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Surgical approaches in pediatric neuro-oncology

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Abstract
Tumors of the central nervous system comprise nearly a quarter of all childhood cancers and are the most frequent solid tumor in the pediatric population. The most common location is in the posterior fossa, but tumors can occur anywhere intracranially. The spectrum of lesions encountered varies, from being completely benign and requiring surveillance alone to being highly malignant and requiring aggressive treatment in the form of surgery and adjuvant therapy. The extent of resection plays a crucial role in the oncological outcome of many of these tumors. A variety of surgical approaches are available for the spectrum of lesions encountered. This review focuses on summarizing the location, types, and neurosurgical management strategies for pediatric brain intracranial brain tumors. Here, we discuss neurosurgical approaches for a variety of brain tumors and regions, including the management of tumors of the posterior fossa, brainstem, pineal region, intraventricular region, sellar and suprasellar regions, optic pathway and hypothalamus, and supratentorial hemispheres.

Keywords Posterior fossa · Brainstem · Pineal · Intraventricular · Sellar · Suprasellar

1 Introduction

Tumors of the central nervous system (CNS) compromise 20–25% of all childhood cancers and are the most common solid malignancies in the pediatric population [1, 2]. They range from being benign to malignant and can occur in a variety of locations across the infra- and supra-tentorial compartments of the brain. Surgical management may include biopsy for histological diagnosis, cytoreduction/debulking, and complete excision for both local oncological control as well as managing sequelae such as hydrocephalus and insertion of ventricular access devices (VADs) to facilitate intra-thecal/intraventricular adjuvant chemotherapy. Extent of resection (EOR) plays a significant role in the oncologic outcome of many of these tumors. Understanding the principles and concepts that underpin neurosurgical strategy and approaches in the management of these tumors is crucial for all medical professionals involved in caring for these patients. This review will utilize case vignettes to summarize the locations types, and surgical strategies used in pediatric intracranial malignancies.

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2 Posterior fossa tumors

The majority of pediatric CNS tumors (60–70%) arise in the posterior fossa [3], and despite significant advances in diagnosis, molecular profiling, and surgical and adjuvant therapies, they still cause the most cancer-related deaths in children [4, 5]. The three most common tumors are medulloblastoma, ependymoma, and pilocytic astrocytoma. Tumors that are less common include atypical teratoid/rhabdoid tumor (AT/RT), hemangioblastoma, and ganglioglioma. EOR is arguably the most important predictor of oncologic outcome in most of these tumors (Table 1) [11].

2.1 Management strategy

Anatomical proximity to vital structures within the small volume compartment of the posterior fossa means that deterioration can rapidly occur, even with small tumor sizes. Surgical strategy also needs to account for the presence or absence of acute hydrocephalus due to blockage of 4th ventricular outflow—the presence of which is reported in 70–80% cases of posterior fossa tumors—and is frequently the cause of clinical deterioration [12]. The surgical approach is dependent on whether the tumor lies in the midline, laterally within the cerebellar hemisphere or laterally with extension to or arising within the CPA.

Table 1 Surgical principles and the impact of the extent of resection (EOR) for the most common posterior fossa pediatric tumors

Posterior fossa tumors and the extent of surgical resection	
Pilocytic astrocytoma (PA)	• Complete resection is considered curative [6] • Resection of the mural nodule is crucial; resection of the non-enhancing cyst wall is not necessary • STR increases local recurrence rates, but does not affect survival • Risk of neurological deficits with STR should be considered when deciding between STR and surveillance imaging
Ependymoma (EP)	• Complete resection is the surgical aim and the primary predictor of oncological outcome [7, 8] • Re-operation to resect residual tumor seen on post-operative imaging is strongly advocated if safely possible • Surgical resection is also advocated for recurrences; These often occur at the primary tumor site • Encasement of neurovascular structures within tumors of the cerebello-pontine angle (CPA) or ventral brainstem are associated with significant morbidity • Benefits: GTR is highly beneficial as these tumor have higher recurrence/metastasis rates [9] • Limitations: neurovascular injury
Medulloblastoma (MB)	• Surgical aim is GTR without inducing significant neurological morbidity • GTR versus NTR: recent evidence demonstrates no difference in overall survival (OS) [10] • GTR/NTR versus STR: Progression free survival (PFS) is decreased with STR [10]
Atypical teratoid /rhabdoid tumor (AT/RT)	• GTR is feasible in ~ 30% of patients [4] • The surgical plane between normal cerebellum and AT/RTs can be difficult to delineate • Encasement of neurovascular structures in tumors with CPA extension increases surgical difficulty and morbidity

STR, subtotal resection (> 1.5 cm^2 residual tumor); *GTR*, gross total resection; *NTR*, near total resection (< 1.5 cm^2 residual tumor)

2.2 Preoperative hydrocephalus treatment

Acute hydrocephalus in the presence of acutely deteriorating consciousness requires prompt stabilization with an immediate cerebrospinal fluid (CSF) diversion procedure. This is often by way of external ventricular drain (EVD) placement in crisis, which can be rapidly performed and allows predictable CSF drainage and clinical assessment, although CSF drainage should be carefully controlled to avoid complications related to rapid changes in intracranial pressure (ICP) from over-drainage that can precipitate upward cerebellar herniation and subdural hematoma formation. Given that only 15–30% of patients ultimately require permanent CSF diversion following tumor resection [13, 14] and the potential morbidity from complications associated with permanent ventriculoperitoneal shunt insertion (VPS), most neurosurgeons avoid routine preoperative shunt placement. Most patients can be temporized with a course of corticosteroids while awaiting surgical debulking/resection on a semi-urgent basis. The advancement of endoscopic optics and visualization has allowed the implementation of endoscopic 3rd ventriculostomy (ETV) in managing pre- and postoperative hydrocephalus. ETV results in the formation of a stoma in the floor of the 3rd ventricle bypassing any obstruction to CSF outflow in the aqueduct and 4th ventricle, allowing direct drainage of CSF into the basal cisterns and subarachnoid CSF spaces. When performed prior to posterior fossa tumor surgery, it may reduce incidence of hydrocephalus due to normalization of CSF hydrodynamics and decreasing the risk of permanent postoperative CSF dysfunction [15]. However, given the small number of patients needing definitive treatment for hydrocephalus after tumor resection, preoperative ETV is also not routinely performed, unless the patient presents with hydrocephalus associated with vision-threatening papilledema and there exists a delay while awaiting definitive surgery. In these cases, ETV is a useful option to temporarily treat the raised ICP associated with hydrocephalus while avoiding a temporary EVD or permanent VPS.

2.3 Microsurgical approaches

The midline or paramedian suboccipital craniotomy is most commonly performed in the prone position. Care is taken to ensure pressure areas are padded and protected and the head is flexed forward with two fingerbreadths space between the chin and chest to avoid raised venous pressure secondary to obstruction of jugular veins which can precipitate intraoperative posterior fossa swelling and/or hemorrhage. The lateral suboccipital craniotomy/retromastoid craniotomy is usually conducted in the lateral decubitus position, but the supine position with significant head rotation can also be used.

2.4 Suboccipital craniotomy

The suboccipital craniotomy is the workhorse for the majority of pediatric posterior fossa tumors. A midline linear incision is performed extending from the external occipital protuberance down to the posterior arch of C2. The bone of the posterior fossa is exposed together with the foramen magnum. It is important to leave the muscular attachments of C2 intact for cranio-cervical stability. Burr holes are placed on either side of the midline just below the transverse sinuses and a wide craniotomy is performed to expose both cerebellar hemispheres. The posterior arch of C1 is exposed but most often preserved.

The decision to open the foramen magnum as part of the craniotomy depends on the caudal extent of the tumor and the degree of posterior fossa swelling anticipated.

At this point, prior to opening the dura, the degree of swelling within the posterior fossa is anticipated based on the preoperative imaging, the degree and extent of hydrocephalus, and the palpation of the dura. The concern with dural opening in the presence of a swollen posterior fossa is rapid herniation of cerebellar and/or tumor tissue through the defect, which can be complicated by vascular injury and/or brainstem compression and subsequent cardiorespiratory instability. Strategies to reduce swelling include optimizing anesthetic conditions, opening the foramen magnum to decompress the cerebellar tonsils, and early CSF release *via* a dural incision overlying the cisterna magna or creating an intraoperative EVD *via* occipital burr hole (in the case of severe hydrocephalus). For a tumor associated with a large cystic cavity (e.g., pilocytic astrocytoma (PA) or hemangioblastoma with large cyst), collapsing the cyst with a needle puncture and aspiration of cyst fluid reduce swelling. This is performed in a controlled manner leaving the cannula within the cyst as a guide. The dura is opened (this is commonly a Y-shaped opening with a midline linear stem) and hitched laterally exposing the cerebellar hemispheres and the cerebellar tonsils.

For tumors in the midline, two subsequent strategies are available. The transvermian approach utilizes a midline incision through the inferior vermis with subsequent lateral retraction of the two halves of the vermis and the tonsils to expose the dorsal aspect of the tumor. The predominant advantage of this approach is better visualization of the rostral extent of the tumor. The disadvantage is the limited lateral exposure and the occurrence of postoperative cerebellar mutism is believed to be higher with vermian splitting/manipulation. The telovelar approach avoids a vermian split and utilizes the opening of the foramen of Magendie—the outflow of the 4th ventricle—to access the tumor. The cerebello-medullary fissure is entered after dividing the arachnoid between the vermis and one of the cerebellar tonsils. The inferior medullary velum and tela choroidea of the caudal 4th ventricular roof are encountered and opening this allows access to the floor of the 4th ventricle from below. Advantages are the lateral exposure to both lateral foramina of Luschka as well as avoidance of the vermian split. The disadvantage is the limitation to access the most rostral part of the 4th ventricle with tumors extending towards the aqueduct. Of note, this is also the approach to access dorsal brainstem lesions with dorsal exophytic components that breach through the ependymal lining of the 4th ventricle floor and extend into the 4th ventricle (see later). The incidence of cerebellar mutism is also believed to be lower with this approach [16].

For lateral hemispheric tumors, a variant is to perform a paramedian incision with a unilateral craniotomy. This is certainly suitable for smaller laterally placed hemispheric tumors to one side. However, with larger unilateral tumors, most surgeons still perform a midline exposure and a bilateral craniotomy with greater extent of bony exposure on the side of the tumor. This is to allow a larger operating window and decompression, anticipating and accommodating brain swelling. For hemispheric tumors, an incision is then made along the cerebellar folia—transcerebellar approach—and the dissection extends along the trajectory of the shortest distance from the cerebellar surface to the tumor.

2.5 Retromastoid/lateral suboccipital craniotomy

This approach is utilized predominantly for tumors within the CPA. An incision is performed behind the ear with the superior border being the transverse sinus and the lateral border being the sigmoid sinus. The asterion is a bony landmark that approximates the meeting point of the transverse and sigmoid sinuses. A craniotomy is performed, and the dura is opened with flaps pedicled on the transverse and the sigmoid sinuses. Brain relaxation is achieved with CSF release from the cisterna magna and the lateral cerebellopontine cisterns. The cerebellum is retracted, and this provides visualization of the CPA, cranial nerves V through to XII, and the arteries and veins, together with tumors lying within this region.

2.6 Surgical adjuncts

A variety of surgical adjuncts are used to achieve the goal of maximal safe resection. Frameless stereotactic neuronavigation may be used to aid tumor localization although in the posterior fossa, the presence of uniform bone and anatomical landmarks means that this is not used as much as with supra-tentorial tumors. Intraoperative imaging is certainly very useful and may take the form of ultrasound scanning (USS) or increasingly in many centers, intraoperative magnetic resonance imaging (MRI). USS has numerous benefits—it is reasonably inexpensive and fast, provides real-time perioperative guidance, and can help guide tumor resection from start to finish (with visualization of any residual tumor before closure) as well as help with localization of ventricles to insert EVDs. A disadvantage is that interpretation can be difficult and is highly operator-dependent. Intraoperative MRI has been safely utilized in the pediatric population and many proponents suggest that it reduces the number of patients that require early return to theater for revision surgery following postoperative imaging that demonstrates surgically accessible residual tumor. Disadvantages are the expensive infrastructure necessary for its implementation and use within an operating theater. The role of intraoperative neurophysiological monitoring and mapping (IONMM) is crucial in maximizing safe resection by minimizing iatrogenic injury to crucial neurological structures that can compromise functional outcome. It is not routinely required with cerebellar hemispheric tumors.

