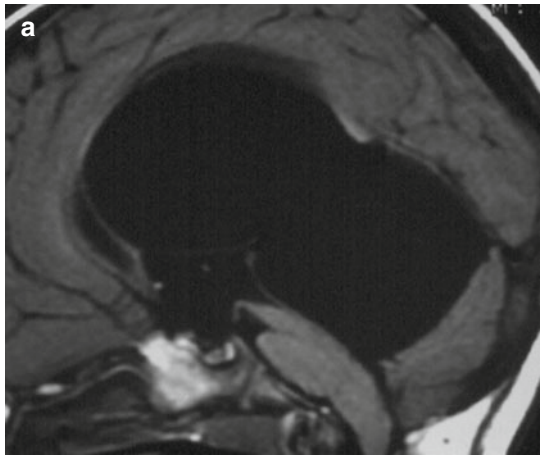
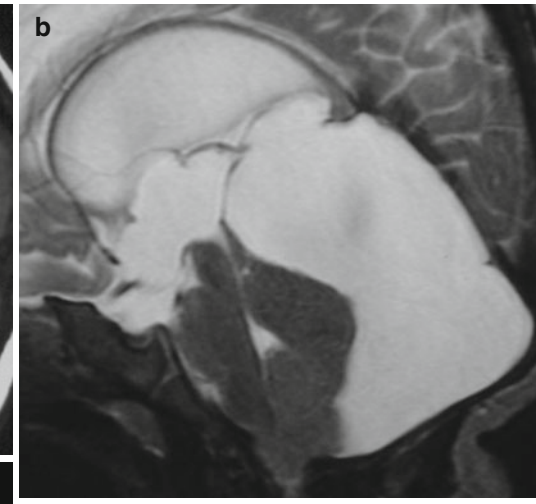


Fig. 16.5 (**a**) Surgical plan for the patient shown on Fig. 16.4. Cyst membranes are open using Thullium laser (**b**). Endoscopic view of the two internal cerebral veins and of Galen complex (**c**). Endoscopic view of the third ventricle (**d**), note the clear ependyma and the two mammillary bodies. Postoperative CT scan (**e**) showing reduction of the cyst volume

according to the presence of loci minoris resistentiae, such as the region of the trigone cranially, the supracerebellar cistern caudally, the third ventricle anteriorly, and the ambient cisterns laterally. Most of them extend both in the supratentorial (at the level of the trigone) and the infratentorial regions (in the supracerebellar cistern) (type I). More rarely, they have only infratentorial extension, in the supracerebellar or supraretrocerebellar regions (type II), or lateral extension in the ambient cisterns toward the temporal lobe (type III) (Fig. 16.6a–c) [16, 27]. Because of their intimate relationship with the dorsal midbrain, quadrigeminal cysts produce distortion or compression of the cerebral aqueduct at an early stage [27]. Symptoms are usually related to the associated hydrocephalus





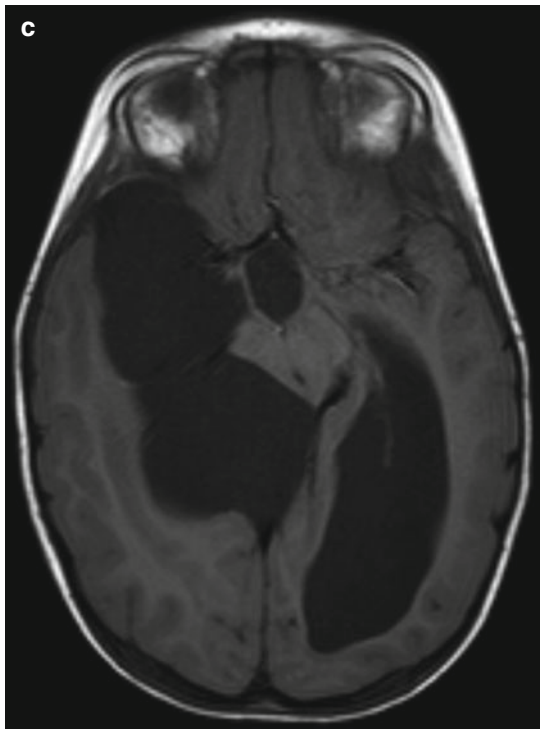


Fig. 16.6 Arachnoid cysts of the quadrigeminal cistern. Type 1 (**a**) has a dumbbell shape and occupies equal volume in the supra and infratentorial space. Type 2 (**b**) is mostly confined to the infratentorial space, in the retrocerebellar or supracerebellar space. Type 3 (**c**) presents a very significant lateral expansion through the choroidal fissure

(macrocrania, headache, vomiting, lethargy, papilledema) and/or compression of the dorsal midbrain (impairment of upward gaze and other ocular disorders).

