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RESEARCH

Classification of the relationship between suprasellar arachnoid cyst and hydrocephalus based on treatment modalities: shunting versus neuroendoscopic approaches

Hongbin Cao[1] · Genrui Guo[2] · Wenjing Wu[1] · Zhenghai Cheng[1]

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Abstract

Purpose Children diagnosed with suprasellar arachnoid cysts often concurrently have hydrocephalus. This study aims to classify the relationship between suprasellar arachnoid cysts and hydrocephalus, discussing surgical strategies—shunting or neuroendoscopic approaches—and their sequence, based on this classification.

Methods A retrospective analysis was conducted on 14 patients diagnosed with suprasellar arachnoid cysts and hydrocephalus, treated surgically by the first author between January 2016 and December 2020. Clinical features, radiological findings, surgical strategies, and outcomes were reviewed. The classification of the relationship between the suprasellar arachnoid cysts and hydrocephalus was developed and illustrated with specific cases. Recommendations for future surgical management based on this classification are provided.

Results We classified the relationship between suprasellar arachnoid cysts and hydrocephalus into three categories. SACH-R1, the direct type, represents cases where the cysts cause obstructive hydrocephalus. Here, neuroendoscopic ventriculocystocisternostomy (VCC) effectively treats both conditions. SACH-R2, the juxtaposed type, involves concurrent occurrences of cysts and hydrocephalus without a causative link. This is further subdivided into SACH-R2a, where acute progressive communicating hydrocephalus coexists with the cyst, initially managed with a ventriculoperitoneal shunt, followed by VCC upon stabilization of hydrocephalus; and SACH-R2b, where the cyst coexists with chronic stable communicating hydrocephalus, first addressed with VCC, followed by monitoring and potential secondary shunting if needed. Key factors differentiating SACH-R2a from SACH-R2b include the patient's age, imaging signs of fourth ventricle and cisterna magna enlargement, and the rapid progression or chronic stability and severity of hydrocephalus symptoms. SACH-R3, the reverse type, describes scenarios where shunting for hydrocephalus leads to the development or enlargement of the cyst, managed via neuroendoscopic VCC with precautions to prevent infections in existing shunt systems.

Conclusion The simultaneous presence of suprasellar arachnoid cysts and hydrocephalus requires a nuanced understanding of their complex relationship for optimal surgical intervention. The analysis and classification of their relationship are crucial for determining appropriate surgical approaches, including the choice and sequence of shunting and neuroendoscopic techniques. Treatment should be tailored to the specific type identified, rather than blindly opting for neuroendoscopy. Particularly for SACH-R2a cases, we recommend initial ventriculoperitoneal shunting.

Keywords Suprasellar arachnoid cysts · Hydrocephalus · Neuroendoscopy · Ventriculocystocisternostomy · Ventriculoperitoneal shunting · Classification · Relationship

Introduction

Suprasellar arachnoid cysts in children are a specific and rare condition [1], accounting for 10% of intracranial arachnoid cysts and 1% of intracranial lesions [2]. These cysts often coexist with hydrocephalus in children. With advancements in neuroimaging techniques such as cranial MRI and prenatal ultrasound, there has been an increase in the detection

✉ Hongbin Cao
hugh.cao@163.com

[1] Department of Neurosurgery, Hebei Children's Hospital, Hebei Medical University, Shijiazhuang, Hebei, China

[2] Department of Anesthesiology, Hebei Children's Hospital, Hebei Medical University, Shijiazhuang, Hebei, China

of suprasellar arachnoid cysts [3]. Moreover, significant progress in neuroendoscopic techniques has established suprasellar arachnoid cysts as one of the primary surgical indications for neuroendoscopy. The endoscopic treatment of these cysts has become a widely discussed topic and is well recognized within the neurosurgical community and the general public [1, 4, 5]. However, this often leads to a misconception where the focus is primarily on the cyst itself and the favorable outcomes of neuroendoscopic treatment, overlooking the analysis of the relationship between suprasellar arachnoid cysts and concurrent hydrocephalus. Understanding this relationship and its impact on treatment decisions and patient prognosis is crucial, as it may result in neglecting the timely and effective management of associated hydrocephalus, potentially leading to treatment delays.

Miyajima et al. postulated two different types of suprasellar arachnoid cysts based on radiological and anatomical findings: a noncommunicating intraarachnoid cyst of the diencephalic membrane of Liliequist and a communicating cyst, which is a cystic dilation of the interpeduncular cistern [6]. However, no clear distinctions concerning clinical presentations and therapeutic strategies have been proposed according to these subtypes [7]. André A and Di Rocco F proposed a novel classification of SAC in 2015, established on clinical and radiological findings, modulating the therapeutic approach based on their experience. They classified suprasellar arachnoid cysts into three types: SAC-1 may come from an expansion of the diencephalic leaf of the Liliequist membrane; SAC-2 shows a dilatation of the interpeduncular cistern and corresponds to a defect of the mesencephalic leaf of the Liliequist membrane; SAC-3 corresponds to the asymmetrical forms expanding to other subarachnoid spaces. Surgical treatment is not always necessary [7]. This new classification is of great clinical significance for the management of this rare and heterogeneous disease [3, 8].

However, the classification by André A and Di Rocco F did not address the relationship between suprasellar arachnoid cysts and hydrocephalus. In clinical practice, we have found that the choice of surgical approach for this disease largely depends on the management of hydrocephalus. Therefore, based on treatment modalities, we propose a new concise classification of the relationship between suprasellar arachnoid cysts and hydrocephalus, elucidating each type with specific cases, and offering recommendations for surgical management of future cases based on this classification.

Methods

A retrospective analysis was conducted on children with suprasellar arachnoid cysts who underwent surgery between January 2016 and December 2020. Cases without concurrent hydrocephalus were excluded from the study, resulting in a total of 14 cases included in the analysis. Based on the clinical characteristics, imaging findings, surgical strategies, and prognosis of these cases, a classification of the relationship between the suprasellar arachnoid cysts and hydrocephalus was developed and illustrated with specific cases. Recommendations for future surgical management based on this classification are provided. The primary employed surgical strategies were neuroendoscopic ventriculocystocisternostomy (VCC) and ventriculoperitoneal shunting.