However, it is particularly relevant during resection of CPA and brainstem tumors, or tumors with proximity to or infiltration of the floor of the 4th ventricle, that therefore require manipulation or working in between cranial nerves and/or their nuclei and ascending/descending white matter tracts, to successfully maximize EOR.

2.7 Postoperative care

Following tumor resection, meticulous hemostasis is obtained. The dura is closed primarily to try and achieve watertight closure (to reduce the risk of complications such as wound leaks together with their antecedent risk of meningitis or infection, and pseudomeningocoeles), or is augmented with autograft (pericranium) or synthetic allograft (e.g., bovine pericardium). The bone flap is replaced and secured with sutures or screws and a careful closure of the muscle, fascia, and skin is performed. If an EVD was placed, it is often kept in place during the immediate postoperative period to allow CSF drainage to optimize wound healing (to avoid CSF leaks) and allow normalization of CSF pathways. The volume of CSF drainage output is a marker of the adequacy of the function of the patient's normal CSF pathways and once it decreases, the EVD is clamped and if the patient tolerates this challenge successfully, it is removed.

2.8 Postoperative hydrocephalus

Despite successful tumor resection, 30% of children will ultimately require permanent CSF diversion [17]. Risk factors for post-resection hydrocephalus requiring surgical treatment include the following: age < 2 years, presence of transependymal edema, pre-resection moderate or severe hydrocephalus, presence of cerebral metastases, diagnosis of medulloblastoma, ependymoma or dorsal exophytic brainstem glioma, and incomplete tumor resection. Patients requiring permanent CSF diversion often present in the early postoperative period with either clinically symptomatic deterioration or in the presence of a perioperative EVD, difficulty with weaning off the EVD. The presence of a postoperative wound leak or tense pseudomeningocele is also a marker of impaired CSF drainage and should initiate assessment for hydrocephalus. Surgical options for permanent CSF diversion include a trial of ETV in the first instance followed by placement of a permanent VPS should this fail. The benefit of this strategy is the avoidance of having a permanent shunt if the ETV is successful. The alternative is the placement of a VPS without trial of ETV. This decision is often based on surgical experience and taking into consideration the probability of ETV success in this context. Factors to consider include the age of the child, tumor type, and ability of arachnoid granulations to reabsorb CSF (something which may be compromised in the very young due to immaturity of development, and with tumors with CSF seeding). The benefit of permanent VPS without ETV allows the child to move directly onto adjuvant therapy without interruption or delay, as opposed to requiring a second procedure owing to failure of ETV occurring during adjuvant therapy and impacting on safety of surgical VPS insertion due to concerns with clotting, hematologic parameters, immune-compromise, and wound-healing.

2.9 Case illustrations

The first patient was a 5-year old girl that presented with headaches and ataxia. Imaging confirmed a right-sided cerebellar hemispheric tumor with superior extension to the tentorium (Fig. 1a, b). She underwent a midline sub-occipital craniotomy (greater exposure on right compared to left) and resection of the tumor. Postoperative imaging confirmed complete excision (Fig. 1c, d) and histology was consistent with PA. The second patient was a 5-month-old infant presenting with irritability, vomiting, and increasing head circumference. Imaging confirmed a lesion arising from the vermis, roof of the 4th ventricle, and possibly infiltrating or effacing the dorsal aspect of the brainstem, eccentric to the right (Fig. 2a). He underwent a midline sub-occipital craniotomy with telovelar approach for complete excision of the tumor (Fig. 2b). Histology was consistent with AT/RT.

3 Brainstem tumors

Primary brainstem tumors encompass approximately 10–20% of primary CNS tumors in children [18]. The majority of brainstem gliomas are diffuse intrinsic pontine gliomas (DIPG), which unfortunately carry a very poor prognosis [19]. The rest are often focal and low grade and with advancements in MRI, image-guided surgery, microsurgical techniques, and IONMM, good long-term results have been achieved after resection of focal gliomas and adjuvant therapy [20, 21]. As well as distinguishing between midbrain, pontine, and medullary lesions, a useful radiological classification subdivides brainstem tumors into 4 types [22] and helps stratify these lesions in terms of the likely diagnosis (and therefore behavior) and also helps determine surgical strategy (Table 2).

3.1 Anatomy

The brainstem (midbrain, pons, and medulla) runs through the supra- and infra-tentorial spaces and with the nexus of cranial and autonomic nuclei, descending (mainly motor) and ascending (mainly sensory) white matter tracts, and is arguably the most eloquent structure within the intracranial compartment. The majority of cranial nerves extend ventrally or laterally into their respective skull base foramina except the 4th cranial nerve which originates from the dorsal midbrain with a long

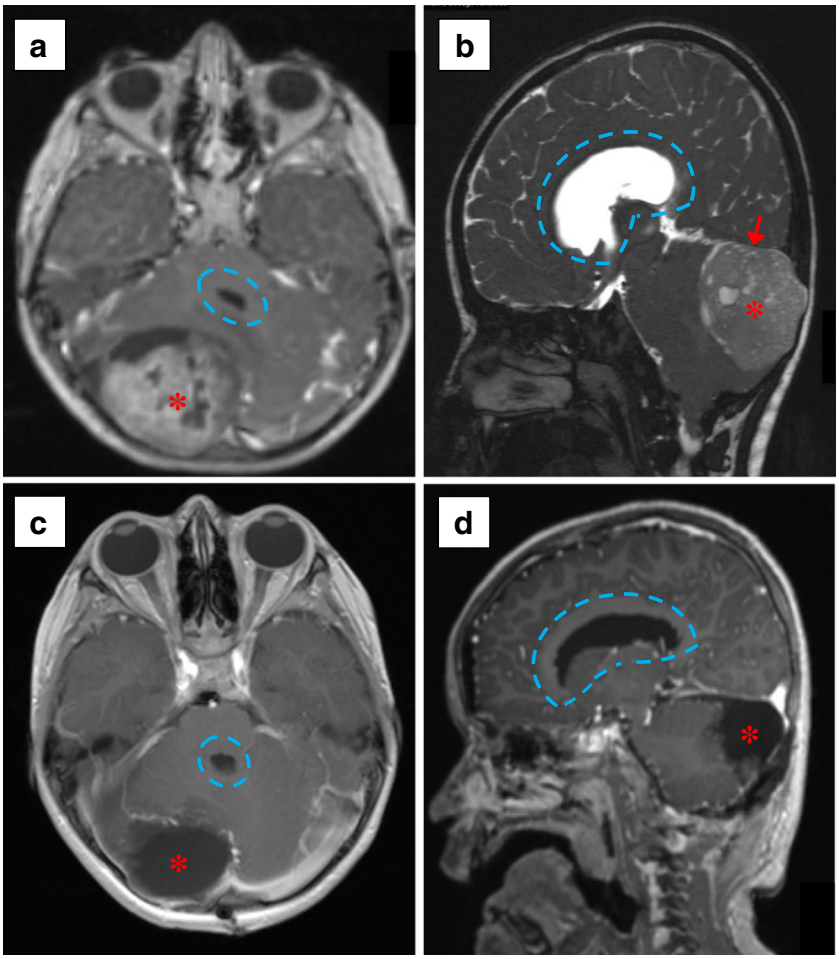

Fig. 1 **a** Axial T1 post-contrast MRI confirming a right cerebellar hemispheric tumor (red asterisk). The 4th ventricle is partially effaced (dashed blue line). **b** Sagittal T2 MRI confirming the cerebellar hemispheric tumor (red asterisk) with upward extension into the tentorium (red arrow). There is dilatation of the supratentorial ventricles (dashed blue line). Histology was consistent with a pilocytic astrocytoma. **c** Axial T1 post-contrast MRI confirming excision of the right cerebellar hemispheric tumor (red asterisk). The 4th ventricle has re-opened and is no longer effaced (dashed blue line). **d** Sagittal T2 MRI confirming the excision of the cerebellar hemispheric tumor via a transcerebellar folia approach (red asterisk). There is no longer dilatation of the supratentorial ventricles confirming resolution of the hydrocephalus (dashed blue lines)

intracranial course passing around the midbrain and cerebral peduncles before moving anteriorly. The arteries and veins of the posterior fossa lie in close relation to the cranial nerves within the subarachnoid CSF cisterns around the brainstem.

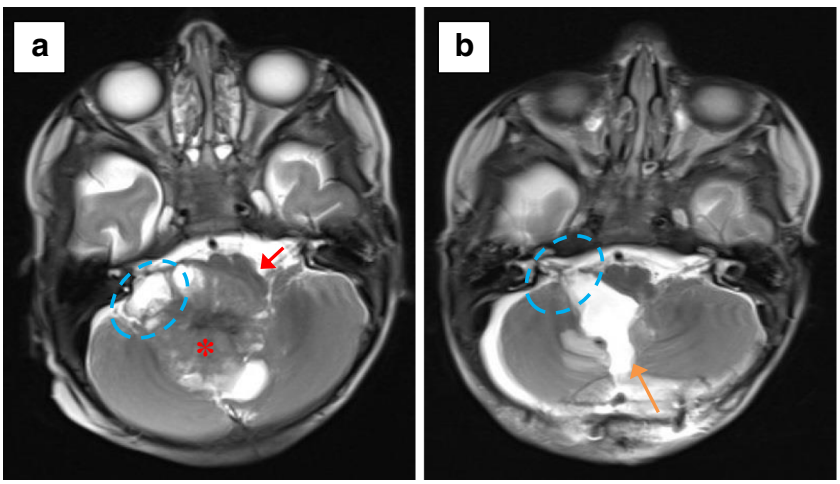

Fig. 2 **a** Axial T2 MRI confirming tumor arising from the vermis and entering the 4th ventricle eccentric to the right (red asterisk) with compression of the dorsal brainstem at the medulla (red arrow) and lateral extension of the tumor through the right foramina of Luschka into the cerebello medullary cistern on the right (dashed blue line). Histology was consistent with atypical teratoid rhabdoid tumor (AT/RT). **b** Axial T2 MRI post-operative confirming complete excision of the tumor via a sub-occipital craniotomy and telo-velar tonsillar approach. The 4th ventricle is open as is the excision of the tumor including the part extending through right Luschka (dashed blue line, top). The space between the right tonsil and vermis was opened to perform the telo-velar approach (orange arrow)

The cerebellum lies on the dorsal aspect of the brainstem and is joined to it *via* three cerebellar peduncles. Two ependymal lined cavities lie between the dorsal brainstem and the cerebellum—the cerebral aqueduct of Sylvius (connects the 3rd to 4th ventricle and lies at the level of the midbrain) and the 4th ventricle (lies at the level of the pons and medulla). Lying just rostral the cerebellum and forming the immediate dorsal surface of the aqueduct is the quadrigeminal plate with the superior and inferior colliculi. The "floor" of the 4th ventricle is essentially therefore the ependyma-lined dorsal brainstem with a number of recognizable undulations and prominences that signify cranial nerve nuclei, white matter tracts, and sulcal separations. The "roof" of the 4th ventricle is formed by superior and inferior medullary veli and base of the cerebellum.

3.2 Management strategy

Surgery is considered predominantly in the context of focal lesions if there is a questionable diagnosis and the lesion is easily accessible (usually presents and breaches through a pial or ventricular ependymal surface) and in the context of a symptomatic patient with or without documented change in interval imaging. A useful treatment paradigm subdivides strategy based on location [23].