16.4.1 Indication for Surgery

Large quadrigeminal cyst usually need treatment, because they distort CSF pathways at an early stage and become symptomatic. Untreated cysts may lead to macrocephaly and neurodevelopment delay. Only small cysts not associated with hydrocephalus and with minimal distortion of the aqueduct can be managed conservatively, with close clinical and radiological follow-up, especially in young children.

Careful evaluation of MR preoperative exams should be made in order to distinguish quadrigeminal cysts from pulsion diverticula, which are formed by herniation of the medial wall of the ventricular atrium into the quadrigeminal cistern through the tentorial hiatus in case of severe triventricular hydrocephalus. Anatomically the diverticula are not cysts but communicate with the ventricles; therefore, the treatment is different: endoscopic third ventriculostomy (ETV), curing the hydrocephalus, is usually sufficient to resolve also the associated diverticula, without additional procedures at the level of the quadrigeminal cistern.

16.4.2 Selection of Candidates to Endoscopic Procedure Versus Alternative Treatments

Because quadrigeminal cysts are almost invariably associated with hydrocephalus, endoscopic treatment is usually technically feasible because of the possibility of working in large spaces and the presence of an area of contiguity between the cyst wall and the ventricular ependyma (at the level of the trigone of the lateral ventricle, or at the level of the posterior third ventricle) or subarachnoid spaces. Third ventriculostomy can be also performed (Fig. 16.7a, b) [8, 32–34].

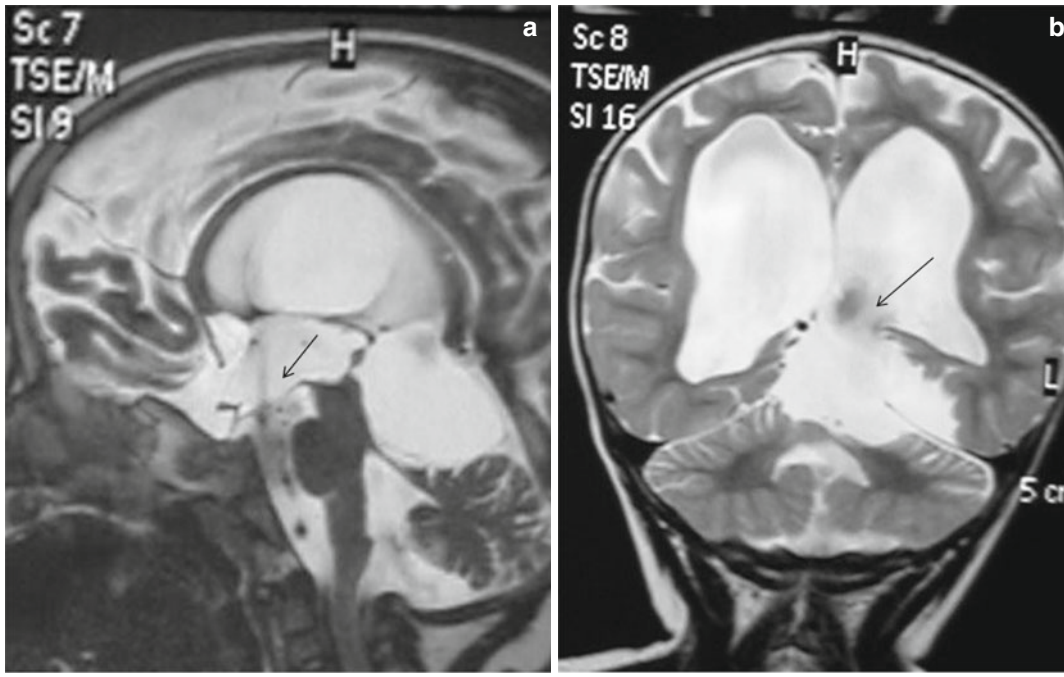


Fig. 16.7 Arachnoid cysts of the quadrigeminal cistern, type 2. Postoperative MRI. Third ventriculostomy is well visible (**a**) as well as the flow artifact through the ventriculocystostomy on coronal cuts (**b**), performed through the lateral ventricle following coronal approach. *Arrows* indicate flow artifact through the stomy

Extrinsic aqueductal stenosis arising from long-standing compression by the cyst may persist despite cyst opening.

