Hydrocephalus and Arachnoid Cysts

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Abstract

The prevalence of intracranial arachnoid cysts (ACs) in children is comprised between 1% and 3%. Most of them are asymptomatic and no treatment is needed.

ACs can be divided into seven types: suprasellar, temporal, intraventricular, quadrigeminal, posterior fossa, convexity, and interhemispheric.

History, pathophysiology, epidemiology, clinical presentation, radiological diagnosis, classifications, association with hydrocephalus, surgical management and techniques, their outcome, and complications are discussed.

Three main techniques are available: open microsurgery, cyst and/or ventriculoperitoneal shunting, and endoscopic fenestration or a combination of these techniques.

Suprasellar, intraventricular, and quadrigeminal ACs appear to be the best indications for endoscopic fenestration with better outcome because of their position in the midline, in intimate relationships with the basal cisterns and ventricles. For a durable endoscopic treatment, the key point is to perform the largest and more fenestrations as possible, allowing a better restoration of cerebrospinal fluid (CSF) pathways. Cortical cysts seem to be best treated by open microsurgery or shunting. Shunt implantation showed a high rate of complications such as malfunctions, shunt dependency, slit cyst syndrome, or chronic tonsillar herniation that can be severe but appears to be the best treatment when indications are well chosen.

When ACs are symptomatic or if hydrocephalus is present, the best treatment remains to create communications between the cyst and normal CSF cisterns or ventricles, each time it

is possible with the best mini-invasive single procedure. The amendment of neuroendoscopy tends to treat most of ACs, but microsurgical and shunting procedures still keep a place in selected cases.

Keywords
Arachnoid cyst · Hydrocephalus · Endoscopy · Shunt · Suprasellar · Quadrigeminal · Interhemispheric · Temporal · Slit cyst syndrome · Chronic tonsillar herniation

Introduction

Arachnoid cysts are fluid-filled sacs that are located between the brain or spinal cord and the arachnoid membrane. Their prevalence in children is comprised between 1% and 3%. With the advent and increasing use of magnetic resonance imaging, arachnoid cysts have been diagnosed with increasing frequency. Most of them are asymptomatic. The symptoms depend on the size, the location, the growth rate, and the obstruction to CSF circulation. Hydrocephalus can be associated with different frequency according to the location.

It is generally accepted that surgical treatment is not indicated in the case of arachnoid cysts that are asymptomatic or incidentally discovered. Hydrocephalus, raised intracranial pressure, subdural hematoma, and focal neurological deficit are usually considered straightforward surgical indications. Open debate remains about other symptoms like headache, macrocrania, associated pericerebral collections, developmental delay, and seizures. The ideal option to manage them surgically would consist of finding the technical method that offers the highest success rate with a single procedure for treating hydrocephalus and the arachnoid cyst simultaneously diminishing the complication rate. Different options in the surgical management exist: open surgery, cyst and/or ventriculoperitoneal shunting, endoscopic fenestration, or a combination of these techniques, and the choice among them can depend upon patient-related factors like age, size, and location and surgeon-related factors like surgical experience in different techniques (i.e., endoscopy) or personal preferences.

Classification of Intracranial Cysts

Pathology

A cyst is a cavity, closed sac-like structure having a distinct membrane, defined by absence of solid component, containing fluid or semifluid matter.

Two classifications of intracranial cysts can be proposed. The first one, radiologic, emanates from the fetal diagnosis by ultrasound scan. The second one comes from postnatal histological diagnosis.

Prenatal Classification (Intracranial Cysts Diagnosed in Fetus)

Intracranial cystic lesions are frequently diagnosed by fetal ultrasound scan (Malinger et al. 2008), and a useful classification was proposed according to these findings (Table 1).

Extra-Axial Cysts

Arachnoid cysts are the most common extra-axial cysts found in prenatal ultrasound (Malinger et al. 2008). Their prenatal diagnosis has been

Table 1 Differential diagnosis of fetal intracranial cystic lesions (Malinger et al. 2008)

Extra-axial cysts	Intraparenchymal cysts	Intraventricular cysts
Arachnoid cyst	Periventricular pseudocyst	Choroid plexus cysts
Dural separation	Cystic periventricular leukomalacia	Choroid plexus hemorrhage
Glioependymal cyst	Porencephalic cyst	
Endodermal cyst	Brain cystic tumor	
Cystic teratoma		

reported in literature. Pilu et al. report 21 cases (Pilu et al. 1997) and Pierre-Khan and Sonigo 54 cases (Pierre-Kahn and Sonigo 2003). In their study of 54 cases, 63% of the patients, the arachnoid cysts were supratentorial, mostly in the interhemispheric fissure (25%) and then in the infratentorial region (22,2%). All the cysts were diagnosed after the 20th week of gestation. All cases were confirmed by a magnetic resonance imaging (MRI), allowing better localization and relationship with adjacent structures. Hogge et al. published a case of infratentorial arachnoid cyst earlier, in the 18th week associated with an unbalanced X;9 translocation (Hogge et al. 1995). Bretelle et al. reported an infratentorial arachnoid cyst at the 13th week, confirmed histologically after termination of pregnancy at 15 weeks (Bretelle et al. 2002). Sometimes those cysts are associated with cortical malformation (Malinger et al. 2007), metabolic disorders (glutaric aciduria type 1), congenital hypothyroidism (Malinger et al. 2008), neurofibromatosis, tuberous sclerosis, autism, dystonia, leukodystrophy, and microcephaly. Huang Huang et al. found in 25% of their 488 patients with arachnoid cysts an association with other congenital abnormalities (Al-Holou et al. 2010). Malinger et al. recommend to follow up longitudinally throughout the pregnancy searching for other brain anomalies, performing MRI and glutaric aciduria test (arachnoid cyst in the opercular area particularly).

Differential and rare diagnosis of extra-axial fetal cysts are glioependymal cysts (neuroectodermal cysts), endodermal cysts, and cystic teratoma (Hirano and Hirano 2004; Cassart et al. 2008).

Intraparenchymal Cysts

Periventricular pseudocysts hemorrhage, infection, ischemia, or tumoral process can generate cystic lesions in the brain. Often found close to the central gray nuclei, they can be unique or multiloculated and unilateral or bilateral. They are found in 1% of newborns and can develop after a small hemorrhage in the germinal matrix that secondary liquefies (Malinger et al. 2008). They are more rarely a consequence of Cytomegalo virus (CMV) infection, metabolic or chromosomal disorders, and cardiac malformation. Half of them are asymptomatic after birth.

Cystic periventricular leukomalacia touches preferably premature newborns or later after hypoxic or ischemic events (Volpe 2008). They are basically found on the top of the lateral ventricles. They are formed by a focal necrosis of the periventricular white matter. Prenatal maternal infection has been recently studied as a possible cause.

Porencephalic cysts occur after focal necrosis due to an ischemia in the territory of a major brain artery (Volpe 2008). Pilu et al. studied ten fetuses suffering of severe porencephaly. In 90% of them, the cyst was connected to the lateral ventricles. 60% of the patients were dead due to prenatal problems or as a result of medical termination. The remaining 40% of the patients suffered of severe neurodevelopment delay (Pilu et al. 1997). One case of porencephalic cyst caused by a car accident during pregnancy has been reported (Malinger et al. 2008).

Brain cystic tumors defined by the absence of solid components are very rare. Malinger et al. found as a possible cause the intraparenchymal choroid plexus papilloma (Malinger et al. 2008).

Intraventricular Cysts

Choroid plexus cysts are the most common intraventricular cysts with a prevalence estimate between 1% and 3.6% of pregnancies (Chinn et al. 1991). They can be unilateral or bilateral and found in all places where the choroid plexus is present but are more prominent in lateral ventricles. This hyperechogenic structure of choroid plexus is easily recognizable from the 8th week of pregnancy (Malinger et al. 2008), but cysts are not observed before 17 weeks and most of the time disappear before the 26th week. They are considered as benign lesions, but when they are multiple, one should search for other lesion because of the possible association with trisomy 18 and 21 (Malinger et al. 2008).

Intraventricular hemorrhage inside the choroid plexus, colloid, or ependymal cysts is rarely diagnosed by fetal ultrasound.

Postnatal Classification (Intracranial Cysts Diagnosed After Birth)

Nowadays intracranial cysts can be diagnosed thanks to computerized tomography (CT) and MRI. But many different types of them have been identified based on clinical and pathological studies. Hirano and Hirano propose a simple classification of intracranial cysts based on the histological etiologic point of view (Hirano and Hirano 2004). They divide the cysts in two groups: one is derived from central nervous system tissue and the other group derived from extracranial tissue.

Cysts Derived from Central Nervous System (CNS) Tissue

Infarcts, **traumatic injuries**, and **infection** can result into loss of brain tissue. The destroyed parenchyma is replaced by a fluid cavity, which often communicates with ventricular or subarachnoid spaces, and is surrounded by arachnoid membranes (Hirano and Hirano 2004). When the parenchymal cavity reaches the brain surface, they are called porencephalic cysts.

Arachnoid cysts, unlike cysts resulting from damage to the CNS tissue, are entirely surrounded by a transparent arachnoid membrane and a more or less severely flattened cerebral cortex.

Ependymal cysts are deep within the brain parenchyma and have a single layer of ependymal cells and an extracellular space containing collagen fibers. The underlying white matter made by astrocytes may have gliotic reaction, and the cells show spongy changes (Hirano and Hirano 2004).

Neuroectodermal cysts reported on case reports are localized in subarachnoid space and look like arachnoid cysts, but histologically they distinguish from them by a layer of cuboidal epithelial cells more or less part of choroid plexus on the walls.

Tumoral cyst of the cerebellar hemispheres has three known causes: cystic astrocytoma, hemangioblastoma, or focal angiomas. The wall of these cysts contains nodules with blood vessels and tumor cells. The rest of the wall is made by gliotic astrocytes containing Rosenthal fibers and overlying white matter (Hirano and Hirano 2004).

Cysts caused by infectious agents have two main origins: cysticercosis and toxoplasmosis. The walls of cysticercosis cyst are very similar to the bacterial abscesses with a layer of multinucleated giant cells and inflammatory cells surrounded by a layer of connective tissue and reactive gliosis (see part "Main Etiologies", chapter ► "Neurocysticercosis: Pharmacological and Surgical Handling in Open and Endoscopic Surgery"). The cyst wall of toxoplasmosis is surrounded by cytoplasmic processes of astrocytes containing abundant glial fibrils. Those cysts are intracytoplasmic inclusion bodies (bradyzoites) distinguished from cysticercosal cysts. One can observe tachyzoites in macrophages, reactive astrocytes, and in extracellular space (Hirano and Hirano 2004).

Cysts Derived from Extracranial Tissue

These cysts derive from the intrusion of non-nervous tissue into the neuroaxis and often originate in one of these subarachnoid regions: the pineal gland region, the suprasellar cistern, and the cerebellopontine angle. All of these lesions may grow to considerable size before clinical symptoms appear (Hirano and Hirano 2004).

Teratomas are rarely purely cystic and often associated with keratin-forming epidermis, hair follicles, and sweat glands.

Dermoid and epidermoid cysts result from aberrant inclusion of ectodermal elements during neural tube closure, between the 3rd and 5th week of pregnancy. Epidermoid cyst with a hilly appearance and a white pearly coloration is made by a squamous stratified and keratinized epithelium delimiting a cavity in which desquamated cell degradation products accumulate as an amorphous material, rich in keratin and cholesterol. Dermoid cysts do not have the same milky white appearance characteristic. The epithelium is similar, but its wall contains dermal elements as hair follicles, sebaceous and sweat glands, and sometimes fat lobules. The cyst contains heterogeneous mixture of keratin, cholesterol, sebaceous and sweat secretions, and very often hair. Both of them wall are avascular and may contain islands of calcification.

Craniopharyngiomas (8–13% of primary brain tumors in children), typically from the suprasellar region, are classically considered as developing from epithelial remnants of the Rathke's cleft. This tumor contains in varying proportions: a thick part, cysts, and calcifications. Sometimes the tumor is essentially made of macrocysts that contain a yellow, brownish, or green liquid and often "glitter" of cholesterol. Their wall is smooth and translucent or dotted of fine calcifications.

Endodermal cysts result from invagination or displacement of endodermal tissue into the neuroaxis. They are more frequent in the spine in intradural extramedullary space. The cyst wall is made by a single layer of cuboidal epithelium containing cilia and microvilli in direct contact with connective tissue. Many similarities can be found with the upper respiratory tract (Hirano and Hirano 2004).

Colloid cysts of the third ventricle (0.5–2% of primary brain tumors) are sharply bordered oval masses below the fornix of the septum pellucidum, made of a thin wall, clear, or even translucid containing a gelatinous elastic substance of variable density. They occupy the extension of the subarachnoid space called velum interpositum as the neuroectodermal tumors in the pineal region (Hirano and Hirano 2004). The size of the lesion is variable, ranging from few millimeters to several centimeters. The possible origin of these dysembryoplastic cysts may be neuroectodermal or endodermal. Microscopically the wall consists of a conjunctive capsule covered with a unilayered or multilayered epithelium made of cubic or cylindrical cells that may contain cilia. The mucus content is rich in carbohydrates and acellular and can contain microcalcifications.

Rathke's cleft cysts, literally placed in the pituitary region, may have endodermal and epidermal features. Desmosomal connections between the cells of the superficial layer of respiratory-type epithelium and the underlying cells of squamous epithelium have been observed (Hirano and Hirano 1988). They have also showed junctions between epithelial cells lining a cyst in the pars intermedia of the pituitary gland with anterior pituitary cells containing secretory granules.

Table 2 Neuropathological classification of intracranial cystic lesion (Hirano and Hirano 2004)

Derived from CNS tissue	Derived from extracranial tissue
Cysts resulting from damaged CNS tissue: infarcts, traumatic injuries, infection, porencephalic cysts	Teratomas, dermoid and epidermoid cysts
Arachnoid cysts	Craniopharyngioma
Ependymal cysts	Endodermal cysts
Neuroectodermal cysts (glioependymal)	Colloid cysts
Obstructive hydrocephalus	Rathke's cleft cysts
Cysts of the cerebellar hemispheres	Enterogenous cysts
Cysts caused by infectious agents: cysticercosis, toxoplasmosis	

CNS central nervous system

Enterogenous cysts can be found within the neuroaxis (Hirano and Hirano 1988). Similar to the gastrointestinal tract, the epithelial cyst walls do not contain cilia but goblet cells with secretory granules and occasional small dense Kultschitzky's cells containing neurosecretory endocrine vesicles (Table 2).

This chapter will be focused mainly on intracranial arachnoid cysts and their relation with hydrocephalus.

Arachnoid Cysts

Epidemiology

For this epidemiological analysis, we will particularly insist on the two largest series found in the literature: the one of Al-Holou et al. with 309 arachnoid cysts found on 11,738 MRI (Al-Holou et al. 2010) and the one of Huang et al. with 488 cases of arachnoid cysts (Huang et al. 2015).

With the improvement in quality and quantity of diagnostic imaging realized, the number of

incidentally founded arachnoid cysts has increased. Older studies focusing on adults estimate arachnoid cysts prevalence between 0.2% and 1.7% (Katzman et al. 1999; Weber and Knopf 2006; Naidich et al. 1985; Eskandary et al. 2005). Pediatric studies especially the one of Al-Holou et al. who reviewed MRI of 11,738 patients under 18 years of age found a prevalence rate of 2.6% (Al-Holou et al. 2010). Pascal-Castroviejo et al. reported a higher prevalence in their series (13%) (Pascual-Castroviejo et al. 1991). Al-Holou et al. found two peaks of prevalence at 1 year (3.8%) and 5 years (4.6%). Most patients with arachnoid cysts often present during the first two decades of life. Large ones and those symptomatic are frequently seen in younger ages contrary to incidental ones that do not have age preference. Some appear with spontaneous bleeding inside the cyst or bleeding/rupture in subdural or extradural spaces (Martínez-Lage et al. 2011).

The sex ratio is nearly 2:1 in favor of males, especially in middle fossa arachnoid cysts (Oberbauer et al. 1992; Pierre-Kahn and Sonigo 2003; Al-Holou et al. 2010; Candela et al. 2015; Martínez-Lage et al. 2011). There is no sex predilection for location. Most of the time, the arachnoid cyst is single (93% in the series of Huang et al.).

Twenty percent of the cysts in Al-Holou et al. 's series were larger than 5 cm. Those in the anterior fossa were larger than cysts found in other locations.

Supratentorial cysts are found in 62–85% and infratentorial in 15–38% (Tables 3 and 4).

The middle fossa is the most common location with a rate between 25% and 72% according to the series (Huang et al. 2015; Al-Holou et al. 2010; Campistol Plana et al. 1983; Oberbauer et al. 1992; Spacca et al. 2010; Wester 1999), with a left side predominance.

Clinical Presentation

Most of arachnoid cysts are asymptomatic and discovered incidentally. The apparition of clinical symptoms due to the cyst depends on its location, evolution, etc. It is difficult to attribute a percentage of symptomatic cysts according to the literature because of unspecific, insidious, non-related symptoms that lead to diagnostic imaging. Intracranial arachnoid cysts typically become symptomatic in patients before the age of 20 years but mostly during the first decade (Oberbauer et al. 1992). With respect to the largest series, clinical symptoms attributed to the arachnoid cyst were present in 5–25% of the cases (Huang et al. 2015; Al-Holou et al. 2010).

Hydrocephalus is not the main cause of symptoms in arachnoid cysts, and there is an abundance of clinical manifestations. The commonest and

Table 3 Clinical manifestations in arachnoid cysts (except incidental)

Signs/symptoms	Combined literature data (%)	Huang et al. (2015) (488 patients) (%)	Al-Holou et al. (2010) (309 patients) (%)
Hydrocephalus	2–80	–	33.3
Macrocephaly	7–50	11.4	38.1
Headache	33–49	48.8	33.3
Increased intracranial pressure	8–49	8.1	19
Focal signs (motor, phasic)	2–38	–	4.8
Developmental delay	2–25	–	23.8
Seizure	5–24	23.6	4.8
Visual disorder	2–14	1.6	–
Behavioral/psychiatric disorders	7–10	6.5	9.5
Endocrine disorders	5–6	–	–

Table 4 Anatomical distribution of arachnoid cysts

Location		Combined literature data (%)
Supratentorial		**62–85**
	Middle cranial fossa	25–72
	Convexity	4–17
	Interhemispheric	0.4–15
	Suprasellar	1.4–21
	Quadrigeminal	1–10
	Intraventricular	0–5
Infratentorial		**15–38**
	Midline cerebellar region (Retrocerebellar, supracerebellar)	5–37
	Cerebellopontine angle	4.3–27
	Lateral cerebellar region	7–24

unspecific presenting symptom is headache, which can be due to the direct cyst pressure, intracranial hypertension, or hydrocephalus. Headache is present in 14–20% of arachnoid cyst cases but exists in 50–90% of symptomatic cysts.

Infants and young children may present with macrocrania, split sutures, developmental delay, and irritability. Older children may manifest intracranial hypertension's signs such as headaches, nausea and vomiting, blurred vision, abducens nerve paresis, dizziness, papilledema, behavioral problems, and psychiatric disorders (Martínez-Lage et al. 2011).

Temporal arachnoid cysts seem to present mostly before the age of 16 (Boop et al. 1998). They can produce headache, dizziness, psychomotor delay, behavioral abnormalities or other problems in school, phasic disorders, hemiparesis, partial or general seizures, and ventriculomegaly (Arai et al. 1996; Rocco et al. 2003; Fewel et al. 1996; Levy et al. 2004; Naidich et al. 1985; Oberbauer et al. 1992; Pascual-Castroviejo et al. 1991; Shim et al. 2009; Tamburrini et al. 2008; Zada et al. 2007). More than in other locations, cysts of the middle cranial fossa can undergo a posttraumatic or spontaneous intracystic or subdural hemorrhage or realize a hygroma by communication or rupture in the subdural space. Spontaneous resolution of the cyst following rupture has been described.

Suprasellar cysts usually produce hydrocephalus, macrocephaly, chiasm, optic or oculomotor nerve compression (papilledema, optic atrophy, bitemporal field cuts), endocrine alterations by compression of the pituitary stalk or the hypothalamus, developmental delay, motor deficits, and sometimes the "bobble-head doll" syndrome, which is pathognomonic (Erşahin et al. 2008; Hinojosa et al. 2001; Pierre-Kahn et al. 1990b; André et al. 2016; Gui et al. 2011).

Posterior fossa cysts usually manifest by hydrocephalus and intracranial hypertension and cerebellar syndrome such as ataxia, dysmetria, intention tremor, and nystagmus or less frequently with diverse cranial nerve paresis (vertigo, diplopia, hearing loss, trigeminal neuralgia, facial paresis, or hemifacial spasm) and brain stem compression (pyramidal signs, respiratory disorders) (Di Rocco et al. 1981; Galarza et al. 2010; King et al. 2010; Oberbauer et al. 1992; Zada et al. 2007). Cysts of the cerebellopontine angle usually have a long history of slowly evolving symptoms referable to stretching of cranial nerves and distortion of the cerebellum.

Quadrigeminal cysts may present, besides hydrocephalus that can appear early, nystagmus, Parinaud's syndrome, hearing disturbances, or, rarely, motor deficit.

Intraventricular cysts may present with macrocephaly and hydrocephalus in infants, focal seizures, ataxia or other gait disturbances, blurred vision or diplopia, and with position-related headaches in older patients (Martinez-Lage et al. 1992; Tamburrini et al. 2007).

Interhemispheric cysts can have a unique clinical presentation pattern with macrocrania, seizures, and developmental delay, in relation with the frequently complex associated cerebral malformations such as focal or diffuse cerebral dysplasias, tonsillar descent, agenesis of the corpus callosum, and abnormal midline septi (Martínez-Lage et al. 2011).

General Anatomical Classification

Different classifications of arachnoid cysts are found in the literature and are based on localization, morphology, and evolution of the cyst and will be treated in each subchapter. It is important to take into account the anatomical variability of arachnoid membranes and cisterns and the possible occupation of several cisterns or regions according to the size of the cyst.

First of all, arachnoid cysts are divided between supratentorial and infratentorial. Those supratentorial contain arachnoid cysts of middle fossa (temporal), convexity, hemispheric, interhemispheric, suprasellar, intraventricular, and quadrigeminal. Those infratentorial have many classifications in the literature.

All authors agree to differentiate arachnoid cysts of the posterior fossa from other malformations such as mega cisterna magna or Dandy-Walker and its variant (see part "Prenatal and Neonatal Hydrocephalus", chapter ► "Hydrocephalus Associated with Cerebral Malformations").

Radiology

Diagnosis of arachnoid cysts can be easily made by currently imaging modalities, mainly CT and MRI.

Ultrasound (Fig. 1)

The echographic pregnancy monitoring has allowed a major detection of intracranial lesions where they appear as hypoechogenic (Bretelle et al. 2002; Cassart et al. 2008; Chinn et al. 1991; Hogge et al. 1995; Malinger et al. 2007, 2008; Pilu et al. 1997). Prenatal ultrasound can detect hydrocephalus in early periods of gestation. Figure 1 shows an interhemispheric and a posterior fossa arachnoid cyst on prenatal ultrasound.

CT Scan (Fig. 2)

Arachnoid cysts appear hypodense, with CSF density, without contrast enhancement. Adjacent calvarial remodeling and hypoplasia of the adjacent brain parenchyma are common, especially in middle cranial fossa arachnoid cysts. Figure 2 shows a suprasellar arachnoid cyst associated to hydrocephalus on a CT Scan.

MRI (Figs. 3 and 4)

They appear hypointense on T1 sequences and hyperintense on T2, because they are usually isointense with CSF. The demarcation of the cyst surface is well contoured and in intimate relationship with subarachnoid cisterns, without enhancement after gadolinium injection, which distinguish them from other cystic lesions. The signal of the adjacent brain is of normal intensity. In diffusion they reveal a low signal secondary to high water diffusivity and high apparent diffusion coefficient (ADC). It has been demonstrated that diffusion-weighted sequences can help to distinguish arachnoid cysts and epidermoid tumors from cysts of the middle cranial fossa, clivus, and cerebellopontine angle (Tsuruda et al. 1990). Suprasellar arachnoid cysts need to be differentiated from craniopharyngiomas and Rathke's cleft cysts. Intraventricular arachnoid cysts must be differentiated from epidermoid, dermoid, and parasitic cysts.

Concerning the pathophysiology of CSF flow perturbation, MRI with CSF flow sequences can be very useful. Several MRI sequences can qualitatively assess and quantify pulsatile CSF flow in regions of interest. MRI coupled to the neuronavigation is a very useful technique, helping the intraoperative orientation inside cysts, ventricles, and cisterns, because of the distorted anatomy.

As important as the preoperative planning, the postoperative imaging by MRI allows the assessment of the patency of stomas through the various fenestrations and the global pathway of CSF with resolution of eventual preexisting hydrocephalus. The second reason to perform postoperative MRI is the detection of possible complications: subdural hygroma or subdural, intracystic, and intraventricular hematoma. Long-term radiological follow-up allows the observation of the slow decrease of the cyst's size and excludes

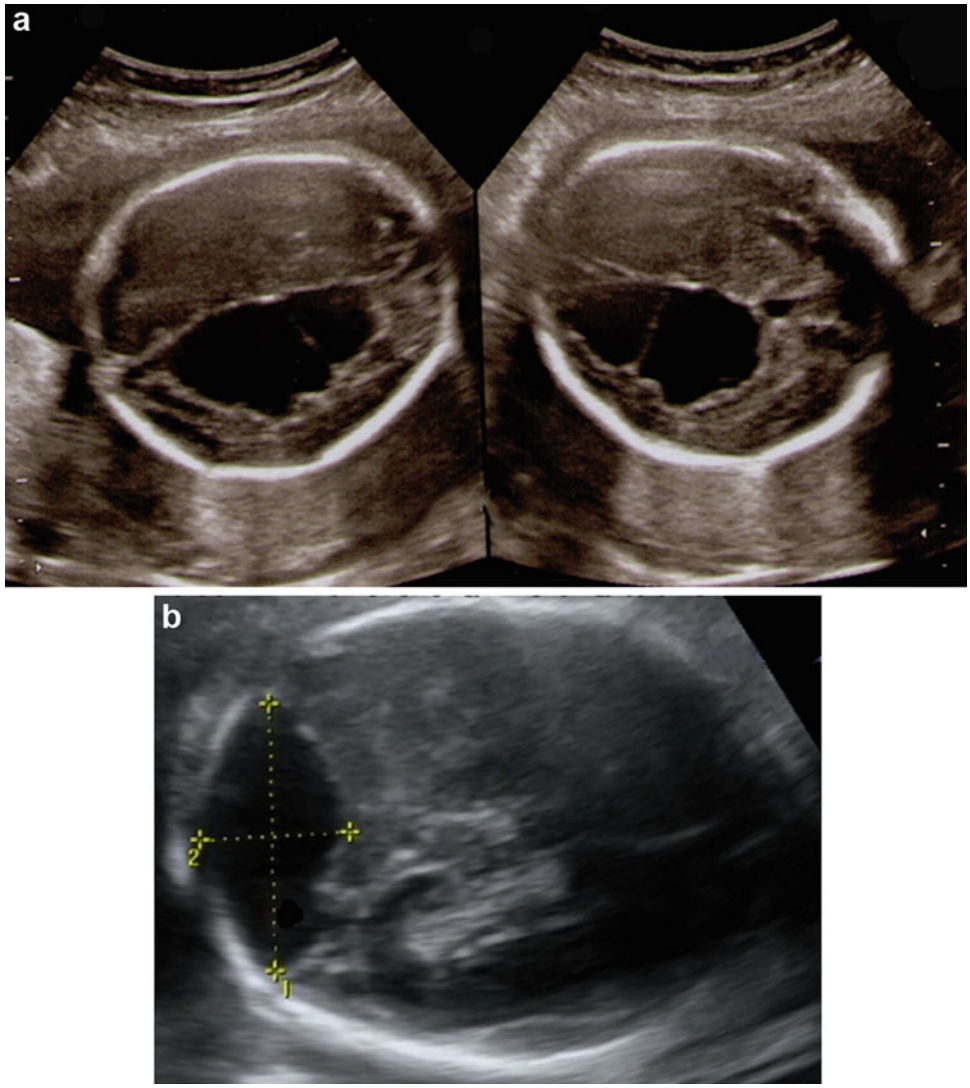


Fig. 1 Axial sections of prenatal ultrasounds showing (**a**) an interhemispheric arachnoid cyst. (**b**) A posterior fossa arachnoid cyst

recurrences. Figure 3 shows a posterior fossa arachnoid cyst on pre- and postnatal MRI. Figure 4 shows an interhemispheric arachnoid cyst on prenatal MRI.

Cisternography

Isotope cisternography and ventriculography were first described during the 1960s to study the cerebrospinal fluid circulation. A radionuclide is injected into the lumbar subarachnoid space, the progress of the tracer's diffusion through the CSF is recorded by a nuclear medicine gamma camera immediately, at 6 h and at 24 h. The first who described the technic for an arachnoid cyst was Goluboff (1973). Indeed, he discovered a posterior fossa arachnoid cyst obstruction of natural CSF pathways and stagnation of the tracer in a large volume of the posterior fossa, confirmed by subsequent surgery.

CT cisternography has been described for the first time during the 1970s. Metrizamide is a non-ionic low-osmolar radiopaque contrast agent that is

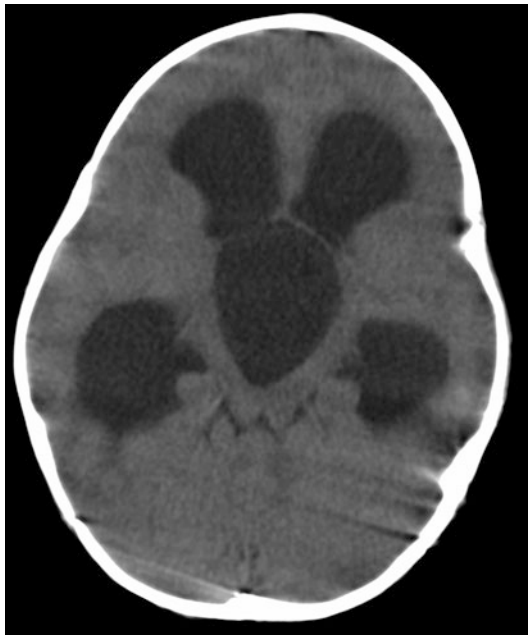


Fig. 2 Axial section of computed tomography realized in emergency showing acute obstructive hydrocephalus with a suprasellar arachnoid cyst

injected into thecal sac by lumbar puncture. Then a computed tomography is performed. Its application for arachnoid cysts was described at the end of the 1970s and beginning of 1980s. Galassi et al. used it to erect a classification and understand the pathophysiology of arachnoid cysts of the middle cranial fossa (Galassi et al. 1982).

Radionuclide cisternography is more sensitive but has poor anatomic resolution compared to CT cisternography.

These methods have long been used for preoperative strategies but are now almost obsolete.

Epidemiology of Hydrocephalus in Arachnoid Cysts

The incidence of hydrocephalus in arachnoid cysts goes from 2% to 80% of cases depending on the cyst location. About prenatal diagnosed cysts, Pierre-Khan and Sonigo found an incidence of 2% of hydrocephalus and 16.6% of ventriculomegaly (Pierre-Kahn and Sonigo 2003). During postnatal follow-up of the same patients, they found up to 17% of hydrocephalus.

Posterior fossa and midline arachnoid cysts often evolve with symptoms of hydrocephalus with respect to other locations such as Sylvian cysts where it is exceptional (Pierre-Kahn and Sonigo 2003) (5% of 54 children). Marinov et al. reported hydrocephalus in 75% of their series of 58 patients, only in the cysts of the midline and posterior fossa (Marinov et al. 1989). Fewell et al. found 40% of hydrocephalus in 102 arachnoid cysts, with 80% of their posterior fossa and 90% of their suprasellar cysts (Fewel et al. 1996). So 60% of the midline arachnoid cysts of their series developed hydrocephalus. In a series of 170 pediatric cases, all the suprasellar and all the quadrigeminal cysts had hydrocephalus but only half of those in posterior fossa (Shim et al. 2009). Al-Holou et al. reported only 2% of hydrocephalus requiring treatment in the follow-up of their series of 309 patients, but 93% of the cysts were asymptomatic (Al-Holou et al. 2010). Galarza et al. found also only 30% of hydrocephalus of ten patients with posterior fossa cysts and tonsillar descent (Galarza et al. 2010). In a series of eight patients operated on for infratentorial arachnoid cyst, 25% was for arrested hydrocephalus, and all of them had macrocephaly (Di Rocco et al. 1979). Zada et al. reported an incidence of hydrocephalus of 14% in 44 patients under 2 years of age (Zada et al. 2007). Martínez-Lage et al. had 32% of hydrocephalus in a series of 99 cysts. André et al. reported an incidence of 43% of hydrocephalus in a series of 35 suprasellar arachnoid cysts.

Pathophysiology of Arachnoid Cyst Formation

Normal dural anatomy is extensively described in part, "Basics, Anatomy, Embryology, Physics, and Morphology" chapter ▶ "Anatomy of the Cranial and Spinal Meninges."

It is important to note that subarachnoid spaces are crossed by networks of fine, continuous, sheet-like trabeculae that divide the space into compartments, facilitating a more directional CSF flow. Trabeculae enclose small blood vessels and

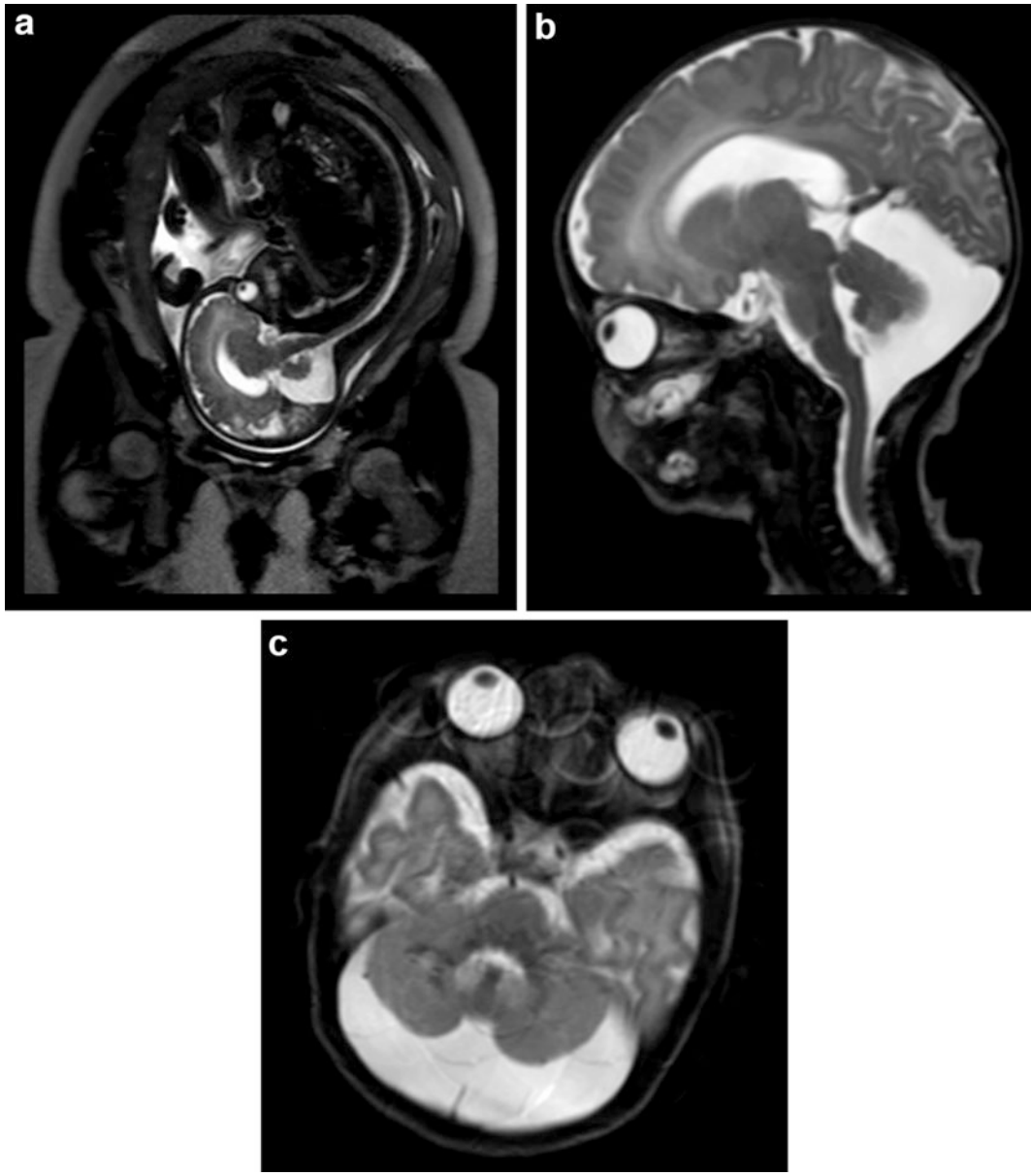


Fig. 3 Posterior fossa arachnoid cyst on (**a**) prenatal T2-weighted MRI, sagittal section (**b**, **c**) Completed by a postnatal MRI. (**b**) sagittal T2-weighted section. (**c**) axial T2-weighted section

adhere to the surface of larger blood vessels and nerves in the subarachnoid space. Liliequist's membrane is the most important part of the trabecular arachnoid and is directly concerned by suprasellar arachnoid cysts. Various descriptions about its morphology, orientation, attachment, classification, and relationship with surrounding structures have been studied.

History

In 1831 Bright makes the first description of an arachnoid cyst in his volume *Diseases of the brain and nervous system* in the *serious arachnoid cysts* section (Bright 1831). He describes them as cysts connected to and surrounded by layers of arachnoid. In his book he reported two cases and believed that they were chronic formations with

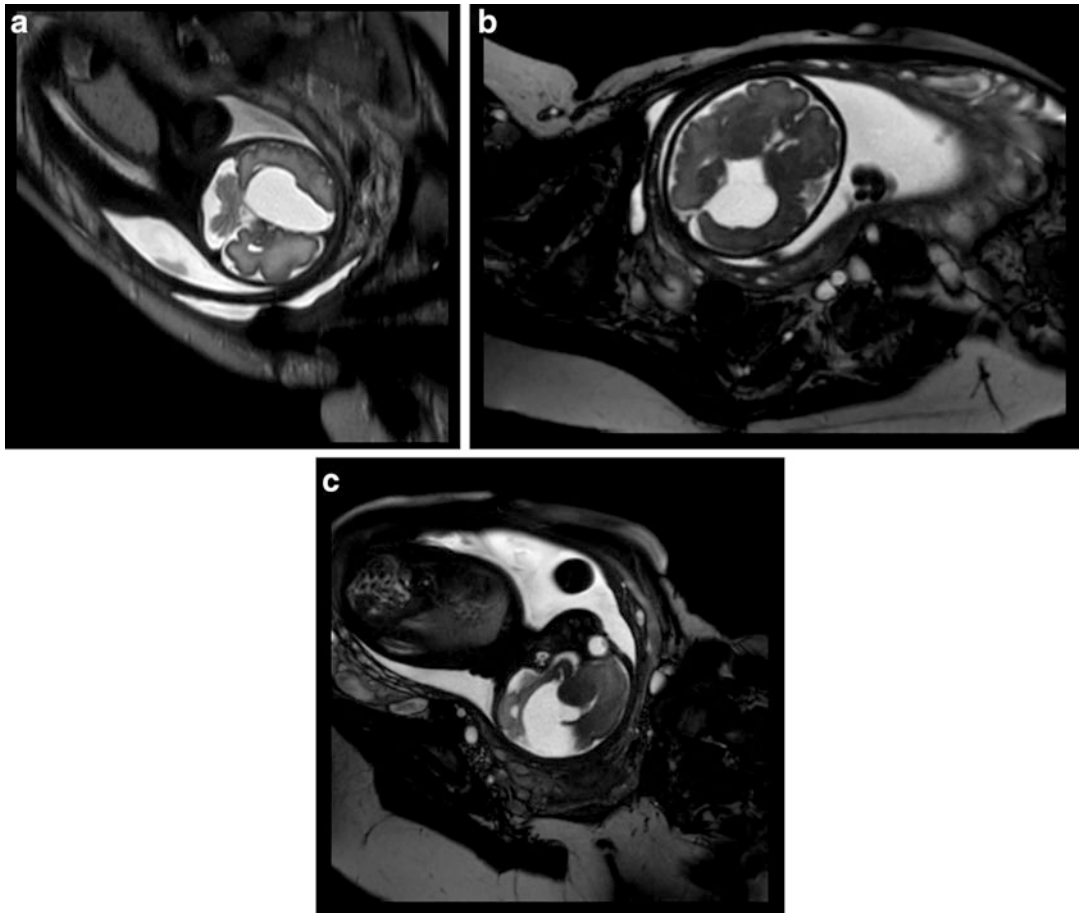


Fig. 4 Interhemispheric arachnoid cyst on a prenatal T2-weighted MRI. (**a**) Coronal section. (**b**) Axial section. (**c**) Sagittal section

a low potential for growth. Later, in the twentieth century, the use of microscopic techniques will confirm that these cysts are really intraarachnoidal (Starkman et al. 1958). In 1978, Rengachary et al. showed a photomicrograph illustrating Bright's observation (Rengachary et al. 1978). They observed splitting of the arachnoid membrane at the margin of the cyst and a lack of trabecula within the cyst, whose walls contained hyperplastic arachnoid cells and a thick layer of collagen. So they confirmed Bright's finding that arachnoid cysts are raised within arachnoid membrane and not within the subarachnoid space.

However, the pathogenesis has been more controversial. Several theories appear about the origin of these cysts, considering secondary to trauma, infection, or bleeding (Trowbridge and French 1952).

The pathogenesis of arachnoid cysts is not really known. Often the adjacent brain is hypoplastic, and sometimes the cyst is accompanied by deformity of the adjacent cranial bone, even with apparently normal intracranial pressure (Conde Sardón 2015). It would imply that arachnoid cysts could be secondary to a defect in the fetal development. The middle fossa cysts are often accompanied by abnormalities of the Sylvian veins, which imply a failure between weeks 6 and 10 of the fetal development (Sato et al. 1983, 1991; Sugata et al. 2003). Sugata supports this theory of the early failure because giant posterior fossa cysts are accompanied by an elevation

of the tentorium, the straight sinus, and the torcular. With the same point of view, Robinson describes arachnoid cysts as congenital but secondary to agenesis/hypoplasia of the temporal lobe. It is his concept of "Temporal agenesis syndrome" (Robinson 1964), but this theory is now considered obsolete.

Trowbridge and French have also attributed to arachnoid cysts a congenital origin. Their hypothesis is that they are formed by remnants of embryonic tissue with secretory capacity (Trowbridge and French 1952). Starkman et al. proposed that the problem doesn't come from anomaly of brain development but from an aberrant CSF flow. The CSF produces a tear in the developing arachnoidal membrane, creating diverticula and eventually capturing those diverticula and CSF (Starkman et al. 1958). Miyajima et al. propose that some suprasellar cysts are caused by dilatation of the interpeduncular cistern (Miyajima et al. 2000). Fox and Al-Mefty said that the origin of suprasellar cysts is from a diverticulum of the Liliequist membrane, imperforated because of inflammation (Fox and Al-Mefty 1980). Wester suggests that the middle fossa cysts result from an incomplete fusion of the arachnoid membrane of the frontal and temporal lobes during the development of the Sylvian fissure in early fetal life, thereby creating a non-communicating fluid compartment entirely surrounded by arachnoid membranes (Wester 1999). It is also showed that some arachnoid cysts develop around tufts of ectopic choroid plexus (Schuhmann et al. 2000). A more unusual location is the ventricular system (Nakase et al. 1988; Martinez-Lage et al. 1992; Di Rocco et al. 1979). The arachnoid cysts of the lateral ventricles could be formed from the remnants of arachnoid cells trapped in the developing ventricles from the vascular mesenchyme that formed the choroid plexus through the choroid fissure. The mechanism could be similar for the cysts of the fourth ventricle. Suprasellar cysts could be formed through fissures of arachnoid making hernia through the diaphragma sellae or arachnoid remnants in the sella (Harter et al. 1980).

Most of the cysts do not increase. Becker et al. found no correlation between the age of the patient and the size of the cyst (Becker et al. 1991). Increased pressure inside the cyst has been documented (Kumagai et al. 1986). Several theories try to explain this fact. Growth of cysts has been explained by the existence of an osmotic gradient (Dyck and Gruskin 1977) or production of liquid from its walls. Go et al. have shown in their study a high level of activity of the Na/K ATPase pump and many microvilli with similar form to the secreting mesothelium of the arachnoid granulations and villi (Go et al. 1986). In a few cases, it has been shown plumes of choroid plexus incorporated into the cyst wall (Handa and Bucy 1956; Schuhmann et al. 2000).

More recently, a valvular mechanism with liquid flow in one direction called slit-valve mechanism has been demonstrated. This valve would allow entry in the cyst during cardiac systole and accumulation during the periods of elevation of intracranial pressure. This communication has been shown by cine MRI and direct visualization by endoscopic procedures (Santamarta et al. 1995; Schroeder and Gaab 1997). In the case described by Schroeder and Gaab, a suprasellar cyst was penetrated by the basilar artery with a slit-valve mechanism formed by an arachnoid membrane surrounding the artery. Cysts may or may not communicate with subarachnoid space. Probably the non-communicants result from communicant ones where the small entry point has been closed secondarily. This would explain why many of the large cysts of the middle fossa commonly are not communicant (Galassi et al. 1982). Some articles report that coexistent or preceding hydrocephalus could play an important role in arachnoid cyst expansion (Martínez-Lage et al. 1999; Di Rocco et al. 1981). In fact, increased hydrocephalus or ventriculomegaly after the placement of a cyst shunt or, on the contrary, increasing size of arachnoid cysts after a ventricular shunt placement has been described. It may be because of CSF dynamic perturbation of the physiological pressure balance between subarachnoid spaces and ventricular system.

Embryopathogenesis

The endomeninx or primitive meninx, which is the precursor of the dura mater and arachnoid mater, surrounds as a loose layer of mesenchyme,

the neural tube during the early embryonic period. At the 15th week of gestation, the subarachnoid space can be detected, and then the rupture of the rhombic roof allows the CSF circulation. This flow contributes to the arachnoid development creating trabeculae that form the "spider web" structure of the subarachnoid space. An anomaly in this process could explain the accumulation of CSF within the arachnoid and form an arachnoid cyst (Martínez-Lage et al. 2011; Pascual-Castroviejo et al. 1991; Naidich et al. 1985).

Histological Aspects (Fig. 5)

Histological studies (García-Conde and Martín-Viota 2015) show arachnoid cysts as abnormal accumulation of CSF between the two layers that constitute arachnoid (Zada et al. 2007). The walls of arachnoid cysts are composed of a vascularized collagen membrane with flattened arachnoid cells without epithelial or glial wall (Naidich et al. 1985). They are slightly thicker because of this collagen accumulation and sometimes also because of hyaline changes. The subarachnoid space below arachnoid cysts is obliterated, and no signs of inflammation are observed, unless if a prior hemorrhage exists which relates to the presence of blood vessels, either on the cyst walls or inside it. At the edges of the cyst, both layers are merged to constitute the normal arachnoid membrane (Rengachary and Watanabe 1981). The underlying brain is normal but can be compressed depending on the size and topography of the cyst. When the compression is important, the brain parenchyma may present fibrillar gliosis and neuronal atrophy, without evidence of a primary brain tissue lesion. However, in posterior fossa cysts remnants of choroid plexus are observed. In suprasellar and prepontine cysts, we can find neuroglial elements which remind those found on the floor of the third ventricle (Pradilla and Jallo 2007). Suprasellar arachnoid cysts have been attributed to an upward extension of the membrane of Liliequist (Fox and Al-Mefty 1980). Figure 5 shows histological sections of the wall of a posterior fossa arachnoid cyst.

