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Lateral skull base surgery in a pediatric population: A 25-year experience in a referral skull base center

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ABSTRACT

Objective
To analyze the pathology and surgical outcomes of lateral skull base (LSB) procedures in a pediatric population.
Study design
Retrospective case review in a referral skull base center.
Methods
Charts of pediatric patients who underwent defined LSB procedures from 1983 to 2015 for various pathologies were evaluated at our center. A systematic review of literature was performed and our results were compared with the literature.
Results
63 patients presented with 65 diseased ears. The mean age was 13 years. 29 (44.6%) presented with hearing loss and 28 (44.4%) and chronic otorrhea. The most common pathology was petrous bone cholesteatoma (27, 42.5%) followed by vestibular schwannoma (10, 15.8%). Subtotal petrosectomy (24, 35.8%) was the most common surgical procedure followed by, transotic (18, 26.8%). The facial nerve function was preserved in 45 (67.1%) and the hearing in 28 (41.7%) cases respectively. No major complications, including mortality was encountered in our series.
Conclusion
In rare and extensive pathologies involving the skull base in a pediatric population, the surgeon is posed with the dilemma of trying to achieve facial and hearing preservation while dealing with total tumor clearance. Mastery over LSB procedures can ensure complete disease clearance with optimal functional outcomes.
Level of evidence
2b.

1. Introduction

Pathology that involves the deep parts of the LSB like the cochlea-vestibular system, facial nerve (FN), internal auditory canal and jugular bulb not only causes functional disturbances that can be devastating in children but also makes extirpation of such tumors a challenging proposition. Considering the early age of the patient, the treating practitioner will always be posed with the dilemma of whether to achieve functional preservation (hearing and FN function) or disease clearance. Fortunately, over the last few decades due to rapid advances in neuroradiology and neuroanesthesia, development of rational surgical approaches and better instrumentation, the objective of LSB surgery has moved from solely being focused on tumor removal to also preservation of cranial nerve functions. Children with LSB pathology have benefitted most from this development because in them, any functional deficit at that age and which remains over a very long period of their life, has serious social and psychological consequences.

Pediatric LSB surgery pathology and procedures are relatively rare and there are very few series dealing with this subject [1–8]. We present one of the largest series of pediatric LSB surgery published in English literature and discuss the special considerations in dealing with children with pathologies of the LSB.

2. Material and methods

Of the 4523 skull base surgeries performed from January 1991 to August 2015, charts of all patients less than 18 years of age undergoing LSB surgery in our center, were reviewed. The inclusion criterion was surgical treatment of extensive pathology that extended beyond the confines of the middle ear cleft and involved the petrous bone, the jugular bulb or destroyed large areas of the middle and posterior dural plates, that required at least a Subtotal Petrosectomy (STP) or any other procedure that involved a more extensive dissection. Exclusion criteria were: 1) Surgery for routine middle ear conditions, 2) LSB pathology in children that were not operated upon, 3) Malignant tumors (due to necessity of adjuvant treatment that could alter the outcome of the study), 4) Patients with incomplete case records, follow-up of less than one year or those lost for follow up.

All patients underwent complete preoperative otoneurologic evaluation followed by audiometric exam. FN function was graded preop-

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eratively according to the House-Brackmann (HB) grading system. Audiometric studies included four-frequency (500, 1000, 2000 and 4000 Hz) pure tone average (PTA) for bone-conduction (BC), air-conduction (AC) and speech discrimination scores (SDS). The modified Sanna classification of hearing scores were used for documenting and analyzing audiological data [9,10]. A high resolution computed tomography scan of the temporal bone and magnetic resonance imaging were obtained as a part of diagnostic battery.

Petrous bone cholesteatomas (PBC), were classified according to the Sanna classification [11,12]. In Vestibular schwannomas (VS), the tumor was graded according to the Tokyo consensus meeting [10]. The surgical procedures included STP, transotic approaches (TOA), translabyrinthine approaches (TLA), transcochlear approaches (TCA), infratemporal fossa approaches (ITFA), transmastoid-infralabyrinthine approaches (TM-IL). In some cases, cochlear and auditory brainstem implants were performed along with the abovementioned procedures. All the procedures have been described in detail elsewhere [13–15].

Intraoperatively, operating time and complications were recorded. Postoperatively, average length of hospital stay and complications were recorded. FN and hearing status at the end of one year follow-up were obtained from the charts.

