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Endoscopic Skull Base Surgery in the Pediatric Patient

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Abstract

Pathology along the anterior and posterior skull base has long posed challenges to surgeons due to the difficulty accessing these locations and complexity of the surrounding neurovascular anatomy. Initial surgical management of these disorders included open craniofacial approaches and/or craniotomy and these approaches are still utilized today for selected cases. However, advances in the later half of the 20th century in optics, endoscope design, and high definition digital images allowed the development of microscopic and, more recently, endoscopic approaches to the skull base. This technology was initially developed and used in adults, but has been adapted over time for use in the pediatric patient. In this chapter, the salient points regarding pediatric endoscopic skull base surgery will be reviewed to provide the reader a framework for understanding the indications, pertinent anatomy, preoperative evaluation, and intraoperative management of skull base pathology, highlighting challenges and circumstances unique to the pediatric population.

Keywords: skull base, pediatric surgery

1. Introduction

Pathology along the anterior and posterior skull base has long posed challenges to surgeons due to the difficulty of accessing these locations and the complexity of the surrounding neurovascular anatomy. Initial surgical management of these disorders included open craniofacial approaches and/or craniotomy, and these approaches are still utilized today for

select cases. However, advances in the later half of the 20th century in optics, endoscope design, and high-definition digital images allowed the development of microscopic and, more recently, endoscopic approaches to the skull base. This technology was initially developed and used in adults, but has been adapted over time for use in the pediatric patient. In this chapter, the salient points regarding pediatric endoscopic skull base surgery will be reviewed to provide the reader a framework for understanding the indications, pertinent anatomy, preoperative evaluation, and intraoperative management of skull base pathology, highlighting the challenges and circumstances unique to the pediatric population.

2. History of endoscopic skull base surgery

Transnasal access to the skull base was introduced over a century ago by Schloffer, and Hirsch and Cushing respectively popularized the sublabial and transnasal approaches [1–3]. After Cushing's initial experience with transsphenoidal pituitary surgery, he turned to the subfrontal intracranial approaches [2]. The intracranial approaches remained popular until the introduction of the operating microscope in the 1960s allowed for the refinement of the transseptal transsphenoidal approach [4]. While modest efforts were made in addressing the anterior skull base and clival pathology endonasally with the microscope, it was not until the advent of the endoscope that the field expanded its scope to address pathology outside the sella, extending from the crista galli to the foramen magnum [4, 5]. In the 1990s endoscopic endonasal approaches to address pituitary lesions and cerebrospinal fluid leaks were proposed, which led to the gradual expansion of the field [6–11]. In 2005, the Center for Cranial Base Surgery at the University of Pittsburgh published several landmark papers highlighting the expanded endonasal endoscopic approaches to the skull base [4, 5, 12] and the use of these techniques has expanded steadily since the early 2000s. One key point emphasized by the Pittsburgh collaborative was the idea of "team surgery" in which the neurosurgeon and otolaryngologist—head and neck surgeon work simultaneously throughout all phases of the procedure [4]. With this binostril, four-handed approach, the surgeons can both visualize and manipulate the full extent of the midline skull base. In the last decade, the extent of the expanded endonasal approaches have continued to grow, pushing the limits of endonasal access anteriorly, laterally, superiorly, and posteriorly.

3. Applications to pediatric patients

The use of expanded endonasal endoscopic approaches in pediatric populations also developed as the field expanded. The application of open skull base approaches in the pediatric patient was described following its use in adults [13, 14]. The earliest case reports focusing on the use of endoscopic skull base surgery in children were published in the late 1990s [10, 11, 15] and steadily increased in the following decades. Though the techniques of endoscopic skull base surgery were carried on to the pediatric population, there has been limited publication on the topic compared to adult literature. There are several case reports outlining the use of

endoscopic techniques in the management of congenital meningoencephaloceles [16-19], benign tumors [20, 21], and bony lesions [22]. As juvenile nasopharyngeal angiofibroma (JNA) is exclusive to the adolescent age group, endoscopic management of this lesion is more widely described [15, 23-28]. There have also been several case series highlighting the application of endoscopic skull base surgery in the pediatric patient [29-31]. In Kassam et al.'s 2007 review, access to the entire skull base via a fully endoscopic approach demonstrated multiple advantages of over the previously utilized open approaches for these lesions. Key advantages discussed included minimal disruption of growth centers, lack of brain retraction with associated morbidity, and improved pituitary stalk visualization and potential preservation of the pituitary function [30].

4. Key differences between pediatric and adult skull base surgery

Though the general principles of endoscopic skull base surgery remain the same for both adults and pediatric patients, some critically important differences must be carefully considered in the pediatric patient preoperatively. These differences center around the development and aeration of the paranasal sinuses, size constraints encountered in the pediatric nose, and concern regarding skull base growth centers. A more complete review of developmental anatomy as it relates to endoscopic skull base surgery is described later in this chapter. Regarding size constraints, it is generally accepted that the nasal aperture is of sufficient size at the age of 6–7 years to accommodate a binostril endoscopic skull base approach [32]. Prior to this age, the interpyriform distance can preclude the introduction of endoscopes and instruments together and must be addressed on a case-by-case basis. Even with adequate nasal access, the intercavernous carotid distance may preclude adequate access in pediatric patients [32, 33] and must be evaluated preoperatively, especially if sellar or suprasellar access is the goal of the procedure. Lastly, the pediatric skull base is a dynamic structure and continues to expand and remodel until it reaches adult configuration following puberty. Careful attention must be paid to minimize disruptions to skull base growth centers in the prepubertal patient to abate growth disruptions [33, 34].

5. Developmental anatomy review

5.1. Sinonasal

First noted in the fourth week of development, the nasal capsule's lateral walls enlarge and fold by the ninth week, forming six ethmoturbinals, all derived from ectoderm from the frontonasal process. The first ethmoturbinal becomes the uncinate process while the second develops into the ethmoid bulla. The developmental end point of the third ethmoturbinal is the ground lamella of the middle turbinate and the fourth ethmoturbinal becomes the superior turbinate. The fifth and sixth ethmoturbinals may regress or fuse to become a supreme turbinate.

The four paired paranasal sinuses each have a distinct developmental trajectory. The maxillary sinuses project from the ethmoid infundibulum between the first and second ethmoturbinals and are first noted at week 9 of development. Present at birth, the maxillary sinus expands most significantly between ages 0–3 and 6–12, reaching adult size at approximately age 18. The ethmoid sinuses are also present at birth and continue to mature in an anterior to posterior fashion in the first 12 years of life. The frontal sinus is last to begin development in the fourth month of gestation as a superior evagination of the frontal recess. However, the frontal sinus is not distinguishable radiographically until approximately the third year of life and does not reach the frontal bone until age 5. The frontal sinus reaches adult size toward the end of puberty and can continue to aerate the frontal bone throughout adult life [35–37]. The sphenoid sinus is of primary importance in the endoscopic skull base surgery. It is first noted in the fourth week of gestation as an evagination from the sphenoethmoid recess and slowly aerates over the first 10–15 years of life. Aeration begins in the inferior and anterior aspect of the sinus and extends posteriorly and superiorly such that the dorsum sellae is last to aerate. At age 6–7, the sphenoid face is typically fully pneumatized and the planum is aerated in the majority of the patients. By age 10, the clival recess is typically pneumatized but dorsum sellae is infrequently pneumatized, 16% of the time [32, 33].

