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Facial Nerve Preservation and Surgical Outcomes of Retrosigmoid Approach to Large Vestibular Schwannoma- an Eight-year Single Institution Experience

Objective: To evaluate the result of microsurgical excison of vestibular schwannoma by retrosigmoid approach and to correlate the facial nerve outcome with the tumor size.

Methods: Retrospective analysis of 84 patients with cerebello-pontine angle lesions (57 vestibular schwannomas) operated at National Institute of Neurological and Allied Sciences, Nepal, from Baisakh 2066 to Chaitra 2073 (eight years). Facial nerve outcomes as per the House and Brackmann grading at six months follow-up were recorded and corelated with tumor size.

Result: The mean tumor size was 4.32 ± 1.23 cms and 48 (84.2%) of the patients had tumor size more than 3cms. Patients presented late with papilloedema documented in 42.1% and preoperative ventriculoperitoneal shunting required in 31.57% patients. Of the patients whose facial nerve functions could be followed up, all patients with tumors < 3cm diameter had good House and Brackmann facial nerve outcome (grade I to III). Larger tumors had poorer facial nerve outcomes; however statistical significance could not be reached. (Fischer Exact test, p-value: 0.077). There were two mortalities (3.5%), one following cavity rebleed and one following malignant brain swelling.

Conclusion: Retrosigmoid approach is a versatile surgical corridor to excise large vestibular schwannomas with minimal complications and larger tumors have poorer facial nerve outcomes.

Keywords: Facial nerve, House and Brackmann, Retrosigmoid, Vestibular Schwannoma

Vestibular schwannomas are said to arise from the glial-Schwann cell junction – „Obersteiner-Redlich zone“ of the Vestibular nerve located at or medial to the porus of the internal auditory canal (IAC).[4,20] Acoustic neuroma is a commonly used misnomer, as the tumor arises from Schwann cells, mostly the vestibular

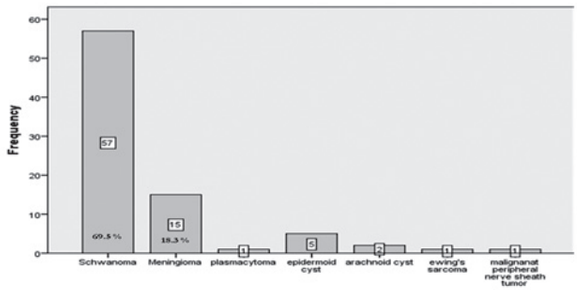

Figure 1: Histopathology of cerebello-pontine angle tumors

branch of the 8th cranial nerve. [7] Small tumors present with vestibular symptoms as tinnitus[18] and otological symptoms as hearing impairment which may not be noticed by the patients. [24] As the tumor grows and compresses the central nervous system, cranial nerve palsies, brain stem compression and hydrocephalus can gradually occur. This is when patients present to us in developing countries like Nepal, due to lack of awareness and accessibility to service-providers.

We do not have a national registry but the overall incidence of vestibular schwannoma has been reported as 1.09 per 100,000 population.[13] Unlike our clinical scenario, increasing availability and frequent use of magnetic resonance (MR) imaging have led to diagnosis of very small tumors worldwide,[8] with neurosurgeons successfully attempting hearing preservation.

Observation with neuro-radiological follow-up, microsurgical resection and stereotactic radiotherapy are the available treatment modalities. [1,19] The three surgical approaches are retrosigmoid, translabrynthine and middle cranial fossa or subtemporal approaches; and though each of these have advantages and limitations, [11,21] we routinely practice retrosigmoid approach in our centre.

Microsurgical excision of large vestibular schwannomas can be associated with serious complications and functional sequlae mainly in the form of facial nerve function, as chances of preserving serviceable hearing in large vestibular schwannomas is very low. The aim of our study was to evaluate the outcome of surgical management of vestibular schwannoma and to determine if the size of the tumor affected the facial nerve outcome.

