Developmental Considerations in Pediatric Skull Base Surgery

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Abstract

Objectives To review developmental surgical anatomy and technical nuances related to pediatric skull base surgery.

Design Retrospective, single-center case series with literature review.

Setting MD Anderson Cancer Center.

Participants Patients undergoing pediatric skull base surgery.

Main Outcome Measures Review developmental anatomy of the pediatric skull base as it relates to technical nuance of various surgical approaches and insight gained from a 25-year institutional experience with this unique patient population.

Results Thirty-nine patients meeting these criteria were identified over a 13-year period from 2003 to 2016 and compared to a previously reported earlier cohort from 1992 to 2002. The most common benign pathologies included nerve sheath tumors (11%), juvenile nasopharyngeal angiofibromas (9.5%), and craniopharyngiomas (4.8%). The most common malignancies were chondrosarcoma (11%), chordoma (11%), and rhabdomyosarcoma (11%). Varied surgical approaches were utilized and were similar between the two cohorts save for the increased use of endoscopic surgical techniques in the most recent cohort. The most common sites of tumor origin were the infratemporal fossa, sinonasal cavities, clivus, temporal bone, and parasellar region. Gross total resection and postoperative complication rates were similar between the two patient cohorts.

Conclusions Pediatric skull base tumors, while rare, often are treated surgically, necessitating an in depth understanding of the anatomy of the developing skull base.

Keywords

- pediatric skull base
- developmental anatomy
- surgery

Introduction

Tumors of the skull base are rare in the pediatric population, and, though many are chemosensitive, neurosurgical interventions continue to play an important role in treatment.¹ Pediatric skull base tumors comprise only 5.6% of all skull base tumors and, when compared to the adult population, occur in distinct locations and have unique histopathologic features.¹ Although surgical interventions are employed in the treatment of these rare lesions,² several important considerations must be kept in mind when approaching the pediatric skull base.

The purpose of this study was to review the normal development of the pediatric skull base, explore the implica-

tions pediatric anatomy plays in open and endoscopic approaches to skull base tumors, and understand how developmental anatomic variants may further impact surgical considerations. Additionally, we review the pediatric skull base experience of a major cancer center (MD Anderson) over a 25-year period, reporting on the types of tumors affecting the pediatric population.

Normal Pediatric Skull Base Anatomy

The embryonic, fetal, and infantile growth and development of the skull base play an important role in understanding pediatric skull base anatomy and its variants. The cranial

vault and skull base grow continuously in the first 10 years of life, with suture fusion, fontanel ossification, and bone development occurring at a variable rate in children over a wide age range.

Craniofacial Development

The human skull is derived from three components: the cartilaginous neurocranium (chondrocranium), the membranous neurocranium, and the viscerocranium. These components develop from neural crest and paraxial mesoderm cells that coalesce on either side of the rostral end of the notochord, surrounding the developing brain beginning in the fourth week of fetal development.^{2,3} While the membranous neurocranium contributes principally to the flat bones of the skull (frontal, parietal, occipital, squamous temporal) and the viscerocranium to the bones of the face, it is the chondrocranium that forms the majority of the skull base, visible by the eighth week of development. The membranous neurocranium forms via intramembranous ossification, a process characterized by spicule formation and the absence of cartilage, whereas the chondrocranium is so named because it forms via endochondral ossification, bone formation in the presence of cartilage.

Chondrocranium

During the fifth and sixth weeks of development, mesenchymal cells coalesce along the rostral termination of the notochord, developing various condensations of cartilage known collectively as chondrocranium.^{3,4} Condensations forming rostral to the terminal end of the notochord develop in the absence of molecular signaling from the notochord and are derived from cranial neural crest cells, while those condensations that form alongside the terminal notochord are derived from paraxial mesoderm. As a point of reference, the mature *sella turcica* corresponds to the rostral termination of the notochord. Thus, as the various cartilages of the chondrocranium ossify and fuse over time to form the skull base, those structures rostral to the *sella turcica* are of neural crest origin, while those structures caudal to this limit are derived from paraxial mesoderm.