5.2. Sella and Skull Base

The sella itself divides the anterior and posterior skull base. The skull base develops primarily from cartilaginous precursors, which ossify over the first two decades of life. The anterior skull base derives from a neural crest cell origin while the clivus and other posterior skull base structures have paraxial mesodermal origins [32, 35]. The remaining details of skull base development and ossification are beyond the scope of this chapter and are described elsewhere [35].

6. Surgical indications

The indications for surgical intervention at the skull base are fairly limited in terms of broad categories and include cerebrospinal fluid (CSF) leak, neoplasms, and congenital abnormalities. While the surgical goal with CSF leak is clear, when addressing neoplasms, the role of surgery can range from biopsy for diagnosis to surgical debulking to complete resection (clear margins) and is largely dependent upon the pathology being addressed and the location. In the setting of malignancies such as rhabdomyosarcoma or other sarcomas, the role of surgery may vary, and it becomes important to pre-operatively define the goals of the surgical treatment in these cases.

6.1. Cerebrospinal fluid leak

Whether spontaneous, posttraumatic, or iatrogenic, the presumptive diagnosis of CSF leak is made when persistent clear rhinorrhea is identified (Figure 1). The confirmation of CSF is made with a laboratory analysis for β-2 transferrin. Imaging with CT can identify areas of bony

discontinuity. In cases where the location is difficult to assess, MR or CT myelography can be performed to help localize the area of CSF egress. The initial approach to CSF leaks often entails conservative management with lumbar drain placement and medical management to decrease intracranial pressures with surgery reserved for refractory leaks or high-flow CSF leaks not likely to resolve with conservative management alone.
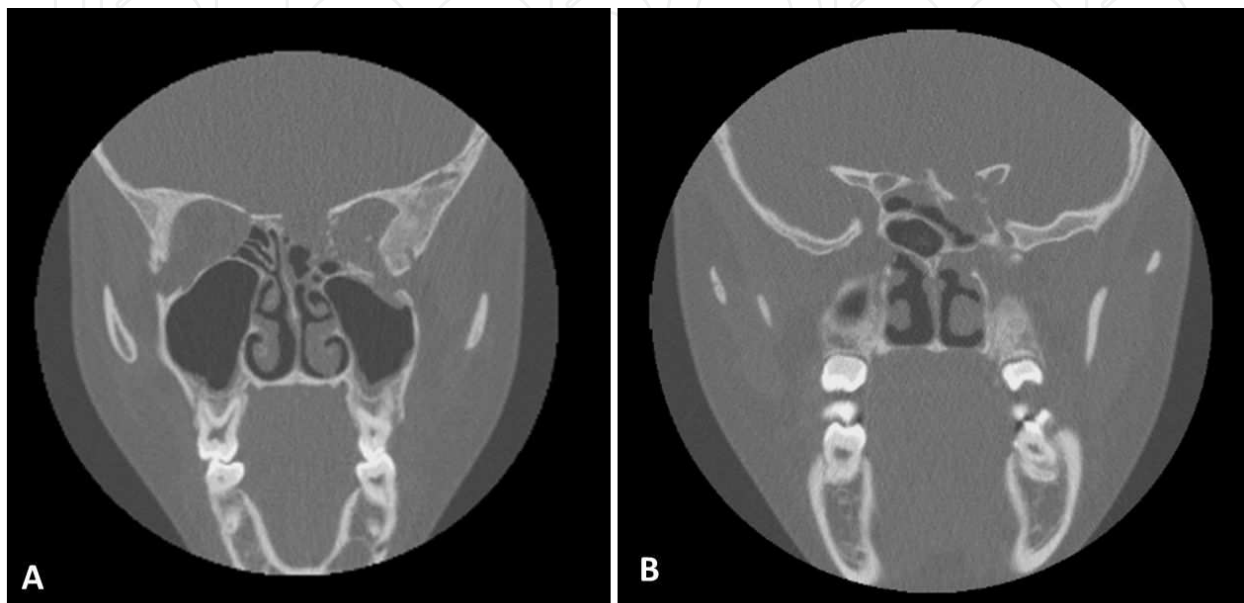

Figure 1. CSF leak from multiple skull base fracture: (A) cribriform plate and (B) left sphenoid. This was successfully repaired using transnasal endoscopic techniques.

6.2. Neoplasms

Neoplasms can vary in both pathology and presentation. A thorough discussion of each tumor is beyond the scope of this chapter, and tumors have been widely described in the literature. A brief overview of the various pathologies and their most common locations are shown in Table 1. Figure 2 demonstrates a rhabdomyosarcoma.

Lesion	Category	Location
Fibrous dysplasia	Benign neoplasm	Anterior skull base, perisellar, clivus
Fibro-osseous tumors	Benign neoplasm	Anterior skull base
Pituitary adenoma	Benign neoplasm	Sella
Craniopharyngioma	Benign neoplasm	Sella
Chordoma	Benign neoplasm	Clivus
Schwannoma	Benign neoplasm	Clivus, infratemporal fossa

Lesion	Category	Location
Juvenile nasopharyngeal angiofibroma (JNA)	Benign neoplasm	Pterygopalatine fossa, infratemporal fossa
Glomus jugulare	Benign neoplasm	Clivus
Chondrosarcoma	Malignant neoplasm	Clivus
Esthesioneuroblastoma	Malignant neoplasm	Anterior skull base
Meningoencephalocele	Congenital abnormality	Anterior skull base, perisellar, clivus
Glioma	Congenital abnormality	Anterior skull base, perisellar, clivus
Nasal dermoid	Congenital abnormality	Anterior skull base
Cerebrospinal fluid leak	Iatrogenic, traumatic, idiopathic	Anterior skull base, perisellar, clivus

