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Review of surgical approaches for posterior fossa tumors in pediatric population and illustrative cases

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Introduction/Background: Posterior fossa tumors are common in the pediatric population, and various surgical approaches have been described to reach them based on their specific location. This paper aims to review the most common posterior fossa approaches in pediatric patients and present illustrative cases to provide insights into their successful application in specific clinical scenarios.
Case report: Three cases are presented, each demonstrating a different approach and surgical outcome. Case 1 involved a telovelar approach for the resection of a medulloblastoma, resulting in complete removal without neurological deficits. Case 2 required a combination of telovelar and supracerebellar infratentorial approaches to achieve gross total resection of a pilocytic astrocytoma. In Case 3, an occipital transtentorial approach was utilized to access and completely resect an atypical teratoid rhabdoid tumor.
Connclusion: The paper discusses the telovelar and transvermian approaches, the supracerebellar infratentorial approach, the transtentorial suboccipital approach, and intraventricular endoscopy as individual or combined approaches for posterior fossa tumors. The presented cases highlight the importance of tailoring the surgical approach to each patient and provide valuable insights for neurosurgeons managing similar clinical scenarios.

Keywords: telovelar, ventricular endoscopy, supracerebellar infratentorial, occiptal transtentorial, combined approches

INTRODUCTION

Posterior fossa tumors are the most common in the pediatric population. They represent 45%–60% of pediatric brain tumors(1–3). These group of tumors englobes a great number of different pathologies, with different pathophysiological characteristics, from WHO grade 1, such as pilocytic astrocytoma, to malignant and infiltrating, like medulloblastoma (WHO grade 4)(3,4).

Given that most of them have their origins in the fourth ventricle and adjacencies, including surrounding cerebrovascular structures, they pose a significant challenge for neurosurgeons. Numerous approaches have been described to reach them according to their specific location. However, there are some patients in which these approaches alone are not enough to reach the goals of surgical management, like maximal safe resection(5–9), especially in young children, that may require surgery in multiples steps.

The aim of this article is to review the most common posterior fossa approaches in the pediatric population, their combination and display illustrative cases.

Background

The transvermian approach, which involves splitting the vermis, is the most traditional surgical approach to access the fourth ventricle. Dandy described the transvermian approach sparing the dentate nucleus to be free of new neurological deficits. However, subsequent papers didn't corroborate the same results(6,10). In 1992, Matdhshima et al., proposed an alternative approach through the cerebellomedullary fissure, commonly known nowadays as telovelar approach, to reduce the risk of complications like cerebellar mutism. Over the past three decades, this has been refined and used successfully.(11,12)

The pineal region is another critical area of the posterior fossa that brings discussion regarding the best approach. The supracerebellar infratentorial approach was first described by Krause in 1911. In 1920, Pendell described the theoretical and practical advantages of this surgical route to the pineal region. And latter, in the microsusrgical era, it was reintroduced by Stein in 1971(13). The transtentorial approach, also known as Poppen's approach, was first described by Heppner in 1959 and popularized by Poppen and Marino in 1968. After that, many authors proposed

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modifications to prevent brain retraction and surgeon discomfort, the most common is to adopt a three-quaters prone position(14).

For over a century, ventricular endoscopy has been used to treat hydrocephalus. In 1973, Fukushima introduced for endoscopic tumor biopsy. But, it was only in 1997, that combined endoscopic biopsy with third ventriculostomy was described in a single step surgical approach(15,16).

CASE REPORTS

Case 1

Six-year-old boy presented with progressive headaches. He was submitted to a Magnetic Ressonance Image (MRI) that exhibit a heterogeneous midline mass occupying the fourth ventricle (Figure 1 A and B). The telovelar approach was used to access the tumor, with gross total resection (Figure 1 C and D). The immunohistochemistry was compatible with Medulloblastoma group 3. He was referred to adjuvant treatment in another unit.