Treatment

General Principles

Surgical treatment is not indicated in case of asymptomatic or incidentally discovered cysts or in case of symptoms clearly unrelated with the cyst. Almost all investigators agree in treating patients with associated hydrocephalus, raised intracranial pressure, subdural hematoma, or focal clinical signs. Other situations such as associated subdural hygromas, development delay, or epilepsy remain debated and should be evaluated case by case. The choice of the most appropriate surgical approach for the treatment of pediatric arachnoid cysts remains widely debated. No prospective randomized (controlled) clinical trial exists comparing treatment options.

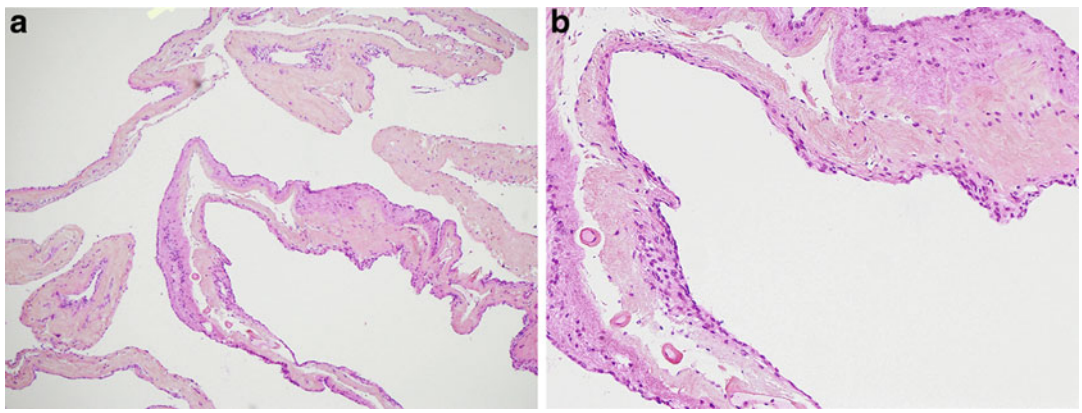


Fig. 5 Histological sections of the wall of an arachnoid cyst of the posterior fossa (with HES stain), comprising a fibrovascular stroma and a cubo-cylindrical lining. (**a**) 5× magnification. (**b**) 10× magnification

Different options include open surgery with cyst fenestration or excision, cyst and/or ventriculoperitoneal shunting, endoscopic fenestration, or a combination of these techniques.

In the 1980s, most of the articles discussing the treatments favored the use of shunts, probably because of a high rate of morbidity and mortality of operative craniotomies during the 1960s and 1970s. Then micro neurosurgery has taken over large craniotomies, and the techniques have evolved, reducing significantly the complications. So, the trend reversed in favor of microsurgery because the belief was that fenestrate a cyst is to cure it (Fewel et al. 1996) while complications of shunting revealed in the forefront, with its burden of malfunction, shunt dependency, etc. In the end of the 1990s, neuroendoscopy offered a mini-invasive alternative to fenestration, accelerating the trend to avoid shunts and craniotomies whenever possible.

Microsurgical Procedure

Authors in favor of open surgery with cyst fenestration or excision defend a success rate (good outcome) of 75% avoiding the risks of shunt dependence and complications (Oberbauer et al. 1992; Fewel et al. 1996; Galarza et al. 2002; Gangemi et al. 2011) but with a recurrence rate around 30%. Probably open surgery offers a better anatomical control and the possibility of using microsurgical instruments for better excision of the membranes; the capacity to open larger fenestrations of the cyst in basal cisterns, subarachnoid space, and ventricles; and better hemostasis. But it is important to think about the risks of a craniotomy in the benefit-risk balance, mostly in pauci symptomatic patients. In old studies, the cyst recurrence rate was not so low (Choux et al. 1978; Sprung and Mauersberger 1979), and several complications were reported such as meningitis, subdural hematoma, seizure, hemiparesis, and death (Choux et al. 1978; Aoki and Sakai 1990; Ciricillo et al. 1991; Fewel et al. 1996). Open approach is aggressive also because of brutal decompression of the brain, due to the necessary intraoperative drain of the cyst, which can provoke intraparenchymal hemorrhage and subdural hematoma/hygroma. In case of associated hydrocephalus, there is no proof of utility of this kind of single approach, and the necessity of secondary CSF derivation is estimated between 24% and 80% (Raffel and McComb 1988; Ciricillo et al. 1991; Fewel et al. 1996).

Shunting

Some authors recommend to use initially a ventricular shunt especially in children younger than 2 years who present with hydrocephalus or macrocephaly (Marinov et al. 1989; Zada et al. 2007). Concerning cystoperitoneal shunts, one of the advantages is the possibility to perform a gradual reduction of cyst size probably better than sudden microsurgical decompression, helping the brain re-expansion.

The Necker's team, in 2002, advocated the inutility of the addition of a ventriculoperitoneal shunt in case of implantation of a cystoperitoneal shunt, adding a double-risk of postoperative shunt complications, with pressure differences between the various compartments (Pierre-Kahn et al. 2002). They showed the regression of an infratentorial cyst, the reopening of the aqueduct, and the decrease of hydrocephalus. Pierre-Khan reported also the case of expansion of a suprasellar cyst after the derivation of hydrocephalus (Pierre-Kahn et al. 1974).

The weaknesses of shunting are infections and hemorrhage (Arai et al. 1996; Aoki and Sakai 1990; Raffel and McComb 1988). But the major drawback of extracranial CSF shunting is the propensity of these methods for developing over drainage (Martínez-Lage et al. 2011), especially in middle fossa cysts with shunt dependency like the slit cyst syndrome, cranioencephalic disproportion, and acquired Chiari malformation (Caldarelli et al. 2009; Di Rocco 2010; Martínez-Lage et al. 2009; Shim et al. 2009; Sunami et al. 2002; Zada et al. 2007; Alexiou et al. 2010). Also we should take into account the risk for surrounding structures of a shunt placement in critical locations such as suprasellar or posterior fossa (Ali et al. 2014). The propensity for valve dysfunction with a need of reoperation is estimated at 30% in the literature, but Germano et al. had a lower rate using programmable valves (Germanò et al. 2003).

Endoscopic Procedure

In 1974 Pierre-Kahn et al. described the procedure of fenestration of a suprasellar cyst with a leukotome, under ventriculographic/radioscopic control (Pierre-Kahn et al. 1974). The same author described four cases of cyst fenestration under endoscopic control. In 1988 Auer had already described endoscopic treatment of an arachnoid cyst. Endoscopic neurosurgery has since been increasingly used in the management of arachnoid cysts especially with associated hydrocephalus or for the insertion of intracystic catheters (Abbott 2004; Talamonti et al. 2011). Abott described amply the technique in his article (Abbott 2004). He reminds us that the walls of arachnoid cysts, rich in collagen, may be difficult to penetrate but also to see through them and avoid lesion of vascular-nervous structures. The necessity of scissors or knifes is usual, followed by inflations of a balloon in the hole or its enlargement by instrumental dissection. The goal is to perform the largest possible safe fenestrations. According to him, as for any minimal invasive surgery, the most important thing is not to hesitate to convert to an open surgery if craniotomy is needed, because of failure, hemorrhagic complication, etc. He reminds us of the need to anticipate the possibility of conversion to open surgery, informing the patient and his family preoperatively. Authors referred also the possibility, if necessary, of the introduction of a second operative channel, to use more instruments, or to place a catheter precisely in the cyst or ventricles (Abbott 2004; Jallo et al. 1996; Fujio et al. 2016).

Neuroendoscopy is nowadays considered the best compromise of definitive treatment and shunt independence with least surgical morbidity (Nowosławska et al. 2006; Shim et al. 2009; Tamburrini et al. 2007; Pierre-Kahn et al. 2002). With respect to open surgery, there is less surrounding tissue injury and a shorter length of stay due to minimally invasive treatment. It is possible to perform fenestration to the basal cisterns, the ventricles, or both (cysto-ventriculocisternostomy). Whereas microsurgical procedures require an open environment work, endoscopic procedures are conducted in liquid environment within or around the cyst and cisterns, characterized by the absence of cystic draining and sudden cerebral decompression.

Sophisticated advances include the use of intraoperative neuronavigation or ultrasonography. These operating aids can help to manage multiloculate hydrocephalus. The principal drawbacks are the need of specialized training which requires a learning curve (Martínez-Lage et al. 2011; López et al. 2015; Abbott 2004) and difficulties in identifying anatomical structures like blood vessels in middle fossa and suprasellar cysts (Cinalli et al. 2007; Martínez-Lage et al. 2011). According to Abbott, two very important success factors are the preoperatory planning, to find the optimal trajectory of the channel and endoscope, and the experience of the assistant, who will navigate the scope, while the surgeon use the instruments (Abbott 2004). The complication rate of neuroendoscopic procedures is low in the literature, but in children some complications can considerably deteriorate the outcome, such as infections, subdural hematoma/hygroma, and CSF leaks (Cinalli et al. 2007; Navarro et al. 2006). The percentage of these complications, which is less than 15%, may be reduced by the transcortical entrance rather than intracystic direct entrance.

Discussion on Treatment

Other surgical possibilities are reservoir placement (Kawamoto et al. 2007), a stereotactic puncture, or a shunt to make a communication between the cyst and the ventricles to prevent overdrainage (McBride et al. 2003).

To guide the surgical decision, it is very important to keep in mind the outcome assessment focusing on quality of life, considering the need for reoperation and morbidity/mortality of each technique. Any of the three main techniques has its advantages and disadvantages that vary between the cysts' characteristic and location, habits, preferences, and experience of the surgeon/center. In the literature, these three techniques, taken individually, have a success rate with good clinical and radiological outcome around 80% or more. Maybe the limitations of neuroendoscopy will overcome with advancements in neuroendoscopy instruments and techniques, and surgical treatment for

arachnoid cysts may become entirely endoscopic (Ali et al. 2014). It is important to keep in mind that each situation with arachnoid cyst is different because of their characteristics in location, size, extension, and clinical consequences. Advantages of one approach for one particular cyst may be a disadvantage in another one.

Ali et al. have not found any significant differences in outcome and quality of life of patients between treatments in their 5-year follow-up of a review of 36 series in the literature including 1324 cases (Ali et al. 2014).

Suprasellar Cysts

Pathophysiology of Hydrocephalus

Hydrocephalus may be produced by mechanical obstruction of CSF pathways by the arachnoid cyst. They can block the foramina of Monro if there is an extension through the third ventricle (Fox and Al-Mefty 1980) or rarely the aqueduct of Sylvius.

Subtypes

Some controversies persist in the literature regarding the classification and the treatment of suprasellar arachnoid cysts (Özek and Urgun 2013; Mattox et al. 2010; Gui et al. 2011; André et al. 2016; Hinojosa et al. 2001; Erşahin et al. 2008). The first description of a suprasellar arachnoid cyst was made by A. Barlow in 1935. Fox and Al-Mefty were the first to propose an origin from the Liliequist membrane (Fox and Al-Mefty 1980). Later, Miyajima et al. made a distinction between these cysts. Several subtypes of suprasellar arachnoid cysts have been described based on the anatomy (Özek and Urgun 2013; Miyajima et al. 2000; Lyu 2014). Rhoton et al. and Froelich et al. made a very complete and complex anatomical description of the basal cisterns and variability of the Liliequist membrane (Rhoton 2000; Froelich et al. 2008). The Liliequist membrane, anatomically, is located between the interpeduncular and chiasmatic cisterns. It covers the upper edge of the posterior clinoid process and the dorsum sellae. It is separated into two leaves extending upward from the dorsum sellae and across the inter oculomotor nerve's space. The interpeduncular cistern is located between cerebral peduncles and Liliequist membrane with its two leaves. Most of the above-cited authors propose a simple anatomical classification in two types: prepontine or interpeduncular. This classification made on radiological and intraoperative findings has a great impact on the surgical approach because of the position of the basilar artery. In fact, the prepontine cyst realizes a dilatation of the interpeduncular cistern, and the basilar artery would remain inside the cyst, whose walls are made by the diencephalic membrane upward and the mesencephalic one downward. The interpeduncular cyst is formed by the diencephalic membrane of Liliequist that will lead a compression of the interpeduncular cistern, leaving the basilar artery behind the posterior wall of the cyst. Usually, in all the suprasellar arachnoid cysts, the pituitary stalk and the optic chiasm are pushed upward and forward while mammillary bodies upward and backward.

André et al. have proposed an original classification not only based on the anatomical origin but also according to the clinical and radiological presentations (André et al. 2016) (Table 5). Their classification in three subtypes should permit to estimate the evolution of the cyst according to each subtype and help to choose the best treatment, if necessary. It may be also useful in the management of prenatal discovered suprasellar arachnoid cysts. Hydrocephalus can be present only in the subtype 1 and was found in all cases of this series.

Radiology (Figs. 6, 7, and 8)

MRI is key to evaluate the exact cyst location, its intimate relationships, and compression of adjacent structures, such as basilar artery, pituitary stalk, optic chiasm/tracts, mammillary bodies, third ventricle, and foramen of Monro. Classical sequences are made as T1-weighted, T2-weighted, diffusion-weighted, and, nowadays, 3D, CISS, FIESTA, or DRIVE sequences (see section "Radiology").

Table 5 Classification of suprasellar arachnoid cysts (SAC) according to André et al. (2016)

Subtype (% of cases)	Anatomical origin	Radiological presentation	Clinical consequences
SAC-1 (43%)	From an expansion of the diencephalic leaf of the Liliequist membrane or chiasmatic cistern	Blockage of both the interventricular foramen of Monro (Fig. 6)	Hydrocephalus
SAC-2 (31.5%)	From a defect of the mesencephalic leaf of the Liliequist membrane	Dilatation of the interpeduncular or prepontine cisterns (Fig. 7)	No hydrocephalus
			Severe compression of the brain stem
SAC-3 (25.5%)	Origin from other subarachnoid spaces	Asymmetrical forms (temporopolar, Sylvian fissure) (Fig. 8)	Macrocephaly without hydrocephalus

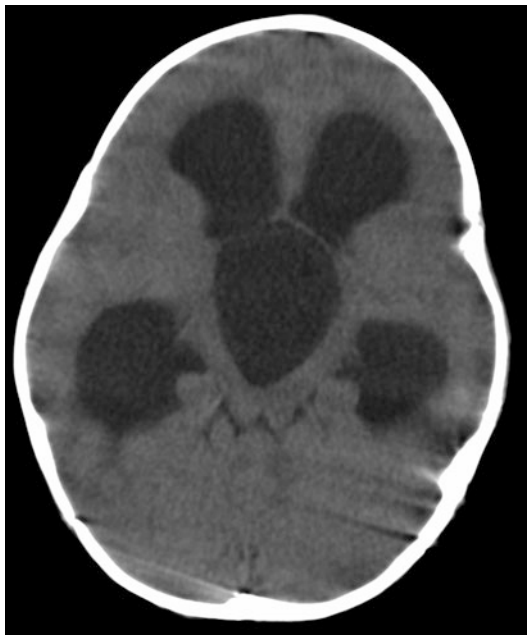


Fig. 6 Axial section of computed tomography realized in emergency showing acute obstructive hydrocephalus with a suprasellar arachnoid SAC-1 cyst. Note the Mickey Mouse appearance

These latter allow a better delineation of the cyst walls with structures cited above and particularly with the roof of the third ventricle. The so-called Mickey Mouse appearance can be seen on axial images because of the severe dilatation of the third and lateral ventricles (Özek and Urgun 2013) (Fig. 6). The preoperative imaging permits the study of the best trajectory in case of endoscopic treatment.

Obviously, MRI must have a major role in the follow-up of these patients and helps to differentiate arachnoid cysts from craniopharyngiomas or Rathke's cleft cysts. They typically have no calcifications, are larger, and grow more symmetrical. So CT scan may warrant this absence of calcifications.

Treatment/Surgical Techniques

Suprasellar arachnoid cysts with and without hydrocephalus are totally different because of their origin but also because of the surgical approach, if a treatment is needed. These cysts are on the midline and the skull base, which gives them relationships with the ventricles and the basal cisterns that are surgically natural possibilities of CSF derivation.

Microsurgical Procedure

Historically, the first technique described was the open microsurgical approach, which is efficient to obtain a collapsus of the cyst walls. During the 1980s, two studies advocated cyst wall resection as radical as possible (Konovalov et al. 1988; Jones et al. 1989). They performed transventricular-transcortical or combined subfrontal and transcallosal approaches or transcallosal approach alone (Murphy 1985). Pterional approach is also described. Other authors proposed a coagulation retraction or a simpler marsupialization (Raimondi et al. 1980).

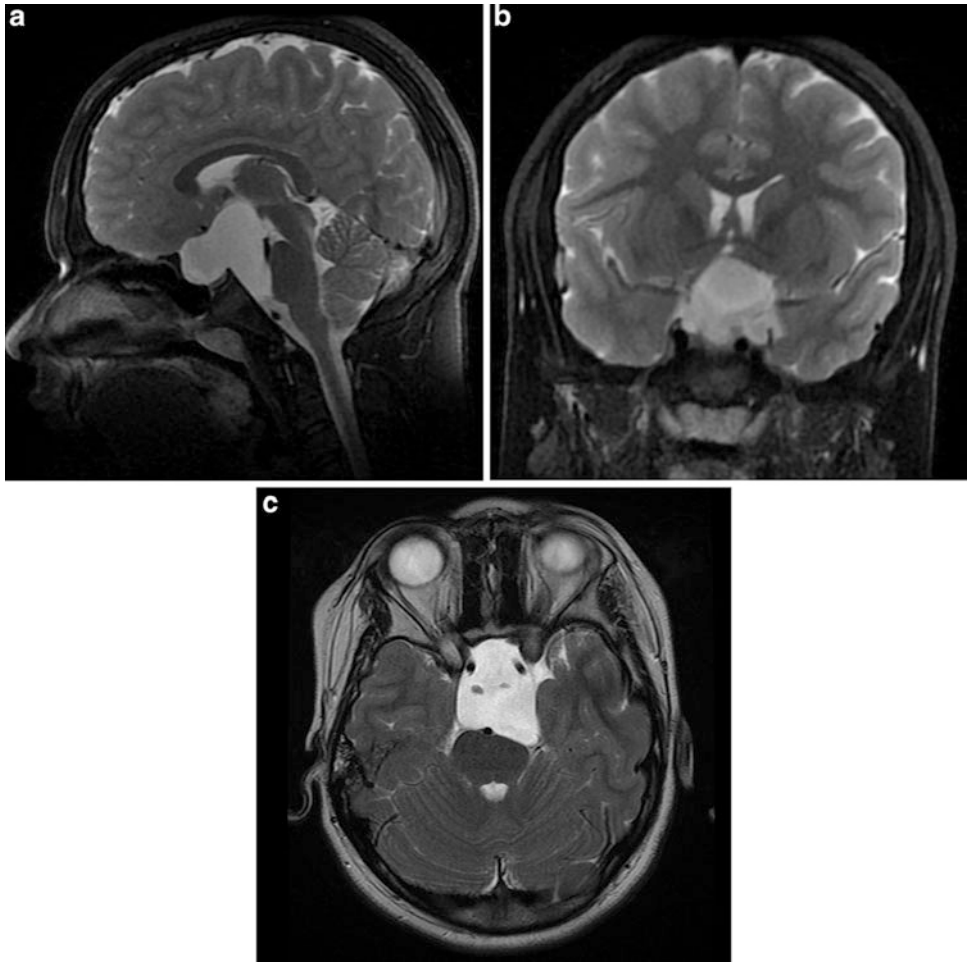


Fig. 7 T2-weighted MRI showing a suprasellar SAC-2 arachnoid cyst according to André et al. Note the dilatation of the interpeduncular and prepontine cisterns, without associated hydrocephalus (no upper extension). (**a**) Sagittal section. (**b**) Coronal section. (**c**) Axial section

Shunting

The second option is the CSF diversion by shunt. There are three possibilities: a ventriculo-peritoneal shunt, a cystoperitoneal shunt, or a ventriculo-cystoperitoneal shunt. Shunting was often used as a secondary treatment when open craniotomy was performed on suprasellar arachnoid cysts with hydrocephalus (Raimondi et al. 1980), the latter not always allowing a microsurgical ventriculocisternostomy. But shunts are burdened with morbidity attributed to the over-shunting and shunt dependence risks. The main problem of shunts in suprasellar cysts derives by the fact that the proximal catheter cannot perforate the thick cyst wall, remaining into the ventricular cavity. The progressive ventricular drainage induces a slow enlargement of the suprasellar cyst with increasing mass effect on the suprasellar structures, namely, optic chiasm and pituitary stalk. Progressive growth upward induces obstruction of both foramina of Monro, leading to the necessity of shunting also the contralateral ventricle. As a matter of fact, this technique usually ended up in a patient with bilateral drainage and slit-like ventricles and a suprasellar cyst that remained not treated.

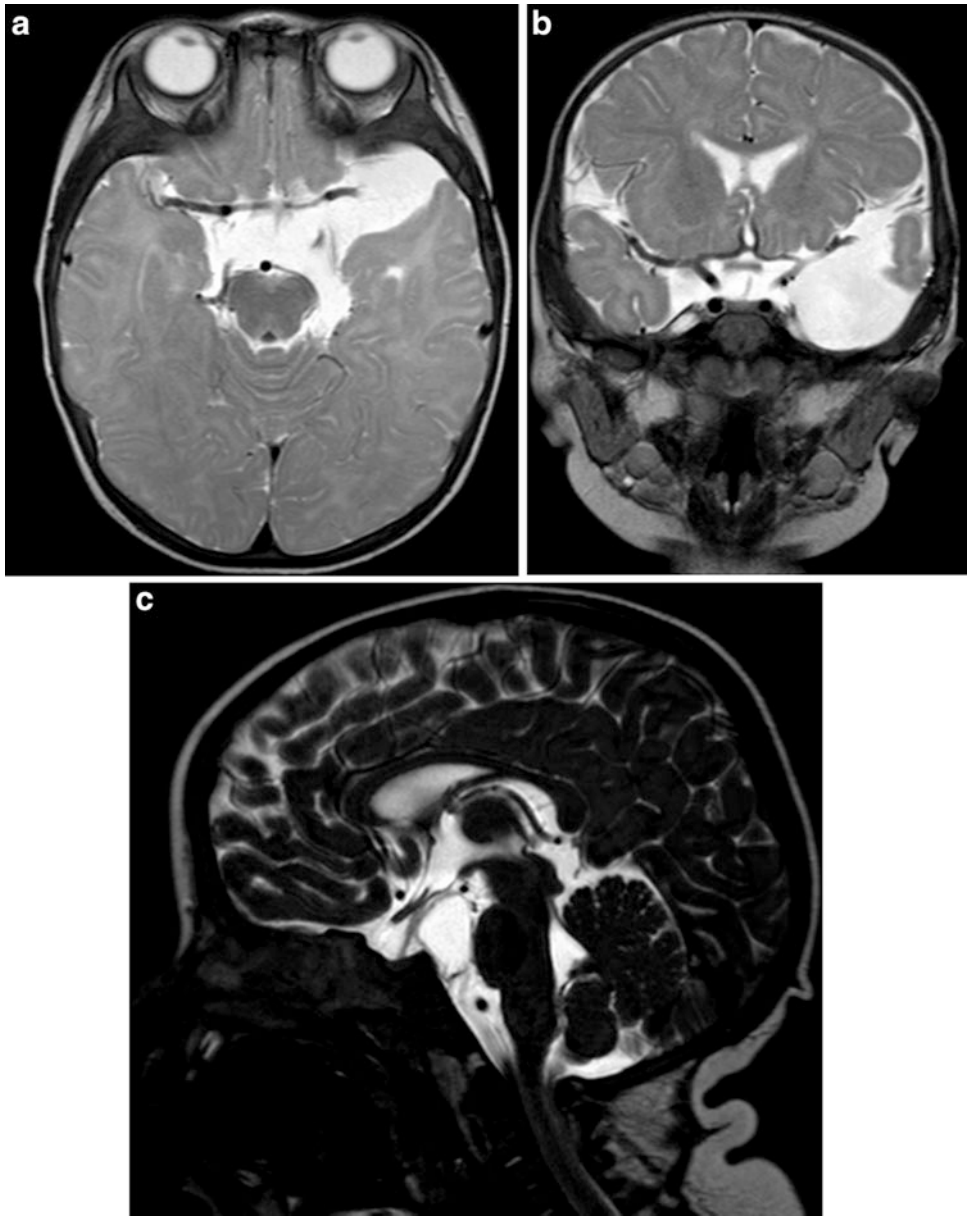


Fig. 8 MRI showing a suprasellar SAC-3 arachnoid cyst that seems to come from the interpeduncular and prepontine cisterns with a lateral extension to the middle cranial fossa. (**a**) T2-weighted axial section with the basilar artery flattened against the brain stem. (**b**) T2-weighted coronal section showing the entire occupation of the base of the middle cranial fossa. (**c**) CISS-weighted sagittal section that eliminates an extension above the midbrain (no mass effect on the third ventricle)

Endoscopic Procedure

In the 1990s, the progress of endoscopic neurosurgery (Pierre-Kahn et al. 1990b) allowed the widespread usage of the technique, and the direct surgical approach remained reserved to the suprasellar arachnoid cysts without hydrocephalus as subtypes SAC-2 or SAC-3 of André et al. (André et al. 2016). Neuroendoscopic treatment is preferred by most of the authors, in case of associated hydrocephalus (Mattox et al. 2010; Di Rocco and Zerah 2011;

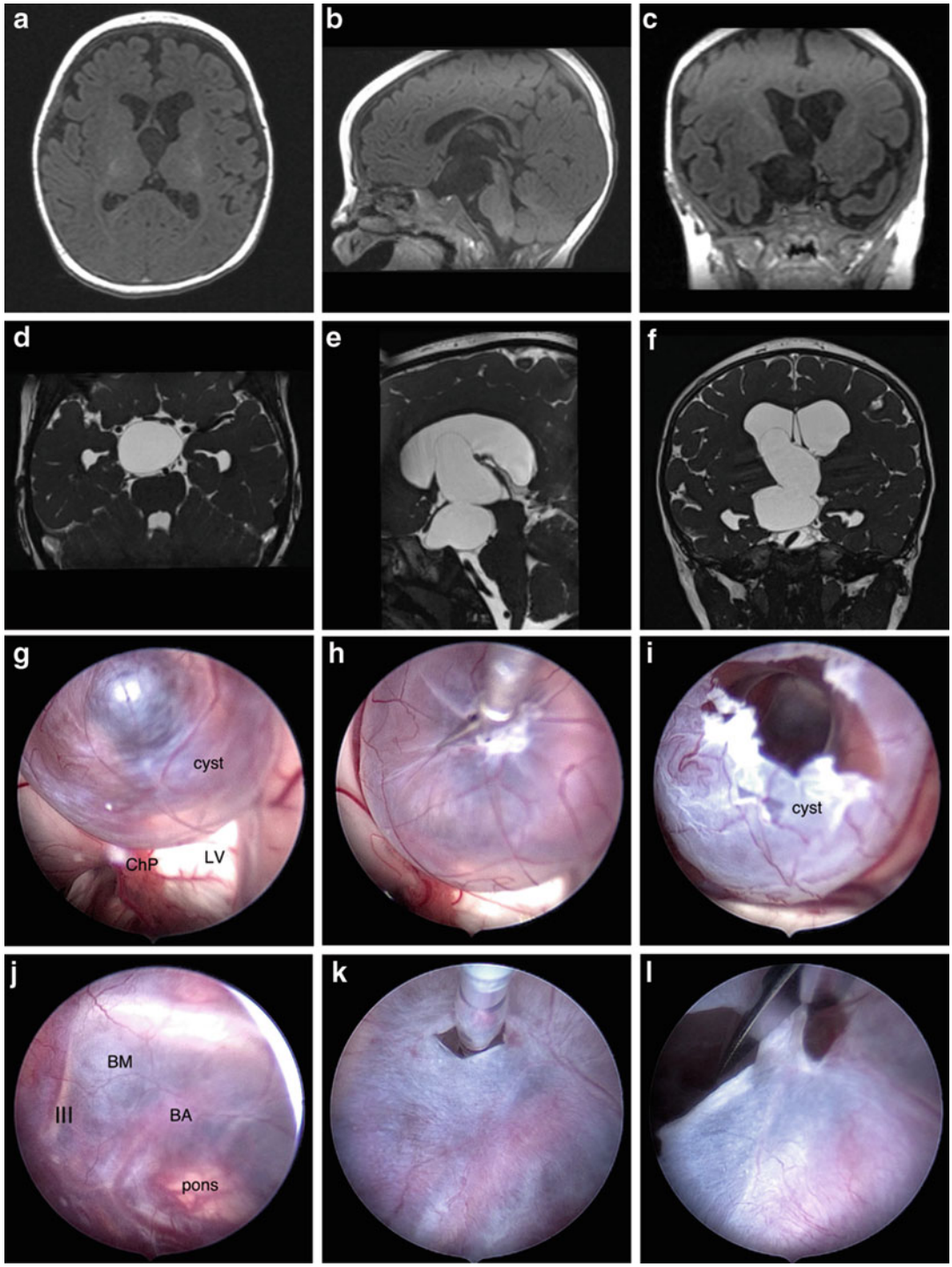
André et al. 2016; Erşahin et al. 2008; Gangemi et al. 2011; Hinojosa et al. 2001; Özek and Urgun 2013). Nowadays, the debate is placed on where to perform the fenestration. Some authors consider that a simple ventriculocisternostomy by opening the roof of the cyst (the floor of the third ventricle) is sufficient (Ogiwara et al. 2011). But the most recent comparative series have showed a significant superiority of the ventriculocystocisternostomy on the recurrence rate, on the reoperation rate, and on the MRI results with CSF flow sequences (Decq et al. 1996; Gui et al. 2011; Erşahin et al. 2008; Maher and Goumnerova 2011; Wang et al. 2004; André et al. 2016; Rizk et al. 2013; Di Rocco and Zerah 2011). One of the reasons of the success of the ventriculocystocisternostomy is that it seems to prevent the secondary aqueduct occlusion in case of incomplete decompression of the midbrain. Decq et al. noticed in their long follow-up that the upper perforation often closed with time, while the lower remained permeable, avoiding recurrences. So, the best option of fenestration allows the treatment of both the hydrocephalus and the mass effect of the cyst. The endoscopic procedures for SAC-1 are detailed in Figs. 9 and 10, while the procedure for SAC-3 is detailed in Fig. 11.

Standard Endoscopic Procedure (for Suprasellar Arachnoid Cyst Associated to Hydrocephalus) (Figs. 9 and 10)

The patient is positioned supine with the neck slightly flexed. The use of the neuronavigation system is particularly recommended as it can help to access the ventricle and estimate the presence of neurovascular structures under cystic membranes. A burr hole is made on the mid-pupillary line on the coronal suture. The right side is preferred, but in case of cyst asymmetry, the focus will be on the side where the cyst bursts into the lateral ventricle by the foramen of Monro, as it will provide more workspace and a better reopening of the ipsilateral interventricular foramen. An incision of the dura mater is made allowing its subsequent watertight closure. The rigid endoscope is introduced in the frontal horn of the lateral ventricle. The upper wall of the cyst is visualized through the foramen of Monro. In suprasellar arachnoid cysts, the upper wall consists of the floor of the third ventricle, but a fenestration can be safely performed on its translucent part. The way to achieve the fenestration is chosen according to the preoperative planning, having considered the anatomical structures lying below the membrane. As the walls of arachnoid cysts are often thick, we usually perforate them with the laser, ideally as a grid. The stoma thus produced is expanded using the double-balloon probe. Usually it is necessary to enlarge the stoma using scissors between two holes because of the hardness of the membranes. We recommend not to coagulate too much the upper wall at first, because this would reduce the size of the cyst and consecutively the overall vision of the skull base and the bottom wall of the cyst. The endoscope is introduced inside the cyst allowing the identification of anatomical structures and the bottom wall of the cyst, which is also perforated to obtain cysto-cisternal fenestration. A special attention should be paid to open the Liliequist's membranes. Large fenestration warrants the highest long-term permeability. The endoscope is removed, and attention is paid to not drain the CSF, avoiding brain, ventricular, or cystic collapse while maintaining flows through the fenestrations. The dura is sutured.

Discussion on Treatment

There is no study comparing prospectively the three main surgical methods to treat suprasellar arachnoid cysts. Reviewing the literature, few series have isolated the success rate strictly on hydrocephalus of each technique. André et al. on their series of 15 patients with SAC-1 and hydrocephalus reported that 14 (93%) were treated endoscopically with ventriculocystocisternostomy initially and one (7%) had a ventriculoperitoneal shunt (André et al. 2016). The radiological follow-up showed that 12 (80%) reduced the cyst and ventricular volumes with a good flow through the stoma after primary endoscopy and 2 (13%) after a second one. One patient (7%) had the persistence of dilatation despite a visible CSF flow. Two SAC-1 were diagnosed prenatally. One patient had a prenatal treatment with fetoscopy with the need of a second endoscopy in postnatal period because of persistence of hydrocephalus. The clinical results were more modest because eight patients (53%) presented abnormalities, five patients (33%) with persistence of endocrine disorders, two (13%) with developmental



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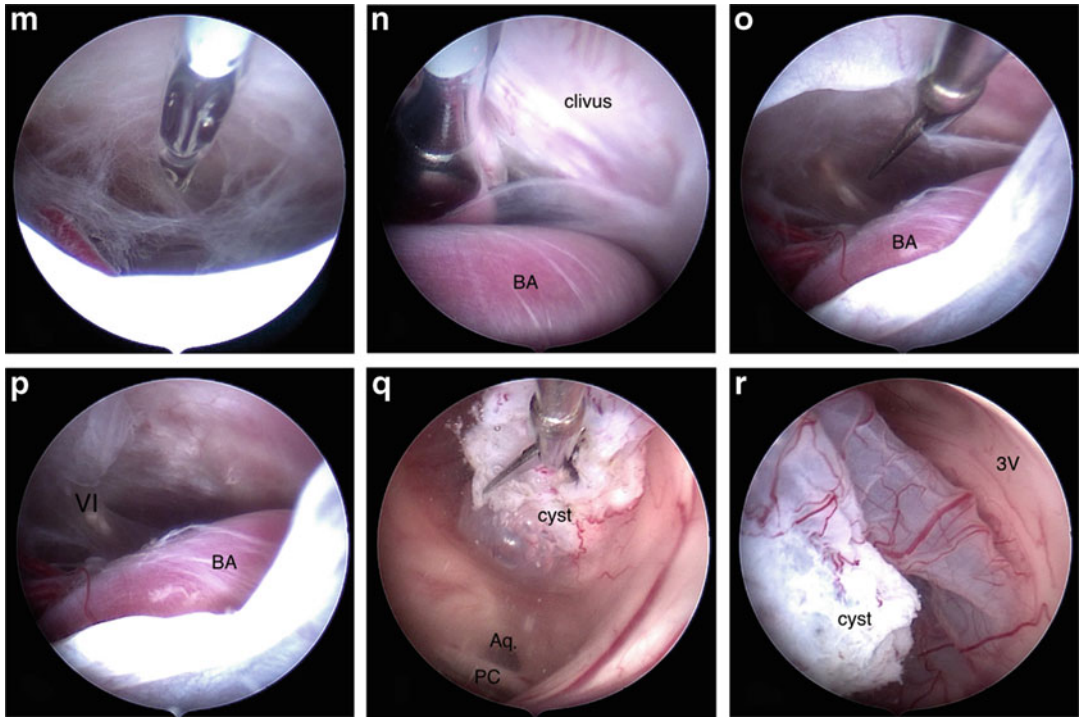


Fig. 9 **Case 1**. (**a**–**c**) T1-weighted MRI of a 3 months old girl with a suprasellar arachnoid cyst type **SAC-1**. (**d**–**f**) Increasing cyst 18 months later on a T2-weighted MRI. Note the extension through the right foramen of Monro and the ventriculomegaly. (**g**–**r**) Endoscopic procedure from the right lateral ventricle. (**g**) Upper wall of the cyst inside the right lateral ventricle. (**h**, **i**) Upper wall fenestration. (**j**–**l**) Fenestration of the bottom wall of the cyst. (**m**–**p**) Section and laceration of Liliequist membranes. (**q**) Coagulation and retraction of the cyst, enlargement of the superior stoma. (**r**) Note the penetration of the cyst through the floor of the third ventricle

delay, three (20%) with minor neuropsychological disturbances, two (13%) with oculomotor palsy, and one (7%) with epilepsy.

In 2011, Gangemi et al. have performed a comprehensive review of 61 reports in the literature comparing the clinicoradiological outcome of the 3 surgical procedures in the treatment of 169 suprasellar arachnoid cysts (Gangemi et al. 2011) (Table 6).

Temporal Cyst

Pathophysiology of Hydrocephalus

Hydrocephalus is rarely associated to temporal arachnoid cysts but can exceptionally be found with huge hemispheric cysts (type Galassi III), with a shift of the midline, occluding CSF pathways.

Subtypes

Galassi et al. classified in 1982 the temporal arachnoid cysts based on CT cisternography, according to their volume and characteristics in terms of communication with the arachnoid space, in three categories (Galassi et al. 1982) (Table 7). No significant correlation between this classification and the symptomatology is clearly apparent in the literature.

More recent studies tried to make another classification of Sylvian arachnoid cysts based mainly on the communicating character of the cyst with the subarachnoid spaces and cisterns rather than cyst size, using CT cisternography or cine MRI. But no one really measured the association of his technique with the evolution of the cyst over time and clinical complains.

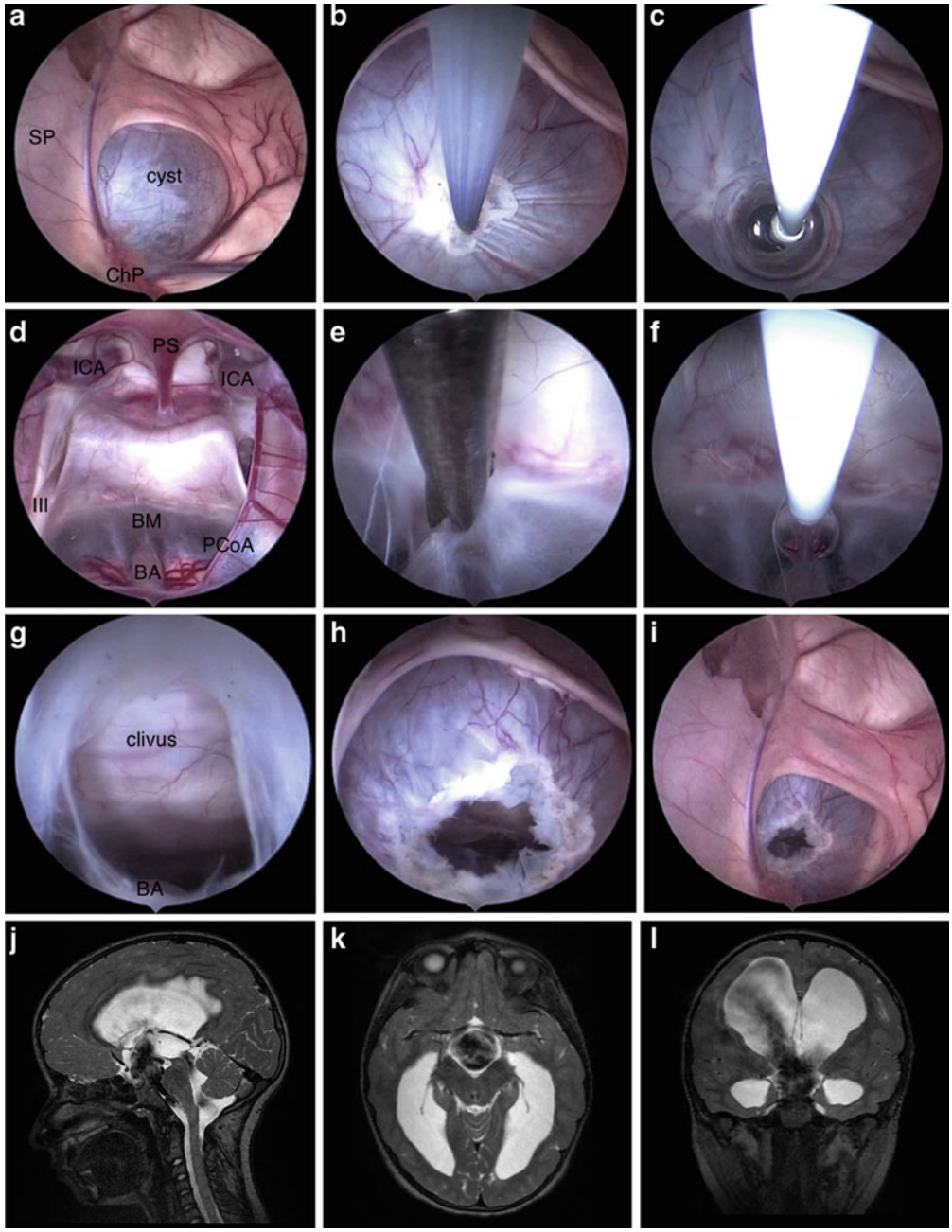


Fig. 10 **Case 2**. (**a**–**i**) Endoscopic procedure by the right lateral ventricle, for a suprasellar arachnoid cyst type **SAC-1**, with severe hydrocephalus. (**a**) Upper wall of the cyst through the right foramen of Monro. Note the spontaneous laceration of the anterior part of the septum pellucidum due to the emergence of mono- and/or biventricular hydrocephalus. (**b**, **c**) Fenestration of the upper wall of the cyst. (**d**) Overview of the interior of the cyst and the skull base anatomy. (**e**–**g**) Fenestration of the bottom wall of the cyst. (**h**, **i**) View of the stoma and decompression of the column of the fornix. (**j**–**l**) Postoperative T2-weighted MRI showing the flow artifacts. (**j**) Sagittal section showing flow artifact into the cyst and through the cysto-cisternal stoma. (**k**) Axial section showing intracystic flow artifact. (**l**) Coronal section showing cystoventricular flow artifact

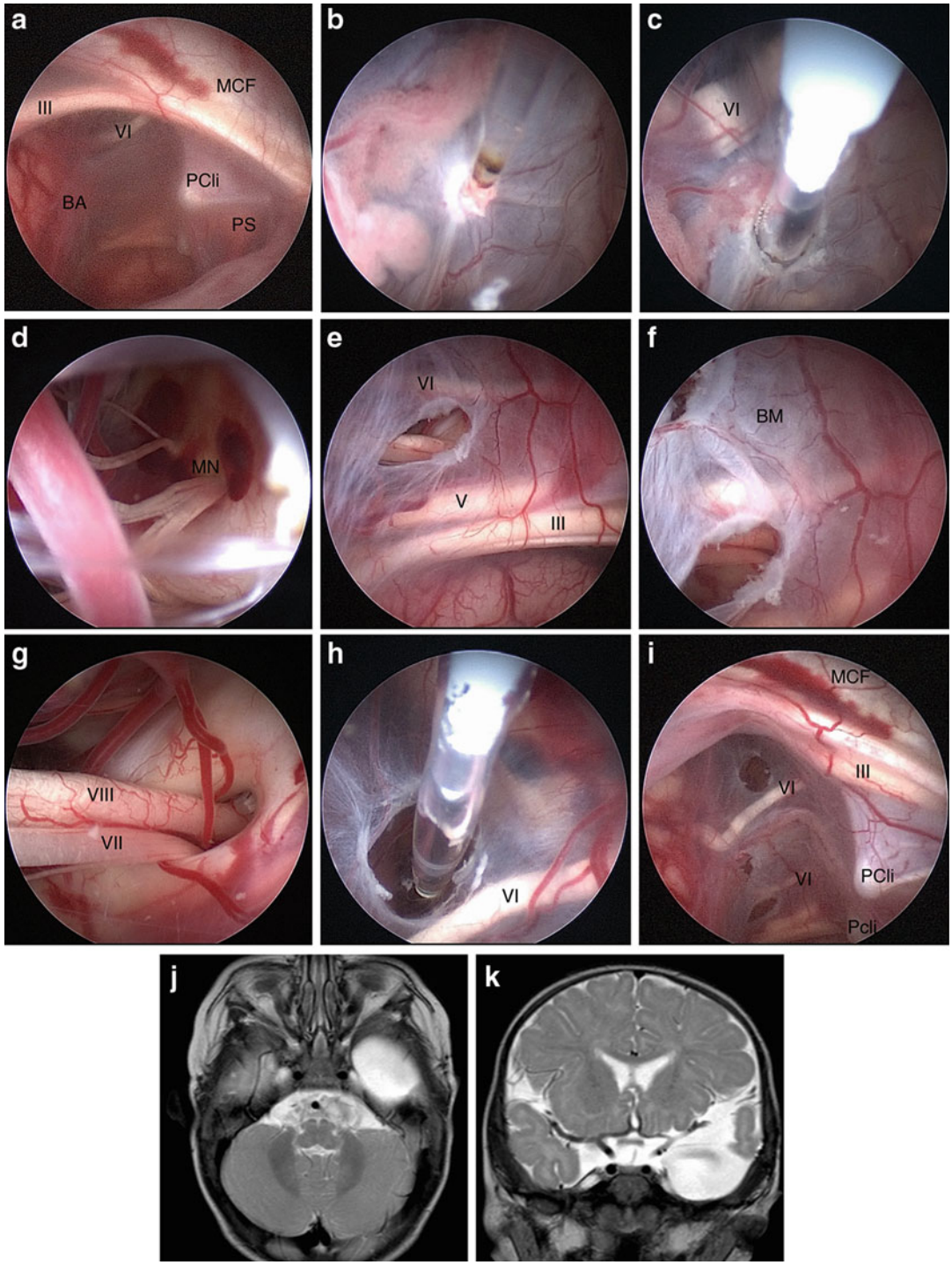


Fig. 11 **Case 3**. (**a**–**i**) Endoscopic procedure for a suprasellar arachnoid cyst type **SAC-3**. Preoperative images on Fig. 7. The patient is supine and head turned to the right. With the help of neuronavigation, the optimal trajectory is calculated with a left temporal entry point. The view of the endoscopic operative field is reverse respected to an anatomical view (up is down and vice versa; posterior is on the left and anterior on the right). (**a**) Overview of the prepontine and interpeduncular dilated spaces, with a left-right cranio-caudal angle, seeing the contro-lateral

Table 6 Success rate between the three surgical options for suprasellar arachnoid cysts treatment without isolating hydrocephalus (Gangemi et al. 2011)

Technique	Good outcome (%)
Microsurgery	79
***Cyst* shunting**	66
Endoscopic fenestration	89.7
VC	86.3
VCC	92.5

Good outcome = complete or partial clinical remission, *VC* ventriculocisternostomy, *VCC* ventriculocystocisternostomy

Radiology

See Figs. 12, 13, 14, 15, and 16

Treatment/Surgical Techniques

The best surgical management of Sylvian arachnoid cysts is the subject of controversies for a long while, particularly if we add hydrocephalus in the balance and also because of the rarity of association.

In most of the articles found in the literature, craniotomy with cyst fenestration has been compared to shunting procedures, whether with programmable valves or not. Some authors proposed the combination of the two procedures. Tamburrini et al. performed a survey on the management of Sylvian arachnoid cysts in 45 pediatric neurosurgical centers (Tamburrini et al. 2008) and found a large disparity of practices.

Microsurgical Procedure

The successful rate of open surgery in some study is low with a failure rate of 24–67% (Ciricillo et al. 1991; Pulido-Rivas et al. 2005). According to Ciricillo et al. on 40 patients, 15 were treated initially by fenestration (Ciricillo et al. 1991). Sixty-three percent showed no clinicoradiological improvement and have undergone cystoperitoneal shunting in 80%, ventriculoperitoneal shunting (10%), or revision of a ventriculoperitoneal shunt placed because of hydrocephalus before cyst fenestration (10%). The two patients with preexisting ventriculoperitoneal shunts required no further procedures. On all patients treated initially by cyst fenestration, only 20% remained shunt-independent during the follow-up. Other authors found that shunt malfunction rates could be up to 40% (Raffel and McComb 1988), with the risk of shunt dependency like the slit ventricle syndrome (Aoki et al. 1990; Kim et al. 2002; Sunami et al. 2002). On more recent series, no significant differences in perioperative morbidity and mortality have been reported in the comparison of fenestration and shunt (Kang et al. 2000; Lena et al. 1996; Pulido-Rivas et al. 2005; Raffel and McComb 1988; Levy et al. 2003). The risk of subdural effusion after microsurgical or endoscopic procedures varies between 2% and 40% (Di Rocco et al. 2010; Cinalli et al. 2007; Tamburrini et al. 2003).

