

# Endoscopic Fenestration of Posterior Fossa Arachnoid Cysts: four Surgical Cases and Literature Review

Patricia Dastoli, Marcos Devanir S. Costa, Jardel M. Nicácio, Sergio Cavalheiro

Department of Neurology and  
Neurosurgery, Universidade Federal de  
São Paulo, São Paulo- SP, Brazil

To whom correspondence  
should be addressed: Patricia  
Dastoli, MD, PhD

e-mail:  
paty.dastoli@uol.com.br

Available at:  
<http://www.archpedneurosurg.com.br/>

*Introduction:* Posterior Fossa arachnoid cysts (PFAC) are rare lesions in the childhood. Owing to the wide variety of cystic malformations within the posterior cranial fossa, PFAC have long attracted the attention of pediatric neurosurgeons because they can produce not only neurological symptoms but also behavior and learning deficits, in addition to presenting major surgical challenges. Surgical treatment is warranted in symptomatic patients. Nowadays, neuroendoscopic techniques have established themselves as the treatment of choice of these lesions.

*Patients and Methods:* We present four cases of symptomatic PFAC which were treated using endoscopic neurosurgery. Endoscopic approaches were defined by the presence of hydrocephalus, and discussed in each case.

*Results:* Two cases of quadrigeminal plate cysts, predominantly occupying the infratentorial compartment, one case of supracerebellar cyst and one case of cerebellopontine angle were treated with good results. However, one case of quadrigeminal plate cyst developed a communicating hydrocephalus, requiring placement of cyst-ventricle peritoneal shunt, guided by the neuroendoscope.

*Discussion and Conclusion:* A literature review was performed. The clinical features, the advantages and disadvantages of several surgical treatments are discussed. Symptomatic PFAC should be treated. The relatively good results using endoscopic fenestration buttresses our use as a first-line surgery.

**Keywords:** Posterior fossa, Arachnoid cysts, Hydrocephalus, Neuroendoscopy, Children, VP shunt

## INTRODUCTION

Cystic lesions of the posterior fossa correspond to congenital malformations, to infectious, hemorrhagic complications or after excision of tumors[1]. Among the congenital malformations, there is a wide variety of cystic lesions whose differential diagnoses include posterior fossa arachnoid cysts (PFAC)[2,3,4].

Arachnoid cysts represent congenital lesions that are characterized by separation or duplication of the arachnoid membrane, which cause expansive lesions with cerebrospinal fluid (CSF) inside, compressing the structures around them[1,3,4]. In the posterior fossa, compression of the cerebellum, brainstem, and fourth ventricle accounts for the signs and symptoms of intracranial hypertension, characterized in childhood by hydrocephalus and

macrocrania. Neurological deficits such as ataxia, vertigo, tinnitus, hearing loss and swallowing disorders might also occur[3,4,5,6]. Cerebellar impairment is also connected to behavioral, psychiatric disorders and learning deficits presented by these children[7,8].

Although the indication of surgical treatment for PFAC remains controversial in cases of incidental diagnosis, there is no discussion when it comes to symptomatic patients. Currently, endoscopic fenestration has established itself as a safe and efficient treatment method for the control of hydrocephalus and the mass effect caused by PFAC.

The aim of this study is to discuss four cases of posterior fossa cysts separately located (two quadrigeminal plate cysts, one supracerebellar cyst, and one retrocerebellar



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cyst), which were treated neuroendoscopically. Our findings will be compared with literature data.

### MATERIALS AND METHODS

#### Patients and methods

Between January 2021 and March 2022, we treated four patients with diagnosis of PFAC. All four patients arrived at the emergency room and were referred to the Pediatric Neurosurgery wing of the Neurosurgery course at the Federal University of São Paulo - Escola Paulista de Medicina (UNIFESP/EPM). After the initial clinical evaluation, the patients underwent cranial tomography. Once the diagnostic hypothesis of a cystic lesion of the posterior fossa was raised, the patients underwent magnetic resonance imaging of the brain to study the anatomy of the cyst and differential diagnosis of cystic lesions of the posterior fossa. Endoscopic fenestration of the cyst was the treatment of choice. According to the anatomy of the cyst and the association with hydrocephalus, the endoscopic access was made directly through the posterior fossa or through the frontal horn of the right lateral ventricle, via the ventricular system.

The effectiveness of the treatment was confirmed by the patients' clinical improvement, and when possible, by a control MRI, showing reduction of the cyst dimensions, the presence of flow through the fenestrations of the cyst or the reduction of ventricular dimensions.