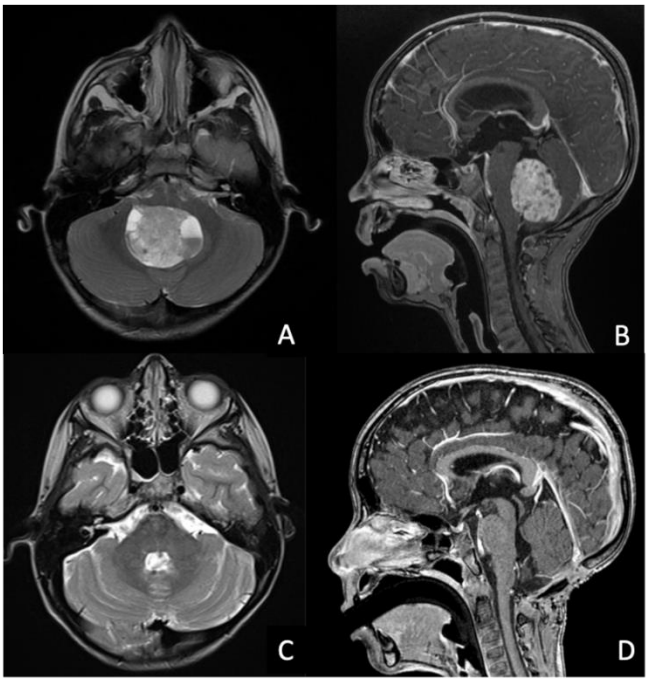

Figure 1- Preoperative MRI showing a heterogenic fourth ventricle mess in Axial T2-weighted (A) and sagittal contrast enhanced images(B), and postoperative exam showing gross total resection of the medulloblastoma Axial T2-weighted (C) and sagittal contrast enhanced images (D).

Case 2

Ten-year-old girl presented with headaches for a year and recent vomits. During investigation, she had an MRI that revealed a lesion that occupied the superior aspect of the fourth ventricle and cerebellomesencefalic fissure (Figure 2A and B). The hormone profile was normal. We initially decided to operate via telovelar approach to perform diagnosis, once the tumor could have a germinative nature. During surgery was noticed that the lesion was very adherent to the parenchyma, IV ventricle floor and a partial resection was possible without the risk of neurological deficits (Figure 2C). The immunohistochemistry was positive for pilocytic astrocytoma. Therefore, the patient was later undertaken to another surgical procedure through supracerebellar infratentorial midline approach in the prone position to complete the removal (Figure 2D). She had no permanent neurological deficits and had no recurrence after 3 years of follow up.

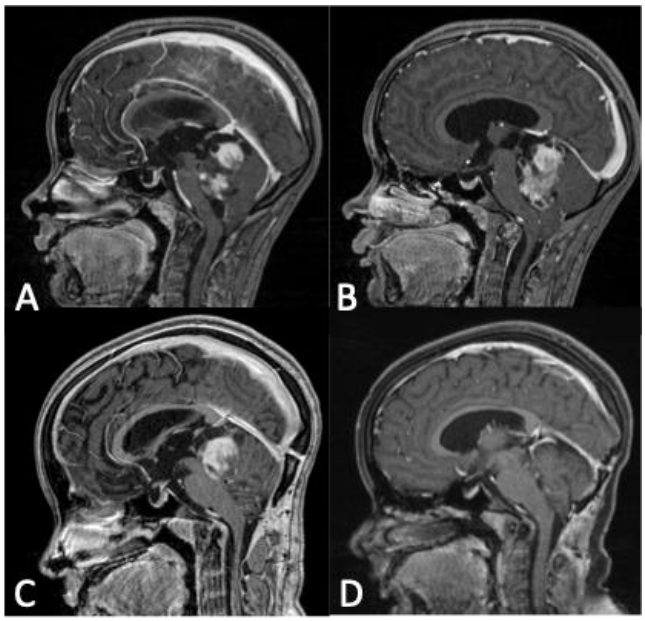

Figure 2- Sagittal contrast-enhanced MRI. A e B) Preoperative. C) Residual tumor after telovelar resection. D) Complete extraction after combination with supracerebellar infratentorial approach.

