Lateral Skull Base Approaches in Pediatric Skull Base Surgery

Ken Kazahaya^{1,2}

¹ Division of Pediatric Otolaryngology, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States

² Department of Otorhinolaryngology/Head and Neck Surgery, University of Pennsylvania, Philadelphia, Pennsylvania, United States

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Address for correspondence Ken Kazahaya, MD, MBA, FACS, Division of Pediatric Otolaryngology, Department of Skull Base Surgery Program, The Children's Hospital of Philadelphia, 3401 Civic Center Blvd, Philadelphia, PA 19104, United States (e-mail: kazahaya@email.chop.edu).

Abstract

Keywords

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- cholesteatoma
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- temporal bone

Lateral skull base pathology is rare in children. Awareness of the potential for lateral skull base lesions in children is imperative for timely identification and appropriate management. Some of the common presentations and pathologies shall be presented, as well as a variety of approaches that may be utilized to access the lateral skull base in the pediatric patient. Although the lateral skull base approaches utilized in adults may also be considered for management of pediatric lesions, some special considerations given the small developing anatomy need to be kept in mind.

Introduction

The management of pediatric skull base lesions requires understanding of the developing anatomy of the skull base, the different histopathologic diagnoses in children, special surgical considerations and also need to be particularly cognizant of the long-term morbidity, and complications that the disease and its treatment may bring. A conservative “wait and see” or radiation therapy for a benign lesion may not be appropriate in a child who could have a long life span.

Although there have been significant technical advances in adult cranial base surgery, there has been slow translation to the pediatric population. Since there are smaller numbers of affected children than adults, few centers specialize in the evaluation and management of pediatric cranial base lesions.

Commonly with a favorable outlook of a long life after treatment, the potential morbidity (particularly facial nerve paralysis and hearing loss) from different treatment choices and potential complications need strong consideration. Facial nerve deficits and hearing loss both have significant impact on the quality of life for a child.

The histopathology in children tends to be more benign than in adults—tending to have better overall outcomes than in adults. Lesions of the lateral skull base may cause functional problems that usually lead to evaluations that will reveal an indolent disease process. Management and surgical resection of these lesions is often challenging and may lead to further functional impairments. With the advances on technologies and techniques, the objectives of management of the lesions have evolved from focusing on eradication of the lesion to also including consideration for the preservation and restoration of function.

Imaging in children needs to be interpreted with an understanding of the developing cranial base. The cranial base bone is not fully ossified in infants and younger children. Also, their small size may make even thin imaging sections relatively “thick.” The developing anatomy will have findings of the lack of pneumatization and marrow in the mastoid and petrous bones. Furthermore, young children often have otitis media with effusion so if there is temporal bone or middle ear pathology to be evaluated, an otoscopic evaluation of the middle ear may be prudent.

Some imaging modalities, in particular, magnetic resonance imaging (MRI), may also require sedation or even general anesthesia to obtain an adequate study. Radiation exposure consideration to the developing brain also has come into scrutiny over the past few years. Therefore, computed tomography (CT) scans should be performed with pediatric dose reduction. Also, rather than perform a CT without and with contrast, often would also consider performing one or the other not both scan acquisitions.

Image guidance in the lateral skull base approaches can be challenging in children. Given the lack of prominent landmarks such as the nose and brow of the face, which are used for registering the image-guidance system (IGS) for an anterior approach, often fiducial markers are utilized while obtaining preoperative imaging of the lateral skull base. Children tend to pick and remove the fiducial markers; therefore, the IGS-images with the fiducial markers may need to be obtained the day of the surgical procedure. The child can be anesthetized, the fiducial markers placed, imaging obtained, and then proceed directly to the operating room for the surgery. Intraoperative flat panel or other imaging modalities can be useful especially if the IGS can be integrated with the imaging modality to autoregister the scans with the patient.

Developing Anatomy

At birth, the otic capsule, the middle ear, and ossicular chain are completely developed. The temporomandibular joint (TMJ), external ear canal, and mastoid still have to undergo significant development.

After birth, the mastoid and petrous bone grows slowly and pneumatizes. Marrow that fills the mastoid and petrous bones gives way to enlarging air cells. There are theories that the mastoid air cell development may be restricted by early or persistent middle ear disease. All portions of the temporal bone, except for the petrous bone, continue to enlarge after birth.

In addition to the obvious difference in size of the skull base in children, there are significant inconsistencies in the

usual anatomic landmarks in children when compared with the fully developed adult cranial base anatomy.¹

In infants, the outer surface of the temporal bone is markedly different from an adult temporal bone, as there is lack of an inferior portion of the bony ear canal and there is no mastoid process. The mastoid process begins to appear around 1 year of age. The mastoid process is relatively well developed by 3 years of age. Lack of the mastoid process in infants can result in the stylomastoid foramen to be exposed and the facial nerve tends to be very superficial and is more susceptible to trauma such as by forceps used during delivery or injury during extratemporal surgical approaches (►Fig. 1).

The majority of skull base growth of the temporal bone occurs during the first half of the first decade of life, and continues through the first decade of life.² Effects upon bone growth and skull development after lateral skull base approaches are not well described. The disturbance of ossification and growth centers and unfused suture lines may affect the development of the skull; however, the effects of the various approaches have not been well described and will require further investigation.