2.1. Review of the literature

A review of the literature was done using a PubMed Search using relevant search words. The demographic data, pathology, performed surgical procedures, duration of surgery, FN function and hearing status both pre- and postoperatively, other cranial nerve status, complication rates, and follow-up time were tabulated and compared with our data.

This study was approved by the Institutional Review Board of the hospital for ethical research.

3. Results

A total of 63 pediatric patients with 65 pathological ears were included in the study. 37 patients were males and 26 were females. The age ranged from 18 months to 324 months (mean, 156 months). All patients in this series were operated. The incidence of pathology in the LSB in children was 1.4% of all the 4523 patients who presented to us with such pathology at our center. The mean duration of follow-up was 43.6 months (range:12–125). Patient characteristics are shown in Table 1.

3.1. Symptomatology

The mean duration of symptoms at the time of presentation in our center was 26.5 months. 32.3% of the patients were referred to our center for surgical intervention after a prior surgery elsewhere (Table 1). On presentation, the most common symptoms were hearing loss (44.6%) and chronic otorrhea (43.1%). Preoperatively, FN was normal (HB I) in 53 (81.5%), and HB IV and above in nine (13.8%) cases. Clinically, 23 ears (35.4%) were totally deaf. The FN function and hearing status are shown in Table 3.

3.2. Pathology

25 (38.5%) cases were of tumoral pathology, mostly VS and FNTs. 31 cases were non tumoral pathology most common being PBCs seen in 26 (40%). Inflammatory and infective pathology was seen in four (6.2%) cases, the rest of them being surgeries for hearing related pathology (Table 2). Among the VS, five (50%) of the tumors were sporadic and the rest were NF II. The VS were extrameatal in all but one (90%) case. All NF II cases had bilateral and larger tumors (mean 3.6 cm) compared to sporadic type (mean 2.9 cm). Four (66.7%) of the FNTs were schwannomas and all of them involved multiple segments. Most of the PBCs, 14 (53.9%), were of the infralabyrinthine type [16].

Table 1
Patient characteristics and symptoms of the study population.

Population characteristics	
Patients	63
Procedures	65
Mean age	13.0 (range 1.5–18)
Males, females	37, 26
Left side, right side	38, 29
Mean symptom duration (range)	25.6 days (range 2–360)
Mean duration of follow-up	42.8 months (range 12–125)
Symptoms	
Hearing loss	29 (44.6%)
Chronic otorrhea	28 (43.1%)
Dizziness/Vertigo	17 (26.2%)
Tinnitus	9 (13.8%)
Facial nerve palsy	7 (10.8%)
Trigeminal palsy	3 (4.6%)
Lower cranial nerve palsy	6 (9.2%)
Headache	4 (6.2%)
Recurrent meningitis	2 (3.1%)
Treatment details	
Patients previously operated elsewhere	21 (32.3%)
Revision surgeries in this series	2 (3.1%)

3.3. Surgical approaches

STP was the most common approach performed in 23 (35.4%), followed by TOA and TLA in 18 (27.7%) and 13 (20.0%) cases, respectively. The surgical approaches are listed in Table 2.

STPs were used commonly in PBCs, in 17 (65.4%) cases. TLA was used mainly for VS in eight (80%) cases. Three cases of petrous apex cholesterol granulomas were treated by the TM-IL approach. In one of them, the procedure was abandoned due to a prominent sigmoid sinus and a high jugular bulb. An infracochlear approach was then attempted in the same sitting but this was also abandoned due to unfavorable anatomy. Two cases of class C1 tympanojugular paragangliomas (TJPs) were treated by the ITFA type A with complete tumor removal. In another case of extensive class C3Di2 a labyrinthectomy was added to the ITFA type A. The patient had a residual tumor and after five months a TCA was performed with a sural nerve grafting despite incomplete tumor resection.

In 55 out of 60 procedures (91.7%) gross total removal was achieved. Of the incomplete resections, one was a case a 4 × 3.7 cm NF II VS where the surgery had to be stopped due to bleeding. Other partial resections included a C3Di2 TJP, chordoma (involving the clivus, infratemporal fossa, orbit and the parapharyngeal space), posterior fossa meningioma (involving the parapharyngeal space and the internal carotid artery). Another case of unsuccessful drainage of petrous apex cholesterol granuloma is asymptomatic and stable on follow-up for the last 28 months.