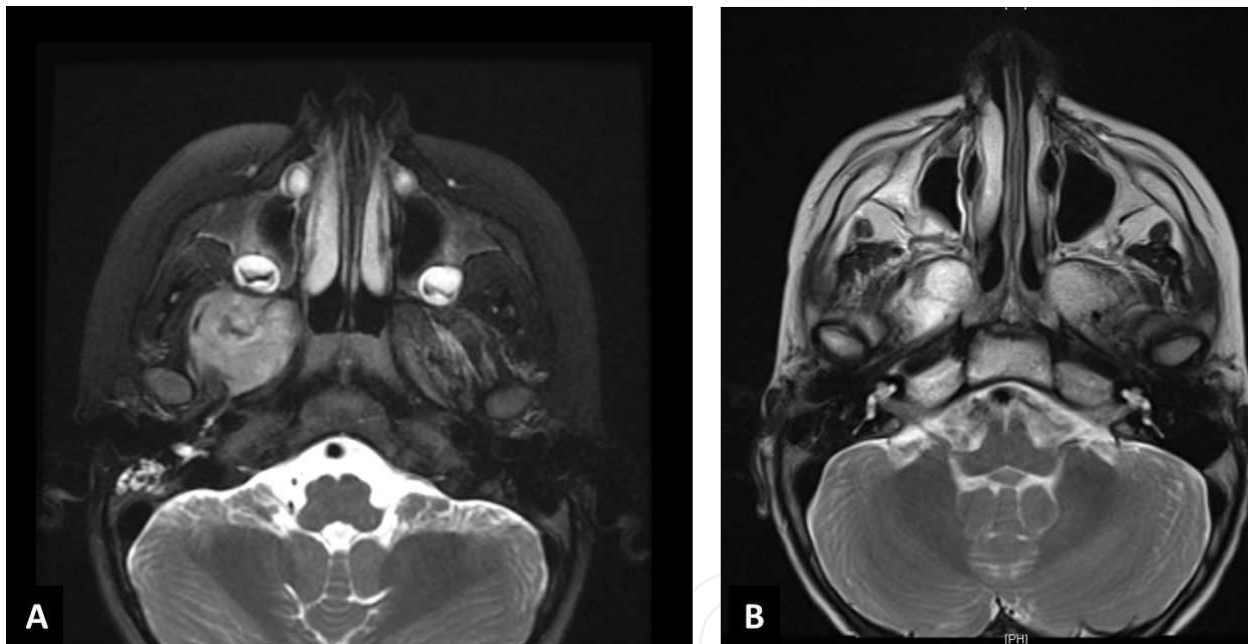**Table 1.** Differential Diagnosis of Pediatric Skull Base Abnormalities

Figure 2. A 3-year-old male with right pterygopalatine fossa rhabdomyosarcoma. (A) Pretreatment and (B)Post-chemoradiation therapy. Given some residual enhancing tissue, multiple endoscopic biopsies were performed after treatment to ensure no residual disease and were consistent with radiation changes.

7. Developmental abnormalities

Disruptions in the normal development of skull base structures may result in meningoencephaloceles (Figure 3), gliomas, and nasal dermoids. These lesions can be characterized by their location of origin and contents [18, 38–40], but all share a common origin in aberrant ectodermal differentiation and/or closure of the neural folds [18].
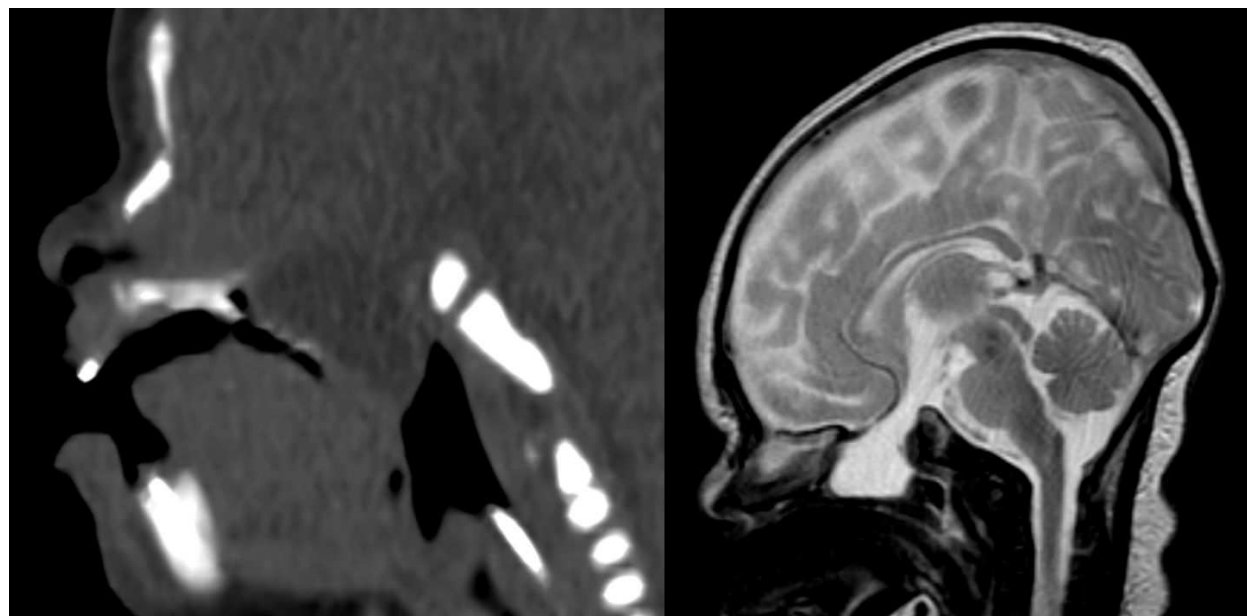
Figure 3. Newborn child with a large sphenoethmoidal meningoencephalocele. With a skull base defect this large in a newborn, open repair is the only option. The left image is a sagittal noncontrasted CT scan performed for initial diagnosis. On the right is a T2-weighted sagittal MRI to further characterize this lesion, identifying no neural components within the sac.

8. Clinical presentation

Clinical presentation varies for each lesion and is dependent upon the lesion's origin and compression of the surrounding structures. Pituitary lesions that are hormonally active may also present with endocrine disturbances [30]. Compression of the optic nerve or tracts may present with unilateral or bilateral ocular symptoms depending upon the location of the compression [30]. Sellar lesions extending into the cavernous sinus may compress cranial nerves 3 or 4 and the ophthalmic branch of 5 or 6. The sixth nerve is the most sensitive to compression and lateral rectus palsy is often the first presenting symptom of cavernous sinus involvement. Clival lesions may also compress cranial nerves 9, 10, 11, and rarely 12. Meningoceles and meningoencephaloceles may present with a nasal obstruction, which can become more significant with agitation due to increased intracranial pressure expanding the extracranial portion of the sac [18]. Osseous lesions such as fibrous dysplasia may present with narrowing of neural foramina or with externally visible skeletal deformities depending upon the bone(s) involved [30]. Vascular lesions, such as juvenile nasopharyngeal angiofibroma, present with unilateral nasal obstruction and epistaxis, a diagnosis exclusive to adolescent males. Other presenting symptoms may include headache, proptosis, nausea, hydrocephalus, or nondescript symptoms such as drooling or torticollis [29, 30].

9. Preoperative assessment

9.1. History and physical examination

A thorough history and physical examination is essential for any patient. Careful attention to progression and timing of symptoms in the history and to the cranial nerve examination on physical examination can help to direct the differential diagnosis. Nasal endoscopy can be helpful in the presence of nasal obstructive symptoms or olfactory disturbances to assess intranasal involvement and characterize the extent and potential origin of any lesion in question. A biopsy of any pediatric nasal mass should *not* be performed prior to imaging, and if tissue diagnosis is needed, the procedure should be done in the operating room. Figures 4 and 5 demonstrate examination findings as to why imaging is necessary prior to interventions. In the setting of CSF leak, a thorough history of any traumatic injuries or previous sinonasal or intracranial procedures is valuable in helping to identify the etiology and potential location of the CSF leak.