Pediatric Lateral Skull Base Pathologies

Pathology of the lateral skull base can be separated into neoplastic, non-neoplastic, inflammatory, and infectious categories (►Table 1). Tumors of the temporal bone are rare—approximately 6 per million in adults. Temporal bone malignancies in children are even rarer. In children, benign tumors are about three times more common than malignant tumors.^{1,3} In one review article of 241 pediatric head and neck malignancies, the temporal bone was the primary site of only 1.5% of cases.⁴ Temporal bone lesions may have a delayed diagnosis in children due to their insidious nature and also as many of the symptoms mimic other common pediatric otologic processes such as otitis media. A high degree of suspicion is necessary to

Fig. 1 Comparison of infant temporal bone (left) and adult temporal bone (right). In infants, there is a lack of an inferior portion of the bony ear canal and there is no mastoid process. The mastoid process begins to appear at around 1 year of age. The mastoid process develops relatively well by 3 years of age. Lack of the mastoid process in infants can result in the stylomastoid foramen to be exposed and the facial nerve tends to be very superficial and is more susceptible to injury.

Table 1 Some tumors/masses of the temporal bone in children

Benign tumors/Masses	Malignant
Aberrant carotid artery	Adenocarcinoma
Adenoma	Adenoid cystic carcinoma
Aneurysmal bone cyst	Burkitt lymphoma
Carcinoid tumor	Chondrosarcoma
Cholesteatoma (middle ear/mastoid, petrous)	Desmoid tumor
Cholesterol granuloma	Endolymphatic sac tumor
Chondroblastoma	Ewing sarcoma
Chondroma	Fibrosarcoma
Chondromyxoid fibroma	Fibrous histiocytoma
Choristoma	Ganglioneuroblastoma
Endodermal sinus tumor	Granulocystic sarcoma
Encephalocele	Hemangiosarcoma
Eosinophilic granuloma	Juvenile fibromatosis
Fibrous dysplasia	Liposarcoma
Giant cell tumor	Lymphosarcoma
Glomus tumor	Metastatic neuroblastoma of adrenal gland origin
Granular cell tumor	Rhabdomyosarcoma
Granuloma	Squamous cell carcinoma
Hamartoma	
Hemangioma	
Hemangiopericytoma	
Langerhans cell histiocytosis	
Lipoma	
Lymphangioma	
Melanotic neuroectodermal tumor of infancy	
Meningoencephalocele	
Meningioma	
Neurofibroma	
Ossifying fibroma	
Osteoblastoma	
Osteoma	
Paraganglioma	
Reparative granuloma	
Schwannoma (acoustic, facial, chorda tympani, jugular foramen, trigeminal, vestibular)	
Teratoma	
Tuberculosis	
Wegener's granulomatosis	

appropriately diagnosis lesions of the temporal bone, especially if the child does not respond to treatment or if pain is out of proportion or unrelated to an inflammatory or infectious etiology. Suspicion should also be raised if there are symptoms such as facial nerve paralysis, sensorineural hearing loss, or vertigo (►Table 2). There are also characteristic

findings on radiologic studies that can assist with the diagnosis of various lesions (►Table 3).

Non-neoplastic lesions of the lateral skull base include pathologies such as cholesteatoma, cholesterol granuloma, and eosinophilic granuloma. Infectious and inflammatory lesions of the lateral skull base are rare and include lesions

Table 2 Some presenting symptoms for lateral skull base lesions

– Hearing loss
– Chronic otorrhea
– Dizziness/vertigo
– Tinnitus
– Facial nerve palsy
– Headache
– Recurrent meningitis

such as tuberculosis of the temporal bone and granulation from chronic infection.

In a recent retrospective study over 25 years, there were 4,523 skull base surgeries performed of which there were 63 patients 18 years and younger who underwent lateral skull base procedures (1.4% of all skull base surgeries).⁵ Most common presenting symptoms were hearing loss (44.6%) and chronic otorrhea (43.1%). Other symptoms included dizziness or vertigo (26.2%), tinnitus (13.8%), and facial nerve palsy (10.8%). Facial nerve function was normal in 81.5% of subjects. Twenty-three patients (35.4%) had complete hearing loss at presentation. In Grinbalt's study, 38.5% of subjects had neoplasms, most common were vestibular schwannomas (15.4%), facial nerve tumors (7.7%), and paragangliomas (4.6%); about a half of the subjects had nonneoplastic pathology—most commonly cholesteatoma (42.8%). There were three patients with cholesterol granulomas.

Congenital Cholesteatoma—Middle Ear/Mastoid and Petrous Apex

Congenital middle ear cholesteatoma is relatively uncommon comprising ~2 to 5% of cholesteatoma.⁶ Typically presents as an asymptomatic white mass in the middle ear space—most often in the anterior superior quadrant. By

definition, they are present at birth and there should be no prior history of surgery in the ear. Most commonly diagnosed around 4 years of age. There is a male preponderance of 3:1.⁷ Early diagnosis is important for optimal outcomes. If the cholesteatoma is small and encapsulated, it can often be successfully removed without hearing loss or risk of residual or recurrent cholesteatoma.

Imaging of cholesteatoma usually reveals a lytic lesion on CT often with smooth convex edges in the bone. MRI appearance of cholesteatoma is typically hypointense on T1, with variable T2 intensity, does not enhance, and hyperintense in diffusion.

Surgical approach depends upon the extent of the cholesteatoma. Mastoid involvement commonly requires a mastoidectomy. Whether the posterior ear canal is left intact or taken down to create a large mastoid bowl is dependent upon the extent of the cholesteatoma and the surgeon. Some surgeons feel that they will initially perform an intact canal wall procedure to avoid a mastoid bowl in a child and the need for routine debridements. The intact canal wall procedure may require a “second-look” procedure to look for recurrent/residual cholesteatoma. Diffusion-weighted MRI imaging may provide an alternative to the “second-look” procedure for potential recurrence/persistent cholesteatoma. There are proponents of the canal-wall down procedure and creating an easily monitored cavity without sacrificing hearing outcomes.

