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Treatment and Outcome of Posterior Fossa Congenital Cystic Lesions in Pediatrics

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Abstract:

Posterior fossa congenital cystic lesions in pediatric patients represent a heterogeneous group of anomalies that vary in clinical presentation, natural history, and management strategies. These lesions, including arachnoid cysts, Dandy–Walker malformation, mega cisterna magna, and other cystic entities, may remain asymptomatic or present with hydrocephalus and neurological deficits. The optimal treatment approach remains controversial and depends on multiple factors such as lesion type, presence of hydrocephalus, patient age, and symptom severity. Surgical options include cerebrospinal fluid diversion, endoscopic or open cyst fenestration, and lesion resection, while conservative management is considered in selected asymptomatic cases. Understanding the indications, outcomes, and complications of different treatment modalities is essential for improving patient prognosis.

Keywords: Posterior fossa; congenital cystic lesions; pediatric neurosurgery; arachnoid cyst; hydrocephalus; endoscopic fenestration; ventriculoperitoneal shunt.

Introduction:

The posterior fossa is the second most common location after the middle fossa. Posterior fossa arachnoid cysts (PFACs) can be diagnosed incidentally in utero, or postnatally, either incidentally or due to symptoms. Progressing PFACs or hydrocephalus usually lead to neurological symptoms and are therefore often treated surgically. Surgical options include CSF diversion procedures (e.g., ventriculoperitoneal shunt [VPS] or cystoperitoneal shunt [CPS] placement), endoscopic cyst fenestration, combined endoscopic fenestrations and shunts, or craniotomy with cyst marsupialization or fenestration. For PFACs in children, especially in infants, indications for surgical treatment and the ideal surgical technique remain undetermined. The risk factors for cyst growth leading to surgery are also still unknown. Very few studies have analyzed surgical treatment of PFACs in children and/or adults **(Beresford et al., 2020).**

For PFACs, the surgical options include open or endoscopic cyst fenestration/marsupialization into the CFS space, shunting (VPS or CPS), Endoscopic Third Ventriculostomy (ETV), or a combination of these. However, surgery is not always indicated, since these cysts can remain asymptomatic despite radiological cyst progression or even regress spontaneously. Whether to place a shunt in or fenestrate Arachnoid Cysts (ACs) in general has been an ongoing debate over the last few decades. Modern neurosurgery has introduced endoscopic (AC) treatment to the mix, adding a respected technique which must be considered, making the debate that much more complex. Should we fenestrate openly, fenestrate endoscopically, place a shunt, perform an (ETV), leave an Ommaya reservoir behind as a stent or as a fallback in case of failure **(Liby et al., 2018).**

Based on results, the surgical technique selected typically depends on the cyst type, taking into consideration the interface with the third ventricle and the presence of hydrocephalus, and the surgeon's personal preference based on positive and negative experiences **(Soleman et al., 2021).**