#### Case 1

A 15-year-old girl, complaining of progressive difficulty in swallowing, frequent choking and drooling. She progressed with moderately intense headache, hypoactivity and apathy, in addition to difficulty in supporting the torso. At six months of age, she underwent placement of a ventriculoperitoneal shunt (VPS) to treat hydrocephalus diagnosed after birth. Physical examination was characterized by global delay in neuropsychomotor development, without focal neurological deficits. MRI showed the presence of a giant quadrigeminal plate cyst, with compression of the brainstem and obliteration of the fourth ventricle (Fig.1). The patient underwent endoscopic fenestration of the cyst through a right frontal trepanation via the frontal horn of the right lateral ventricle. The lateral ventricles were chronically enlarged and the cyst insinuated itself between the thin columns of the fornix. The walls were distended, showing the accumulation of CSF inside. Seeing as the patient already had a VPS, we placed a catheter of approximately 15 cm in length, communicating the interior of the cyst with the ventricular cavity (Fig.2). A few days after the surgery, the

patient showed significant neurological improvement, returning to active interaction with her family members, with a decrease in drooling and choking, and no difficulty swallowing solid and liquid foods.

#### Case 2

3-year-old boy with drowsiness and irritability for about a week. Upon neurological examination, the child showed a head circumference of 53 cm (macrocrania), was sleepy and unresponsive to stimuli, and had no focal neurological deficits. Fundoscopy revealed papilledema. Cranial CT-scan showed a hypertensive hydrocephalus, caused by a voluminous cyst of the quadrigeminal plate, which obliterated the aqueduct. The initial treatment was performing a neuroendoscopy for cyst fenestration and third ventriculocisternostomy, through a right frontal trepanation orifice. After fenestration and decompression of the cyst, it was possible to identify the mammillary bodies and the floor of the third ventricle, which was also fenestrated. About a week after the surgery, the patient returned presenting symptoms of irritability and drowsiness, accompanied by vomiting. A cranial CT-scan was performed, which showed a reduction in the dimensions of the cyst and ventricles. However, due to the persistence of signs and symptoms of intracranial hypertension, we placed a neuroendoscopically guided cysto-ventricular peritoneal shunt to ensure that the proximal catheter maintained communication between the cyst and the ventricular cavity (Fig.3). The proximal catheter was multiperforated, so that the cystic and ventricular cavities were simultaneously derived.

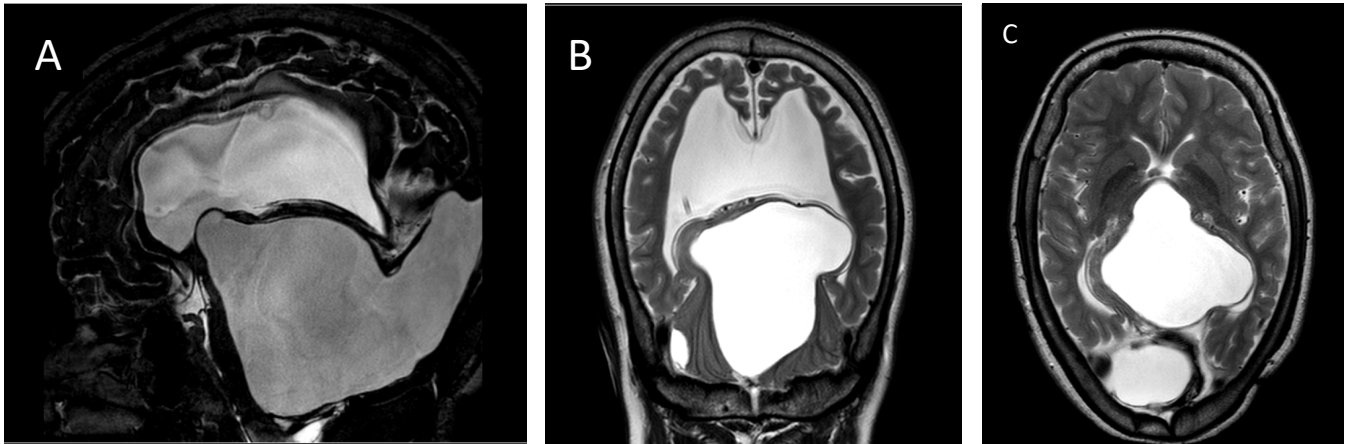
#### Case 3

A 3-year-old male patient complained of severe headache. Upon physical examination, we arrived at the diagnosis of macrocrania, in addition to evident bone bulging in the occipital region. The MRI showed a large cystic lesion located above the cerebellum, with compression of the cerebellum and determining occipital bone remodeling (Fig.4). We performed neuroendoscopic fenestration of the cyst and established its communication with the cisterna magna. Surgical access was made through a suboccipital, paramedian trepanation orifice, with the patient in the prone position (Fig. 5). The patient progressed well, with improvement of the headache and head circumference control.