Schwannoma

Vestibular schwannomas are the most common neoplasm requiring a lateral skull base approach. Often pediatric patients present with more advanced stage of disease due to late presentation. These often present with increased intracranial pressure, cerebellar symptoms, or cranial neuropathies. Pediatric schwannomas tend to be more vascular and difficult to remove than in adults.⁸ As in adults, the size, position, and hearing will help determine which approach is appropriate for management of the vestibular schwannoma.

Table 3 Differential radiologic findings for different tissue types

	CT scan	MRI	MRI	MRI
		T1	T2	+Gadolinium
Marrow	May be dense	Hyperintense	Hypointense	May enhance
Cholesteatoma	Usually has bony erosion/destruction	Hypointense	Hyperintense	No enhancement
Cyst – mucous	Opacification without bony destruction	Reduced intensity	Increased intensity	No enhancement
Cholesterol granuloma	Cystic expansion, bony remodeling	Hyperintense	Increased intensity	No enhancement
Fibrous dysplasia	Bony enlargement with sclerotic or classic ground-glass lesion	Low to intermediate signal	Low to intermediate signal	Enhance mildly
Neoplasm	Bony erosion/destruction	Decreased intensity	Decrease intensity	Enhances

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.

CT imaging of schwannomas typically find a mass along a nerve or with enlargement of a neural foramen. Bone erosion has a regular margin. MRI images reveal a lesion hypointense on T1, isointense on T2, and enhances.

Meningioma

Meningiomas are tumors that arise from the membranes that surround the brain—the meninges. They are the most common tumors that arise in the head. Most are slow growing, and often have an insidious onset of symptoms. Since they are slow growing, they may not require immediate intervention if asymptomatic and without significant signs. CT scans may reveal hyperostosis and bone erosion. MRI imaging reveals intermediate signal on T1 and variable signal intensity on T2, the dura is thickened, and enhances.

Fibrous Dysplasia

Fibrous dysplasia (FD) makes up ~7% of all benign bone tumors.⁹ Most common presentation of FD in the temporal bone is stenosis of the external auditory canal (EAC). FD reflects an arrest of bone maturation. There is a formation of collagen with little production of cancellous tissue. The process is proliferative and will often extend beyond the normal anatomical limits of the affected bone and cause bony expansion, compression of surrounding tissues and structures, and weakness of the bone. FD grows from within the bone eroding the outer cortex of the affected bone, leaving a weak thin shell of cortical bone. FD has an average age of onset of around 10 years of age, and can be monostotic (one bone) or polyostotic (multiple bones affected). FD often presents with painless bony swellings. About one-third of children will present with cutaneous nevi. Monostotic form is four times more common than the polyostotic form and ~30 times more common than Albright syndrome. Monostotic FD will often arrest growth during puberty, the polyostotic form usually continues beyond the third decade of life. Albright syndrome is a triad of FD, pigmented areas of skin, and precocious puberty. Head and neck involvement is ~10% in monostotic FD, and 50 to 100% in polyostotic disease. The temporal bone is involved in ~20% of cases of head and neck bony involvement. About 2:1 monostotic to polyostotic forms. Diagnosis should be confirmed with biopsy of the lesion if easily accessible. There is potential for malignant transformation in FD, with the incidence malignant transformation the highest in patients with monostotic cranial disease. Malignant transformation is often associated with pain, rapid swelling, and elevation of alkaline phosphatase. Many of the case of malignant transformation are in cases that have received prior radiotherapy treatment

Langerhans Cell Histiocytosis (Histiocytosis X)

Eosinophilic granuloma, Hand–Schuller–Christian disease, and Letterer–Siwe disease are different presentations of Langerhans cell histiocytosis (histiocytosis X). Eosinophilic granuloma is a more localized form, limited to bone and typically monostotic. Hand–Schuller–Christian disease comprises a classic triad of skull lesions, exophthalmos, and diabetes

insipidus. Letterer–Siwe disease is a disseminated form of histiocytosis X that involves multiple visceral organs. The incidence of Langerhans cell histiocytosis in the United States is ~0.05 to 0.5 cases per 100,000 children. Temporal bone involvement is identified in 15 to 61% of cases with the initial presentation in the temporal bone in as many as 25% of cases. Temporal bone involvement is often confounded by similar signs of more common disorders such as mastoiditis, otitis externa, chronic suppurative otitis media, or cholesteatoma.

Surgery in these cases is typically confined to surgical biopsy and curettage of the affected bone. If the mastoid is involved, a simple mastoidectomy may be all that is needed. For larger symptomatic temporal bone lesions, radiotherapy alone or combined with surgical curettage may be undertaken. In younger children, due to the undesirable late effects of radiotherapy, chemotherapy is commonly considered.

Parangliomas

Parangliomas are tumors of neural origin. Majority of these are located in the abdomen. They represent ~30% of pediatric pheochromocytomas. Parangliomas are usually slow-growing and benign, however, can undergo malignant transformation. Most parangliomas are sporadic; however, there are some syndromes associated with parangliomas such as von Hippel–Lindau (VHL) syndrome, succinate dehydrogenase (SDH) gene mutations, and multiple endocrine neoplasia type 2. About half of the parangliomas secrete catecholamines and may cause hypertension, tachycardia, flushed skin, sweating, headaches, and tremors.