The parachordal cartilage forms in a symmetric fashion on either side of the rostral termination of the notochord (mesoderm origin) and extends caudally, eventually fusing with occipital somites to form the basal plate. Endochondral ossification of the basal plate begins early in the seventh week of development eventually forming the posterior fossa, the occipital bone, hypoglossal canals, and foramen magnum.²

The prechordal cartilage forms from paired mesenchymal consolidations beyond the rostral termination of the notochord (neural crest origin). This prechordal cartilage, in turn, develops into hypophyseal and trabecular cartilages, which go on to form the sphenoid and ethmoidal bones, respectively.³

The hypophyseal cartilages form around the developing pituitary and are comprised of the presphenoid cartilage, which forms the anterior body of the sphenoid; the basisphenoid cartilage, which forms the sella turcica and posterior body of the sphenoid; the ala orbitalis, which forms the lesser wing of the sphenoid as well as the planum sphenoi-

dale; and the ala temporalis, which forms the greater wing of the sphenoid.

The trabecular cartilages (*trabeculae cranii*) and tissues surrounding the nasal placodes chondrify to form the ethmoid bone and nasal concha, as well as portions of the nasal septum. The paired trabecular cartilages fuse anteriorly to form the *trabeculae communis*, which with the prechordal cartilage is the foundation for the anterior cranial fossa.

Additionally, as the otic placode invaginates to form the otic pit, mesodermal chondrification fuses with the parachordal cartilages to form the petrous and mastoid portions of the temporal bone. As fusion occurs, the inner ear structures become embedded within the petrous ridge, and the jugular foramen develops between the otic capsule and basal plate.

Membranous Neurocranium

The membranous neurocranium is the precursor to most of the flat bones of the cranium, which eventually form the cranial vault and are initially unfused, yielding the patent fontanelles and sutures characteristic of newborns. Cranial vault growth is driven by the expansile growth of the neural elements which displace bones outward, increasing tension at bony sutures, thus promoting growth of new bone at the suture edge.

Viscerocranium

The face develops from the viscerocranium, exclusively from neural crest cells, via derivation from the pharyngeal arches.⁵ The membranous viscerocranium comes from the first arch, where it divides into a maxillary and mandibular process, allowing for intramembranous ossification to form the maxilla, zygoma, squamosal temporal bone, vomer, palatine bone, and a part of the mandible. Meckel's cartilage forms in the mesoderm of the mandibular process, and the chondral viscerocranium develops forming the ossicles of the middle ear and part of the mandible.⁶ Reichert's cartilage, from the second parapharyngeal arch, divides into two cartilaginous segments, one of which forms the stapes, the other which develops into the styloid process and hyoid.⁷ This development is uniquely important when understanding maxillary and mandibular growth, which begins with membranous ossification of Meckel's cartilage, and later undergoes endochondral ossification of the mandible resulting in ossification by 6 months into fetal development.⁶

The significance of facial development is not simply aesthetic, but important in function in regard to the mechanism of mastication, appropriate organ function, and function of bony articulations, but also muscle insertions. Zygomatic arch formation and function is a prime example of this. Additionally, formation of the orbit holds significance for not only visual reception and pathways but also for shape and development of the face and skull base structures. The rapid development of the eye impacts the position and shape of the orbit as the eye ball develops from ectoderm, neural crest cells, and mesoderm, and completes half of its postnatal

growth within the first 2 years of life, with adult size achieved by 7 years of age.⁸ The orbital bones begin formation via two routes, migration of the frontonasal process over the prosencephalon, approaching the orbit superiorly, giving rise to the lacrimal and ethmoid bones, while the maxillary process curves from below, giving rise to the lateral and inferior orbital walls, fusing by the sixth or seventh month of gestation.⁸

Skull Base Approaches

While these rudimentary structures form in utero, the skull base continues to develop and change for at least 10 years after birth, and the cranial vault is not fully fused for even longer.² Therefore, any surgical approaches to the pediatric cranial base should consider these variations and anatomical relationships.

Common skull base surgical approaches, with appropriate adjustments, can be safely and effectively implemented in pediatric patients.⁹ Respecting anatomic age-dependent relationships and employing a multidisciplinary approach are crucial to optimally expose and resect lesions while minimizing injury and manipulation of critical structures.