3.4. Analysis of hearing outcomes according to pathology

Among the 26 PBC, four were deaf preoperatively. In the remaining 22 cases the mean preoperative PTA-AC, PTA-BC, ABG and SDS were, 71.3 dB (±14.1), 39.7 dB (±9.8), 31.6 dB (±11.5) and 87.5% (±10.9), respectively. Postoperatively, seven (31.8%) out of 22 cases were deaf. In the remaining 15 cases, the mean PTA-AC, PTA-

Table 2
Characteristics of various pathologic conditions and performed surgical approaches.

Pathology		No (%)	Surgical approaches
Tumors (n = 25)			
Vestibular schwannoma	Sporadic	5 (7.7%)	TLA (3), TLA (5), TOA (2)
	NF II	5 (7.7%)	
Facial nerve tumors	Schwannoma	4 (6.2%)	TO (1), TO-TPA (1), TC (1), STP (1)
	Neurofibroma	1 (1.5%)	TM-ILA
	Involved by pleomorphic adenoma	1 (1.5%)	STP-TPA
Tympanojugular Paraganglioma		3 (4.6%)	ITF- A (1), ITF- A (1), ITF- A + SN graft (1)
Lipoma of cochlear nerve		1 (1.5%)	TLA
Chordoma		1 (1.5%)	ITF- D + OZ + TC
Osteoblastoma		1 (1.5%)	STP
Meningioma meningoteliale		1 (1.5%)	TO-TC
Endolymphatic sac tumor		1 (1.5%)	TLA
Juvenile nasopharyngeal angiofibroma (IIIB)		1 (1.5%)	IFT-D + OZ + SFC
Non-tumoral pathology (n = 31)			
Petrous Bone Cholesteatoma	IL	14 (21.5%)	STP (17), TLA (1), TOA (8)
	SL	5 (7.7%)	
	Massive	7 (10.8%)	
Middle ear and mastoid cholesteatoma		1 (1.5%)	STP + CI
Cholesterol granuloma		3 (4.6%)	TM-ILA (3)
Eosinophilic granuloma		1 (1.5%)	TOA
Inflammatory and infectious pathology (n = 4)			
Tuberculosis of temporal bone		1 (1.5%)	TOA
Inflammatory pseudotumor		1 (1.5%)	TOA
Meningoencephalic herniation		1 (1.5%)	STP
Granulation tissue		1 (1.5%)	TM-ILA
Hearing related pathology (n = 5)			
Post-meningitis deafness		1 (1.5%)	TLA + ABI
Bilateral cochlear aplasia		1 (1.5%)	TLA + ABI
Congenital deafness with COM		1 (1.5%)	STP + CI
Mondini deformity, recurrent meningitis		1 (1.5%)	TOA
Traumatic petro-occipital fracture		1 (1.5%)	TOA
Total		**65**	
Gross total disease removal[χ]		55 (91.7%)	
Disease free survival at the end of 3 years		63 (96.9%)	
Duration of surgery (hours) (mean, range)		4.2 (1.5–12)	
Post-operative period (days) (mean-range)		5.5 (4–14)	

[^]According to modified Sanna classification,[9,10] [χ]Excluding the hearing related pathology, STP: Subtotal Petrosectomy, TLA: Translabyrinthine Approach, TOA: Transotic Approach, TPA: Transparotid Approach, TM: Transmastoid, TC: Transcervical, ILA: Infralabyrinthine Approach, OZ: Orbitozygomatic, SFC: Subfrontal Craniotomy, SN: Sural Nerve, NF: Neurofibromatosis, ABI: Auditory Brainstem Implant, COM: Chronic Otitis Media, CI: Cochlear Implant, ITF-A: infratemporal fossa approach type A, ITF-D: infratemporal fossa approach type D.

BC, ABG and SDS was 69.5 dB (±13.5), 34.5 dB (±7.5), 35 dB (±10.5) and 91% (±8.9), respectively. One case with sporadic VS was deaf on VS ear and all NF-2 cases were deaf bilaterally preoperatively. The mean PTA-AC, PTA-BC, ABG and SDS in the remaining four sporadic VS cases were 51.25 dB (±26.1), 40.6 dB (±18.1), 10.6 dB (±11.2) and 73.75% (±15.6), respectively.