Methods

Patients with tumors in the cerebello-pontine angle region operated between Baisakh 2066 to Chaitra 2073 (8 years) at the National Institute of Neurological and Allied Sciences, a 100-bed, tertiary care neuro centre in Kathmandu, Nepal was retrospectively analyzed. Among them a detailed study of patients who were histopatholgically proven to have vestibular schwanomma was done. This study was approved by the ethical board of the institution. The demographic profile of the patients, the presenting symptoms, the cranial nerve involvement, radiological investigations, surgical intervention and complications were analyzed. The size of the tumor was classified according to the Koos classification[12]on the basis of the maximum transverse diameter on MR imaging. Pure tone audiometry hearing assessment records where available were retrieved. Patients requiring CSF diversion procedures before and after surgery was assessed. The surgical approach to the tumor was routinely retrosigmoid but positioning varied from supine oblique, lateral park bench to sitting. For facial nerve preservation we used a two-channel Neurosign 100, a nerve monitoring system with electrodes embedded in the ipsilateral orbicularis oculi and orbicularis oris muscle. The surgical technique employed was intra-capsular dissection and avoiding the use of bipolar coagulation close to structures suspected to be the cranial nerves. The morbidity and mortality following the surgery was analyzed. The outcome of patients in terms of facial nerve preservation at six months post surgery as per the House and Brackmann (HB)[9] facial nerve grading system was determined. A Pearson's chi square (Fischer's Exact) test was done to see if the size of the tumor had any bearing on the facial nerve outcome at six months. Data entry and analysis was done using IBM SPSS (version 20; SPSS Inc., Chicago, IL, USA)

Results

Demographics

A total of 88 patients with cerebello-pontine angle (CPA) tumors were operated over eight years, averaging around 11 cases every year. Out of these, records of 84 patients could be retrieved. 57 out of the 84 tumors (69.5%) were schwannomas. Two of them as confirmed by radiology were trigeminal schwanommas and the rest vestibular. Menigioma, epidermoid, arachnoid cyst accounted for the rest and there was a case each of plasmacytoma, manlignant peripheral nerve sheath tumor and ewing's sarcoma (Fig. 1)

The mean age of the patients with vestibular schwannoma was 39.96 ± 10.95 years and they ranged from 22 years to 68 years. 25 (43.86%) were males and 32 (56.14%) were females. 24 (42.1%) were Aryans (Brahmins and Chettris), 22 (38.6%) Mongolian and the rest 11 (19.3%) were Madhesis. 32 patients (56.1%) had the tumor on the left side, 23 (40.4%) had it on the right side and two patients had bilateral vestibular

Koos Classification		Number of Patients
Stage 1	≤1cm	0
Stage 2	≤2cm	1 (1.75%)
Stage 3	≤3cm	8 (14.04%)
Stage 4	>3cm	48 (84.2%)

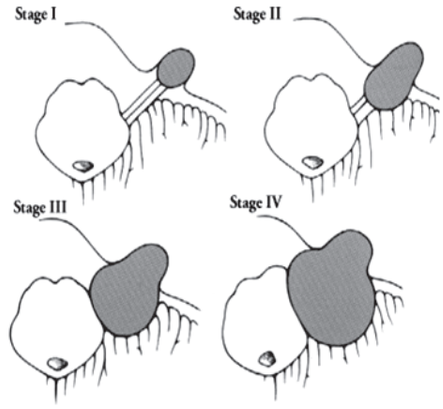

Figure 2: Classification of tumors according to their size (Koos classification) StageI: tumor confined to internal auditory canal, diameter1—10mm; stage II: tumor protruding at the CPA, less than 20mm; stage III: tumor occupies the CPA cistern with no displacement of the cerebral trunk, less than 30mm or touching the brainstem without causing compression; stage IV: large tumor with displacement of the cerebral trunk or cranial nerves, greater than 30mm or causing brainstem compression (reproduced from Koos et. al.)[12]

schwannomas. These two patients with Neurofibromatosis type 2 were a 22 year old gentleman with a left vestibular schwanomma, hypoglossal neuroma, right trigeminal schwannoma and multiple schwannomas in spine and a 46 year old gentleman with right parafalcine meningioma, B/L vestibular schwannomas and a C3-4 schwannoma.

Clinical Features and Investigations

The most common symptoms were headache and hearing loss, present in 80% and 91.3% patients respectively. Tinnitus and vertigo were complained by 45.2% and 40.6% patients respectively. In Nepalese scenario, presentation was mostly late, when the tumor had grown to a large size with nearly half of the patients having visual symptoms and papilloedema documented in 42.1% of cases. Involvement of the 5th cranial nerve and the lower cranial nerves were seen in 65% and 29.6% cases respectively.