3.3 Surgical approaches

The surgical approach depends on the location in the brainstem (midbrain, pons, medulla) as well as whether it is predominantly ventral/anterior, lateral, or dorsal/posterior (Table 3). The most common approach involves access to lesions within the 4th ventricle, and those that are focal and exophytic, breaching and presenting through pial or ventricular ependymal surfaces. For lesions that do not present to the pial or ependymal surface, the combination of anatomic and fiber dissection cadaveric studies integrated with advanced MRI techniques has led to more precise anatomical definition within the brainstem and the delineation of "anatomical safe entry zones" to access these lesions [24–28]. Regardless of the surgical approach used, there are some important tenets in operating on brainstem tumors. The use of surgical adjuncts is critical.

3.4 Advanced imaging techniques

Implementation of advanced MRI such as diffusion tensor imaging (DTI) and fiber tractography prior to surgical decision-making provides useful information on the position of key motor tracts such as the corticospinal tract and how they are displaced or affected by the tumor. These help determine surgical feasibility, guide the surgical approach, and when there is no pial/ependymal breach, locate the optimum and safest place for pial/ependymal incision.

3.5 Intraoperative neurophysiological monitoring and mapping

IONMM is mandatory when operating on intrinsic brainstem lesions with monitoring of motor evoked potentials (MEPs) for corticospinal tract integrity and somatosensory evoked potentials (SSEPs) for dorsal column function and cranial nerves. The cranial nerves can be monitored and mapped (*via* direct nerve stimulation) and include V (temporalis, masseter muscles), VII (orbicularis oris and oculi), VIII (brainstem auditory evoked responses (BAER)), IX and X (laryngeal muscles, soft palate, pharynx), XI (trapezius and sternocleidomastoid), and XII (intrinsic tongue muscles). The VI (lateral rectus) can also be monitored as can the III nerve function, although this is rarely performed. Despite the presence of anatomical safe entry zones in the 4th ventricular floor and dorsal brainstem to avoid cranial nerve nuclei and

Table 2 Choux radiological classification of brainstem tumors

Type	Location, appearance	Likely who grade	Surgical strategy
I Diffuse	• Usually pontine • Infiltrate ascending and descending white matter tracts and cranial nerve nuclei	High grade (III, IV)	Typical cases: radiologic diagnosis, no surgical intervention necessary Atypical cases: biopsy when atypical imaging
II Focal	• Anywhere in brainstem • Localized, clear margins • Solid and/or cystic	Low grade (I, II)	Debulking, especially if present on the pial/ependymal surface
III Dorsally exophytic	• Focal, exophytic • From subependymal glial tissue of the 4th ventricle • Grow dorsally, laterally	Low grade (I, II)	Debulking
IV Cervicomedullary	• Similar to intramedullary spinal cord gliomas	Low grade (I, II)	Debulking, as they usually displace rather than infiltrate tracts

WHO, World Health Organization (indicates grading system referenced)

Table 3 Surgical approaches to brainstem tumors

Location		Surgical approach	Safe entry zone
Midbrain	Ventral/anterior	Cranio-orbito-zygomatic: pterional craniotomy with removal of orbital rim, roof, and zygomatic process; wide Sylvian fissure split to access optic chiasm, carotid, and oculomotor nerve; oculomotor nerve followed back to brainstem.	Anterior mesencephalic: lateral aspect of the cerebral peduncle, lateral to the oculomotor nerve and Medial to the Corticospinal tractl tract
	Lateral	Subtemporal: access superolateral pons and ponto-mesencephalic junction *via* temporal craniotomy and elevation of temporal lobe; splitting the tentorium enables access to incisural space.	Lateral mesencephalic: vertical incision extending from the medial geniculate body to the ponto-mesencephalic sulcus
	Dorsal/posterior	Infra-tentorial supracerebellar or occipital transtentorial Similar to the pineal approaches to the dorsal midbrain and quadrigeminal plate.	–
Pons	Ventral/anterior	Far lateral or endoscopic transclival: both rarely used	–
	Ventro-lateral	Middle fossa subtemporal with anterior petrosectomy (Kawase approach): middle fossa subtemporal craniotomy with elevation of temporal lobe. Drilling of petrous apex and ridge and subsequently splitting the tentorium. Provides access to the petro-clival junction.	Epitrigeminal: medial to the trigeminal root, lateral to the corticospinal tract fibers
	Lateral	Retromastoid craniotomy with CPA approach: excellent access to the lateral CPA cisterns and space	Dorsal/lateral to trigeminal root
	Dorsal/posterior	Midline suboccipital craniotomy with 4th ventricular approach: transvermian or telovelar approach	Suprafacial triangle: above the facial colliculus prominence on the 4th ventricle floor, lateral to the medial longitudinal fasciculus, medial to sulcus limitans Intrafacial triangle: below the facial colliculus prominence on the 4th ventricle floor, lateral to the medial longitudinal fasciculus, superior to the stria medullares
Medulla	Ventral/anterior	Far lateral: suboccipital craniotomy with partial or complete resection of occipital condyle	Anterolateral olivary sulcus: lateral to the pyramids and medial to the superior olive Posterior olivary sulcus: medial to the inferior olive, medial to the inferior cerebellar peduncle
	Lateral	Retromastoid craniotomy with CPA approach	Inferior cerebellar peduncle or lateral medullary zone: inferior to the cochlear nuclei, dorsal / posterior to the vagus and glossopharyngeal nerves
	Dorsal/posterior	Midline suboccipital craniotomy with 4th ventricular approach	Posterior median sulcus at obex: represents superior extension of the midline approach to the spinal cord, access limited by gracile nuclei (lateral)

n/a, not available

whiter matter tracts, the presence of tumor can cause significant distortion and displacement of these structures. Electrophysiological mapping with direct nuclei stimulation is an important adjunct to identify functionally silent entry point on the dorsal brainstem or 4th ventricular floor. Generally speaking, a decrease in amplitude of response by $\geq 50\%$ or increase in latency of response by $\geq 10\%$ should cease surgical manipulation and prompt optimization of anesthetic conditions (e.g., increase blood pressure) until responses recover.

3.6 Tumor removal

The safest entry point for a brainstem tumor is where it is closed to the pial/ependymal surface or, if relevant in those tumors that are exophytic, where the tumor has breached the pial/ependymal surface. Internal debulking of tumor by staying within the tumor margins is critical. There is typically no clear plane between tumor tissue and the normal brainstem tissue and therefore, attempts to create such a plane can lead to significant neurological injury and morbidity.

3.7 Anesthetic considerations

Lastly, when working near the brainstem, especially cranial nerves V, IX, and X, cardiorespiratory instability can occur with the precipitation of arrhythmias (especially bradycardia) and hypo- or hypertensive episodes. Anesthetic support and collaboration can optimize operative conditions, maintain and modulate strict blood pressure control to augment recovery

during neural manipulation-induced IONMM deterioration, and rapidly respond to any perioperative crises.

3.8 Case illustrations

The first patient is a 15-year-old girl presenting with ataxia, multiple cranial nerve palsies, and progressive pyramidal weakness. Imaging confirmed a diffuse lesion in the pontine region. Radiologically, a DIPG was in the differential diagnosis but given her age and presence of some pial breach ventrally, a biopsy was performed to exclude the possibility of a low-grade glioma. Figure 3 demonstrates the lesion within the pons with diffuse infiltration. A transcerebellar biopsy through the middle cerebellar peduncle was performed *via* a sub-occipital burr hole, which confirmed a DIPG. She was started on adjuvant therapy. The second patient presented with progressive upper and lower limb neurology in a pyramidal pattern as well as ocular movement dysfunction and an upper motor neuron facial weakness. Imaging demonstrated a contrast-enhancing lesion that was dorsal and exophytic breaching through the ependymal surface and into the 4th ventricle (Fig. 4a, b). A midline sub-occipital craniotomy with telovelar approach was performed with IONMM. Postoperative imaging demonstrates significant debulking of the tumor, with care to remain within the enhancing capsule of the tumor to avoid iatrogenic brainstem injury (Fig. 4c). The patient made a good clinical recovery and low-grade glioma was confirmed.

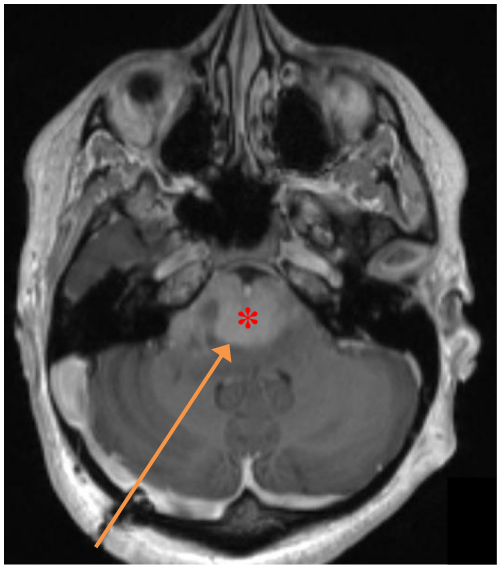

Fig. 3 Axial T1 post-contrast MRI confirming a heterogeneous diffusely enhancing lesion within the pons (red asterisk). The lesion was biopsied via a transcerebellar approach with a right suboccipital burr hole (orange arrow) to confirm the diagnosis of a diffuse intrinsic pontine glioma (DIPG)

4 Pineal region tumors

Tumors of the pineal region account for a higher proportion of CNS tumors in children compared with adults (Table 4), but still only make up 0.4–2% of all primary CNS tumors in children [29, 30]. The pineal gland arises from a diencephalic diverticulum during embryogenesis and is predominantly composed of pinealocytes (melatonin producing neuro-secretory cells) believed to regulate circadian rhythms. Located deep in the brain, it is surrounded by numerous critical structures and thus, an intimate and working knowledge of which is crucial in safe navigation to and resection of tumors from this region. These include the following.

4.1 Management strategy

Management paradigms depend on the acute nature of the clinical presentation, the presence and severity of hydrocephalus, imaging appearances, and presence or absence of serum and/or CSF tumor markers to diagnose nonoperative GCTs. Pathognomonic presentations highly suggestive of diagnosis may allow application of therapeutic paradigms without the need for initial tumor tissue biopsy. For instance, synchronous bifocal tumors in the pineal and sellar/suprasellar regions in the absence of elevated GCT markers suggest germinoma. By contrast, elevation of malignant GCT markers (α-fetoprotein for yolk sac tumor, β-human chorionic gonadotrophin for choriocarcinoma) suggests a malignant GCT. Bilateral retinoblastomas with a pineal lesion otherwise known as "trilateral retinoblastomas" are often pineal tumors and are managed accordingly. Importantly, classic radiological findings of meningioma and epidermoid cyst enable upfront surgical resection without the need for diagnostic biopsy.

In almost all other scenarios where diagnosis is uncertain, tissue biopsy is necessary to establish histologic diagnosis prior to proceeding with treatment. Treatment options include surveillance, diagnostic tissue biopsy, surgical resection, focal and/or craniospinal radiation, and chemotherapy, with these modalities used either in isolation or combination depending on histopathological diagnosis. CSF sampling can be performed *via* lumbar puncture in the absence of obstructive hydrocephalus but most commonly, the presence of compression of 3rd ventricular outflow contraindicates lumbar puncture and necessitates the need of cranial access for CSF.