Imaging of parangliomas on CT often shows irregular bony erosion. MRI imaging reveal a lesion hypointense on T1, hyperintense on T2, pathopneumonic salt and pepper pattern (from vessel voids and hemorrhage), and intense enhancement.

Surgery is the recommended treatment of choice if feasible. Lower cranial nerve (CN) deficits are not uncommon with tympanojugular paranglioma resections. Jackson et al noted that complication rates were higher and had reduced preservation of lower CN function in pediatric cases.¹⁰

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common soft tissue sarcoma in infants and children—comprising ~5 to 15% of all neoplasms in children with an annual incidence of 5 to 6 per million children under 15 years of age. Peak occurrences happen between 2 and 5 and between 15 and 19 years of age. The head and neck being the most common sites of involvement with ~35% of cases, however, less than 10% of these begin in the temporal bone. Rhabdomyosarcoma is the most common pediatric malignancy of the temporal bone.

Treatment of rhabdomyosarcoma: If localized and the mass can be excised completely or with microscopic residual disease without creating significant morbidity or disability, then surgical resection should be pursued as the initial treatment modality. If complete resection can be achieved, then postoperative radiation treatment may be avoided. Extensive surgery should be avoided as a first line of treatment if it involves sacrifice of vital structures or creates

significant morbidity. Often in cases of temporal bone rhabdomyosarcoma, the surgery is limited to a biopsy to confirm the diagnosis. Aggressive lateral skull base surgery may be necessary to treat residual or recurrent disease after completing combined chemotherapy and radiotherapy.

Leukemic Involvement of the Temporal Bone

About 20% of patients with leukemia have involvement of the temporal bone, and infrequently is the initial or presenting symptom or sign of leukemia. It can present with lesions of the skin in the ear, red or thickened tympanic membrane, middle ear effusions, mastoiditis/mastoid opacification, or hearing loss. Very rarely, it may present with facial nerve paralysis.

Petrous Apex Lesions

Petrous Apex Cholesteatoma

Petrous apex cholesteatoma is a much less common lesion than congenital cholesteatoma, and often grow to a large size prior to becoming symptomatic—presenting most commonly as headaches, however, may also present with hearing loss or facial nerve paralysis. The etiology of petrous apex cholesteatoma is not known. Gacek theorized that these lesion may arise from epithelial remnants trapped in the region of the foramen lacerum.¹¹ Petrous apex cholesteatomas are often large by the time symptoms begin to become evident—headaches, hearing loss, facial nerve spasms, or paralysis are common initial symptoms. CT findings usually show bone erosion with clearly defined edges. MRI reveals a lesion that is nonenhancing and low intensity on T1 while high intensity on T2. These are difficult lesions to access given their location.

Other petrous apex lesions include cholesterol granulomas, mucous cysts, encephalocoeles, bone marrow, and tumors such as chordomas, chondroma, or chondrosarcomas.

Approach to petrous apex lesions is challenging due to its location. If hearing is intact, may need to approach the lesion with a cochlea sparing technique such as a middle cranial fossa (MCF) approach or retrolabyrinthine. Transcochlear approach may be considered if hearing is lost. For drainage of cholesterol granulomas, a subarcuate or infracochlear approach may be undertaken depending upon the pneumatization of the temporal bone. In adults with well pneumatized sphenoid bones, it is sometimes possible to drain the petrous apex via a transsphenoidal pathway.

Cholesterol Granuloma

Cholesterol granulomas are benign cystic collections, usually found in the petrous apex. On CT scan, they are often rounded and have regular margins with an intensity similar to brain tissue. MRI findings are hyperintense signal on T1 and T2 imaging and does not enhance with gadolinium. There are various theories as to their origin. If they grow and are left untreated, they can cause hearing loss, headaches, facial pain, diplopia, dizziness, and tinnitus. Management of asymptomatic, stable lesions may be surveillance, while treatment of enlarging lesions involves drainage of the cyst.

Adenocarcinoma

Endolymphatic sac tumors (ELSTs) may present in children with VHL syndrome—involves the VHL gene on chromosome 3p25.¹² Patients with VHL syndrome have predisposition for developing benign and malignant neoplasms, such as paragangliomas, hemangioblastomas, clear cell renal carcinoma, pheochromocytoma, and pancreatic neuroectodermal tumors. ELSTs are also described with VHL syndrome. ELST is a papillary adenocarcinoma of the endolymphatic sac. ELSTs are rare tumors of the temporal bone that may cause extensive destruction of the temporal bone and invasion of the cerebellopontine angle (CPA). Hearing loss and tinnitus are frequent presenting symptoms. The radiologic findings include heterogeneous signal on T1 with peripheral rim enhancement. Flow voids are common in ELST that are greater than 2 cm in size.¹³

Approaches

Preoperative assessment of audiologic and facial nerve function should be attempted to be obtained. Depending upon the age of the child, audiologic assessment may be limited and various measures of audiologic function may be necessary to be utilized by experienced audiologists. Vestibular function may also be difficult to assess depending upon the age of the child. Facial nerve function must also be carefully assessed as the musculature tone in children may allow for passive eye closure even with full facial nerve paralysis. Electromyography (EMG) may be used to assess facial nerve function.

In children, the mastoid air cells are less developed than in adults. Sometimes, there is solid bone and marrow that makes the identification of otic capsule structures and other important temporal bone structures more challenging. Since the mastoid bone is not well developed and pneumatized in a child, the tegmen and sigmoid sinuses are lower and more anterior, respectively, narrowing the window of access through the mastoid. However, the lateral exposures in children often require less retraction of neural structures than in adults.