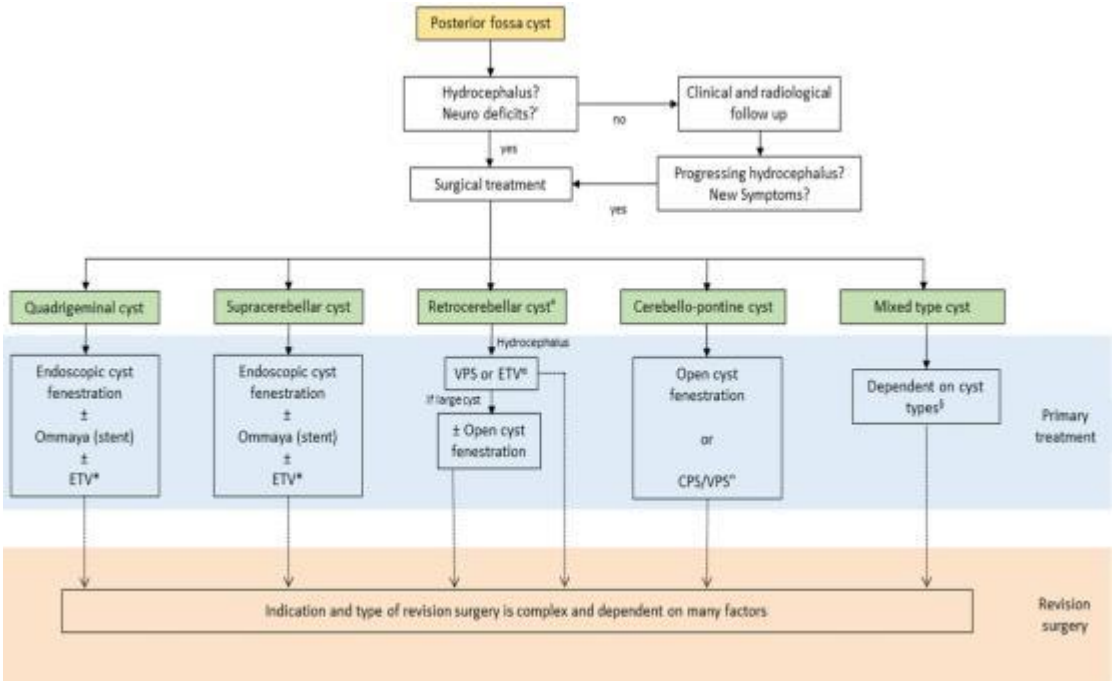

Figure (1): Suggested flow chart for the treatment of PFACs in infants. 'For most cases, hydrocephalus is the actual indication for surgery (and, thus, the focus of this algorithm). Only rarely do neurological deficits in the absence of hydrocephalus lead to surgery. The surgical approach for non–hydrocephalus-related neurological issues will differ, and usually includes an open fenestration or rarely a CPS. *Additional ETV is indicated if hydrocephalus is apparent. Generally, for RetroCerebellar (RC) cysts if hydrocephalus is apparent the hydrocephalus should be treated first. A combined approach (CSF diversion and open cyst fenestration) should be considered in cases where the child presents with neurological symptoms attributed to the cyst accompanied by hydrocephalus. Soleman et al 2021 suggested , most patients were treated with a VPS and showed good outcome with low revision rates. "Shunting is dependent on the extent of the cyst, extent of hydrocephalus (if apparent), and surrounding anatomy in relation to the cranial nerves. §In their series, mixed cysts were either (RC) and SupraCerebellar cyst (SC), or (RC) and Quadrigeminal cyst (QR); usually the (SC) or (QR) cysts were primarily treated (endoscopic fenestration with or without ETV) since they cause hydrocephalus, while the RC cysts could be observed. If, however, the RC cyst was symptomatic or grew in size and no hydrocephalus was apparent, it was treated primarily **(Soleman et al., 2021).**

Treating PFACs endoscopically can be as challenging as treating them through open surgery (posterior fossa craniotomy). The surgeon should master the technical nuances of each method, as well as their potential complications. For large PFACs addressed endoscopically via the supratentorial ventricular system, navigation is advisable for better trajectory planning, but bleeding control, creating a sufficient fenestration, or accessing the posterior fossa through a frontal burr hole can be very challenging as well. Posterior fossa craniotomy in an infant carries the burden of a lengthy operation in the prone position. In addition, excessive blood loss from the much-vascularized dura mater of the infant may be very difficult to control, posing a great challenge not only for the surgeon but for the anesthesiologist as well **(Schulz et al., 2021).**

CSF issues after craniotomy in an infant (presenting with hydrocephalus to begin with) can also be demanding. For these reasons, some authors advocate the use of VPSs or CPSs as effective methods for treating hydrocephalus as well as ACs. However, shunts are known to have a rather high rate of complications and revisions, especially if implanted at an early age. Shunt insertion was associated with significantly higher surgical morbidity rates. On the other hand, endoscopic fenestrations at a young age (typically < 6 months) seem to carry a high risk of closure (**Rei et al., 2017).**

The natural history, surgical outcome, and surgical morbidity associated with PFACs seem different from those of supratentorial, intraventricular, or suprasellar cysts. It also seems that PFACs in different locations behave differently and might respond differently to the various treatment options. Apart from the anatomical location, other factors, such as

age at the time of surgery and the type of surgical procedure, might also play a role. Data seem to indicate that SC cysts showed progression in size more often than other cyst types, especially RC cysts **(Cuny et al., 2017).**

Data also seem to indicate that most of the treated cysts requiring revision surgery were of the RC type. This might be due to the fact that RC cysts were often treated through open craniotomy and cyst marsupialization, which, at times, can be challenging within the posterior fossa and is associated with more morbidity. In addition, many patients with RC cysts presented with hydrocephalus, which was not addressed when preforming craniotomy and cyst fenestration. In many cases, this led to additional surgery addressing the progressing hydrocephalus (**Ali et al., 2015).**