Many surgical considerations come into play when planning surgical procedures in the pediatric population. In pediatric cases, the smaller, thinner skull lends to varying decisions in pinning, drills, handling, and plating of the bone. Cranial fixation, for example, is widely used in patients under 5 years of age, though no standard guidelines are established in regard to pin size or pressure, and a complication rate as high as 54% has been reported.¹⁰ The use of rigid microplate fixation in our institutional series has previously been reported by the senior author.¹ In this report, no complications related to the use of nonabsorbable mini-plates such as plate translocation or plates becoming palpable through the skin were noted, though these are often reported complications of the technique.^{11,12} Nonetheless, the use of resorbable microplate fixation is a reasonable alternative that can yield excellent results, particularly in children younger than 5 years of age.^{13,14}

Of particular importance is diligent monitoring of blood loss during surgery. Pediatric patients have significantly smaller circulating blood volumes than in the adult population, thus seemingly small amounts of intraoperative blood loss can require intraoperative transfusion. In addition, the higher ratio of body surface area to volume in children can result in precipitous falls in body temperature, which among other pediatric anesthetic concerns is an important consideration that must be accounted for and controlled by the operative team.

Open Surgery

Pterional/Frontotemporal Craniotomy

The pterional craniotomy coupled with a transsylvian approach is the benchmark surgical approach to the parasellar region as well as the lateral and anterior brain stem. Its usefulness in both adults and children is widely reported. However, several important considerations remain when

employing this technique in children. The pterion, for example, is displaced anteriorly in children, making a posterior adjustment in placement of the “keyhole” burr hole important to ensure appropriate access to the anterior cranial fossa.⁹ Specific to the transsylvian approach, the pediatric brain is full and less relaxed, with less generous subarachnoid spaces in the fissure, which results in more difficult fissure dissection.⁹

Orbitozygomatic Craniotomy

The orbitozygomatic (OZ) craniotomy and its variations were first developed in the adult population to provide a greater degree of exposure to the anterior and middle skull base than seen in the traditional pterional approach, thus minimizing brain retraction.^{15–17} In children, the OZ craniotomy has similarly been proposed and employed to provide access to the parasellar region, orbital apex, and anterior and middle fossae, with reports of a one-piece bone flap reducing reconstruction at the orbital rim and improving viability, all the while maintaining temporalis anatomy without added morbidity.^{18,19}

When planning this approach, special consideration to anatomical variations with age is important. The supraorbital foramen or notch is absent until age eight and the zygoma is poorly developed in young children.⁹ In adults, the supraorbital notch is an important landmark in planning both the medial extent of frontal craniotomy and the orbital roof osteotomy in the OZ approach. Its absence in children necessitates use of other landmarks. Intraoperative navigation is particularly useful in this regard. The small size of the zygomatic arch may be a concern during attempted mini-plate reconstruction of the zygomatic osteotomy.

An additional consideration is the impact that orbital or zygomatic osteotomies may have in altering craniofacial growth patterns. In patients younger than 4 years of age, craniofacial growth is only at 80% of the mature state.²⁰ In our previously reported series, altered facial growth patterns were noted following zygomatic craniotomy in an 18 months old.¹ No disruptions in orbital growth patterns were associated with orbital osteotomies. While zygomatic arch complications were not observed in series by Teo et al. and Lang et al, none of their patients undergoing orbital or zygomatic osteotomies were under 4 years of age.^{21,22} These findings suggest that zygomatic osteotomies may result in noticeable deformity in patients under 4 years of age, more subtle alterations in patients aged 4 to 5 years, and are unlikely to result in craniofacial asymmetry in patients older than 5 years. Of note, no evidence of perturbed orbital growth has been observed in children undergoing orbital osteotomies.

Transfacial Procedures

A variety of surgical approaches have been developed to approach midline lesions of the anterior and middle skull base, often combining traditional craniotomies with osteotomies of the facial bones; although variations on these “trans-facial” approaches abound and the nomenclature of such approaches is not standardized, common examples include the transsphenothmoidal, transmaxillary transnasal, transfacial, transmaxillary, midfacial and facial translocation,

and LeFort osteotomy approaches.²³ In the adult population, these approaches are able to achieve adequate surgical access and exposure to allow for appropriate oncologic resection of midline skull base lesions. Appropriate surgical exposure allows for preservation of uninvolved anatomic structures, such as the blood supply to the overlying skin and the surrounding muscle, both of which are vital to allow for adequate and viable soft tissue reconstruction. While myriad anatomic considerations come to play in the execution of “trans-facial” approaches in the adult, specific considerations must be kept in mind when they are used in the pediatric population, specifically the effect of facial osteotomies on midface development and dentition (►Fig. 1). The majority of these approaches, thankfully, have been supplanted by modern endonasal endoscopic techniques.