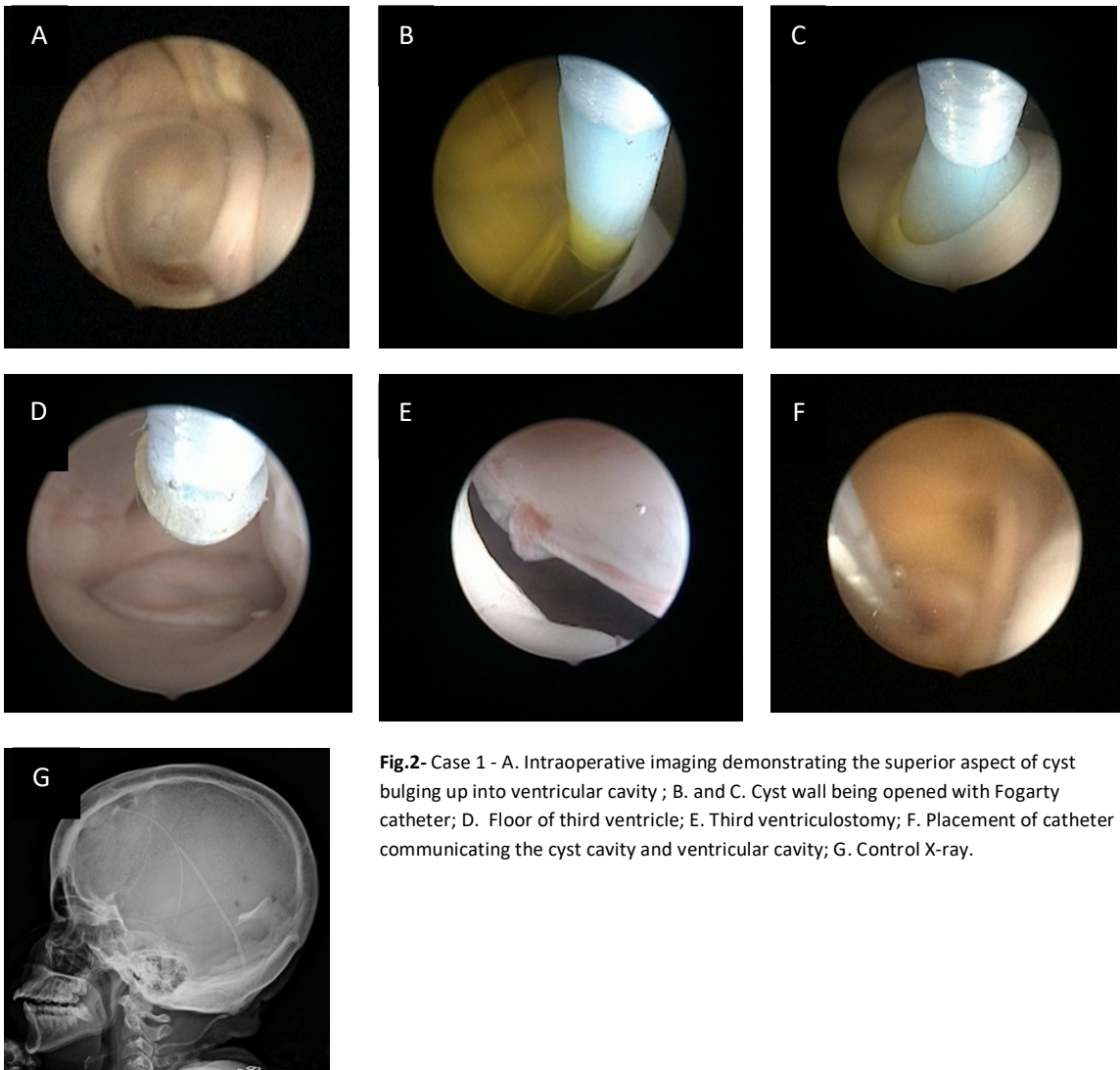
#### Case 4

A 17-year-old female adolescent complaining of vertigo, sudden episodes of malaise and headache for about three months. Physical examination showed no neurological deficit. The MRI showed the presence of a large cyst occupying the left cerebellopontine angle, with compression of the left cerebellar hemisphere, shifting it and displacing