The fact that some PFACs remain stable over time should encourage neurosurgeons to choose to monitor infants presenting with PFACs that are asymptomatic and do not lead to hydrocephalus, since the natural history may be benign, even if the cyst shows progression at some point. In addition, when counseling parents who are carrying a fetus with a PFAC, the surgical options presented in this discussion, which offer a high probability of low permanent morbidity, no mortality, and good clinical outcome, must be taken into consideration **(Pesaresi et al., 2024).**

Still, how and whether PFACs can and should be discriminated from Mega Cisterna Magna (MCMs), Dandy Walker Syndrome (DWS), and Black Pouch Cyst (BPCs), as well as what are the clear radiological characteristics defining MCMs, DWS, and BPCs, still remain a matter of debate. For this study, the differentiation between PFACs and other entities (e.g., MCM, DWS, and BPS) was based on the definitions of AC, MCM, DWS, and BPS described by Wüest et al. A clear definition of the different types of PFACs, based on radiological images, does not exist. Vaquero et al. divided the PFACs into SC (including cysts of the quadrigeminal region), RC, laterocerebellar (the same as CPA cysts), clival, and mixed types. Many of the cysts are very large, crossing anatomical boundaries and making it difficult to distinguish between the different types (**Roth et al., 2019).**

The treatment of PFACs in infants varies, depending on the course and behavior of the cyst, primarily based on the cyst location and anatomical nuances. QR and SC cysts can often be treated exclusively by neuroendoscopic techniques, while RC and CPA cysts are often treated by open surgery or shunt insertion. SC cysts often show progression in size over time, while RC cysts usually cause hydrocephalus leading to surgery. PFACs of infancy often require surgical treatment before the age of 6 months, leading (in the majority of patients) to good clinical and radiological outcomes. Additional surgery is required in up to 30% of patients, with a mean of 2.4 surgeries per patient; younger age and nonprogressive cysts might be risk factors for requiring revision surgery. The fact that various surgical options exist for effective treatment of PFACs in infants, leading to low permanent morbidity and mortality rates, should guide prenatal counseling of parents **(Wüest et al., 2017).**

The treatment of congenital cystic lesions in the posterior fossa of pediatrics is influenced by factors such as the presence of hydrocephalus, lesion type, patient age, and symptom severity. A multidisciplinary approach involving neurosurgeons, neurologists, and radiologists is essential for optimal outcomes **(Samkari et al., 2021).**

1. Treatment Based on Key Factors:

A. Presence or Absence of Hydrocephalus:

With Hydrocephalus:

- **Ventriculoperitoneal (VP) Shunt or Endoscopic Third Ventriculostomy (ETV):** These are used when hydrocephalus is either symptomatic or worsening **(Tamburrini et al., 2020).**
- **Complications:** Shunt infections, shunt malfunction, overdrainage leading to slit ventricle syndrome or subdural hematoma, and potential failure of ETV **(Sufianov et al., 2019; Tamburrini et al., 2020).**
- **Cyst Fenestration:** In cases where the cyst obstructs cerebrospinal fluid (CSF) flow, endoscopic or surgical fenestration may be necessary **(Brockmeyer, 2015).**
- **Complications:** CSF leakage, infection, incomplete fenestration resulting in cyst recurrence, and damage to surrounding structures **(Sufianov et al., 2019).**

- **Lesion Resection:** Surgical removal may be required if the lesion itself is contributing to hydrocephalus **(Samkari et al., 2021).**
- **Complications:** Brainstem injury, neurological deficits, infection, and bleeding **(Sufianov et al., 2019).**

Without Hydrocephalus:

- **Observation:** If the lesion is small, asymptomatic, and does not cause significant mass effect **(Sufianov et al., 2019).**
- **Complications:** Minimal risk; however, there may be a chance of lesion progression requiring future surgical intervention **(Tamburrini et al., 2020).**
- **Surgical Excision:** Recommended if neurological deficits or symptoms from mass effect are present **(Dincer et al., 2017).**
- **Complications:** Neurological deficits, infection, bleeding, and risk of damage to adjacent brain structures **(Dincer et al., 2017).**