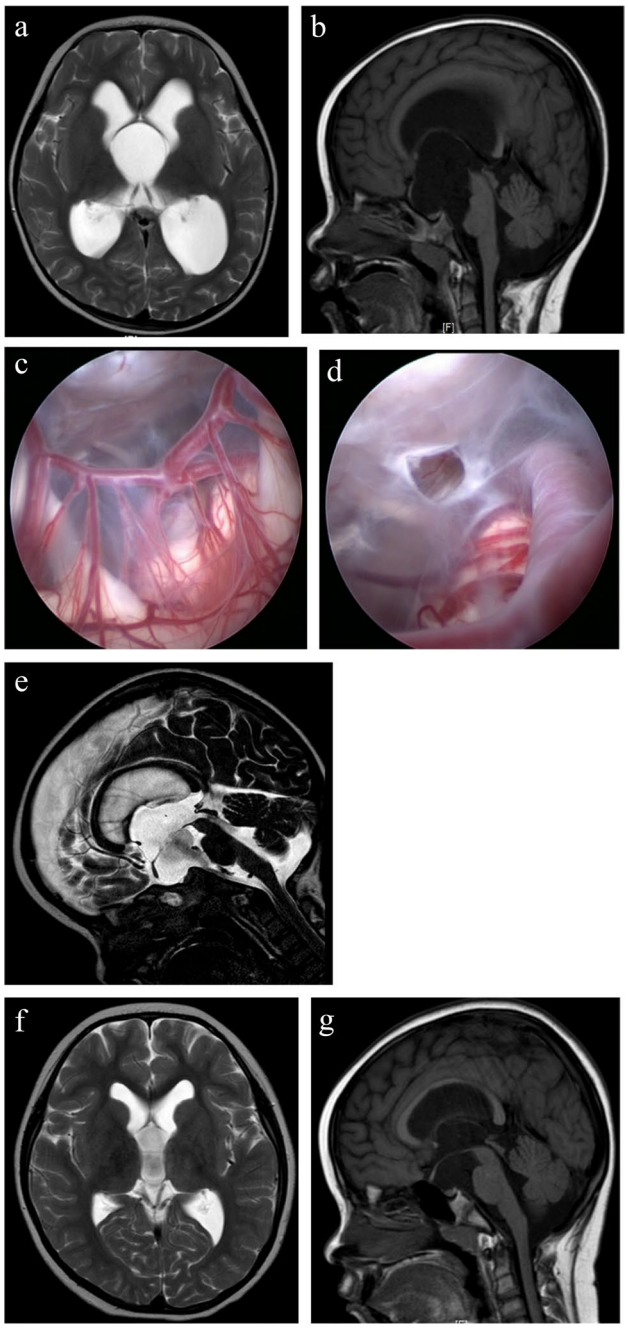
Regarding neuroendoscopic surgery for suprasellar arachnoid cysts, the following techniques and considerations are highlighted: In all cases, ventriculocystocisternostomy (VCC) was performed rather than just ventriculocystostomy (VC). The use of electrocoagulation on the cyst wall was minimized to avoid unnecessary thermal damage to surrounding neural tissues and was only employed conservatively when there was hemorrhage from small vessels within the cyst wall. The opening on the superior wall of the cyst was created and enlarged by mechanical shearing with a scissor. These techniques were employed to minimize the risk of postoperative endocrine symptoms caused by thermal damage to surrounding neural tissues [9]. The opening might close following the reduction of the cysts [10], and these techniques can minimize the contraction and scar closure of the opening. For the fenestration on the inferior wall of the cyst, a combination of a scissor, a grasping forcep, and a Fogarty balloon was employed. The procedure was deemed satisfactory when the dura on basilar clivus and the deep and profound space of prepontine cistern were visualized.

All shunt procedures were ventriculoperitoneal shunts. The ventricular catheter was placed into the frontal horn of the lateral ventricle via a trans-occipital approach. Programmable shunt valves were utilized in all procedures and adjusted postoperatively based on the patient's clinical status and follow-up imaging.

Results

The relationship between suprasellar arachnoid cysts and hydrocephalus was classified into 3 categories: SACH-R1, the direct type; SACH-R2, the juxtaposed type, which can be subdivided into SACH-R2a and SACH-R2b; and SACH-R3, the reverse type. They are described below and illustrated with cases.

SACH-R1, the direct type, also known as the classic type, represents cases where the cysts cause obstructive hydrocephalus (Fig. 1). On the midline sagittal view of cranial MRI, a large suprasellar arachnoid cyst intrudes into the third ventricle, occupying almost all of its space, and obstructing the foramen of Monro and the aqueduct of Sylvius. Hydrocephalus is characterized by enlargement of



◂**Fig.1** Case 1 for SACH-R1 illustration. A 2-year and 9-month-old female child presented with delayed motor skills and was prone to stumbling. After a head trauma, a CT scan revealed hydrocephalus, and further cranial MRI indicated a suprasellar arachnoid cyst coexisting with hydrocephalus. The sagittal MRI image confirmed the SACH-R1 classification. The child underwent neuroendoscopic VCC. Postoperatively, the suprasellar arachnoid cyst decreased in size, and the floor of the third ventricle returned to a more normal position. A follow-up MRI three years after surgery showed resolution of hydrocephalus and stabilization of the suprasellar arachnoid cyst. Currently, eight years post-surgery, the child exhibits normal neurological development and leads a normal daily life. **a** and **b** Preoperative MRI shows a large suprasellar arachnoid cyst intruding into and occupying almost all of the third ventricle, obstructing the foramen of Monro and the aqueduct of Sylvius, leading to obstructive hydrocephalus. However, the fourth ventricle and cisterna magna are not enlarged. **c** and **d** During neuroendoscopic VCC, after fenestration of the superior cyst wall, the circle of Willis was clearly visible. Fenestration of the inferior wall established communication between the cyst and the prepontine cistern. **e** Ten days post-VCC, the suprasellar arachnoid cyst had reduced, with visible flow voids on MRI at the fenestration sites. **f** and **g** Three years post-VCC, follow-up MRI shows a nearly normal ventricular system and a stable suprasellar arachnoid cyst

the bilateral lateral ventricles and third ventricle, without enlargement of the fourth ventricle and cisterna magna. Symptoms of hydrocephalus and increased intracranial pressure are relatively mild. Treatment for SACH-R1 typically involves neuroendoscopic VCC, which often leads to cyst reduction, alleviation of obstructive factors, and improvement in hydrocephalus. Further ventriculoperitoneal shunting is usually unnecessary.