The records of pure tone audiometry (PTA) could only be retrieved in 17 patients. PTA on the affected ear was < 30db in 2 (11.76%), 30-50dB in 5 (29.41%), 51- 70 db (11.76%) and >70db in 8 (47.05%) patients. Speech discrimination study was not done in our centre.

The transverse, antero-posterior and the craniocaudal dimensions of the tumor were recorded from the MRI reports of the patients; and the largest transverse dimension was used for the Koos classification. The mean tumor size was 4.32 ± 1.23 cms, the largest being 7.5 cm and the smallest 2 cm. 48 (84.2%) of the patients had tumor size more than 3cms (fig. 2)

Surgical Intervention and Complications

As the patients presented late and had hydrocephalus and as many as 18 (31.57%) patients required a pre-operative ventriculoperitoneal shunt on the contra-lateral side. In eight (14.04%) patients an external ventricular drain was placed immediately prior to the surgery. The duration of the surgery was noted from the anaesthetic chart, from the time of induction to recovery and it was 6.07 ± 1.72 hours.

Though trans-labryinthine and sub-temporal (middle fossa) approaches have their advantages, retrosigmoid approach was routinely employed at our centre. 43 (75.44%) patients were operated in supine oblique position, eight (14.03%) in lateral park-bench position with a hockey stick incision and six (10.53%) in sitting position.

The mean hospital stay was 25.44 ± 14.71 days. This long hospital stay is partly affected by the need of pre-operative VP shunt in as many as 18 (31.57%) patients where the major surgery followed a week later once the patient recovered from the CSF diversion procedure.

Eight (14.04%) patients developed post operative hydrocephalus and had to be shunted. Nine (15.79%) patients developed pseudomenigocele at the operative side, all of whom responded completely with placement of lumbar drain for five days. Two (3.5%) patients developed CSF fistula, who were also treated with lumbar drain. The incidence of post operative meningitis could not be confirmed by this retrospective study; as in the post-operative period antibiotics were empirically used when the patient's state and CSF analysis were dubious. Records showed the use of antimeningitic treatment in 14 (24.56%) patients; but this was not based on culture positivity. Post operative hematoma requiring re-exploration occurred in one patient.

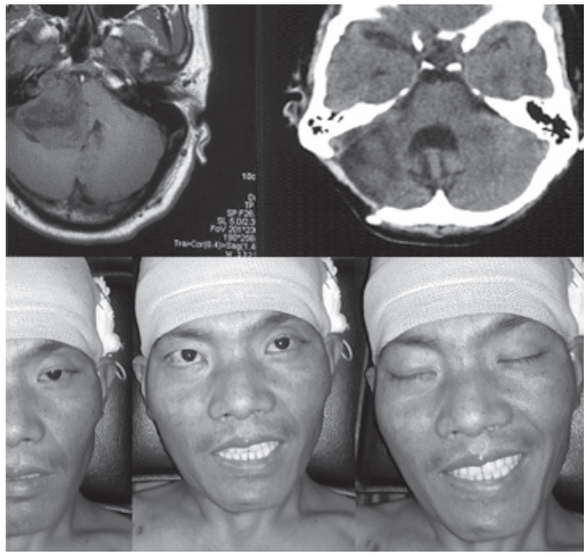

Figure 3: Facial nerve preservation in a35/M with Right Vestibular Schwannoma 5.1 cm X 4.5 cm X 4 cm

(Written consent obtained for the use of these photographs)

There were two (3.5%) mortalities. The first was a 66 year old gentleman with recurrent right cystic vestibular schwannoma, who underwent a near total excision a week after ventriculo-peritoneal shunting. After prolonged surgery, the patient had cavity hematoma, which was evacuated the same night; but the patient expired a week later. Another was a 40 year old gentleman who was reported to have a 5 cm X 4 cm X 3.5 cm left CP angle mass, likely Glossopharyngeal Schwannoma. He was operated in a supine oblique position, a week after initial CSF shunting. Brain swelling occurred before the tumor could be reached. External ventricular drain had to be kept which had to be changed multiple times due to infection; the patient expired after 37 days of ICU stay.