Figure 4. Right nasal endoscopy in an infant with nasal obstruction symptoms, which was found to have a nasopharyngeal/skull base mass, consistent with a meningocele on MRI imaging.

9.2. Labs

Laboratory assessment is pathology-driven. In the presence of a suspected hormonally active pituitary lesion, a "pituitary panel" may be ordered to assess cortisol response, TSH, prolactin,
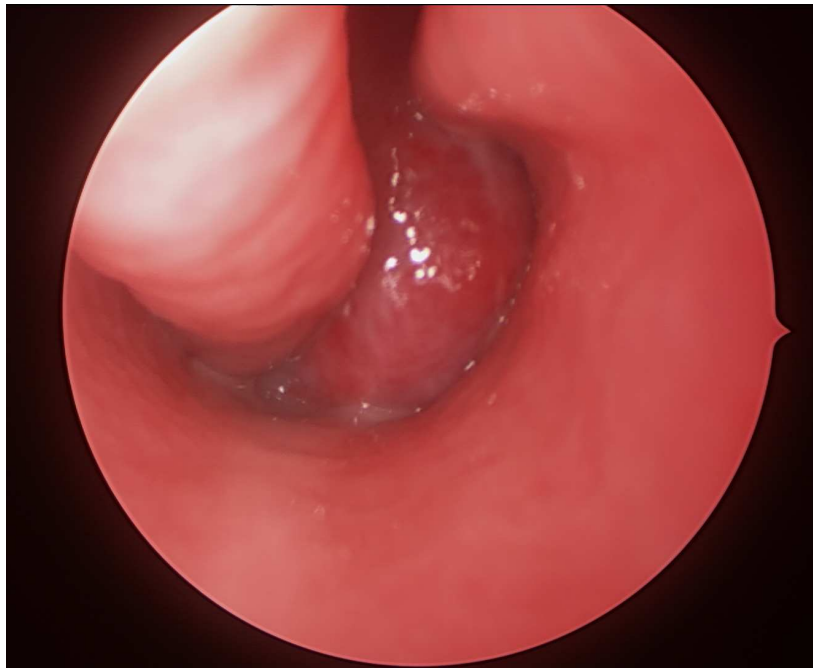
Figure 5. Right nasal endoscopy demonstrating a sinonasal mass in a 15-year-old male, consistent with JNA.

FSH, and LH levels. In the presence of a vascular tumor, preoperative hematologic labs including blood typing and cross-matching are indicated, as surgical resection can entail significant blood loss requiring transfusion.

9.3. Imaging

Preoperative imaging is integral to surgical planning in any endoscopic skull base procedure. Imaging allows the characterization of the extent of the lesion and enables identification of carotid artery involvement or encasement, intracranial extension, orbital invasion, and infratemporal fossa extension.

9.3.1. Computed Tomography (CT)

CT provides excellent bony detail and can help to identify stable surgical landmarks for intraoperative use. While soft tissue resolution is not as clear as with magnetic resonance imaging (MRI), the overall extent of the lesion can be characterized. While a non-contrast CT scan is often the first performed test with which a skull base lesion is diagnosed, a scan for intraoperative surgical navigation use should be a fine-cut, axially oriented contrast-enhanced scan that includes the maxillary alveolus, nasal tip, and extends superiorly far enough to capture the optic tracts or cranial aspect of the lesion in question, whichever is more cranial. With these images, coronal and sagittal reconstructions can be created and the images can be loaded onto a surgical navigation system for intraoperative use. The use of contrast clearly outlines the carotid arteries in their tortuous course through the clivus and cavernous sinus.
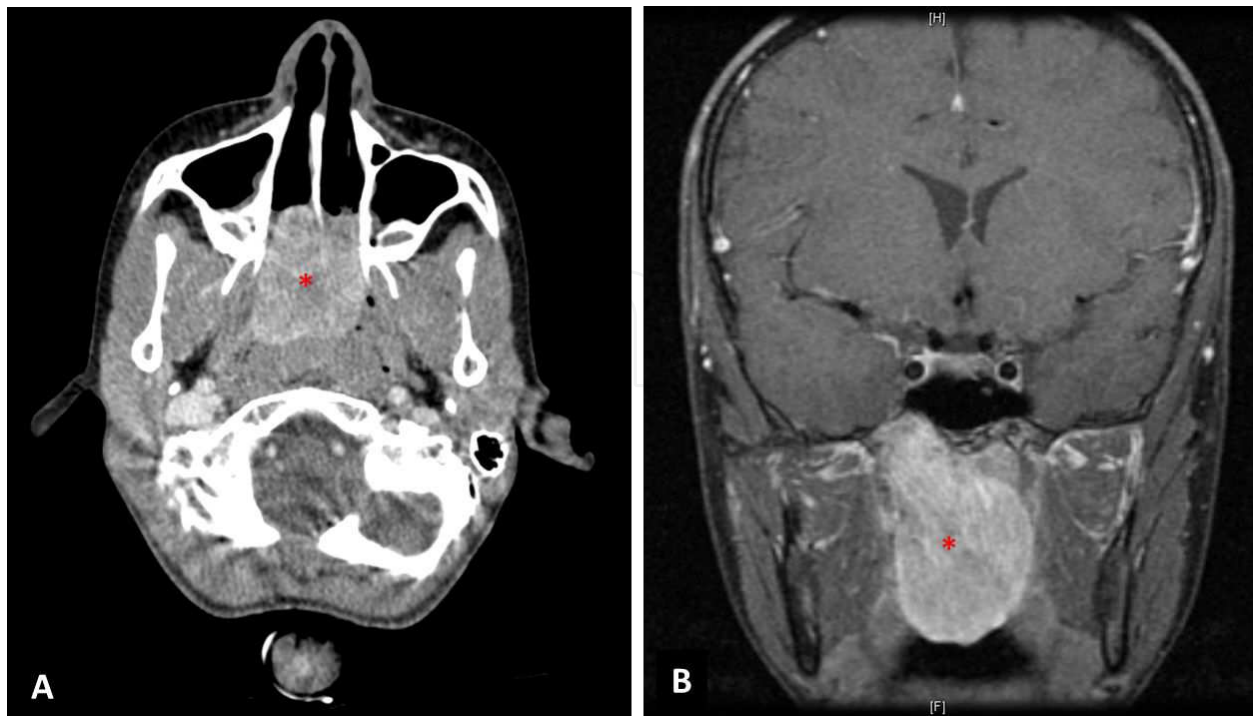
Figure 6. (A) Axial CT scan showing JNA, arising from right sphenopalatine foramen. (B) Coronal MRI showing proximity to sphenoid sinus and extension into nasopharynx and oropharynx.