Case 3

One-year-and-five-months-old girl arrived at the pediatric emergency with a seizure. At neurological exam she presented with Parinaud sign, bulged fontanelle, and developmental delay (motor and speech). On image exams, supratentorial hydrocephalus associated with pineal lesion was noticed as shown in figures 3 and 4. The lesion was iso to hypointense in T1 weighted image and hyperintense in T2, with a dull heterogeneous contrast enhencement. She underwent one-burr-hole endoscopy for a EVT and biopsy in the same procedure (Figure 4). The immunohistochemistry revealed the diagnosis of Atypical Teratoid Rhabdoid Tumors (ATRT). She was later submitted to an occipital transtentorial approach in the ¾ prone position, and gross total resection was reached (Figure 5). A chemotherapy protocol with Temozolamide, Cyclophosphamide, Doxorubicin, Vincristine and Etoposide was performed for one year. Unfortunately, she died 2 years after with pulmonary infection.

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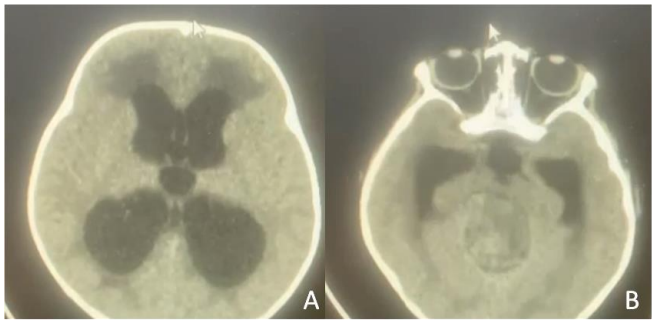

Figure 3- Emergency head CT-scan showing obstructive hydrocephalus (A) by a mass lesion obliterating the aqueduct (B).

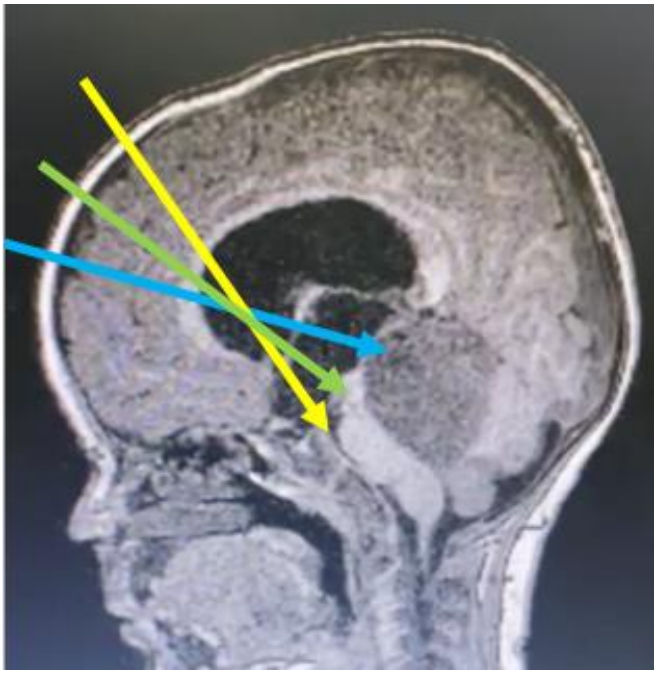

Figure 4- Endoscopic trajectories. Blue: optimal entry-point for biopsy. Yellow: optimal entry-point for ETV. Green: Midway entry-point for endoscopy.

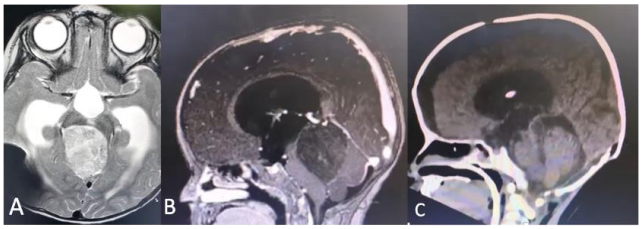

Figure 5- Post-endoscopy MRI. A) Axial T2-wheighted image showing a heterogenic lesion in the topography of quadrigeminal cistern. B) Sagittal T1 contrast-enhenced image showing that the mass dis not enhanced with contrast. C) Post-operative sagittal CT-scan showing total resection after occipital transtentorial approach

DISCUSSION

The telovelar and transvermian approach

The most classical approaches for tumors of the 4th ventricle are the telovelar and the transvermian approach, both traditionally performed on the prone or concorde position(6,10,17,18). The transvermian approach consist in a suboccipital craniotomy follow by “y” shaped durotomy and a midline incision on vermis at the level of the pathology. It provides an extensive cranio-caudal angle. Although Dandy described the transvermian approach sparing the dentate nucleus to be free of new neurological deficits, subsequent papers didn’t corroborate the same results(6,10).