4.2 Hydrocephalus treatment

Acute hydrocephalus in the presence of an acutely deteriorating conscious level requires prompt stabilization and immediate CSF diversion. This is often by way of EVD

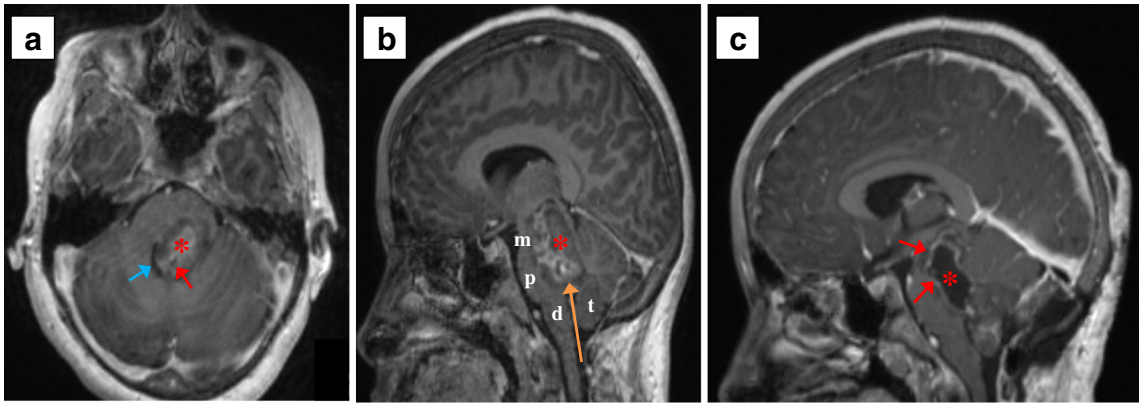

Fig. 4 **a** Axial T1 post-contrast MRI confirming brainstem tumor (red asterisk) with a dorsally exophytic component (red arrow) breaching through the ependymal lining of the 4th ventricle filling most of the ventricle. A small part of the compressed 4th ventricle is seen (blue arrow). Histology confirmed a pilocytic astrocytoma. **b** Sagittal T1 post-contrast MRI demonstrating the brainstem tumor (red asterisk). The cerebellar tonsil is visualized ("t"), as are the parts of the brainstem: midbrain ("m"), pons ("p"), and medulla ("d"). The trajectory for the telo-velar-tonsillar approach via the foramen of Magendie is demonstrated (orange arrow). **c** Sagittal T1 post-contrast MRI confirming near complete excision of the brainstem tumor (cavity indicated by red asterisk) via a sub-occipital craniotomy and a telo-velar-tonsillar approach. Per the principles of intrinsic tumors in the brainstem, debulking has been performed intracapsularly leaving a thin rim of enhancing tumor capsule which is adherent to the brainstem (red arrows). This technique avoids traction injury to the brainstem

placement in crisis. If surgical resection is not likely to occur soon, the primary concern should be interval CSF diversion, and in a clinically and neurologically more stable patient, ETV is the procedure of choice, which often avoids the need for implantation of a permanent VPS. Often definitive surgical or non-surgical treatment of the tumor results in decreased tumor size with restoration of normal CSF flow dynamics [31].

4.3 Tissue diagnosis

When tumor markers in the serum and CSF are negative, tissue biopsy is required to determine how the tumor should be managed. Tumor biopsy is often undertaken endoscopically at the same time as an ETV. Open microsurgery, as a prelude to debulking/excision, is also used for tissue diagnosis and also provides a larger specimen. This advantage with the latter approach is particularly valuable given the heterogenous and often mixed nature of pineal tumors. Open microsurgery for biopsy may be necessary in the absence of hydrocephalus. Stereotactic needle biopsies although demonstrated to be safe in large patient series [32, 33] are now rare with the proximity of critical structures limiting its use together with the high risk of sampling error or non-diagnostic yield in the absence of direct visualization. Trajectory planning needs to account for avoidance of deep veins and can be performed with a frontal paramedian technique or lateral temporoparietal approach. In cases of clearly resectable tumors, the risks of biopsy should be avoided. The preferred approach is open biopsy and resection at the same time.

Table 4 Spectrum of lesions within the pineal region. Roman numerals indicate grade according to World Health Organization (WHO) classification

Pineal tumors and local lesions		
Germ cell tumors (GCTS)	Germinomatous	• Germinoma • Atypical teratoma
	Non-germinomatous	• Choriocarcinoma • Embryonal cell carcinoma • Yolk-sac/endodermal sinus tumor
Pineal parenchymal tumors	• Pineocytoma (I) • Pineal parenchymal tumor of intermediate differentiation (II, III) • Pineoblastoma (IV)	
Neural support structure origin (e.g., glial, arachnoid cap cells)	• Glioma/glioblastoma • Meningioma • Hemangiopericytoma	
Non-neoplastic lesions (to exclude)	• Pineal cyst • Arachnoid cyst • Dermoid/epidermoid cyst • Vascular (e.g., vein of Galen aneurysmal malformation)	

4.4 Endoscopic approaches

Endoscopic transventricular surgery is well established and has added to the neurosurgeon's armamentarium in management of pineal region tumors [34] allowing direct visualization for tumor biopsy and tissue sampling, allowing CSF sampling for tumor markers, and allowing CSF diversion to be established with ETV to treat obstructive hydrocephalus. In the setting of the classically observed aqueductal compression and 3rd ventricular outflow obstruction, ETV creates a stoma in the floor of the 3rd ventricle to allow CSF to bypass the obstruction and drain directly into the basal cisterns allowing reabsorption *via* the arachnoid granulations. This may obviate the need for permanent CSF diversion techniques such as a VPS together with its antecedent complications.

The ETV is usually performed first rather than the biopsy as establishment of CSF diversion is critical and there is a risk that bleeding following a biopsy if performed first may prevent the subsequent creation of the stoma in the 3rd ventricle floor by obscuring visualization, necessitating the need to insert an EVD for CSF diversion. The availability of flexible endoscopes allows both procedures to be performed using a single burr hole due to improved maneuverability; however, the visualization and optics may not be as high as that of a rigid endoscope. The disadvantage of the latter is that very often, two burr holes are required, one for the ETV and a second for the biopsy *via* a different trajectory.

The patient is positioned supine in the neutral position. Frameless stereotactic neuro-navigation can be used for guidance. An incision is made in the right frontal region 3 cm lateral to the midline and 1 cm anterior to the coronal suture. A burr hole is sited with a drill and the endoscope is introduced into the body of the lateral ventricle. After identification of anatomical landmarks, the endoscope is passed through the foramen of Monroe and into the 3rd ventricle, with care to not damage the fornix or critical diencephalic draining veins (the thalamostriate vein and internal cerebral vein). The perforation is made in the region of the floor of the 3rd ventricle to enter the interpeduncular and basal cisterns, confirmed usually through visualization of the basilar artery and the posterior border of the clivus. A second burr hole is then placed anterior to the coronal suture and the endoscope is introduced in a posterior trajectory to enter the frontal horn of the lateral ventricle. Careful passage through the foramen of Monroe into the 3rd ventricle and subsequently inferior to the inter-thalamic adhesion (massa intermedia) within the 3rd ventricle allows visualization of the tumor for tissue sampling.

Risks of surgery include excessive hemorrhage and subsequent obstruction of the 3rd ventricular floor stoma with need for an external ventricular drain, retraction injury to the ipsilateral forniceal column during endoscope passage with possible short-term memory impairment, and most seriously, damage to the basilar artery or its perforators, which can result in catastrophic life-threatening hemorrhage. The latter is fortunately extremely rare.

4.5 Open microsurgery

There are three predominant approaches to the pineal region, each with its own advantages and disadvantages: infratentorial supracerebellar, occipital transtentorial, and posterior interhemispheric *via* a transcallosal or retrocallosal route. The route is determined by extent of the lesion and its relationship to critical neuro-vascular structures (in particular the great veins of the region), the aims of surgery, and surgeon familiarity and comfort [35, 36]. The more common anatomical arrangement is for the large veins to be displaced superiorly, which makes the infra-tentorial supracerebellar approach amenable. Inferior displacement of the veins is less common, and this arrangement is more conducive to the supratentorial approaches [37], especially in the context of tumors with an extensive supratentorial component and extension into the lateral ventricles and atrium. Rarely, a combination of approaches in staged fashion is required.

4.6 Infra-tentorial supracerebellar approach

This approach is performed either in the sitting or the prone position. A midline incision is made, and a sub-occipital craniotomy exposes the undersurface of the transverse sinuses and torcula. The dural flap is retracted superiorly, and bridging veins between the tentorium and superior cerebellar surface are divided. This technique provides the most direct route to the tumor epicenter and the natural plane below the tentorium/cerebellum with minimal brain retraction. There is also minimal risk of damage to deep venous structures when displaced superiorly by the tumor. The dorsal midbrain structures (superior and inferior colliculi) lie inferiorly. Surgical resection when complete allows ventricular entry *via* the posterior 3rd ventricular wall. The disadvantages are the limited lateral exposure and long working distance [38]. Operating in the sitting position has the advantage of brain relaxation but at the increased risk of VAE that can have significant cardiac complications including perioperative cardiorespiratory arrest. It is contraindicated in the presence of a patent foramen ovale that must be excluded preoperatively.

This approach is performed either in the sitting or prone position. A midline incision is made and a sub-occipital craniotomy exposes the inferior surface of the transverse sinuses and torcula. The dural flap is retracted superiorly, taking care not to occlude the transverse sinus. Bridging veins between the tentorium and superior cerebellar surface are divided, allowing the cerebellum to fall as the tumor is approached. It provides the most direct route to the center of the tumor, and the natural plane below the tentorium and cerebellum with minimal brain retraction results in minimal injury to normal

tissue. There is also minimal risk of injury to deep venous structures when they are displaced superiorly by the tumor. Surgical resection, when complete, allows entry into the third ventricle through its posterior wall; the tectum and colliculi of the dorsal midbrain are inferiorly located. The disadvantages are the limited lateral exposure and long working distance [38].

Operating in the prone position has advantages such as the comfortable operating position for the surgeon and the low risk for venous air embolism (VAE). A disadvantage is the swelling within the posterior fossa owing to the higher venous pressures and the need to retract the superior cerebellar surface inferiorly. This is the main advantage of operating in the sitting position where gravity allows the brain the fall away without retraction with significantly relaxed operating conditions, in addition to gravity-assisted venous drainage meaning venous pressures are low and the operative field is reasonably dry with little blood pooling. The predominant risk with the sitting position is the high risk of VAE especially in the context of large osseous and occipital sinuses (especially in children) that can have significant cardiac complications including perioperative cardiorespiratory arrest. The sitting position is therefore contraindicated in the presence of a patent foramen ovale which must be excluded preoperatively. Excessive CSF drainage away from the ventricular system can also cause significant pneumocephalus and subdural hematomas.

4.7 Occipital transtentorial approach

The occipital transtentorial approach is usually performed in the prone or lateral position. A unilateral occipital parasagittal craniotomy is made, and the operative corridor is developed between the medial occipital lobe laterally, the falx cerebri medially, and the tentorium inferiorly. Division of the anterior two-thirds of the tentorium, 1 cm lateral to the straight sinus, allows a wide exposure of the superior cerebellum, as well as the tumor in the pineal region, with the deep veins located anteriorly and superiorly. The disadvantages of this approach include the high risk of visual field deficits due to retraction of the highly eloquent occipital lobe and damage to the calcarine visual cortex; this can be avoided by adequate drainage of ventricular CSF, positioning the patient in the lateral position with the craniotomy side down, allowing the occipital pole to fall by gravity, and minimizing any direct retraction on the brain parenchyma. There is also a risk to the trochlear nerve as it arises posterior from the dorsal midbrain and enters the tentorium.

4.8 Posterior interhemispheric trans- or retrocallosal approach

This approach is rarely used and is primarily the option for lesions located within the posterior part of the 3rd ventricle, with inferior displacement of the deep venous system, such that the lower angle of approach with an infra-tentorial supracerebellar approach requires excessive manipulation and risk to these structures. It is usually performed in the lateral position *via* a parasagittal craniotomy centered over midline and extending laterally over the medial parietal region. The dissection between the falx and medial parietal lobe allows access to the interhemispheric fissure and subsequently *via* either a small callosotomy (transcallosal) or behind the splenium of the corpus callosum (retrocallosal), access to the pineal region. The major disadvantages are risk to bridging veins between the superior sagittal sinus and parietal lobe (which if damaged could lead to significant venous infarction), medial parietal lobe retraction, and risk of disconnection syndromes that occur with the need for a posterior callosotomy in the transcallosal approach.

4.9 Case illustration

An 8-year-old boy presented with features of raised ICP and ocular movement abnormalities consistent with Parinaud's syndrome. Initial MRI confirmed a large pineal mass (Fig. 5a) with hydrocephalus. He underwent ETV and endoscopic tumor biopsy with CSF markers confirming AFP-secreting teratoma. He was started on chemotherapy but interval imaging 4 weeks showed no reduction in tumor size despite resolution of hydrocephalus with patent ETV (Fig. 5b). He underwent surgical excision with an infra-tentorial supracerebellar approach with postoperative imaging confirming complete excision (Fig. 5c).