9.3.2. Magnetic Resonance Imaging (MRI)

MRI utilizes the differential response of hydrogen atoms to magnetic fields to generate images with higher resolution of the soft tissue characteristics than can be obtained with CT. Whereas inspissated secretions within a sinus would be indistinguishable from the tumor invasion of that sinus on a CT scan, these two scenarios are clearly differentiated with a MRI scan. With the use of multiple image sequences including T1-weighted, T2-weighted, and contrast enhancement, soft tissue interfaces can be delineated and signal characteristics can narrow the differential diagnosis of the lesion. MRI images can also be utilized for intraoperative surgical navigation and should be formatted in a similar fashion to that described for CT. Of note, MRI requires the patient to remain still for up to 2 hours to obtain multiple sequences without image distortion from movement. For this reason, pediatric patients often require sedation or general anesthesia to obtain adequate images. Figure 6 shows a JNA and Figure 7 a meningocele on MRI.

9.3.3. Angiography and possible embolization

Several circumstances benefit from preoperative angiography and possible endovascular embolization (Figure 8). First, in the case of juvenile nasopharyngeal angiofibroma, preoperative embolization 24–48 hours prior to the planned resection enables significant devascularization of the contribution of the external carotid artery to the lesion, which leads to a decreased intraoperative blood loss [41, 42], though this finding is seen primarily when the

Figure 7. A nasopharyngeal/skull base mass, consistent with a meningocele in an infant who presented with nasal ob-struction and feeding difficulties as seen with T2-weighted sagittal MRI scan.

JNA is supplied exclusively by the external carotid system. Nevertheless, preoperative angiography identifying the location of internal carotid feeding vessels can guide dissection for vascular control [24].

The second situation in which preoperative angiography and possible embolization are beneficial is in the case of suspected carotid artery encasement. In this situation or similar situations in which the integrity of the carotid wall is in question, angiography allows for preoperative characterization of the intracranial blood flow and affords the opportunity to perform balloon occlusion testing to determine the impact of unilateral carotid sacrifice [43]. As this intervention still carries a 6% risk of cerebrovascular accident [44, 45], the decision for preoperative occlusion is reserved for situations in which intraoperative carotid injury is highly likely.

10. Surgical planning

Pediatric skull base surgery is a rapidly developing field involving multiple specialties. The collaboration between otolaryngology, neurosurgery, neuroradiology (including interven-
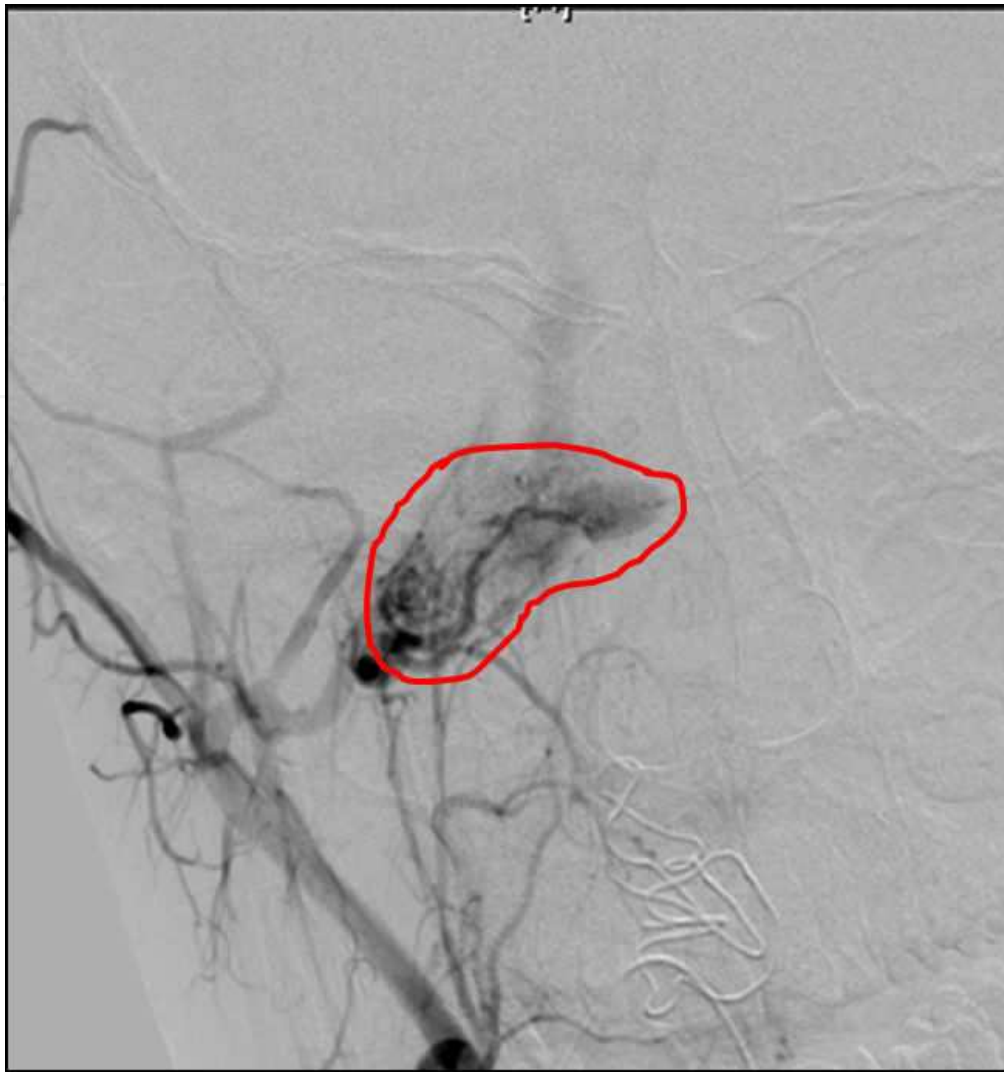
Figure 8. Angiography of external carotid artery, showing tumor blush. Blood supply from sphenopalatine artery, a branch of the internal maxillary artery. This vessel was embolized preoperatively by the interventional radiology team. Sometimes there can be feeding vessels off branches of the ipsilateral internal carotid artery or contralateral carotid system so complete assessment is necessary.

tional radiology), critical care, pediatrics, nursing, and surgical staff has evolved to allow for the optimal management of pediatric patients. Technology has driven the advancement of the management of neoplasms of the nasal cavity and skull base. Endoscopy has become critical in surgical approaches and its applications to neoplasms of the nasal cavity have been a major advance in otolaryngology and neurosurgery. Endoscopic approaches are the standard of care for most sinonasal problems and the instrumentation has been well developed.

10.1. Endoscopes and camera

Currently, rigid endoscopes are the standard for use during a surgical intervention. Although flexible scopes are used and available, they are typically used for diagnostic purposes. Rigid endoscopes allow visualization with a wide field of view, as opposed to a microscope. The

rod-lens rigid scopes are the most commonly used as they provide a high-quality image. They consist of three parts: shaft, glass fiber bundle, and optics. The endoscopes are available in various diameters but the goal is to use the largest diameter that can be accommodated. The most common rigid scopes are 2.7 and 4 mm in diameter. Endoscopes larger than 4 mm become difficult to use transnasally with other instruments. Although the use of a smaller diameter endoscope allows for greater degrees of freedom, the amount of light transmitted is decreased, which can make surgery at the skull base difficult. Several different angles are available with rigid endoscopes. Standard endoscopes part of a surgical set should include 0°, 30°, 45°, and 70°. The more angled the scope, the more disorienting the image may be. The standard approach is with the 0° to maintain the image projected as anatomic and changing to angled endoscopes as necessary. The endoscopes are attached to a light source that should be xenon. The cable is made of glass fibers, which poorly transmit heat, preventing the endoscope from causing thermal damage. The endoscopes are also attached to a HD camera that transmits the images to be projected onto monitors strategically placed in the operating room. The advancement in image technology has allowed for high resolution necessary to operate around neurovascular structures.