The defenders of open surgery approach also advocate that good outcomes (80%) can be obtained using a keyhole microsurgical approach, having few complications (Levy et al. 2003; Pulido-Rivas et al. 2005).

Nowadays, microsurgical procedures are preferably indicated when the cyst does not open enough the Sylvian fissure and does not reach the surface.

Standard Microsurgical Procedure (Figs. 17 and 18)

The patient is positioned supine; the head is turned versus the contralateral side of the temporal cyst.

Fig. 11 (continued) ponto-cerebellar angle. (**b**–**i**) Completion of three fenestrations through the bottom wall of the cyst, between cranial nerves. (**d**) View of the right vertebral artery and mixed nerves. (**g**) View of the right facial and vestibulocochlear nerves. (**i**) Overview of the three stomas at the end of the procedure. (**j**, **k**) T2-weighted axial and coronal sections of postoperative MRI showing artifact flows forward the midbrain. Note that the basilar artery is not pushed anymore against the brainstem on the axial section. The aim of the intervention was mostly to restore the communications with the basal cisterns, to prevent the expansion of the cyst, and to promote its relaxation

Table 7 Classification of temporal arachnoid cysts (Galassi et al. 1982) and percentage of cases on 145 temporal arachnoid cysts (Al-Holou et al. 2010)

Subtypes (% of cases)	Radiological presentation
Galassi I (68%)	Small fusiform cyst limited to the most anterior portion of the middle cranial fossa, below the sphenoid ridge
	The temporal pole is com-subsequently compressed. Absence of mass effect. Cisternography shows free communication with subarachnoid spaces (Figs. 12 and 13)
Galassi II (15%)	Cyst of medium size and triangular morphology or square with straight inner margin. Occupies the anterior and middle portions of the temporal fossa and superiorly extends along the Sylvian fissure, which wildly exposes the insula
	The temporal lobe is displaced (or hypoplastic). Cisternography shows slow communication with subarachnoid spaces (Fig. 14)
Galassi III (17%)	Huge ovoid cyst occupying the temporal fossa almost completely and extends to a wide area of the cerebral hemispheres opening cap of the Sylvian fissure
	The temporal lobe is severely compressed (and atrophic), and both the frontal and parietal lobes are widely tableted (Fig. 15)
	The great mass effect can cause a large ventricular distortion and shift of the midline. Cisternography shows a little communication with subarachnoid spaces
	It produces cranial deformities and macrocephaly in children
	When temporal cysts become fronto-temporo-parietal, they are called Hemispheric arachnoid cysts, and are part of subtype Galassi III (Fig. 16)

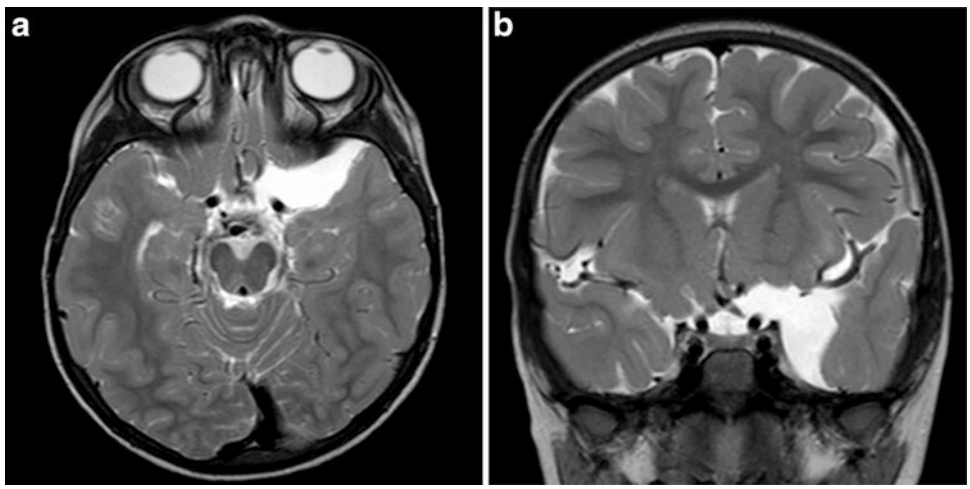


Fig. 12 T2-weighted MRI showing a left temporal cyst **Galassi I** (Galassi et al. 1982). (**a**) Axial section. (**b**) Coronal section

The incision is made according to the habits of the surgeon. The size of the craniotomy is adapted to the size and location of the cyst. The dura is then opened, revealing the external wall of the cyst, which is opened. Attention is paid to decompress slowly the cyst avoiding sudden neurovascular re-expansion with risks of bleeding, brain contusion, nerve trauma, etc. A gentle excision of the cyst is performed leading to the wall in contact with basal cisterns, which is often very thick. With

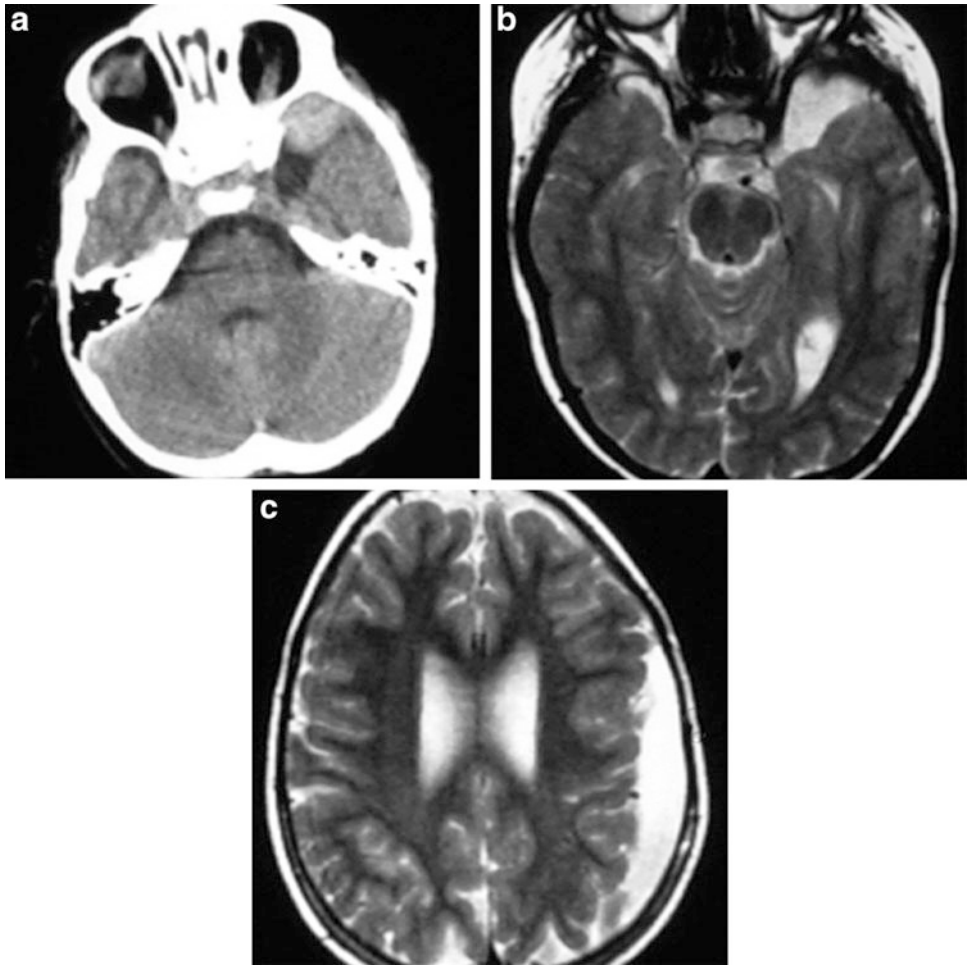


Fig. 13 Left temporal arachnoid cyst **Galassi I** reveled after a head trauma. (**a**) Axial section of computed tomodensitometry showing a temporo-polar intracystic hematoma. (**b**) Axial section of T2-weighted MRI confirming the arachnoid cyst. (**c**) Axial section of T2-weighted MRI showing a subdural hygroma probably due to a traumatic rupture of the cyst

microsurgical instruments, the largest possible openings are performed in the basal cisterns (inter optics, opto-carotid, retro-carotid windows, etc.). The closure of the dura mater is watertight. Figures 17 and 18 shows the microsurgical procedure for treating right and left Temporo-Sylvian arachnoid cysts.

Shunting

The shunt placement in case of temporal arachnoid cyst is rarely indicated. The placement of the intracranial catheter should be maturely meditated, providing for its future position when the brain will take back its place. Indeed, the catheter may cause neurovascular injuries.

Supporters of shunt procedures claim that craniotomy is very aggressive mainly for young children, with major complications such as meningitis, hemiparesis, oculomotor palsy, subdural hematoma/hygroma, seizures, and death (Arai et al. 1996; Ciricillo et al. 1991). Arai et al. treated 77 patients by shunting: one (1%) had a material infection, and eight (10%) had a shunt malfunction with the need of revision (Arai et al. 1996). Half of these malfunctions were because of a shunt dependency syndrome, but none for hydrocephalus.

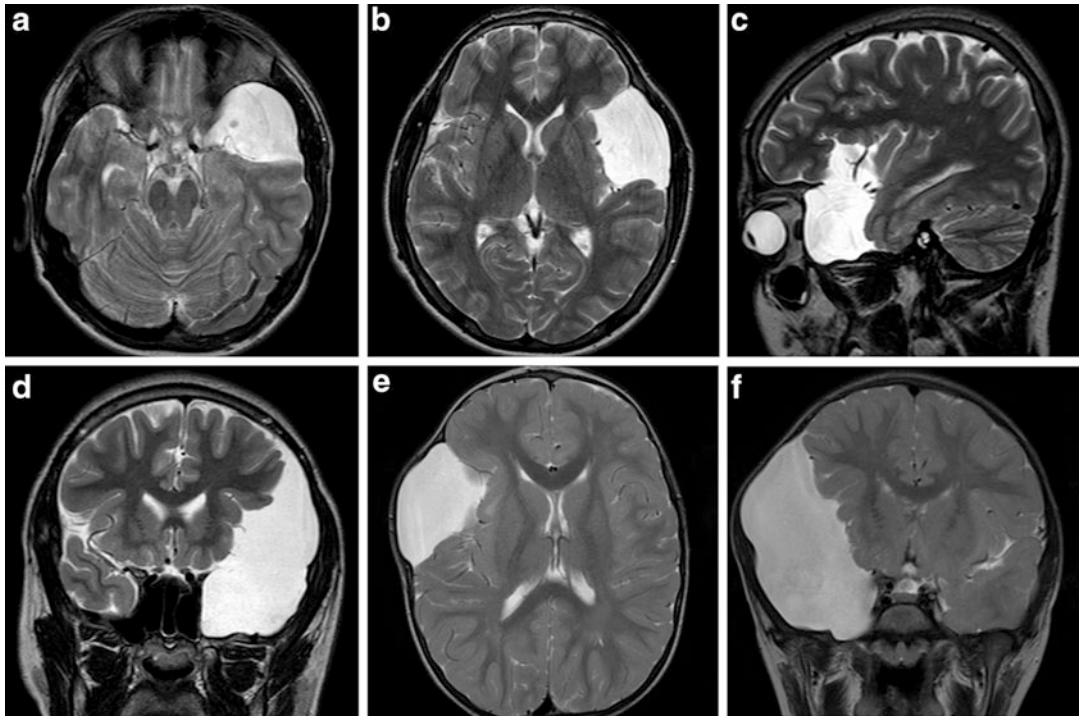


Fig. 14 (**a**–**d**) Left temporal arachnoid cyst **Galassi II** on a T2-weighted MRI. (**a**, **b**) Axial sections. (**c**) Sagittal section. (**d**) Coronal section. (**e**, **f**) T2-weighted MRI showing a right temporal arachnoid cyst **Galassi II** on (**e**) axial and (**f**) sagittal sections. Note the chronic bone lysis and distortion

Standard Procedure for Cystoperitoneal Shunting (Figs. 19 and 20)

The use of the neuronavigation system is highly recommended. We will not develop the implantation techniques of the valve itself and the distal catheter, as it is the same procedure of the implantation of a ventriculoperitoneal shunt. In the literature, the use of a programmable valve is advisable as it can perform a controlled slowly re-expansion in the time. It is preferable to initially leave a high opening pressure.

The patient is positioned supine with the head slightly extended and turned to the contralateral side. A burr hole is made at the top of the normal hypothetical Sylvian fissure. The dura mater is incised together with the external wall of the cyst with dimensions only to let the catheter inside and not more, in order to limit the formation of hygroma. Attention is paid not to empty the cyst, so the catheter must be inserted quickly. The neuronavigation is used to aim the direction just under the ridge of the great wing of the sphenoid, with a tangential trajectory to the bone. This position also helps to avoid subdural detachment. Then the catheter is connected to the rest of the system. Figure 19 shows the recommended position of the intracranial catheter for shunting a tempora-Sylvian arachnoid cyst, while Fig. 20 shows the consequence of a misplaced catheter.

Endoscopic Procedure

In the era of endoscopic procedures, its introduction has extended the debate. Fully endoscopic procedure should be distinguished from endoscope-assisted procedure.

One thing which should be considered before performing neuroendoscopy is that often the Sylvian cyst is large and the intracystic anatomy can totally confuse the surgeon. Neuronavigation applied to the endoscope, avoiding the risk of the brain shift with CSF loss, can be particularly useful to indicate the right direction to follow. Literature is controversial about success rate of endoscopy (Couvreur et al. 2015; Fernández 2013;

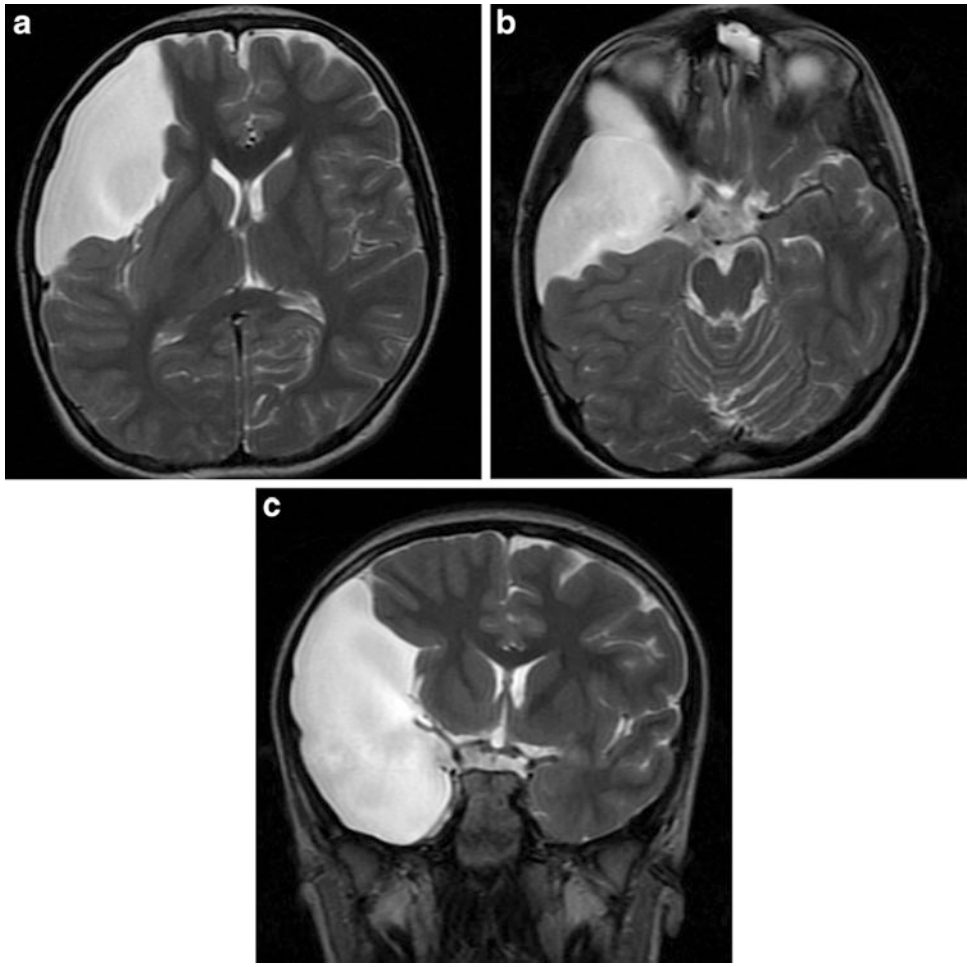


Fig. 15 Right temporal arachnoid cyst **Galassi III** on a T2-weighted MRI. (**a**, **b**) Axial sections. (**c**) Coronal section

Cinalli et al. 2007; Elhammady et al. 2007; Huang et al. 2007; Karabatsou et al. 2007; Nowosławska et al. 2006; Paladino et al. 1998; Pradilla and Jallo 2007; Schroeder et al. 1996).

Examining the results, complications and limitations of neuroendoscopy on middle fossa arachnoid cysts, Di Rocco said that the main limitation of the endoscopic approach is the anatomical proximity of the cyst to the basal cisterns and critical structures behind the wall of the cyst (Di Rocco 2010). He advocated the use of a small-diameter endoscope to avoid major external membrane opening with its consequences. Cinalli et al. reported that among the five patients with Sylvian cysts treated by endoscopic procedure, 50% developed a subdural hygroma and 20% a CSF leak. Eighty percent of them have required the secondary implantation of a shunt (Cinalli et al. 2007). Elhammady et al. recommend to realize, each time it is possible, a transcortical access to keep some impermeability on the approach, but this solution is, of course, highly questionable (Elhammady et al. 2007).

Indeed, using mono- or bipolar coagulation, inflation of the balloon can be dangerous in the deep wall of the cysts because of the presence of the internal carotid artery, the posterior communicating artery, and the oculomotor nerve (Johnson et al. 2011). Figures 21 and 22 shows the endoscopic procedures of a left Galassi II and a right hemispheric arachnoid cyst. Figure 23 shows the procedure for a left temporal arachnoid cyst of the choroid fissure, which does not belong to Galassi classification.

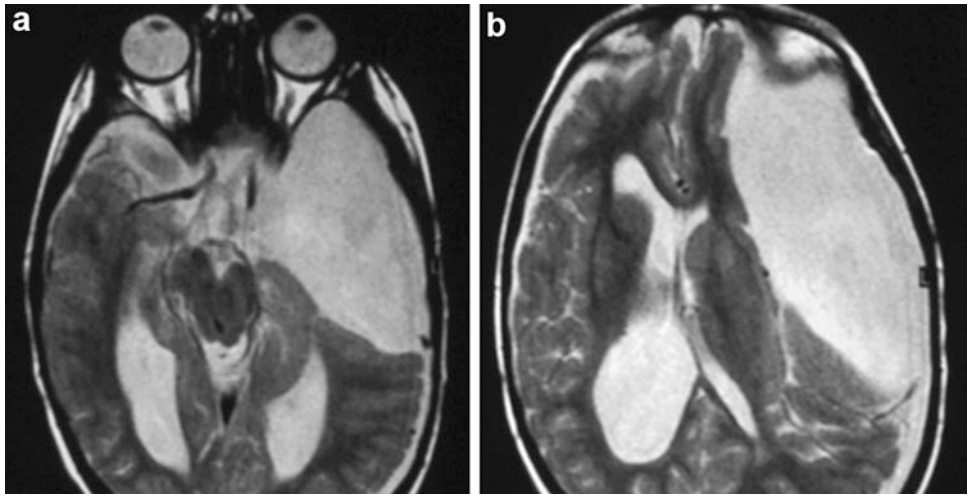


Fig. 16 (**a**, **b**) Left hemispheric arachnoid cyst on axial T2-weighted MRI showing an important brain compression, a shift of the midline, and an associated hydrocephalus

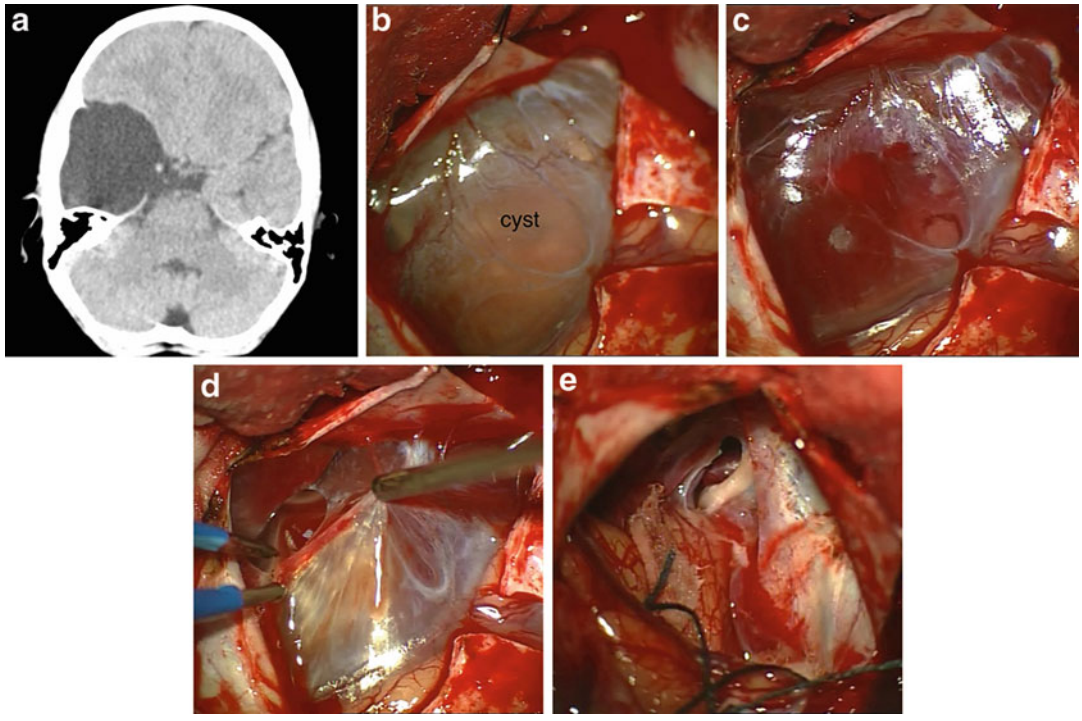


Fig. 17 **Case 1**. (**a**) Right temporo-Sylvian arachnoid cyst on axial section of computed tomodensitometry. (**b**–**e**) Microsurgical procedure. (**b**) Arachnoid cyst with its intact external wall. (**c**, **d**) Opening and excision of the cyst. (**e**) Fenestration of the inner membrane into basal cisterns

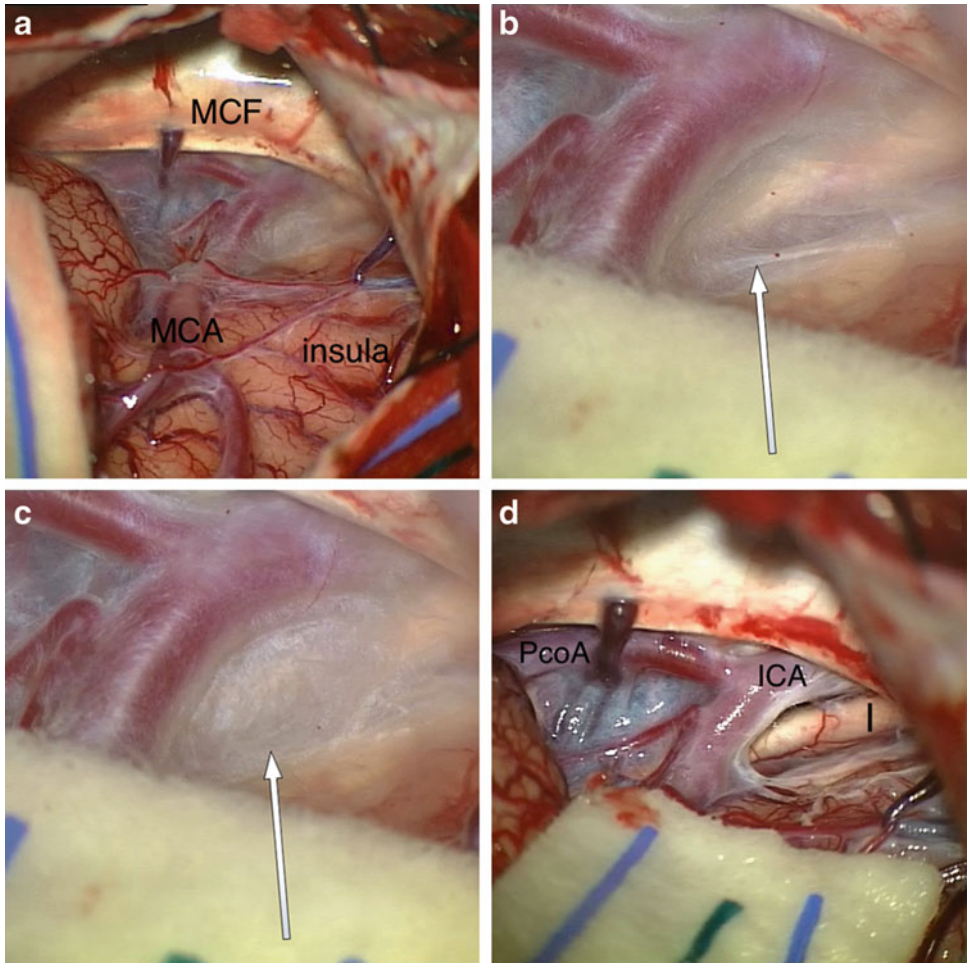


Fig. 18 **Case 2**. (**a**–**d**) Microsurgical procedure on a left temporo-sylvian arachnoid cyst. With a view on the inner wall of the cyst, the basal cisterns, the left anterior axe of the circle of Willis, and the left optic nerve. Note the enlargement of the Sylvian fissure, showing skull base of the middle cranial fossa and the course of the middle cerebral artery on the insula. (**b**, **c**) We can see the pulsatility, the thickness, and the trapped aspect of the inner wall. (**d**) Final aspect after wide opening of the optical-carotid and inter-chiasmatic windows

Endoscopic Standard Procedure (Figs. 21, 22)

The patient is positioned supine; the head is turned versus the contralateral side of the temporal cyst. The use of the neuronavigation system is highly recommended. A cutaneous muscular linear incision is performed. A burr hole is made according to the preoperative planning, depending on the characteristic of the cyst and above all the position of its inner wall and the basal cisterns. The hole will be slightly enlarged by considering that basal cisterns are not in the axis of the craniotomy. The dura is then opened, revealing the external wall of the cyst, which is secondarily opened. Attention is paid to not empty and decompress the cyst avoiding sudden neurovascular re-expansion with risks of bleeding, brain contusion, nerve trauma, etc. and keeping in mind that neuroendoscopy is a procedure to be performed in a fluid environment. The rigid endoscope is quickly introduced into the cyst, and anatomical landmarks are identified with the help of neuronavigation. Once the inner wall and basal cisterns are identified with their neurovascular structures, the largest possible fenestrations are performed, using endoscopic instruments (inter optics, opto-carotid, retro-carotid windows,

etc.). The way to achieve fenestrations is chosen according to the preoperative planning, having considered the anatomical structures lying below the membrane. As the walls of arachnoid cysts are often thick, we usually perforate them with the laser, ideally as a grid. The stoma thus produced is expanded using the double-balloon probe. Usually it is necessary to enlarge the stoma using scissors between two holes because of the thickness of the membranes. The endoscope is then removed without emptying the cyst. The closure of the dura mater is watertight.

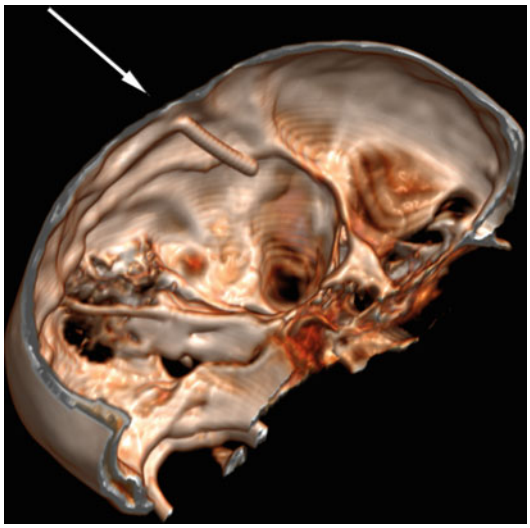


Fig. 19 3D bone reconstruction of a computed tomodensitometry. Recommended position of the intracranial catheter for a left hemispheric arachnoid cyst

Discussion on Treatment

In 2011, Gangemi et al. have performed a comprehensive review of 61 reports in the literature comparing the clinicoradiological outcome of the 3 surgical procedures in the treatment of 304 middle fossa arachnoid cysts (Gangemi et al. 2011) (Table 8).

Hydrocephalus in Sylvian arachnoid cysts has been studied in a dedicated paper (Levy et al. 2004). In the series of 40 middle fossa cysts, the authors found 15% of associated hydrocephalus. All patients required CSF shunting following the cyst fenestration because of persistent progressive hydrocephalus after cyst marsupialization. They reported almost the same need with the patients presenting macrocephaly. These papers mix together all kinds of cysts and are not specific for Sylvian cysts.

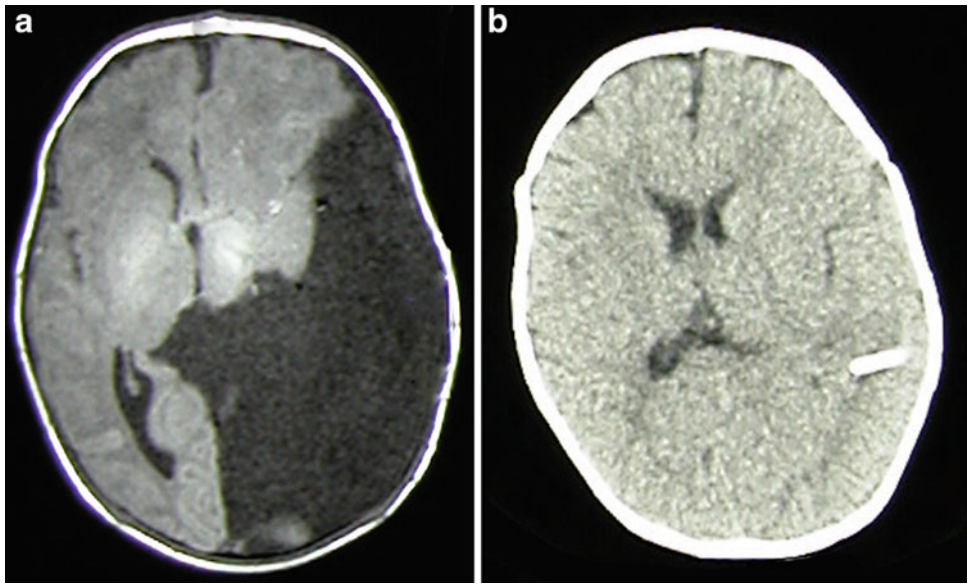


Fig. 20 (**a**) T1-weighted MRI showing a huge left hemispheric arachnoid cyst with mass effect and shift of the midline. (**b**) Computed tomodensitometry performed 1 year after implantation of a cystoperitoneal shunt. The catheter, correctly placed at surgery, finally ended up embedded in the brain parenchyma due to the unexpected complete disappearance of the cyst

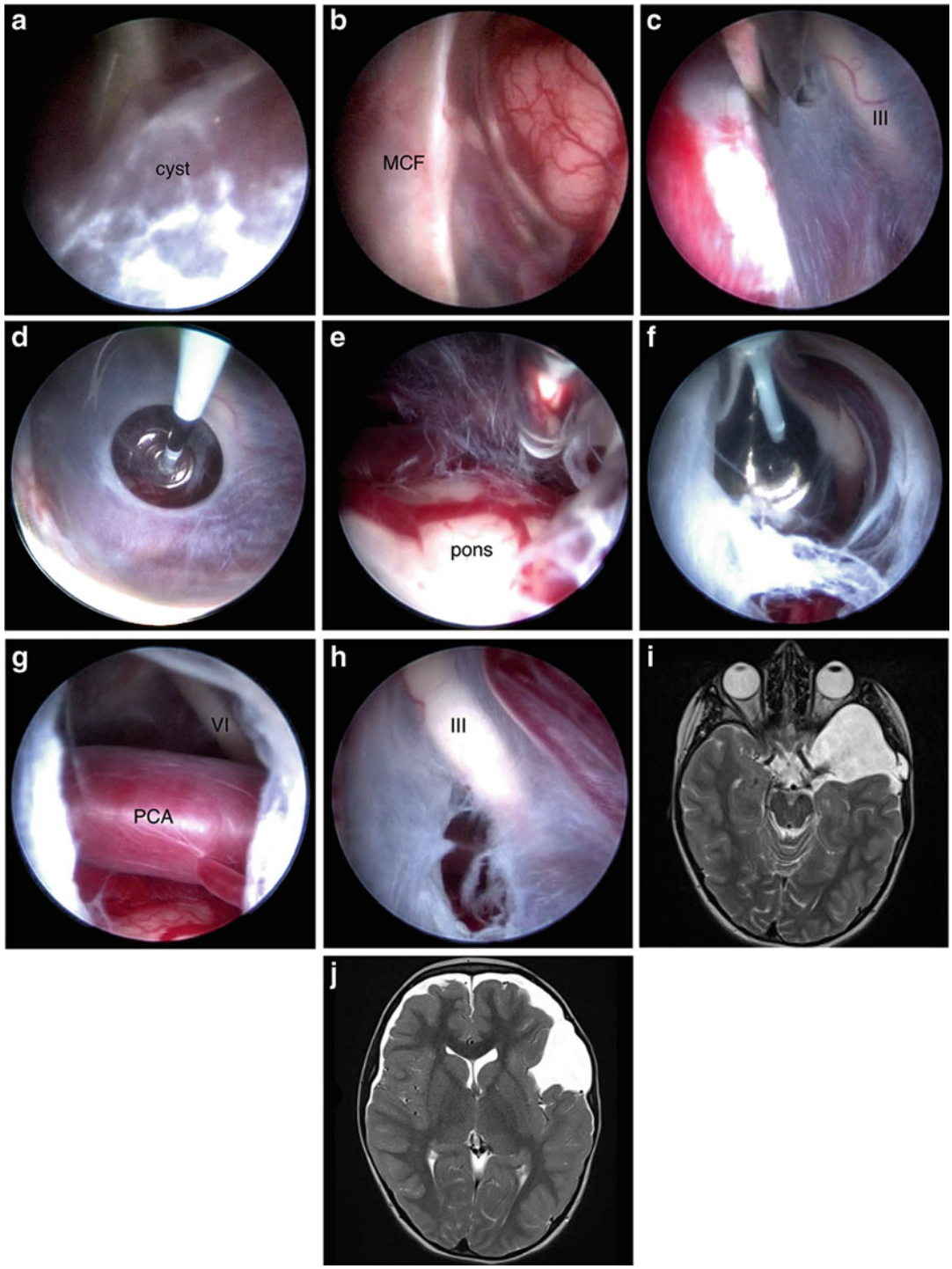


Fig. 21 **Case 1**. (**a**–**h**) Endoscopic procedure for a left temporo-Sylvian **Galassi II** arachnoid cyst. (**a**) Vision from the subdural space with opening of the external wall of the cyst. (**b**–**h**) Free margin of the tentorium on the left with fenestration of the inner wall of the cyst and the basal cisterns. (**g**) View of the prepontine cistern. (**i**, **j**) T2-weighted axial sections of postoperative MRI showing (**i**) artifact flows through the inner wall. (**j**) Classical post-operative bilateral subdural hygromas

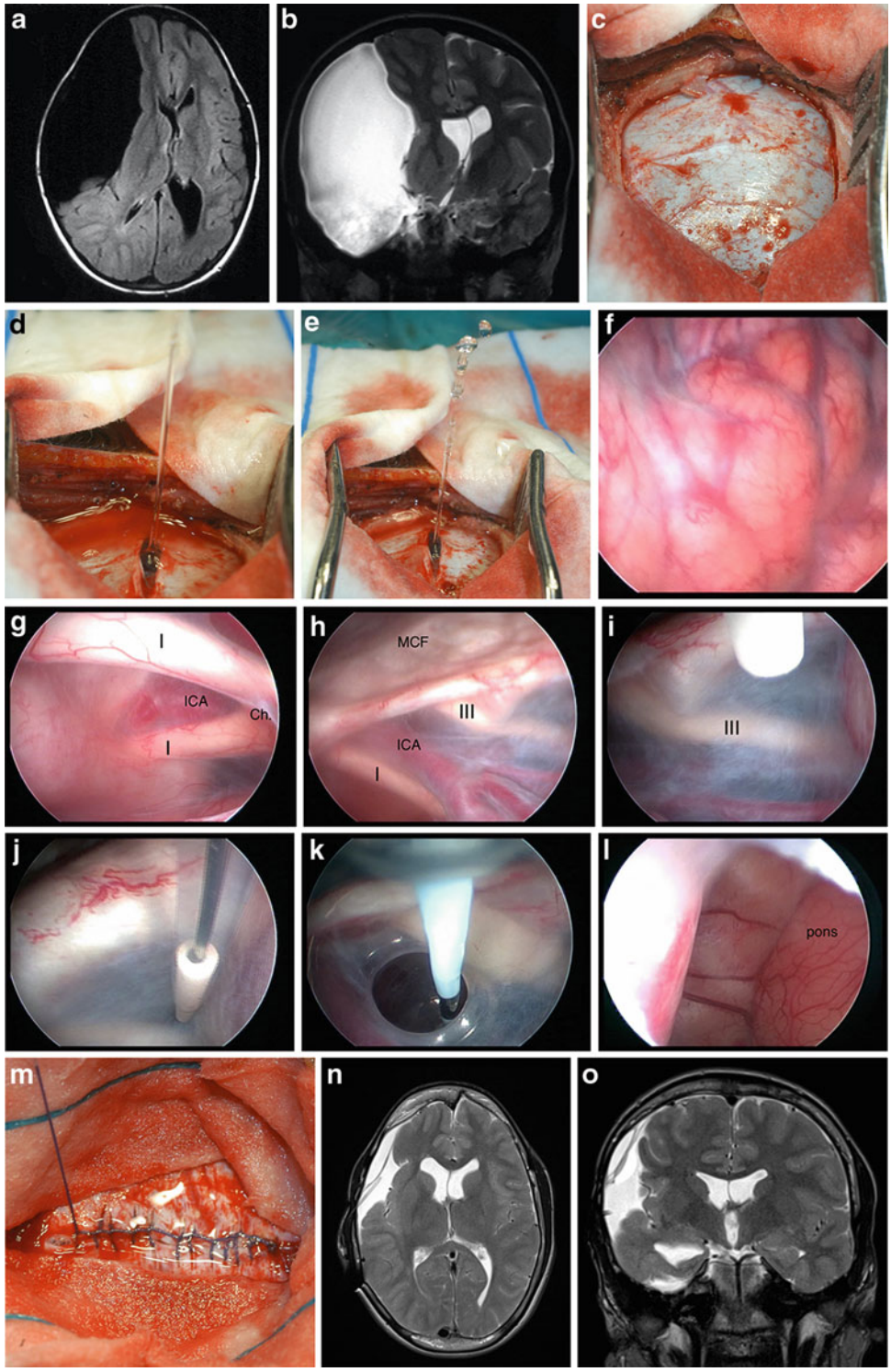


Fig. 22 **Case 2**. Endoscopic procedure for a right hemispheric arachnoid cyst. (**a**) Axial section of T1-weighted MRI and (**b**) coronal section of T2-weighted MRI showing a huge cyst with mass effect on the parenchyma and shift of

Table 8 Success rate between the three surgical options for temporal arachnoid cysts treatment without isolating hydrocephalus (Gangemi et al. 2011)

Technique	Good outcome
Microsurgery	88%
	(7.5% of secondary shunt placement)
***Cyst* shunting**	96.8%
Endoscopic fenestration	70%
	(24% of secondary shunt placement)

Good outcome = complete or partial clinical remission

Performing microsurgical fenestration on temporo-Sylvian arachnoid cysts, Okano and Ogiwara present, on a series of 28 patients, a good clinical outcome of 90.5% and a good radiological outcome of 100% with regression or disappearance of the cyst (Okano and Ogiwara 2016). However, one patient younger than 2 years needed the placement of a shunt, because of preoperative hydrocephalus. So, on the two patients (7%) with preoperative hydrocephalus, 50% required shunt placement. Postoperatively, 82.1% of the patients had a subdural hygroma, one of them symptomatic. At longer term, 83% of hygroma disappeared and 13% decreased. One case (4%) needed a subduroperitoneal shunt. In the following literature, the clinical evidence of supremacy of endoscopy over microsurgery is still lacking.

According to Martínez-Lage, very often there is the need to perform both procedures (fenestration by open craniotomy or by endoscopy, followed by shunt procedure) in case of hydrocephalus, particularly in young children (Martínez-Lage et al. 2011). They prefer to use programmable valves instead of single-pressure ones, to allow more gradual reduction of both cyst and ventricle size. They also advocate the utility of a ventriculocystoperitoneal shunt, preventing differences in pressure between the diverse compartments (Martínez-Lage et al. 1999). Silav et al. and Kimiwada et al. advocate for the combine procedure including fenestration followed by shunt placement (Silav et al. 2015; Kimiwada et al. 2015).

Slit Cyst Syndrome (Figs. 24 and 25)

As showed above, shunt placement in the treatment of middle fossa arachnoid cyst is often considered as a minor procedure with low risks and few complications. Slit ventricle syndrome, also called shunt dependency syndrome, is a serious complication quite rare but difficult to diagnose of shunting an arachnoid cyst. Slit ventricle syndrome was initially defined by Epstein et al. during the 1970s as an important reduction in size of the lateral ventricles associated to clinical symptoms of intracranial hypertension (Epstein et al. 1974) (Figs. 24 and 25). He reported the origin to an excessive CSF drainage after placement of a ventricular shunt in hydrocephalus, whose ventricles were not able to re-expand even after intracranial hypertension. Several articles describe the slit ventricle syndrome but less differ the syndrome due to a cystoperitoneal shunt (Aoki et al. 1990; Fang et al. 2012; Sunami et al. 2002). This latter entity began to be described in the early 1990s. But other authors prefer the term of shunt dependency syndrome (Kim et al. 2002; Laviv and Michowitz 2010; Li et al. 2014). So, we focus on these syndromes associated to cyst shunting, keeping some references to the original one.

Ahn et al. advocated an anomaly of CSF absorption mechanism (Ahn et al. 1997). Someway, according to them, the valve replaces the physiologic pathways of reabsorption leading to this phenomenon of shunt dependency. Reviewing the literature, Li et al. think that the drainage of the

Fig. 22 (continued) the midline. (**c**) Burr hole. (**d**, **e**) Opening of the dura and the external wall of the cyst, showing the pressure of liquid inside the cyst. (**f**–**l**) Endoscopic view. (**f**) View of the insula from inside the cyst. (**g**) View of the inter-chiasmatic space. (**h**) View of posterolateral internal carotid structures. (**i**, **j**) Fenestration laterally to the oculomotor nerve. (**k**) Fenestration between the internal carotid artery and the oculomotor nerve. (**l**) View of the prepontine cistern. (**m**) Closure of the dura. (**n**, **o**) T2-weighted MRI showing the secondary implantation of a cystoperitoneal shunt because of insufficient clinical-radiological postoperative response

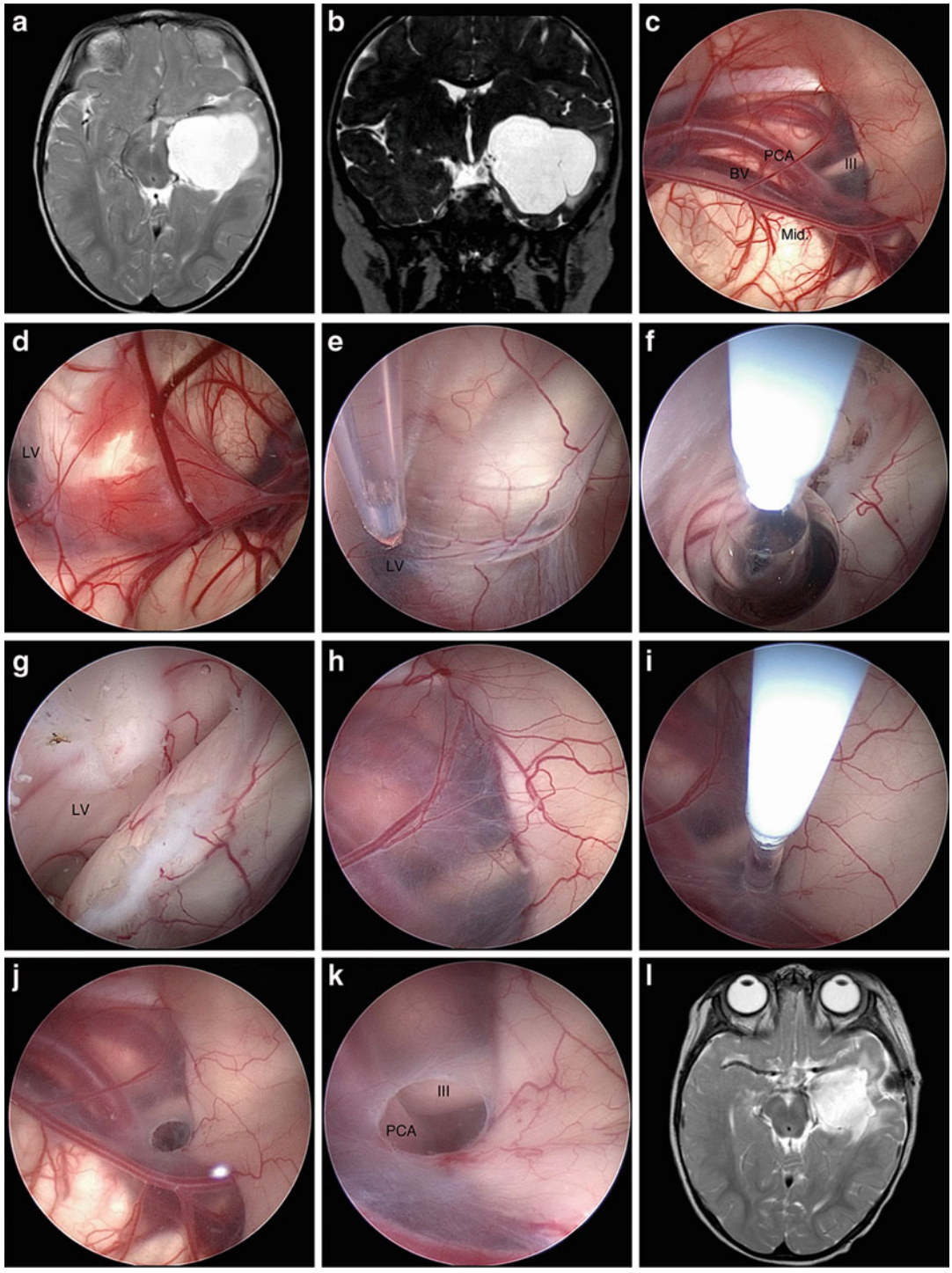


Fig. 23 (continued)

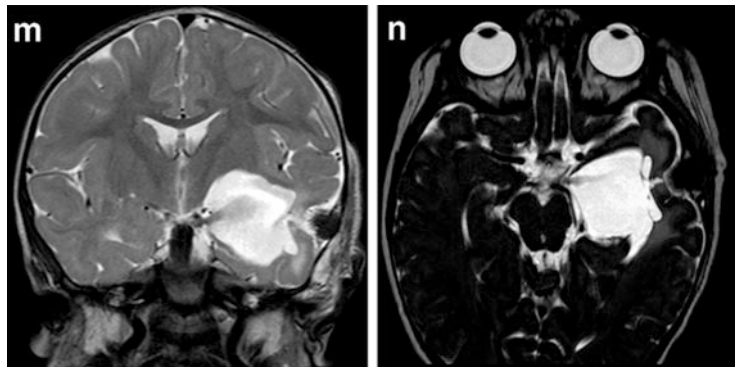


Fig. 23 **Case 3**. (**a**, **b**) MRI showing a left temporal arachnoid cyst of the choroid fissure. Note the mass effect on the temporal lobe and on the midbrain by dilatation of the ambient cistern. This kind of cyst is rare and does not belong to any other type (not precisely to lateral intraventricular or quadrigeminal type 3). (**a**) Axial T2-weighted section. (**b**) Coronal CISS-weighted section. (**c**–**k**) Endoscopic procedure. (**c**) Superolateral view of the midbrain and diencephalon from inside the cyst, which took the place of the ambient cistern. (**d**–**g**) Cystoventricular fenestration of the temporal horn of the left lateral ventricle. (**h**–**k**) Fenestration of the prepontine cistern above the oculomotor nerve. (**l**–**n**) Postoperative MRI showing brain decompression and cysto-cisternal artifacts of flows. (**l**) T2-weighted axial section. (**m**) T2-weighted coronal section. (**n**) CISS-weighted axial section

cyst can lead, sometimes, to its collapse, and then the adhesions created inside the cyst obstruct the communication between the cyst and subarachnoid spaces (Li et al. 2014). This condition creates initially an intermittent failure of the shunt resulting in intermittent intracranial hypertension. This chronic hypertension induces a dural and suture fibrosis, which conduce to a decrease of the brain elasticity and compliance, similarly to the slit ventricle syndrome. In 2002, Del Bigio et al. performed an autopsy on a 10 years old girl for whom a slit ventricle syndrome was diagnosed (Del Bigio 2002). She had a ventriculoperitoneal shunt in infancy, following a meningitis. She died because of a severe intracranial hypertension attributed to the slit ventricle syndrome. The autopsy revealed large skull and brain, small ventricles with glial adhesions, obstruction of the cerebral aqueduct and the ventricular catheter, and a reactive astrogliosis in the periventricular white matter. Jang et al. advocated the theory of malabsorption of CSF with a direct consequence of brain edema or swelling and eventually distortion or herniation (Jang and Yoon 2013).