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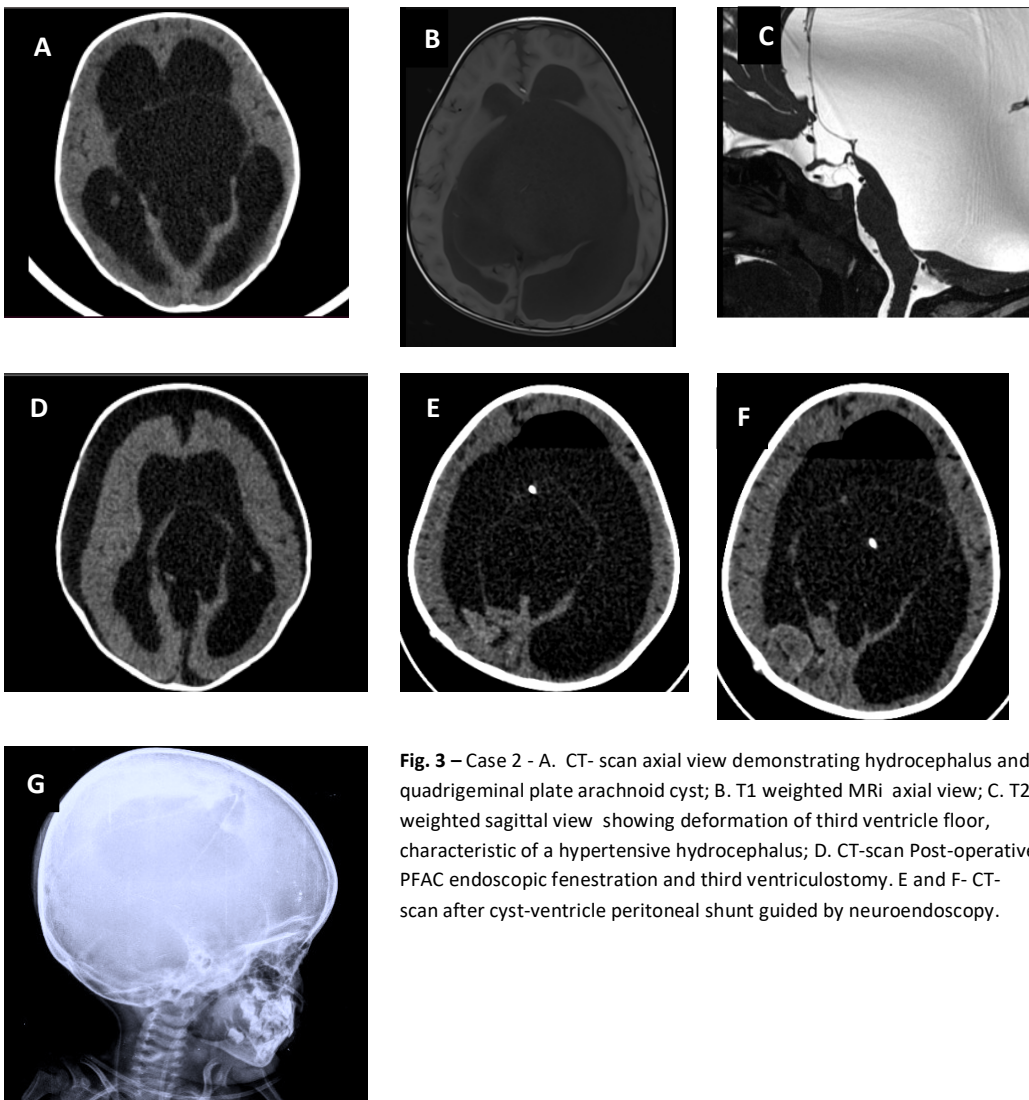
**Fig 1-** Case 1 – T2-weighted MRI showing a huge quadrigeminal plate arachnoid cyst with compression of brain stem and obliteration of fourth ventricle. A. Sagittal view; B. Coronal view; C. Axial view.



**Fig.2-** Case 1 - A. Intraoperative imaging demonstrating the superior aspect of cyst bulging up into ventricular cavity; B. and C. Cyst wall being opened with Fogarty catheter; D. Floor of third ventricle; E. Third ventriculostomy; F. Placement of catheter communicating the cyst cavity and ventricular cavity; G. Control X-ray.