3.5. Analysis of hearing outcomes according to surgical approach

Preoperatively, 22 (33.8%) of the cases were deaf. Of the remaining 43, the mean PTA-AC, PTA-BC, ABG and SDS were 56.3 dB (±26.4), 33.6 dB (±16.4), 22.7 dB (±16.5) and 88.9% (±13.2), respectively. Postoperatively, 38 (58.5%) were deaf. The postoperative mean PTA-AC, PTA-BC, ABG and SDS were 56.3 dB (±32.4), 26.7 dB (±16.0), 29.5 dB (±18.0) and 94.6% (±46.1), respectively (Table 3).

In the STP group, five (21.7%) cases were deaf preoperatively. In the remaining 18, the mean preoperative PTA-AC, PTA-BC, ABG and SDS was 61.5 dB (±20.7), 35.3 dB (±9.7), 26.2 dB (±14.1) and 92.7% (±7.6), respectively. The mean postoperative PTA-AC, PTA-BC, ABG and SDS was 67.9 dB (±14.2), 32.9 dB (±9.6), 35 dB (±9.7) and 91.9% (±8.7), respectively. In the TOA group, nine (50%) cases were deaf preoperatively. In the remaining nine, the mean preoperative PTA-AC, PTA-BC, ABG and SDS was 71.6 dB (±23.3), 41.4 dB (±9.7), 30.3 dB (±15.2) and 82.7% (±12.9), respectively. Postoperatively, all cases were deaf. In the TLA group, seven (53.8%) of cases were deaf preoperatively. In the remaining six, the mean preoperative PTA-AC, PTA-BC, ABG and SDS were 60.8 dB (±26.7), 43.3 dB (±15.3), 17.5 dB (±15.5) and 70% (±13.8), respectively. Postoperatively, all cases were deaf (see Table 4).

3.6. Analysis of FN outcomes according to pathology

Among PBC, the mean preoperative and postoperative HB grading was 1.2 and 1.2 respectively. Among VS, the mean preoperative and postoperative HB grading was 1.5 and 2.9 respectively. Among FNT, the mean preoperative and postoperative HB grading was 4.2 and 3.3 respectively. Among the hearing related pathologies, the mean preoperative and postoperative HB grading were one and one respectively.

3.7. Analysis of FN outcomes according to surgical approach

Among STP, all cases retained their normal FN function postoperatively. Among TOA, the mean preoperative and postoperative HB grading was 2.1 and 2.1 respectively. Among TOA, 13 (72.2%) cases presented with HB I and II; one presented with HB III and four cases presented with HB VI. Postoperatively, 11 of 13 cases preserved HB I and HB II. Five had HB III of whom three improved from HB VI after a sural nerve graft in the same setting. In the remaining two cases, one NF II case retained the same grade. The other case was that of a huge sporadic VS, wherein the FN was pressed and convoluted by the tumor and a decision was made to sacrifice the nerve with consecutive treatment by VII-XII anastomosis. Of the two with FN schwannomas treated with TOA and a sural nerve graft; one improved from HB VI to IV and the other remained HB IV. In a case of

Table 3
Preoperative and post-operative facial nerve and hearing status at the end of one year of follow-up (63 patients).

Status	Pre-operative; no (%)	Post-operative; no (%)
Facial nerve status		
HB I	52 (80.0%)	44 (67.7%)
HB II	2 (3.1%)	3 (4.6%)
HB III	3 (4.6%)	11 (16.9%)
HB IV	4 (6.2%)	4 (6.2%)
HB V	0 (0%)	0 (0%)
HB VI	4 (6.2%)	3 (4.6%)
Hearing status		
Total deafness	22 (33.8%)	38 (58.5%)
Mean PTA AC	56.3 dB ± 26.4[a]	56.3 dB ± 32.4[b]
Mean PTA BC	33.6 dB ± 16.4[a]	26.7 dB ± 16.0[b]
Mean ABG	22.7 dB ± 16.5[a]	29.5 dB ± 18.0[b]
Speech Discrimination Score	88.9% ± 13.2[a]	94.6% ± 46.1[b]

PTA: Pure Tone Audiogram.

[a] Out of the 43 cases with hearing, all had measurable results.

[b] Only 27 cases with hearing preservation procedures included.

Table 4
Review of literature for LSB surgery in a pediatric population.