Gil et al noted that a child's brain is tighter than the adults, and typical maneuvers, such as ventriculostomy or opening of the basal cistern, that can be used to provide relaxation in an adult brain are not effective in reducing the brain tightness in children.¹

Approaches described for use in pediatric lateral skull base surgery include subtotal petrosectomy, translabyrinthine approach (TLA), retrosigmoid approach, combined translabyrinthine/retrosigmoid approach, transotic approach, infralabyrinthine approach, infratemporal fossa approaches, and MCF approach.

Retrosigmoid approach and TLA typically will be able to preserve facial nerve function. Cerebrospinal fluid CSF leaks have been reported for these approaches in children.¹⁴ Subtotal petrosectomy was noted to also have preservation of facial nerve function in Grinblat et al's series.⁵

Teo et al noted the transcochlear allows for access to lesions that are located in both the middle and posterior

cranial fossa and also lesions anteromedial to the internal auditory canal (IAC). The transcochlear approach can provide a limited exposure to the lateral clivus.³

Infralabyrinthine, infracochlear approach has been utilized for drainage and aeration of petrous apex cholesterol granulomas.¹⁵

MCF approach: Cunningham et al reported on utilizing predominantly a MCF approach (78.6%) in their study for neurotologic pediatric skull base surgery.¹⁶ Tumor removal in 60 cases and debulking in 10 patients. The vast majority of their cases were vestibular schwannomas. MCF approach was hearing sparing for all the subjects.

The MCF approach is useful for tumors of the IAC and the CPA, such as for management of acoustic or vestibular neuromas, lipomas, gliomas, etc. The facial nerve can be exposed and reanastomosed in the CPA and labyrinthine segment via the MCF approach. The MCF approach can also access the basal region of the MCF, as well as anterior surface of the petrous pyramid, the IAC, and the CPA.

There are various scalp incisions utilized to approach the MCF, all involve the scalp just superior to the ear. Burr holes are created in the squamosal portion of the temporal bone and a window is created in the skull and the dura is elevated from the temporal bone and off the petrous pyramid. The IAC can be approached by utilizing landmarks and drilling through the anterior surface of the petrous portion of the temporal bone. The principle lateral landmark is the arcuate eminence of the superior semicircular canal; the anterior landmark is the greater petrosal nerve. The IAC will bisect the angle formed by the superior semicircular canal and the greater petrosal nerve.

Transmastoid approach: This can be useful for approaching the endolymphatic sac, access to the posterior cranial fossa with endoscopy for an evaluation, biopsy, or drainage of abscess, etc. Through a postauricular incision, the mastoid cortex is exposed and opened. A complete mastoidectomy can be performed with the tegmen as the superior extent, the bony ear canal anteriorly, and posteriorly the sigmoid sinus and posterior cranial fossa dura. Trautmann's triangle is a triangular area of the bone bordered by the sigmoid sinus posteriorly, the labyrinthine capsule anteriorly, and the tegmen superiorly. The bone in this area can be drilled medially to expose the posterior cranial fossa dura. Drilling between the posterior semicircular canal and the sigmoid allows for an exposure of the posterior semicircular canal dura. The position of the endolymphatic sac can be isolated. The semicircular canals and facial nerve are identified and preserved.

TLA: The TLA is appropriate for large CPA tumors and can also be used for the reconstruction of the intratemporal facial nerve. The TLA approach is good when there is hearing loss. The TLA provides broad access to the IAC and petrous apex. The TLA is an extension of the transmastoid approach. After performing the complete mastoidectomy and exposing the posterior semicircular canals, the facial nerve is identified in its fallopian canal. The semicircular canals (posterior, then lateral, and superior last) and vestibule are drilled out, going medially to identify the IAC.

Transotic approach: The transotic approach is useful for extensive tumors and lesions of the petrous apex and IAC, such as petrous cholesteatomas or cholesterol granulomas. This approach is a good choice when there is already profound hearing loss and vestibular loss. A mastoidectomy is performed, followed by the removal of the bony EAC and tympanic membrane. The ear canal is oversewn and closed. A complete labyrinthectomy is performed, and much like the TLA the IAC is exposed. The approach proceeds anteriorly to the facial canal, the promontory of the cochlea is drilled, and the cochlea is removed. The extents of the drillout are the jugular bulb and inferior petrosal sinus inferiorly and the genu of the internal carotid artery anteriorly. Medially and anterosuperiorly, the dura of the middle and posterior fossa can be exposed. The IAC is also entered and the petrous apex is approachable. The facial nerve is left in place. Care must be taken with the facial nerve, especially the labyrinthine segment is at risk for injury with this approach. Because the facial nerve is left in place, it limits the access to the anterior CPA.

Transcochlear approach: The transcochlear approach allows for a wide access to the CPA, and for lesions that have an anterior extension such as into the prepontine cistern. It provides perhaps the widest approach to the lateral skull base. It is an extension of the TLA after drilling of the labyrinth, the external ear canal is transected and everted and oversewn as a blind pouch. The bony external ear canal is removed as well as the tympanic membrane. The cochlea is drilled and removed. Drilling continues to remove the entire petrous bone to the clivus. The facial nerve is transposed posteriorly, but is related to a weakened House–Brackmann (HB) III as the best outcome. If hearing needs to be preserved, the transpetrosal approach may be more appropriate.³