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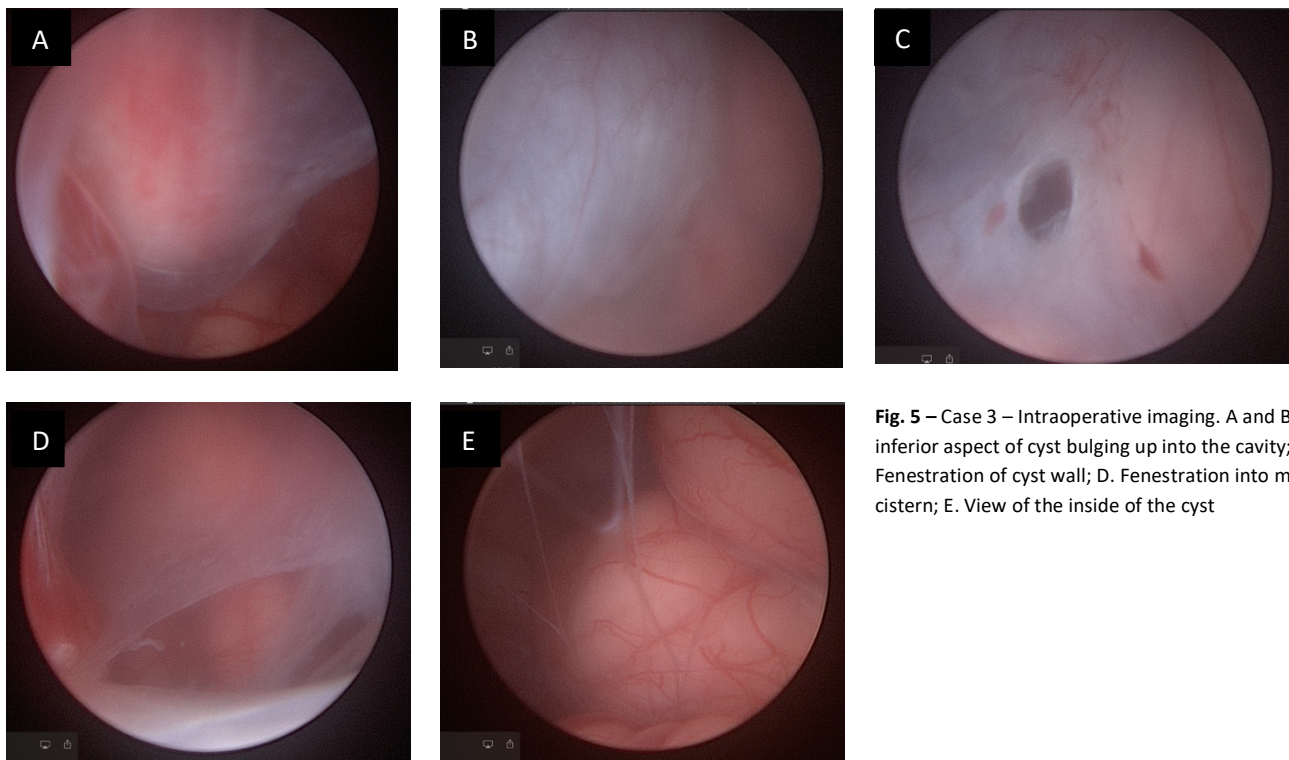
**Fig. 3** – Case 2 - A. CT- scan axial view demonstrating hydrocephalus and quadrigeminal plate arachnoid cyst; B. T1 weighted MRI axial view; C. T2 weighted sagittal view showing deformation of third ventricle floor, characteristic of a hypertensive hydrocephalus; D. CT-scan Post-operative PFAC endoscopic fenestration and third ventriculostomy. E and F- CT-scan after cyst-ventricle peritoneal shunt guided by neuroendoscopy.



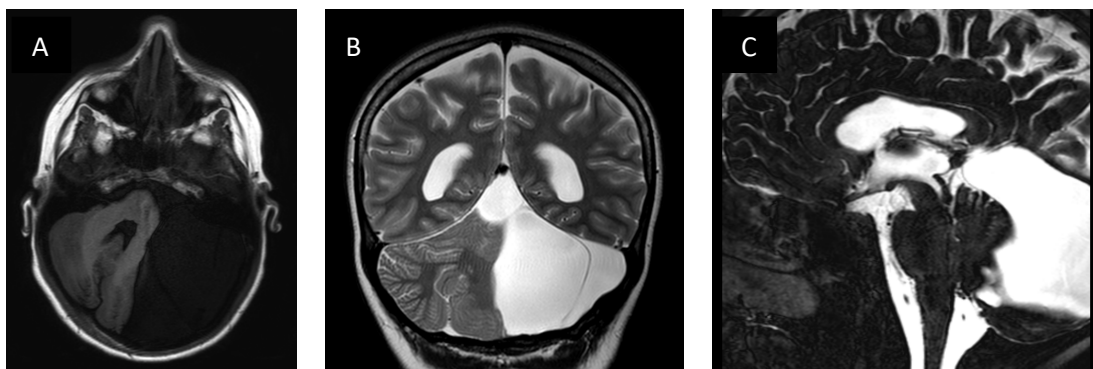
**Fig. 4** – Case 3 - MRI T2-weighted demonstrating a supracerebellar arachnoid cyst. A. Axial view; B. Sagittal view; C. Coronal view



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**Fig. 5** – Case 3 – Intraoperative imaging. A and B. inferior aspect of cyst bulging up into the cavity; C. Fenestration of cyst wall; D. Fenestration into magna cistern; E. View of the inside of the cyst



**Fig. 6** – Case 4 – Cerebellopontine angle arachnoid cyst A. MRI T1-weighted axial view. B. and C. MRI T2-weighted coronal and sagittal view respectively.

the fourth ventricle (Fig.6). We performed a neuroendoscopic treatment for cyst fenestration. The patient was placed in lateral decubitus position and access was performed through a suboccipital paramedian trepanation hole on the left. In the postoperative period, the patient sustained complete resolution of symptoms.