5 Intraventricular tumors

Intraventricular tumors are rare (0.8–1.6% of all CNS tumors) but occur more commonly in the pediatric population and make up to 16% of all pediatric intracranial tumors [3]. They are subdivided into primary (arise from neuroglial tissue within the ventricle) or secondary/paraventricular (arise from brain parenchyma outside the ventricle and bulge into the ventricle, with by definition greater than two-thirds of tumor volume in the ventricle) tumors (Table 5) [39, 40].

The commonest tumors in the youngest patients are choroid plexus papillomas (CPPs) of the lateral ventricles, while in the older pediatric age group, low-grade gliomas (LGG) such as ependymomas and astrocytomas predominate [41, 42]. Genetic predisposition has been shown to be associated with certain ventricular tumors including subependymal giant cell astrocytoma (SEGA) in tuberous sclerosis and choroid plexus carcinoma (CPC) in rhabdoid predisposition syndrome [43]. The majority of tumors are benign and given that they occupy a non-functional space, are able to grow into a considerable size before clinical manifestations, often presenting

Table 5 Categorization of intraventricular tumors

Intraventricular tumors		
Primary: Neuroglial tissue within the ventricle	Ependyma	Ependymoma
	Subependymal glia	Subependymoma
	Choroid Plexus Epithelium	Choroid plexus papilloma (CPP), choroid plexus carcinoma (CPC)
	Astrocytes	Intraventricular astrocytoma, subependymal giant cell astrocytoma (SEGA)
	Neuronal	Central neurocytoma
	Arachnoid supporting tissue	Intraventricular meningioma
Secondary (paraventricular): surrounding brain parenchyma and bulge into ventricle (by > 2/3 of tumor volume)	• Medulloblastoma (extension into 4th ventricle) • Astrocytoma – hypothalamic-optic pathway, thalamic, tectal plate lesions (extension into 3rd ventricle) • Craniopharyngioma (extension into 3rd ventricle) • Pineal gland tumors – including germ cell tumors and pineal parenchymal tumors (extension into posterior 3rd ventricle) • Meningioma	

with acute or chronic hydrocephalus. In the neonatal period, this may manifest with a tense fontanelle or rapidly increasing head circumference. Given their benign nature, they are often treated surgically but their deep-seated and central location including close relationship to deep structures and vasculature as well as being enclosed by sometimes eloquent cerebral cortex means that they pose a significant challenge to neurosurgeons [44].

5.1 Management strategy

Maximal safe resection is often the aim with the majority of ventricular tumors and GTR can often be curative. Ependymomas most commonly originate in the floor of the 4th ventricle in children, and EOR is the most significant prognostic factor [45]. Subependymomas are benign lesions with a low recurrence rate after GTR. Complete resection is known to be curative for CPPs, with a 5-year survival of nearly 100%. By contrast, CPCs are frequently invasive with relatively poor prognosis [46] with a 5-year overall survival (OS) of 40–50%. Uniquely, SEGAs have been effectively treated with everolimus (mTOR inhibitor) without significant toxicity. Patients often do not require surgical intervention post-everolimus treatment [47].

5.2 General principles

By virtue of their central location, a myriad of surgical approaches is available to access the ventricles (Table 6). They all have advantages and disadvantages given the complex

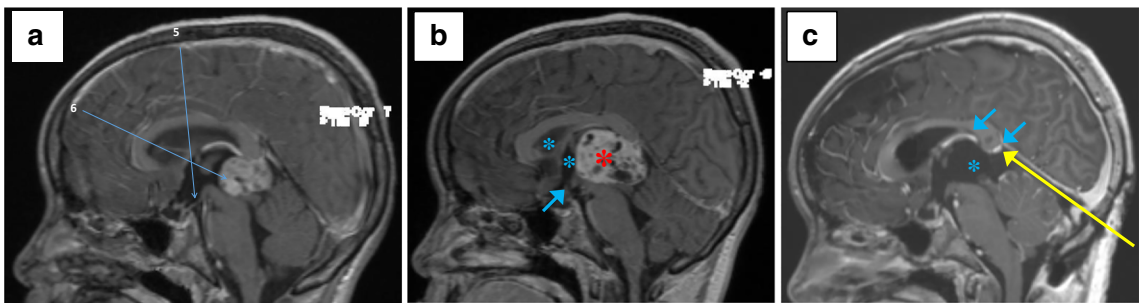

Fig. 5 **a** Arrow 5 – Represents the direction and trajectory for an Endoscopic third ventriculostomy (ETV) through the foramen of Monroe through the floor of the 3rd ventricle to communicate CSF with the basal cisterns bypassing the pineal tumour related obstruction of the Aqueduct. Arrow 6 – Represents the direction and trajectory for an Endoscopic guided Biopsy of a Pineal Region tumour utilising a 2nd frontal pre-coronal suture Burr hole when performing this using a rigid endoscope. Note, Both Biopsy and 3rd Ventriculostomy can also be performed via a single Burr-hole utilising a Flexible endoscope. **b** Interval imaging 4 weeks following ETV, endoscopic biopsy with subsequent diagnostic confirmation of teratoma, and initiation of chemotherapy. The 3rd ventricular stoma is visualized (blue arrow) together with a decrease in size of the lateral and 3rd ventricles (blue asterisks). Tumor size has increased consistent with a Growing Teratoma Syndrome (red asterisk). **c** Post-operative imaging following resection of tumor via an infra-tentorial/supra cerebellar approach. The surgical corridor between the tentorium and superior cerebellar surface is visualized (yellow arrow). The posterior wall of the 3rd ventricle has been opened (blue asterisk) with no evidence of residual tumor. The deep draining veins are visualized and preserved (3)

Table 6 Surgical approaches to intraventricular tumors

Location	Approach	Benefits, limitations	Complications
Lateral ventricle			
Frontal horn and anterior body	Transcortical (superior frontal gyrus)	Benefits: enlarged ventricles (hydrocephalus) facilitates access, orientation Limitations: difficult with small ventricles, difficult to access contralateral ventricle, cannot be used to access mid-body (eloquent sensorimotor cortex overlies with risk of hemiparesis)	Seizures (with any trans-cortical approach)
Mid-body	Transcallosal (anterior)	Benefits: avoids need for cortical transgression, access possible even with small ventricles, bilateral ventricular access Limitations: risk of injury to cortical draining veins to sagittal sinus	Disconnection syndromes
Atrium and trigone	Transcortical (superior parietal lobule)	Benefits: enlarged ventricles with hydrocephalus facilitates access Limitations: orientation challenging, difficult with small ventricles	Seizures (with any trans-cortical approach) Optic radiation injury (visual field deficit) Parietal lobe dysfunction (apraxia, acalculia)
	Transcallosal (posterior)	Benefits: avoids need for cortical transgression Limitations: risk to cortical draining veins to sagittal sinus	Disconnection syndromes
Temporal horn	Transcortical (middle temporal gyrus)	Benefits: short trajectory, relative non-eloquence, early identification of anterior choroidal artery, enlarged ventricles with hydrocephalus facilitates access Limitations: poor visualization posterior choroidal artery, risk of injury to temporal horn structures (hippocampus, optic tract/radiation), mesial border is near eloquent structures (brainstem, posterior cerebral artery, deep veins)	Seizures (with any trans-cortical approach) Optic radiation injury (visual field deficit) Dominant hemisphere speech and memory dysfunction
3rd ventricle			
Anterior	Endoscopic	Benefits: minimally invasive, enables biopsy, enables fenestration to establish CSF drainage pathways Limitations: difficulty to control bleeding, manipulation of fibrous/vascular tumors difficult	Seizures (as utilizes a trans-cortical access)
	Transcallosal (foramen of Monro, interforniceal, trans-choroid fissure)	Benefits: avoids need for cortical, transgression, orientation Limitations: risk to cortical draining veins to sagittal sinus, long working depth, lesions anterior to foramen of Monro / inferior to 3rd ventricle difficult	Disconnection syndromes Injury to fornix (memory complications) Injury to deep diencephalic draining veins
	Pterional Lateral sub-frontal	Benefits: lesions inferior to 3rd ventricle, suprasellar lesions within anterior 3rd ventricle Benefits: midline suprasellar lesions with encroachment of 3rd ventricle	Central neurovascular structure dissection (optic chiasm, carotid vessels and bifurcation) Injury to 3rd ventricular floor (thin)
Posterior	Infra-tentorial supracerebellar	Benefits: midline access Limitations: difficult if significant lateral extension or supra-tentorial extension, long working depth	See discussion on pineal approaches
	Occipital transtentorial	Benefits: good for accessing supra- and infra-tentorial compartments with extensive lesions Limitations: tentorial splitting carries risks (e.g., 4th cranial nerve injury), mesial occipital lobe retraction with risk of visual field deficit	See discussion on pineal approaches
4th ventricle			
Floor/roof	Suboccipital craniotomy and telovelar approach	Benefits: access *via* opening of 4th ventricle, avoids vermian splitting, familiarity of approach Limitations: rostral part of 4th ventricle (including Aqueduct of Sylvius) difficult to access, cistern beyond foramina of Luschka difficult to access laterally	Risk to posterior inferior cerebellar artery (PICA)

functional anatomy and vasculature surrounding the ventricles [40]. The optimal approach is determined following careful preoperative evaluation of the imaging, tumor size and location within the ventricle, ventricular size and dilatation, tumor vascular supply, and surrounding eloquent neural structures including the cerebral cortex. The ideal approach provides sufficient exposure to allow complete tumor removal, early identification of eloquent traversing vessels that must be preserved and tumor vascular supply for interruption to de-vascularize the tumor, minimal damage to or transgression of functional eloquent neural parenchyma, and the most direct shortest trajectory to the tumor.

5.3 Endoscopic approaches

Endoscopic approaches are highly attractive by virtue of their minimally invasive approach, ability to biopsy under direct visualization, and ability to open up and restore normal CSF pathways by fenestration. However, limitations such as difficulty in removing tumors with high vascularity and fibrous consistency, difficulty mobilizing tumors away from neurovascular structures safely, and difficulty with stopping bleeding mean that endoscopic resection is very selectively used. Their predominant use is in fenestration of the ventricular septum pellucidum to communicate ventricular CSF pathways (allowing the use of single rather than multiple CSF shunts if needed), tumor biopsy with or without ETV, and aspiration of tumor cysts [48].

5.4 Open microsurgery

Open microsurgical approaches are often favored for the majority of intraventricular tumors (Table 6). The lateral ventricles are surrounded by the cerebral parenchyma and transgression of eloquent cortex should be avoided if possible. Approaches to the lateral ventricle are generally transcortical (through non-eloquent parenchyma) or *via* the interhemispheric fissure (avoiding a cortical incision) through a small corpus callosotomy (transcallosal). The structures within the lateral ventricles to be identified include the thalamus and caudate nucleus, the veins lining the ependyma (thalamostriate and septal veins that together form the internal cerebral vein lying proximal to the foramen of Monroe) and the fornix. The 3rd ventricle is in intimate relationship with the hypothalamus, thalamus, fornix, and deep draining veins. The access to the 3rd ventricle is *via* the lateral ventricle (transcortical or transcallosal) or for the posterior 3rd ventricle, *via* approaches similar to the pineal gland (located on the posterior 3rd ventricle wall). The 4th ventricle floor consists of the dorsal aspect of the brainstem and contains a number of prominences that relate to the cranial nerve nuclei and ascending and descending white matter pathways. The base of the cerebellum and superior and inferior medullary veli form the 4th ventricular roof. It is usually accessed *via* a posterior fossa sub-occipital craniotomy and through the natural opening of the 4th ventricle into the subarachnoid cisterns—the foramen of Magendie—*via* the telovelar approach.

5.5 Adjuncts

The deep location of tumor vascular supply means it is encountered relatively late during removal of intraventricular tumors. Preoperative endovascular embolization of tumor vessels if possible is greatly beneficial to de-vascularize the tumor and should be always considered in intraventricular tumors, especially with choroid plexus tumors where in a very young infant, significant blood loss can be fatal.