Endoscopy can be also considered an alternative to shunt revision in patients already shunted, in order to remove the shunt, or to simplify the shunt system in those patients in whom only one compartment expands (the cyst or the ventricular system), while the other is adequately drained [16, 27].

16.4.3 Authors' Preferred Surgical Technique

In the most frequent presentation (type I), the cyst extends upward in the lateral ventricle, thinning the floor of the ventricular trigone. In these cases, a standard precoronal burr hole is indicated and the upper pole of the cyst, which appears below the ependyma medial to the choroid plexus, can be fenestrated (lateral ventricle –cystostomy). Neuronavigation is useful but not mandatory. The side of incision should be chosen according to the position of the internal cerebral veins: the cyst, in fact, has usually an asymmetric expansion, and the cerebral veins are displaced contralaterally. The ependyma and the cyst wall can be opened by monopolar or bipolar laser coagulation; the fenestration can be enlarged by grasping forceps and 3-F Fogarty balloon or double-balloon catheter (Lighttouch balloon; Integra Neuroscience, Biot, France). To avoid reclosure of the stoma, large openings, at least 10–15 mm in diameter, should be performed. After decompression of the arachnoid cyst, the endoscope can be advanced through the foramen of Monro to perform a standard endoscopic third ventriculostomy. Fenestration of the deep wall of the cyst, if ETV is successfully performed, seems to not add significant advantages.

In case of cysts bulging in the posterior aspect of the third ventricle (type II), a precoronal trans-Monro approach to the third ventricle allows fenestration of both the anterior wall of the cyst (third ventricle – cystostomy) and the floor of third ventricle. In this case, a steerable endoscope should be preferred, allowing both fenestrations through the same burr hole. With a rigid endoscope, a more anterior burr hole should be drilled, to reach the posterior part of the third ventricle: cyst fenestration and ETV can be both performed only when the ventricular system and the foramen of Monro are markedly dilated.

In the case of laterally extending cysts in the ambient cistern (type III), (Fig. 16.8a–h) a parietal burr hole with the patient head rotated 90° on the contralateral side is usually indicated to approach the lateral ventricle at the level of the trigone. The upper pole of the cyst will appear just in front of the endoscope. In these cases, third ventriculostomy cannot be performed; therefore, further fenestration of the deep wall in the basal cistern (usually in the cisterna ambiens) should be attempted. In type III cyst, neuronavigation is very useful, to plan the correct entry point and to guide the endoscope toward the target in the basal cistern.

16.4.4 Results

In the last decades, endoscopic treatment has become the first-line option in the treatment of quadrigeminal cysts in several centers all around the world [11, 32–39]. The results are very encouraging results, especially when double fenestration was performed (ventriculocystostomy + third ventriculostomy, ventriculocystocisternostomy, double ventriculocystostomy).

In our series of 14 cases [30], double fenestration was realized in 12 cases, with only one failure (success rate 91 %). All seven cases in which only ventriculocystostomy was performed failed and required a second procedure (repeat endoscopy or shunting).

In our experience, the patients, already shunted, who present with cysts enlarging, despite functioning VP shunts, were the most difficult to treat. The shunts were externalized, and the drainage bag progressively elevated to obtain enough space to allow safe endoscopic navigation. However, their ventricular system was not so compliant and there were symptoms of increased ICP before dilation occurred. The endoscopic procedure was uneventful in only one out of three cases. In the remaining two cases

(one of these was complicated by venous bleeding from the cyst walls and also from the third ventricular floor) the cysts recurred and the shunts were reimplanted.