### RESULTS

The age of these patients was concentrated in two extremes: two patients were 3 years old, one was 15 and

another was 17 years. This small sample was equally distributed between the sexes.

Although the patients sought the emergency department, none of them presented acute intracranial hypertension. In adolescents, symptoms of compression of the posterior fossa structures, such as swallowing deficit, ataxia, malaise and vertigo, were predominant. In the 3 year-old patients, predominant symptoms were headache, hypoactivity and somnolence. However, both showed increased head circumference, which can be interpreted as a slowly evolving picture of intracranial hypertension.

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Adolescent patients did not develop acute hydrocephalus. One of them had been shunted at six months of age, sustaining ventriculomegaly, and the other had general malaise and vertigo as predominant symptoms, without hydrocephalus.

All patients underwent brain MRI for surgical planning. In cases 1 and 2, a quadrigeminal plate cyst was diagnosed; in case 3, a supracerebellar cyst and, in case 4, a cerebellopontine angle cyst.

In Cases 1 and 2, which showed respectively ventriculomegaly and hydrocephalus, cyst fenestration was performed through a frontal trepanation orifice. In cases 3 and 4, with no hydrocephalus, neuroendoscopic access was performed through a trepanation hole in the posterior fossa, fenestrating the cyst directly.

Cases 1, 3 and 4 progressed well, with almost immediate improvement of symptoms. However, case 2, which presented a giant quadrigeminal plate cyst, with obstruction of the third ventricle and aqueduct, progressed with problems of CSF circulation, requiring a VPS for the treatment of hydrocephalus, then communicating. The placement of the proximal catheter was performed with neuroendoscopic view, so that the cystic cavity and the ventricular cavity remain in communication, avoiding an eventual closure of the cyst wall fenestration. This catheter was multi-perforated.

In case 1, since the hydrocephalus had already been treated by placing a VPS, we decided to place a multi-fenestrated stent through the cyst fenestration orifice, keeping the communication between the cystic cavity and the ventricular cavity patent.

None of the patients showed signs of fistula in the postoperative period.

### DISCUSSION

Arachnoid cysts account for 1,3 - 2,6% of space occupying intracranial lesions in the pediatric population[3,4,5,6]. The middle cranial fossa is the most frequent location, with about 47% of cases[5,6]. In the posterior fossa, we find 10%[6], 22-27%[3,4], and up to 38% of pediatric series[11]. The posterior fossa is the second most common location: 1/3 of the cysts occupy the cerebellopontine angle, 1/3 occupy the midline, and 20% correspond to cysts in the cerebellar hemisphere[3,4]. Even more rare are cysts that extend above the tentorial notch or into the superior cerebellar cistern[3,4]. Still, in the series by Holou et. al, a higher number of midline cysts were found - about 50% of cases in this location[12]. In a 1992 European cooperative study, 285 childhood arachnoid cysts were reviewed : 15,5% occupied

the cerebellomedullary cistern, 2,3% the cerebellopontine angle and 1,3% the clival and interpeduncular area [13].

In the literature, there is a paucity of large series of patients with PFAC. In 2007, Marin-Sanabria et al. described a series of 23 patients treated for 27 years at their institution. 54 patients with posterior fossa arachnoid cysts were pooled in a 1992 European cooperative study from 17 institutions for 8 years. Although these cysts are predominantly diagnosed in the pediatric population, there is a lack of studies with long follow-up, or that have followed the evolution of patients diagnosed incidentally. For that reason, the development and natural history of PFAC remain unknown[9]. Holou et al. followed 309 patients with incidentally diagnosed arachnoid cysts and observed that PFAC is negatively associated with the onset of symptoms and with surgical treatment[12].

In the series of 23 patients by Manrin-Sanabria et al, there was a predominance of females[9], while most studies show that arachnoid cysts are considerably more frequent in the male population[3,4,5,6,14]. In the study by Holou et al. no predilection for location or laterality was found between the sexes[12]. In our small series, there was equal distribution between the sexes.

PFAC can cause symptoms at any age, but children are most affected in the first decade of life[3,4,5]. In the series by Galassi et al, published in 1985, among the 10 patients diagnosed with PFAC, 4 of them were children aged 17 and 18 months, 2 and 7 years, and boys were more affected than girls. In this group, the predominant symptom was macrocrania and delayed neuropsychomotor development. An increase in head circumference was observed from birth to the first months of life[5].