Transpetrosal approach: The transpetrosal approach is sometimes called a posterior petrosal approach or retrolabyrinthine approach. This approach is appropriate for lesions centered in the petrous apex. A wide complete mastoidectomy is performed. The mastoidectomy is extended posteriorly to expose the sigmoid sinus and the dura posterior to the sigmoid sinus. The entire sigmoid sinus is retracted posteriorly to expose the posterior fossa dura. It is important that the mastoidectomy include the retrofacial air cells down toward the jugular bulb to allow for exposure of the CPA cistern. The petrous temporal bone is drilled with preservation of the labyrinth and the facial nerve. The mastoid is opened and the labyrinth and facial nerve identified. Retrofacial air cells are opened, sigmoid sinus skeletonized, the sinodural angle is opened, and the exposure extended medially to the superior petrosal sinus. The posterior and superior semicircular canals are skeletonized. There is preservation of hearing and facial nerve function. This approach also results in minimal retraction of the posterior temporal lobe and the anterolateral surface of the cerebellar hemisphere.^{17,18}

Infracochlear approach: This approach is through the external ear canal for access to the petrous apex, and can be used for biopsy of a lesion of the petrous apex or drainage of a cholesterol granuloma. A tympanomeatal flap is

elevated. The inferior portion of the annular ring is drilled to expose the cochlear promontory and the jugular bulb is skeletonized in the hypotympanum. Continue drilling through the triangle of bone between the jugular bulb (posteroinferior), internal carotid artery (anteroinferior), and the basal turn of the cochlea (superior), proceeding medially to the petrous apex.

Dr. Yugo Fisch developed a four-part system of surgical approaches for the lateral skull base and infratemporal fossa. These approaches are useful for lesions of the petrous temporal bone and the lateral skull base, such as glomus tumors, cholesteatomas, dermoid cysts, meningiomas, malignant tumors, etc. Fisch type A approach is useful for working on lesions of the temporal bone centered on the jugular foramen, sigmoid sinus, and the internal carotid artery near the skull base. Fisch type B and C are extensions of the type A approach with additional exposure and access of the infratemporal fossa. Type C approach provides access to the lateral orbit and pterygopalatine fossa (→Fig. 2). The type D approach is centered more on the anterior portions of the infratemporal fossa, lateral orbit, and retromaxillary spaces as well as the nasopharynx.

Fisch Type A approach: For the Fisch type A approach, a large incision is made from the temporal region, postauricularly and extended into the neck. This allows access to the temporal region, the mastoid, and the superior aspect of the carotid sheath. A periosteal flap is created over the mastoid and the cartilaginous EAC is divided and oversewn creating a blind pouch. The auricle is displaced anteriorly. The extra-temporal facial nerve is identified as it exits the stylomastoid foramen. The lateral aspect of the parotid gland is removed and the facial nerve is dissected to the bifurcation and the branches in the parotid are dissected. Going inferiorly into the superior neck, the great vessels (internal and external carotid arteries, internal jugular vein) and associated lower CNs (X, XI, and XII) are identified and dissected. Some temporal bone and lateral skull base malignancies may require a neck dissection to be performed. The inferior aspect

of the incision can be extended to provide access to the lateral neck compartments. A complete mastoidectomy is performed, exposing the sigmoid sinus, vertical segment of the facial nerve, and the osseous labyrinth. The bony EAC and tympanic membrane are resected, continuing medially to resect the ossicular chain and cutting the stapes crura leaving the stapes footplate in place. The mastoid tip is completely drilled out with exposure of the entire vertical segment of the intratemporal facial nerve from the genu to the stylo-mastoid foramen. The chorda tympani is divided and the facial nerve is transposed anteriorly and passed through a newly created foramen in the anterior attic wall. The jugular foramen and the nerves passing through it are exposed and dissected. The internal carotid artery is also exposed and down into the cervical region in the skull base. The sigmoid sinus and internal jugular vein may need to be ligated depending on the lesion to be resected. The inferior petrosal sinus can cause troublesome bleeding as it enters the anteromedial aspect of the jugular bulb.

Fisch Type B approach: Fisch type B extends the Fisch type A approach anteriorly. The temporalis muscle is mobilized and retracted anteriorly. Care is taken to try to preserve the neurovascular supply to the temporalis muscle. The root of the zygomatic arch is exposed, dissecting anteriorly almost to the orbit. With the prior dissection of the facial nerve from the parotid gland, the superior branches of the facial nerve can be identified and care is taken to preserve the facial nerve. The zygomatic arch is divided, one cut just anterior to the TMJ and the other just anterior to the origin of the masseter muscle. The glenoid fossa is entered and the TMJ is disarticulated. The condyle is displaced inferiorly. A heavy retractor (e.g., Fisch intratemporal fossa retractor) is used to retract the mandibular condyle, the zygomatic arch, and the temporalis anteriorly. This provides a wide access to the infratemporal fossa. The zygomatic root and the glenoid fossa are drilled. The greater wing of the sphenoid bone and the inferior temporal bone squamosa are drilled to provide wide access to the infratemporal fossa. The middle meningeal artery may need to be divided. The pterygoid process is the anterior extent of Fisch type B approach. If the lesion is primarily anterior and in the infratemporal fossa, the facial nerve may not need to be skeletonized and transposed. In that case, the bony fallopian canal may be left intact to help protect the nerve.

Fisch Type C approach: Fisch type C is used to gain access to extensive lesions that involve more anterior areas such as the lateral orbit and lateral aspects of the pterygopalatine fossa and nasopharynx. Much like with Fisch type B approach, the temporalis muscle is elevated, but in the Fisch type C approach, the soft tissues are dissected off the zygoma completely to the lateral orbital margin. The zygomatic arch is divided, but the anterior cut is made close to the orbit rather than at the origin of the masseter muscle. The bone cut should be made as anterior as possible to allow for adequate exposure to the anterior aspect of the infratemporal fossa, and into the pterygopalatine fossa, the lateral orbit, and down into the nasopharynx.