As an alternative, the telovelar route benefits from the opening of the non-eloquent tela choroidea and inferior medullary velum and retraction of cerebellar tonsils to access the fourth ventricle. By removal of C1 posterior arch, one or both tonsils, it can be tailored to have better access of the fourth ventricle extremities and adjacent areas, such as the lateral recess, foramen of Lushkae, and aqueduct(6,7,18,19). Besides the greater working area, the telovelar is indeed less associated with cerebellar mutism than the transvermian approach(6,7,20).

In case 1, the telovelar approach was sufficient to completely remove the medulloblastoma without the precipitation of new neurological deficit, including cerebellar mutism. Figure 1B, demonstrates that the superior aspect of the rhomboid fossa was spared, a fact that facilitates the resection through this approach. On the other hand, the telovelar approach was not enough to perform a gross total resection in case 2. The adhesions hindered the dissection on the superior aspect of the astrocytoma.

The supracerebellar infratentorial approach

The posterior incisural space, with the quadrigeminal cistern, is another region of posterior fossa tumors in children. With some variants, the supracerebellar infratentorial approach can access the posterior incisural space, without the deep venous system in the way, through at least three different routes: the midline, paramedian and extreme lateral. Although it has been well reported to be done in prone position, the most traditional is the semi-sitting because it offers the advantage of the gravity working in the surgeon’s favor, which decreases the need for active cerebellar retraction. The cranial approach is tailored accordingly to the intracranial route and should always include exposing of the transverse sinus.(16,21–23).

For the midline route, a standard midline suboccipital craniotomy is enough to adequate visualization. The surgeon must coagulate the precentral cerebellar vein, the superior vermian vein, and the vermian-tentorium bridging veins to retract the culmen for a better view of the quadrigeminal cistern. However, the access to the inferior colliculus is still

limited(21,24,25). Another limitation of this approach is the dependency of the tentorium angle. When the tentorium angle is high, the access to the quadrigeminal cistern is harder(25).

The paramedian route is accessed via paramedian suboccipital craniotomy. It allows exposure of the inferior part cerebellomesencefalic fissure and ambiant cistern of the ipsilateral side, however the contralateral visualization is restricted(21,24)

Finally, the extreme lateral provides good exposure of the posterolateral aspect of the mesencephalon, including the ipsilateral tectal plate and superior cerebellar peduncle, from a more lateral angle. The recommended cranial access is the retrosigmoid. The more lateral the approach, less bridging veins exists(21,23,24).

In case 2, we used the supracerebellar infratentorial midline approach to complete the removal of the pilocytic astrocytoma. Because of the risks of semi-siting position and the upper level of lesion on the posterior fossa, we didn't predict a need for intense cerebellar retraction and preferred to use the prone position. The gross total removal without new neurological deficits, corroborated our hypothesis.

The transtentorial subocciptal approach

An alternative to the access the posterior incisural space is the suboccipital transtentorial approach, also known as Poppen's approach. An average of positions have been described for this route, the most common are the prone and 3/4 prone positions. Although relatively simpler and anatomically more favorable, the prone position is associated with significant more retraction of the occipital lobe. On the other side, the three quarters prone position with the operated side facing downward, has the advantage of minimal occipital lobe retraction, and higher comfort for surgeon and assistant(16,25,26).

Whatever the position, the approach begins with a parieto-occipital craniotomy beyond the midline with careful exposition of the superior sagittal sinus, transverse sinus, and the superior sagittal sinus-straight sinus corner(16,26).