Hearing and Facial nerve preservation

In the Nepalese setting, hearing preservation could never be our aim due to delayed presentation; however, we were able to preserve hearing in a patient with a 2.7 cm sized vestibular schwannoma, which has been reported as a case report elsewhere. [15]

We then assessed the facial nerve outcomes of patients at or after six months. Facial nerve functions as per the House and Brackmann grading was done by authors PS and NK. Out of the 55 surviving patients, only 26 could be followed despite repeated attempts. Photographic recording of the facial nerve function was done whenever possible with adequate consent at rest, while smiling and while closing eyes. Tumors less than 3 cms (Koo's classification; largest dimension) were grouped as small tumors and those more than 3 cms as large. Patients with HB grading of 3 or below (upto moderate dysfunction), i.e. those patients who did not have gross facial asymmetry and those that could close eyes completely on effort were regarded as good outcome. HB grade 4-6 were regarded as bad facial nerve outcomes. All patients, seven, with tumors less than 3 cm in size had good facial nerve outcome at 6 months; however 7 out of 19 patients with tumors larger than 3 cm had poor facial nerve outcome. (Figure 3) The findings have been presented in the table below and a Fischer Exact test was done to test for statistical significance. The p-value was 0.077, which approached significance but was still clinically significant.

Discussion

Lesions in the CPA may be lesions originating in the CPA, lesions primarily located in adjacent anatomical sites and extending to the CPA and intraventricular and brain stem pathologies showing exophytic expansion to the CPA. Vestibular schwannomas (80-94%), meningiomas (3 to 10%), Epidermoids (2-4%) are the common tumors occurring in the CPA; 3 the list however is exhaustive. In our series Schwannomas accounted for 69.5 % and meningiomas 18.3 %.

There have been many large series of vestibular schwannomas published worldwide with skewed demographic details as presentation to clinicians in different parts of the world differ, so does the management ranging from watchful waiting to microsurgical resection. We had an adult subgroup of patients with the oldest being 68 years and the youngest, a patient with NF2, 22 years.

		House and Brackmann Grading		Total	Fischer's Exact Test (p-value)
		Good (1-3)	Bad (4-6)		
Size of tumor	≤ 3cms	7	0	7	0.077
	>3cms	12	7	19	
Total		19	7	26	

Figure 5: Cross tabulation of the size of the tumor with House and Brackmann facial nerve grading

The mean age was 39.96 ± 10.95 years and 56.14% were females. In Sami's series of 1000 patients 54 percent were females and the mean age of the patients was in the mid forties. 18 NF2 accounted for two (3.5%) cases in our series and it was 6.6% of 7017 patients operated elsewhere.16 These patients presented early, required multiple operation; hence our practice was to delay surgery as long as the patient had some useful hearing.

According to Sami's series, the most frequent clinical symptoms were disturbances of the acoustic (95%), vestibular (61%), trigeminal (9%), and facial (6%) nerves. 18 In our series, however, patients presented late with papilloedema documented in 42.1% of cases and many presenting with involvement of Trigeminal (65%) and the lower cranial nerves (29.6%).

Our review had some limitations. We were unable to elicit details regarding the extent of excision from the retrieved records. Sub-total resection, gross total resections were not adequately recorded; however, in most patients, total excision of the tumor with drilling of the internal auditory canal was attempted by senior author UPD. Post-operative MR imaging was not done routinely to lessen the financial burden to family, so we are not able to document the extent of resection. Anatomical intactness of the facial nerve at the end of the surgery was also not adequately recorded. In a series of 979 tumors operated by Sami, 23major neurological complications included a case of tetraparesis, 10 cases of hemiparesis, and caudal cranial nerve palsies in 5.5% of the cases. Operative site hematoma occurred in 2.2%, cerebrospinal fluid fistulas in 9.2%, hydrocephalus in 2.3%, bacterial meningitis in 1.2%, and wound revisions in 1.1%. We had one (1.8%) operative site hematoma, two (3.5%) CSF fistulas, hydrocephalus in eight (14%) and pseudomeningocele in nine (15.79%). Though antimeningitic treatment was used in 14 (24.56%) patients postoperatively, we were unable to identify the incidence of true bacterial meningitis. In Sami's series, there were 11 deaths occurring at 2 to 69 days postoperatively (1.1%); we lost two patients (3.5%) one following operative site hematoma and another to unexplained brain swelling before the tumor could be reached.