Lateral, subtotal, and total petrosectomy: Zanoletti et al defined various petrosectomy approaches which are useful

Fig. 2 Fisch type A, B, and C surgical approaches.

for enblock resection of lesions of the lateral temporal bone and petrous bone.¹⁹ Lateral petrosectomy allows for an enblock resection of the ear canal and a portion of the mastoid as it is an excision of the lateral aspect of the temporal bone. The posterior wall of the bony ear canal is removed as well as the middle ear contents and tympanic membrane. The cartilaginous portion of the ear canal is everted and oversewn as a blind pouch. The labyrinth serves as the medial extent of the resection and the facial nerve is left in place. Subtotal petrosectomy extends the resection medial to the labyrinth and transecting the IAC, thereby sacrificing facial nerve function and hearing. The inferior extent of the resection is the carotid canal and internal jugular vein. The total petrosectomy extends even more medially to include the petrous apex.

Retrosigmoid approach: The retrosigmoid approach is useful for the management of CPA pathology. The approach is via a craniotomy behind the sigmoid sinus and between the angle of the lateral and sigmoid sinuses. The apex of the internal auditory meatus is visible with drilling and removal of the porous portion of the temporal bone. The retrosigmoid approach can be combined with a complete mastoidectomy and drilling retrolabyrinthine to create a combined retrosigmoid/presigmoid approach. If the labyrinth is drilled out, then this becomes a TLA.^{1,3}

Suboccipital approach: The suboccipital approach is useful for resecting large CPA tumors, lesions along the posterior aspect of the petrous pyramid, and for decompression of the lower CNs and surgery in the region of the tentorium cerebelli. For access, the occipital muscles are incised and elevated off the bone. An occipital craniotomy is created medial and posterior to the sigmoid sinus and inferior to the transverse sinus. The dura of the posterior fossa is exposed and incised. The cerebellar hemisphere is elevated medially and superiorly. The IAC is exposed from the posterior aspect. The lower CNs can also be identified and decompressed via this approach. The facial nerve is often at greater risk via this approach as the nerve is typically displaced anterosuperiorly and is often not able to be visualized until much of an acoustic neuroma is debulked.

Endoscopes in Lateral Skull Base Surgery

Endoscopes were described for use in the middle ears in 1967 by Mer et al.²⁰ With recent technological advances in endoscopes, video equipment, and techniques, there has been a recent increased use of endoscopes in the middle ear, mastoid, and lateral skull base. There are very few reports of endoscopic lateral skull base surgery, and the pediatric literature is currently predominantly reporting on the endoscopic management of the middle ear and mastoid cholesteatomas.^{21–23}

Marchioni et al described their experience with exclusive transcanal approach to pediatric middle ear cholesteatoma. They reported the feasibility of this approach for the management of middle ear cholesteatoma and ossicular reconstruction as well. They had a statistically significant larger number of ossicular chain preservation (42%) that with the

microscope control group (10%). They had no statistical significance of residual or recurrent disease results between the microscope controls and the transcanal endoscopic approaches.²¹

In 2016, Hunter et al reported on the use of endoscopes in pediatric cholesteatoma surgery.²² They compared the results of traditional management of cholesteatoma with the microscope versus endoscopic transcanal approach. They also showed similar outcomes between both modalities looking at hearing results, rates of recurrence and recidivism, and complications. Thomassin et al reported that cholesteatoma was noted in 47% of second-look surgeries where the microscope only was used at the primary surgery, versus 6% of cases where the endoscope was used in an adjunctive manner to the microscope.²⁴ Hunter et al noted no difference in disease found on second look, the total endoscopic group having 10.3% of patients with cholesteatoma and the microscope group with 8.5%, not statistically significant difference.²²

There are critics of the use of endoscopes in otologic and lateral skull base surgery citing similar complaints that were made when endoscopes started being used for anterior skull base surgery—arguments such as perceived loss of binocular vision and depth perception, one handed surgery, and obscuring visualization due to soiling of the endoscope. However, endoscopes have advantages such as a wider field of view, angled scopes can provide visualization “around a corner,” as well as better illumination and magnification at the target site. Therefore, the endoscopes could be utilized with the goal of reducing the need for wide external incisions and wide exposures to access pathology, and also improving visualization in hidden areas while sparing structures.

In 2004, Mattox reported on a series of patients with petrous apex lesions (four cholesterol granulomas and one petrous cholesteatoma) who were treated with a combined microscope and endoscopic techniques. Using traditional infracochlear and/or retrofacial approaches, endoscopic examination and debridement of the interior of the petrous apex lesions were performed. Mattox reported that with the use of the endoscopes, residual disease and cyst septae were found where it had not been evident with the operating microscope alone.²⁵

Presutti et al described the use of endoscopes for the management of inner ear and lateral skull base pathologies.²⁶ They describe a transcanal exclusive endoscopic approach to the lateral skull base. With this approach, they were able to manage lesions involving the petrous apex, IAC fundus, and lesions with extensions intracochlear, intravestibular, and pericarotid. Depending upon intracochlear or intravestibular extension, a transcochlear or transvestibular approach can be used to gain entry to the IAC. If a lesion involved the mastoid, an exclusive transcanal endoscopic approach is not feasible. After performing the traditional exposures with a microscope, the endoscopes were then utilized depending upon the extent of the pathology.