In conclusion, neuroendoscopic approaches can be considered an effective alternative to traditional methods in the management of quadrigeminal cistern cysts either in primary presentation or in patients presenting with shunt malfunction. In case of failure, because of the high success rate also in secondary cases, endoscopic reoperation can be considered before considering alternative treatments. The indications for further intervention depend on the persistence of the patient's symptoms and progressive enlargement of the cyst on postoperative imaging at follow-up.

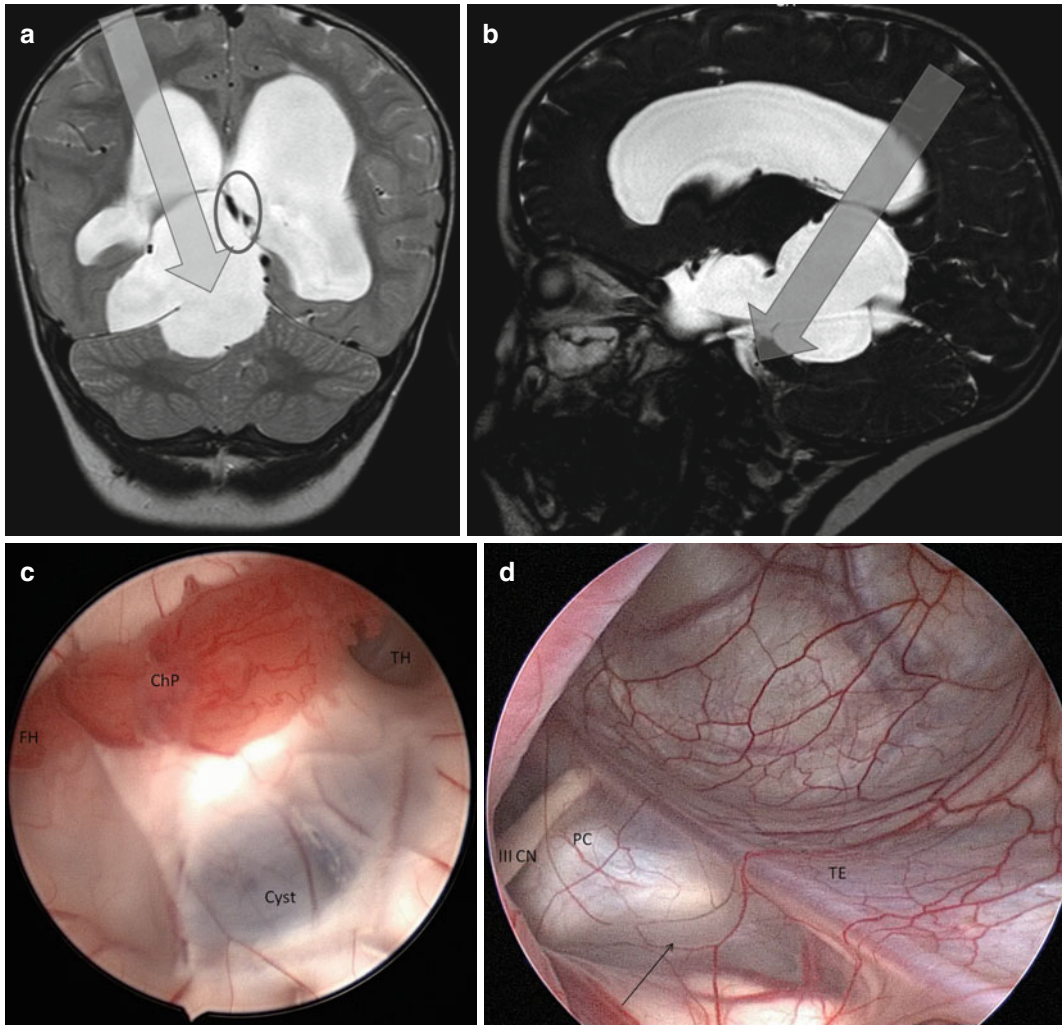


Fig. 16.8 Arachnoid cysts of the quadrigeminal cistern, type 3. Preoperative MRI showing surgical trajectory through a parietal burr hole on coronal (**a**) and sagittal (**b**) views to create a ventriculocystocisternostomy. Endoscopic vision of the cyst dome as seen through a right parietal approach (**c**), the head of the patient is in left lateral decubitus. The cyst wall is easily recognized as a dark layer visible through the attenuated ependyma medial to the choroid plexus. Endoscopic vision of the anterior pole of the cyst as seen after entrance into the cyst lumen (**d**). An area of perforation is identified behind the posterior clinoid (*arrow*). Basal cistern as seen endoscopically through the cyst-cisternostomy (**e**). Post operative MRI showing the flow artifact through the ventriculocystostomy (**f**) and through the cyst-cisternostomy (**g**). Both flow artifacts are visible on this sagittal T2 MRI (**h**). *Arrows* indicate the surgical trajectory, *Circle* indicates the internal cerebral veins