SACH-R2, the juxtaposed type, involves concurrent occurrences of cysts and hydrocephalus without a causative link. On the midline sagittal view of cranial MRI, a small to medium-sized suprasellar arachnoid cyst may extend into or partially occupy half to two-thirds of the third ventricle, without obstructing the foramen of Monro and the aqueduct of Sylvius. In some instances, it may fully occupy the third ventricle, causing some degree of obstructive factors. Hydrocephalus is characterized by enlargement of all four ventricles and the cisterna magna. The fourth ventricle communicates with the enlarged cistern magna via its trumpet-like dilatation of the outlet, serving as a significant radiological criterion. Based on the severity and urgency of hydrocephalus symptoms, it can be further classified into SACH-R2a and SACH-R2b.

SACH-R2a involves acute progressive communicating hydrocephalus (Figs. 2 and 3). The recommended surgical strategy is that ventriculoperitoneal shunting is initially performed to manage the communicating hydrocephalus, followed by neuroendoscopic VCC in a subsequent surgery once the hydrocephalus condition stabilizes. Typically, the duration between the two surgeries ranges from several months to two years.

SACH-R2b involves chronic, relatively stable communicating hydrocephalus (Fig. 4). Treatment typically begins with neuroendoscopic VCC, followed by close monitoring of disease progression. Depending on the development of the hydrocephalus, a decision may later be made either to perform or not to perform a secondary surgery involving ventriculoperitoneal shunting.

It should be noted that cases featuring a large suprasellar arachnoid cyst occupying almost all of the third ventricular space, accompanied by chronic, relatively stable communicating hydrocephalus, are also classified under SACH-R2b. The initial choice of treatment in these cases is neuroendoscopic VCC.

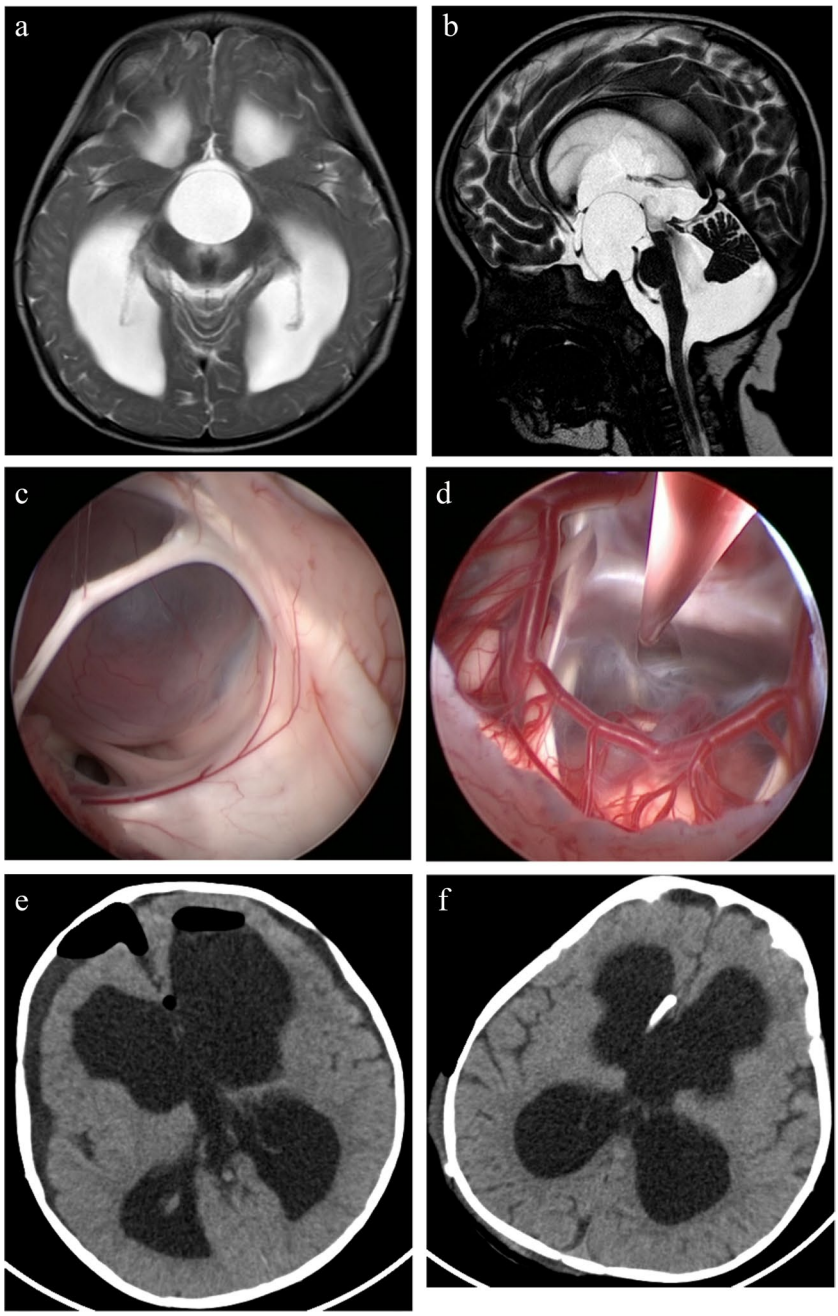
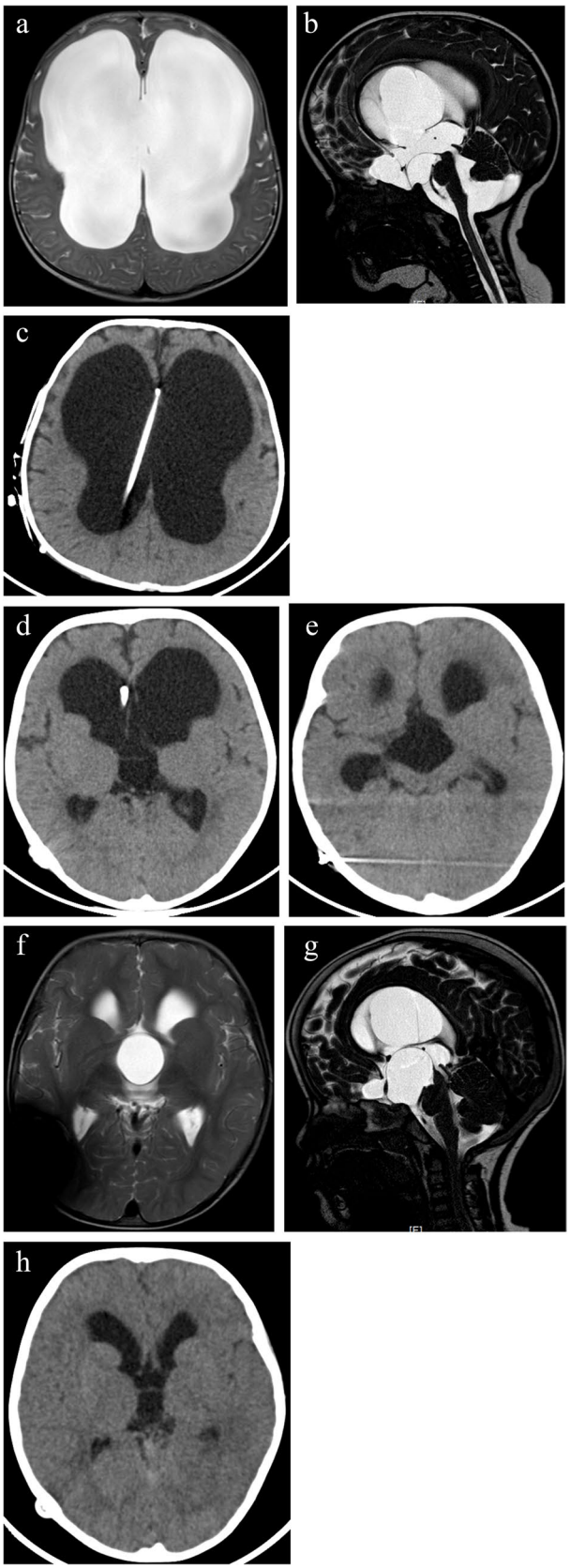