Clinically, both syndromes are characterized by various manifestations, such as ataxia, obnubilation, nausea, vomiting, lethargy, irritability, decreasing eyesight, and headache (Benzel et al. 1992).

ICP monitoring may confirm the diagnosis when there are no significant findings (Li et al. 2014), and various studies include ICP monitoring following chronic implantation of ICP transducers. For example, Rekate et al. made a five-step classification of slit ventricle syndrome based on ICP monitoring, for each recommendation of treatment (Rekate 1993).

Albright et al. postulate that this overdrainage, particularly in young children, may understimulate the calvaria and cause abnormal early ossification of the sutures, leading to an induced shunt craniostenosis (Albright and Tyler-Kabara 2001).

The treatment of slit cyst syndrome may differ from that of slit ventricle syndrome associated to ventricular shunting, because of its physiopathology and the original utility of the valve, aiming to derivate an arachnoid cyst. Focusing on the case of arachnoid cyst, the literature shows the possibilities of revision of the shunt catheter, placement of a high-resistance valve, addition of an anti-siphon device, change for a programmable valve, or placement of a lumboperitoneal shunt.

Fan et al. recommend to change the valve for a programmable one, avoiding low-pressure shunts in first intention in the management of arachnoid cysts (Fang et al. 2010). All symptoms of the six

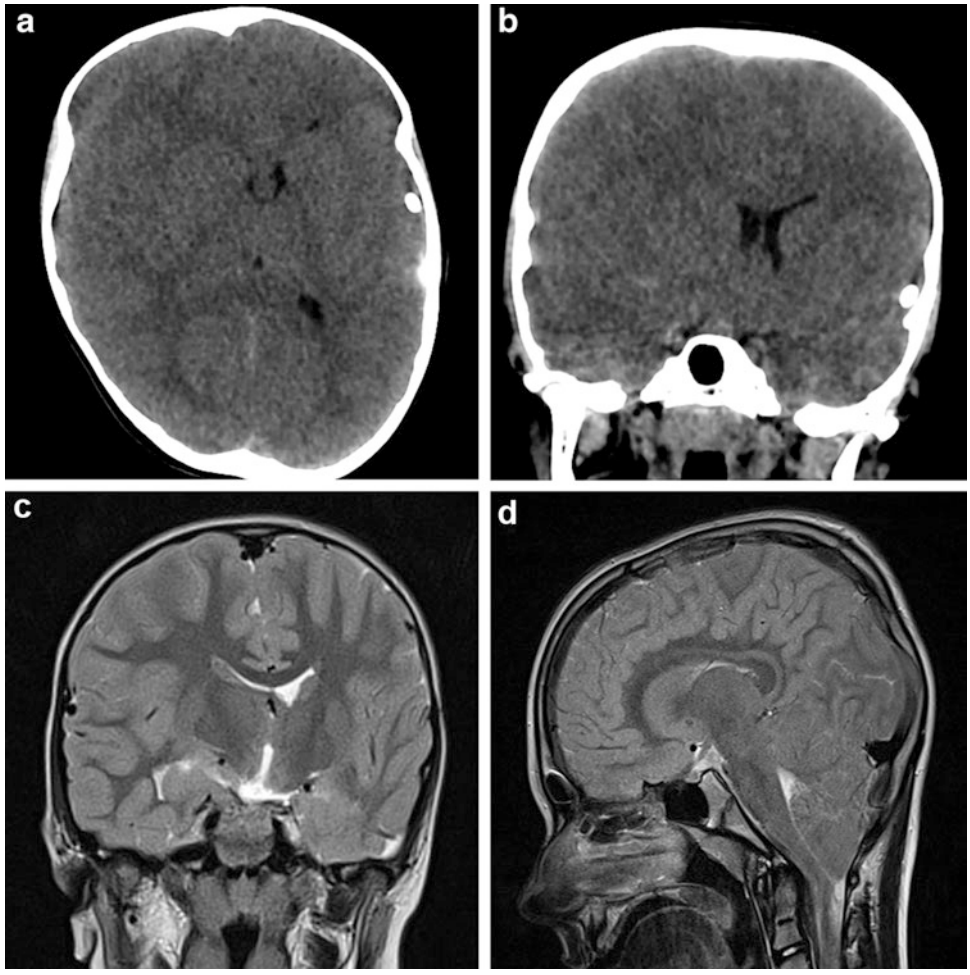


Fig. 24 **Case 1**. (**a**–**d**) Patient who previously received a cystoperitoneal shunt for a left Sylvian arachnoid cyst Galassi II. Slit cyst syndrome characterized by chronic signs of intracranial hypertension. (**a**, **b**) Axial and coronal sections of a computed tomodensitometry and (**c**, **d**) coronal and sagittal T2-weighted sections of MRI, showing the collapse of the cyst, small ventricles, and ipsilateral brain shift. (**d**) The sagittal section shows an induced chronic tonsillar herniation

patients disappeared in the 2 years follow-up. Li et al. present 13 patients with the syndrome, with a mean age of 12,3 years and 20 months of follow-up (Li et al. 2014). Twenty-three percent of them improved after shunt replacement because of non-collapsed cyst, 31% underwent ventriculoperitoneal shunt placement because of cyst collapse but normal ventricle size, and 46% had a lumboperitoneal shunt placement because of cyst and ventricle collapse. All of them presented a resolution of their symptoms. Kim et al. reported that implanting a lumboperitoneal shunt, there is a risk of hindbrain herniation (Kim et al. 2002). But in case of small ventricles, it is still possible to place a ventricular catheter helped by neuronavigation (Fang et al. 2010, 2012).

Concerning the non-cystic slit ventricle syndrome, many other treatments have been tried and benefited, such as in severe presentations of intracranial hypertension with cranial decompression and more recently the realization of cranial expansion like the treatment of craniostenosis or

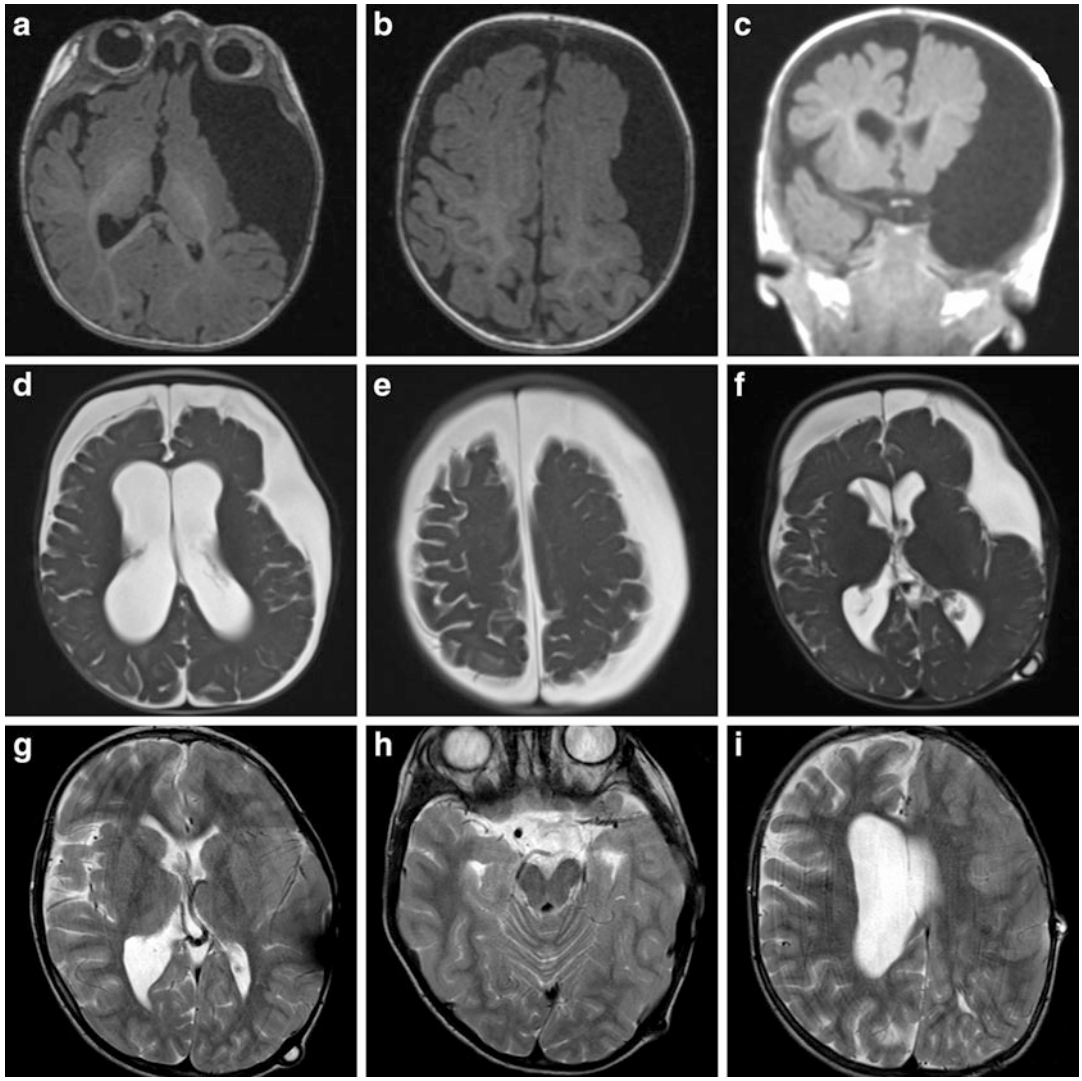


Fig. 25 **Case 2**. Complex case of a slit cyst and ventricle syndrome. (**a**–**c**) Axial and coronal T1-weighted MRI showing a left hemispheric arachnoid cyst that underwent a microsurgical fenestration procedure. (**d**, **e**) Axial T2-weighted MRI sections showing postoperative bilateral hygromas and hydrocephalus. Then the patient received a subduroperitoneal shunt associated to a ventriculoperitoneal shunt in a second time, because of an insufficient effect. (**f**) Axial T2-weighted MRI of postoperative shunt implantations. Afterward, the patient had several valve settings and was fine. (**g**–**i**) Axial T2-weighted MRI realized 6 years later for a symptomatology of progressive intracranial hypertension (cephalea, papilledema, etc.), showing a slit cyst and ventricle syndrome. (**g**) Catheter into the ventricle. (**h**) Catheter into the temporal cyst. (**i**) Cranial asymmetry and cerebral parenchyma hypertrophy. Then the patient received a lumboperitoneal shunt, which resolved the symptoms

even the placement of a bold cisterna magna-ventriculoperitoneal shunt, or the use of oral corticosteroids. But all of these treatments relative to this specific pathology will be detailed in the dedicated chapter (See in Treatment: Shunts, chapter ► "Slit Ventricle Syndromes").

Quadrigeminal Cyst

Quadrigeminal arachnoid cysts, like the interhemispheric cysts, may be associated with other central nervous system malformations (holoprosencephaly, Chiari type II, encephaloceles),

and they tend to be present in young children (Erşahin and Kesikçi 2009; Spennato et al. 2013).

Pathophysiology of Hydrocephalus

Because of the intimate relationship of quadrigeminal arachnoid cyst with the midbrain tectum, there is rapidly a compression or distortion of the aqueduct of Sylvius, which leads to an early triventricular obstructive hydrocephalus. Several authors suspect also the possibility of an associated deficiency of CSF reabsorption (El-Ghandour 2013; Gui et al. 2016). For the infants or neonates, it could be due to immaturity of the subarachnoid spaces. For the patients who had prior shunts, it could be due to loss of the ability of CSF absorption, characterized by shunt dependency. Nevertheless, the high rate of shunt independency in different series such as the one of Cinalli et al. can suggest that hydrocephalus associated with quadrigeminal cysts may be purely obstructive (Cinalli et al. 2010).

Subtypes

Quadrigeminal arachnoid cysts are heterogeneous and may have different extensions in the surrounding structures, according to the area of less resistance through which the cyst will expand: cranially in the region of the trigone, caudally in the supracerebellar cistern, anteriorly in the third ventricle and laterally to the ambient cistern.

Cinalli et al. proposed this classification based on the anatomical extension of the cyst, resulting in different possible surgical techniques for each type (Cinalli et al. 2010) (Table 9). Type 1 is the most frequent one.

Radiology (Figs. 26, 27, and 28)

Quadrigeminal cysts are seen as midline, supracerebellar, infratentorial, and/or supratentorial cysts abut the quadrigeminal cistern. They are occasionally paramedian. The enlarging cyst projects downward and backward to lie over

Table 9 Classification of quadrigeminal cysts according to Cinalli et al. (2010)

Subtype	Extension	Region of extension
Type 1	Supra- and infratentorial (Fig. 26)	Trigone upward and supracerebellar cistern downward
Type 2	Infratentorial (Fig. 27)	Supracerebellar or supra-retrocerebellar
Type 3	Lateral (Fig. 28)	Ambient cistern toward the temporal lobe

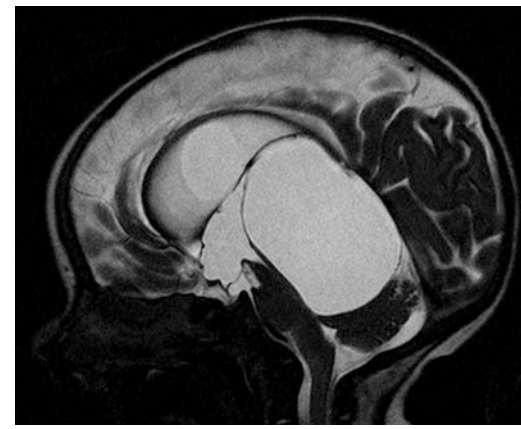


Fig. 26 Quadrigeminal arachnoid cyst **type 1**, characterized by infra- and supratentorial extension with severe aqueductal compression and hydrocephalus, on MRI, T2-weighted sagittal section

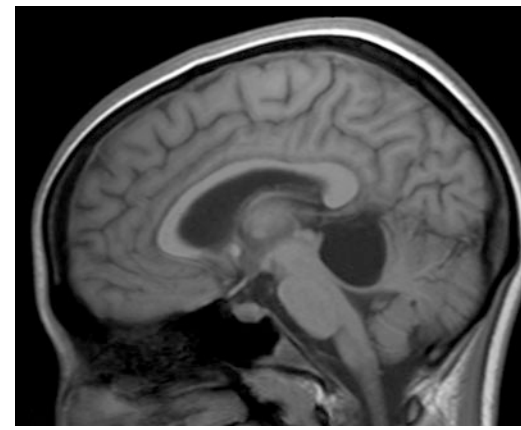


Fig. 27 Quadrigeminal arachnoid cyst **type 2**, characterized by strictly infratentorial extension, on MRI, T1-weighted sagittal section

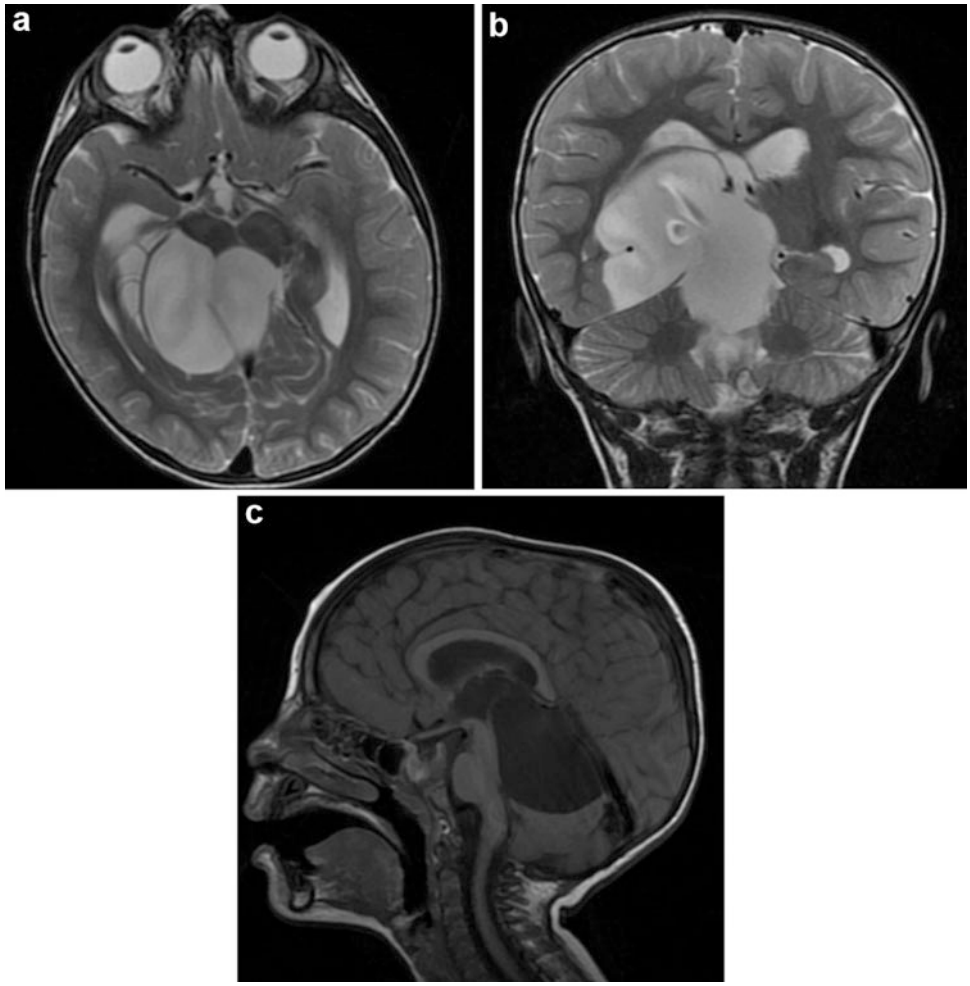


Fig. 28 Quadrigeminal arachnoid cyst **type 3** with a right extension to the middle cranial fossa and the ambient cistern, on MRI. (**a**) T2-weighted axial section. (**b**) T2-weighted coronal section. (**c**) T1-weighted sagittal section

the superior surface of cerebellum. The aqueduct and the fourth ventricle are displaced downward and forward and tectal compression occurs. MRI shows also the dilated third and lateral ventricles in addition to the quadrigeminal arachnoid cyst.

Their main differential diagnosis is the pulsion diverticula, often observed in severe triventricular hydrocephalus and consisting of a hernia of the medial wall of the ventricular atrium into the quadrigeminal cistern through the tentorial hiatus, induced by the pressure differential between supra- and infratentorial compartments (Spennato et al. 2013).

They can be differentiated from the cysts of the cistern of the velum interpositum (Fig. 29), because in the latter hydrocephalus is rare (Cinalli et al. 2010). Quadrigeminal cysts displace the internal cerebral veins upward, while the other displaces the veins downward (Figs. 29 and 30).

Treatment/Surgical Techniques

Microsurgical Procedure

Midline suboccipital craniotomy in the surgical approach of quadrigeminal arachnoid cysts is a more extensive procedure compared to endoscopic

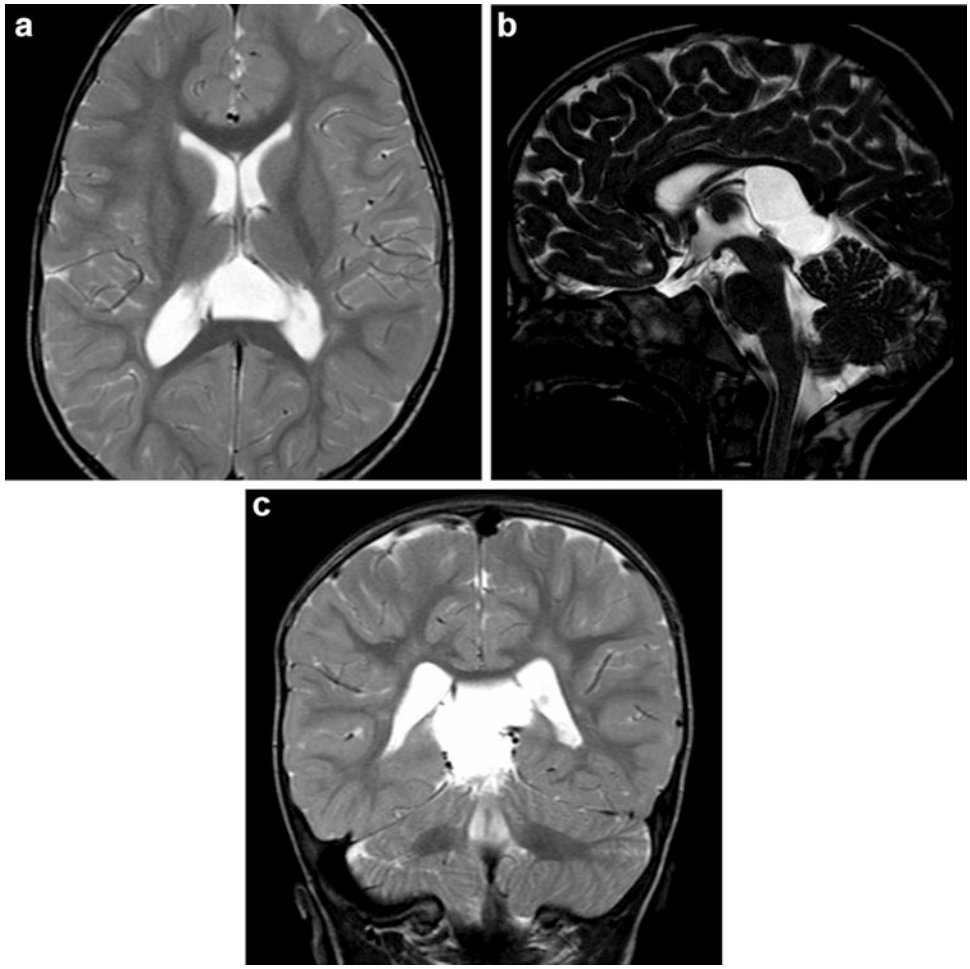


Fig. 29 Differential diagnosis: cyst of the velum interpositum, on MRI. (**a**) T2-weighted axial section. (**b**) CISS sequence (Constructive Interference in Steady State) on sagittal section. (**c**) T2-weighted coronal section

techniques. It will directly expose the cyst, allowing its complete or subtotal resection, and easier in type 2 cysts because of the absence of supratentorial component. However, the need in a second time of the implantation of a shunt seems to be high (75%) (Fewel et al. 1996). The advantage of craniotomy compared to endoscopy is the better control of hemostasis, but it is marked by significant more severe complications in historical series, such as neurological deficits (oculomotor palsy, hemiparesis), meningitis, seizures, and subdural hematomas (Ciricillo et al. 1991).

As quadrigeminal cysts are almost always associated to hydrocephalus, microsurgical procedure has difficulties to treat both problems.

Standard Microsurgical Procedure (Except Type 3)

The patient is in prone position with the head slightly bent. The approach is a classical infratentorial supracerebellar one for type 2 cysts and median interhemispheric trans-tentorial one for type 1.

A median occipital (or parieto-occipital) cutaneous incision is performed. The midline is kept through the dissection of the nuchal ligament allowing a comfortable craniotomy. The dura is opened according to anatomical target needs. A very great attention is paid to the venous sinuses and the torcular.

The aim of the procedure is to fenestrate the cyst through the posterior basal cisterns and the

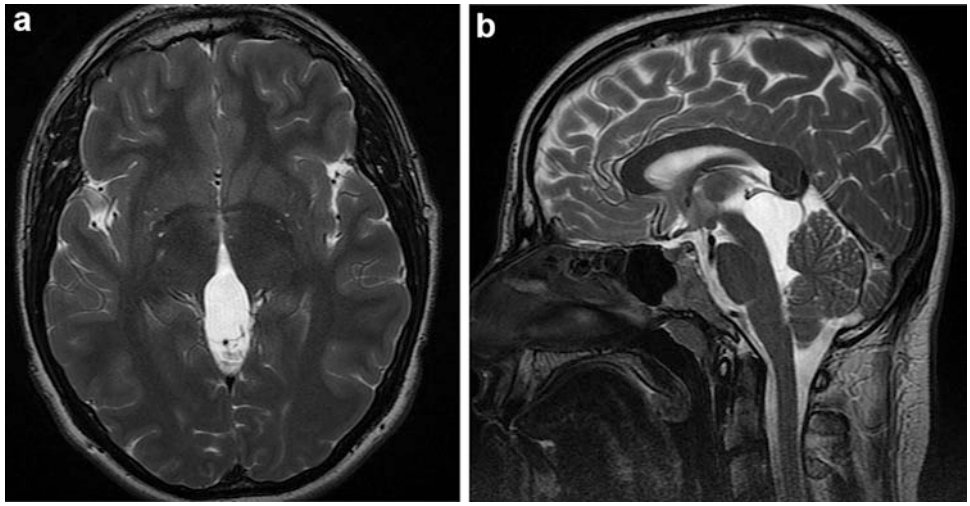


Fig. 30 Differential diagnosis: very rare case of intra-aqueductal cyst, on MRI. (**a**) T2-weighted axial section. (**b**) T2-weighted sagittal section. Note the tectal plate displaced posteriorly and superiorly

ventricles, trying to excise the more possible cystic wall. Then the dura, muscles, and skin are sutured the most watertight way possible.

Shunting (Fig. 31)

The placement of a cystoperitoneal shunt is an easy procedure but is marked by a high risk of cyst recurrence and undertreatment of hydrocephalus (Raffel and McComb 1988). Cystoperitoneal shunting or cysto-cisternal shunting may lead to early obliteration of the cyst but is associated with a high shunt malfunction rate of 40% (Ohnishi et al. 2007). Figure 31 shows the position of the catheter into a quadrigeminal arachnoid cyst, passing also by the right lateral ventricle.

Endoscopic Procedure

In Ersahin et al.'s series, 100% of 17 patients presented with associated hydrocephalus (Erşahin and Kesikçi 2009). They were all treated by endoscopic fenestration and associated endoscopic third ventriculostomy, while five have been previously shunted. Two of them also required an aqueductal stent. The procedure was successful in 53% of the children, although 29% of them, younger than 6 months, later necessitated a ventriculoperitoneal shunt. Only one patient younger than 6 months remained shunt free, while endoscopy was successful for all patients older than 6 months.

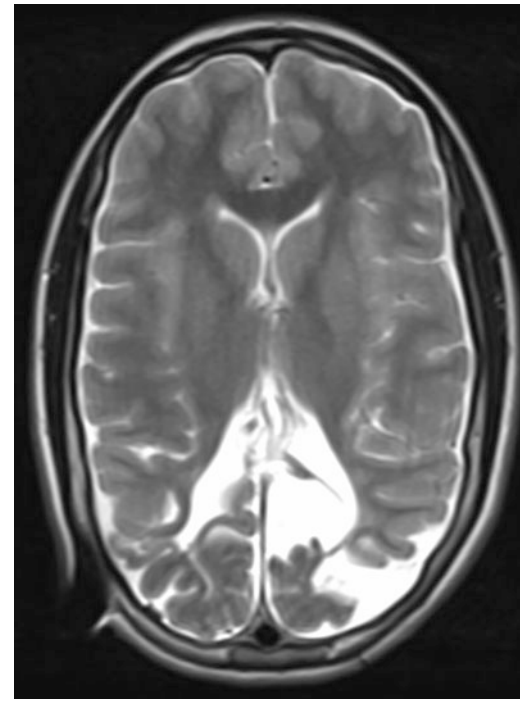


Fig. 31 Axial T2-weighted section of MRI showing the position of the catheter into a quadrigeminal arachnoid cyst together with the right lateral ventricle. This patient presented a clinicoradiological slit cyst/ventricle syndrome

Postoperative subdural hygromas developed in 18% of the patients with implantation of a subduroperitoneal shunt in two on three of them.

This study showed a very good success rate for children older than 6 months, who were preoperatively more symptomatic than the others.

In Cinalli et al.'s series of 14 patients, the global success rate of endoscopic procedure was 78%, which increased until 90% in the case of use of endoscopy as the first surgical option (Cinalli et al. 2010). The combined cystoventriculostomy and ventriculocisternostomy was performed in 43% of the children with the best results, while 88% of the patients treated with cystoventriculostomy alone required reoperation, including three ventriculoperitoneal shunts (37.5%). Fifty percent of the patients, who already had a shunt during the endoscopic procedure, became shunt independent.

In 2013, El-Ghandour published his series of 18 children with hydrocephalus (El-Ghandour 2013). He had a success rate of 83.3%, increasing up to 92.9% in the case of using endoscopic approach as the first line of treatment. Seventy-seven point eight of the cysts and 88.9% of the ventricles decreased in size. According to Cinalli et al., they noticed that the fact to have a previous shunt before endoscopic procedure decreased the possibility to become shunt independent and consequently the success rate of the intervention. They had also a better success rate when third ventriculostomy was feasible, associated to cystoventriculostomy. They suspect the possible decrease of CSF flow disturbances inside the third ventricle and so through the aqueduct, performing an ETV.

The most recent and the largest series is the one of Gui et al., including 28 hydrocephalic patients (25 children) who had undergone endoscopic treatment (Gui et al. 2016). No one benefited of shunting before the neuroendoscopic procedure. They performed 64% of combined lateral ventriculocystostomies and ETV, 11% of combined third ventriculocystostomies and ETV, and 25% of combined lateral and third ventriculocystostomies and ETV. The success rate was 89.3%, the cyst's size decreased in 78.6% of cases, and the ventricular system's size in 82.1%. 14.3% patients developed a subdural collection including 7.2% requiring a subduroperitoneal shunt.

As Cinalli et al. did (Cinalli et al. 2010), Garg et al. suggest to perform different endoscopic procedures according to the type of quadrigeminal arachnoid cyst (Garg et al. 2014). With their 18 patients treated, they propose to treat type 1 cysts by endoscopic fenestration of the cyst with ETV. In type 2 cysts, they performed craniotomy and cyst fenestration followed by the placement of a shunt in 75% of patients (cystoperitoneal or cystoperitoneal and ventriculoperitoneal shunt with Y connector). For type 3 cysts, they realized endoscopic cyst fenestration by ventriculocystostomy and ETV.

Standard Endoscopic Procedure (Cinalli et al. 2010; El-Ghandour 2013; Erşahin and Kesikçi 2009; Spennato et al. 2013; Lechanoine et al. 2018)

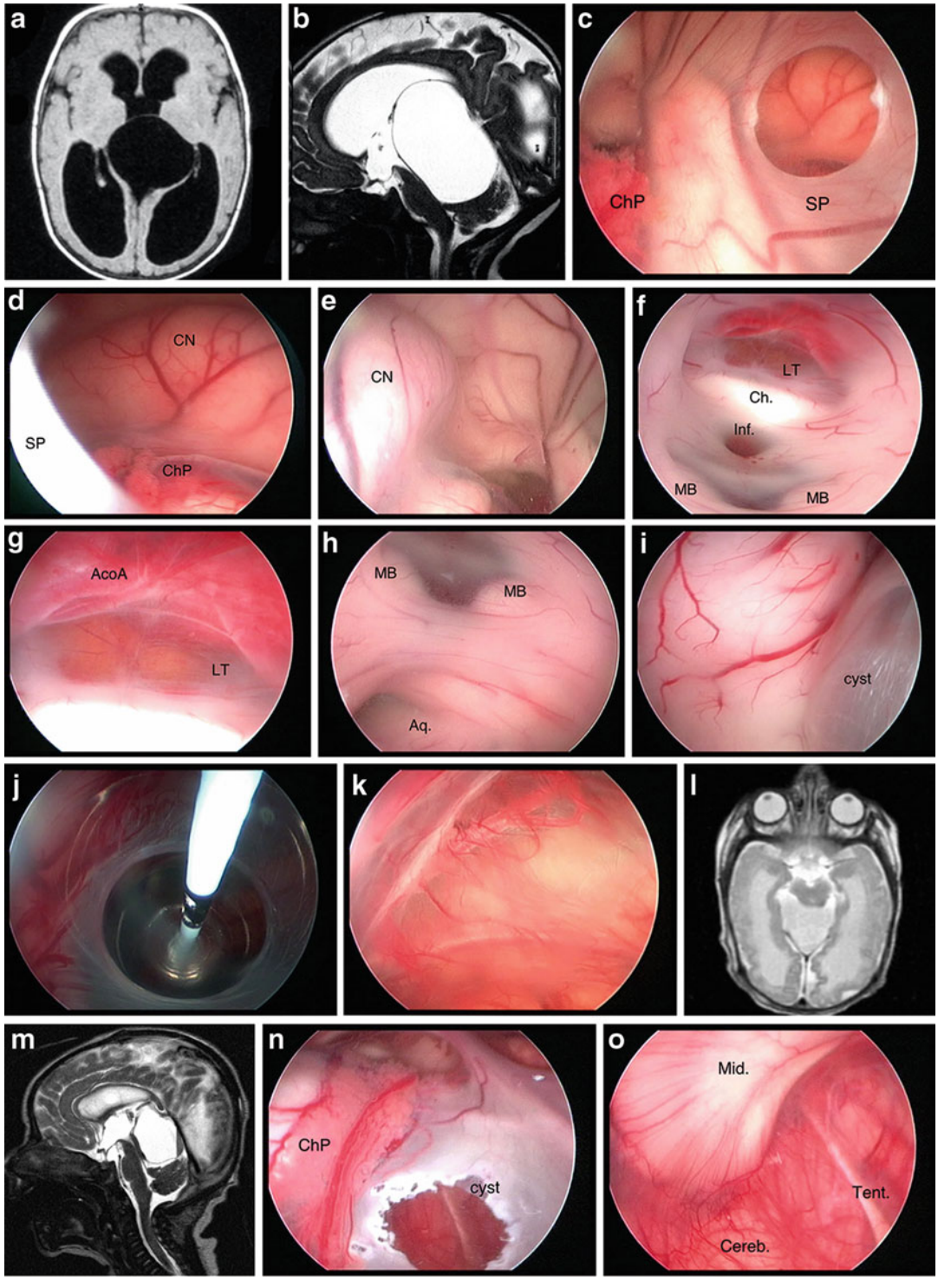
With respect to the subtype of quadrigeminal cyst, with different possible extensions, we must adapt our surgical approach for an optimal endoscopic trajectory intended during the perioperative planning. The use of neuronavigation system is highly recommended.

Type 1 (Figs. 32 and 33)

In this most frequent presentation, the cyst extends upward in the lateral ventricle, thinning the floor of the ventricular trigone, appearing below the ependymal, medially to the choroid plexus. A standard precoronal burr hole is made on the side of the biggest asymmetrical expansion of the cyst, allowing a ventriculocystostomy as a large opening of the upper pole of the cyst into the lateral ventricle, associated to an ETV. As said below, always control preoperatively the side of displacement of the cerebral veins to avoid their injury. To avoid closure of the stoma, the fenestrations should be at least of 10–15 mm. In infants, the penetrance point is made in the lateral angle of the fontanel. Figure 32 shows the endoscopic procedure performed on a quadrigeminal arachnoid cyst type 1 associated to hydrocephalus, with expected postoperative MRI results (Fig. 33).

Type 2

The cyst grows in the posterior part of the third ventricle. A standard precoronal burr hole is made, the endoscope passes through the foramen



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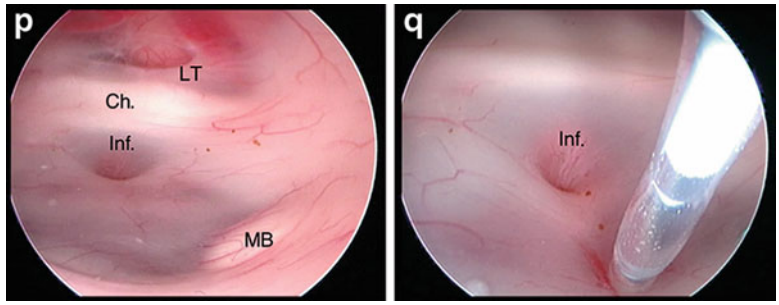


Fig. 32 **Case 1**. Two consecutive endoscopic procedures on a newborn with a quadrigeminal arachnoid cyst **type 1** associated to hydrocephalus. (**a**, **b**) Axial T1-weighted and sagittal T2-weighted MRI showing this median cyst with supratentorial extension. (**c**–**k**) First endoscopic procedure consisting of a ventriculocystostomy through a left lateral trans-fontanellar approach. (**c**) View from the left lateral ventricle of the contralateral caudate nucleus through the septum pellucidum. (**d**) Right caudate nucleus. (**e**) Left caudate nucleus. (**f**) Overview of the anterior part of the third ventricle. (**g**) View of the subcallosal area through the lamina terminalis. (**h**) Posterior part of the third ventricle with the aqueduct. (**i**) The endoscope is rotated posteriorly showing the wall of the cyst. (**j**) Fenestration of the cyst. (**k**) View from inside the cyst. (**l**, **m**) Axial T2-weighted and sagittal CISS-weighted postoperative MRI showing an incomplete decompression of the cyst and predominately the persistence of hydrocephalus. (**n**–**q**) Second endoscopic procedure, 1 month later for an associated ventriculocisternostomy. (**n**) View of the precedent fenestration with a better angle, because of the cyst and brain decompression. (**o**) Overview of the tectum of the midbrain, the cerebellum, and the right brain parenchyma, lying on the tentorium. (**p**, **q**) Ventriculocisternostomy

of Monro, and a double fenestration is performed through the anterior wall of the cyst and the floor of the third ventricle. The use of a steerable endoscope is recommended to perform both stomas through the same burr hole.

Type 3 (Lechanoine et al. 2018) (Fig. 34)
The cyst extends laterally toward the ambient cistern, and the standard approach is not appropriated. The head of the patient is rotated 90° to the contralateral side, a parietal burr hole is performed to approach the lateral ventricle at the level of the trigone, showing us just in front, the upper pole of the cyst. Then, several fenestrations should be made in the deep wall of the cyst, toward the basal cisterns. This procedure can be dangerous because the fenestration is made close to the internal cerebral veins and should be performed only if the wall of the cyst is avascular and translucent or with the help of neuronavigation. ETV is not possible with this approach. Figure 34 shows the endoscopic procedure performed twice on a quadrigeminal arachnoid cyst type 3 with lateral extension in the middle cranial fossa associated to hydrocephalus. The case insists on the necessity to achieve the largest possible fenestrations to avoid any recurrence. Lechanoine et al. made a video describing the step by step endoscopic procedure (Lechanoine et al. 2018).

Role and Importance of ETV in the Treatment

As we have seen above, many authors advocated the need of combining ETV with fenestration of the cyst. The argument is that, fenestrating only the cyst is insufficient to relieve the aqueductal compression, because of a likely alteration of the CSF pathway by extrinsic aqueductal stenosis arising from long-standing high pressure exerted by the cyst, which may persist despite the cyst opening (Di Rocco et al. 2005). It is made by fenestration of the anterior wall of the cyst through the third ventricle and then the fenestration of its posterior wall at the surface of the cerebellum, providing communication between the cyst and the supracerebellar cistern. With ETV, the aqueduct is bypassed into the anterior basal cisterns.

Gangemi et al., in a study of 16 patients, recommend to treat this cyst by endoscopic

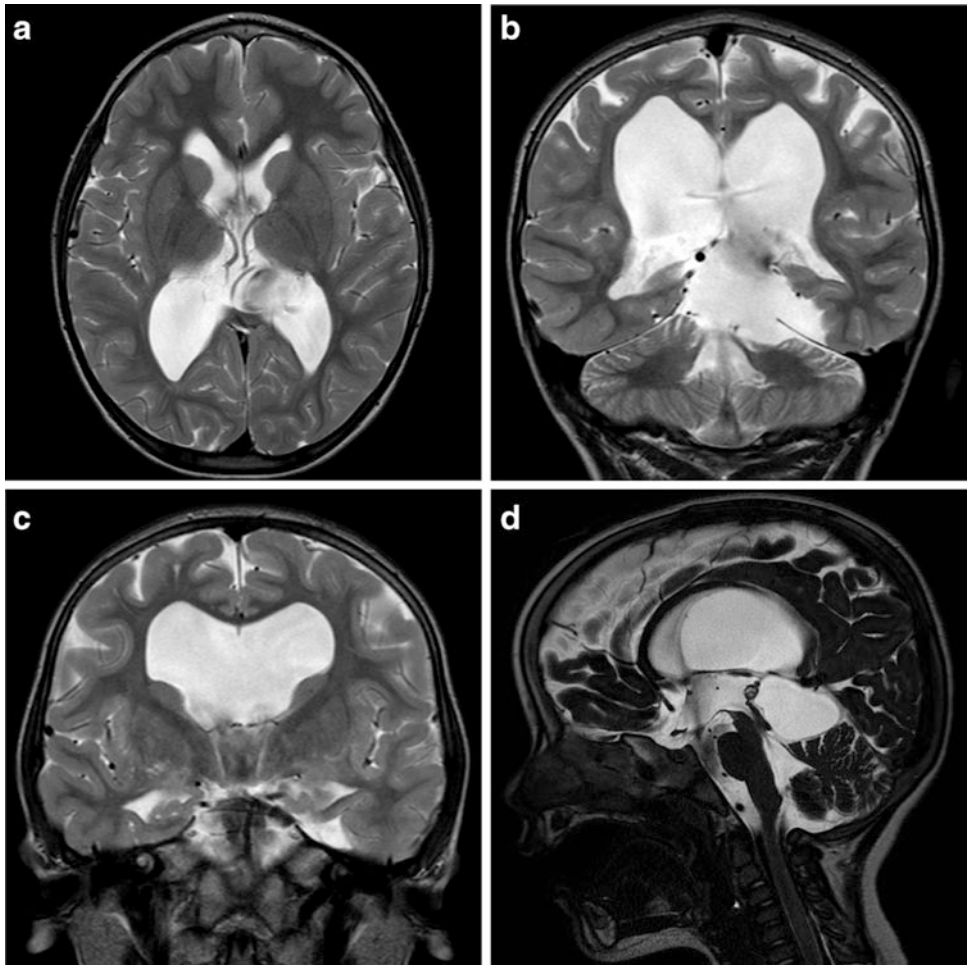


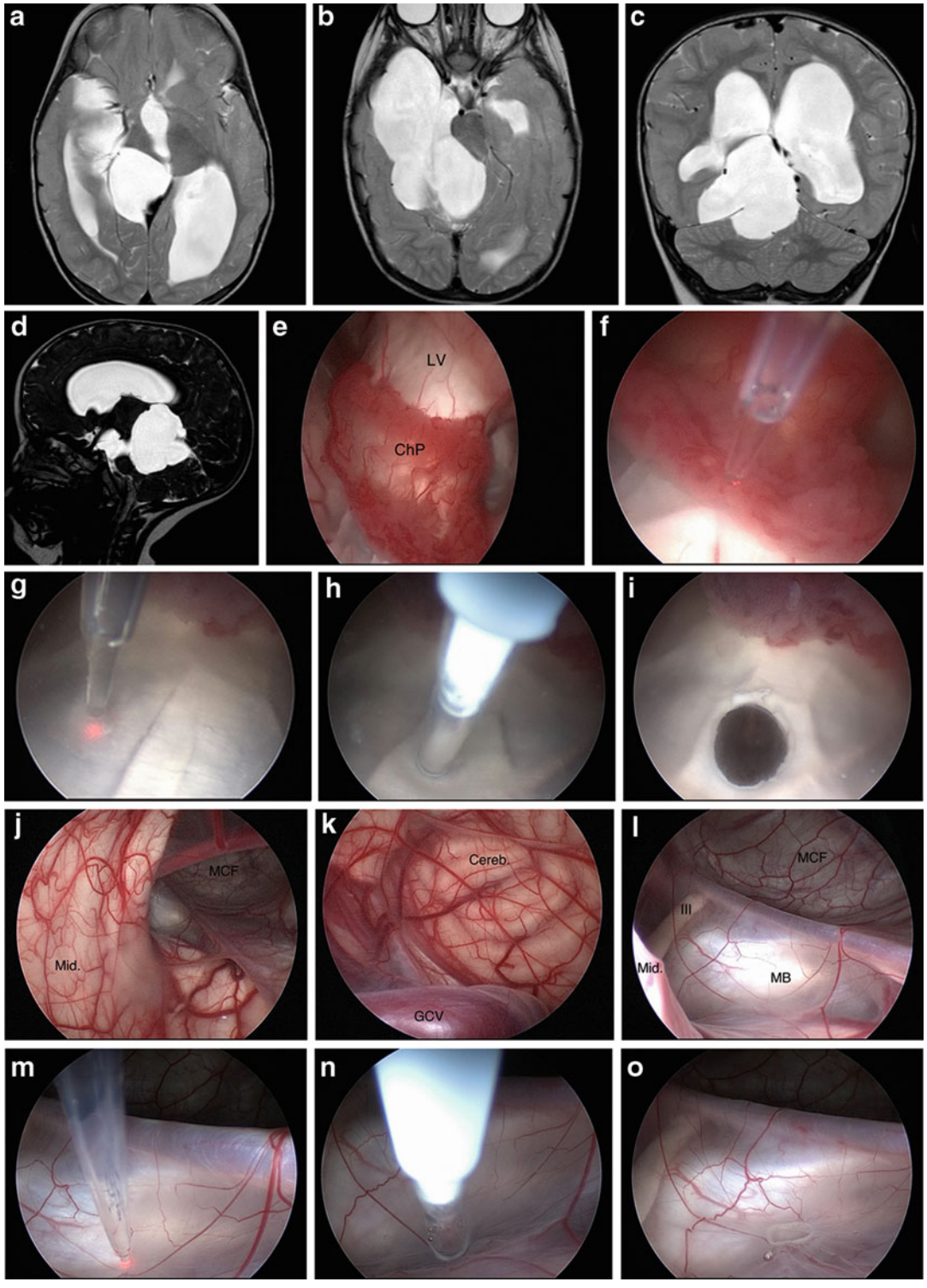
Fig. 33 **Case 2**. Kind of expected results on another patient (than CASE 1) with quadrigeminal arachnoid cyst **type 1** and hydrocephalus. (**a**, **b**) T2-weighted MRI with axial and coronal sections, showing the artifacts of flows through the cystoventricular fenestration. (**c**, **d**) Coronal T2-weighted and sagittal CISS-weighted sections showing the artifacts of flows inside the third ventricle and through the third ventriculocisternostomy into the interpeduncular and prepontine cisterns

procedures, because they found an equivalent success rate between patients treated by neuroendoscopy (87.5%) and treated by traditional surgery such as craniotomy and cyst excision and/or shunt (85%) (Gangemi et al. 2005). They advocated that endoscopic interventions are less invasive and avoid shunt dependency.