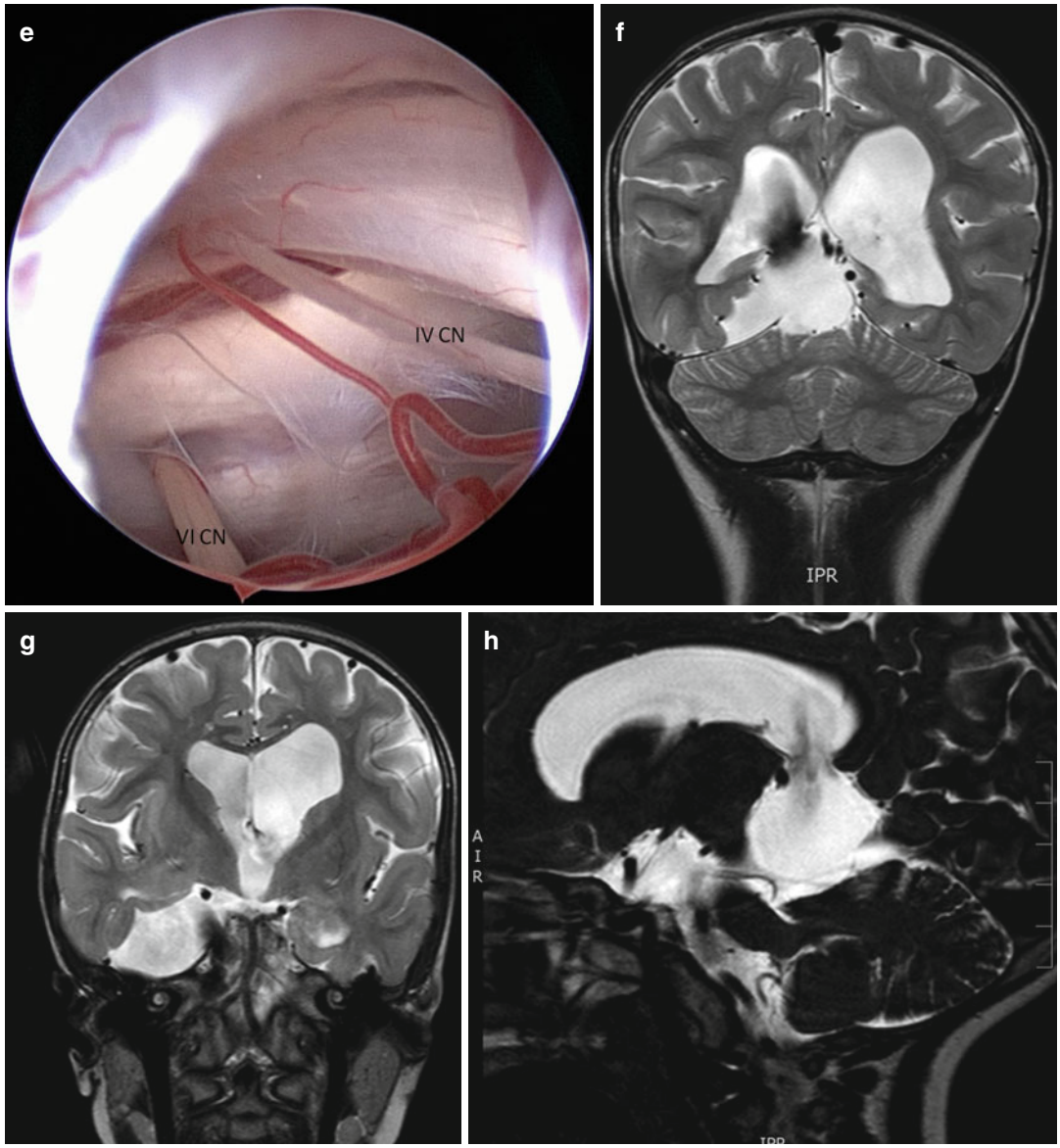


Fig. 16.8 (continued)