Hearing preservation could never be our aim as 84.2% of patient presented to us after the tumor had grown beyond 3 cms. We still do not do speech discrimination study at our centre, a sign that our priorities in vestibular schwanomma surgery differs for many centers worldwide attempting hearing preservation. Hopefully, in the years to come, with greater public awareness and earlier referral from ENT surgeons, we will be able to move in the right direction. A rare case where we could preserve hearing in a patient with a 2.7 cm vestibular schwanomma keeps us reminding this fact. 15

We were able to document the facial nerve outcome at 6 months of only 26 of the 57 (45.6%) patients; as patients operated at our centre were from various places all over the country. Among them, seven patients who had tumors less than 3 cms, all had good outcome (HB grade I to III). 12 out of 19 patients who had tumors larger than 3cms also had good outcome; the rest 7 did not do well. Facial-hypoglossal anastomosis was attempted in one of these patients; but he too did not have a good outcome. Fischer exact test could not, however, establish statistical significance; though our findings suggest, facial nerve preservation is more difficult with larger tumors. Bloch O et al. found out that only preoperative tumor size predicted the facial nerve outcome significantly; age, extent of resection, surgical approach did not.2 Kim J et al. also agree with the fact that preoperative size of the tumor is one of the most important prognostic factors; but surgeon's experience and preoperative facial nerve injury by the tumor can affect the post operative facial nerve outcome. 10

The facial nerve outcome during resection of large vestibular schwannomas has been variously reported in literature. Mild facial nerve weakness (HB grade I and II) only, when tumors larger than 3cm were operated was seen in 52.6 % of 178 patients by Lanmann et al. (1999) 14 , 65% of 40 patients by Wu H et al. (2000) 27 , 45% of 30 patients by Mamikoglu B et al. (2002) 7 , 84% of 50 patients by Yamakani I et al. (2004) 28 , 44% of 239 patients by Darrouzet V et al. (2004) 5 , 62% of 90 patients by Deveze A et al. (2004) 6 , 94% of 35 patients by Patni AH et al. (2005) 22 and 63% in 87 patients by Talfer S et al.(2010) 24. We could follow up only 19 of our 48 patients with tumors larger than 3 cm. Of them 9 out of 19 (47.4%) had HB grade I and II on follow-up, which was comparable to the work of Mamikoglu and Darrouzet. 5,17

We were unable to analyze the difference in preservation of facial nerve in the solid and cystic varieties of the vestibular schwannoma, due to inadequate recording. A study by Thakur JD et al. concluded facial nerve outcomes were significantly better in the cohort of patients with solid Vestibular Schwannomas (SVSs) than in those with cystic Vestibular Schwannomas (CVSs) (52.1% vs 39%, p = 0.0001) 26. In another study by Tang IP et al there was no such difference noted. Postoperative facial nerve outcomes at 1-year follow-up were good (HB Grade I-III) in 116 (88.5%) of 131 cystic Vestibular Schwannoma 118 (90.1%) of 131 solid Vestibular Schwannoma. 25

Conclusion

Retrosigmoid approach is a versatile surgical corridor to excise large vestibular schwannomas with minimal complications. Small tumors < 3cm can be safely microsurgically excised with good facial nerve outcomes; however, lack of awareness and accessibility to service-providers, cause patients to present late, endangering the facial nerve function and making hearing preservation next to impossible in Nepalese context.