Fig. 2 Case 2 for SACH-R2a illustration. A 9-month-old female child presented with delayed motor development, an enlarged head circumference, and a bulging anterior fontanelle for three months. Cranial MRI revealed a suprasellar arachnoid cyst coexisting with hydrocephalus, classified as SACH-R2a on the sagittal view. Initially, neuroendoscopic VCC was performed, which significantly improved clinical symptoms. However, one month postoperatively, the condition reversed, necessitating subsequent ventriculoperitoneal shunting. The child's condition gradually stabilized following shunting. After seven years of follow-up, both the hydrocephalus and the intracranial cyst remain stable. The child exhibited bilateral lower limb spasticity and, after years of rehabilitation, is now able to walk. **a** and **b** Preoperative MRI shows a medium-sized suprasellar arachnoid cyst extending upward to the upper part of the third ventricle, without obstructing the foramen of Monro and the aqueduct of Sylvius. The enlarged fourth ventricle communicated with the enlarged cistern magna via its trumpet-like dilatation of the outlet. **c** and **d** Neuroendoscopic VCC was initially performed successfully. During the surgery, the foramen of Monro and the aqueduct of Sylvius were found to be unobstructed. **e** Postoperative cranial CT showed an improvement in hydrocephalus, accompanied by alleviation of the clinical symptoms. **f** One month post-VCC, intracranial pressure increased again, the anterior fontanelle bulged, necessitating a ventriculoperitoneal shunting procedure



◂**Fig. 3** Case 3 for SACH-R2a illustration. A 6-month-old male child presented with an enlarged head circumference, bulging fontanelle, setting-sun sign, and delayed motor development. Cranial MRI revealed a small suprasellar arachnoid cyst coexisting with severe hydrocephalus, classified as SACH-R2a on the sagittal view. Initially, ventriculoperitoneal shunting was performed, significantly improving clinical symptoms. Over subsequent follow-ups, the hydrocephalus gradually improved, while the suprasellar cyst progressively enlarged. 1.5 years post-shunting, the cyst had significantly enlarged, causing obstruction, which necessitated neuroendoscopic VCC. Two years post-VCC, follow-up cranial MRI showed regression of the suprasellar arachnoid cyst. Currently, 7 years post-shunting and 5.5 years post-VCC, the child is attending school and living a normal life. **a** and **b** Preoperative MRI shows severe hydrocephalus with a very thin cerebral cortex, and a small-sized suprasellar arachnoid cyst extending upward to the lower part of the third ventricle, without obstructing the foramen of Monro and the aqueduct of Sylvius. The enlarged fourth ventricle communicated with the enlarged cistern magna via its trumpet-like dilatation of the outlet. **c** Ventriculoperitoneal shunting was initially performed. One month post-shunting, cranial CT showed the well-positioned shunt catheter and improved hydrocephalus. **d** and **e** Six months post-shunting, cranial CT showed further reduction in the ventricular system but an enlargement of the suprasellar cyst. **f** and **g** 1.5 years post-shunting, cranial MRI showed significant enlargement of the suprasellar cyst, causing obstruction of the third ventricle and the foramen of Monro. The cerebral cortex had significantly thickened, prompting neuroendoscopic VCC. **h** Two years post-VCC, cranial CT showed regression of the suprasellar arachnoid cyst and stabilization of the hydrocephalus

The primary distinguishing factors between SACH-R2a and SACH-R2b are the age of the child and the severity and urgency of the hydrocephalus symptoms. SACH-R2a most commonly occurs in infants under one year old and is characterized by acute, progressive communicating hydrocephalus. In contrast, SACH-R2b more frequently occurs in infants and toddlers over one year old, presenting with chronic, relatively stable hydrocephalus.