Through the corridor between the occipital lobe and falx cerebri, the tentorium is divided slightly obliquely to the straight sinus. The medial cut edge of the tentorium is pulled up and retracted to expose the posterior fossa widely. By standing at the parietal side of the patient, the aqueduct can also be visualized besides the deep cerebellomesencefalic fissure. The approach offers the surgeon the opportunity to incise on the superior medullary velum to access the superior aspect of the fourth ventricle(8,16,23,24,26).

The disadvantage of this approach is that the deep venous system may obstruct the access to the tumor and requires retraction of the occipitoparietal lobes, which can cause sensory or visual field deficits. The latter maybe prevented by positioning the patient with the head rotated with the lesion side downward, which diminishes the need for retraction(16,21,23).

In case 3, we decided for this approach because we had a lesion occupying the quadrigeminal cistern pushing the deep veins upward (Figure 3 and 4). The ¾ prone position provided a comfort for the surgeon and enabled full visualization of the tumor.

Intraventricular endoscopy

Whether to biopsy or treat obstructive hydrocephalus with third ventriculoscopy, intraventricular endoscopy is used to manage of many tumors of posterior fossa. (16,25). First described by Ellenbogen and Moores in 1997(27), the combined approach of endoscopic biopsy (EB) with third ventriculostomy (ETV) can be performed in a single step. The advantages include therapeutic and diagnostic functions and reduced number of procedures for the patient. Most authors advocate for performing the biopsy after the ETV because of the possibility of visual obstruction due to bleeding associated with tumor biopsy(15,16,28).

Although it can be done with two burr holes, in our center we introduce the rigid endoscope through one burr hole in the midway between the optimal entry site for the third ventriculoscopy and aqueduct biopsy (Figure 4). Another strategy is the incorporation of neuronavigation to help plan the best trajectory. This approach has the leverage of a single trajectory in the brain parenchyma and one incision behind the hair line. The major anticipated issues with this approach are more pressure to the fornix and on the plexus choroid on the posterior border of the foramen of Monro that may cause bleeding. Moreover, this strategy is limited by the massa intermedia, tumor and ventriculomegaly size. Therefore, it is best employed in patients with small massa intermedia, larger tumors that presents anterior to it and in those with severe ventriculomegaly. It is possibly contraindicated in small tumors located extremely posterior to the third ventricle(15,16,29,30).

In case 3, we demonstrated an indication for this approach. Tumor size, small interthalamic adhesion, and massive hydrocephalus made the simultaneous one-burr-hole endoscopy for third ventriculostomy and aqueduct biopsy possible.

Combined approaches

The combination of two or more approaches to intracranial lesions have been vastly reported in the literature. Although the telovelar approach with C1 laminectomy provides enlargement of superior fourth ventricle working area, the tumors located in this portion

remain difficult to reach without transecting at least a portion of the vermis(5,6,19).

As an alternative, the telovelar and supracerebellar infratentorial approaches combined is a factual option for large midline tumors that occupies the fourth ventricle with extension to the tectal region (as in case 2). It provides a safe way to prevent neurologic deficits associated with transgression of the vermis(5,7). It is important to notice that care must be taken when coagulating bridging veins in both approaches once there is a risk of diminishing collateral drainage. Piatt and Kellogg, correlated this to postoperative edema, venous infarction and hemorrhage(31).

CONCLUSION

Posterior fossa tumors in pediatric population represent a great challenge for the neurosurgeon due to their diverse pathologies and complex anatomical location. Many approaches to this region have been described in the literature. In cases in which single surgical approach can not lead to maximal safe resection by itself, combined approaches offer a valuable strategy for addressing the challenges associated with these complex lesions. By presenting these illustrative cases, we aimed to contribute to the existing literature and provide valuable insights for neurosurgeons facing similar clinical scenarios.

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DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki.The paper is a review with illustrative cases with no identifiable data

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

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

Conflict of interest

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper

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CONTRIBUTIONS

-**Alick Durão Moreira**: Conceptualization, Data curation, Project administration, Writing – original draft
-**Antonio Bellas**: Conceptualization, Data curation, Project administration, Supervision, Validation
-**Marcelo Pousa**: Data curation, Methodology, Validation
-Tatiana Protzenko: Conceptualization, Data curation, Formal Analysis, Project administration, Validation, Writing – review & editing

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