Facial development, including permanent dentition formation, is of concern in “trans-facial” approaches. The LeFort I osteotomy, which extends from the nasal septum along the tooth apices to the pterygomaxillary junction, is one of the most commonly used osteotomies in the treatment of dento-facial deformities.³⁰ The LeFort I osteotomy, historically, was commonly employed in transfacial approaches to skull base tumors. Of key importance to this technique is a horizontal osteotomy that starts just lateral to the piriform aperture and extends to the maxillary tubercle just above the tooth roots, a trajectory that preserves the internal maxillary artery.³⁰ Lewark et al have described the loss of tooth buds in patients younger than 5 years of age undergoing LeFort I osteotomies, likely due to the horizontal transmaxillary osteotomy interfering with developing dentition.³¹ While permanent dentition occurs between age 10 and 12, younger aged children are not precluded from a LeFort I approach, though a modified, high osteotomy to spare unerupted teeth should be employed. In these instances, screw and plate placement are planned between tooth roots to avoid dental injury.³⁰

Transtemporal Procedures

Transtemporal approaches provide access to midline skull base lesions of the posterior fossa and clivus ventral to the brain stem. Transtemporal approaches to the skull base

include transmaxillary, transmandibular, infratemporal, and transtemporal,³² as well as translabyrinthine, middle fossa, transcochlear, transpetrosal, and retrolabyrinthine approaches.³³ These approaches allow access to difficult lesions in close relation to the brain stem, cranial nerves III to XII, and the major posterior circulation vessels, via direct lateral access through the mastoid, labyrinth, or cochlea (►Fig. 2), from above through a subtemporal middle fossa approach, from posterior in the retrosigmoid approach, or from varying directions in combined approaches.³³

Several important features of pediatric posterior fossa anatomy must be taken into consideration when discussing transtemporal approaches to the posterior fossa. The petroclival angle appears to decrease (become less obtuse) in children less than 9 years of age, a favorable anatomic feature that allows for improved access via presigmoid retrolabyrinthine approaches to pathology in the central clival depression—the space between the internal acoustic meatus and the jugular tubercle, due to a shallower clival depression.³⁴

Complications common to these procedures in the adult population—cerebrospinal fluid (CSF) leakage, hearing loss, transient facial neuropathy—are seen in the pediatric population as well, a finding borne out by our previous institutional experience¹ as well as the report of Jackson et al, who reported on a series of 15 patients undergoing transtemporal approaches with the assistance of a neuro-otologist.³⁵ The role of an experienced neuro-otologist is of particular importance in children because the mastoid air cells are less well developed, which make identification of the facial nerve and bony labyrinth more difficult³⁶ (►Fig. 3).

Far Lateral Approach

The far lateral, or far lateral transcondylar, approach extends the retrosigmoid approach, exposing the posterior fossa superiorly and laterally, by extending inferiorly to the foramen magnum with removal of the posterior third to half of the occipital condyle to allow adequate exposure to see the contralateral jugular tubercle without brainstem retraction.³⁷ Common considerations in this approach include incidence

Fig. 1 Sagittal and coronal computed tomography scans in a 3-year-old with clival chordoma. Note the lack of aeration of the sphenoid sinus and the multiple unerupted teeth. These anatomic findings may significantly impact the choice and difficulty of the operative approach.

Fig. 2 Axial magnetic resonance images of 16-year-old with left acoustic neuroma. This child had absence of functional hearing and a primarily intratemporal location of his acoustic neuroma. A translabyrinthine approach was utilized for complete tumor resection.