Author, year	No of patients No of LSBS	Mean age (Range)	Duration of symptoms (months)	Most common presenting symptoms (%)	Pathologies (%)	Procedures (%)	Mean surgical time (hrs)	FN status (no, %)						Hearing status		Post-op cranial nerve palsies except VII, VIII (%)	Complications (%)	Follow-up (months)
								HB I-II		HB III		HB IV-VI						
								Pre-op	Post-op	Pre-op	Post-op	Pre-op	Post-op	Pre-op	Post-op			
Jackson et al. [1], 1996	15, 15	13.5 (1–21)	NA	Tinnitus (33), HL (26), Otalgia (13) Vertigo (6), FNP (6)	TJP (33), malignant TJP, (13), VPG (7), VS (20), malignant VS (7), meningioma (7), lipoma (7), AC (7)	ITFA-A (73), ITFA + PCFA (7), TM (7), TM + MCFA (13)	NA	NA	9 (60)	NA	NA	NA	NA	NA	NA	LCN (40)	Wound breakdown (13.3), Dysphagia (6.6), Meningitis (6.6), CSF leak (6.6), Stroke (13.3), Death (6.6)	82.8
Cunningham et al. [5], 2005	89, 115	13.8 (7–18)	28	HL (60), Tinnitus (40), Vertigo (19), Headache (19), Paresthesia (6)	VS (86), FNT (7), meningioma (3), Others (3)	MCFA (63), TLA (35), TCA (2), ITFA-A (2), RSA (0.8)	3.5	105 (91.3)	85 (79.4) (n = 107)	6 (5.2)	8 (7.5)	4 (3.5)	14 (13.1)	(A + B)[a] 62 (88.6%) (C + D)[a] 8 (11.4%)	(A + B)[a] 40 (57.2%) (C + D)[a] 30 (42.8%) HPS = 70	LCN (4.3), CN VI (0.8), CN V (0.8)	Wound infection (0.8), CSF leak (6), Meningitis (0.8)	>12
Mazzoni et al. [6], 2007	10, 10	16 (12–18)	2–60	HL (100), Tinnitus (40), Vertigo (30)	VS (100)	TLA (50), RSA (50)	NA	NA	8 (80)	NA	1 (10)	NA	1 (10)	(D + E + F)[b] 7 (100%) n = 7	(C)[b] 1 (20%) (F)[b] 4 (80%)	–	CSF leak (10), Meningitis (10), Brain edema (10), Hemiparesis (10)	3–22
Slattery III et al. [7], 2007	35, 47	12.6 (8–17)	NA	NA	VS (100)	MCFA (100)	NA	4 (97.8)	38 (81)	1 (2.1)	5 (10.6)	–	4 (8.5)	(A + B)[a] 40 (89%) (C)[a] 5 (11%) n = 45	(A + B)[a] 23 (51%) 22 (48.9%) HPS = 45	NA	NA	>12
Walcott et al. [8], 2009	7, 7	15.1 (9–18)	31.2	HL (86), Gait instability (86), Tinnitus (43), Paresthesia (29)	VS (100)	TLA (14), RSA (14), combined TLA + RSA (72)	12.5	5 (71)	7 (100)	1 (14)	–	1 (14)	–	NA	NA	LCN (14.3)	Psychosis (28.6)	75.6
Present study	63, 65	13.2 (1.5–18)	26.5	HL (44.6), Otorrhea (43.1), Vertigo (26.2), Tinnitus (13.8), FNP (10.8), Paresthesia (4.6), LCNP (9.2), Headache (6.2), Meningitis (3.1)	PBC (40), VS (15), FNT (9), TJP (5), PACG (5), Other (26)	STP (35), TOA (27.7), TLA (20), ITFA-A (4.6), ITFA-D (3), ILA (4.6), TM-ILA (3.1), TCA (1.5)	4	54 (83.1)	47 (72.3)	3 (4.6)	11 (16.9)	8 (12.4)	4 (6.2)	(C)[b] 43 (66.2%) (F)[b] 22 (33.8%)	(B)[b] 27 (42%) (F)[b] 38 (58.5%) HPS = 27	–	Abdominal hematoma (3), Wound infection (1.5)	43.2