Discussion on Treatment

In 2011, Gangemi et al. have performed a comprehensive review of 61 reports in the literature comparing the clinicoradiological outcome of the traditional procedures (microsurgical fenestration and/or shunting) with endoscopic fenestration in the treatment of 63 reviewed cases of quadrigeminal arachnoid cysts (Gangemi et al. 2011) (Table 10).

According to the last studies, neuroendoscopy seems to be very effective in the treatment of quadrigeminal arachnoid cysts, with the best success rate in children older than 6 months, when combined ETV and cystoventriculostomy can be performed as a first surgical option, allowing shunt independence, in a single procedure. It can also be an alternative treatment of shunt revision in patients already shunted, in order to remove the shunt.



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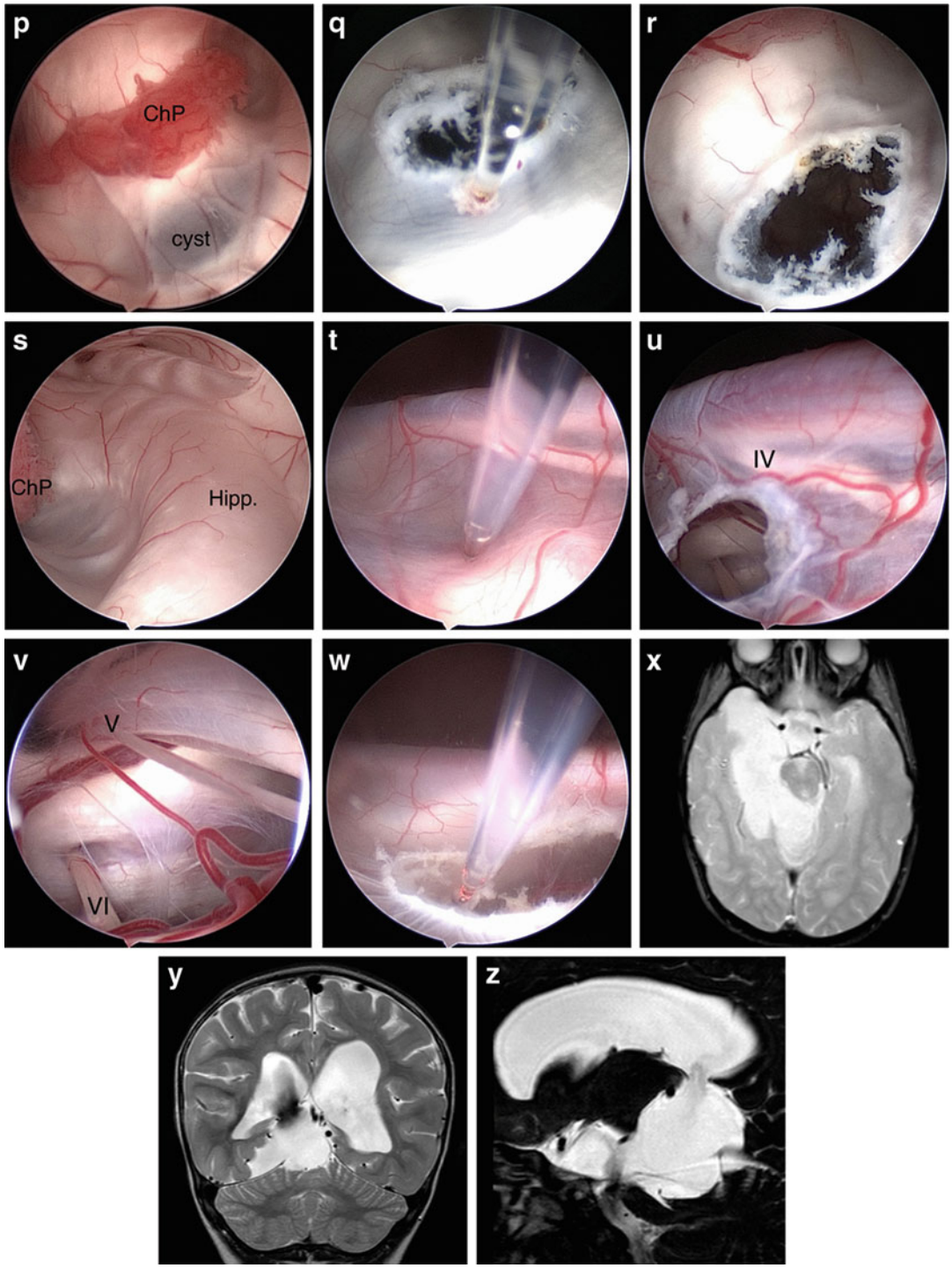


Fig. 34 **Case 3** (Lechanoine et al. 2018). Two endoscopic procedures spaced of 6 months on a patient with a quadrigeminal arachnoid cyst **type 3** with right lateral extension in the middle cranial fossa and around the midbrain; associated hydrocephalus. (**a**, **b**) Axial section of T2-weighted MRI. (**c**) Coronal section of T2-weighted MRI. (**d**) Sagittal section of CISS-weighted MRI. Note the quadrigeminal cyst following the right portion of the midbrain

Table 10 Success rate between surgical options for quadrigeminal arachnoid cysts treatment without isolating hydrocephalus (Gangemi et al. 2011). Note that microsurgical procedure and shunting are placed on the same line

Technique	Good outcome (%)
Microsurgical fenestration and/or shunting	85
Endoscopic fenestration	88.5

Good outcome = complete or partial clinical remission

Posterior Fossa Cyst

Pathophysiology of Hydrocephalus

Hydrocephalus in posterior fossa arachnoid cysts may be caused by an obstruction of CSF pathways by compression of the aqueduct but may be also caused by obstructing the openings of the fourth ventricle or trapping them at the foramen magnum (Di Rocco et al. 1981, 1979; Galarza et al. 2010; King et al. 2010). Maybe the location of these cysts in the posterior fossa could also provoke restriction of natural CSF flows into the basal cisterns.

Subtypes

Infratentorial arachnoid cysts have plenty of classifications in the literature with no guideline. Little et al. made a classification simply based on the localization: prepontine (extension of suprasellar cases), cerebellopontine angle, tentorial, inferior midline, superior midline, intraventricular, and hemispheric (Little et al. 1973).

Vaquero et al. proposed another one based on the localization but also on the extension possibility (Vaquero et al. 1981).

All authors agree to differentiate arachnoid cysts of the posterior fossa from other malformations such as mega cisterna magna or Dandy-Walker and its variant (see in part "Prenatal and Neonatal Hydrocephalus", chapter ▶ "Hydrocephalus Associated with Cerebral Malformations").

The difference with mega cisterna magna must also be done. It consists of a developmental defect resulting from a splitting of the falx cerebelli and the absence of the transverse arachnoid fold, which normally constitutes the upper limit of the cisterna magna. Mega cisterna magna is usually asymptomatic and does not need any treatment. Factors for the diagnosis of retrocerebellar arachnoid cyst are the enlargement of the posterior fossa and thinning of the occipital bone and hydrocephalus.

Most of the prepontine or retroclival cysts are part of suprasellar cysts (treated in the subchapter), as with recent publications they seem to come from the same entity.

Fig. 34 (continued) instead of the ambient cistern. (**e**–**o**) First endoscopic procedure for a ventriculocystocisternostomy, through a right parasagittal trans-parietal approach. (**e**) Posterior view of the right ventricular atriums and its choroid plexus. (**f**) Gentle coagulation of the choroid plexus to retract it, revealing the inter-trigonal velum interpositum, which has a common wall with the inner membrane of the cyst. (**g**–**i**) Ventriculo-cystic fenestration. A posteriori this stoma turns out to be too small. (**j**) Posterior overview from inside the cyst, showing the tectum of the midbrain and the middle cranial fossa. (**k**) Superior view of the cerebellum, overlooked by the great cerebral vein. (**l**) Posterior view of the right petro-clival angle in lieu of ambient cistern. (**m**–**o**) Cysto-cisternal fenestration. A posteriori this stoma turns out to be too small as well. (**p**–**w**) 6 months later it turned out that the clinicoradiological response of the patient was not enough; a second endoscopic intervention was performed to reopen widely the fenestrations. (**p**) Posterior view of the right ventricular atrium. Note the occlusion of the previous stoma. (**q**, **r**) Bigger new ventriculo-cystic fenestration. (**s**) Posterior view of the frontal horn of the right lateral ventricle showing amply the hippocampus, its anterior winding, and its indented aspect. Not that the cyst has discarded the connection of the hippocampus with the fimbria and the choroid fissure that are pushed to the left side. (**t**–**w**) Wide new cysto-cisternal fenestration in the petro-clival right angle, below the fourth cranial nerve and the superior petrosal sinus. (**v**) View, through the stoma, of the right sensitive and motor roots of the trigeminal nerve and the Dorello's canal of the sixth cranial nerve. (**x**–**z**) Postoperative MRI showing a satisfying decreasing of the cyst, brain decompression and decreasing of hydrocephalus. (**x**) T2-weighted axial section. (**y**) T2-weighted coronal section showing artifacts of flows through the ventriculocystic fenestration. (**z**) CISS-weighted sagittal section showing artifacts of flows through the cysto-cisternal stoma

Table 11 Classification of posterior fossa arachnoid cysts

Subtype	Extension
Midline	Pushes the vermis anteriorly, separating the two cerebellar hemispheres
Retrocerebellar	Pushes both cerebellar hemispheres downward (Figs. 35 and 36)
Supracerebellar	
Lateral cerebellar	Compresses one cerebellar hemisphere (Figs. 37 and 38)
Cerebellopontine angle	Displaces the cerebellum and brainstem contralaterally (Fig. 39)
Large cysts	Cysts occupying several above-cited compartments
Intraventricular	Within the fourth ventricle
Others (retroclival, tentorial notch)	

Often, supracerebellar arachnoid cysts are mingled with quadrigeminal cysts, which are treated in the section "Quadrigeminal Cysts."

We propose to use a classification based on anatomical location and extension (Table 11).

Radiology (Figs. 35, 36, 37, 38, and 39)

As said above, it is very important to differentiate arachnoid cysts of the posterior fossa from other malformations like the Dandy-Walker malformation and its variants (see chapter ▶ "Hydrocephalus and the Dandy-Walker Malformation").

Treatment/Surgical Techniques

Symptomatic posterior fossa arachnoid cysts are associated with hydrocephalus in around 90% of cases. Specific treatment of the cyst or of hydrocephalus should be part of preoperative strategy. Ideally the treatment of the cyst should resolve hydrocephalus.

According to the literature, the option of the best treatment should depend on the relationships of the cyst with ventricles and basal cisterns. Cysts can be communicating or not communicating (Juan F. Martínez-Lage et al. 2011).

Some authors consider that, even with associated hydrocephalus, the treatment of choice remains microsurgical fenestration or excision, because according to them, hydrocephalus is secondary and obstructive and will be consequently resolved.

Microsurgical Procedure

Cysts of the cerebellopontine angle usually require microsurgical fenestration because of this extremely delicate location that forbids the possibility of shunt implantation, with a high risk of injury of neurovascular structures after decompression. Using of microsurgical instruments is more than recommended for dissection of thick arachnoid membranes through cranial nerves and vascular structures of the cerebellopontine angle, with an associated better overview of the angle.

Di Rocco et al. treated with good outcome eight children by large fenestration and excision of posterior fossa cysts using common microsurgical techniques (Di Rocco et al. 1981). He has also treated successfully by open craniotomy an arachnoid cyst of the fourth ventricle (Di Rocco et al. 1979).

Samii et al. used microsurgical procedure to treat 12 patients affected by cerebellopontine angle cysts for the most part of them or lateral cerebellar cysts (Samii et al. 1999). It was possible to perform a complete cyst excision only in one case. But his series includes children and adults. A suboccipital retrosigmoid approach was performed for all patients; the outcome was good for all except one patient.

Retrocerebellar cysts of the midline can also be considered as eligible to open craniotomy procedures.

Standard Microsurgical Procedure

For all cases, a suboccipital craniotomy should be performed. According to the location of the cyst, we will adapt it.

Cerebellopontine Angle and Lateral Cerebellar (Fig. 40)

A suboccipital retrosigmoid approach is needed. The patient is positioned ventrally with the head turned ipsilaterally or in lateral derivative positions. The smaller the cyst the more tangential should be the working axis to the intracranial face of the petrous bone. This position is obtained by a gentle

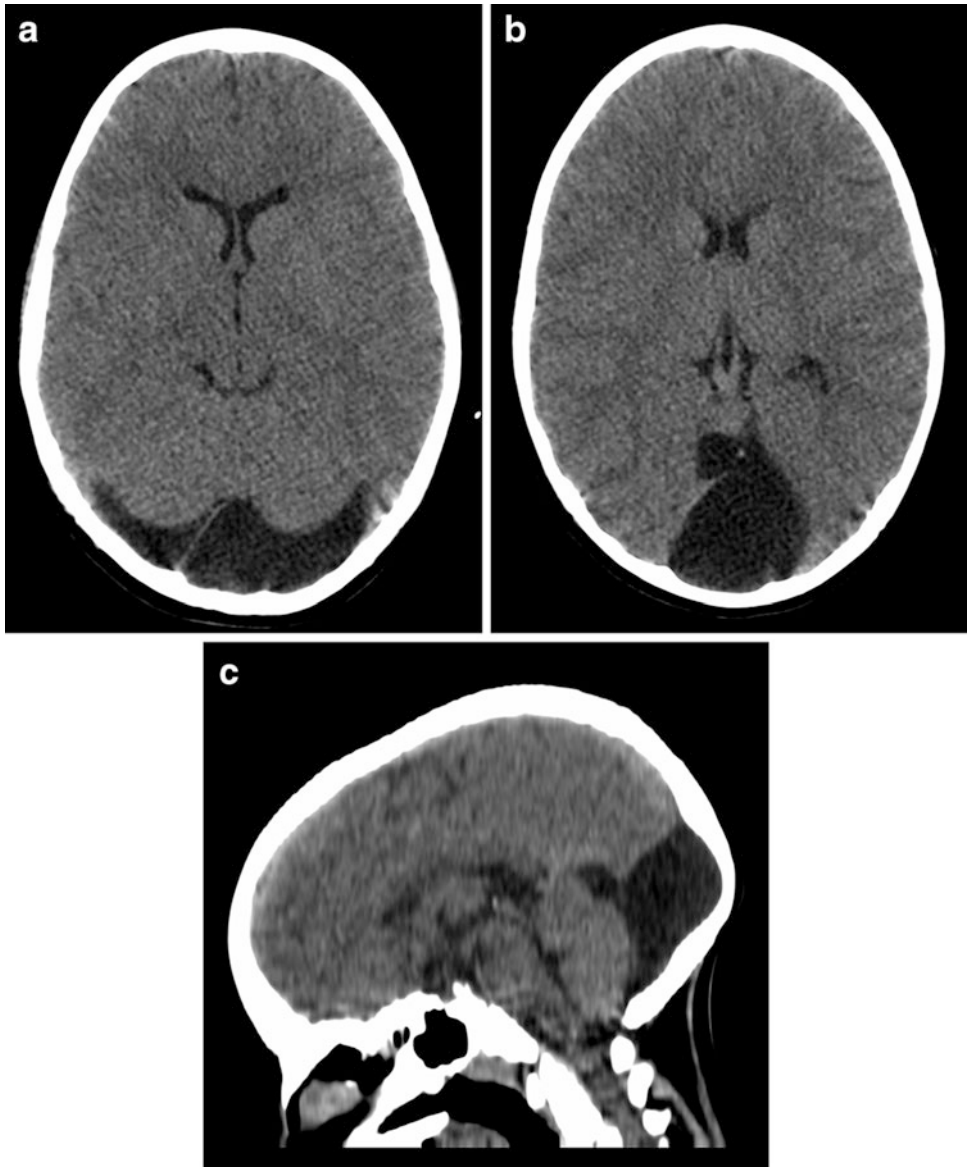


Fig. 35 Computed tomodensitometry of a retrocerebellar arachnoid cyst of the midline. (**a**, **b**) Axial sections. (**c**) Sagittal section

flexion, eversion, and lateral rotation of the head. The skin incision is adapted to the location of the cyst but is standardly 1 in. behind the mastoid process, one third above, and two third below the projection of the lateral venous sinus. A keyhole craniotomy is performed classically in the angle of the lateral and sigmoid sinuses but also adapted to the cyst location. In the particular case of arachnoid cyst of the cerebellopontine angle, no brain retraction is necessary. The use of microsurgical instruments allows large fenestrations of the cyst between the neurovascular structures, which have to be recognized and gently dissected. The ideal situation is a complete excision of the wall of the cyst. Fenestrations consist of cystocisternostomy and cysto-fourth-ventriculostomy if necessary.

The suture of the dura mater has to be particularly watertight because of high risks of CSF

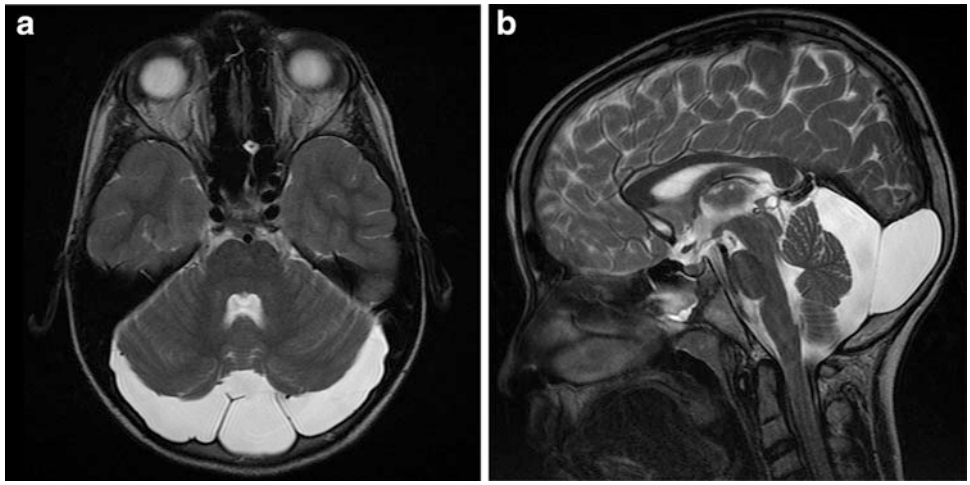


Fig. 36 T2-weighted MRI of a retrocerebellar arachnoid cyst. (**a**) Axial section. (**b**) Sagittal section

flowage. Figure 40 shows the microsurgical procedure performed on a left lateral cerebellar arachnoid cyst.

Midline

A median suboccipital infratentorial approach is performed and also adapted to the location of the cyst. Figure 40 shows the microsurgical procedure performed on a left lateral cerebellar arachnoid cyst.

Shunting

Different options are available such as implantation of a ventriculoperitoneal shunt alone, a cystoperitoneal shunt alone, or combined shunting of both ventricle and cyst. In case of combined shunt, it is highly recommended to use a three-way connector between the intracranial catheters and the valve.

King et al. reported the successful treatment of giant retrocerebellar arachnoid cysts by ventriculocystostomy in combination with direct hydrocephalus treatment (endoscopic third ventriculostomy or shunt placement) (King et al. 2010).

The implantation of a shunt in a posterior fossa arachnoid cyst exposes to the risks of shunt malfunctions, shunt dependency, slit cyst syndrome, and a proper entity as an induced chronic tonsillar herniation (see below). In the literature, the percentage of shunt malfunction is around 10–25.

Standard Shunting Procedure (Fig. 41)

Because of the need of shunting the posterior fossa, careful installation of the patient has to be done. The most comfortable position is a supine or slightly lateral rolled position with the head turned contralaterally at the maximum in order to correctly expose the occipital bone. The use of neuronavigation system is highly recommended as it will allow finding the cyst but mainly finding the ideal intracranial position of the catheter, thinking to the future position of the cerebellum after cystic decompression. Figure 41 shows a retrocerebellar arachnoid cyst leading to acute hydrocephalus of a newborn, treated by shunting procedure.

Endoscopic Procedure

One should pay attention to the communicating character or not (with the fourth ventricle) of arachnoid cyst of the posterior fossa. According to King et al., communicating arachnoid cysts may be first treated by endoscopic way (King et al. 2010). As usual, the preoperative MRI with CISS-weighted and flow sequences has to be carefully analyzed to look at the membranes of the cyst. For these cases, King et al. recommend to perform a third ventriculostomy

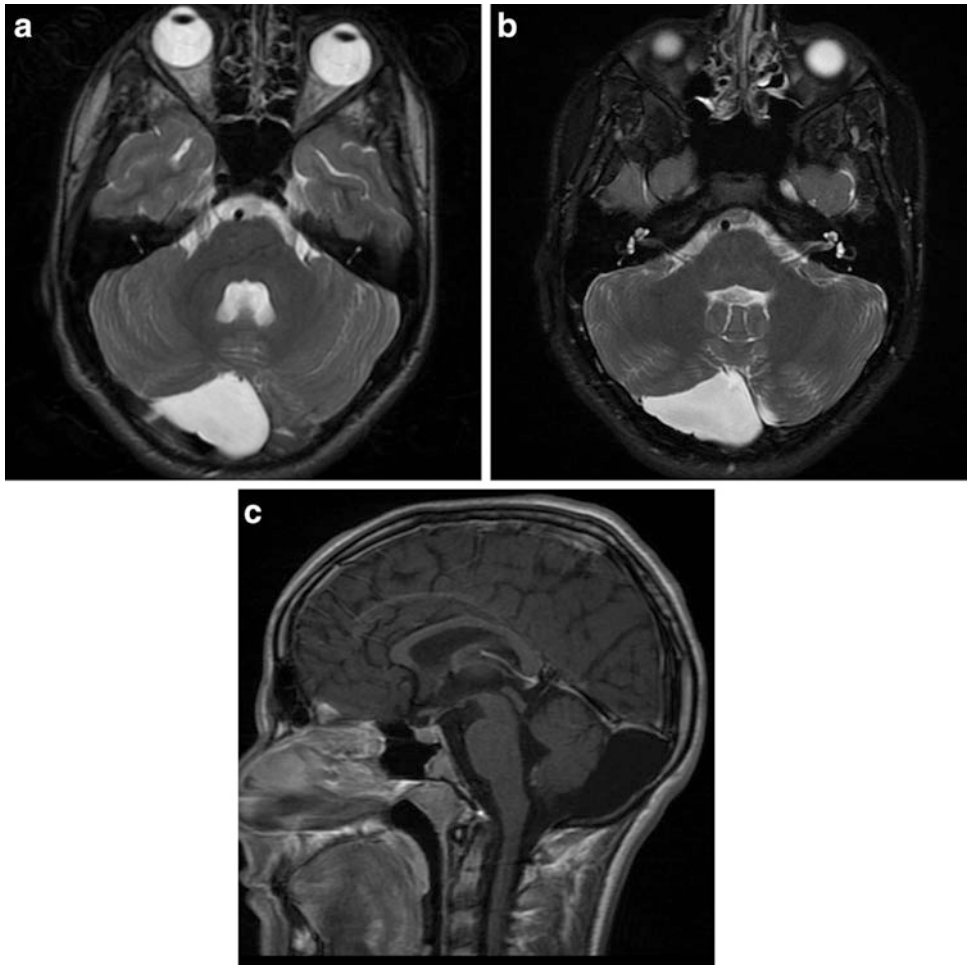


Fig. 37 Right paramedian arachnoid cyst of the posterior fossa. (**a**, **b**) T2-weighted axial sections of MRI. (**c**) T1-weighted sagittal section

(ETV) or a ventriculocystostomy combined with a third ventriculostomy, with good results on both the hydrocephalus and the cyst volume.

Idris et al. described three cases of endoscopic fenestrations by three different surgical approaches: transfrontal transaqueductal, transtrigonal, and suboccipital infratentorial supracerebellar with good outcome (Idris et al. 2016).

The transaqueductal trans-Magendie fenestration of arachnoid cyst of the posterior fossa has also been described (Feletti et al. 2016). They claimed that an endoscopic one-time treatment is better, performing an ETV and a cystocisternostomy in the same time with a flexible endoscope.

Standard Endoscopic Procedure (Figs. 42 and 43)

The use of the neuronavigation system is particularly recommended as it can help in the disturbed anatomy, to locate the position into the cyst and the ventricle and estimate the presence of neurovascular structures under cystic membranes.

ETV and/or Ventriculocystostomy

The patient is positioned supine with the neck slightly flexed, as for an endoscopic third ventriculostomy procedure. The burr hole is made on the mid-pupillary line according to the preoperative planning. If only ETV is needed, one

can use a coronal/precoronal burr hole. If a transaqueductal trajectory is desired, the burr hole will be more anterior. In case of combined procedures, the location of the burr hole has to be adapted, helped by neuronavigation. The orientation of the aqueduct is an important point that has to be studied on preoperative MRI.

An incision of the dura mater is made allowing its subsequent watertight closure. The rigid or flexible endoscope is introduced in the frontal horn of the lateral ventricle.

The ETV procedure is treated in (See in Treatment: Endoscopy, chapter ▶ "Endoscopic Third Ventriculostomy").

The transaqueductal passage can be performed only with a perfect axis through it with the rigid endoscope or with the flexible one, to avoid damages to the periaqueductal substance and particularly to the tegmentum of the midbrain. Then, ventriculocystostomy and/or cystocisternostomy is performed.

The endoscope is removed, and attention is paid to avoid cerebrospinal fluid loss, avoiding brain, ventricular, or cyst collapse while maintaining flows through the fenestrations. The dura is sutured.

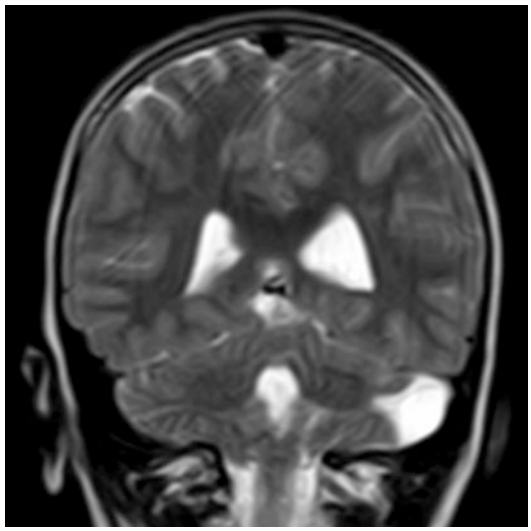


Fig. 38 Small left lateral arachnoid cyst of the posterior fossa on a T2-weighted MRI on coronal section

Suboccipital Infratentorial Supracerebellar Approach

The patient is in ventral position. The orientation of the head depends on the preoperative planning.

If a cystoventriculostomy and/or an inferior aqueductoplasty is needed, the midline is kept at the zenith, and the head is maximally flexed to catch the axis of the aqueduct.

If the cyst is lateral to the cerebellum, the head will be turned to the ipsilateral side; the cutaneous incision and burr hole will be lateral.

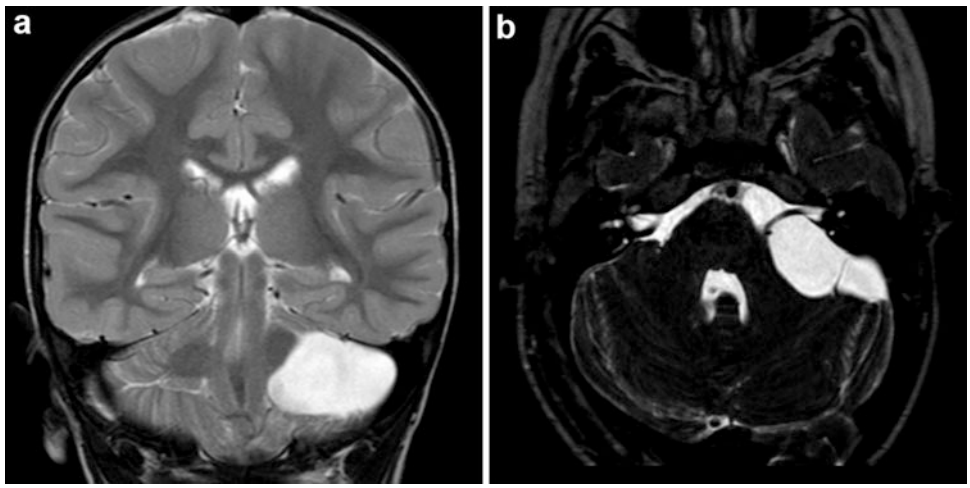


Fig. 39 MRI of a left arachnoid cyst of the cerebellopontine angle. (**a**) T2-weighted coronal section. (**b**) CISS-weighted axial section passing by the left trigeminal nerve, which is pushed forward by the cyst

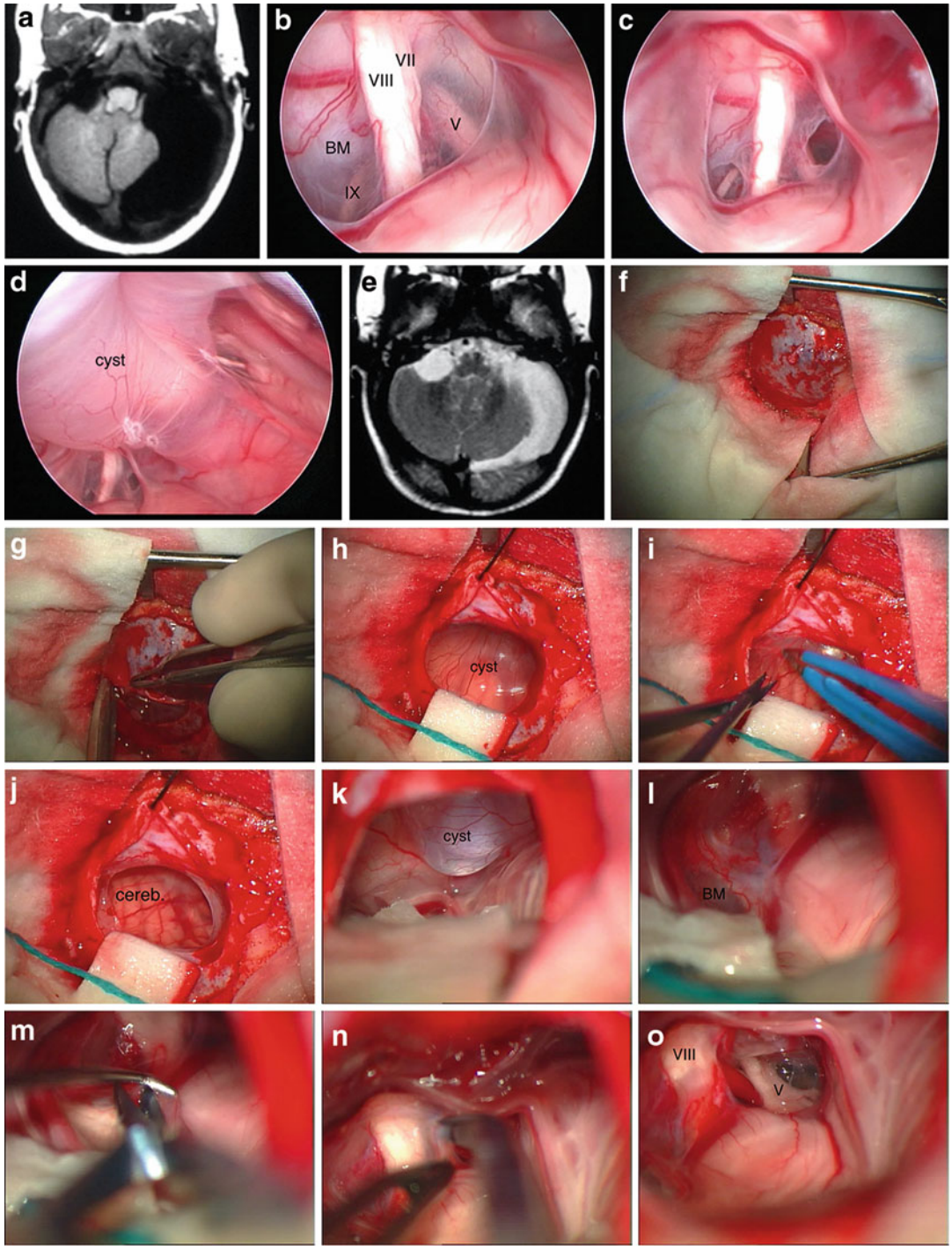


Fig. 40 (continued)

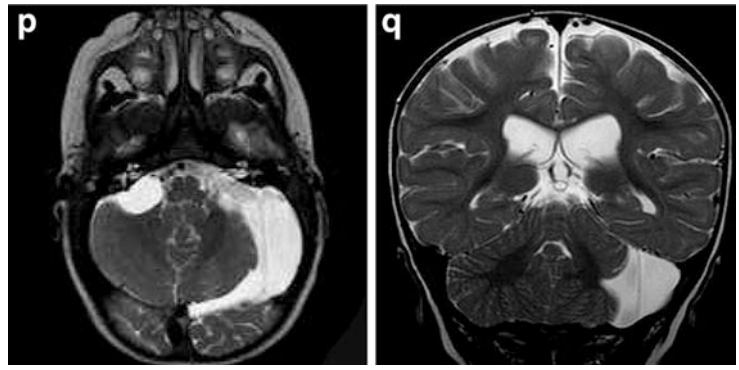


Fig. 40 Left lateral cerebellar arachnoid cyst. (**a**) Preoperative axial T1-weighted MRI showing the compression of the left cerebellar hemisphere. (**b**–**d**) The patient underwent first an endoscopic procedure with fenestrations. (**e**) The clinical and radiological results were not sufficient as seen on an axial T2-weighted MRI. (**f**–**o**) Secondary microsurgical procedure. (**f**) Left suboccipital retrosigmoid craniotomy of 2 cm of diameter. (**g**) Dura opening. (**h**) View of the external wall of the cyst. (**i**, **j**) Excision of a part of the wall. (**k**, **l**) Way down to the cerebellopontine angle. (**m**–**o**) Microsurgical dissection and fenestration of the inner wall of the cyst and basal cisterns between cranial nerves and vascular structures. (**p**, **q**) Postoperative T2-weighted MRI showing artifacts of flows through the cysto-cisternal stoma. Note the decompression of the external cystic wall but the presence of an ipsilateral subdural hygroma. (**p**) Axial section. (**q**) Coronal section

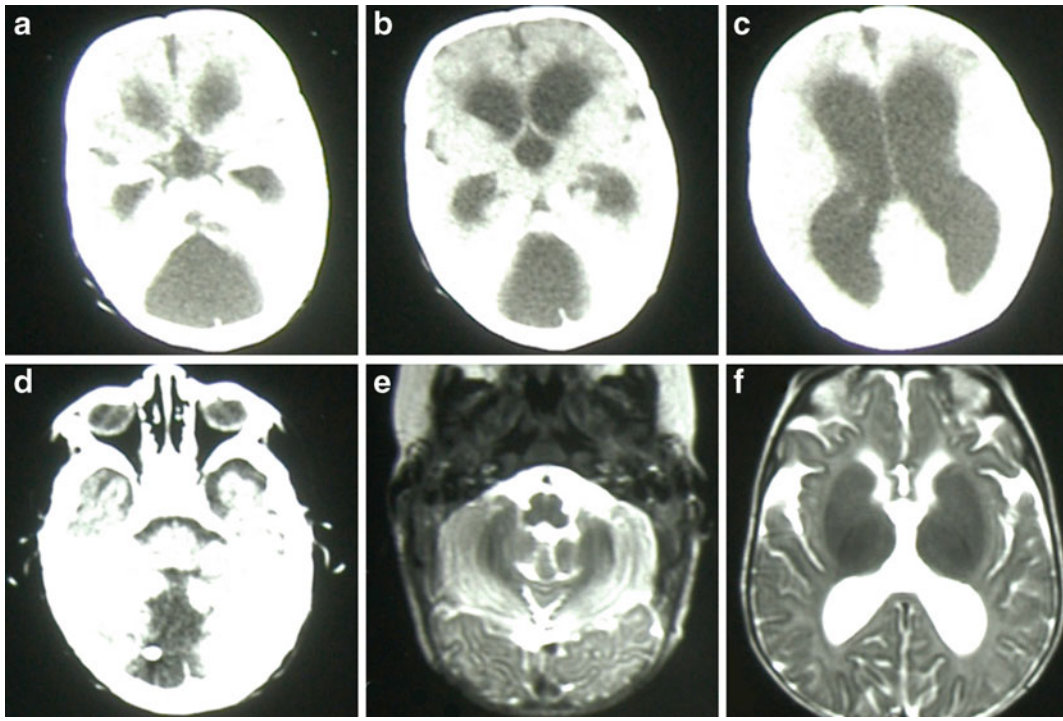


Fig. 41 (**a**–**c**) Computed tomodensitometry on axial sections of a newborn developing acute intracranial hypertension. Note the severe acute hydrocephalus on obstructing process by a retrocerebellar arachnoid cyst. (**d**) Axial postoperative computed tomodensitometry showing the implantation of a cystoperitoneal shunt. (**e**, **f**) T2-weighted axial MRI performed 1 month later, showing a satisfying collapse of the cyst and the subsequent resolution of the obstructive hydrocephalus

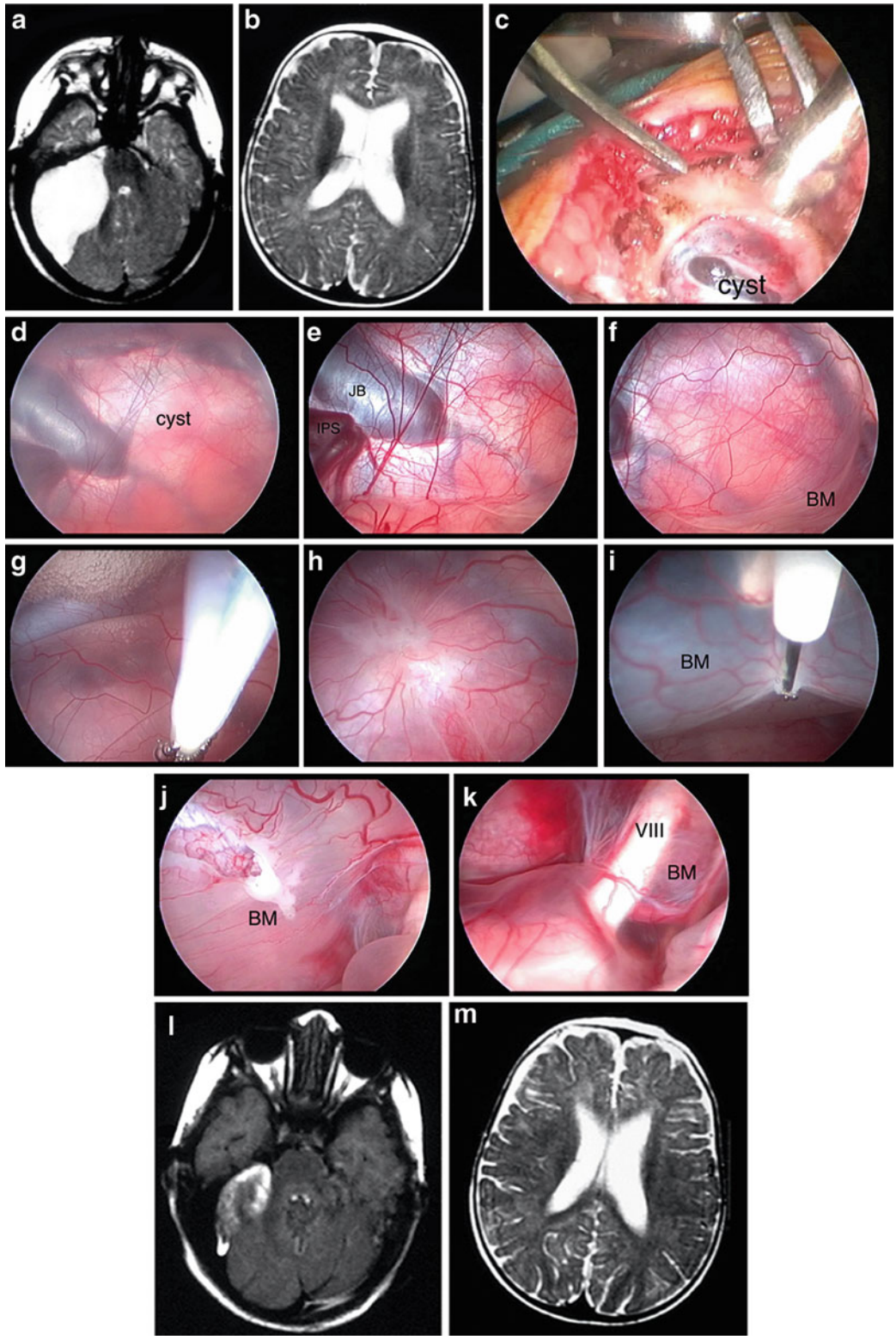


Fig. 42 **Case 1**. Right lateral cerebellar arachnoid cyst. (**a**, **b**) T2-weighted axial sections on MRI, showing the cyst and associated hydrocephalus. (**c**) Extracranial view of the operative field showing a right occipital bur hole, with dura

If the cyst is supracerebellar, the head will not be much flexed, keeping an axis tangential to the tentorium.

An incision of the dura mater is made allowing its subsequent watertight closure. The rigid endoscope is then introduced. The cysto-cisternal and cystoventricular fenestrations are made with conventional endoscopic techniques. Figure 42 shows the endoscopic procedure performed on a right lateral cerebellar arachnoid cyst. Figure 43 illustrates the endoscopic procedure on a large right lateral infracerebellar arachnoid cyst associated to hydrocephalus.

Discussion on Treatment

In 2011, Gangemi et al. have performed a comprehensive review of 61 reports in the literature comparing the clinicoradiological outcome of the 3 surgical procedures in the treatment of 83 posterior fossa arachnoid cysts (Gangemi et al. 2011) (Table 12). The distribution of cases was as follows: 37% in the midline cerebellar region (77% retrocerebellar, 23% supracerebellar), 24% in the lateral cerebellar region, 27% in the cerebellopontine angle, and 10% of large cysts with extension in both midline and lateral regions.

Hydrocephalus and Arachnoid Cyst Within the Fourth Ventricle

Arachnoid cysts of the fourth ventricle are extremely rare entities. Only 14 case reports including children and adults can be found in the literature. In 1979, Di Rocco reported the first case of a 7 years old boy presenting with hydrocephalus caused by a fourth ventricle's cyst (Di Rocco et al. 1979).

Clinically, most of the reported cases presented with the similar triad symptoms of a normal pressure hydrocephalus, often with ataxia in the foreground.

Radiologically, they may provoke an apparently quadriventricular hydrocephalus with a fourth ventricle dilated by a cyst, making mostly a huge compression of the brainstem and cerebellum descending through the foramen magnum.

Radiological examination must differentiate them from ependymal, dermoid, epidermoid, and astrocytoma cysts and a trapped fourth ventricle. Most of the authors agree to say that the MRI flow sequences are very useful to differentiate various cysts and guide the operative technique.

Some of these cysts were treated first by a ventriculoperitoneal shunt, which provided insufficient clinical improvement. All of them needed, in first or second intention, a microsurgical operation by median suboccipital approach to resect or fenestrate the cyst. No endoscopic procedure has been described.

Secondary Chiari Following Cystoperitoneal Shunt (Fig. 44)

Acquired chronic tonsillar herniation as a postoperative complication has been first described during the 1990s, as a consequence of lumboperitoneal shunting or multiple lumbar punctures, performed for pseudotumor cerebri or communicating hydrocephalus. Payner et al. explained this phenomenon by the appearance of a pressure gradient across the cranio-spinal compartments, causing the tonsillar herniation (Payner et al. 1994).

In 1994, Hassounah et al. reported the case of an acquired Chiari malformation, induced by the shunt implantation for a supratentorial arachnoid cyst (Hassounah and Rahm 1994). Then, several other cases were reported (Caldarelli et al. 2009; Di Rocco and Tamburrini 2003; Di Rocco and Velardi 2003; Lazareff et al. 1998; Mendonça et al. 2006; Kim et al. 2002; Martínez-Lage et al. 2009; Osuagwu et al. 2009). This form of acquired Chiari malformation followed early

Fig. 42 (continued) opened and herniation of the cyst through it. (**c**–**k**) Endoscopic procedure. (**d**) View of the inside of the cyst through the outer membrane. (**g**–**k**) Fenestrations of the basal membrane of the cyst. (**l**, **m**) Postoperative axial T2-weighted MRI showing an important decompression of the cyst with a hematoma inside. Diminution of hydrocephalus (less transependymal resorption, enlargement of the cortical sulci)

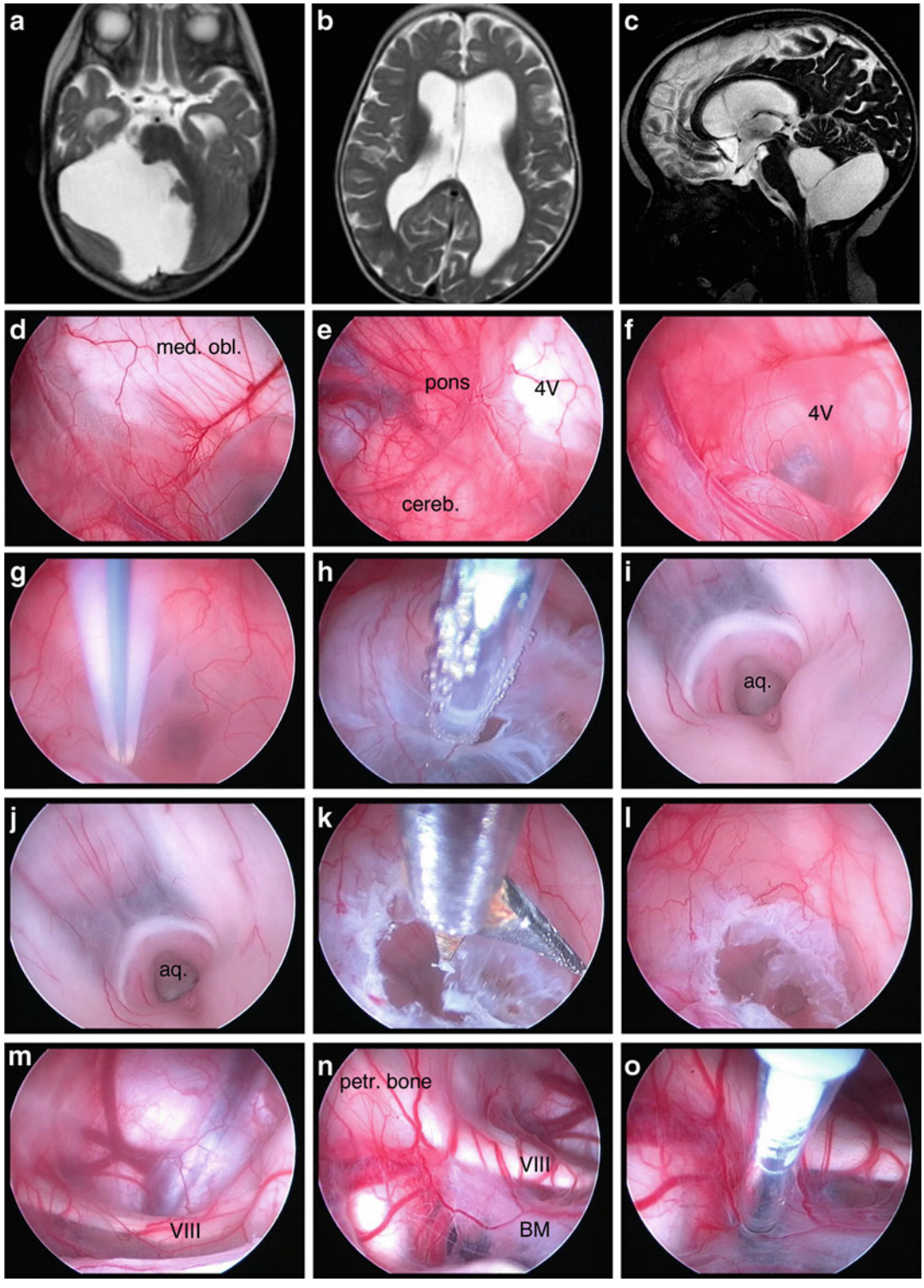


Fig. 43 (continued)

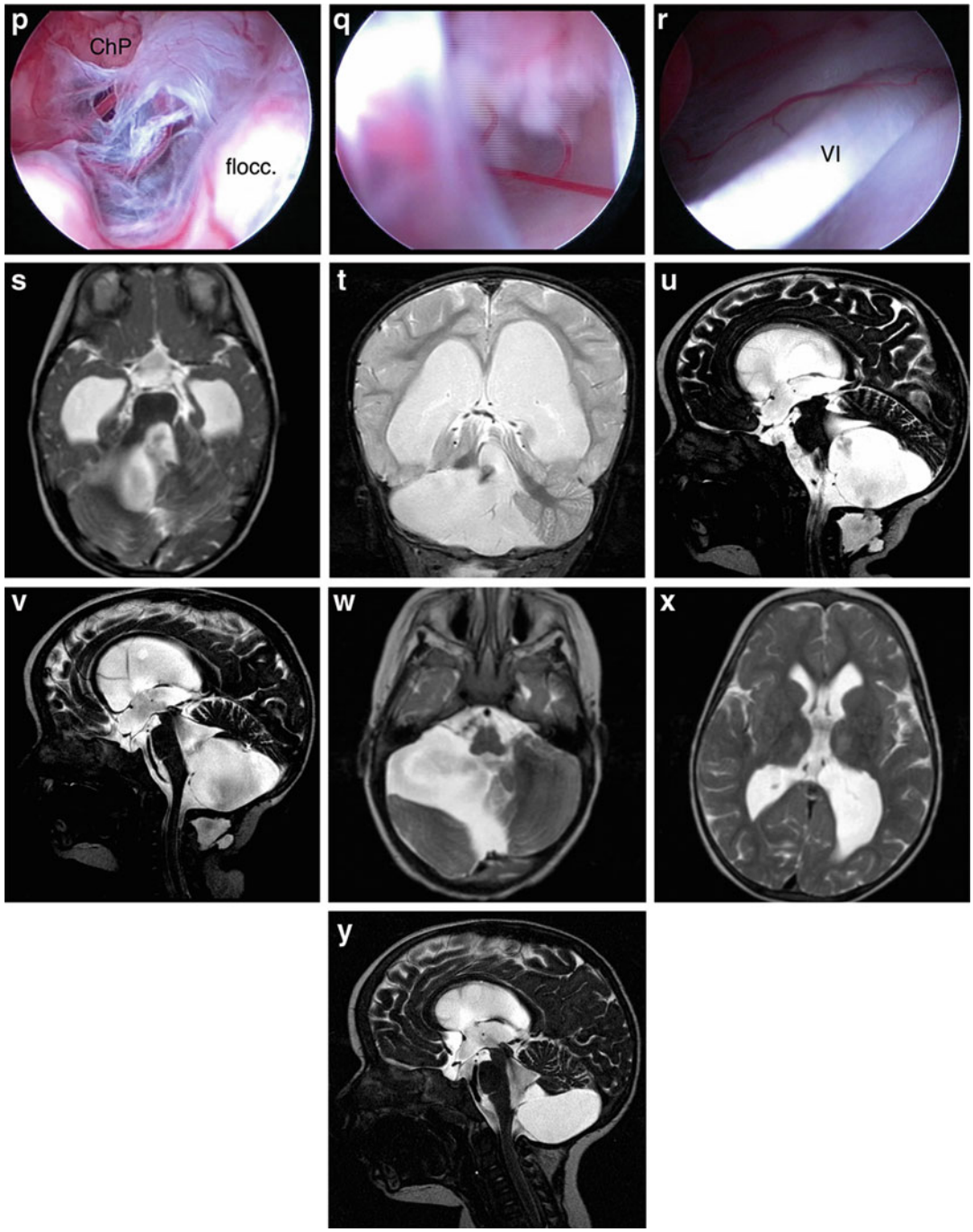


Fig. 43 **Case 2**. Large right lateral infracerebellar arachnoid cyst. (**a**–**c**) T2-weighted MRI. (**a**) Axial section showing the mass effect on the parenchyma. (**b**) Axial section with chronic hydrocephalus. (**c**) Sagittal section showing the infracerebellar location of the cyst with an isolated dilated fourth ventricle. (**d**–**r**) Endoscopic procedure. The patient is in ventral position; the head very flexed. The burr hole is made on the horizontal part of the occipital bone close to the foramen magnum, with a vertical trajectory and superior direction from the virtual cerebellomedullar cistern (upper part of images represents anterior anatomical part, lower part, the posterior part; right side is on the left).