Fig. 3 (A) Sagittal computed tomography scans in 14-year-old demonstrating the preoperative presence of platybasia. Following endoscopic resection of the chordoma, the craniocervical alignment remained stable. Three months later cranial settling was noted and there was evidence of asymptomatic but significant instability necessitating occipitocranial fixation. (B) These magnetic resonance images are from the same patient. Imaging reveals a gross total tumor resection. Note the increased kinking of the brainstem associated with the cranial settling.

of CSF leak, vertebral artery injury, hypoglossal nerve injury, and in children, significant blood loss secondary to posterior condylar vein and venous plexus bleeding; this is in addition to, based on the extent of bony resection, concern for occipitocervical instability.³⁷

The occipital condyle develops from two parts of the occipital bone, the basioccipital and the exoccipital portions, divided by the synchondrosis intraoccipitalis anterior, which ossifies by the age of 4 to 6 years old, progressing from medial to lateral.³⁸ In comparison to adults, the occipital condyle's subchondral compact substance is furrowed and fissured in children, mimicking a subdivision of the articular surface without correlation to the cartilage-free strip, as the subdivision furrows do in adults.³⁸ Understanding the development of the occipital condyles, as well as their changes over time, can help shed light on the impact surgical removal of them will have. For example, the expanded removal of the occipital condyle or C1 lateral mass in extended far lateral approaches may introduce instability at the craniocervical junction. Biomechanical studies in adult cadavers have suggested that removal of 50% or more of the condyle can result in significant hypermobility at the Oc-C1 junction.³⁹

Endoscopic Surgery

Kassam et al established the feasibility and safety of endoscopic surgery in pediatric patients in a review of 430 patients, 5.8% of which were pediatric, where a variety of endoscopic approaches were performed to access medial skull base lesions.⁴⁰ There are several considerations in pediatric endoscopic skull base surgery. The nares and piriform aperture are often large enough to accommodate common endoscopes and instrumentation, though in patients younger than 5 years old, the piriform aperture may limit passage, requiring modification through a sublabial incision.⁴¹ Furthermore, pneumatization of the sphenoid sinus, a stepwise process beginning anterior and progressing posterior, molds the skull base and impacts the relationship between bony structures and related neurovascular elements, making their surgical exposure much more difficult and thus greatly impacting surgical planning⁴² (►Fig. 1) While ethmoid sinuses are fully developed by birth, the maxillary sinus is fully pneumatized at 2 to 4 years of age, and the sphenoid sinus continues to pneumatize into adulthood.⁸ While normal anatomy varies, abnormal anatomy from infection or congenital lesions may also distort anatomical landmarks, making the use of intraoperative navigation particularly useful in endoscopic endonasal cases.⁴³

Fig. 4 (A) Preoperative and (B) postoperative axial computed tomography and coronal post-contrast magnetic resonance imaging of 8-year-old with primary right temporal bone osteoblastoma. A total resection was accomplished. An experienced neuro-otologist is critical to safely navigate the pediatric temporal bone (All images are courtesy of the University of Texas M.D. Anderson Cancer Center Department of Neurosurgery and are used with permission.)

Expanded Endonasal Approaches

Expanded endoscopic endonasal approaches allow less invasive access to the anterior cranial fossa and clivus through a variety of expanded approaches including transplanar, transsphenoidal, transclival, transpterygoid, and transcribiform approaches.⁴⁴ These approaches allow access to the pituitary, as well as mobilization of it to access lesions posterior to the gland and inferior in the clivus; additionally, they allow for anterior exposure to the crista galli and lateral exposure to the infratemporal fossa and temporal bone, all while minimizing brain retraction to improve outcomes.⁴⁴

Endoscopic endonasal approaches to the skull base have significantly improved morbidity when compared with open procedures.⁴⁵ While this holds true for adult and pediatric populations, several considerations are important in the pediatric population. In addition to the aforementioned considerations, special consideration during reconstruction with a nasoseptal flap with respect to age is encouraged. Shah et al demonstrated sufficient coverage with nasoseptal flaps in pediatric patients age 10 to 17 years of age, though for those age younger than 10, it is insufficient to cover the skull base defect.⁴⁶ They note that this is especially evident with anterior approaches reaching to the cribriform plate, where the defect is larger than flap length.