NA: not available, LSBS: lateral skull base surgery, HL: hearing loss, FNP: facial nerve palsy, TJP: tympanojugular paraganglioma, VPG: vagal paraganglioma, VS: vestibular schwannoma, AC: adenocarcinoma; FNT- facial nerve tumor; PBC-petrous bone cholesteatoma; PACG- petrous apex cholesterol granuloma, PCFA: posterior cranial fossa approach, TM: transmastoid, MCFA: middle cranial fossa approach, TLA: translabyrinthine approach, TCA: transcochlear approach, RSA: retrosigmoid approach, STP: subtotal petrosectomy, TOA: transotic approach, ITFA-A: infratemporal fossa approach type A, ITFA-D: infratemporal fossa approach type D, ILA: infralabyrinthine approach, HB: House Brackmann grading system, PTA: pure tone audiometry, SDS: speech discrimination score, HPS: hearing preservation surgery.

[a] According AAO-HNS (American Academy of Otolaryngology–Head and Neck Surgery hearing classification).

[b] According to Modified Sanna classification, LCN: lower cranial nerve, CN: cranial nerve, CSF: cerebrospinal fluid.

meningioma, the tumor was found to be compressing the nerve and in this case, the preoperative HB I deteriorated to HB IV.

Among TLAs, the mean preoperative and postoperative FN status was HB grade 1.2 and 2.2 respectively. Among 11 cases, (78.6%) presented with normal FN function. Postoperatively, eight out of 11 (72.7%) preserved normal function. Of the remaining three, the tumor was found to be stretching a very anteriorly or medially positioned FN, leading to a postoperative HB III and HB II respectively. One case with NF II suffered from bilateral VS had bilateral FN palsy. Intraoperatively, on both sides the tumor tissue intricate FN without a respond to 0.1 mA electric stimulus, leading to its sacrifice. Among ITFAs, the mean preoperative and postoperative HB grading was 1.6 and 2.6 respectively. Of the five cases where other approaches were employed, four preserved normal FN function, while one case improved from HB IV to III (in a case of neurofibroma of the FN).

3.8. Long term outcomes and complications

There were no major complications in our series. There were no incidences of CSF leakages. The incidence of minor complications included abdominal hematoma, postauricular swelling, postauricular fistula with infected fat tissue. They were managed by additional local interventions. The patient with massive C3Di2 TJP and a patient with chordoma died of the disease nine and three years respectively after surgery.

4. Discussion

LSB surgery in a pediatric population is a challenging proposition because due consideration must be given to hearing and FN preservation in the decision making process. Treatment challenges become even greater when adopting these procedures to population with longer life expectancies [1]. Most series regarding pathology involving the LSB in the existing literature deal with adult population with very little data regarding the same in children. A review of literature shows that only seven series accounting to up to 156 cases have been presented in the existing peer-reviewed literature [1,2,4–8]. Our series with 65 cases adds substantially to existing literature.

4.1. Anatomical considerations in pediatric population

It has been established that most of the growth in the skull base takes place in the first five years of life and continues for at least 10 years after birth [17,18]. Hence adult surgical approaches to the cranial base require modification when implemented in a child [18]. The smaller size and thinness of the bones of the cranial base requires lesser drilling. It is also well known that the mastoid process is absent at birth and is not fully developed until three years of age. This renders the FN which is more superficial and inferior vulnerable to surgery. The anatomy of the inner ear, though, once formed, changes little in structure or growth throughout life into adulthood. The effect of extensive bony removal and of ossification centers and unfused suture lines may have an unfavorable effect on the growth of the surrounding structures and it is necessary that further studies are focused on this.

4.2. Clinical presentation in pediatric population

As seen with other series [5,8], unfortunately, the duration between onset of symptoms and intervention in children in our series was quite high. This is due to multiple factors like inability of children to express their symptoms adequately, misdiagnosis due the rarity of pathology of this nature or due to reluctance of performing a radical intervention by the treating practitioner due to the tender age of the patients. This is reflected by the fact that prior to surgery at our center, total deafness was seen in 35.4% of cases and a high grade of hearing deterioration in the rest (Table 3). After a diagnosis, 32.3% of the cases were treated elsewhere by less extensive procedures before they were referred to our center. Fortunately, the FN function fared better with 83.1% of the cases presenting with a HB grade I or II. Lower cranial nerve dysfunction was noted in 9.2% of cases.