Table 12 Success rate between the three surgical options for posterior fossa arachnoid cysts treatment without isolating hydrocephalus (Gangemi et al. 2011)

Technique	Good outcome (%)
Microsurgery	90
***Cyst* shunting**	86
Endoscopic fenestration	83.3

Good outcome = complete or partial clinical remission

implantation of shunt, either ventriculoperitoneal or cystoperitoneal, for arachnoid cysts. The mean age of the patients in the literature was 6,6 years, the shunt being initially implanted at ages comprised from 2 days to 28 years (Martínez-Lage et al. 2009). According to this review, the interval to the appearance of tonsillar herniation was comprised between 6 months and 10 years (mean age 4,5 years) after shunt implantation.

Same findings have been seen following shunt treatment of Dandy-Walker malformation.

Embryologically, the posterior fossa originates by endochondral ossification of the cartilaginous scaffold that forms the cranial base as well as the first four somites that model the occipital bone.

The pathogenesis of this acquired malformation could have different explanations according to the authors. Most of them suppose a modification and reduction of skull growth induced by CSF shunting.

As said above, in section "slit cyst/ventricle syndrome," the overdrainage in arachnoid cysts shunting seems to induce an intracranial CSF hypotension. Martínez-Lage et al. describe the following process, with an early enlargement of the ventricles, which are displaced together with the brain toward the cysts as CSF is drained away (Martínez-Lage et al. 2009). The venous system becomes distended and engorged, leading to brain edema, meningeal swelling, and at term to dural and suture fibrosis. Probably the absence of pulse pressure of the brain and CSF within the cisterns, because of the drainage, leads to increase of this pathophysiological mechanism. The skull bones become thicker with paranasal sinuses expansion. Although these skull modifications are uniformly distributed, they seem to be prominent in the posterior fossa, finally inducing cranioencephalic disproportion, tonsillar herniation, and displacement of the superior vermis into the great vein of Galen cistern or even herniating through the tentorial notch toward the supratentorial space. Mendonça et al. think that shunting causes anyway an alteration of CSF flow dynamic, impairing the mechanism involved in the brain development (Mendonça et al. 2006). Figure 44 illustrates chronic tonsillar herniation following the treatment of a left temporal arachnoid cyst by shunting.

Clinical manifestations are as various as headaches, signs of raised ICP (nausea, vomiting, papilledema, sixth nerve palsy, somnolence/coma), dizziness, and syncope. Before considering any treatment of this induced pathology, one has to carefully evaluate the pathogenesis of each case. Di Rocco et al. realized prolonged ICP recordings without showing significant ICP increasing for most of the patients (Di Rocco and Velardi 2003).

Various managements have been described. Some authors proposed to increase the level of CSF drainage, adding a programmable or antisiphon valve to a valveless CP shunt. Others proposed to enlarge the skull capacity, which was the first option

Fig. 43 (continued) (**d**, **e**) Views of the tegmentum of the pons and of the medulla oblongata; the right cerebellar hemisphere is pushed upward. (**f**–**h**) Fenestration of the cystic fourth ventricle (cystoventriculostomy). (**i**, **j**) Views in the axis of the aqueduct, with the tectum of the midbrain posteriorly and the tegmentum anteriorly. (**k**, **l**) Enlargement of the stoma. (**m**, **n**) View of the vestibulocochlear nerve from its origin up to the petrous bone. (**o**) Cysto-cisternal fenestration through the basal membrane of the cyst. (**p**) View of the choroid plexus through the lateral foramen of Luschka. (**q**, **r**) View of the pontine cistern and the abducens nerve (VI) entering into the Dorello's canal. (**s**–**v**) Early postoperative MRI showing mostly artifacts of flows through the cystoventricular fenestration. Note the presence of a small non-compressive submuscular meningocele posteriorly to the foramen magnum. (**s**) Axial T2-weighted MRI. (**t**) Coronal T2-weighted MRI. (**u**, **v**) Sagittal CISS-weighted sections showing artifacts of flows through the cystoventricular stoma and through the aqueduct. (**w**–**y**) 1-year postoperative MRI showing better decompression and diminution of hydrocephalus. The stoma is still permeable. (**w**, **x**) T2-weighted axial sections. (**y**) Sagittal CISS-weighted showing a better flow through the aqueduct

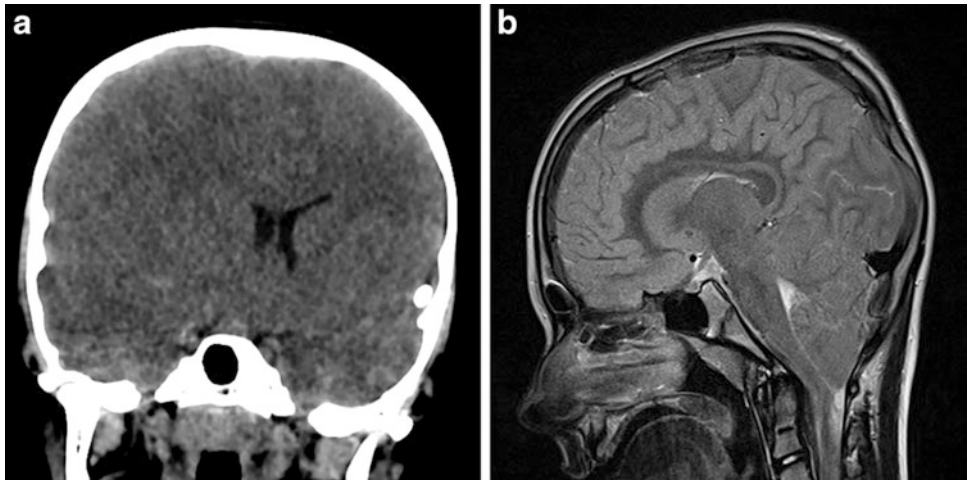


Fig. 44 Same images as Fig. 22 showing a slit cyst syndrome together to a secondary chronic tonsillar herniation in a patient treated of left temporal arachnoid cyst by cystoperitoneal shunt. (**a**) Coronal section of a computed tomodensitometry showing the slit cyst. (**b**) Sagittal section of a T2-weighted MRI showing the induced chronic tonsillar herniation. Note the cranioencephalic disproportion with a small posterior fossa and large cerebellum, virtual subarachnoid spaces

in case of severe symptomatic tonsillar herniation. Two possible surgical enlargements are feasible: foramen magnum decompression (Kim et al. 2002; Mendonça et al. 2006), or supratentorial cranial enlarging procedures (Caldarelli et al. 2009; Di Rocco and Velardi 2003; Martínez-Lage et al. 2006). Supporters of the supratentorial cranial enlargement claim that it treats the causal problem of hindbrain herniation, which is the cranioencephalic disproportion. Some others preferred to place a lumboperitoneal shunt, with the reason that this device drains preferably the subarachnoid spaces. But the initial reason was to treat a slit cyst syndrome.

There is a benefit/risk balance to keep intact when treating an arachnoid cyst, particularly by shunting procedures.

Interhemispheric Cyst

Arachnoid cysts of the interhemispheric fissure may represent an extension of preexisting hydrocephalus and can be considered as congenital malformations, especially in cases of association with corpus callosum dysgenesis (Mori 1992; Martínez-Lage et al. 2011; Caldarelli and Di Rocco 1996; Cinalli et al. 2006). One can also find gyral abnormality, glial heterotopia, holoprosencephaly, and encephalocele as differential diagnoses. They usually are diagnosed during childhood with a head enlargement, sometimes asymmetrical, seizures, and psychomotor retardation. Neuroepithelial cysts are often associated with polymicrogyria and neuronal heterotopia. These associated anomalies might be helpful in preoperative differentiation of neuroepithelial or glioependymal cysts from arachnoid cysts. Ependymal cysts may be intra-axial or extra-axial in location. Gross psychomotor retardation has been reported in many patients with such an anomaly.

They are distinguished from other arachnoid cysts by lack of direct communication with the ventricles, except in cases of associated callosal agenesis.

Pathophysiology of Hydrocephalus

Interhemispheric arachnoid cysts may distort the CSF pathways by displacement downward of one or both the foramina of Monro, obstructing the aqueduct. This may result in a mono- or biventricular dilatation. Because of the location of the cyst, the ventricular dilatation is

Table 13 Classification of interhemispheric arachnoid cysts according to Mori (1992)

Types	Subtypes	Associated malformations	Relationship with the ventricles
Extra-axial	Unilateral parasagittal cysts (Fig. 45)	None	No communication
	Midline cysts (bilateral extension)	Intact corpus callosum	Possible mass effect
		Partial/total agenesis of corpus callosum	
Intra-axial	(Figures 46 and 47)	Dorsal cyst of holoprosencephaly	Communication
		Diencephalic cyst (upward extension of the third ventricle). Porencephalic cyst	

asymmetrical and marked in the body and occipital horn. The frontal horn is compressed and displaced laterally. The second possible explanation of associated hydrocephalus may be the impairment of the resorption mechanism over the convexity (Spennato et al. 2013).

Subtypes

In the literature, the most interesting classification, based on histological discoveries, clinicoradiological presentation, associated congenital anomalies, and prognosis, is the one of Mori (1992) (Table 13). Histologically, he considered interhemispheric cysts as arachnoid cysts but also and more rarely neuroepithelial or glioependymal cysts.

Radiology (Figs. 45, 46, and 47)

Imaging should first exclude other interhemispheric cysts such as the severe holoprosencephaly (Young et al. 1992; Lena et al. 1995). Lena et al. reported antenatal diagnosis by echography (Lena et al. 1995; Vergani et al. 1999).

Preoperative DRIVE and CSF flow sequences on MRI allow the detection of the thinner point of the cyst wall, where the stoma should be done, with minimal brain damage (Cinalli et al. 2006; Spennato et al. 2013).

Treatment/Surgical Techniques

Cysts in this location may or may not communicate with the ventricles, depending of the association of callosal agenesis, holoprosencephaly, encephalocele, gyral abnormalities, and glial heterotopia (Mori 1992; Caldarelli and Di Rocco 1996; Cinalli et al. 2006; Lena et al. 1995; Spennato et al. 2013). Their relationship with the ventricular system and subarachnoid spaces makes the choice of fenestration in the foreground, also if most studies treated about shunting management (Mori 1992; Lena et al. 1995; Caldarelli and Di Rocco 1996; Spennato et al. 2013).

Microsurgical Procedure

Caldarelli and Di Rocco reported 25 children with interhemispheric arachnoid cysts, 14 of them diagnosed prenatally (Caldarelli and Di Rocco 1996). Sixteen were treated by microsurgical excision of the wall associated to fenestration through the midline cisterns or the ventricles. Six benefited from cystoperitoneal shunting, and three were treated by ventriculoperitoneal shunting and cyst fenestration in a second intervention. During the follow-up, 68% of their patients were clinically asymptomatic; the others suffered from psychomotor delay.

Shunting (Fig. 48)

Lena et al. treated 16 patients with interhemispheric arachnoid cysts associated to callosal agenesis by ventriculoperitoneal shunting: "The outcome was good in the large majority of cases" (Lena et al. 1995). Ulu et al. treated seven patients, four of them with callosal agenesis and three with callosal hypogenesis, by cystoperitoneal shunting (Ulu et al. 2010). Three patients had associated hydrocephalus, two diagnosed prenatally, and needed an additional

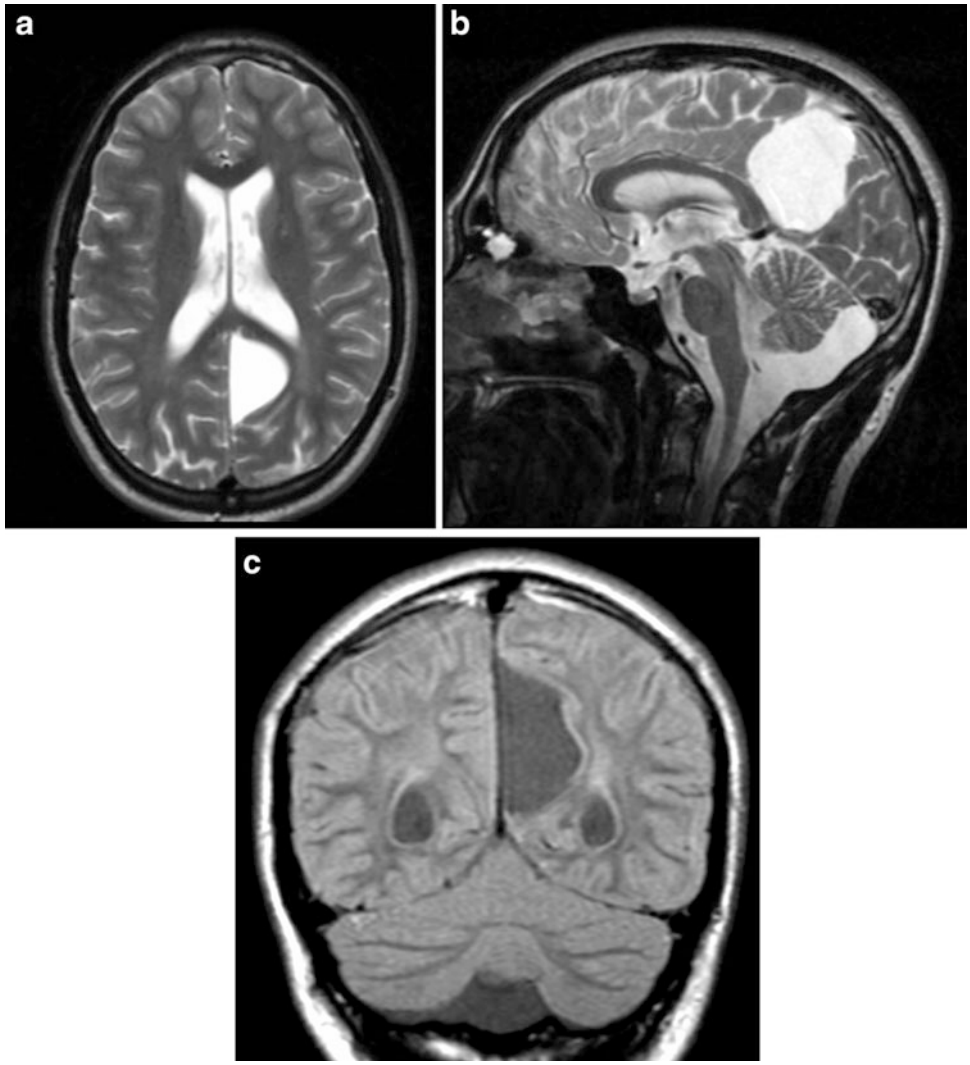


Fig. 45 Small left parasagittal arachnoid cyst on MRI. (**a**) Axial T2-weighted section. (**b**) Sagittal T2-weighted section. (**c**) Coronal T1-weighted section. Note that there is also a median retrocerebellar arachnoid cyst

ventricular catheter, which was connected to the valve by a Y-shaped connector during the same intervention. The clinical outcome was good in 86% of the patients, including the three children with hydrocephalus. The cysts were nearly or totally resolved for 71% and partially for 29% of the children. Figure 48 shows the implantation of a ventriculo-cystoperitoneal shunt on an interhemispheric arachnoid cyst. It illustrates the clinicoradiological appearance of a slit ventricle syndrome after a few years.

Endoscopic Procedure

During the last decade, neuroendoscopic procedures have obtained a prominent place, but only one article talks about neuroendoscopic management of interhemispheric cysts. Cinalli et al. reported seven cases of interhemispheric cysts, including three patients with hydrocephalus, all of them had an endoscopic fenestration from the cyst to the cisterns and/or the ventricles (Cinalli et al. 2006). Six children had agenesis of the corpus callosum and one a hypogenesis. The

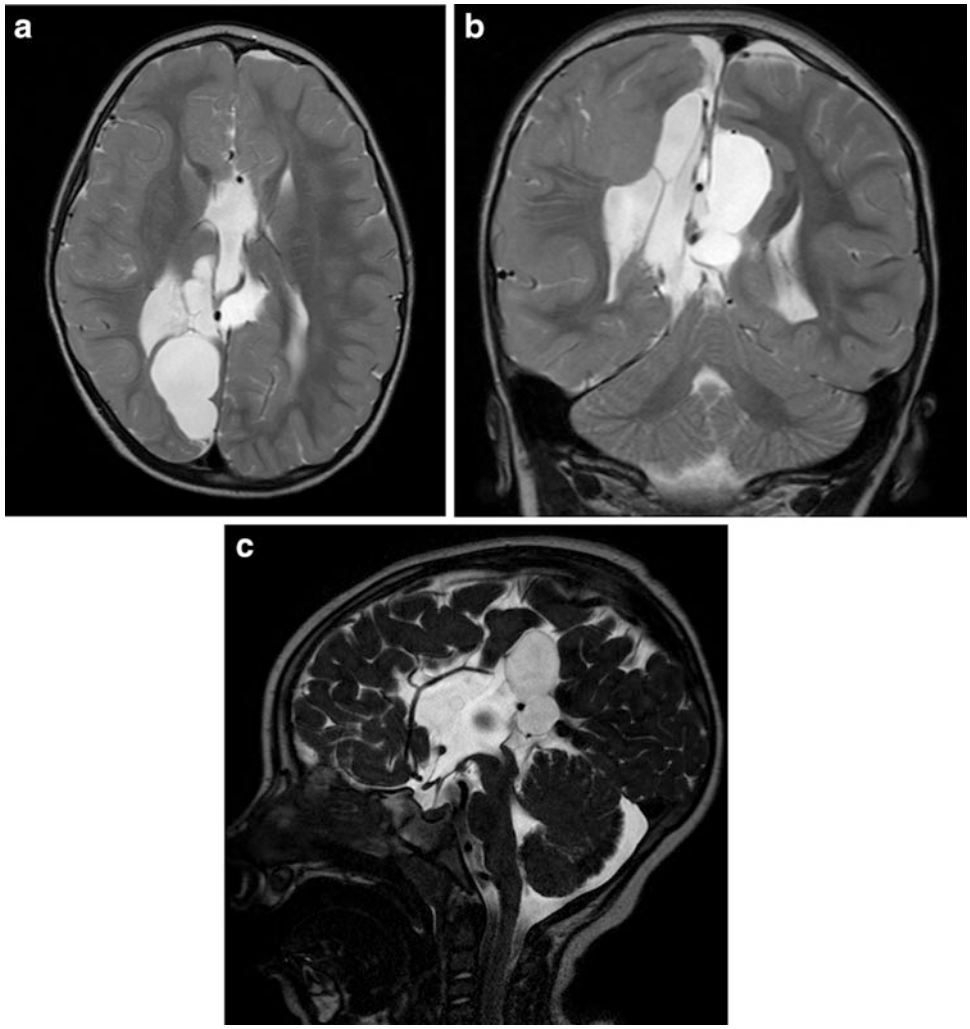


Fig. 46 Intra-axial interhemispheric arachnoid cyst that benefited of from endoscopic multiple fenestrations; MRI. (**a**) T2-weighted axial section. (**b**) T2-weighted coronal section. (**c**) CISS-weighted sagittal section. Note the complete agenesis of corpus callosum and the aspect of continuous third ventricle, which has no roof and is in direct communication with the vertex

success rate was 71%, and the two remaining patients benefited from a second endoscopic fenestration, because of the closure of the initial stoma, and the other one had a lumboperitoneal shunt because of subcutaneous collection of CSF. Three children had a postoperative subdural hygroma, one requiring a subduroperitoneal shunt. Forty-three percent of them had normal psychomotor development and the other had delay. Only two patients had preoperative hydrocephalus, and both had disappearance of ventricle dilatation by reopening of the aqueduct caused by size reduction of the cyst. This allowed the absence of shunt dependency for all patients.

Standard Endoscopic Procedure (Cinalli et al. 2006) (Figs. 49, 50, and 51)

Of course, the very good knowledge of the intraventricular anatomy (choroid plexus especially) and intracisternal nerves and arteries will greatly help for the orientation. However, neuronavigation is highly recommended to find the best placement

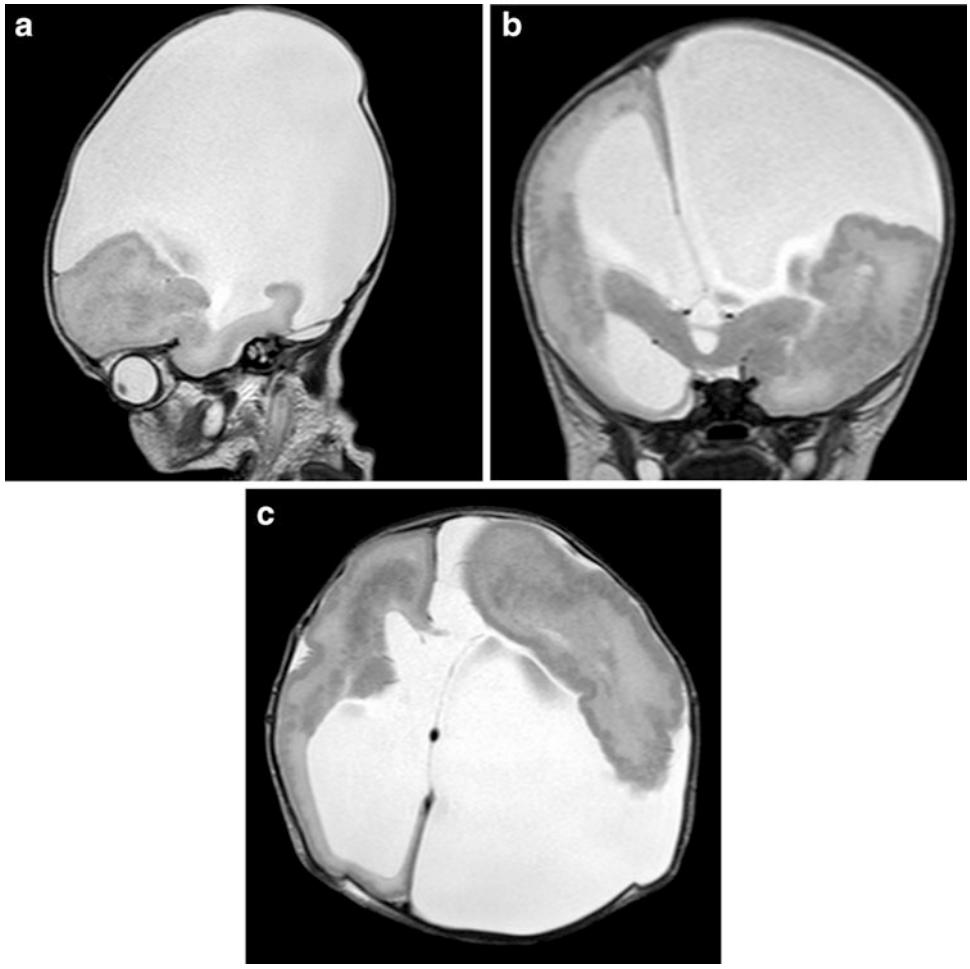


Fig. 47 Huge intra-axial interhemispheric cyst on T2-weighted MRI. (**a**) Sagittal section. (**b**) Coronal section. (**c**) Axial section

of the burr hole to perform the ideal trajectory with the endoscope, according to the cyst's characteristics in size and location, the size of the ventricles, and the relationships with the third ventricle. The objective is the communication between the cyst and ventricles and/or the subarachnoid spaces of the basal cisterns (quadrigeminal, interhemispheric, prepontine cisterns). Usually the entrance, first into the cyst, is easier, using a paramedian burr hole (the anteroposterior placement depends on the preoperative planning). Then the ventricular wall is recognized with the help of the neuronavigation system and perforated, thus achieving the realization of a cystoventriculostomy. If the size of the ventricular system permits, one can perform ventriculocisternostomy, through the third ventricle into the interpeduncular cistern (third ventriculostomy). If fenestration with the ventricles is impossible or risky, one can perform a cystocisternostomy, toward the interhemispheric or quadrigeminal cisterns. Figures 49, 50 and 51 illustrate the endoscopic procedure performed on a left frontoparietal parasagittal arachnoid cyst (Fig. 49), a left parasagittal arachnoid cyst associated to partial corpus callosum agenesis (Fig. 50), and an extra-axial arachnoid cyst of the midline associated to complete agenesis of the corpus callosum (Fig. 51).

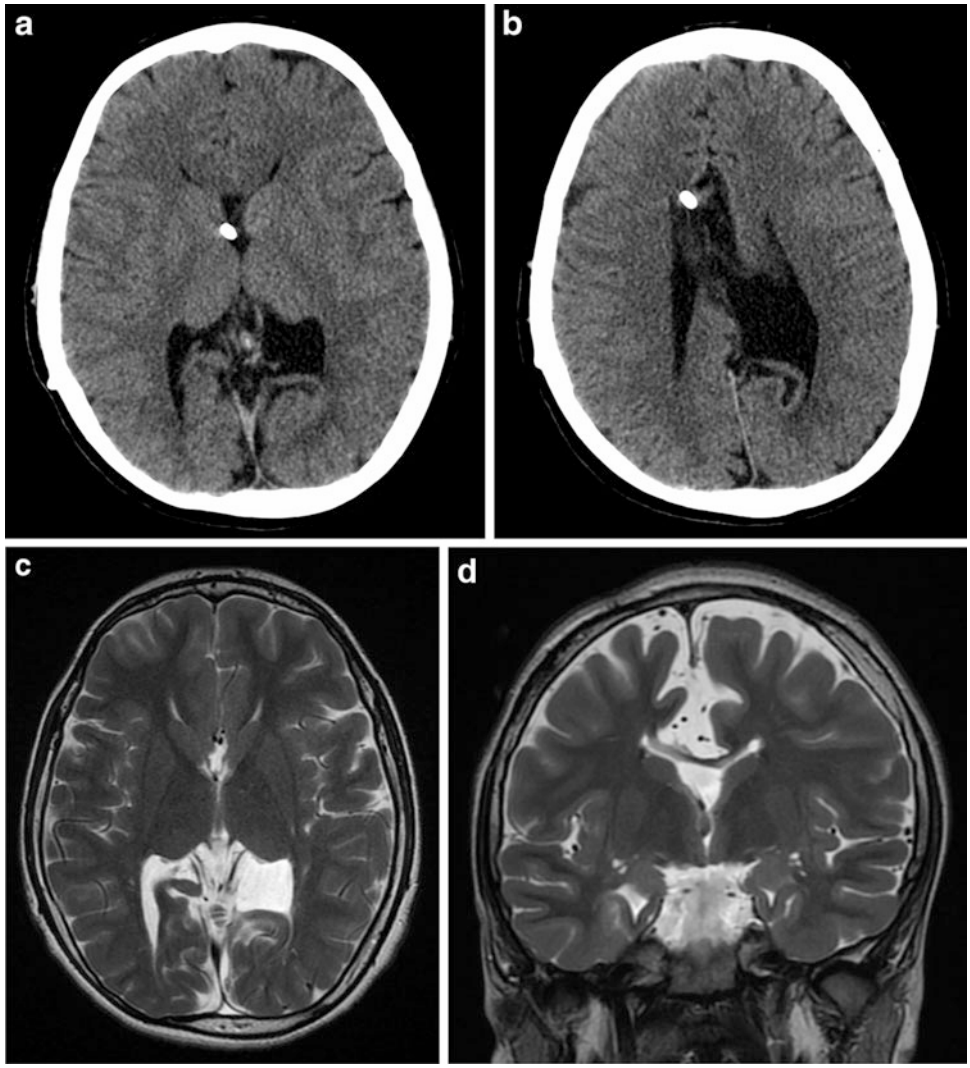


Fig. 48 Interhemispheric arachnoid cyst, treated by a ventriculo-cystoperitoneal shunt years ago. Currently the patient presented a slit cyst/ventricle syndrome with intracranial hypertension symptoms (headache, visual loss, and papilledema). (**a**, **b**) Axial sections of computed tomodensitometry showing collapse of the ventricles around the catheter. (**c**, **d)** T2-weighted axial and coronal MRI sections

Discussion on Treatment

In 1992, Mori advocated for a combined surgery for patients with associated hydrocephalus, microsurgical large fenestration and excision of cyst's wall, and placement of a ventriculo-peritoneal shunt (Mori 1992).

In 2011, Gangemi et al. have performed a comprehensive review of 61 reports in the literature comparing the clinicoradiological outcome of the 3 surgical procedures in the treatment of 44 interhemispheric together with convexity arachnoid cysts (Gangemi et al. 2011) (Table 14).

Intraventricular Cyst

Intraventricular arachnoid cysts are, by definition, developed from a ventricle and should be differentiated from other arachnoid cysts that have an extension through a ventricle, such as

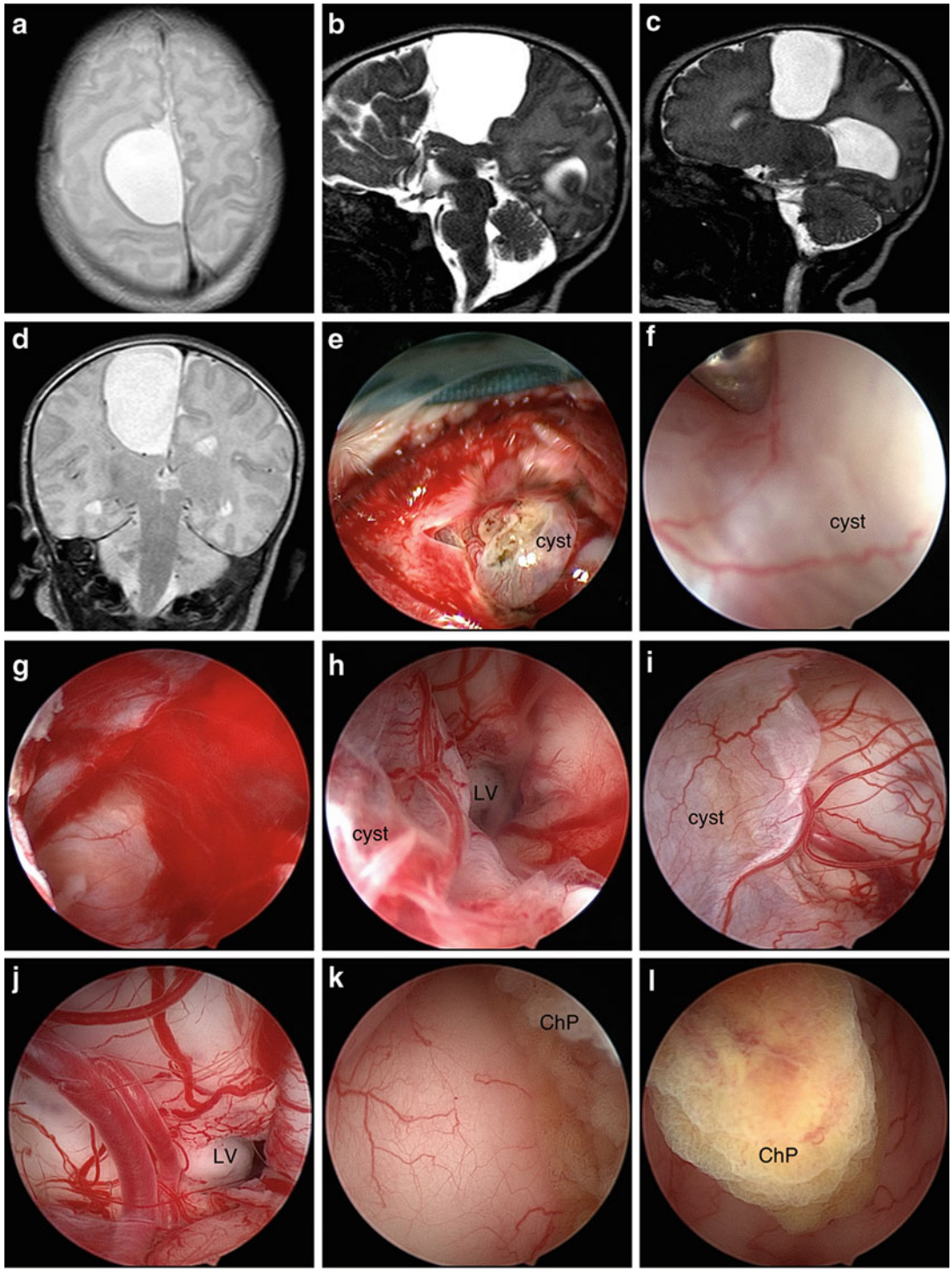


Fig. 49 (continued)

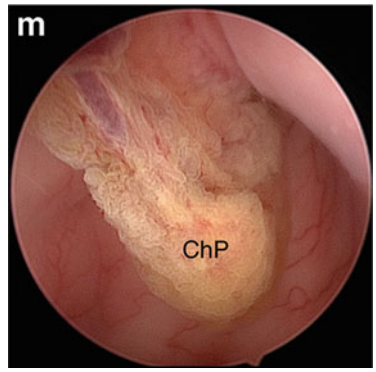


Fig. 49 **Case 1**. Left frontoparietal parasagittal arachnoid cyst of a premature newborn. (**a**) Axial T2-weighted MRI. (**b**) Sagittal CISS-weighted MRI showing the intact corpus callosum. (**c**) Parasagittal CISS-weighted MRI showing a trapped left occipital ventricular horn. (**d**) Coronal T2-weighted MRI. (**e**–**m**) Endoscopic procedure for cystoventricular fenestration. (**e**) The patient is supine; the head is slightly flexed. Left parasagittal burr hole with hernia of the upper wall of the cyst. (**f**) Fenestration of the upper wall. (**g**) View from inside the cyst, noting a small intracystic hemorrhage. (**h**, **i**) Peeling of the external wall of the cyst by rinsing with physiological serum outside the cyst. (**j**) This opens spontaneously the left lateral ventricle. (**k**–**m**) View from the entry point of the left ventricular atrium with its choroid plexus. The occipital horn (not seen) is posterior to us

suprasellar cysts within the third ventricle and quadrigeminal and interhemispheric cysts toward a lateral ventricle. Arachnoid cysts developed from the fourth ventricle have also been described. Often, in the literature, these cysts are not really individualized and are often associated to the ones cited above. Most of them are case reports, so a consensual therapeutic management is difficult to extract.

Differential diagnoses of intraventricular arachnoid cyst are posttraumatic, hemorrhagic, and infectious cysts. Some authors consider cysts of the choroid plexus as arachnoid cysts because of histological findings (Nakase et al. 1988; Pelletier et al. 1990; Shou et al. 2015). According to these authors, during the formation of arachnoid cysts, the mesenchyme in the blood vessels probably folds into the cavity of the neighboring ventricle, such as choroid fissure, covered by a layer of glial and ependymal cells (Shou et al. 2015). Histological examination confirms that the top of the cyst wall is often so thin that there is no more glial or ependymal tissue, allowing its safe fenestration.

Pathophysiology of Hydrocephalus

Intraventricular arachnoid cysts may arise from the displacement of arachnoid cells by the vascular mesenchyme, through the choroid fissure, during the process of choroid plexus development (Martinez-Lage et al. 1992). The mechanism of hydrocephalus is probably due to obstruction of the lateral ventricle with mono- or biventricular dilatation, or the third ventricle with a biventricular hydrocephalus, and to accumulation within the cyst of CSF merging around the plexus vessels (Juan F. Martínez-Lage et al. 2011).

Subtypes

There is no classification in the literature based on clinical presentation or extension as other arachnoid cysts, but we can simply classify them according to the ventricle wherein they grow, as:

- *Lateral ventricle*: often developed in the trigone area, the body of the ventricle, temporal or occipital horn
- *Third ventricle*
- *Fourth ventricle* (treated in the section "Posterior Fossa Cysts")

Radiology (Fig. 52)

CISS sequences allow visualization and delimitation of intraventricular cysts and their relationships with the ventricular walls. CSF flow

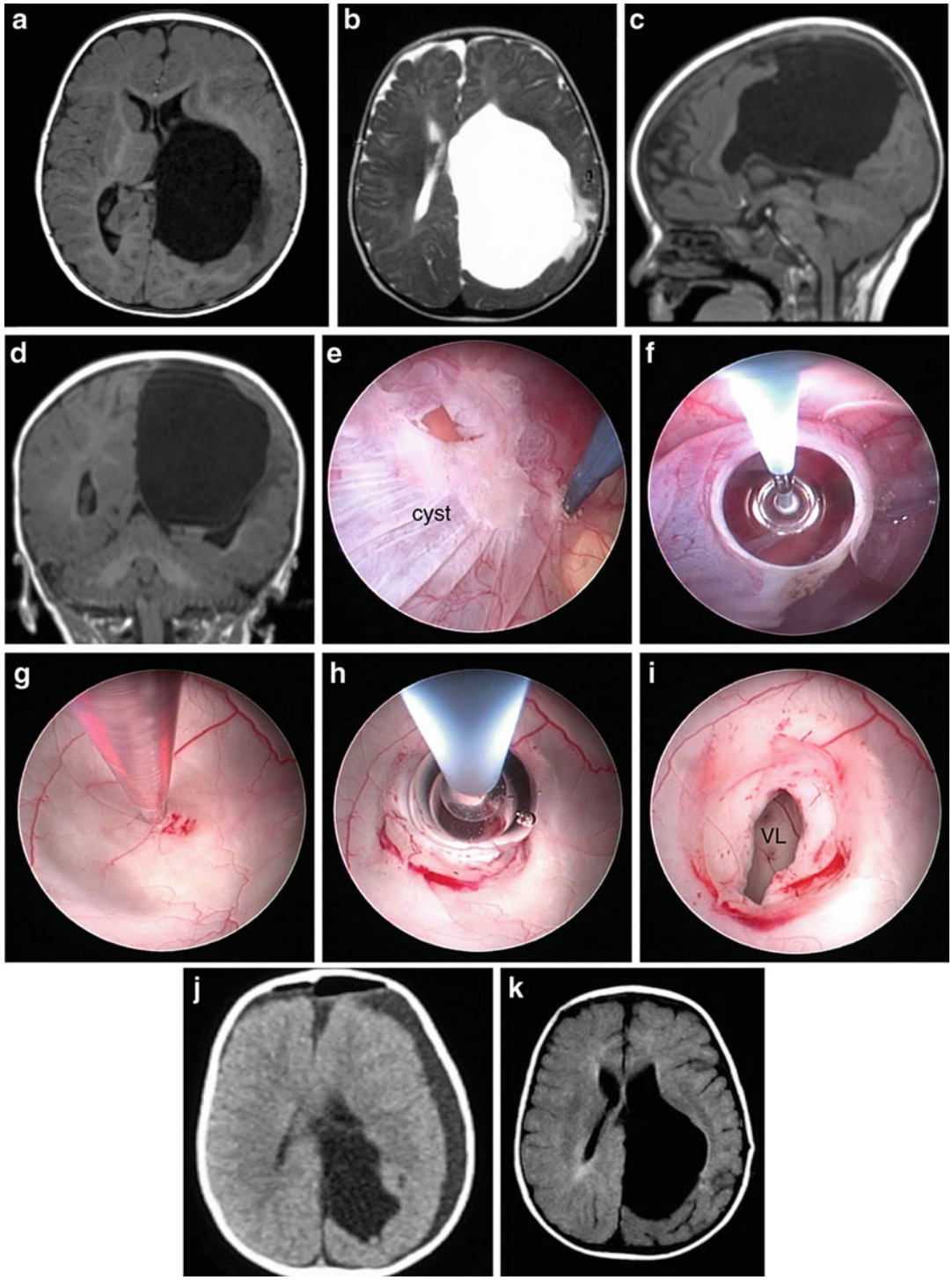


Fig. 50 **Case 2**. Left parasagittal arachnoid cyst with partial agenesis of the corpus callosum. (**a**, **b**) T1- and T2-weighted axial MRI sections. (**c**, **d**) Sagittal and coronal T1-weighted sections showing the upper compression of the ventricle (third and lateral) by the cyst. (**e**–**i**) Endoscopic procedure for cystoventricular fenestration. (**e**, **f**) Upper view from outside the external wall of the cyst, which has been fenestrated and retracted. (**g**–**i**) View of an unfolded parenchyma from inside the cyst that is fenestrated to the third ventricle. (**j**) Early

sequences may be useful during the preoperative planning to diagnose a non-communicating cyst.

Treatment/Surgical Techniques

Historically, intraventricular arachnoid cysts have been treated by microsurgical fenestration and/or cystoperitoneal shunt procedures.

In 1992, Martínez-Lage et al. published three cases of patients with intraventricular arachnoid cysts. One of them was treated by craniotomy with cyst excision associated to a cystoperitoneal shunt, one was treated by endoscopic fenestration and catheter placement, and the last one was treated with a single cystoperitoneal shunt (Martínez-Lage et al. 1992).

The largest series of purely intraventricular arachnoid cysts with retrospective compared study of the different techniques is the one of Zhao et al. with 39 patients with microsurgical resection and 28 with endoscopic fenestration or resection (Zhao et al. 2013). They reported, for these lateral ventricle cysts, a success rate (clinical remission or improvement) in 93% of neuroendoscopic procedures, versus 67% for the microsurgical group. They found also significantly less blood loss, operative time, and complications in the endoscopic branch, with a recurrence rate of 0% versus 21% in the group with craniotomy. The total resection rate followed the same values that the clinical success rate has.

Microsurgical Procedure

Microsurgery for resection of intraventricular arachnoid cysts consists of a craniotomy with a transcortical or sometimes interhemispheric transcallosal approach. The main problem with this technique for deep-seated lesions is the high morbidity rate because of the necessity of several manipulations through the operative channel and parenchymal injuries, with also a worse visualization. According to the literature with mainly case reports, the clinicoradiological success rate of open surgery procedures is included between 54% and 96% but with a non-negligible rate of complications (up to 30%) and rate of reinterventions.

Shunting

Single cystoperitoneal shunting or ventriculoperitoneal shunting seems to cause an expansion of the other undrained compartment. There is the same rate of malfunction (40%) as other shunt procedures (Zhao et al. 2013).

Endoscopic Procedure

The second largest series of purely intraventricular arachnoid cysts is the one of Shou et al. with 21 patients, including 11 children (Shou et al. 2015). All of the patients were treated by endoscopic procedure guided by a neuronavigation system, allowing a cystoventricular fenestration in most of the cases or cyst resection in the others. The clinical success rate was 100% and the ventricle size was smaller in all cases. There was no difference of outcome between a total resection and a single large fenestration. Seven histopathological diagnoses were done in this series, and two of them found a choroid plexus cyst.

Standard Endoscopic Procedure (Shou et al. 2015) (Figs. 53, 54)

The advantage of procedures within the ventricles is the capacity for the operator to keep his landmarks thanks to the normal intraventricular anatomical structures. But, when an arachnoid cyst grows inside, this helpful anatomy may be distorted or absent.