SACH-R3, the reverse type, represents cases where ventriculoperitoneal shunting for hydrocephalus leads to the development and/or enlargement of a suprasellar arachnoid cyst (Fig. 5). On the midline sagittal view of cranial MRI, no suprasellar arachnoid cyst was observed, or only an enlarged suprasellar cistern was present before the shunting. However, years after the shunting procedure, a large suprasellar arachnoid cyst gradually appeared and grew. This is believed to result from an imbalance of pressures between the ventricles and the suprasellar cistern, where the ventricular pressure falls below that of the suprasellar cistern. Over time, this pressure imbalance leads to the formation and subsequent enlargement of the suprasellar arachnoid cyst. As the cyst enlarges, it can occupy the entire third ventricle and even extend into the lateral ventricles, causing obstructive hydrocephalus or dysfunction of the shunt. Treatment typically involves neuroendoscopic VCC, which is effective in managing these cases. During the surgery, strict precautions against infection are necessary to protect the existing shunt system.

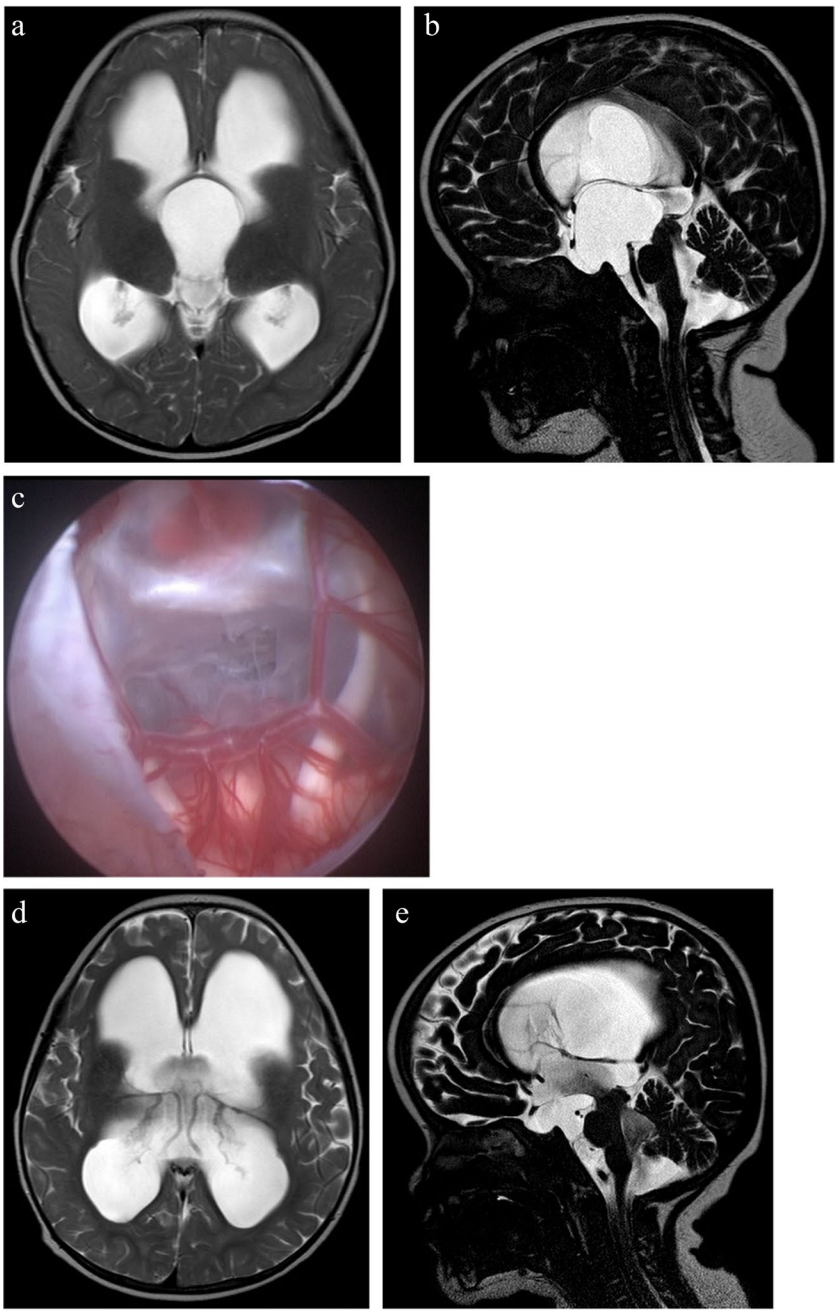


Fig. 4 Case 4 for SACH-R2b illustration. A 14-month-old female child presented with an enlarged head circumference and delayed motor development for three months. Cranial MRI revealed a suprasellar arachnoid cyst coexisting with hydrocephalus, classified as SACH-R2b on the sagittal view. Initially, neuroendoscopic VCC was performed, leading to a reduction of the suprasellar arachnoid cyst; however, the child's communicating hydrocephalus showed no significant improvement. Subsequent cranial CT/MRI follow-ups indicated no progression of the hydrocephalus. The child experienced residual motor developmental delays, which improved with rehabilitation. Currently, after eight years of follow-up, the child's motor skills remain mildly behind her peers but are adequate for normal daily activities and schooling. **a** and **b** Preoperative MRI shows a large suprasellar arachnoid cyst intruding into the third ventricle, obstructing the foramen of Monro and the aqueduct of Sylvius, and leading to obstructive hydrocephalus. However, the enlarged fourth ventricle communicated with the enlarged cistern magna via its trumpet-like dilatation of the outlet, distinguishing this case from SACH-R1. **c** Neuroendoscopic VCC was initially performed successfully. **d** and **e** Six months post-VCC, cranial MRI showed a reduction in the suprasellar arachnoid cyst, but no significant improvement in hydrocephalus

In this case series, a total of 14 patients were included. Of these, 9 cases were classified as SACH-R1, 2 as SACH-R2a, 2 as SACH-R2b, and 1 as SACH-R3.