B. Type of Lesion:

1. Arachnoid Cysts:

- **Conservative Management:** In asymptomatic cases, periodic imaging is used to monitor the cyst **(Tamburrini et al., 2020).**
- **Complications:** None, as this involves observation and imaging **(Tamburrini et al., 2020).**
- **Surgical Options:**
- **Endoscopic Fenestration:** Preferred for accessible cysts **(Brockmeyer, 2015).**
- **Complications:** Infection, cyst recurrence, and injury to surrounding structures **(Brockmeyer, 2015).**
- **Cystoperitoneal Shunt:** If CSF accumulation continues **(Tamburrini et al., 2020).**
- **Complications:** Shunt infection, malfunction, peritoneal complications, and overdrainage **(Tamburrini et al., 2020).**

2. Dandy-Walker Malformation:

- **VP Shunting:** If hydrocephalus is present **(Sufianov et al., 2019).**
- **Complications:** Infection, shunt failure, overdrainage, and cerebral hemorrhage **(Sufianov et al., 2019).**
- **Posterior Fossa Decompression:** In severe cases with significant compression of the brainstem **(Samkari et al., 2021).**
- **Complications:** Cerebellar injury, bleeding, infection, and possible worsening of neurological deficits **(Sufianov et al., 2019).**

3. Mega Cisterna Magna:

- **Observation:** Typically benign and requires no intervention unless symptoms develop **(Tamburrini et al., 2020).**
- **Complications:** Minimal risk; however, there is a potential for symptomatic progression requiring intervention **(Tamburrini et al., 2020).**

4. Epidermoid or Dermoid Cysts:

- **Surgical Resection:** Recommended to prevent rupture or secondary infection **(Dincer et al., 2017).**
- **Complications:** Infection, bleeding, CSF leakage, and damage to nearby cranial nerves **(Dincer et al., 2017).**
- **Complete Excision:** To minimize recurrence **(Tamburrini et al., 2020).**

- **Complications:** Recurrence of the cyst if incomplete excision occurs, injury to surrounding brain structures **(Tamburrini et al., 2020).**

5. Encephalocele:

- **Surgical Repair:** The primary approach is surgical closure to avoid CSF leakage and infection **(Tamburrini et al., 2020).**
- **Complications:** Infection, wound dehiscence, CSF leakage, and potential neurological deficits **(Tamburrini et al., 2020).**
- **Hydrocephalus Management (VP Shunting):** If hydrocephalus is present **(Sufianov et al., 2019).**
- **Complications:** Shunt infections, malfunction, and overdrainage **(Sufianov et al., 2019).**
- **Multistage Surgery:** In large encephaloceles, multiple surgeries may be needed to preserve neurological function **(Tamburrini et al., 2020).**
- **Complications:** Infection, neurological deficits, and prolonged recovery times **(Tamburrini et al., 2020).**

C. Age of the Patient:

Neonates & Infants

- Minimally invasive procedures or shunting are preferred to reduce surgical risks **(Sufianov et al., 2019).**
- Complications: Infection, shunt failure, and developmental delay due to prolonged ICU stays or surgical risks **(Sufianov et al., 2019).**

Older Children:

- More aggressive surgical options may be considered for symptomatic lesions **(Samkari et al., 2021).**
- Complications: Higher risk of complications such as infection, bleeding, and neurological deficits due to more complex procedures **(Samkari et al., 2021).**

D. Severity of Symptoms:

Mild/Asymptomatic Cases

- **Observation** with periodic MRI scans **(Tamburrini et al., 2020).**
- **Complications:** Minimal risk **(Tamburrini et al., 2020).**

Moderate Symptoms

- **Surgical decompression** or cyst fenestration **(Brockmeyer, 2015).**
- **Complications:** Infection, recurrence of cyst, and damage to brain tissue **(Brockmeyer, 2015).**
- **Severe Cases** (e.g., Brainstem Compression, Progressive Symptoms)

Urgent Surgical Decompression:

- **Complications:** Cerebellar injury, brainstem damage, infection, bleeding, and prolonged recovery **(Tamburrini et al., 2020).**

CSF Diversion if Hydrocephalus is Present:

- **Complications:** Shunt infections, overdrainage, and ventricular collapse **(Tamburrini et al., 2020).**

2. Postoperative Care and Follow-up:

- **Neurological Monitoring:** For signs of raised intracranial pressure, infections, or recurrence **(Samkari et al., 2021).**

- **Imaging Surveillance:** Regular MRI scans to monitor for complications or recurrence **(Sufianov et al., 2019).**
- **Physical Therapy:** If motor deficits are present **(Dincer et al., 2017)**.

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