The neuronavigation system should be used to define the optimal entry point and trajectory. Cysts in the ventricular atrium can be approached by a trans-trigone approach; those of the body of the lateral ventricle can be joined by a transfrontal

Fig. 50 (continued) postoperative axial section of a computed tomodensitometry, showing an important cyst decompression and a hemispheric hygroma. (**k**) Axial computed tomodensitometry performed 1 year later, showing a satisfying balance with absence of compression or hydrocephalus. Note that the cortical sulci are well seen

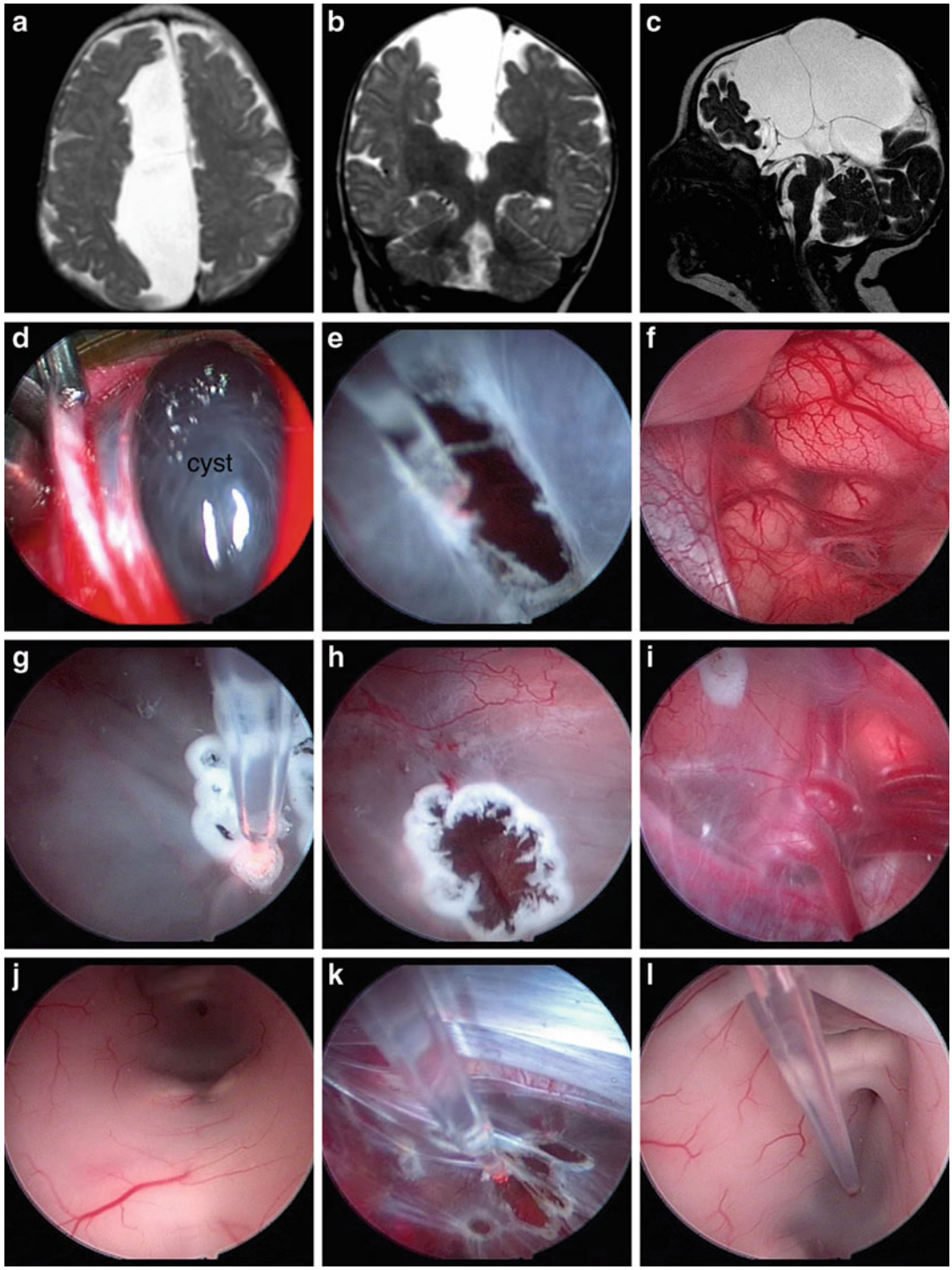


Fig. 51 (continued)

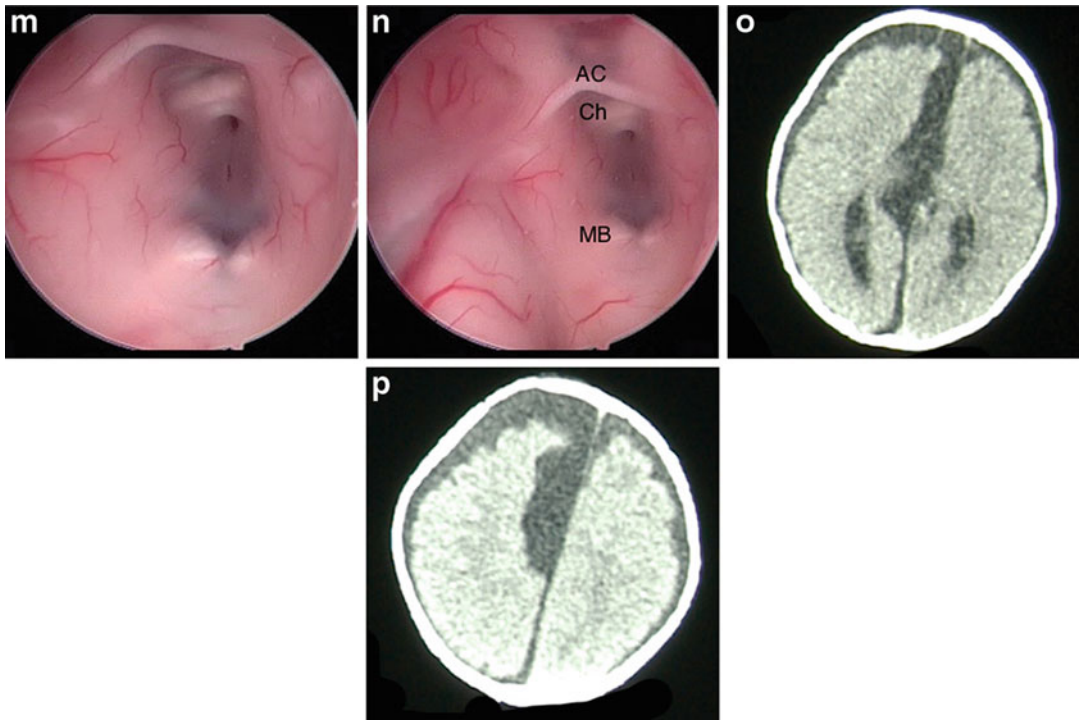


Fig. 51 **Case 3**. Extra-axial arachnoid cyst of the midline associated to complete corpus callosum agenesis. (**a**) T2-weighted axial MRI. (**b**) T2-weighted on coronal section showing the symmetrical extension of the cyst, compressing downward the third ventricle. (**c**) Sagittal CISS-weighted section showing the polylobed character of the cyst. (**d**–**n**) Endoscopic procedure for cystoventricular fenestrations and third ventriculostomy. (**d**) The patient is positioned supine and the head slightly flexed. The entry point is the right lateral angle of the fontanel. (**d**, **e**) Extracranial view of the burr hole and the herniated external wall of the cyst that is opened. (**f**) Upper view of the brain parenchyma from inside the cyst. Anteriorly we can see the middle intracystic septum (seen on (**c**)). (**g**, **h**) Fenestration of the inner wall of the cyst, being the postero-superior roof of the third ventricle (on (c)). (**i**) Penetration through the stoma, showing the venous confluent to the great cerebral vein in front of the free edge of the falx. (**j**) Posterior view of the third ventricle. (**k**) Second fenestration anteriorly on the roof of the third ventricle. (**l**–**n**) Postero-superior view of the third ventricle and third ventriculostomy. (**o**, **p**) Computed tomodensitometry with axial sections, performed 6 days postoperative, showing a symptomatic bilateral hygroma, with the need of secondary implantation of a subdural-peritoneal shunt

Table 14 Success rate between the three surgical options for interhemispheric together with convexity arachnoid cysts treatment without isolating hydrocephalus (Gangemi et al. 2011)

Technique	Good outcome (%)
Microsurgery	89
***Cyst* shunting**	100
Endoscopic fenestration	75

Good outcome = complete or partial clinical remission

horn approach. Then a standard burr hole is performed, and the endoscope can be inserted guided by neuronavigation into the ventricle. When using a posterior approach for cysts of the trigone, after entering the ependyma, the small ventricular lumen can collapse, and the whole view of the endoscope can be occupied by the wall of the cyst that is impossible to penetrate with the endoscope itself. Cystoventricular fenestration should then be performed at this point and can be technically challenging because of the lack of working space due to the collapse of the ventricular lumen (Fig. 52e). Two techniques can be used at this time: with the first, withdrawal of 1 cm of the whole endoscope inside the parenchymal track usually allows to better see the cyst wall and offers a small working chamber for instrument manipulation. With the second the

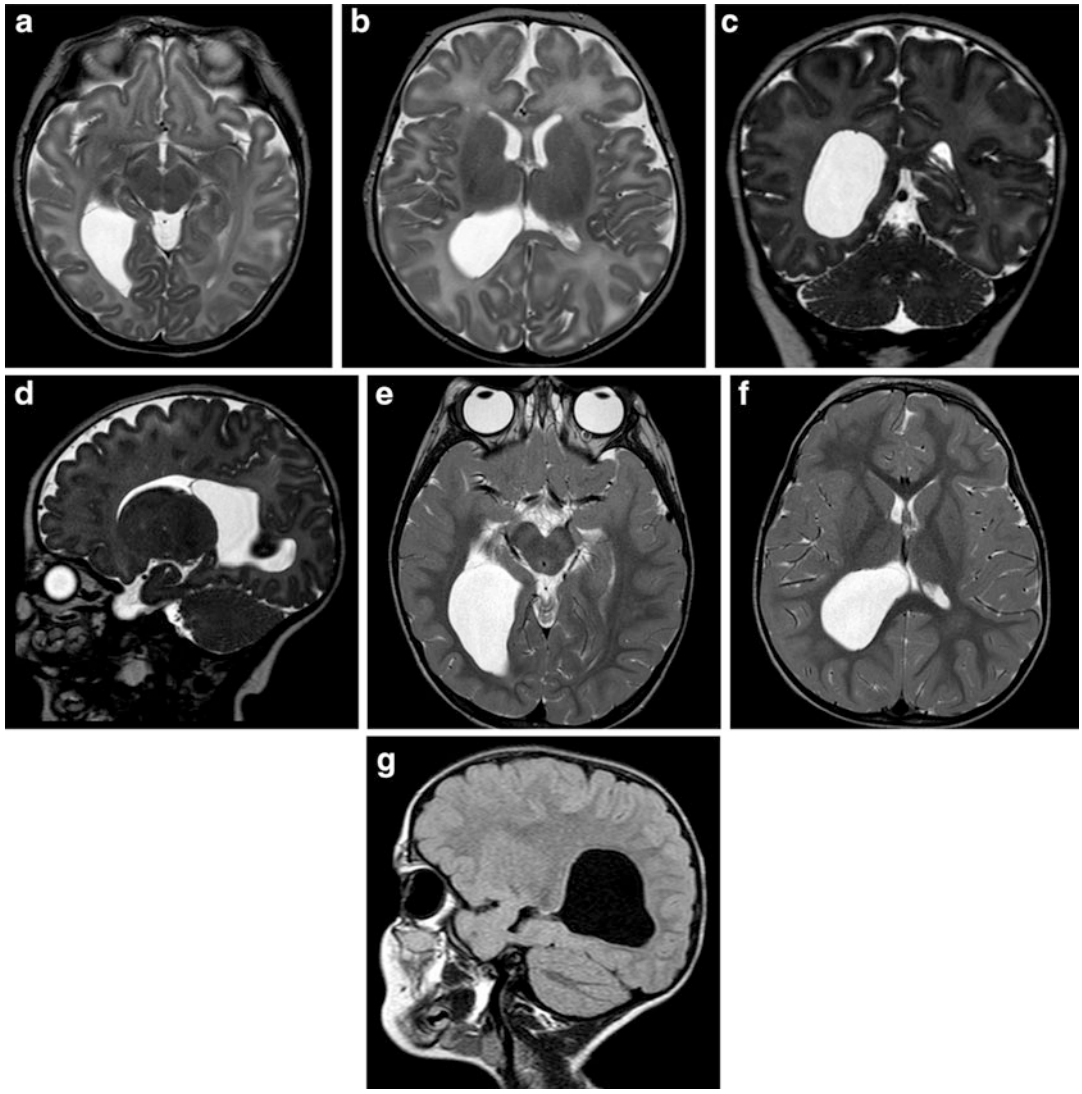


Fig. 52 Growth of an arachnoid cyst of the right ventricular atrium. (**a**–**d**) First MRI showing the cyst. (**a**, **b**) T2-weighted axial sections showing anterior and medial walls of the cyst. (**c**) Coronal CISS-weighted section. (**d**) Sagittal CISS-weighted section showing the anterosuperior wall of the cyst. (**e**–**g**) Second MRI performed 2 years later. (**e, f**) T2-weighted axial sections. (**g**) T1-weighted sagittal section

outer sheath of the endoscope is hold in contact with the cyst wall, while the optic is unlocked from the sheath and withdrawn of few millimeters, creating an artificial working space where instruments can be manipulated to create an opening in the cyst wall. After the first small opening in the cyst wall, the cyst usually collapses and detaches itself from the ependymal wall, allowing to work in a much larger space. As said before, it is better to perforate the top of the cyst because of a thinner and non-gliotic or ependymal layer. The fenestration should be at least from 10 to 15 mm. The deeper wall of the cyst should be fenestrated too, to prevent cyst recurrence or postoperative obstructive hydrocephalus. In case of a non-adherent cyst's wall to the ependyma, one can consider a resection of the cyst. Care should be taken during dissection of the cyst from

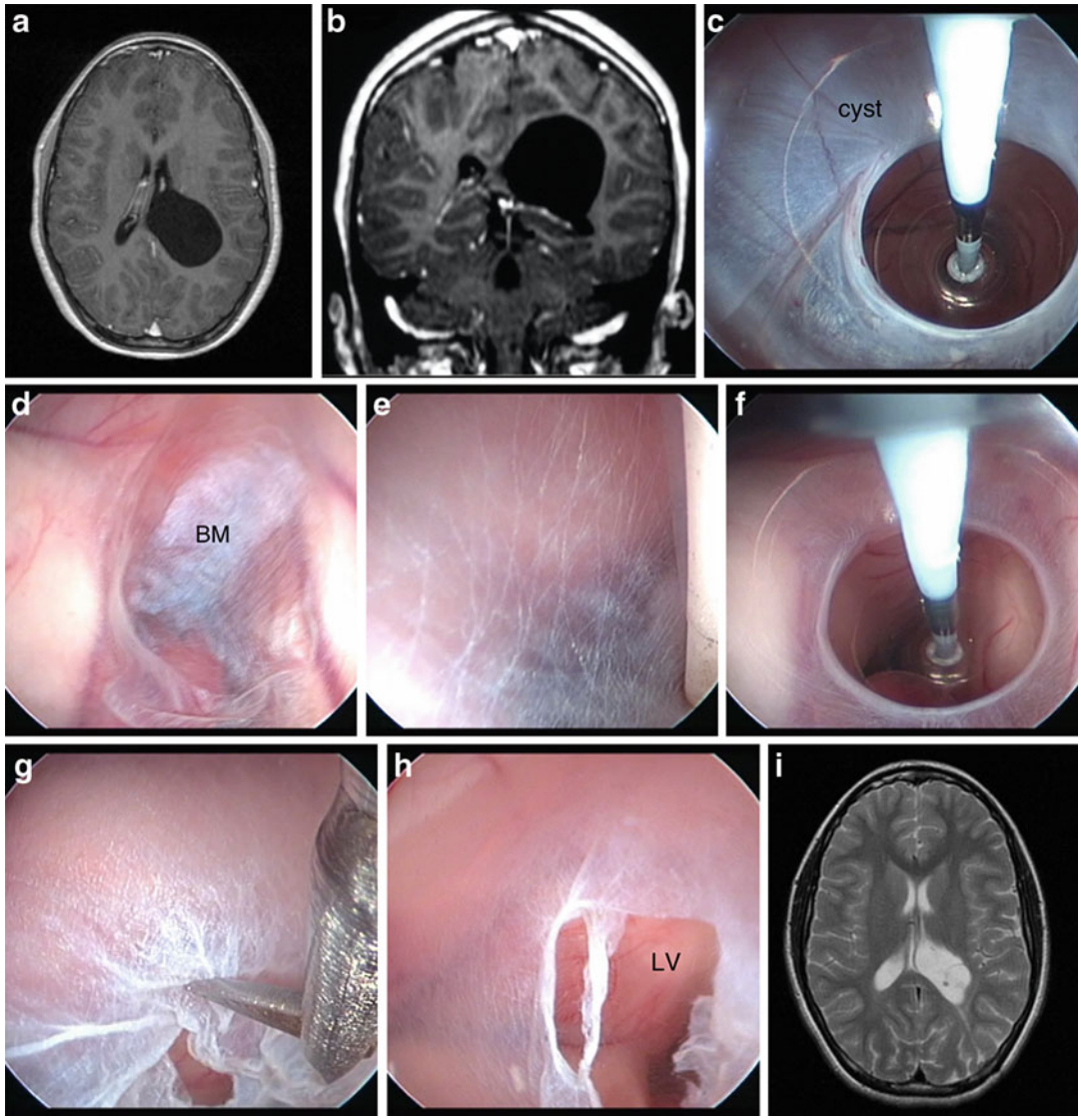
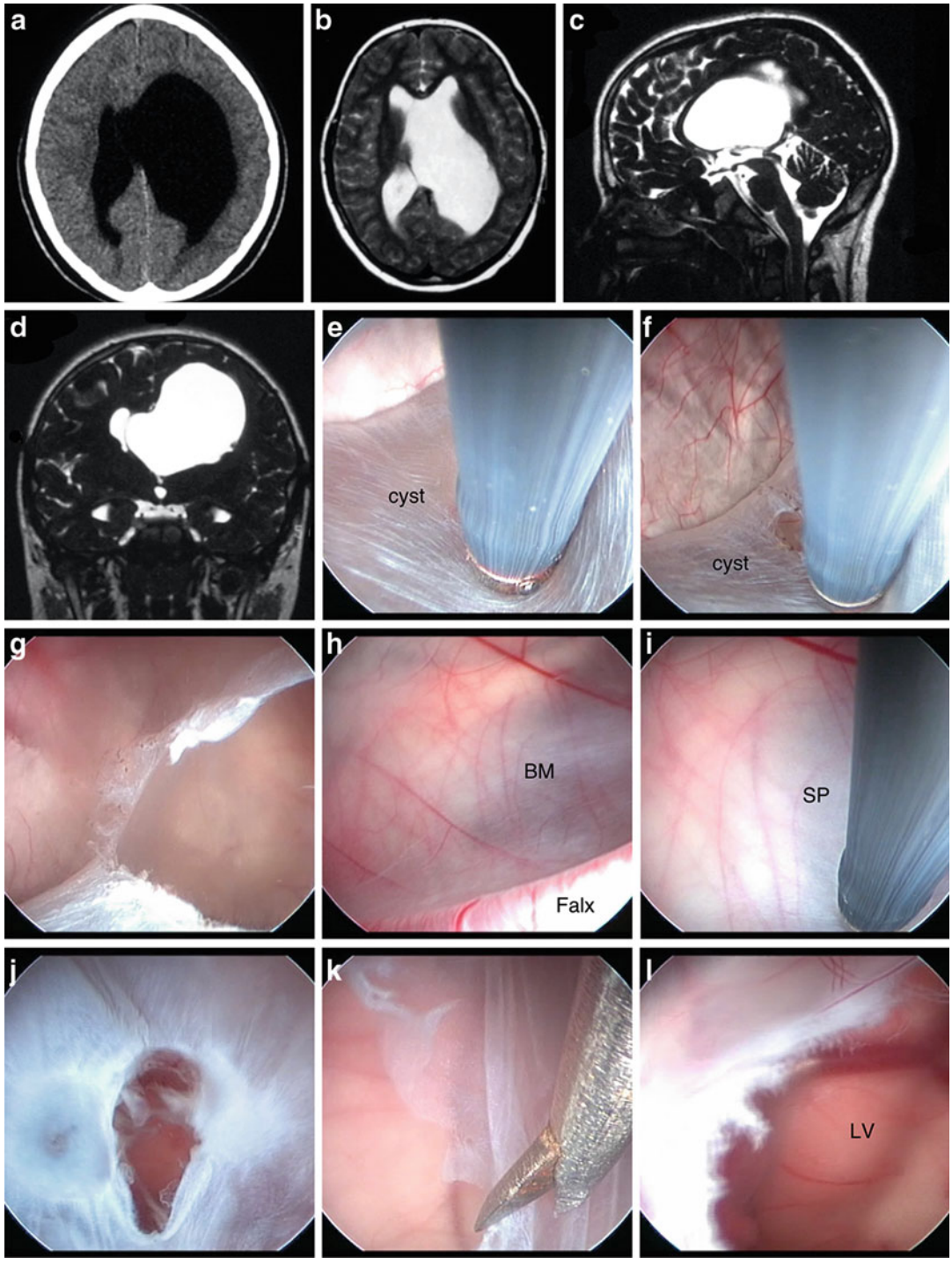


Fig. 53 **Case 1**. Arachnoid cyst of the left ventricular atrium on T1-weighted MRI. (**a**) Axial section. (**b**) Coronal section. (**c**–**h**) Endoscopic procedure consisting of ventriculo-cystoventricular fenestrations. The patient is positioned supine, with the head turned to the right side. The burr hole is made on the parietal bone for kind of the same approach than a classical ventriculoperitoneal shunt. (**c**) Lateral view of the cyst from the parenchymal entry point. Fenestration with the balloon. (**d**) View from inside the cyst of the left frontal horn through the internal wall of the cyst. (**e**–**h**) Cystoventricular fenestration. (**i**) Postoperative T2-weighted axial MRI showing a satisfying cyst decompression inside the atrium

ependyma on the midline and from the choroid plexus, because of the risk of injuring the surrounding vessels. Figures 53 and 54 detail the endoscopic procedure for an arachnoid cyst of the left ventricular atrium (Fig. 53) and a cyst of the left lateral ventricle (Fig. 54).

Discussion on Treatment

In 2013, Zhao et al. compared, in their series, the success rate between microsurgery (39 patients) and neuroendoscopy (28 patients) in the treatment of intraventricular arachnoid cysts (Zhao et al. 2013) (Table 15).



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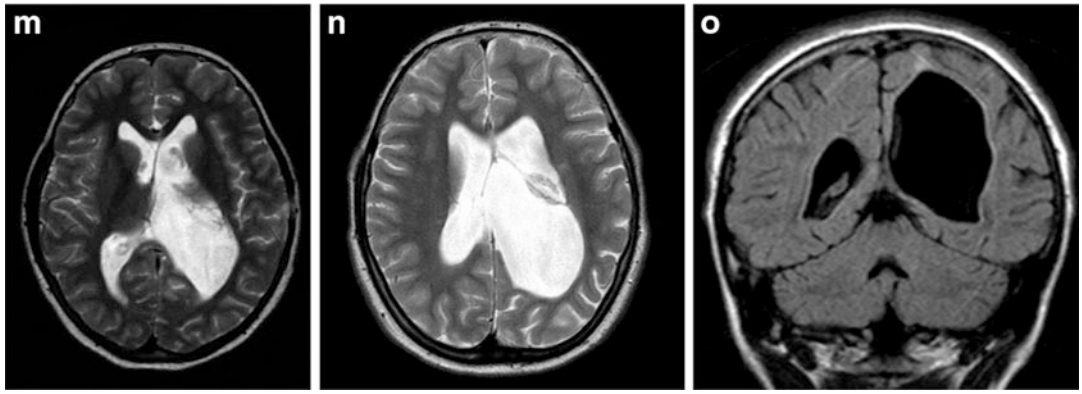


Fig. 54 **Case 2**. Arachnoid cyst of the left lateral ventricle. (**a**, **b**) Axial sections of computed tomodensitometry and T2-weighted MRI showing severe hydrocephalus. (**c**, **d**) Sagittal and coronal CISS-weighted sections showing hernia of the cyst under the falx and the corpus callosum. (**e**–**l**) Endoscopic procedure consisting of maximum cyst wall fenestration and excision and septostomy. (**e**–**g**) Large fenestration and peeling of the lateral wall of the cyst from the ependyma. (**h**) View of the falx and the herniation of the cyst. (**i**–**l**) Cystoventriculostomy of the right ventricle through the horizontal septum pellucidum. (**l**) View of the contralateral ventricle. (**m**) 3 months postoperative axial T2-weighted MRI showing a good decompression and artifacts of flows. (**n**, **o**) 6 months postoperative MRI showing a relaxed cyst wall inside the ventricle. (**n**) T2-weighted axial section. (**o**) T1-weighted coronal section

Table 15 Success rate between microsurgery (39 patients) and neuroendoscopy (28 patients) for intraventricular arachnoid cysts treatment without isolating hydrocephalus, according to Zhao et al.'s series (Zhao et al. 2013)

Technique	Good outcome (%)
Microsurgery	67
Endoscopic fenestration	93

Good outcome = complete or partial clinical remission

Whatever the procedure, the persistence of a ventriculomegaly in certain cases in the literature does not mean a failure of the treatment as the patient becomes asymptomatic.

Intraventricular arachnoid cysts seem to represent a very good indication for endoscopic fenestration because of a simple and safe procedure.

Hydrocephalus and Arachnoid Cyst Within the Third Ventricle (Fig. 55)

Arachnoid cyst of the third ventricle, which does not come from suprasellar or quadrigeminal areas and causing hydrocephalus, is uncommon and fairly described in the literature. Most of them arise from the posterior portion of the third ventricle, eventually growing from the choroid plexus. The most common intraventricular cysts are suprasellar cysts, but they really have to be differentiated because of their embryologic and anatomical origins, their extension and repercussion on surrounding structures such as optic nerves/chiasm, pituitary stalk, and floor of the third ventricle. In fact, the latter one has an important operative consequence, because when performing an endoscopic approach from the lateral to the third ventricle, the wall of the cyst is made of an arachnoid cell layer, and not by the floor of the third ventricle as this is the case with suprasellar cysts. Figure 55 shows the endoscopic procedure for an arachnoid cyst of the third ventricle.

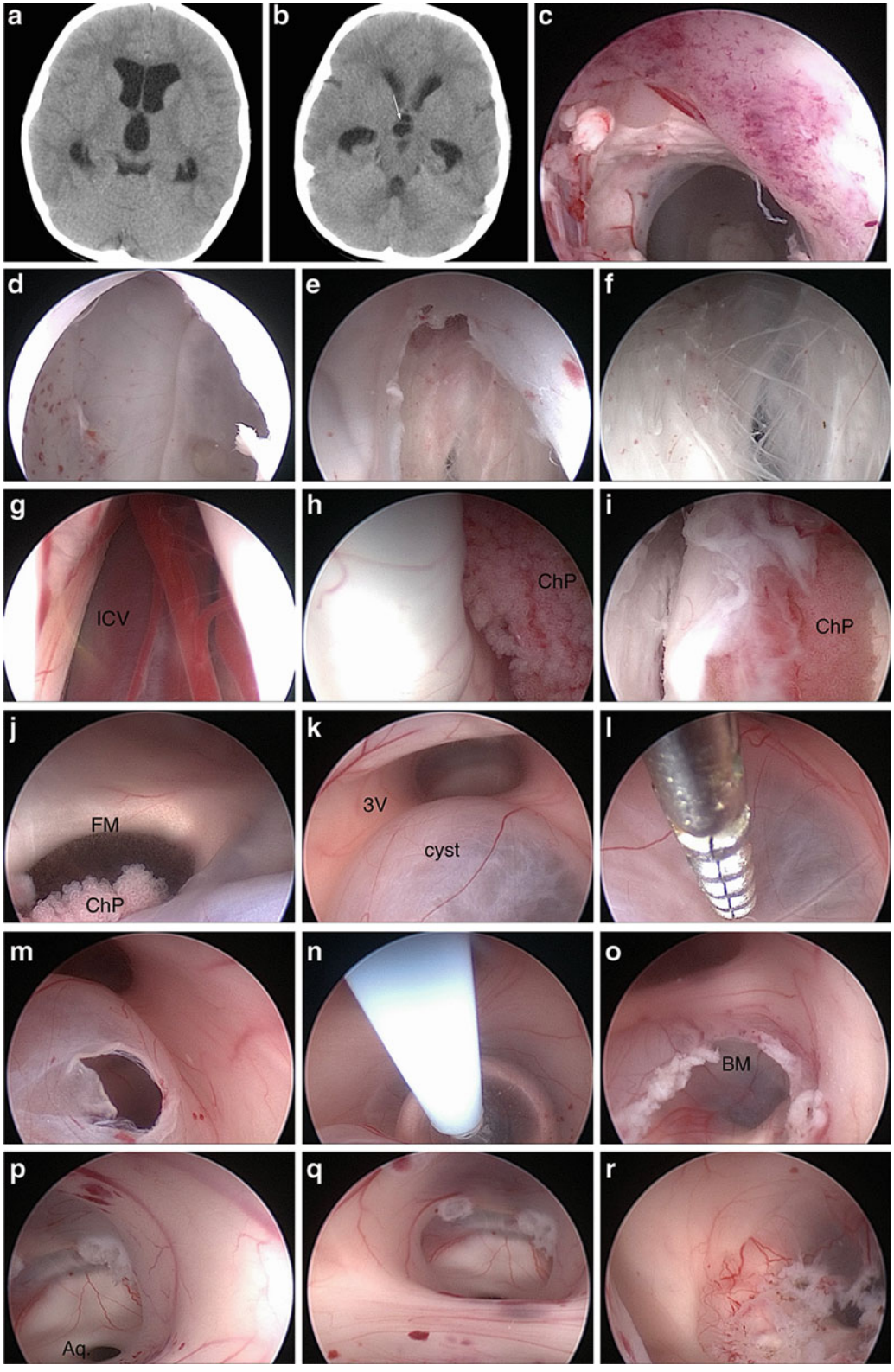
Radiologically, this kind of cysts can easily be missed on a CT, because it molds the third ventricle and can be taken for its simple dilatation. Diagnosis is only allowed by targeted CISS or DRIVE sequences on MRI.

They can be treated easily by endoscopic procedure, performing a cyst fenestration or resection by coagulation, associated with an ETV.

As choroid plexus cysts are often small cysts, they cause obstructive hydrocephalus by occlusion of the aqueduct opening and more rarely by occlusion of foramen of Monro.

Hydrocephalus and Choroid Plexus Cysts

Choroid plexus cysts are frequent, usually small, and asymptomatic intraventricular lesions. They are more frequent in the lateral ventricles,



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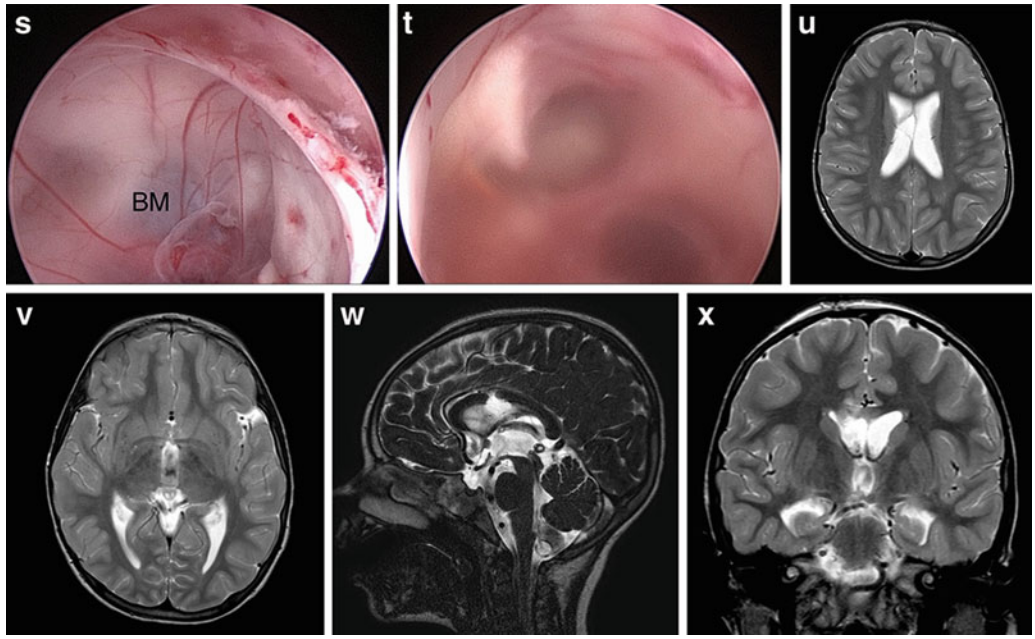

Fig. 55 **Case 3**. Arachnoid cyst of the third ventricle. (**a**, **b**) Axial computed tomodensitometry performed in emergency for a child in a sudden coma. The CT shows acute triventricular hydrocephalus. Actually it is an obstructive biventricular hydrocephalus caused by a third ventricle cyst, which is seen and materialized by the white arrow. (**c**–**t**) Endoscopic procedure consisting of ventriculocystocisternostomy, by a conventional right parasagittal approach through a coronal burr hole. (**c**) View through the parenchymal chimney of the interior of a cyst of the septum pellucidum. (**g**) Inter-fornix approach to an internal cerebral vein. (**h**–**j**) View of the right choroid plexus, which is followed to the right blocked foramen of Monro that is opened by a gentle pression downward. (**k**) Posterior rotation allowing the view of the third ventricle with a large cyst. (**l**–**n**) Large ventriculo-cystic fenestration. (**o**–**q**) Exploration of the third ventricle, which is finally free, as the aqueduct. (**r**, **s**) Rotation of the endoscope, showing arteries of the posterior complex. (**t**) Superior view of the third ventricle

especially in the atrium, although they can be found in the third one. They are attached to the plexus, usually at one point, which make them floating freely within the ventricle and can provoke intermittent obstruction of the CSF circulation, closing the aqueduct opening or the foramen of Monro by strangulation through the latter (Azab et al. 2015). A dozen of other cases are described. Azab et al. showed this particular movement of a cyst within the ventricles, during an endoscopic procedure. If the obstruction happens in a foramen, the hydrocephalus may be asymmetrical. The floating movement has also been showed in some studies with ultrasonographic videos. It is important to note that choroid plexus cysts are frequently found during fetuses' ultrasonography and increase the incidence of other pathological findings such as trisomies.

The three main surgical options are applicable and described for choroid plexus cysts. During the past years, many authors reported successful endoscopic procedures to perform cyst fenestration or resection by cauterization. Usually, an ETV was performed too. As these choroid plexus cysts come from the posterior portion of the third ventricle, where they may cause obstruction of the aqueduct, it is common to use a 30° endoscope after performing the ETV. Others will prefer to realize a double approach with two burr holes, one before the other, to particularly avoid fornix injuries (Ho et al. 2015).

Choroid plexus cysts of the third and lateral ventricles have been treated by cystoventriculoperitoneal shunting, endoscopic fenestration/resection, and microsurgical fenestration/-resection, using transcortical or transcallosal

approaches. After the 2000s, only endoscopic techniques have been described with very high success rates, making them the preferred treatment (Azab et al. 2015; de Lara et al. 2013; Ho et al. 2015).

Convexity Cyst

Pathophysiology of Hydrocephalus

The origin of arachnoid cysts of the convexity is not very known, because they grow far from the basal cisterns and ventricles. Probably they arise from the arachnoid membranes of the convexity, trapped in a sulcus. Arachnoid cysts of the convexity are seldom associated with hydrocephalus, except for those of huge size.

Subtypes

There is no real classification made in the literature for arachnoid cysts of the convexity. Usually, they can be classified into two main groups. The first one includes *focal cysts*, according to the cerebral lobe with which they are in contact and their expansion: frontal, temporal, parietal, occipital, and a combination of two/three lobes. The second group includes *hemispheric cysts*. In the Rengachary and Watanabe classification, these cysts were sometimes extensions of Sylvian fissure cysts and interhemispheric cysts (Rengachary and Watanabe 1981). The hemispheric ones exercise a compression of the cerebral parenchyma and the lateral ventricle, which can induce a dilatation of the contralateral ventricles.

Radiology (Fig. 56)

On a computed tomodensitometry, in infants, it may be difficult to differentiate a hemispheric cyst with a chronic subdural hematoma/hygroma or a hemiaplasia cerebri caused by an arrest in the development of one carotid artery and its branches, but these diagnoses are often more symptomatic, and there is no cranial asymmetry. The mass effect on the ipsilateral sulci is typically stronger with hygroma or hematoma. The MRI will differentiate the diagnosis.

Focal cysts are often associated with a cranial bone deformity, but on a computed tomography, they can assimilate to a low-grade glioma because of a focal hypodensity revelated often by seizures. Figure 56 shows the MRI of a patient with small arachnoid cysts of the convexity.

Treatment/Surgical Techniques

The data of literature about the treatment of these lesions is difficult to extract because of the frequent confusion of convexity cysts and interhemispheric, or sometimes with temporo-Sylvian cysts, in older articles (Matsuda et al. 1982; Chernov et al. 2004; Karabatsou et al. 2007; D'Angelo et al. 1999; Nowosławska et al. 2006; Oertel et al. 2009; Shim et al. 2009; Sommer and Smit 1997; Tamburrini et al. 2007; Gui et al. 2013). Most of them talk about endoscopic technique. The other possibilities are the implantation of a cystoperitoneal shunt, microsurgical fenestration/excision, and cystoventricular shunting. Basically, the surgical technique aims to fenestrate the cyst in a contiguous ventricle, forcing to have a huge convexity's cyst, or in the subarachnoid spaces, which must be widely opened. Sometimes the cyst can be close to a cistern, as the pericallosal, the Sylvian, or a basal cistern.

Following the microsurgical technique of fenestration, the endoscopic procedure appeared. Oertel et al., in 2009, reported 11 cases of convexity's cysts, all treated by endoscopic procedure with fenestration into the lateral ventricles (Oertel et al. 2009).They had 58% of total resolution of the symptoms and 34% of partial amelioration. Ninety-two percent of the cysts decreased in size, 8% needed a repeated intervention for closure of the stoma at 7 years follow-up, and 8% had a subdural hematoma that needed surgical evacuation. Gui et al. reported eight cases of arachnoid cysts of the convexity treated endoscopically (Gui et al. 2013). All of them showed a clinical resolution or improvement. All of the cysts decreased on the radiological controls. One

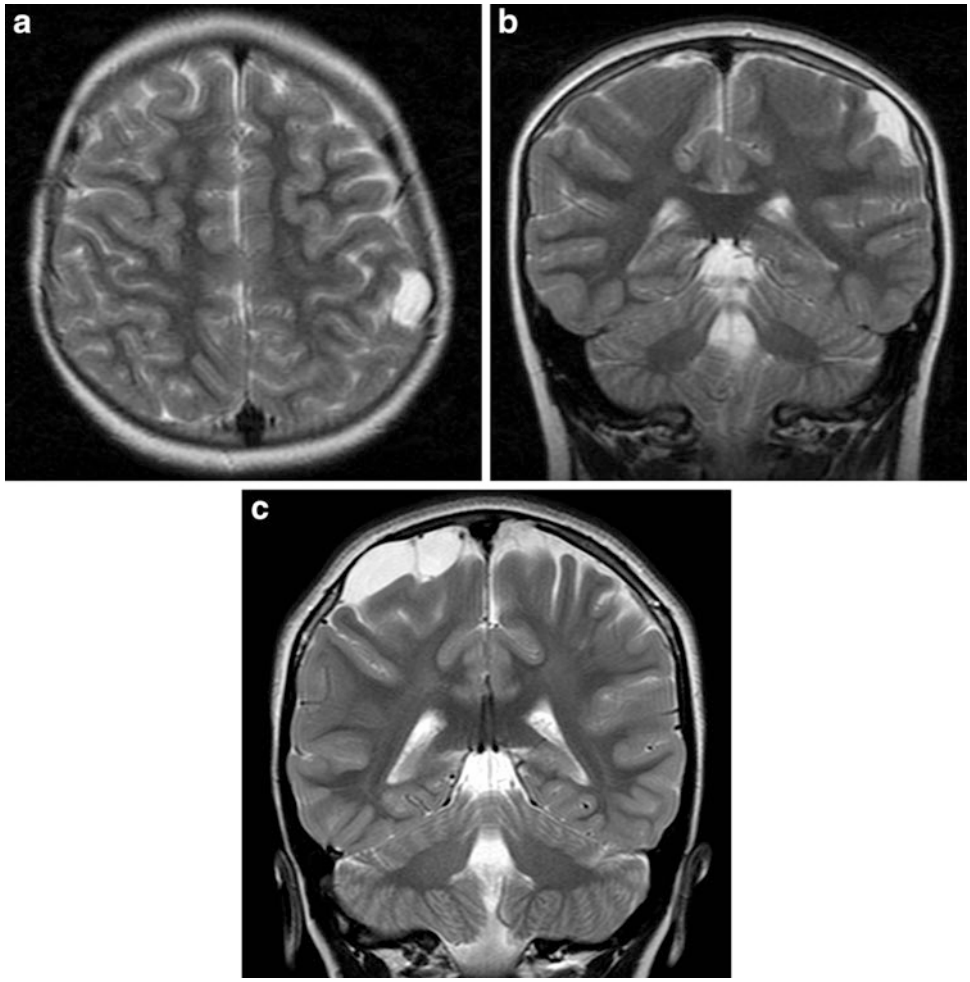

Fig. 56 (**a**–**c**) T2-weighted MRI showing small arachnoid cysts of the convexity, without any mass effect. (**a**) axial section. (**b**, **c**) coronal sections

patient had associated hydrocephalus with a large parieto-occipital arachnoid cyst, adjacent to the occipital horn of the left ventricle. For all of the cases, they performed a fenestration between the cyst and the adjacent ventricle, with addition of a third ventriculocisternostomy for cases with hydrocephalus, which disappeared. One patient had a spontaneously resolvent subdural hygroma. Galarza et al. reported 12 patients with a symptomatic cyst of the convexity, but no one had hydrocephalus (Galarza et al. 2002). Microsurgical fenestration was performed on four of them; the others were treated by cystoperitoneal shunt. Only 25% had radiological disappearance of the cyst; all of them have benefited of shunt. One hundred percent of the patients were asymptomatic at the follow-up.

To the three main treatments, cystoventricular shunt can be added.

Standard Microsurgical Procedure (Figs. 57 and 58)

Microsurgical technique is mostly indicated when a convexity arachnoid cyst is far away from any ventricle or cistern.

The position of the patient is adapted to the location of the cyst. The size of the craniotomy depends on the size of the cyst. The aim of the

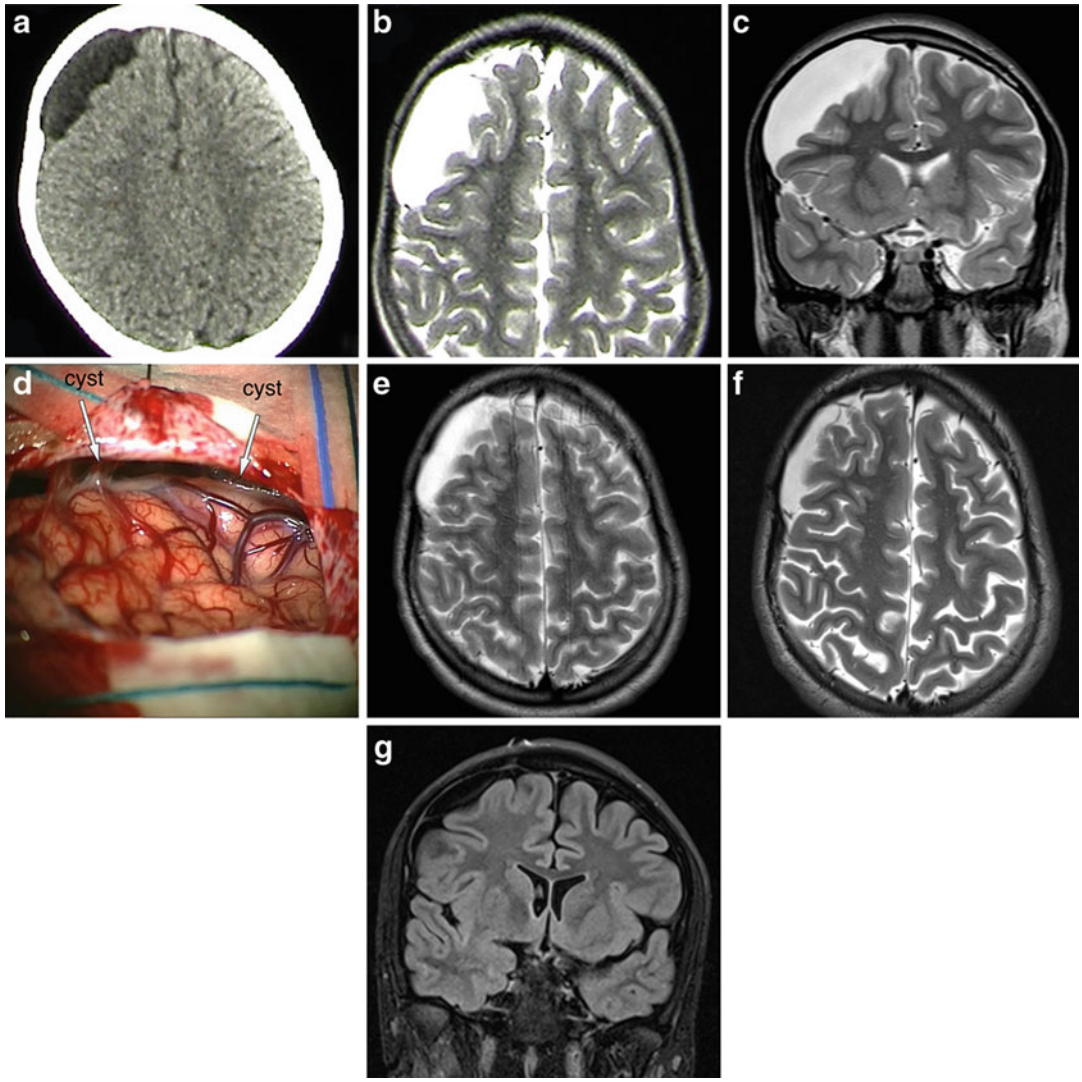

Fig. 57 Case 1. Microsurgical resection of a progressively growing right frontal convexity arachnoid cyst. (**a**) Computed tomodensitometry in axial section showing the cyst. Note the bone lysis. (**b**) Axial T2-weighted MRI 4 years later. (**c**) Coronal T2-weighted MRI 6 years after first imaging. (**d**) Microsurgical excision of the cyst walls. White arrows: rest of arachnoid membranes on bridging veins. (**e**) T2-weighted axial MRI 4 months after surgery. (**f**, **g**) 1-year postoperative MRI showing a satisfying regression of the cyst (probably complete, taking into account the bone lysis). (**f**) T2-weighted axial section. (**g**) T1-weighted coronal section

procedure is a complete removal of the cyst. Pay attention to the bridging veins that may be included into the walls of the cyst. Watertight closing is always very important. Figures 57 and 58 illustrate the microsurgical procedure for the treatment of a right frontal convexity arachnoid cyst (Fig. 57) and a left fronto-polar arachnoid cyst (Fig. 58).

Standard Endoscopic Procedure (Gui et al. 2013)

As usual, the preoperative planning and the perioperative use of the neuronavigation system are very important. They help to choose optimal locations for fenestrations.