10.2. Surgical instrumentation

Instruments designed for endoscopic sinus surgery are standard for use in skull base procedures. The design of the instruments allows for intranasal use with a concurrent endoscope. The instruments are inserted transnasally and the nostril is used as a fulcrum about which the endoscope is pivoted. Angled instruments are part of a standard set for anterior skull base and frontal sinus and are used often with the angled endoscopes. Straight instruments are designed to be low profile and allow for easy visualization of the tip of the instrument and the endonasal structures. Instruments including curettes, elevators, scissors, forceps with and without cutting capability, and drills are each designed specifically for working intranasally. Powered drills and microdebriders are used often to remove tissue for access and for the resection of certain lesions. Bipolar forceps are designed with appropriate size and length to work intranasally in order to achieve hemostasis. Monopolar cautery is to be avoided close to neurovascular structures but is integral to harvesting of pedicled vascular flaps.

10.3. Use of surgical navigation systems

The use of intraoperative surgical navigation systems has become commonplace in the field of endoscopic skull base surgery. Often neoplasms obscure normal landmarks, and navigation systems allow for greater spatial awareness during these dissections. Image-guidance systems use both optical and magnetic field technology and both have advantages and disadvantages. Image guidance for skull base is typically based on preoperative high-resolution CT scans but can be fused with MRI images as well. The information provided by this technology has been referred to as "image guidance," but this is a misnomer when utilized appropriately. The use of surgical tools connected to the navigation system allows additional confirmation of visually identified landmarks and assists in determining the extent of resection in complex skull base lesions or anatomy in a field distorted by the disease process or previous surgery [46]. In the

management of CSF leaks, intraoperative surgical navigation can confirm visually suspected areas of bony dehiscence. While not a sine qua non in endoscopic skull base surgery, intraoperative surgical navigation is a valuable tool that should be utilized when available. Of utmost importance in the use of these systems is ensuring accurate registration of the patient and to periodically confirm accuracy with navigation to known landmarks [46, 47].

11. Operating suite

Endoscopic skull base surgery is often a two-surgeon procedure and the setup of the operating room must accommodate this arrangement. With two surgeons on either side of the patient with a surgical technician, a nurse, or an assistant, it is necessary to have the patient in the 180° position from anesthesia. Communication with the anesthetist for the control of the airway is necessary as these procedures can take several hours. Establishing and securing the airway, intravenous vascular access, arterial lines, and monitors are necessary to prevent adjustments or interruptions of the procedure. The head of the patient can be elevated slightly as well to facilitate venous drainage. The HD monitors and image guidance system can be integrated in the operating room and each surgeon and assistant must have a monitor in a direct line of site. This necessitates a suite designed with multiple monitors linked to a central control center. The instrument table and assistant should be positioned equidistant from the two surgeons without physical obstruction to passing of instruments.

12. Surgical approaches

Total endoscopic approach is appropriate for many sinonasal tumors. The neoplasms can be removed through one or both nostrils depending on the extent. The use of both nostrils requires a trans-septal approach either by creating a perforation or by resection of the posterior septum. The choice is dependent on the location of the tumor. The uninostril (through one nostril) procedure is less destructive to normal structures but can limit the degrees of freedom and the number of instruments placed in the nose. Binostril approach allows for one surgeon to work bimanually and another surgeon to control the suction and the endoscope. For extensive lesions, this is the preferred technique.

The approaches have variations in the trajectory to the skull base and four different endonasal approaches have been described: transnasal, transethmoid, transsphenoidal, and transpterygoidal.

The transnasal approach does not traverse the sinuses but essentially keeps the septum and the middle turbinate as the boundaries. Lesions of the cribiform plate such as encephaloceles can be managed in this manner. In addition, clival and odontoid pathology can be addressed through the choana.

The transethmoid approach requires a total ethmoidectomy and can expose the skull base from the frontal to sphenoid sinus. The lamina papyracea is exposed as well. This approach is useful for encephaloceles or orbital apex lesions.

The transsphenoidal approach requires removal of the anterior face of the sphenoid and often the sphenoid rostrum and posterior septum. Removal of the middle turbinate can increase the degrees of freedom and optimize visualization. The transsphenoid approach is used for sellar, pituitary, upper clival, and cavernous sinus lesions. Figure 9 shows a craniopharyngioma which was managed in this manner.

The transpterygoidal approach requires a corridor through the maxillary sinus and the removal of the posterior wall of the sinus. A medial maxillectomy and ligation of the spheno-palatine artery are necessary which is often done with JNA resection (Figure 10). This approach is useful for the infratemporal fossa. Figure 11 required the pterygoid bone to be removed for access to a rhabdomyosarcoma.
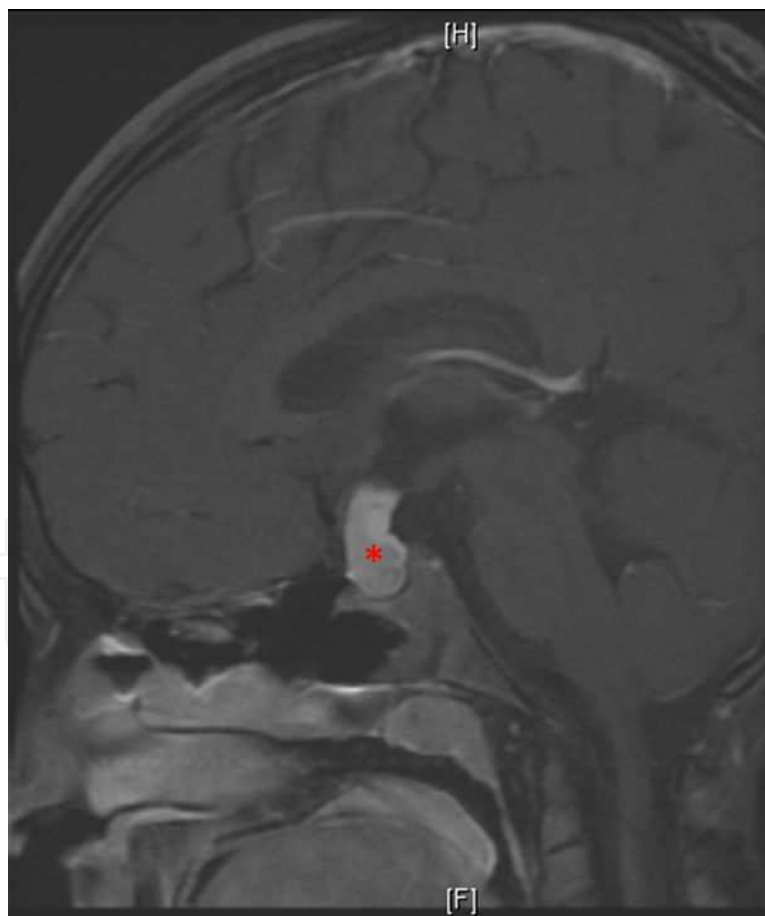

Figure 9. A 13-year-old male who presented with new onset vision loss was found to have a craniopharyngioma on this sagittal T1-weighted MRI with contrast. An endoscopic, transsphenoid approach was performed for removal.