Discussion

Since the 1990s, the application of neuroendoscopy has become increasingly widespread [11]. Suprasellar arachnoid cysts, as a classic indication for neuroendoscopy, have been extensively reported as diseases that previously required craniotomy, stereotactic approaches, or shunting, but can now be managed with minimally invasive techniques [4, 5, 12, 13]. Suprasellar arachnoid cysts often coexist with hydrocephalus, and there has been a significant decline in the use of ventriculoperitoneal shunting for managing these conditions. However, previous reports have primarily emphasized the reduction in shunting, without sufficiently addressing that some patients still require this procedure [14]. Consequently, the subset of patients for whom neuroendoscopic management proves ineffective remains insufficiently analyzed. This leads to a critical question: For which patients is shunting unavoidable? If shunting is indeed inevitable, should ventriculoperitoneal shunting be prioritized over

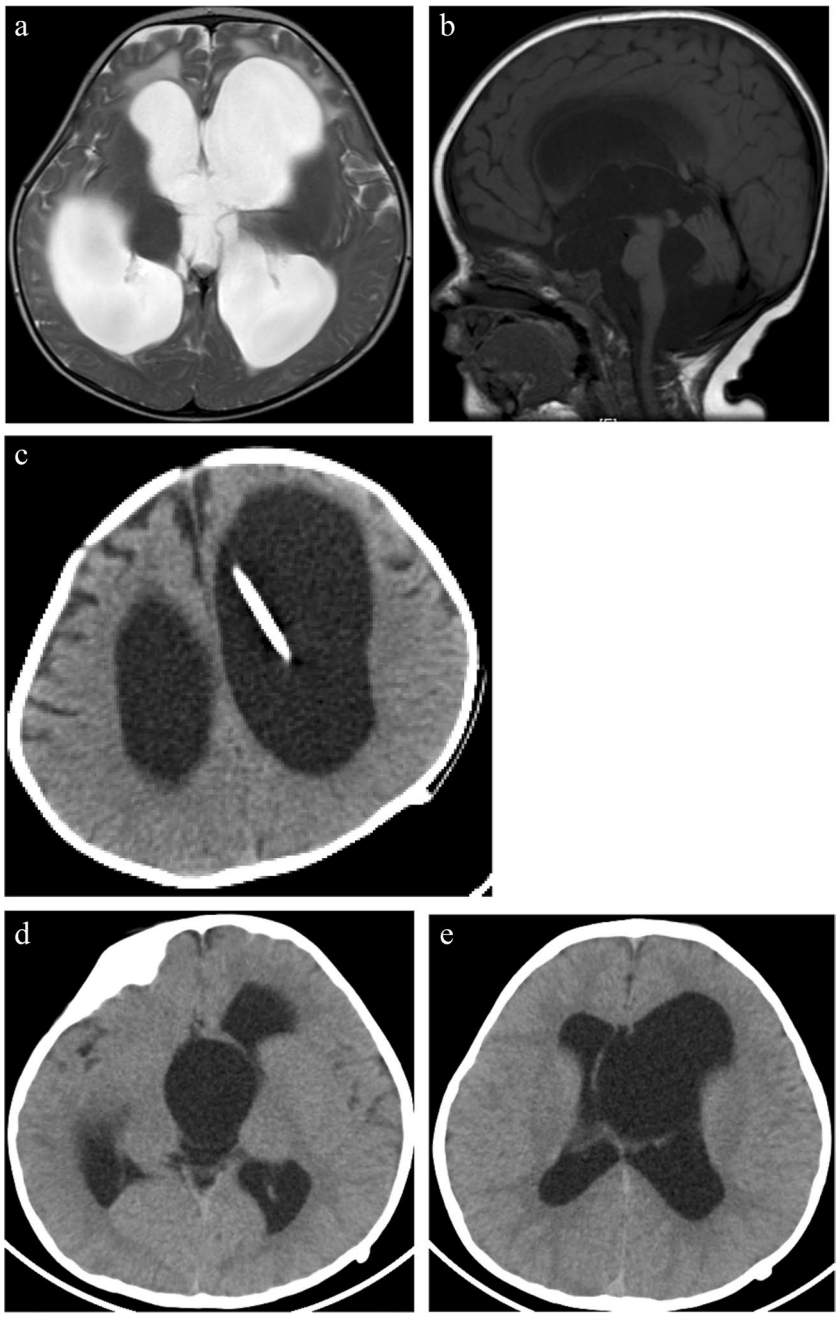


Fig. 5 Case 5 for SACH-R3 illustration. A 6-month-old female child presented with an enlarged head, measuring 49 cm in circumference, with a bulging and tense fontanelle. Cranial MRI revealed severe communicating hydrocephalus. The sagittal view showed an expanded suprasellar cistern elevating the floor of the third ventricle. Initially, ventriculoperitoneal shunting was performed, leading to significant clinical improvement and near-normal neurological development. Follow-up imaging detected the development and gradual enlargement of a suprasellar arachnoid cyst extending into the left lateral ventricle, causing shunt obstruction. Three years after shunting, neuroendoscopic VCC was conducted, relieving the shunt obstruction and leading to the gradual disappearance of the suprasellar arachnoid cyst. Currently, ten years post-shunting and seven years post-VCC, the child is attending school and living a healthy life. **a** and **b** Preoperative MRI revealed severe communicating hydrocephalus with an enlarged suprasellar cistern visible on the sagittal view. **c** Post-shunting, cranial CT confirmed the shunt catheter was well-positioned. **d** One year post-shunting, cranial CT revealed a suprasellar arachnoid cyst occupying the third ventricle. **e** Two years post-shunting, cranial CT showed the suprasellar arachnoid cyst continuing to expand upward and into the left lateral ventricle, where the shunt catheter is located. **f**, **g** and **h** Three years post-shunting, cranial MRI revealed a large suprasellar arachnoid cyst occupying the left lateral ventricle, compressing the shunt catheter against the ventricular wall and causing shunt obstruction. The sagittal view showed significant upward expansion of the suprasellar arachnoid cyst. **i** and **j** Neuroendoscopic VCC was performed. During surgery, the cyst was observed occupying the left lateral ventricle and causing compressive obstruction to the shunt catheter. Post-VCC, the cyst collapsed, eliminating the pressure on the catheter. **k** and **l** Three years post-VCC, cranial MRI showed the disappearance of the suprasellar arachnoid cyst, with the third ventricle restored to its normal morphology. The shunt in the left lateral ventricle was free from obstruction