References

1. Abbott R (2004) The endoscopic management of arachnoidal cysts. Neurosurg Clin N Am 15:9–17
2. Artico M, Cervoni L, Salvati M, Fiorenza F, Caruso R (1995) Supratentorial arachnoid cysts: clinical and therapeutic remarks on 46 cases. Acta Neurochir 132:75–78
3. Galassi E, Gaist G, Giuliani G, Pozzati E (1988) Arachnoid cysts of the middle cranial fossa: experience with 77 cases treated surgically. Acta Neurochir Suppl 42:201–204
4. Cinalli G (1999) Alternatives to shunt. Childs Nerv Syst 15:718–731
5. Tirakotai W, Schulte DM, Bauer BL, Bertalanffy H, Hellwig D (2004) Neuroendoscopic surgery of intracranial cysts in adults. Childs Nerv Syst 20:842–851
6. Gaab MR, Schroeder HWS (1998) Neuroendoscopic approach to intraventricular lesions. J Neurosurg 88:496–505
7. Hopf NJ, Perneczky A (1998) Endoscopic neurosurgery and endoscope-assisted microneurosurgery for the treatment of intracranial cysts. Neurosurgery 43:1330–1337

8. Decq P, Brugieres P, Le Guerinel C, Djindjian M, Keravel Y, Nguyen JP (1996) Percutaneous endoscopic treatment of suprasellar arachnoid cysts: ventriculocystostomy or ventriculocystocisternostomy? Technical note. J Neurosurg 84:696–701
9. Caemaert J, Abdullah J, Calliauw L, Carton D, Dhooge C, van Foster R (1992) Endoscopic treatment of suprasellar arachnoid cyst. Acta Neurochir 119:68–73
10. Choudhri O, Feroze AH, Nathan J, Cheshier S, Guzman R (2014) Ventricular endoscopy in the pediatric population: review of indications. Childs Nerv Syst 30:1625–1643
11. Kirollos RW, Javadpour M, May P, Mallucci C (2001) Endoscopic treatment of suprasellar and third ventricle-related arachnoid cysts. Childs Nerv Syst 17:713–718
12. Spacca B, Kandasamy J, Mallucci CL, Genitori L (2010) Endoscopic treatment of middle fossa arachnoid cysts: a series of 40 patients treated endoscopically in two centres. Childs Nerv Syst 26:163–172
13. Di Rocco F, R James S, Roujeau T, Puget S, Sainte-Rose C, Zerah M (2010) Limits of endoscopic treatment of sylvian arachnoid cysts in children. Childs Nerv Syst 26:155–162
14. Galassi E, Tognetti F, Gaist G, Fagioli L, Frank F, Frank G (1982) CT scan and metrizamide CT cisternography in arachnoid cysts of the middle cranial fossa: classification and pathophysiological aspects. Surg Neurol 17(5):363–369
15. Choi JW, Lee JY, Phi JH, Kim SK, Wang KC (2015) Stricter indications are recommended for fenestration surgery in intracranial arachnoid cysts of children. Childs Nerv Syst 31:77–86
16. Cinalli G, Spennato P, Ruggiero C, Aliberti F, Trischitta V, Buonocore MC, Cianciulli E, Maggi G (2007) Complications following endoscopic intracranial procedures in children. Childs Nerv Syst 23(6):633–644
17. Tamburrini G, Caldarelli M, Massimi L, Santini P, Di Rocco C (2003) Subdural hygroma: an unwanted result of Sylvian arachnoid cyst marsupialization. Childs Nerv Syst 19(3):159–165
18. Levy ML, Wang M, Aryan HE, Yoo K, Meltzer H (2003) Microsurgical keyhole approach for middle fossa arachnoid cyst fenestration. Neurosurgery 53(5):1138–1144
19. Karabagli H, Etus V (2012) Success of pure neuroendoscopic technique in the treatment of Sylvian arachnoid cysts in children. Childs Nerv Syst 28(3):445–452