Figure 10. A JNA s/p endoscopic resection from pterygopalatine fossa, nasal cavity, sphenoid sinus, and nasopharynx.
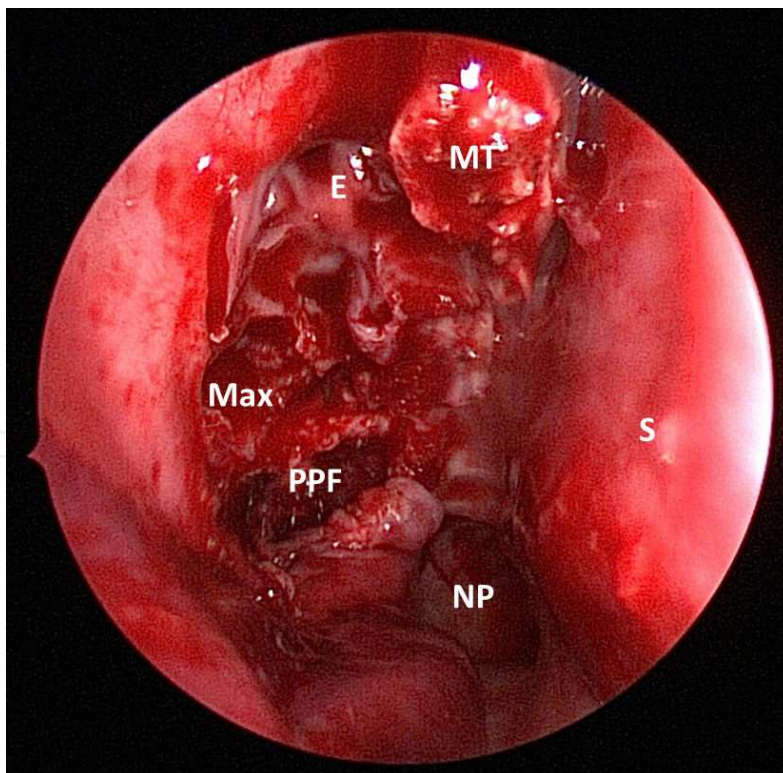

Figure 11. Right endoscopic view for approach and biopsy of pterygopalatine fossa mass that was a rhabdomyosarcoma. The pterygoid bone had to be drilled down medially in order to get access. S=septum, NP=nasopharynx, MT=middle turbinate, E=ethmoid, Max=maxillary sinus, PPF=pterygopalatine fossa.

13. Hybrid approaches

Although many lesions can be approached by endoscopic-only approach, certain lesions necessitate a hybrid approach with the use of open techniques. Lateral lesions of the infra‐temporal fossa may require a Caldwell-Luc combined with transpterygoid. Lesions that are far anterior, such as neoplasms at the anterior pyriform aperture or of the frontal sinus, often require a hybrid approach. Other open approaches such as bicoronal, lateral rhinotomy, or palatal split (Figure 12) can be used depending on the extent of surgery, and endoscopy can be a useful adjunct to ensure complete resection. Transcervical endoscopic-assisted surgery of the skull base can be a useful adjunct to access difficult-to-reach areas as shown in Figure 13.

Endoscopic-assisted trephination of the frontal sinus with a repair of an isolated posterior table fracture with CSF leak has been described in a pediatric patient. This avoided the need for bicoronal flap and craniotomy for open repair [48].
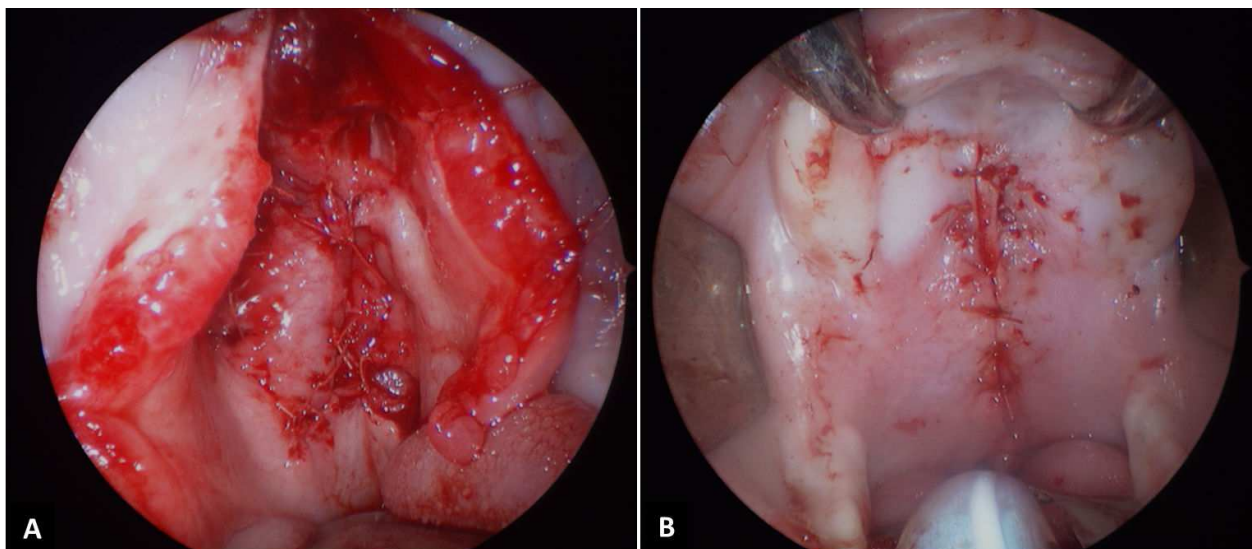

Figure 12. (A) Endoscopic-assisted, transpalatal/palatal split approach to skull base in nasopharynx for excision of me‐ningocele in an infant. The palatal flaps are retracted and a vascularized pharyngeal flap was placed over a cartilage graft on right side of superior nasopharynx to repair the bony defect in skull base. (B) Multilayer repair of the palate.