Craniocervical Anomalies

Craniocervical anomalies carry several implications for access to the skull base. The relationship and articulations between the occiput, atlas, and axis are maintained by critical ligaments, which are susceptible to injury by trauma, inflammation, cancer, or congenital malformations. Craniocervical junction malformations occur when there is hypoplasia of the occipital bone, resulting in posterior fossa and upper cervical anomalies, including Chiari malformations, platybasia, and basilar invagination, among others.⁴⁷ Such abnormalities at the craniocervical junction result in anomalous craniocervical relationships and angles, impacting cervical lordosis, occipitocervical stability, and skull base development throughout childhood.⁴⁷ Their presence may also complicate postoperative care (► Fig. 4).

The relationship between these angles is critical in planning for craniocervical instrumentation. For example, platybasia entails a more obtuse angle between the anterior cranial fossa and clivus. In children who are 10 years and older and may be candidates for anterior clival fixation techniques, extreme platybasia may make such fixation technically challenging or unfeasible altogether.⁴⁸ Generally, in pediatric patients, craniocervical fixation is made more complicated by the diminutive pediatric occiput and cervical spine, the dimensions of which may impact bony purchase and construct strength.⁴⁹ In the case of patient's with congenital anatomic variants and associated syndromes, nonstereotyped bony anomalies such as atlanto-occipital assimilation, os odontoideum, absence of the occipital condyles, or absence of the posterior C1 arch, as well as abnormal vertebral artery location can pose potential challenges, requiring unique screw trajectories and limiting substrate for arthrodesis, all with an increased complication pro-

file.^{49,50} Obtaining rigorous preoperative imaging including vascular studies is essential to planning such procedures; the advent of intraoperative spinal neuronavigation holds promise for further minimizing complications in this challenging patient population. Moreover, craniocervical fixation procedures can have implications for longitudinal growth and occipitocervical alignment as pediatric patients mature, with generally good clinical and patient-reported outcomes but variable growth and alignment reported.^{51,52}

Institutional Review

A database maintained by the Department of Neurosurgery at the University of Texas M.D. Anderson Cancer Center was

Table 1 Skull base tumors by pathology: surgical pathology in current series and series by Hanbali et al

Pathology	LoPresti et al (current study) (n = 39)	Hanbali et al ¹ (n = 24)
	N (%)	N (%)
Benign		
Nerve sheath tumors	4 (10.3)	3 (12.5)
Craniopharyngioma	3 (7.7)	
Juvenile angiofibroma	2 (5.1)	4 (16.7)
Meningioma	2 (5.1)	
Eosinophilic granuloma	1 (2.6)	1 (4.2)
Desmoid tumor		2 (8.3)
Glomus vagale		1 (4.2)
Hemangioma		1 (4.2)
Neurocristic hamartoma		1 (4.2)
Teratoma		1 (4.2)
Osteoblastoma	1 (2.6)	
Rathke's cyst	1 (2.6)	
Malignant		
Chondrosarcoma	6 (15.4)	1 (4.2)
Chordoma	6 (15.4)	1 (4.2)
Rhabdomyosarcoma	4 (10.3)	3 (12.5)
Osteosarcoma	2 (5.1)	
Neuroendocrine carcinoma	2 (5.1)	
Ewing's sarcoma	1 (2.6)	1 (4.2)
Glioblastoma	1 (2.6)	
Metastasis (meningioma, squamous cell)	1 (2.6)	1 (4.2)
Lymphoma	1 (2.6)	
Myofibroblastic sarcoma	1 (2.6)	1 (4.2)
Liposarcoma		1 (4.2)
Olfactory neuroblastoma		1 (4.2)

searched to identify all patients with tumors of the skull base, age 18 or less at time of surgery. From 1992 to 2002, 24 patients meeting these criteria were identified and the details of their management have been previously published.¹ Over the subsequent 13-year period from 2003 to 2016, 39 further children were identified. Patient data was deidentified and pathology, tumor location, and extent of resection were examined to better define tumors of the skull base in this pediatric population.