20. Elhammady MS, Bhatia S, Ragheb J (2007) Endoscopic fenestration of middle fossa arachnoid cysts: a technical description and case series. Pediatr Neurosurg 43(3):209–215
21. Karabatsu K, Hayhurst H, Buxton N, O'Brien DF, Mallucci CL (2007) Endoscopic management of arachnoid cysts: an advancing technique. J Neurosurg 106:455–462
22. El-Ghandour NM (2012) Endoscopic treatment of middle cranial fossa arachnoid cysts in children. J Neurosurg Pediatr 9:231–238
23. Miyajima M, Arai H, Okuda O, Hishii M, Nakanishi H, Sato K (2000) Possible origin of suprasellar arachnoid cysts: neuroimaging and neurosurgical observations in nine cases. J Neurosurg 93:62–67
24. Maher CO, Goumnerova L (2011) The effectiveness of ventriculocystocisternostomy for suprasellar arachnoid cysts. J Neurosurg Pediatr 7:64–72
25. Rizk E, Chern JJ, Tagayun C, Tubbs RS, Hankinson T, Rozzelle C, Oakes WJ, Blount JP, Wellons JC (2013) Institutional experience of endoscopic suprasellar arachnoid cyst fenestration. Childs Nerv Syst 29:1345–1347
26. Mori K (1992) Giant interhemispheric cysts associated with agenesis of the corpus callosum. J Neurosurg 76:224–230
27. Spennato P, Ruggiero C, Aliberti F, Buonocore MC, Trischitta V, Cinalli G (2013) Interhemispheric and quadrigeminal cysts. World Neurosurg 79(2 Suppl):S20.e1–S20.e7
28. Caldarelli M, Di Rocco C (1996) Surgical options in the treatment of interhemispheric arachnoid cysts. Surg Neurol 46:212–221
29. Ulu MO, Kafadar AM, Dashti R, Isler C, Uludağ S, Erdinçler P (2010) Treatment of symptomatic interhemispheric arachnoid cysts by cystoperitoneal shunting. J Clin Neurosci 17:700–705
30. Cinalli G, Peretta P, Spennato P, Savarese L, Varone A, Vedova P, Grimaldi G, Ragazzi P, Ruggiero C, Cianciulli E, Maggi G (2006) Neuroendoscopic management of interhemispheric cysts in children. J Neurosurg 105(Suppl):194–202
31. Giannetti AV, Fraga SM, Silva MC, Gurgel-Giannetti J (2012) Endoscopic treatment of interhemispheric arachnoid cysts. Pediatr Neurosurg 48:157–162
32. Cinalli G, Spennato P, Columbano L, Ruggiero C, Aliberti F, Trischitta V, Buonocore MC, Cianciulli E (2010) Neuroendoscopic treatment of arachnoid cysts of the quadrigeminal cistern: a series of 14 cases. J Neurosurg Pediatr 6:489–497
33. Ersahin Y, Kesikçi H (2009) Endoscopic management of quadrigeminal arachnoid cysts. Childs Nerv Syst 25:569–576
34. Tamburrini G, Di Rocco C, Caldarelli M, Massimi L (2005) Endoscopic management of supratentorial intra and paraventricular cysts. Childs Nerv Syst 21:500
35. Choi JU, Kim DS, Huh R (1999) Endoscopic approach to arachnoid cyst. Childs Nerv Syst 15:285–291
36. Gangemi M, Maiuri F, Colella G, Magro F (2005) Endoscopic treatment of quadrigeminal cistern arachnoid cysts. Minim Invasive Neurosurg 48:289–292
37. Schroeder HWS, Gaab MR, Niendore WR (1996) Neuroendoscopic approach to arachnoid cysts. J Neurosurg 85:293–298
38. Garg K, Tandon V, Sharma S, Suri A, Chandra PS, Kumar R, Mahapatra AK, Sharma BS (2015) Quadrigeminal cistern arachnoid cyst: a series of 18 patients and a review of literature. Br J Neurosurg 29(1):1–7
39. El-Ghandour NM (2013) Endoscopic treatment of quadrigeminal arachnoid cysts in children. J Neurosurg Pediatr 12:521–528