14. Staging surgery

The goal of any approach to a neoplasm is ideally total removal of the tumor with minimal damage to normal structures. The strategy for sinonasal tumors must be defined and the pathology often dictates the approach. Some tumors such as juvenile nasopharyngeal angio‐fibroma can be diagnosed by imaging. Other lesions need initial biopsy. Biopsy of the sinonasal tumors requires adequate imaging to ensure safety of any disruption of the tumor as intra‐cranial extent or prominent vascularity can make limited biopsies potentially dangerous. Consultation with the radiologist can be helpful for planning.
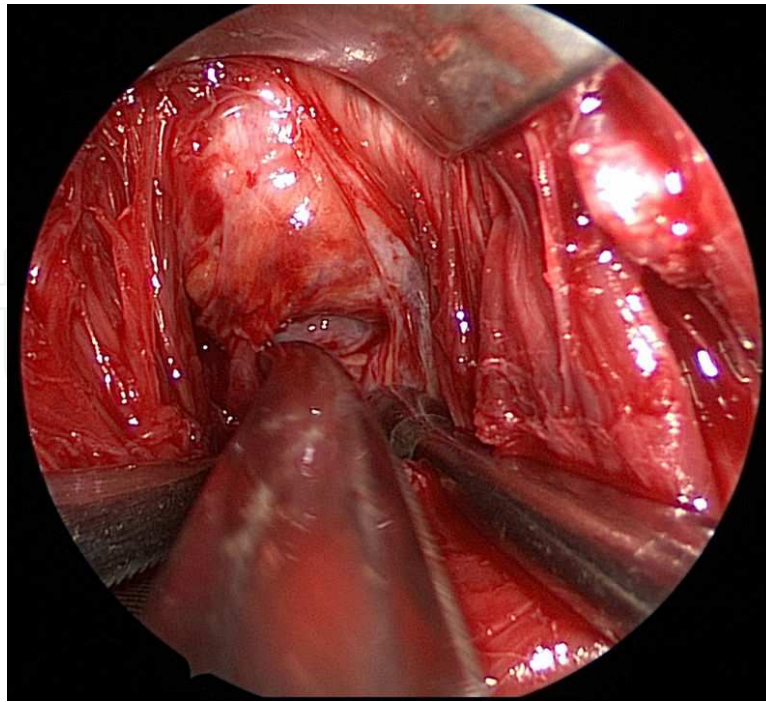
Figure 13. Endoscopic-assisted, transcervical approach to the left parapharyngeal space and skull base for excision of recurrent pleomorphic adenoma after initial excision at age 15. The mass was dissected from the skull base with endoscopic visualization and avoidance of mandibulotomy and plating for surgical access.

Due to the proximity of neurovascular structures such as the optic nerve and carotid artery, incomplete resection may be necessary to preserve function or prevent injury. Certain benign pathologies such as fibrous dysplasia can make this a feasible strategy. In addition, lymphomas and certain subtypes of rhabomyosarcoma are chemotherapy- and radiation-sensitive, and the morbidity of complete resection makes this approach prohibitive.

15. Reconstructive options

The endoscopic approach to resection of sinonasal tumors is only feasible because reconstructive techniques exist to maintain the separation between the nonsterile nasal cavity and the sterile intracranial contents. As the extent of tumors and surgical approach varies, so do the reconstructive techniques. The goal is to create a water- and air-tight closure to prevent CSF leak and meningitis. Preoperative planning includes reconstruction for the anticipated defect. Reconstruction may span from no reconstruction with healing by secondary intention to the use of vascularized grafts. Ideally, the reconstruction should match the tissue resected. If a large bony defect is encountered, rigid support may be necessary to prevent herniation of intracranial contents. Bone, cartilage, fascia, mucosa, and fat can be used as autologous free grafts.

Free tissue grafts can be harvested intranasally. If middle turbinate is resected, the bone and mucosa can be used for small defects of the skull base. The middle turbinate free graft can be

filleted open and used as an onlay graft. If a more rigid tissue is necessary, cartilage can be harvested from the nasal septum and used as an inlay graft to repair skull base defects. Multilayered grafts that combine various types of tissues are often used. Once the grafts are placed, fibrin-sealing agents can be placed over the surface of the reconstruction to provide a watertight closure.

One of the recent advances in skull base reconstruction is the use of the pedicled nasal septal flap, which is based on the posterior septal artery [49]. This flap is useful for defects with a concomitant CSF leak. Planning of this flap is important specifically with transsphenoidal approaches in order not to damage the blood supply to this flap. Other intranasal flaps include the inferior turbinate flap or middle turbinate flap, both based on branches of sphenopalatine artery. Scalp flaps include the pericranial flap (supraorbital and supratrochlear arteries), temporoparietal fascia flap (superficial temporal artery), or temporalis muscle flap (deep temporal arteries).

All of the skull base reconstructions require some packing to keep contact and ensure adequate healing. A combination of absorbable and non-absorbable packing can be used. Typically, gel foam is placed over the reconstruction, and non-absorbable splint or packing is used for additional reinforcement.

15.1. Use of lumbar drain

In the past, lumbar drains were routinely used for decompression during an endoscopic repair of a CSF leak. With the advent of reliable vascularized flaps, the need for lumbar drainage has decreased. It is now thought that low-flow CSF leaks can generally be managed surgically without the use of the lumbar drain, while in some high-flow leaks it should be considered [50].

16. Postoperative management

After surgical intervention, close monitoring in the hospital setting is recommended for pediatric patients. Depending on the extent of surgery and need for hemodynamic monitoring, and frequent neurologic checks, the patient may go to the pediatric ICU setting as opposed to a traditional floor. When there is any violation of the dura or CSF leak repair is preformed intraoperatively, there should be close observation for any CSF rhinorrhea. Activity levels should be determined based on the extent of surgery. Stool softeners can help reduce straining, as constipation may increase intracranial pressure and a risk of CSF leak. Antibiotic prophylaxis postoperatively is considered in a setting of CSF leak repair [51] or when absorbable packing is used to reduce the risk of infection.

17. Surgical complications

There are several potential complications from endoscopic skull base surgery. Given the high vascularity of these operative sites, and even despite "near complete" embolization of a JNA,

there can be significant blood loss intraoperatively, which requires blood transfusion. In a pediatric patient, the decision to stage resection is sometimes made to avoid blood transfusion. A major vascular injury to the cavernous sinus, carotid artery at skull base, or intracranial hemorrhage can occur. Cerebral vascular accident or strokes can also occur. When the muscles of mastication are within the surgical field, trismus postoperatively can occur and patients should begin jaw-stretching exercises at the earliest stages. CSF leak can occur when the dura is violated, but the key is recognizing it and performing an endoscopic repair. The risk of meningitis is significantly reduced with prompt and proper repair of a CSF leak. Infections including sinusitis and meningitis are possible. Endocrinopathy can occur when the pituitary gland is involved. Altered olfaction can result from either a change in nasal airflow dynamics or injury to the olfactory bulb or other special sensory fibers within the nasal cavity. Other cranial nerve injuries, including vision loss or blindness, diplopia, dry eye symptoms, and facial numbness can also occur.

18. Conclusions

The use of pediatric endoscopic skull base surgery has continued to grow as techniques and technology pioneered in the adult population have been applied to children. The need for open surgical approaches has decreased with the advancement of endoscopic techniques that are now available to address pathologic conditions of the skull base in the pediatric population. Experienced surgical teams with multidisciplinary support at tertiary care pediatric facilities can best manage these young patients.

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