5.6 Case illustration

A 3-week-old neonate presented with irritability, tense fontanelle, increasing head circumference, and "sun setting" eyes. CT imaging demonstrated a large right lateral intraventricular tumor radiologically consistent with CPP (Fig. 6a). An emergency left-sided EVD was inserted for CSF diversion. Intracranial angiography demonstrated predominant supply *via* the medial posterior choroidal arteries from the posterior cerebral arteries (Fig. 6b) and vessel embolization was performed (Fig. 6c). The post-embolization CT demonstrated necrotic change in the tumor consistent with the loss of its vascular supply (Fig. 6d). A trans-cortical frontal approach was performed and GTR of the tumor was achieved (Fig. 6e). Histology confirmed CPP.

6 Sellar and supra-sellar tumors

Sellar and supra-sellar tumors constitute 10% of all primary CNS tumors in children [49] with 90% of these being craniopharyngiomas and the remaining 10% pituitary adenomas or Rathke cleft cysts. There are numerous critical neurovascular structures within the sellar and supra-sellar regions, and the relationship of the tumor to these determines surgical strategy and approach, as well as the common clinical phenotypic patterns of presentation of these tumors [50] (Table 7).

6.1 Anatomy

The pituitary gland lies within the sella turcica or pituitary fossa beneath a dural covering known as the diaphragma sellar. The region below the diaphragma is referred to as the infra-diaphragmatic/infra-sellar region (or simply sellar). The region above the diaphragma is the supra-sellar region. The floor of the pituitary fossa is formed by the sphenoid bone; the aeration and degree of pneumatization of which vary. The trans-sphenoidal route utilizes the natural opening of the

Table 7 Open microsurgical approaches to craniopharyngioma

Approach	Technique	Benefits, limitations
Craniopharyngioma		
Pterional	• Following scalp incision and craniotomy, sphenoid ridge is flattened to provide optimal access to the skull base and Sylvian fissure • The Sylvian fissure is widely opened from medial to lateral or *vice versa*. • With a medial to lateral approach, a sub-frontal trajectory is used to identify the optic nerve and internal carotid artery • Opening the chiasmatic cisterns and the optico-carotid cistern for CSF release achieves brain relaxation • Wide opening of the Sylvian fissure allows identification of key neuro-vascular landmarks including the ipsilateral optic nerve, internal carotid artery and its terminal branches and the bifurcation. • The tumor capsule together with cysts is identified • Intra-capsular debulking of solid components and cyst aspiration is alternated with development of arachnoidal plane between tumor and neurovascular structures and progressively debulk the tumor. • Tumor debulking occurs within the arachnoidal cisterns of the optico-carotid (between optic nerve and internal carotid artery) and carotid-oculomotor (between internal carotid artery and oculomotor nerve)	Benefits: familiar approach to most neurosurgeons, unilateral approach with no need to cross the midline, unilateral approach and identification of contralateral neuro-vascular structures (optic nerve and internal carotid artery) occurs late Limitations: Sylvian fissure splitting can be difficult, sub-frontal and temporal lobe retraction may be needed, limited use for suprasellar tumors with a significant vertical component
Sub-frontal	• Similar scalp incision to pterional approach • Approach is more anteriorly orientated and utilizes more frontal bone drilling to flatten the fronto-orbital roof to achieve flatter trajectory along anterior fossa floor	Benefits: unilateral approach with no need to cross midline Limitations: unilateral approach and delayed identification of contralateral neuro-vascular structures, sub-frontal retraction
Bi-frontal anterior interhemispheric	• Utilizes a bi-coronal incision and a craniotomy that crosses the midline following by a dural opening and division of the superior sagittal sinus anteriorly • Approach is *via* the interhemispheric space between the two frontal lobes to initially identify the anterior skull base in the midline followed by the tuberculum sellae and then the optic nerves and chiasm • Facilitated by presence of a post-fixed optic chiasm (the chiasm is far away from the Tuberculum sellae with a large prechiasmatic space) – this allows the tumor to be debulked anterior to the chiasm with relative ease. • Allows access to tumors with significant vertical extension that are difficult to reach with a purely pterional or sub-frontal approach	Benefits: midline trajectory and orientation, bilateral access to neurovascular structures (optic nerves, chiasm and internal carotid artery) early, minimal sub-frontal retraction Limitations: need to cross the midline and divide superior sagittal sinus, small risk of venous infarction affecting mesial frontal lobes, pre-fixed optic chiasm with minimal or no prechiasmatic space is a relative contraindication
Interhemispheric transcallosal	• Utilizes a midline craniotomy and approach *via* the interhemispheric space between the falx cerebri and the mesial frontal lobe to access the corpus callosum • Corpus callosotomy to enter the lateral ventricle from which the 3rd ventricle can be reached *via* a trans-foraminal approach (or transchoroidal, interforniceal) • Cannot be used to access lesions below the 3rd ventricle floor and anterior to the 3rd ventricle	Benefits: predominantly used for tumors within the 3rd ventricle Limitations: need to cross the midline, risk to midline draining veins, risks with callosotomy, risk to fornix with possible short-term memory impairment
Subtemporal	• Rarely indicated in most circumstances • Temporal craniotomy and middle fossa approach to access the retro-chiasmatic space and the interpeduncular cisterns • Tentorium splitting can be performed to subsequently access the petro-clival region	Benefits: access to tumor components and cysts with significant retrochiasmatic extension and into the posterior fossa Limitations: temporal lobe retraction, risk to temporal draining veins including vein of Labbe, risk to 4th nerve if tentorium splitting is needed

sphenoid sinus to access sellar tumors. Laterally, either side of the pituitary fossa, enclosed by dural coverings, is the cavernous sinuses within which lie the cavernous segment of the internal carotid artery; cranial nerves III, IV, and VI; and ophthalmic division of V. The pituitary stalk extends superiorly through an opening in the diaphragma towards the floor of the 3rd ventricle. Within this supra-sellar region, the pituitary stalk is intimately related to a variety of structures including the optic chiasm, nerves, and tract; the internal carotid artery together with its terminal branches (posterior communicating artery, anterior choroidal artery) and bifurcation; and the 3rd ventricle floor together with the hypothalamus. Tumors therefore extending into the supra-sellar region can displace/distort these structures with associated clinical sequelae. There may also be extension from the supra-sellar region to the anterior, middle, and even posterior cranial fossae.

6.2 Clinical presentation

Tumors within this region may present with non-specific features of raised ICP, hydrocephalus secondary to 3rd ventricular compression (with either aqueduct compression or foramen of Monro compression), visual impairment due to compression of the optic apparatus, hyper-secretion or hypo-secretion of pituitary hormones (together with their clinical phenotypes and sequelae), and even pan-hypopituitarism. The proximity of the hypothalamus and its displacement and compression can be a major source of morbidity for patients in the context of these tumors. Clinical features such as behavioral dysfunction, diabetes insipidus, growth failure, metabolic syndrome with obesity, and thermoregulation dysfunction may be the presenting symptoms of these tumors or even be complications following treatment.

6.3 Management strategy

Sellar and supra-sellar lesions are often approached by either open cranial microsurgery, trans-sphenoidal, or a combination approach. Broadly speaking, the degree of supra-sellar extension and relationship of the optic chiasm and its displacement in relation to the tumor govern the approach. Trans-sphenoidal approaches suffice for the majority of pituitary adenomas and Rathke cleft cysts as well as craniopharyngiomas that are predominantly infra-diaphragmatic/infra-sellar, where the diaphragma sella drapes over the tumor and separates it from supra-sellar structures such as the optic chiasm, circle of Willis, and hypothalamus. The presence of significant supra-sellar extension, lateral extension beyond the internal carotid arteries, evidence of significant supra-sellar calcification (in craniopharyngiomas), increased adhesion to vascular structures and hypothalamus, and inferior displacement of the optic chiasm necessitates open microsurgical approaches (e.g., pterional, lateral sub-frontal, or anterior interhemispheric), as does other anatomical constraints (e.g., lack of pneumatization of the sphenoid sinus, anticipated difficulties with skull base reconstruction following extended trans-sphenoidal approaches).

6.4 Trans-sphenoidal approach

While neurosurgeons have routinely used the microscope for the trans-sphenoidal approach, the advancements in optics and illumination have led to the endoscope becoming far more common. The microscope has advantages of depth perception but is limited by a narrow field of view. The endoscope has a significantly wider field of view as well as enhanced illumination capability. The availability of angled endoscopes further increases visualization and access. The trans-sphenoidal approach also has the advantages of being able to reach sellar tumors with minimal disruption to normal neurovascular structures, minimal brain retraction, early decompression and minimal manipulation of the optic apparatus, and avoidance of a cranial incision with less postoperative pain and possibly shorter hospital stays. The major disadvantage of trans-sphenoidal approaches is the degree of skull base bone removal required, particularly in extended approaches, and the associated difficulty in reconstruction of anatomic barriers between the sino-nasal compartment and the intradural space. The result is CSF leakage and risk of pneumocephalus and meningitis. Techniques of reconstruction have improved over the years minimizing the morbidity of this approach and options include multi-layered reconstruction with abdominal fat grafts, tensor fascia lata graft, pedicled nasoseptal mucoperichondrial, and pericranial flaps augmented by synthetic dural allografts and tissue sealants.

One or both nostril approaches are possible. After preparation of the nasal mucosa with vasoconstrictive agents, the endoscope is introduced. The middle turbinate is lateralized to provide working space. A nasal septal mucoperichondrial flap with its vascular pedicle preserved is elevated off the nasal septum and protected and the posterior septum is fractured to expose the sphenoid rostrum. The sphenoid sinus is entered *via* its natural opening (sphenoid ostia) and a combination of bone rongeurs or high-speed drill is used to perform a wide spheno-ethmoidectomy to enter the sphenoid sinus. Once entered and the mucosa stripped, bony prominences overlying the optic nerves and internal carotid arteries are identified, as is the sellar face. The bone over the sellar is drilled off exposing the dura. A dural incision is made exposing the tumor within. For most pituitary adenomas, a combination of ring curettes is used to debulk the tumor progressively and circumferentially. Valsalva maneuvers help deliver the last remaining supra-sellar components into the operative field. Rathke cleft cyst walls are biopsied and opened to drain the cyst fluid and often left open to avoid recurrence [51].

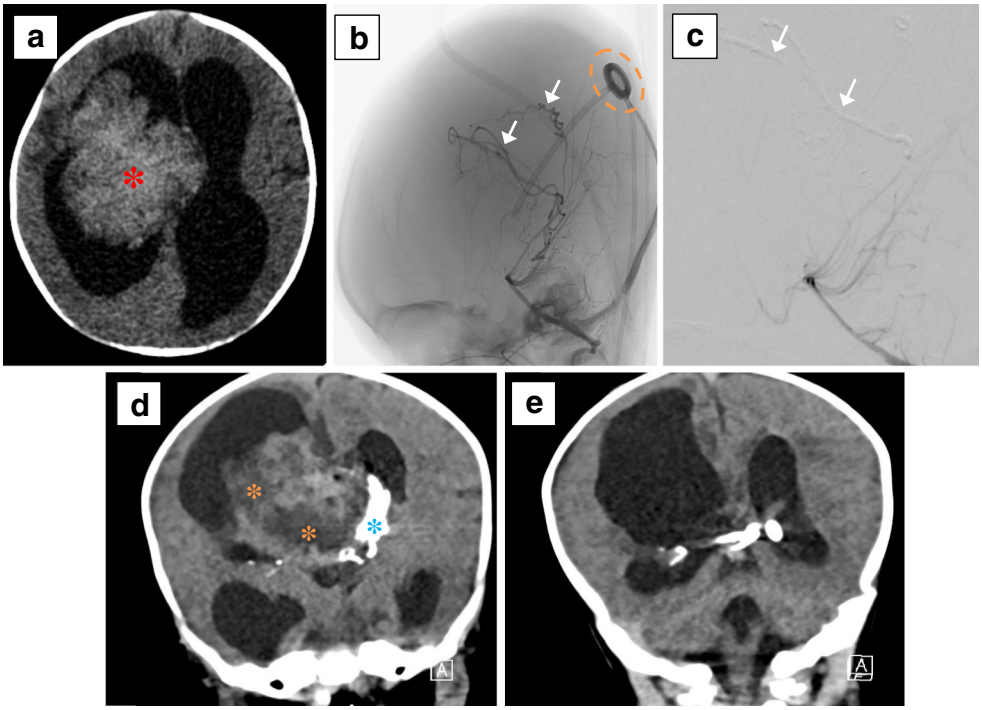

Fig. 6 **a** CT imaging demonstrating a large right sided ventricular tumor with acute hydrocephalus (red asterisk). **b** Cerebral angiography demonstrating an external ventricular drain (orange dashed line) inserted to control/relieve hydrocephalus and prominent vascular supply from the medial posterior choroidal arteries (white arrows). **c** Post-embolization cerebral angiography demonstrating embolic material (white arrows) within the medial posterior choroidal vessels. **d** Post-embolization CT scan demonstrating hypodense necrotic change within the tumor following an interruption of the tumor vascular supply (orange asterisks). The embolic material within the tumor feeding vessels results in hyperdensities seen on imaging (blue asterisk). **e** Post-operative CT following trans-cortical approach to lateral ventricle with complete tumor excision

Resection of the cyst wall may be difficult due to adherence to the infundibulum and therefore not often advised due to the risk of traction injury. After tumor resection and hemostasis, reconstruction of the skull base takes place.