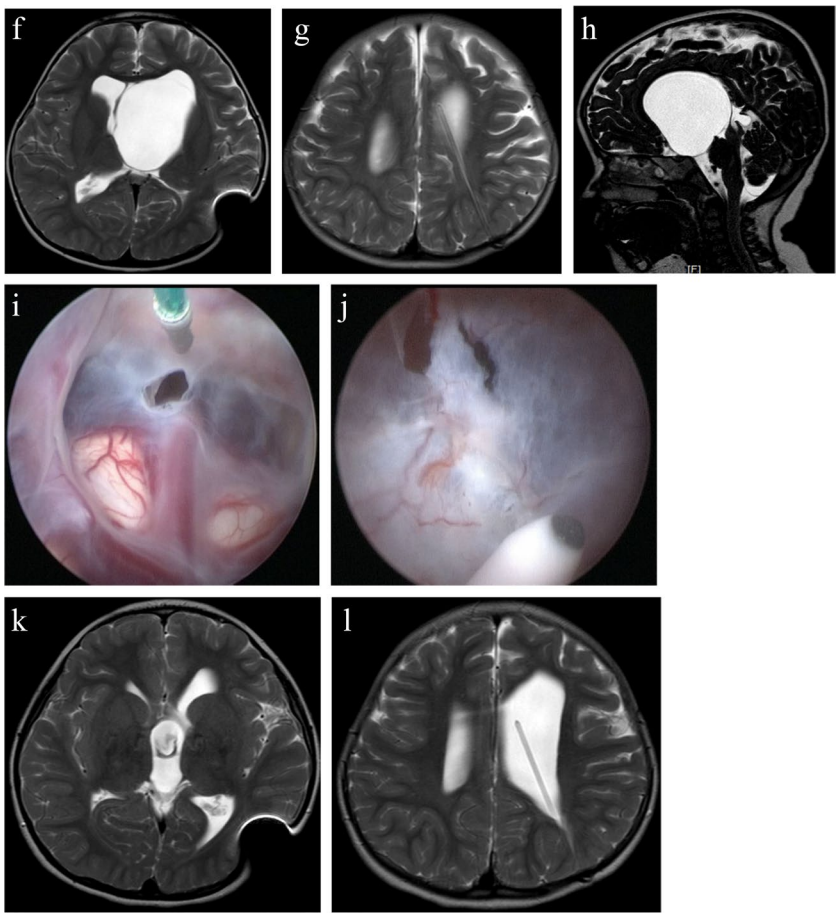


Fig. 5 (continued)

neuroendoscopic VCC to reduce the risk of progressive brain damage due to advancing hydrocephalus during the observational follow-up period? As early as 1988, when discussing whether to manage arachnoid cysts through shunting or fenestration, Raffel C pointed out that the presence or absence of concomitant hydrocephalus is a crucial factor in choosing the treatment approach [15]. This remains a significant issue in today's context, where the endoscopic treatment of suprasellar arachnoid cysts is extensively documented.

André A and Di Rocco F proposed a new classification of suprasellar arachnoid cysts (SAC) based on prognosis and treatment modalities, distinguishing three main subtypes [7]. They noted that SAC-1 and SAC-3 cysts require surgical treatment, whereas SAC-2 cysts can be simply observed. Regarding the type of treatment, they pointed out that there is a limited place for open-surgery procedures. CSF diversion by shunt is considered a complementary treatment in cases of failure or persistent hydrocephalus, while endoscopic treatment is regarded as the gold standard for these lesions [7]. They reported 35 cases of suprasellar arachnoid cysts, 15 of which were accompanied by hydrocephalus, all classified as SAC-1. However, our clinical findings suggest that cysts accompanied by hydrocephalus can also be classified as SAC-2 or SAC-3. Five of their cases underwent shunting, three classified as SAC-1 and two as SAC-3, yet no further analysis was performed on these shunted cases. We believe that André A and Di Rocco F's classification provides a crucial anatomical and radiological framework that enhances our understanding of this disease and offers a preliminary guideline on whether to opt for surgical intervention or observation. However, it does not delineate which surgical strategies—shunting versus neuroendoscopic approaches—and their sequence should be chosen. We contend that the decision regarding surgical strategy should not be based solely on the classification of the suprasellar arachnoid cyst itself, but rather on the classification of the relationship between the cyst and associated hydrocephalus.

In the surgical treatment of suprasellar arachnoid cysts combined with hydrocephalus, the choice of surgical strategy—shunting or endoscopic approaches—and their sequencing is crucial. Drawing from our experience in managing such cases, we have classified the relationship between suprasellar arachnoid cysts and hydrocephalus based on treatment choice, outcome, and prognosis. We hope this

classification will be beneficial in guiding the selection of appropriate surgical strategies.

There is little need for extensive discussion about SACH-R1, in which a suprasellar arachnoid cyst causes obstructive hydrocephalus. This is the most frequently reported condition in the literature [16]. The primary diagnostic criteria, as outlined in the results section, include a cyst that occupies nearly the entire third ventricle while the fourth ventricle and cisterna magna remain normal. Neuroendoscopic VCC has shown favorable results in treating both the suprasellar arachnoid cyst and the hydrocephalus, without requiring further shunting procedures.

A thorough discussion of SACH-R2 is essential, as this category is often subject to misunderstandings. SACH-R2a typically affects infants under one year old. In such cases, suprasellar arachnoid cysts combined with acute progressive communicating hydrocephalus are characterized by significant symptoms such as large head circumference, bulging high-tension fontanels, along with delayed motor development and rapid disease progression. The cyst occupies half to two-thirds of the third ventricle without obstructing the foramen of Monro or the aqueduct of Sylvius. The enlarged fourth ventricle, communicating with the enlarged cistern magna through its trumpet-like dilatation at the outlet, is a typical feature of this subtype. While neuroendoscopic VCC effectively addresses the cyst, it fails to resolve the hydrocephalus. However, influenced by current literature and the hope for a treatment with no shunting, neurosurgeons and parents often opt for VCC as the initial treatment. Despite transient improvements post-surgery due to the release of CSF, the condition typically worsens within two weeks to a month after VCC, necessitating additional shunting procedures. In reality, avoiding shunting in this type is unrealistic. Opting for VCC first, followed by shunting if necessary, complicates the condition and treatment, and the delay in shunting could be detrimental to the recovery of damaged neurological functions. Therefore, for SACH-R2a, we recommend initially performing ventriculoperitoneal shunting to manage the communicating hydrocephalus, followed by neuroendoscopic VCC in a subsequent surgery once the hydrocephalus condition stabilizes.