The position of the patient and the placement of the burr hole are adapted to the location of the

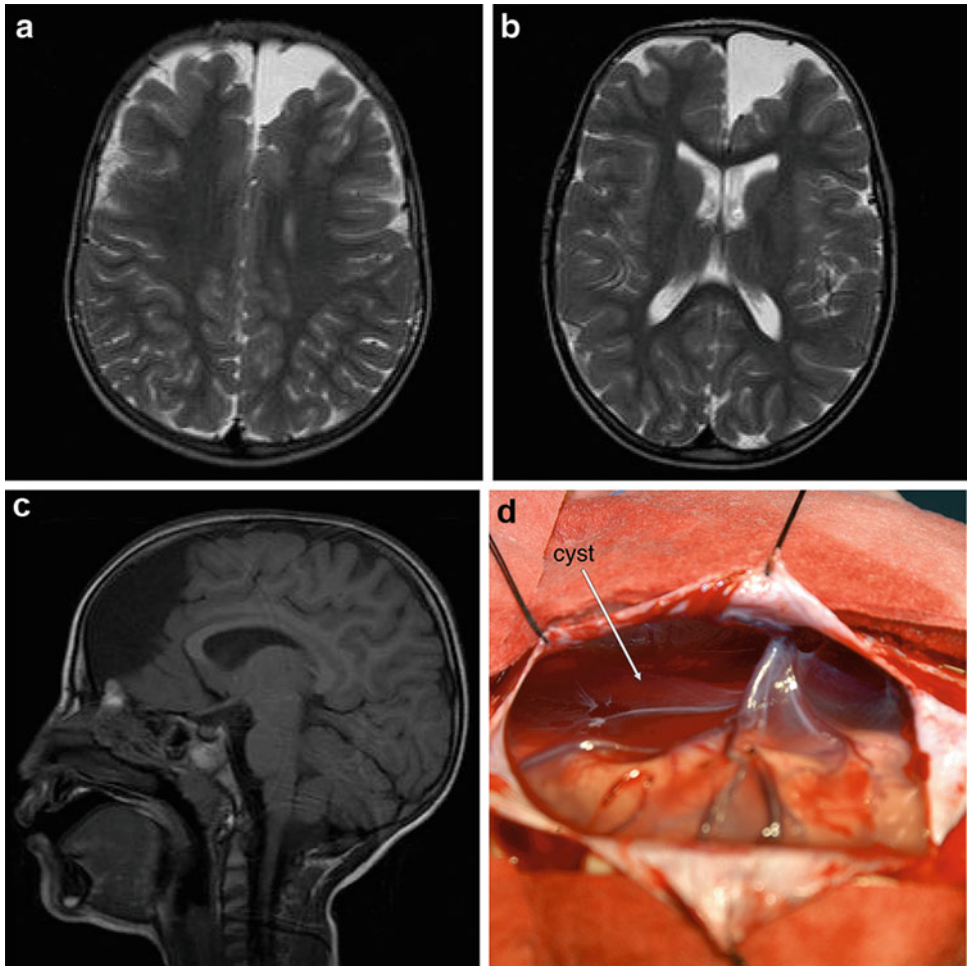

Fig. 58 **Case 2**. Microsurgical resection of a left frontopolar arachnoid cyst. (**a**, **b**) T2-weighted axial sections of the preoperative MRI. (**c**) T1-weighted sagittal section. (**d**) Microsurgical view through the craniotomy and durotomy. Remaining parts of the walls, adherent to bridging veins

Table 16 Success rate between the three surgical options for convexity together with interhemispheric arachnoid cysts treatment without isolating hydrocephalus (Gangemi et al. 2011)

Technique	Good outcome (%)
Microsurgery	89
***Cyst* shunting**	100
Endoscopic fenestration	75

Good outcome = complete or partial clinical remission

cyst. The aim of the procedure is to perform a cystoventricular, a cysto-cisternal, or a cysto-subarachnoid fenestration.

Watertight closing is always very important.

Discussion on Treatment

In 2011, Gangemi et al. have performed a comprehensive review of 61 reports in the literature comparing the clinicoradiological outcome of the 3 surgical procedures in the treatment of 44 convexity together with interhemispheric arachnoid cysts (Gangemi et al. 2011) (Table 16).

Conclusion

Hydrocephalus is mainly associated with posterior fossa and midline arachnoid cysts (Pierre-Khan and Sonigo 2003; Marinov et al. 1989;

Fewel et al. 1996; Shim et al. 2009). No significant differences in outcome and quality of life of patients between treatments have been found in a large meta analysis (Ali et al. 2014). Suprasellar, interhemispheric, quadrigeminal, and posterior fossa arachnoid cysts seem to be the best indications for endoscopic fenestration, because of their position in the midline, in intimate relationships with basal cisterns and ventricles. Cortical cysts are best treated by open surgery or shunting.

Using natural fluid structures as field of approach, each time it is possible, seems to be a key of the success of neuroendoscopy, keeping an overview of anatomy. Preferring ventricles allows recognition of normal anatomical structures.

Keep in mind during neuroendoscopic procedures to use the neuronavigation system after a good preoperative planning (Cinalli et al. 2007; Martínez-Lage et al. 2011). Pay attention to realize the largest possible fenestration (Abott 2004). Large openings prevent subsequent occlusion (Lechanoine et al. 2018). Multiple fenestrations help for the restoration of better CSF pathways. Intraoperative tabulation control of the flows through stoma could give good indices about subsequent efficiency. Each time it is possible and safe, perform a third ventriculostomy (Di Rocco et al. 2005). For such neuroendoscopic procedures the experience of the assistant surgeon is atleast as important as the one of the main surgeon (Abott 2004).

Choosing the implantation of a cyst or ventriculoperitoneal shunt must be carefully weighed, because such a solution implies a lifelong condition, with a high risk of complications, such as shunt malfunctions, shunt dependency, or acquired chronic tonsillar herniation malformation, which can lead to severe clinical manifestations and represent a huge challenge for their management (Kim et al. 2002; Laviv and Michowitz 2010; Li et al. 2014; Martínez-Lage et al. 2009). Anti-siphon or programmable valves seem to decrease such complications.

The appropriate expression "primum non nocere" has to be still kept in mind for the management of benign intracranial lesions such as arachnoid cysts, in case of asymptomatic ones.

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Anatomical Nomenclature of Figures	
3V	Third ventricle
4V	Fourth ventricle
AC	Anterior commissure
AcoA	Anterior communicant artery
Aq.	Aqueduct
BA	Basilar artery
BM	Basal membrane of the arachnoid cyst
BV	Basal vein
Cereb.	Cerebellum
Ch.	Optic chiasm
ChP	Choroid plexus
clivus	–
CN	Caudate nucleus
Cyst	–
Falx	–
flocc.	Flocculus
FM	Foramen of Monro
GCV	Great cerebral vein
Hipp.	Hippocampus
I, II, III, IV, V, VI, VII, VIII, IX, X, XI, XII	Cranial nerves
ICA	Internal carotid artery
ICV	Internal cerebral vein
Inf.	Infundibulum
insula	–
IPS	Inferior petrosal sinus
JB	Jugular bulb
LT	Lamina terminalis
LV	Lateral ventricle
MB	Mamillary bodies
MCA	Middle cerebral artery
MCF	Middle cranial fossa

med. obl.	Medulla oblongata
Mid.	Midbrain
MN	Mixes nerves
PC	Posterior commissure
PCA	Posterior cerebral artery
PCli	Posterior clinoid
PcoA	Posterior communicant artery
Petr. bone	Petrous bone
pons	–
PS	Pituitary stalk
SP	Septum pellucidum
Tent.	Tentorium

References

Abbott R (2004) The endoscopic management of arachnoidal cysts. Neurosurg Clin N Am Clin Neuroendoscopy 15(1):9–17. https://doi.org/10.1016/S1042-3680(03)00071-8

Ahn Y, Cho BK, Wang KC (1997) Bobble-head doll syndrome associated with subduroperitoneal shunt malfunction. Childs Nerv Syst 13(4):234–237. https://doi.org/10.1007/s003810050074

Albright AL, Tyler-Kabara E (2001) Slit-ventricle syndrome secondary to shunt-induced suture ossification. Neurosurgery 48(4):764–769; discussion 769–70

Alexiou GA, Varela M, Sfakianos G, Prodromou N (2010) Shunting for the treatment of arachnoid cysts in children. Neurosurgery 67(6):1632–1636; discussion 1636. https://doi.org/10.1227/NEU.0b013e3181f94476

Al-Holou WN, Yew AY, Boomsaad ZE, Garton HJL, Muraszko KM, Maher CO (2010) Prevalence and natural history of arachnoid cysts in children: clinical article. J Neurosurg Pediatr 5(6):578–585

Ali ZS, Lang S-S, Bakar D, Storm PB, Stein SC (2014) Pediatric intracranial arachnoid cysts: comparative effectiveness of surgical treatment options. Childs Nerv Syst 30(3):461–469. https://doi.org/10.1007/s00381-013-2306-2

André A, Zérah M, Roujeau T, Brunelle F, Blauwblomme T, Puget S, Bourgeois M, Sainte-Rose C, Ville Y, Di Rocco F (2016) Suprasellar arachnoid cysts: toward a new simple classification based on prognosis and treatment modality. Neurosurgery 78(3):370–380. https://doi.org/10.1227/NEU.0000000000001049

Aoki N, Sakai T (1990) Intraoperative subdural hematoma in a patient with arachnoid cyst in the middle cranial fossa. Childs Nerv Syst 6(1):44–46

Aoki N, Sakai T, Umezawa Y (1990) Slit ventricle syndrome after cyst-peritoneal shunting for the treatment of intracranial arachnoid cyst. Childs Nerv Syst 6(1):41–43

Arai H, Sato K, Wachi A, Okuda O, Takeda N (1996) Arachnoid cysts of the middle cranial fossa: experience with 77 patients who were treated with cystoperitoneal shunting. Neurosurgery 39(6):1108–1112; discussion 1112–13

Azab WA, Mijalcic RM, Aboalhasan AA, Khan TA, Abdelnabi EA (2015) Endoscopic management of a choroid plexus cyst of the third ventricle: case report and documentation of dynamic behavior. Childs Nerv Syst 31(5):815–819. https://doi.org/10.1007/s00381-015-2649-y

Becker T, Wagner M, Hofmann E, Warmuth-Meth M, Nadjmi M (1991) Do arachnoid cysts grow? A retrospective CT volumetric study. Neuroradiology 33:341–345

Benzel EC, Reeves JD, Kesterson L, Hadden TA (1992) Slit ventricle syndrome in children: clinical presentation and treatment. Acta Neurochir 117(1–2):7–14

Boop F, Young R, Scott R (1998) Arachnoid cysts of the middle cranial fossa and convexity. In: Kaufmann H (ed) Cerebrospinal fluid collections. American Association of Neurological Surgeons, Park Ridge, pp 67–96

Bretelle F, Senat M-V, Bernard J-P, Hillion Y, Ville Y (2002) First-trimester diagnosis of fetal arachnoid cyst: prenatal implication. Ultrasound Obstet Gynecol 20(4):400–402. https://doi.org/10.1046/j.1469-0705.2002.00813.x

Bright R (1831) Reports of medical cases selected with a view of illustrating the symptoms and cure of diseases by a reference to morbid anatomy. Vol II: Diseases of the brain and nervous system. Longman, Rees, Orme, Brown, Green, and Highley, London, pp 437–439

Caldarelli M, Di Rocco C (1996) Surgical options in the treatment of interhemispheric arachnoid cysts. Surg Neurol 46(3):212–221. https://doi.org/10.1016/0090-3019(96)00155-3

Caldarelli M, Novegno F, Di Rocco C (2009) A late complication of CSF shunting: acquired chiari I malformation. Childs Nerv Syst 25(4):443–452. https://doi.org/10.1007/s00381-008-0760-z

Campistol Plana J, Costa Clara JM, Fernández-Alvarez E (1983) Intracranial arachnoid cysts in children. Review of 34 cases. An Esp Pediatr 19(6):459–470

Candela S, Puerta P, Alamar M, Barcik U, Guillén A, Muchart J, García-Fructuoso G, Ferrer-Rodríguez E (2015) Epidemiología y clasificación de los quistes aracnoideos en niños. Neurocirugia 26(5):234–240. https://doi.org/10.1016/j.neucir.2015.02.007

Cassart M, Bosson N, Garel C, Eurin D, Avni F (2008) Fetal intracranial tumors: a review of 27 cases. Eur Radiol 18(10):2060–2066. https://doi.org/10.1007/s00330-008-0999-5

Chernov MF, Kamikawa S, Yamane F, Hori T (2004) Double-endoscopic approach for management of convexity arachnoid cyst: case report. Surg Neurol 61(5):483–486; discussion 486–87. https://doi.org/10.1016/S0090-3019(03)00574-3

Chinn DH, Miller EI, Worthy LM, Towers CV (1991) Sonographically detected fetal choroid plexus cysts. Frequency and association with aneuploidy. J Ultrasound Med 10(5):255–258

Choux M, Raybaud C, Pinsard N, Hassoun J, Gambarelli D (1978) Intracranial Supratentorial cysts in children

excluding tumor and parasitic cysts. Childs Brain 4(1):15–32

Cinalli G, Peretta P, Spennato P, Savarese L, Varone A, Vedova P, Grimaldi G et al (2006) Neuroendoscopic management of interhemispheric cysts in children. J Neurosurg 105(Suppl 3):194–202. https://doi.org/10.3171/ped.2006.105.3.194

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. https://doi.org/10.1007/s00381-007-0333-6

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: clinical article. J Neurosurg Pediatr 6(5):489–497

Ciricillo SF, Cogen PH, Harsh GR, Edwards MS (1991) Intracranial arachnoid cysts in children. A comparison of the effects of fenestration and shunting. J Neurosurg 74(2):230–235. https://doi.org/10.3171/jns.1991.74.2.0230

Conde Sardón R (2015) Quistes aracnoideos. Evolución histórica del concepto y teorías fisiopatológicas. Neurocirugia 26(4):192–195. https://doi.org/10.1016/j.neucir.2015.03.001

Couvreur T, Hallaert G, Van Der Heggen T, Baert E, Dewaele F, Okito J-PK, Vanhauwaert D, Deruytter M, Van Roost D, Caemaert J (2015) Endoscopic treatment of temporal arachnoid cysts in 34 patients. World Neurosurg 84(3):734–740. https://doi.org/10.1016/j.wneu.2015.04.053

D'Angelo, V', L. Gorgoglione, G. Catapano. 1999. Treatment of symptomatic intracranial arachnoid cysts by stereotactic cyst-ventricular shunting. Stereotact Funct Neurosurg 72(1): 62–69. https://doi.org/10.1159/000029672

de Lara D, Ditzel Filho LFS, Muto J, Prevedello DM (2013) Endoscopic treatment of a third ventricle choroid plexus cyst. Neurosurg Focus 34(Suppl 1):Video 9. https://doi.org/10.3171/2013.V1.FOCUS12332

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(4):696–701. https://doi.org/10.3171/jns.1996.84.4.0696

Del Bigio MR (2002) Neuropathological findings in a child with slit ventricle syndrome. Pediatr Neurosurg 37(3):148–151

Di Rocco C (2010) Sylvian fissure arachnoid cysts: we do operate on them but should it be done? Childs Nerv Syst 26(2):173–175. https://doi.org/10.1007/s00381-009-1041-1

Di Rocco C, Tamburrini G (2003) Shunt dependency in shunted arachnoid cyst: a reason to avoid shunting. Pediatr Neurosurg 38(3):164. https://doi.org/10.1159/000068817

Di Rocco C, Velardi F (2003) Acquired chiari type I malformation managed by supratentorial cranial enlargement. Childs Nerv Syst 19(12):800–807. https://doi.org/10.1007/s00381-003-0837-7

Di Rocco F, Zerah M (2011) Evaluating endoscopic treatment of arachnoid cysts. Neurosurgery 68(4):E1176; author reply E1175–76. https://doi.org/10.1227/NEU.0b013e31820bf77d

Di Rocco C, Di Trapani G, Iannelli A (1979) Arachnoid cyst of the fourth ventricle and 'arrested' hydrocephalus. Surg Neurol 12(6):467–471

Di Rocco C, Caldarelli M, Di Trapani G (1981) Infratentorial arachnoid cysts in children. Childs Brain 8(2):119–133

Di Rocco C, Tamburrini G, Caldarelli M, Velardi F, Santini P (2003) Prolonged ICP monitoring in children with sylvian fissure arachnoid cysts. Minerva Pediatr 55(6):583–591

Di Rocco F, Yoshino M, Oi S (2005) Neuroendoscopic transventricular ventriculocystostomy in treatment for intracranial cysts. J Neurosurg 103(Suppl 1):54–60. https://doi.org/10.3171/ped.2005.103.1.0054

Di Rocco F, James SR, Roujeau T, Puget S, Sainte-Rose C, Zerah M (2010) Limits of endoscopic treatment of Sylvian arachnoid cysts in children. Childs Nerv Syst 26(2):155–162. https://doi.org/10.1007/s00381-009-0977-5

Dyck P, Gruskin P (1977) Supratentorial arachnoid cysts in adults: a discussion of two cases from a pathophysiologic and surgical perspective. Arch Neurol 34(5):276–279

El-Ghandour NMF (2013) Endoscopic treatment of quadrigeminal arachnoid cysts in children. J Neurosurg Pediatr 12(5):521–528. https://doi.org/10.3171/2013.7.PEDS13155

Elhammady MSA, Bhatia S, Ragheb J (2007) Endoscopic fenestration of middle fossa arachnoid cysts: a technical description and case series. Pediatr Neurosurg 43(3):209–215. https://doi.org/10.1159/000098833

Epstein FJ, Fleischer AS, Hochwald GM, Ransohoff J (1974) Subtemporal craniectomy for recurrent shunt obstruction secondary to small ventricles. J Neurosurg 41(1):29–31. https://doi.org/10.3171/jns.1974.41.1.0029

Erşahin Y, Kesikçi H (2009) Endoscopic management of quadrigeminal arachnoid cysts. Childs Nerv Syst 25(5):569–576. https://doi.org/10.1007/s00381-008-0778-2

Erşahin Y, Kesikçi H, Rüksen M, Aydin C, Mutluer S (2008) Endoscopic treatment of suprasellar arachnoid cysts. Childs Nerv Syst 24(9):1013–1020. https://doi.org/10.1007/s00381-008-0615-7

Eskandary H, Sabba M, Khajehpour F, Eskandari M (2005) Incidental findings in brain computed tomography scans of 3000 head trauma patients. Surg Neurol 63(6):550–553. https://doi.org/10.1016/j.surneu.2004.07.049

Fang T, Xu J, Wang S, Ma Z, Xing J (2010) Analysis of therapeutic choices for slit ventricle syndrome after cyst-peritoneal shunting for temporal arachnoid cysts in children. J Neurosurg Pediatr 6(5):474–480. https://doi.org/10.3171/2010.8.PEDS10222

Fang T, Yan R, Xing J, Li Y (2012) Slit ventricle syndrome after cyst-peritoneal shunting for the arachnoid cyst – a report of two cases. Clin Neurol Neurosurg 114(1):87–89. https://doi.org/10.1016/j.clineuro.2011.09.007

Feletti A, Alicandri-Ciufelli M, Pavesi G (2016) Trans-aqueductal trans-magendie fenestration of arachnoid cyst in the posterior fossa. Acta Neurochir 158(4):655–662. https://doi.org/10.1007/s00701-016-2734-3

Fernández Molina G (2013) Neuroendoscopic management of middle fossa arachnoid cysts. World Neurosurg 79(Suppl 2):S19.e19–S19.e23. https://doi.org/10.1016/j.wneu.2012.02.012

Fewel ME, Levy ML, McComb JG (1996) Surgical treatment of 95 children with 102 intracranial arachnoid cysts. Pediatr Neurosurg 25(4):165–173

Fox JL, Al-Mefty O (1980) Suprasellar arachnoid cysts: an extension of the membrane of Liliequist. Neurosurgery 7:615–618

Froelich SC, Abdel Aziz KM, Cohen PD, van Loveren HR, Keller JT (2008) Microsurgical and endoscopic anatomy of Liliequist's membrane: a complex and variable structure of the basal cisterns. Neurosurgery 63(1 Suppl 1):ONS1–8; discussion ONS8–9. https://doi.org/10.1227/01.neu.0000335004.22628.ee

Fujio S, Bunyamin J, Hirano H, Oyoshi T, Sadamura Y, Bohara M, Arita K (2016) A novel bilateral approach for suprasellar arachnoid cysts: a case report. Pediatr Neurosurg 51(1):30–34. https://doi.org/10.1159/000440811

Galarza M, Pomata HB, Pueyrredón F, Bartuluchi M, Zuccaro GN, Monges JA (2002) Symptomatic supratentorial arachnoid cysts in children. Pediatr Neurol 27(3):180–185. https://doi.org/10.1016/S0887-8994(02)00414-9

Galarza M, López-Guerrero AL, Martínez-Lage JF (2010) Posterior fossa arachnoid cysts and cerebellar tonsillar descent: short review. Neurosurg Rev 33(3):305–314; discussion 314. https://doi.org/10.1007/s10143-010-0262-9

Galassi E, Tognetti F, Gais 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 17:363–369

Gangemi M, Maiuri F, Colella G, Magro F (2005) Endoscopic treatment of quadrigeminal cistern arachnoid cysts. Minim Invasive Neurosurg 48(5):289–292. https://doi.org/10.1055/s-2005-915604

Gangemi M, Seneca V, Colella G, Cioffi V, Imperato A, Maiuri F (2011) Endoscopy versus microsurgical cyst excision and shunting for treating intracranial arachnoid cysts. J Neurosurg Pediatr 8(2):158–164. https://doi.org/10.3171/2011.5.PEDS1152

García-Conde M, Martín-Viota L (2015) Quistes aracnoideos: embriología y anatomía patológica. Neurocirugia 26(3):137–142. https://doi.org/10.1016/j.neucir.2015.02.003

Garg K, Tandon V, Sharma S, Suri A, Chandra PS, Kumar R, Mahapatra AK, Sharma BS (2014) Quadrigeminal cistern arachnoid cyst: a series of 18 patients and a review of literature. Br J Neurosurg:1–7. https://doi.org/10.3109/02688697.2014.957646

Germanò A, Caruso G, Caffo M, Baldari S, Calisto A, Meli F, Tomasello F (2003) The treatment of large Supratentorial arachnoid cysts in infants with cyst-peritoneal shunting and hakim programmable valve. Childs Nerv Syst 19(3):166–173. https://doi.org/10.1007/s00381-002-0702-0

Go KG, Houthoff HJ, Hartsuiker J, Blaauw EH, Havinga P (1986) Fluid secretion in arachnoid cysts as a clue to cerebrospinal fluid absorption at the arachnoid granulation. J Neurosurg 65:642–648

Goluboff LG (1973) Arachnoid cyst of the posterior fossa demonstrated by isotope cisternography. J Nucl Med 14(1):61–62

Gui S-B, Wang X-S, Zong X-Y, Zhang Y-Z, Li C-Z (2011) Suprasellar cysts: clinical presentation, surgical indications, optimal surgical treatment. BMC Neurol 11:52. https://doi.org/10.1186/1471-2377-11-52

Gui S, Zong X, Li C, Zhang Y (2013) Endoscopic treatment of convexity arachnoid cysts. Childs Nerv Syst 29(3):505–508. https://doi.org/10.1007/s00381-012-1957-8

Gui S, Bai J, Wang X, Zong X, Li C, Cao L, Zhang Y (2016) Assessment of endoscopic treatment for quadrigeminal cistern arachnoid cysts: a 7-year experience with 28 cases. Childs Nerv Syst 32(4):647–654. https://doi.org/10.1007/s00381-015-2962-5

Handa H, Bucy PC (1956) Benign cysts of the brain simulating brain tumor. J Neurosurg 13(5):489–499

Harter LP, Silverberg GD, Brant-Zawadzki M (1980) Intrasellar arachnoid cyst: case report. Neurosurgery 7:387–390

Hassounah MI, Rahm BE (1994) Hindbrain herniation: an unusual occurrence after shunting an intracranial arachnoid cyst. Case report. J Neurosurg 81(1):126–129. https://doi.org/10.3171/jns.1994.81.1.0126

Hinojosa J, Esparza J, Muñoz y MJ, Valencia J (2001) Tratamiento Endoscópico de Los Quistes Aracnoideos Supraselares. Neurocirugia 12(6):482–488. https://doi.org/10.1016/S1130-1473(01)70662-8

Hirano A, Hirano M (1988) Benign cystic lesions in the central nervous system. Light and electron microscopic observations of cyst walls. Childs Nerv Syst 4(6):325–333

Hirano A, Hirano M (2004) Benign cysts in the central nervous system: neuropathological observations of the cyst walls. Neuropathology 24(1):1–7

Ho AL, Pendharkar AV, Sussman ES, Ravikumar VK, Gordon HL (2015) Dual-trajectory approach for simultaneous cyst fenestration and endoscopic third ventriculostomy for treatment of a complex third ventricular arachnoid cyst. Curēus 7(3):e253. https://doi.org/10.7759/cureus.253

Hogge WA, Schnatterly P, Ferguson JE (1995) Early prenatal diagnosis of an infratentorial arachnoid cyst: association with an unbalanced translocation. Prenat Diagn 15(2):186–188

Huang Q, Wang D, Guo Y, Zhou X, Wang X, Li X (2007) The diagnosis and neuroendoscopic treatment of non-communicating intracranial arachnoid cysts. Surg Neurol 68(2):149–154; discussion 154. https://doi.org/10.1016/j.surneu.2006.10.059

Idris Z, Tan YC, Kandasamy R, Ghani RI, Abdullah JM (2016) Transfrontal transaqueductal, transtrigonal, and suboccipital infratentorial supracerebellar endoscopic

fenestration of posterior fossa arachnoid cysts: three surgical cases. J Neurol Surg A Cent Eur Neurosurg. https://doi.org/10.1055/s-0035-1571161

Jallo GI, Morota N, Abbott R (1996) Introduction of a second working portal for neuroendoscopy. A technical note. Pediatr Neurosurg 24(2):56–60

Jang M, Yoon SH (2013) Hypothesis for intracranial hypertension in slit ventricle syndrome: new concept of capillary absorption laziness in the hydrocephalic patients with long-term shunts. Med Hypotheses 81(2):199–201. https://doi.org/10.1016/j.mehy.2013.05.003

Jian-Huang H, Mei W-Z, Chen Y, Chen J-W, Lin Z-X (2015) Analysis on clinical characteristics of intracranial arachnoid cysts in 488 pediatric cases. Int J Clin Exp Med 8(10):18343–18350

Johnson RD, Chapman S, Bojanic S (2011) Endoscopic fenestration of middle cranial fossa arachnoid cysts: does size matter? J Clin Neurosci 18(5):607–612. https://doi.org/10.1016/j.jocn.2010.10.006

Jones RF, Warnock TH, Nayanar V, Gupta JM (1989) Suprasellar arachnoid cysts: management by cyst wall resection. Neurosurgery 25(4):554–561

Kang JK, Lee KS, Lee IW, Jeun SS, Son BC, Jung CK, Park YS, Lee SW (2000) Shunt-independent surgical treatment of middle cranial fossa arachnoid cysts in children. Childs Nerv Syst 16(2):111–116. https://doi.org/10.1007/s003810050024

Karabatsou K, Hayhurst C, Buxton N, O'Brien DF, Mallucci CL (2007) Endoscopic management of arachnoid cysts: an advancing technique. J Neurosurg 106 (Suppl 6):455–462. https://doi.org/10.3171/ped.2007.106.6.455

Katzman GL, Dagher AP, Patronas NJ (1999) Incidental findings on brain magnetic resonance imaging from 1000 asymptomatic volunteers. JAMA 282(1):36–39

Kawamoto H, Ikawa F, Imada Y, Katoh H, Hasegawa A (2007) Two-step surgical treatment using miniature Ommaya's reservoirs for a neonate with multiple large arachnoid cysts. Childs Nerv Syst 23(5):591–594. https://doi.org/10.1007/s00381-006-0257-6

Kim S-K, Cho B-K, Chung Y-N, Kim H-S, Wang K-C (2002) Shunt dependency in shunted arachnoid cyst: a reason to avoid shunting. Pediatr Neurosurg 37(4): 178–185. https://doi.org/10.1159/000065393

Kimiwada T, Hayashi T, Narisawa A, Shirane R, Tominaga T (2015) Shunt placement after cyst fenestration for middle cranial fossa arachnoid cysts in children. J Neurosurg Pediatr:1–7. https://doi.org/10.3171/2015.3.PEDS14573

King JAJ, Auguste KI, Halliday W, Drake JM, Kulkarni AV (2010) Ventriculocystostomy and endoscopic third ventriculostomy/shunt placement in the management of hydrocephalus secondary to giant retrocerebellar cysts in infancy. J Neurosurg Pediatr 5(4):403–407. https://doi.org/10.3171/2009.10.PEDS09208

Konovalov AN, Rostotskaia VI, Ivakina NI, Simernitskiĭ BP (1988) Surgical treatment of suprasellar cerebrospinal fluid cysts. Zh Vopr Neirokhir Im N N Burdenko 1:11–16

Kumagai M, Sakai N, Yamada H, Shinoda J, Nakashima T, Iwama T (1986) Postnatal development and enlargement of primary middle cranial fossa arachnoid cyst recognized on repeat CT scans. Childs Nerv Syst 2:211–215

Laviv Y, Michowitz S (2010) Acute intracranial hypertension and shunt dependency following treatment of intracranial arachnoid cyst in a child: a case report and review of the literature. Acta Neurochir 152(8): 1419–1423; discussion 1422–23. https://doi.org/10.1007/s00701-010-0658-x

Lazareff JA, Kelly J, Saito M (1998) Herniation of cerebellar tonsils following supratentorial shunt placement. Childs Nerv Syst 14(8):394–397. https://doi.org/10.1007/s003810050252

Lechanoine F, Spennato P, Ruggiero C, Cinalli G (2018) Importance of fenestration size for definitive treatment of a quadrigeminal arachnoid cyst: Endoscopic inspection of the cyst and surrounding anatomy: 2-dimensional Operative video. Oper Neurosurg. https://doi.org/10.1093/ons/opy122

Lena G, van Calenberg F, Genitori L, Choux M (1995) Supratentorial interhemispheric cysts associated with callosal agenesis: surgical treatment and outcome in 16 children. Childs Nerv Syst 11(10):568–573

Lena G, Erdincler P, Van Calenberg F, Genitori L, Choux M (1996) Arachnoid cysts of the middle cranial fossa in children. A review of 75 cases, 47 of which have been operated in a comparative study between membranectomy with opening of cisterns and cystoperitoneal shunt. Neuro-Chirurgie 42(1):29–34

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; discussion 1144–45

Levy ML, Meltzer HS, Hughes S, Aryan HE, Yoo K, Amar AP (2004) Hydrocephalus in children with middle fossa arachnoid cysts. J Neurosurg 101(Suppl 1):25–31. https://doi.org/10.3171/ped.2004.101.2.0025

Li C, Yin L, Jiang T, Ma Z, Jia G (2014) Shunt dependency syndrome after cystoperitoneal shunting of arachnoid cysts. Child Nerv Syst 30(3):471–476. https://doi.org/10.1007/s00381-013-2248-8

Little JR, Gomez MR, MacCarty CS (1973) Infratentorial arachnoid cysts. J Neurosurg 39(3):380–386

López R, Bienvenido ÁMG, Moroño SI (2015) Quistes aracnoideos del sistema nervioso central. Algoritmos y recomendaciones generales de manejo. Neurocirugia. https://doi.org/10.1016/j.neucir.2015.02.002

Lyu J (2014) Possible origins of suprasellar arachnoid cysts based on anatomical considerations. World Neurosurg 82(3–4):e570–e573. https://doi.org/10.1016/j.wneu.2014.05.020

Maher CO, Goumnerova L (2011) The effectiveness of ventriculocystocisternostomy for suprasellar arachnoid cysts. J Neurosurg Pediatr 7(1):64–72. https://doi.org/10.3171/2010.10.PEDS10356

Malinger G, Kidron D, Schreiber L, Ben-Sira L, Hoffmann C, Lev D, Lerman-Sagie T (2007) Prenatal diagnosis of malformations of cortical development by dedicated neurosonography. Ultrasound Obstet Gynecol 29(2):178–191. https://doi.org/10.1002/uog.3906

Malinger G, Sereño EC, Lerman-Sagie T (2008) The differential diagnosis of fetal intracranial cystic lesions. Ultrasound Clin 3(4):553–558. https://doi.org/10.1016/j.cult.2008.09.002. Advanced obstetrical ultrasound fetal brain, spine, and limb abnormalities

Marinov M, Undjian S, Wetzka P (1989) An evaluation of the surgical treatment of intracranial arachnoid cysts in children. Childs Nerv Syst 5(3):177–183

Martinez-Lage JF, Poza M, Sola J, Puche A (1992) Congenital arachnoid cyst of the lateral ventricles in children. Childs Nerv Syst 8(4):203–206

Martínez-Lage JF, Ruíz-Maciá D, Valentí JA, Poza M (1999) Development of a middle fossa arachnoid cyst. A theory on its pathogenesis. Childs Nerv Syst 15(2–3):94–97

Martínez-Lage JF, Vilar AR-E, Pérez-Espejo MA, Almagro M-J, Pedro JR d S, Murcia MF (2006) Shunt-related craniocerebral disproportion: treatment with cranial vault expanding procedures. Neurosurg Rev 29(3):229–235. https://doi.org/10.1007/s10143-006-0022-z

Martínez-Lage JF, Ruíz-Espejo AM, Almagro M-J, Alfaro R, Felipe-Murcia M, López López-Guerrero A (2009) CSF overdrainage in shunted intracranial arachnoid cysts: a series and review. Childs Nerv Syst 25(9):1061–1069. https://doi.org/10.1007/s00381-009-0910-y

Martínez-Lage JF, Pérez-Espejo MA, Almagro M-J, López-Guerrero AL (2011) Hydrocephalus and arachnoid cysts. Childs Nerv Syst 27(10):1643–1652. https://doi.org/10.1007/s00381-011-1481-2

Matsuda M, Hirai O, Munemitsu H, Kawamura J, Matsubayashi K (1982) Arachnoid cysts – report of two adult cases in the interhemispheric fissure and over the cerebral convexity. Neurol Med Chir 22(1):71–76

Mattox A, Choi JD, Leith-Gray L, Grant GA, Cory Adamson D (2010) Guidelines for the management of obstructive hydrocephalus from suprasellar-prepontine arachnoid cysts using endoscopic third ventriculocystocisternostomy. Surg Innov 17(3):206–216. https://doi.org/10.1177/1553350610377212

McBride LA, Winston KR, Freeman JE (2003) Cystoventricular shunting of intracranial arachnoid cysts. Pediatr Neurosurg 39(6):323–329. https://doi.org/10.1159/000075261

Mendonça R, Lima TTF, Oppitz PP, Raupp SF (2006) Herniation of the cerebellar tonsils after suprasellar arachnoid cyst shunt: case report. Arq Neuropsiquiatr 64(2B):523–525

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

Mori K (1992) Giant interhemispheric cysts associated with agenesis of the corpus callosum. J Neurosurg 76(2):224–230. https://doi.org/10.3171/jns.1992.76.2.0224

Murphy L (1985) Parasagittal transcallosal craniotomy. Surgical treatment of suprasellar arachnoid cysts. Can Oper Room Nurs J 3(5):6–12

Naidich TP, McLone DG, Radkowski MA (1985) Intracranial arachnoid cysts. Pediatr Neurosci 12(2):112–122

Nakase H, Hisanaga M, Hashimoto S, Imanishi M, Utsumi S (1988) Intraventricular arachnoid cyst. Report of two cases. J Neurosurg 68(3):482–486. https://doi.org/10.3171/jns.1988.68.3.0482

Navarro R, Gil-Parra R, Reitman AJ, Olavarria G, Grant JA, Tomita T (2006) Endoscopic third Ventriculostomy in children: early and late complications and their avoidance. Childs Nerv Syst 22(5):506–513. https://doi.org/10.1007/s00381-005-0031-1

Nowosławska E, Polis L, Kaniewska D, Mikołajczyk W, Krawczyk J, Szymański W, Zakrzewski K, Podciechowska J, Polis B (2006) Neuroendoscopic techniques in the treatment of arachnoid cysts in children and comparison with other operative methods. Childs Nerv Syst 22(6):599–604. https://doi.org/10.1007/s00381-006-0068-9

Oberbauer RW, Haase J, Pucher R (1992) Arachnoid cysts in children: a European co-operative study. Childs Nerv Syst 8:281–286

Oertel JMK, Baldauf J, Schroeder HWS, Gaab MR (2009) Endoscopic cystoventriculostomy for treatment of paraxial arachnoid cysts. J Neurosurg 110(4):792–799. https://doi.org/10.3171/2008.7.JNS0841

Ogiwara H, Morota N, Joko M, Hirota K (2011) Endoscopic fenestrations for suprasellar arachnoid cysts. J Neurosurg Pediatr 8(5):484–488. https://doi.org/10.3171/2011.8.PEDS11226

Ohnishi Y-i, Fujimoto Y, Taniguchi M, Tsuzuki T, Taki T (2007) Neuroendoscopically assisted cyst-cisternal shunting for a quadrigeminal arachnoid cyst causing typical trigeminal neuralgia. Minim Invasive Neurosurg 50(2):124–127. https://doi.org/10.1055/s-2007-982507

Okano A, Ogiwara H (2016) The effectiveness of microsurgical fenestration for middle fossa arachnoid cysts in children. Childs Nerv Syst 32(1):153–158. https://doi.org/10.1007/s00381-015-2908-y

Osuagwu F, Fagel N, Salamon N, Lazareff J (2009) Symptomatic prepontine cistern flattening after an arachnoid cyst shunting: a report of 2 cases. Surg Neurol 72(4):418–421. https://doi.org/10.1016/j.surneu.2008.09.017

Özek MM, Urgun K (2013) Neuroendoscopic management of suprasellar arachnoid cysts. World Neurosurg 79(2):S19.e13–S19.e18. https://doi.org/10.1016/j.wneu.2012.02.011

Paladino J, Rotim K, Heinrich Z (1998) Neuroendoscopic fenestration of arachnoid cysts. Minim Invasive Neurosurg 41(3):137–140. https://doi.org/10.1055/s-2008-1052029

Pascual-Castroviejo I, Roche MC, Martinez Bermejo A, Arcas J, García-Blázquez M (1991) Primary intracranial arachnoidal cysts. A study of 67 childhood cases. Childs Nerv Syst 7:257–263

Payner TD, Prenger E, Berger TS, Crone KR (1994) Acquired chiari malformations: incidence, diagnosis, and management. Neurosurgery 34(3):429–434; discussion 434

Pelletier J, Milandre L, Péragut JC, Cronqvist S (1990) Intraventricular choroid plexus 'arachnoid' cyst. MRI findings. Neuroradiology 32(6):523–525

Pierre-Kahn A, Sonigo P (2003) Malformative intracranial cysts: diagnosis and outcome. Childs Nerv Syst 19 (7–8):477–483. https://doi.org/10.1007/s00381-003-0773-6

Pierre-Kahn A, Sachis M, Hirsch JF (1974) Hydrocéphalie et Kyste Arachnoïdien Supra-Sellaire. Neurochirurgie 20:179–190

Pierre-Kahn A, Capelle L, Sainte-Rose C, Renier D, Hoppe-Hirsch E, Hirsch JF (1990a) Treatment of suprasellar arachnoid cysts by percutaneous transfrontal ventriculocystostomy. Apropos of a series of 17 cases. Neuro-Chirurgie 36(6):370–377

Pierre-Kahn A, Capelle L, Brauner R, Sainte-Rose C, Renier D, Rappaport R, Hirsch J-F (1990b) Presentation and management of suprasellar arachnoid cysts: review of 20 cases. J Neurosurg 73(3):355–359

Pierre-Kahn A, Carpentier A, Parisot D, Cinalli G, Zerah M, Renier D, Sainte-Rose C (2002) Treatment of intracranial cysts in children: peritoneal derivation or endoscopic fenestration? Neuro-Chirurgie 48(4): 327–338

Pilu G, Falco P, Perolo A, Sandri F, Cocchi G, Ancora G, Bovicelli L (1997) Differential diagnosis and outcome of fetal intracranial hypoechoic lesions: report of 21 cases. Ultrasound Obstet Gynecol 9(4):229–236. https://doi.org/10.1046/j.1469-0705.1997.09040229.x

Pradilla G, Jallo G (2007) Arachnoid cysts: case series and review of the literature. Neurosurg Focus 22(2):E7

Pulido-Rivas P, Villarejo-Ortega FJ, Cordobés-Tapia F, Pascual Martín-Gamero A, Pérez-Díaz C (2005) Surgical treatment of symptomatic arachnoid cysts in children. Rev Neurol 41(7):385–390

Raffel C, McComb JG (1988) To shunt or to fenestrate: which is the best surgical treatment for arachnoid cysts in pediatric patients? Neurosurgery 23(3):338–342

Raimondi AJ, Shimoji T, Gutierrez FA (1980) Suprasellar cysts: surgical treatment and results. Childs Brain 7(2):57–72

Rekate HL (1993) Classification of slit-ventricle syndromes using intracranial pressure monitoring. Pediatr Neurosurg 19(1):15–20

Rengachary SS, Watanabe I (1981) Ultrastructure and pathogenesis of intracranial arachnoid cysts. J Neuropathol Exp Neurol 40(1):61–83

Rengachary SS, Watanabe I, Brackett CE (1978) Pathogenesis of intracranial arachnoid cysts. Surg Neurol 9(2):139–144

Rhoton AL (2000) The posterior fossa cisterns. Neurosurgery 47(Suppl 3):S287–S297

Rizk E, Chern JJ, Tagayun C, Shane Tubbs R, Hankinson T, Rozzelle C, Jerry Oakes W, Blount JP, Wellons JC (2013) Institutional experience of endoscopic suprasellar arachnoid cyst fenestration. Childs Nerv Syst 29(8):1345–1347. https://doi.org/10.1007/s00381-013-2032-9

Robinson RG (1964) The temporal lobe agenesis syndrome. Brain 87:87–106

Samii M, Carvalho GA, Schuhmann MU, Matthies C (1999) Arachnoid cysts of the posterior fossa. Surg Neurol 51(4):376–382. https://doi.org/10.1016/S0090-3019(98)00095-0

Santamarta D, Aguas J, Ferrer E (1995) The natural history of arachnoid cysts: endoscopic and cine-mode MRI evidence of a slit-valve mechanism. Minim Invasive Neurosurg 28:133–137

Sato K, Shimoji T, Yaguchi K, Sumie H, Kuru Y, Isshii S (1983) Middle fossa arachnoid cysts: clinical, neuroradiological, and surgical features. Childs Brain 10:301–316

Sato H, Sato N, Katayama S, Tamaki N, Matsumoto S (1991) Effective shunt-independent treatment for primary middle fossa arachnoid cyst. Childs Nerv Syst 7:375–381

Schroeder HWS, Gaab MR (1997) Endoscopic observation of a slit-valve mechanism in a suprasellar prepontine arachnoid cyst: case report. Neurosurgery 40:198–200

Schroeder HW, Gaab MR, Niendorf WR (1996) Neuroendoscopic approach to arachnoid cysts. J Neurosurg 85(2):293–298. https://doi.org/10.3171/jns.1996.85.2.0293

Schuhmann MU, Tatagiba M, Hader C, Brandis A, Samii M (2000) Ectopic choroid plexus within a juvenile arachnoid cyst of the cerebellopontine angle: cause of cyst formation or reason of cyst growth. Pediatr Neurosurg 32:73–76

Shim K-W, Lee Y-H, Park E-K, Park Y-S, Choi J-U, Kim D-S (2009) Treatment option for arachnoid cysts. Childs Nerv Syst 25(11):1459–1466. https://doi.org/10.1007/s00381-009-0930-7

Shou X, Zhao Y, Li S, Wang Y (2015) Ventriculoscopic surgery for arachnoid cysts in the lateral ventricle: a comparative study of 21 consecutive cases. Int J Clin Exp Med 8(11):20787–20795

Silav G, Sarı R, Bölükbaşı FH, Altaş M, Işık N, Elmacı İ (2015) Microsurgical fenestration and cystoperitoneal shunt through preauricular subtemporal keyhole craniotomy for the treatment of symptomatic middle fossa arachnoid cysts in children. Childs Nerv Syst 31(1): 87–93. https://doi.org/10.1007/s00381-014-2530-4

Sommer IE, Smit LM (1997) Congenital supratentorial arachnoidal and giant cysts in children: a clinical study with arguments for a conservative approach. Childs Nerv Syst 13(1):8–12. https://doi.org/10.1007/s003810050030

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(2):163–172. https://doi.org/10.1007/s00381-009-0952-1

Spennato P, Ruggiero C, Aliberti F, Buonocore MC, Trischitta V, Cinalli G (2013) Interhemispheric and quadrigeminal cysts. World Neurosurg 79(2):S20.e1–S20.e7. https://doi.org/10.1016/j.wneu.2012.02.010

Sprung C, Mauersberger W (1979) Value of computed tomography for the diagnosis of arachnoid cysts and assessment of surgical treatment. Acta Neurochir Suppl 28(2):619–626

Starkman SP, Brown TC, Linell EA (1958) Cerebral arachnoid cysts. J Neuropathol Exp Neurol 17:484–500

Sugata S, Niiro M, Tanioka K, Yano T, Kuratsu J-i (2003) Huge arachnoid cyst of the posterior fossa with cerebellar tentorium dysplasia associated with juvenile polyposis. Pediatr Neurosurg 38:253–257. https://doi.org/10.1159/000069819

Sunami K, Saeki N, Sunada S, Hoshi S, Murai H, Kubota M, Takanashi J-i, Yamaura A (2002) Slit ventricle syndrome after cyst-peritoneal shunting for temporal arachnoid cyst in children – a clinical entity difficult to detect on neuroimaging study. Brain Dev 24(8):776–779

Talamonti G, D'Aliberti G, Picano M, Debernardi A, Collice M (2011) Intracranial cysts containing cerebrospinal fluid-like fluid: results of endoscopic neurosurgery in a series of 64 consecutive cases. Neurosurgery 68(3):788–803; discussion 803. https://doi.org/10.1227/NEU.0b013e318207ac91

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. https://doi.org/10.1007/s00381-003-0724-2

Tamburrini G, D'Angelo L, Paternoster G, Massimi L, Caldarelli M, Di Rocco C (2007) Endoscopic management of intra and paraventricular CSF cysts. Childs Nerv Syst 23(6):645–651. https://doi.org/10.1007/s00381-007-0327-4

Tamburrini G, Fabbro MD, Del Fabbro M, Di Rocco C (2008) Sylvian fissure arachnoid cysts: a survey on their diagnostic workout and practical management. Childs Nerv Syst 24(5):593–604. https://doi.org/10.1007/s00381-008-0585-9

Trowbridge WV, French JD (1952) Benign arachnoid cysts of the posterior fossa. J Neurosurg 9:398–404

Tsuruda JS, Chew WM, Moseley ME, Norman D (1990) Diffusion-weighted MR imaging of the brain: value of differentiating between extra axial cysts and epidermoid tumors. AJR Am J Roentgenol 155(5):1059–1065; discussion 1066–68. https://doi.org/10.2214/ajr.155.5.2120936

Ulu MO, Kafadar AM, Dashti R, İşler C, Uludağ S, Erdinçler P (2010) Treatment of symptomatic interhemispheric arachnoid cysts by cystoperitoneal shunting. J Clin Neurosci 17(6):700–705. https://doi.org/10.1016/j.jocn.2009.09.029

Vaquero J, Carrillo R, Cabezudo JM, Nombela L, Bravo G (1981) Arachnoid cysts of the posterior fossa. Surg Neurol 16(2):117–121

Vergani P, Locatelli A, Piccoli MG, Ceruti P, Patanè L, Paterlini G, Ghidini A (1999) Ultrasonographic differential diagnosis of fetal intracranial interhemispheric cysts. Am J Obstet Gynecol 180(2):423–428. https://doi.org/10.1016/S0002-9378(99)70226-7

Volpe JJ (2008) Hypoxic-ischemic encephalopathy: neuropathology and pathogenesis. In: Neurology of the newborn, 5th edn. Saunders, Philadelphia, pp 347–399

Wang JC, Heier L, Souweidane MM (2004) Advances in the endoscopic management of suprasellar arachnoid cysts in children. J Neurosurg 100(5 Suppl Pediatrics):418–426. https://doi.org/10.3171/ped.2004.100.5.0418

Weber F, Knopf H (2006) Incidental findings in magnetic resonance imaging of the brains of healthy young men. J Neurol Sci 240(1–2):81–84. https://doi.org/10.1016/j.jns.2005.09.008

Wester K (1999) Peculiarities of intracranial arachnoid cysts: location, sidedness, and sex distribution in 126 consecutive patients. Neurosurgery 45:775–779

Young JN, Oakes WJ, Hatten HP (1992) Dorsal third ventricular cyst: an entity distinct from holoprosencephaly. J Neurosurg 77(4):556–561. https://doi.org/10.3171/jns.1992.77.4.0556

Zada G, Krieger MD, McNatt SA, Bowen I, Gordon McComb J (2007) Pathogenesis and treatment of intracranial arachnoid cysts in pediatric patients younger than 2 years of age. Neurosurg Focus 22(2):E1. https://doi.org/10.3171/foc.2007.22.2.1

Zhao P, Wang X, Li C, Gui S, Zong X, Zhang Y (2013) The effectiveness of neuroendoscopic versus non-neuroendoscopic procedures in the treatment of lateral ventricular cysts: a retrospective medical record review study. BMC Neurol 13:59. https://doi.org/10.1186/1471-2377-13-59