For "simple" pituitary adenomas, the risk of dural breach of the diaphragma sellar and therefore entry into the subarachnoid space is low and reconstruction with simple packing is often sufficient. As mentioned above, following drainage of Rathke cleft cysts, the opening is left open to provide persistent drainage to avoid recurrence. Endoscopic debulking/resection of craniopharyngiomas will often require complex skull base reconstruction often with a pedicled naso-septal flap as the risk of dural breech and sub-arachnoid entry and therefore, risk of CSF leak is much higher. This is especially true in tumors where the resection is followed right to the floor of the 3rd ventricle where the risk of a high flow CSF leak is very high. Some surgeons often supplement the repair with a lumbar CSF drain peri-operatively to allow CSF diversion over the immediate postoperative period for a few days until the repair seals. The trans-sphenoidal approach can be extended in either the sagittal or coronal plane to incorporate more extended bone resections to access other more extensive pathologies. Examples include transtuberculum, transplanum, transcribriform, and transclival modifications. The risk of CSF leak and therefore the complexity of the skull base reconstruction required increase as the extent of bone resection increases.

6.5 Craniopharyngiomas

Craniopharyngiomas merit special consideration given they are the most common sellar region tumor in pediatrics and although a histologically benign tumor, it poses significant challenges on account of its highly eloquent location and locally recurrent nature. There is extensive debate regarding the optimal management strategy for craniopharyngiomas, including optimal approach whether trans-sphenoidal or open microsurgical [52] and evaluation of optimal treatment paradigms is difficult owing to selection biases and the degree of morbidity that is present with all forms of treatment [53, 54]. There are two main strategies: GTR or STR with adjuvant radiation therapy. Both have similar rates of disease control and overall survival. Therefore, strategy has evolved and shifted to outcomes that focus on optimizing patient quality of life metrics [55]. The two key contributors to post-treatment morbidity are hypothalamic dysfunction and visual impairment. A variety of clinico-radiological classifications have therefore been developed that assess degree of vertical and horizontal extension of the tumor, displacement of the optic

nerves and chiasm, and, most importantly, the relationship between the dorsal aspect of the tumor and hypothalamus including the extent of deformation and involvement [56–58]. The impact of these classifications is in their prediction of the degree of preoperative hypothalamic dysfunction as well as operative morbidity associated with attempted surgical resection. They therefore also help guide the feasibility of being able to perform GTR of the lesion *versus* opting for a more conservative approach that aims to achieve the primary aims of diagnostic confirmation, and decompression of visual apparatus protecting vision.

6.6 Surgical strategy

Surgical strategies for craniopharyngioma resection are outlined in Table 7. With open surgical approaches, general concepts are minimizing significant brain retraction with use of techniques for brain relaxation (early access to CSF cisterns or ventricles—such as lamina terminalis fenestration in the 3rd ventricle in pterional approaches or lateral ventricular entry for transcallosal approaches—for CSF drainage), early identification and protection of normal neurovascular anatomy (optic nerves and chiasm, internal carotid artery and its major terminal and bifurcating branches), early development of arachnoidal planes between tumor and neurovascular structures, intra-capsular debulking of solid components and aspiration of cyst contents, and finally attempt at cyst wall resection if a decision has been preoperatively made for GTR. It is imperative that the degree of traction of the cyst or tumor capsule in critical areas such as the hypothalamus and optic chiasm be minimized to avoid traction-related neurological injury.

6.7 Case illustration

A 10-year-old boy presented with a progressive impairment predominantly in color vision. MRI confirmed the presence of an extensive predominantly solid lesion with extension into both the infra-sellar and suprasellar space with calcification consistent with radiological diagnosis of craniopharyngioma. The optic chiasm was displaced superiorly. There was no evidence of hydrocephalus (Fig. 7a–c). Preoperatively, there was no suggestion of hypothalamic or pituitary dysfunction. He underwent surgery in staged fashion with an initial trans-sphenoidal approach to remove the infra-diaphragmatic/infra-sellar component followed by an open microsurgical approach with a bifrontal craniotomy and anterior interhemispheric approach to resecting the suprasellar component of the tumor. GTR was successfully achieved (Fig. 7d, e). He made a good recovery postoperatively.

7 Optic pathway and hypothalamic tumors

Optic pathway and hypothalamic tumors occur either in isolation or in association with neurofibromatosis type 1 (NF-1) (approximately 33% are associated with NF-1). The majority are low-grade gliomas with the most frequent being pilocytic astrocytomas although fibrillary astrocytomas, pilomyxoid astrocytomas, and gangliogliomas may also be found and form part of the differential [59]. Their clinical manifestations can be diverse owing to their variable location along the visual pathway from the optic nerves, chiasm, and tract to the hypothalamus (and therefore impact on the pituitary) and suprasellar extension into the 3rd ventricle and along the optic radiations [60, 61]. Perhaps more than any other type of pediatric tumour, owing to the heterogeneity of these tumours, their lack of a well-defined natural history, the significant morbidity associated with not only the primary disease process and its progression, but with all the various surgical and non-surgical adjuvant options, means that management decisions must be tailored to each individual patient taking into account their age, tumour anatomy, clinical presentation and comorbidities. Therapeutic paradigms should be implemented jointly in the context of a multidisciplinary team including neurosurgeons, neuro-oncologists, neuro-ophthalmologists, endocrinologists, and neuro-radiologists [62].

7.1 Management strategy

The primary treatment is chemotherapy. These tumors are rarely excised and surgery has significant risks as they infiltrate the hypothalamus and optic apparatus. The risks of surgical intervention should be balanced against alternative adjuvant therapies or serial surveillance. Therefore, a clear definition of the treatment goals and expectations is essential when planning treatment [63]. The aims of surgical intervention for these tumors include obtaining tissue for histologic diagnosis (especially if atypical imaging or absence of serum NF-1), the treatment of hydrocephalus, and tumor debulking.

7.2 Diagnosis

In tumors discovered incidentally or patients that are asymptomatic with no visual dysfunction and no evidence of mass effect or hydrocephalus, lesions can be monitored with serial surveillance. Evidence of visual deterioration and lesion growth usually becomes an indication for treatment. In the presence of a diagnosis or features of NF-1 and a typical radiological and clinical presentation, there is no indication for surgery for diagnostic purposes. Treatment is up front with chemotherapy usually. In patients without NF-1, diagnostic biopsy may be considered depending on whether radiological appearances are atypical, or one may proceed directly to adjuvant therapy. In non-NF-1 patients presenting with visual

and/or hypothalamic-pituitary dysfunction and typical radiological appearances, in the absence of hydrocephalus or mass effect, surgery is often not required and treatment proceeds directly to adjuvant therapy.

The presence of hydrocephalus or significant mass effect on surrounding neural structure and/or the visual apparatus due to a combination of solid and/or cystic tumor components usually forms an indication for surgery. Diagnostic biopsy can either be endoscopic (in combination with treatment for hydrocephalus), stereotactic, or open microsurgical; the approach to which depends on the tumor topography and location. Although most tumors are low-grade gliomas, the expansion of genotypic, molecular, and pathological description of these tumors together with the development of biological agents targeting specific molecular pathways means that tissue obtained for purposes of diagnosis and histopathological analysis is important and should be considered in all treatment paradigms [63, 64].

7.3 Hydrocephalus treatment

A common presentation of patients with large optic pathway-hypothalamic tumors is hydrocephalus due to extension into the 3rd ventricle with obstruction of the foramina of Monro or aqueduct of Sylvius. The initial procedure often required in these circumstances is CSF diversion; the type of which depends on the clinical urgency. Presentation in neurological crisis is rare, and in these situations, the safest and fastest option is an EVD. In semi-urgent circumstances, as is often the case given the indolent nature of these tumors, an endoscopic approach is useful for not only performing CSF diversion but also for obtaining tissue for tumor diagnosis.

Incisions should always be planned to take into consideration the need for future craniotomy for purposes of surgical debulking or the need for definitive CSF diversion with a shunt (usually VPS). Involvement of the floor of the 3rd ventricle and hypothalamus means that ETV is usually not an option for treatment of hydrocephalus. The endoscope is introduced *via* a frontal burr hole and a biopsy can be performed together with aspiration of tumor cyst contents if needed. This is followed by a septum pellucidotomy, thereby communicating both lateral ventricles as a single compartment (important especially in the presence of bilateral foramina of Monro obstruction) allowing the placement of a single lateral ventricular catheter that may be connected initially to a subcutaneous reservoir or formally converted to a permanent VP-shunt.

7.4 Tumor- and cyst-directed treatment

Surgical approaches to these tumors require careful planning and consideration. Radical excision is only rarely achieved owing to the infiltrative nature and relationship to surrounding structures. Occasionally, an optic pathway tumor restricted to one optic nerve alone that demonstrates progressive growth in a blind eye may be considered for GTR with optic nerve sacrifice and resection in order to preserve vision in the contralateral eye. The excision is performed proximal to the optic chiasm in order to conserve some of the medial retinal decussating fibers of the contralateral normal eye (von Willebrand's knee). In the majority of circumstances, surgical treatment is debulking for relief of mass effect or allowing treatment of hydrocephalus by free communication of CSF pathways to try and avoid the need for definitive shunting.

Approaches may be predominantly *via* the skull base (pterional with or without orbito-zygomatic osteotomies, bifrontal anterior interhemispheric) and *via* the 3rd ventricle (usually midline interhemispheric transcallosal) or endoscopic. Endoscopic approaches rarely allow significant debulking and are predominantly used for the purposes of diagnostic biopsy and fenestration to treat hydrocephalus. Skull base approaches are for tumors that do not have significant 3rd ventricular extension and are reserved for debulking of tumor or cyst to decompress the optic nerve and/or chiasm and brainstem if there is significant retrochiasmatic extension. The approach to the 3rd ventricle provides limited access to tumor within the skull base but is excellent for extensive intraventricular components that cause mass effect and hydrocephalus. The approach is *via* the interhemispheric fissure following a parasagittal craniotomy crossing the midline followed by a corpus callosotomy to enter the lateral ventricle. A septum pellucidum fenestration can be performed microscopically followed by access into the 3rd ventricle *via* a transforaminal, trans-choroidal, or interforniceal approach. Within the 3rd ventricle, the tumor is debulked with care to preserve the lateral walls and floor of the 3rd ventricle and hypothalamus. This can be achieved by leaving a thin carpet of tumor along these structures to avoid injury given these tumors often lack a plane in these regions as a result of their infiltrative nature and origin from these structures. It is also important to communicate as much of the CSF pathways as possible to decrease the need for permanent CSF diversion and therefore, debulking tumor posteriorly within the 3rd ventricle to open the aqueduct should also be considered if possible.