This recommended strategy has proven to be reliable and effective. We illustrate this with two comparative cases of SACH-R2a. In Case 2, the large suprasellar arachnoid cyst occupied more than two-thirds of the third ventricle, combined with acute communicating hydrocephalus. Owing to the parents' hopes and insistence on a non-shunting approach, we initially chose neuroendoscopic VCC. Although the child's clinical symptoms significantly improved postoperatively, the condition progressed within a month, necessitating subsequent shunting to stabilize the situation. In Case 3, guided by our previous experience and under our strong recommendation and insistence, we first opted for shunting and scheduled a future neuroendoscopic VCC. After the shunting, the child's condition improved steadily, with VCC performed 1.5 years later. Compared to Case 2, Case 3 underwent a more predictable and controlled treatment process. Case 3 demonstrated a favorable neurological prognosis, while Case 2 exhibited bilateral lower limb spasticity and, after years of rehabilitation, is now able to walk.

SACH-R2b typically affects infants or toddlers older than one year old. In this subtype, the hydrocephalus is chronic and communicating, having potentially stabilized over the course of the disease, which results in relatively steady symptoms. Initial treatment often involves neuroendoscopic VCC, which can be augmented with choroid plexus cauterization to reduce cerebrospinal fluid production. Rigorous postoperative observation and imaging are crucial to determine the necessity of subsequent shunting. Cases in which suprasellar arachnoid cysts occupy the entire third ventricle and are accompanied by an enlarged fourth ventricle and cisterna magna should be classified into SACH-R2b rather than SACH-R1. Although the initial approach for both types is neuroendoscopic VCC, SACH-R2b requires careful monitoring and follow-up, with a significantly higher likelihood of requiring future shunting compared to SACH-R1.

SACH-R3 represents another unique scenario encountered in clinical practice, where children diagnosed with hydrocephalus, either with no suprasellar arachnoid cyst or only an enlarged suprasellar cistern, are treated by ventriculoperitoneal shunting and may develop a large suprasellar arachnoid cyst years after the surgery. We hypothesize that the mechanism involves a shunt-induced subtle imbalance of pressure between the ventricles and the suprasellar cistern, leading to the formation and gradual enlargement of the suprasellar arachnoid cyst. For instance, in Case 5, a suprasellar arachnoid cyst that developed years after shunt surgery not only filled the third ventricle but also extended into the lateral ventricle containing the shunt catheter, eventually causing obstruction of the catheter. This type can be successfully resolved with neuroendoscopic VCC. However, strict precautions are imperative to avoid infection of the shunt system. For SACH-R3, it is crucial to be proactive and communicate clearly with parents to ensure future follow-ups and surgical planning.

This classification scheme comprehensively covers the relationships between suprasellar arachnoid cysts and hydrocephalus, specifically designed to guide surgical strategy choices. Particularly for SACH-R2a, we strongly recommend that ventriculoperitoneal shunting be initially performed to manage the communicating hydrocephalus, followed by neuroendoscopic VCC in a subsequent surgery once the hydrocephalus condition stabilizes. Emphasizing this approach is increasingly important given the widespread

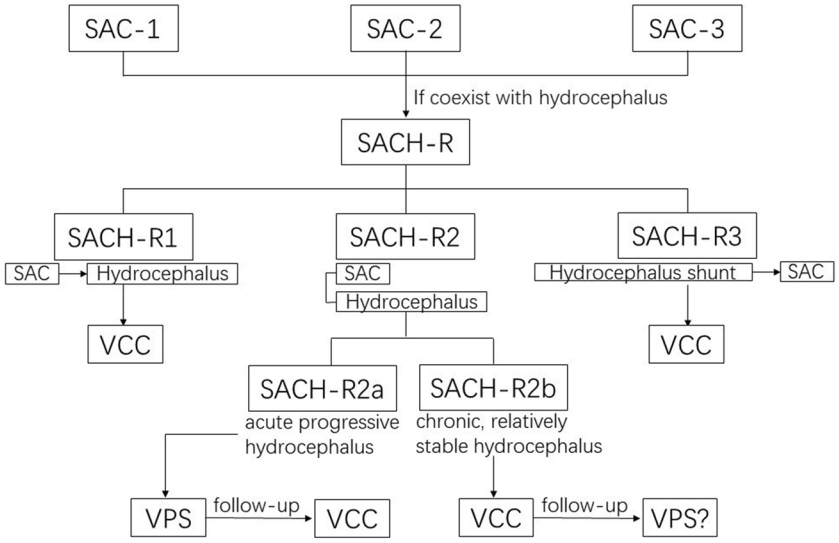


Fig. 6 The comprehensive diagram illustrating surgical strategy selection based on the classification of the relationship between suprasellar arachnoid cyst and hydrocephalus

medical and public perception of the effectiveness of neuroendoscopic treatment for suprasellar arachnoid cysts.

It is important to clarify that the classification by André A and Di Rocco F does not contradict ours but rather addresses different aspects of the condition. Their classification concentrates on the characteristics of suprasellar arachnoid cysts themselves, while ours delves into the relationship between these cysts and hydrocephalus. André A and Di Rocco F's classification provides essential and comprehensive insights into understanding the nuances of suprasellar arachnoid cysts. Each subtype they identified may potentially coexist with hydrocephalus, which seamlessly integrates into our relationship-based classification. We have synthesized both classifications into a comprehensive diagram to facilitate the selection of surgical strategies (Fig. 6).

Conclusion

The simultaneous presence of suprasellar arachnoid cysts and hydrocephalus requires a nuanced understanding of their complex relationship for optimal surgical intervention. The analysis and classification of their relationship are crucial for determining appropriate surgical approaches, including the choice and sequence of shunting and neuroendoscopic techniques. Treatment should be tailored to the specific type identified, rather than blindly opting for neuroendoscopy. Particularly for SACH-R2a cases, we recommend initial ventriculoperitoneal shunting.

Author contributions Hongbin Cao performed the surgeries, managed patient care, designed the classification scheme, and drafted the initial manuscript. Genrui Guo collected data, conducted the literature review, and assisted in drafting the final version of the manuscript. Wenjing Wu and Zhenghai Cheng assisted in the surgeries and patient care, and contributed to the Discussion section of the manuscript.

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Data availability No datasets were generated or analysed during the current study.

Declarations

Ethics approval This study was reviewed and approved by the Ethics Committee of our hospital, ensuring compliance with the institution's ethical standards and adherence to national guidelines for retrospective studies.

Competing interests The authors declare no competing interests.

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