In the mixed series of patients with arachnoid cysts, 60 to 90% of the patients are in the pediatric group and most of them had symptoms before six months of age[12,14]. In the 1986 series of 16 pediatric patients by Hirsch et al, 5 of them had PFAC. The predominant symptoms were intracranial hypertension (3 cases), developmental delay (2 cases), macrocrania (2 cases) and ataxia (1 case)[15].

Most cases of PFAC become symptomatic during childhood, with more than half of cases diagnosed within the first year of life. The average age in pediatric series in the literature is 38.5 months[3,4]. In the European cooperative study, there was a significant peak in the incidence of cases in the first two years of life. However, the average age for onset of symptoms was 6 years old[13].

In the 1981 study by di Roccco et al, 8 cases of PFAC were found, aged between 3 months and 8 years. 6 of these cases were under one year of age, and the other 2 were 16 months and 8 years old. Macrocrania was the sign present in all

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patients, and neuropsychomotor developmental delay was diagnosed in 5 patients aged from 8 months onwards. Hydrocephalus was a finding present in all cases[3].

The vast majority of PFAC cases in the pediatric population present macrocrania and signs and symptoms of increased intracranial pressure sometimes associated with obstructive hydrocephalus[5]. Although hydrocephalus characterizes cysts located in the midline, this complication is also found in patients whose lesions develop over the cerebellar hemispheres[3,4,5]. Later, nystagmus and cerebellar symptoms will be part of the clinical presentation[5,6]. Rarely PFACs determine a localized, unilateral or bilateral bone bulge in the occipital squama, which is more frequently found in patients with Dandy-Walker malformation[3,4,5,6]. A significant portion of affected individuals exhibit varying degrees of neuropsychomotor developmental delay, and epilepsy is relatively rare[1,7,8]. In the study of Arai and Sato, 14 of 18 pediatric patients with PFAC presented mental or psychomotor delay[1].

None of our cases were diagnosed during the antenatal period. In the study by Marin-Sanabria et al, the cysts of only 3 of the 23 patients were recognized during routine gestational ultrasounds[9]. The differential diagnosis includes several cystic malformations of the posterior fossa such as Dandy-Walker malformation, Dandy-Walker variant, Blake's pouch cyst and mega cisterna magna[2,3,4,5,16]. The rarity of antenatal diagnosis supports the hypothesis that CSF collection within the cyst increases during the first years of life[9]. That is, arachnoid cysts are congenital malformations, in which the separation or duplication of the primitive arachnoid membrane during the embryonic period determines the accumulation of CSF in between these membranes, which will continue to occur after birth[[8,9,17].

Thus, the expansion of PFACs early in childhood can exert a mass effect on adjacent structures and alter important functions[7,8]. The neurological symptoms produced by the compression of the cyst in older children are most commonly: headache, ataxia, vertigo, tinnitus and progressive hearing loss[5,17,18,19]. In recent years, learning deficits, behavioral changes and difficulties in social interactions have been described in patients with PFAC[1,7,8]. There are several case reports of patients with PFAC and psychiatric disorders: conversive disorders, affective disorders, obsessive-compulsive disorders, schizophrenia and autistic symptoms[7,8]. The cerebellum has traditionally been related to coordination and balance motor functions. Currently, the consensus has been that the cerebellum plays an important role in the performance of cognitive, emotional and language functions. Rechtmann et al. described PFAC's impact on the functioning of remote

areas of the cerebral cortex of these patients in the eye tracking study. The authors also performed measurements of cerebral blood flow using arterial spin labeling MRI (ASL-MRI) sequence. A significant decrease in cerebral blood flow was observed in the superior temporal regions of patients with PFAC. This study suggests that compression of the cerebellum by PFACs during the early stages of life may have an important functional impact on temporal lobe activity, which would explain the behavioral changes identified as "autistic-like" in these children[8].

The association of PFAC and other congenital abnormalities can occur in about 10 to 20% of cases[3,4].

While the treatment of asymptomatic PFAC patients remains controversial, the need for surgical treatment of symptomatic patients is beyond discussion[9,10]. The goal of the treatment is to eliminate the mass effect of the cyst on the adjacent structures, with the re-expansion of the cerebellum, to reestablish CSF flow and treat hydrocephalus, if present[3,4,9,10]. Surgical techniques for the treatment of PFAC include: craniotomy for fenestration and microsurgical excision of the cyst wall; cyst and hydrocephalus shunting, in association; the combination of microscopic cyst fenestration and placement of a shunt; stereotactic cysto-ventricular shunting; and neuroendoscopy[3,4,9,10,11,15,16,17,18,19,20,21]. The choice of the most appropriate surgical technique depends on the dimensions of the cyst, its location and the presence or absence of hydrocephalus[20,21].