4.3. Pathological considerations in pediatric population

A variety of pathologies afflict young children that require a complex skull base procedure as seen in Table 2. As with most other series published in literature, LSB surgery in children was directed at primarily extirpating tumoral pathology (38.5%) in our series. TJPs were not very commonly seen in our series in children as against the series by Jackson CG et al. [1], in whose series 53.3% of the tumors were TJPs. However, a significant number of cases in our series also involved LSB surgery for non tumoral pathology (PBCs), inflammatory/infectious pathologies and hearing related pathologies that have not been discussed in other series.

4.4. Surgical considerations in pediatric population

The long life span ahead of the patient makes conservative approaches like wait-and-scan and radiotherapy irrelevant in the treatment of majority of benign lesions of the skull base. Petrous apex cholesterol granuloma could be an exception to this as it can be followed up with a wait-and-scan policy. Radiotherapy is ill advised due to the concerns relating to malignant transformation and interference with cranial growth centers that could occur over a long life time [1]. Adult LSB procedures can be safely applied in the pediatric population with minimal comorbidities [18]. While the surgeon is poised with the dilemma of trying to achieve facial and hearing preservation while dealing with total tumor clearance in an extensive disease, it is important to lean towards total disease clearance. Hearing preservation surgeries, though not used in this series due to the advance nature of pathologies that we encountered, must be however considered wherever feasible. Mastery over LSB surgery can enable complete disease clearance with optimal functional outcomes. In our series, the LSB surgery enabled complete tumor removal in 91.7% of the cases.

4.5. Functional preservation

With the advancement of LSB surgery over the last few decades, hearing preservation and FN outcomes have improved tremendously. While many series did not analyze hearing results adequately, in the ones that did hearing preservation rates varied according to the pathology and surgical approach. In case of hearing preservation surgeries for VS, the actual rate of residual hearing after surgery ranged between 20% and 71.4% [5–7]. In our series, we did not perform any hearing preservation surgeries for VS as the tumors were not indicated for such surgeries. However, in all other surgeries, hearing was preserved in 41.5% of the cases. While the mean postoperative AC and ABG worsened, there was improvement in the mean BC and the SDS.

The FN function was well preserved both pre- and postoperatively in most series. Postoperatively, the review of literature showed that FN was HB grade I and II ranged from 79.4% to 100% [1,5,6,8]. In our series, FN HB grade I and II was seen in 72.3%. When the FN tumors were excluded, our FN preservation rates improved to 79.6%.

In the literature, postoperative lower cranial nerve deficits ranged from 4.3% to 40% [1,5–8], however the highest deficit was associated with TJPs which is quite expected considering their close proximity to the nerves [1]. Jackson et al. [1], observes that complication rates are higher along with worser incidence of lower cranial nerve preservation in children compared with adults. On the contrary, in our series, we did not have any case with postoperative lower cranial nerve deficits. This could be attributed to the fact that the series by Jackson et al. dealt predominantly with paragangliomas unlike ours that had a heterogenous pathology.

4.6. Complications

Major postoperative complications included mortality, cerebrovascular accidents, meningitis and CSF leaks and ranged from 6.9% to 43%. Minor complications included wound breakdown, infection and abdominal hematoma. The incidence of complications ranged between 0.8 and 13.3%. At the last follow-up, 62 of the 65 patients survived and only one of the 55 cases with gross disease clearance developed a recurrence. The sole case of recurrence was that in a case of supralabyrinthine PBC that developed a recurrence after five years and subsequently underwent a STP. The three-year disease free survival that could be calculated in 27 cases, was 97.6%. Teo et al. [2], in their series of predominantly tumoral etiology, reported a 2 year tumor free survival of 81%.

LSB surgery is a high risk surgery. However, the quality of life after such surgeries has improved due to developments in subspecialties like phonosurgery, FN reanimation surgeries and hearing implantology. Also, going by literature, surgery on skull bones did not appear to inflict any long term morphological sequelae in children [2].

5. Conclusion

LSB surgery in a pediatric population is a surgical challenge. However, results from experienced centers that routinely perform such procedures have yielded good results in terms of disease clearance and functional outcomes. While on one hand, advances in radiology have helped in earlier diagnosis, the silent nature of pathology in the skull base and the inability of children to express themselves, must lead the treating practitioner to have a high index of suspicion towards such rare pathologies.

Disclosures

The authors have no conflicts of interests to disclose. The authors also do not have any financial disclosures.

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