Treatment of cysts is often performed by endoscopic aspiration and cyst wall biopsy (for diagnostic confirmation), open microsurgical techniques or as a last resort, by way of catheters placed into the cyst and connection either to a subcutaneous reservoir (for frequent aspiration) or to a permanent shunt (cysto-peritoneal shunt). Placement of cyst catheters is performed stereotactically using image guidance, choosing the optimal trajectory and avoiding eloquent structures within the region.

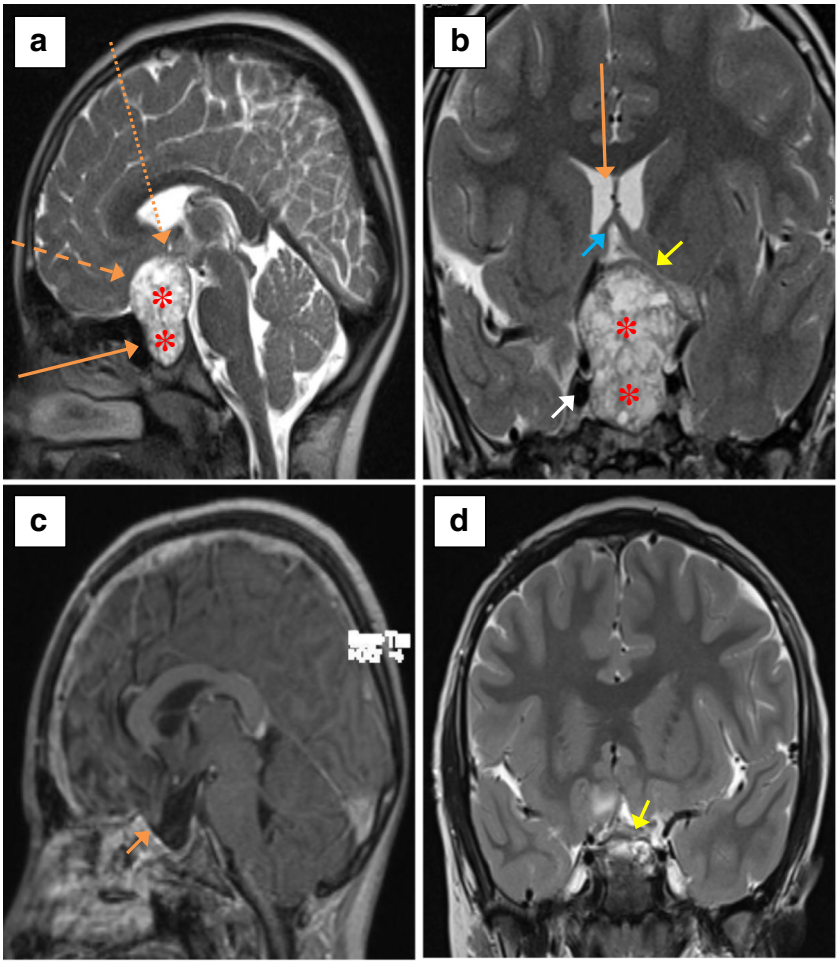

Fig. 7 **a** Sagittal T2 MRI pre-operatively demonstrating large infra-sellar tumor (lower red asterisk) with extensive suprasellar extension (upper red asterisk). The trajectory for a trans-sphenoidal approach (orange arrow) via the sphenoid sinus is demonstrated to access the infra-sellar portion of the tumor, as is the trajectory for an anterior interhemispheric approach (dashed orange arrow) to access the supra-sellar component of the tumor. The trajectory for a transcallosal approach is also demonstrated (dotted orange arrow). **b** Coronal T2 MRI pre-operatively demonstrating large infra-sellar tumor (lower red asterisk) with extensive suprasellar extension (upper red asterisk). The optic chiasm is displaced superiorly (yellow arrow) and the 3rd ventricle which is compressed (blue arrow). The right internal carotid artery is visualized (white arrow) within the cavernous sinus. The trajectory for an interhemispheric transcallosal approach is seen (orange arrow) which provides access into the lateral ventricle from which 3rd ventricle access can be obtained. Notably the tumor has not breached the floor of the 3rd ventricle, and therefore a transcallosal approach is not appropriate. **c** Sagittal T1 post-contrast MRI following complete resection of the tumor via a trans-sphenoidal approach for the infra-sellar component and a bi-frontal anterior interhemispheric approach for the suprasellar component. The pedicled nasoseptal flap used to repair the anterior wall of the sella and the sphenoid sinus is visualized (orange arrow). **d** Coronal T2 MRI demonstrating complete resection of the tumor. The optic chiasm (white arrow) is now fully decompressed

7.5 Postoperative and long-term care

Much like craniopharyngiomas, the long-term morbidity of these tumors is a result of their involvement and infiltration of the hypothalamus, pituitary, and visual pathways. The indolent nature and slow-growing nature of these frequently low-grade tumors mean that 5- and 10-year survival rates are high and therefore, it is imperative that surgical approaches are conducted with caution and care to avoid causing iatrogenic hypothalamic injury and optimize quality of life. The overall management of these patients is truly multidisciplinary with need for treatment for endocrinological and visual complications as a result of both the disease process and iatrogenically due to the treatments employed. Revision surgery is associated with increased neurological and endocrinological complications and should be reserved for clear failures of multiple adjuvant therapies. The management of hydrocephalus in these patients is also complex due to high protein content and viscosity of tumor cysts and ventricular CSF, which lead to frequent malfunction of catheters and associated valves requiring revision surgery, as well as the entrapment and compartmentalization of the ventricular system as a result of scarring due to repeated surgery and/or chemotherapeutic options. The compartmentalization of the ventricular system often requires multiple ventricular shunt systems. Wherever possible, permanent shunt placement should be considered only if

absolutely necessary and if it is indicated, tailoring approaches to minimizing the complexity of shunt systems by communicating ventricular compartments to allow single ventricular catheter and shunt systems where possible.

8 Supratentorial hemispheric tumors

Supratentorial tumors account for between 17 and 31% of pediatric CNS tumors [44, 65] and are commoner in children younger than 2-years compared to older children where infratentorial tumors predominate. Tumors can range from being completely benign with no intervention required and only surveillance to highly malignant tumors requiring aggressive adjuvant therapy. Surgical indications are predominantly for oncological and/or epilepsy-related reasons. The major predictor of survival in the majority of both indolent and malignant tumors is EOR [66]. The most common supratentorial hemispheric tumors are gliomas, which unlike adults are predominantly low-grade (Table 8) [65].

8.1 Management strategy

The goals of surgery are individualized to each patient depending on location and histology and whether the aim is predominantly oncologic, epilepsy-related, or both. Key oncological indications include pathological diagnosis together with molecular and genetic analysis for risk stratification, prognostication and possible targeting of adjuvant therapies, maximal safe cytoreduction of the tumor with the aim of GTR but at the very least, decompression to relieve mass effect on neural structures [77]. The treatment of intractable epilepsy requires patients to be appropriately investigated and discussed in an epilepsy multidisciplinary program and if epilepsy is localized to the lesion, surgical resection of the lesion and associated epileptogenic cortex, if clinically feasible [78].

8.2 Preoperative stabilization

The stabilization of the patient preoperatively consists of initiation of corticosteroids with gastro-protection while appropriate neural axis imaging is performed to plan definitive surgical intervention. Rarely, treatment of life-threatening hydrocephalus requires surgical intervention with CSF diversion, or presentation with significant neurological obtundation and mass effect acutely necessitates urgent craniotomy for decompression and tumor debulking or resection.

8.3 Operative principles

Treatment goals should be established with careful scrutiny of preoperative imaging and if time permits following discussion in neuro-oncology multidisciplinary setting. The decision whether to perform biopsy only, attempt GTR, or only debulking and STR can often be made preoperatively. Positioning and skin incision depend on the tumor location. Once the scalp flap is elevated and the craniotomy is performed, depending on the perceived swelling, techniques

Table 8 Common supratentorial hemispheric tumors in pediatrics and the impact of extent of resection (EOR)

Supratentorial tumors and the extent of surgical resection	
Low-grade gliomas (LGG)	• Maximal safe resection with the aim of GTR with preservation of functional neurological outcome • No role for adjuvant therapy if GTR is achieved [66] • Partial tumor resection/unresectable tumors often require chemotherapy and/or radiotherapy
High-grade gliomas (HGG)	• Surgical aim is GTR with preservation of functional neurological outcome • EOR is the strongest predictor of survival [67] • Adjuvant therapy is always indicated
Pleomorphic xanthoastrocytoma (PXA)	• Surgical aim is GTR with preservation of functional neurological outcome [68] • Role of adjuvant therapy is not well established, consider reserving for tumors with anaplastic features
Primitive neuro-ectodermal tumors (PNETs)	• GTR is controversial, but should be attempted where possible • Adjuvant therapy is almost always indicated, increases survival [69]
Atypical teratoid/rhabdoid tumor (AT/RT)	• Improved OS and PFS with maximal tumor resection [70, 71] • Adjuvant therapy with chemotherapy is almost always indicated, but sometimes delayed due to average patient age (3 years) if benefit does not outweigh risks [72]
Desmoplastic neuro-epithelial tumor (DNET)	• GTR is curative with good outcome
Ganglioglioma	• GTR if possible, does not require adjuvant therapy • PFS is affected by extent of resection [73] • Adjuvant radiotherapy post-STR may improve long-term control in LGG, HGG [74]
Ependymoma (EP)	• GTR is most important prognostic factor with ependymoma [75] • GTR with external beam radiation therapy results in the longest time to recurrence/progression • STR correlated with inferior outcome [76]

GTR, gross total resection; *OS*, overall survival; *PFS*, progression free survival

can help achieve brain relaxation such as infusion of osmotic diuretics, corticosteroids, and anesthetic techniques (control of end-tidal carbon dioxide and head elevation). Following dural opening, tumor debulking is performed using standard microsurgical techniques. Deep tumors are accessed *via* either a trans-cortical or trans-sulcal approach. Importantly, a variety of adjuncts are used to optimize extent of resection.

8.4 Radiological

Multimodal imaging including functional MRI and DTI techniques are used to carefully plan and evaluate the feasibility of achieving GTR. The relationship of the tumor to eloquent areas including motor, speech, and visual areas is defined anatomically and radiologically. Frameless stereotactic neuronavigation is used to guide the resection, as are intraoperative imaging adjuncts such as ultrasound scanning and intraoperative MRI. Preoperative embolization is used in highly vascular tumors to help limit intraoperative blood loss.

8.5 Intraoperative neurophysiological monitoring and mapping

IONMM is used when operating on tumors within or in close relation to eloquent cortex to maximize EOR safely. Transcranial MEPs and SSEPs are frequently monitored and phase reversal can be used to intraoperatively map the location of the central sulcus. Direct cortical stimulation is used to map eloquent functional cortex to plan the cortical incision in functionally non-eloquent areas. Sub-cortical stimulation and mapping is subsequently used to map and monitor the location of the deep corticospinal tract during the subcortical phase of resection. Awake craniotomy and functional mapping are only used in older children as cooperation is required and especially when operating on tumors near language areas. In the context of surgery for intractable epilepsy, pre- and peri-operative invasive mapping of the epileptogenic zone as well as intraoperative electrocorticography (ECoG) may be utilized.

8.6 Immunofluorescence

Use of 5-aminolevulinic acid (5-ALA) has been proven in adults for maximizing EOR and impacting in OS and PFS [79]. Its use in pediatric patients has not been fully established but should be considered for use in recurrent high-grade gliomas (HGG) to maximize resection [80].

9 Conclusions

Pediatric brain tumors cause significant morbidity for this vulnerable patient population. The heterogeneity of tumor types and the wide variety of locations in which they occur mean that all physicians involved in the care and management of these patients require an understanding of the principles of neurosurgical strategy and approaches to these tumors. We hope the above review has summarized the surgical approach and decision-making involved in management of pediatric brain tumors.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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