In the 1980s and 1990s, microsurgical fenestration of the lesion and excision of the cyst membrane through a craniotomy was considered the surgical method of choice, especially in cases without hydrocephalus. However, this treatment does not prevent the recurrence of cysts, or the sudden occurrence of hydrocephalus in the postoperative period. This is evidenced by the frequency with which shunts are required in the postoperative period of microsurgeries with sufficient excision of the cystic membrane by craniotomy. Reconstitution of the cyst wall after incomplete excision, or deficiency of CSF circulation through the subarachnoid or ventricular space where the fenestration was performed may be the factors responsible for treatment failure[3,4,5,9,10,11,15,17,20].

In the 2007 study by Marin-Sanabria, among the 23 patients diagnosed with PFAC, 14 were treated with cyst marsupialization, 5 with cyst marsupialization and placement of a cystoperitoneal shunt, and 3 with endoscopic cisternostomy. Cyst marsupialization was effective in 9 of the 14 patients, the other 5 required an additional shunt. Marsupialization of the cyst with placement of a cystoperitoneal shunt was successful in only 1 of the 5 patients; in the other 4, a cysto-peritoneal shunt or a ventriculoperitoneal shunt was required. About 45% of the



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surgical procedures performed in this study corresponded to surgeries for dysfunction or removal of shunts[9].

Placement of a cysto-peritoneal shunt was considered the method of choice for deeply located cysts. Although the cysto-peritoneal shunt is the technique that presents the highest rate of cyst reduction, the use of shunts is accompanied by the highest rates of additional surgeries for dysfunctions, in addition to the shunt-dependence that is established over time[3,4,9,10,11,15,16,17,20,21].

In the last two decades, different neuroendoscopic techniques have been established as the main method of PFAC treatment: endoscopic fenestration through a trepanation orifice, microsurgery guided by neuroendoscopy, or placement of a catheter guided by neuroendoscopy[10,19,20,21,22].

The challenge of the endoscopic treatment is promoting adequate fenestration of the cyst, and its communication with an anatomical corridor of CSF flow. The use of a rigid endoscope may limit its movement and impair the angulation necessary for fenestration of the cyst in the correct location. The suboccipital trepanation orifice should be carefully planned, providing for this angulation[10,20]. In addition, it is important to consider that the dura mater of the posterior fossa in infants and children may show persistence of the occipital sinus and its numerous anatomical variations[23]. The dimensions of the trepanation orifice might prove inadequate to control bleeding of these venous vestiges of the dura mater, so much so that some groups suggest prior angiographic study[23].

Finding the most appropriate site for fenestration of large posterior fossa cysts can be difficult, since the subarachnoid space is compressed and obliterated by the cyst. Endoscopic exploration within the cyst can also be difficult for inexperienced hands, and the use of neuronavigation may be helpful[10,16,17,18,19,20,21].

Midline infratentorial cysts can be accessed through a paramedian suboccipital trepanation orifice, so that fenestration provides communication of the cyst with the cisterna magna. Lateral, cerebellar, or cerebellopontine angle cysts can be treated with a retromastoid trepanation orifice, so that the cyst communicates with the pre-pontine cistern. The most favorable site for fenestration of these cysts is the space between the trigeminal nerve and the acoustic-facial complex. Here, it is essential to study the dimensions of the pre-pontine cistern and its relationship with the cyst wall in the axial MRI sequences[10,16,17,18,19,20,21].

Quadrigeminal plate cysts, which extend into the third ventricle, causing inferior displacement of the cerebellum, are usually associated with hydrocephalus and can be

accessed through a frontal trepanation orifice. These cysts can be fenestrated via trans-trigonal (trans-lateral-ventricular), communicating the cyst directly with the cavity of the lateral ventricle; or directly with the third ventricle, when the cyst wall extends to the interventricular foramen, or the foramen of Monro. In these cases, fenestration of the floor of the third ventricle is advised, because even with the decompression of the cyst, the aqueduct may remain obliterated, in addition to these anomalies of the subarachnoid space resulting in CSF flow disorders[10,16,17,18,19,20,21].

Endoscopic access for PFAC fenestration has lower complication rates and lower recurrence rates than microneurosurgery. Endoscopic communication of the cystic cavity with the subarachnoid or ventricular space has been shown to be effective in numerous case reports of PFAC, avoiding complications from prolonged use of shunts. The use of shunts should be reserved for complex cases in which the cerebrospinal fluid flow is not restored properly, leading to communicating hydrocephalus[10,16,17,18,19,20,21].

### DISCLOSURES

#### *Ethical approval*

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the local Ethics Committee

#### *Consent to participate*

The patient gave consent to use his information and images for research proposes.

#### *Consent for publication*

The patient gave consent to use his information and images for publication.

#### *Conflict of interest*

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

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