In the earlier cohort of patients, the most common tumors were of mesenchymal origin. Of the malignant tumors 10 of 13 were sarcomas. The most common benign pathologies were juvenile nasopharyngeal angiofibroma and nerve sheath tumors, although craniopharyngioma and pituitary adenoma were excluded from that review (►Table 1). The infratemporal fossa was the anatomic region most commonly affected, followed by the orbit and the sinonasal cavity. Thirty procedures were performed in these 24 patients utilizing a variety of surgical approaches (►Table 2). Endoscopic surgery was not utilized at that time. A gross total resection was accomplished in 77% of patients. Five patients (20%) had postoperative complications. These included sensorineural hearing loss in one patient with a facial neuroma, jaw deviation, and inadequate zygomatic growth in an 18-month-old following zygomatic osteotomies, hypoglossal neuropathy, new ophthalmic and maxillary neuropathies in a patient with a cavernous sinus hemangioma and palatal dehiscence following transmandibular circumglossal retropharyngeal and transpalatal resection of a clival chordoma. In the more recent patient cohort, the most

common pathologies included chondrosarcoma, chordoma, rhabdomyosarcoma, and craniopharyngioma. Of the malignant tumors 20 of 25 were sarcomas. Nerve sheath tumors and craniopharyngioma were the most common benign tumors (►Table 1). The majority of the tumors were localized in the sellar/parasellar region (7 patients, 18%), clivus (6 patients, 15%), temporal bone (6 patients, 15%), sinonasal cavities (5 patients, 13%), and infratemporal fossa (5 patients, 13%). Other sites were the cavernous sinus (4), orbit (4), and petroclival synchondrosis (2). Fifty surgical procedures were undertaken in these 39 patients. These included planned staged surgeries and surgery for recurrence. The surgical approaches utilized were similar over the two time periods save for an increase in the use of endoscopic approaches and the subtemporal/infratemporal approach in the more recent patient cohort (►Table 2). The gross total resection rate in the more recent cohort of patients was 67%. Eight patients (20.5%) had complications that required further treatment. One patient had a significant stroke at the time of preoperative tumor embolization for a giant juvenile nasopharyngeal angiofibroma. Two patients had postoperative CSF leakage, one following a trans-temporal resection of an osteosarcoma and the second following extended endoscopic sinus surgery (ESS) for a clival chordoma. Two patients had delayed pseudomeningoceles: one of the posterior fossa and one extending to the nasopharynx from the medial temporal fossa following resection of an infratemporal fossa mesenchymal chondrosarcoma. One patient developed a corneal ulceration requiring corneal transplant following two-stage resection of a trigeminal neuroma.

Table 2 Skull base resections by approach: surgical approaches employed in current series and series by Hanbali et al

	LoPresti et al (n = 50)	Hanbali et al (n = 30)
Approach	N (%)	N (%)
Cranio-orbital zygomatic and transfacial	4 (8)	8 (26.7)
Cranio-orbital zygomatic	5 (10)	6 (20)
Transfacial/transorbital		2 (6.7)
Bifrontal craniotomy	4 (8) ^a	3 (10) ^c
Endonasal endoscopic approach	5 (10)	
Far lateral/transcondylar approach	2 (4)	
Cranio-orbital	9 (18)	
Midfacial degloving approach	1 (2)	
Total petrosectomy	2 (4)	
Posterior transtemporal approach	5 (10)	5 (16.7)
Preauricular subtemporal and infratemporal fossa approach	5 (10) ^b	2 (6.7)
Sublabial transseptal transsphenoidal approach	3 (6)	1 (3.3)
Suboccipital craniotomy	1 (2)	1 (3.3)
Transmandibular circumglossal retropharyngeal	1 (2)	1 (3.3)
Zygomatic and infratemporal fossa approach	1 (2)	1 (3.3)
Occipitocervical fusion	2 (4)	

^aWith endoscopic sinus surgery in two patients.
^bWith cranio-orbital zygomatic (COZ) in one patient.
^cWith lateral rhinotomy in one patient.
Note: In Hanbali et al. 30 procedures were performed in 24 patients.

Prolonged dysphagia occurred in a 3-year old following ESS and occipitocervical fusion for clival chordoma. Finally, one patient with baseline platybasia developed delayed cranial settling requiring occipitocervical fusion (►Fig. 3).

Conclusions

Pediatric skull base tumors, while rare, are often treated surgically, necessitating the need for further understanding of developmental anatomy when planning surgical approaches to the pediatric skull base. With slight adjustments accommodating for age-related skull base anatomic relationships, many common surgical approaches provide safe and efficacious access to the anterior, middle, and posterior cranial fossae allowing for adequate treatment.

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