In 2014, Presutti et al described various endoscopic-assisted surgery (EAS) approaches using angled rigid endoscopes and curved instruments and burs to dissect around structures and to be able to resect lesions, such as transotic EAS,

infralabyrinthine EAS, and suprameatal translabyrinthine EAS. So for the transotic EAS, though the labyrinth and cochlea may be drilled out, the facial nerve may not need to be disturbed from its fallopian canal with use of the endoscopes. If a lesion extends inferior to the labyrinth and there is good preoperative hearing, an infralabyrinthine EAS procedure may be considered—after identifying the labyrinth, the retrofacial air cells are drilled to allow room for optics and instruments. If there is a lesion inferior to the cochlea, the infracochlear EAS procedure can be used. After elevating a tympanomeatal flap, an endoscope can be used to remove the inferior bony annulus to allow access to the hypotympanum. Using the round window and promontory to help delineate the cochlea (superior extent of exposure), then the carotid (anteriorly) and jugular (posteriorly) are identified. Drilling the bone between the carotid and jugular under endoscopic visualization and using curved burs allowed for access to the inferior portions of the petrous apex. Hearing and facial nerve function can be preserved with this approach.²⁷

In 2016, Kempfle et al reported on feasibility of an endoscopic transcanal retrocochlear approach to the IAC which was cochlea sparing as a cadaveric study. They described a transcanal infracochlear approach to the IAC. They used rigid endoscopes, curved instruments and burs, and stereotactic navigation to perform a transcanal hypotympanotomy (similar to the approach for a traditional infracochlear approach to the petrous apex) and a subcochlear tunnel to access the petrous apex and IAC. The extent of bone removal is the vertical segment of the facial nerve posteriorly, the jugular bulb inferiorly, the petrous carotid anteriorly, and the cochlea superiorly. Navigation was used to help delineate the basal turn of the cochlea and the trajectory to the IAC.²⁸ This approach has been used successfully on patients by the senior author of the paper.²⁹

Intraoperative Considerations

Monitoring

Much as possible in adult patients, CN monitoring is also possible in infants and children. CN VII may be monitored as would be done in adults. For CNs IX, X, XI, and XII, some variations in electrode type may be necessary. Can consider the use of hookwire electrodes in cases where adult-sized electrodes may be too large for the pediatric patient. For monitoring of CN X in children, there are commercially available monitoring endotracheal tubes as small as 5.0 (e.g., Medtronic Trivantage monitoring endotracheal tubes) or there are adhesive monopolar electrodes (e.g., Stryker C2 Nerve monitor system) that can be used. Hookwire electrodes are also able to be placed in the vocalis muscle (via microdirect laryngoscopy) to monitor CN X in younger children with smaller endotracheal tube requirements.³⁰

Blood Loss

In infants and small children, care must be taken to monitor blood loss. Loss of more than ~10% of blood volume may cause significant hemodynamic instability. This will translate to only ~80 mL of blood loss for a 10-kg infant.

Intraoperative Imaging/Computer-Assisted Image Guidance

We have used image-guidance at our institution for some lateral skull base approaches. Since the lateral parts of the skull lack defined anatomical points and surface variation such as found on the face used for registering IGS for anterior skull base procedures, often fiducials are used to give defined points to help with the accuracy of the registration of the scans. Some of the challenges in children are that they often need sedation or general anesthesia to obtain fine-cut high-resolution imaging, and they will often pick and remove fiducials when they are awake. Because of this, imaging under general anesthesia may need to be coordinated with transporting patients to the operating room for surgery. The use of intraoperative imaging can help reduce the need for preoperative imaging or transportation of an anesthetized child. We use the O-arm system with StealthStation IGS (Medtronic Navigation Inc, Louisville, Colorado, United States) to obtain real-time images which are autoregistered. This provides high accuracy image-guidance and real-time imaging while saving time and reducing complexity of obtaining and registering the images. The use of the O-arm does not add significant operating room time compared with the typical registering process needed for using IGS—images are able to be obtained with the O-arm and transferred to the StealthStation in a few minutes. Another advantage of this system is the ability to obtain another image at the conclusion of the procedure to check on the extent and completeness of the surgery.

Complications

Various studies report significantly lower complication rates compared with adults. A few studies reported no cases of CSF leaks, intracranial infections, or pneumocephalus.^{1,5,25} Some reported complications include wound infection, wound breakdown, dysphagia, meningitis, CSF leak, brain edema, hemiparesis, and abdominal hematoma (from harvesting of fat graft).^{5,10,16}

In Grinblat et al's series, patients with subtotal petrosectomy had preservation of facial nerve function. With transotic approach, 11 of 13 cases preserved the preoperative HB I and II. Five had postoperative HB III, three of whom had actually improved from HB VI after sural nerve grafting. With TLAs in Grinblat et al's series, 8 of 11 case preserved normal facial nerve function and 3 cases resulted in a HB II, HB III, and a nerve sacrifice. For infratemporal fossa approaches, the average preoperative HB score was 1.6, and had a postoperative average score of 2.6.⁵

If there is facial nerve weakness or paralysis, the eyelids in children often have sufficient tone to provide some closure of the eyelid. This may give an erroneous impression of facial nerve function. Care must be taken to minimize the risk of corneal abrasion or exposure keratitis.

CSF leaks have been noted in adult lateral skull base surgeries, but in Grinblat et al's series, there were no CSF leaks reported. Also, no major complications were reported. Minor complications included abdominal hematoma,

postauricular swelling, and postauricular fistula. Cunningham et al noted CSF leaks in seven cases (6%)—five translabrynthine and two middle fossa approaches.¹⁶

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