References

1. Arthurs BJ, Fairbanks RK, Demakas JJ, Lamoreaux WT, Giddings NA, Mackay AR, et al. A review of treatment modalities for vestibular schwannoma. **Neurosurg Rev 34(3):**265-77, discussion 77-9, 2011
2. Bloch O, Sughrue ME, Kaur R, Kane AJ, Rutkowski MJ, Kaur G, et al. Factors associated with preservation of facial nerve function after surgical resection of vestibular schwannoma. **J Neurooncol 102(2):**281-6, 2011
3. Bonneville F, Sarrazin JL, Marsot-Dupuch K, Iffenecker C, Cordoliani YS, Doyon D, et al. Unusual lesions of the cerebellopontine angle: a segmental approach. **Radiographics 21(2):**419-38, 2001
4. Bridger MW, Farkashidy J. The distribution of neuroglia and schwann cells in the 8th nerve of man. **J Laryngol Otol 94(12):**1353-62, 1980
5. Darrouzet V, Martel J, Enee V, Bebear JP, Guerin J. Vestibular schwannoma surgery outcomes: our multidisciplinary experience in 400 cases over 17 years. **Laryngoscope 114(4):**681-8,2004
6. Deveze A, Roche PH, Facon F, Gabert K, Pellet W, Thomassin JM. [Functional outcomes after translabyrinthine approach for vestibular schwannomas]. **Neurochirurgie 50:**244-52, 2004
7. Eldridge R, Parry D. Vestibular schwannoma (acoustic neuroma). Consensus development conference. **Neurosurgery 30(6):**962-4, 1992
8. Haines SJ, Levine SC. Intracanalicular acoustic neuroma: early surgery for preservation of hearing. **J Neurosurg 79(4):**515-20. 1993
9. House JW, Brackmann DE. Facial nerve grading system. **Otolaryngol Head Neck Surg 93(2):**146-7, 1985
10. Kim J, Moon IS, Jeong JH, Lee HR, Lee WS. What really decides the facial function of vestibular schwannoma surgery? **Clin Exp Otorhinolaryngol 4(4):**168-73, 2011
11. Koerbel A, Gharabaghi A, Safavi-Abbasi S, Tatagiba M, Samii M. Evolution of vestibular schwannoma surgery: the long journey to current success. **Neurosurg Focus 18(4):**e10, 2005
12. Koos WT, Spetzler RF, Böck FW, Salah S. Microsurgery of cerebellopontine angle tumors. In: Koos WT, Böck FW, Spetzler RF(eds): **Clinical microneurosurgery.** Stuttgart: Thieme; 1976. pp 91—112
13. Kshettry VR, Hsieh JK, Ostrom QT, Kruchko C, Barnholtz-Sloan JS. Incidence of vestibular schwannomas in the United States. **J Neurooncol 124(2):**223-8, 2015
14. Lanman TH, Brackmann DE, Hitselberger WE, Subin B. Report of 190 consecutive cases of large acoustic tumors (vestibular schwannoma) removed via the translabyrinthine approach. **J Neurosurg 90(4):**617-23, 1999
15. Lohani S, Devkota UP. Hearing preservation in 2.7 cm vestibular schwannoma. **JNMA J Nepal Med Assoc 48(174):**158-61, 200916.
16. Mahboubi H, Maducdoc MM, Yau AY, Ziai K, Ghavami Y, Badran KW, et al. Vestibular Schwannoma Excision in Sporadic versus Neurofibromatosis Type 2 Populations. **Otolaryngol Head Neck Surg 153(5):**822-31, 2015
17. Mamikoglu B, Wiet RJ, Esquivel CR. Translabyrinthine approach for the management of large and giant vestibular schwannomas. **Otol Neurotol 23(2):**224-7, 2002
18. Matthies C, Samii M. Management of 1000 vestibular schwannomas (acoustic neuromas): clinical presentation. **Neurosurgery 40(1):**1-9; discussion -10, 1997
19. Muzevic D, Legcevic J, Splavski B, Caye-Thomasen P. Stereotactic radiotherapy for vestibular schwannoma. **Cochrane Database Syst Rev 16(12):**CD009897, 2014
20. Obersteiner H, Redlich E: Über Wesen und Pathogenese der tabischen Hinterstrangserkrankungen. **Arbeit Neurol Inst Wien.** Wien, 1895
21. Ojemann RG. Management of acoustic neuromas (vestibular schwannomas) (honored guest presentation). **Clin Neurosurg 40:**498-535, 1993
22. Patni AH, Kartush JM. Staged resection of large acoustic neuromas. **Otolaryngol Head Neck Surg**

132(1):11-9, 2005

23. Samii M, Matthies C. Management of 1000 vestibular schwannomas (acoustic neuromas): surgical management and results with an emphasis on complications and how to avoid them. **Neurosurgery 40(1):**11-21, 1997
24. Talfer S, Dutertre G, Conessa C, Desgeorges M, Poncet JL. Surgical treatment of large vestibular schwannomas (stages III and IV). **Eur Ann Otorhinolaryngol Head Neck Dis 127(2):**63-9, 2010
25. Tang IP, Freeman SR, Rutherford SA, King AT, Ramsden RT, Lloyd SK. Surgical outcomes in cystic vestibular schwannoma versus solid vestibular schwannoma. **Otol Neurotol 35(7):**1266-70,2014
26. Thakur JD, Khan IS, Shorter CD, Sonig A, Gardner GL, Guthikonda B, et al. Do cystic vestibular schwannomas have worse surgical outcomes? Systematic analysis of the literature. **Neurosurg Focus 33(3):**E12, 2012
27. Wu H, Sterkers J. Translabyrinthine removal of large acoustic neuromas in young adults. **Auris Nasus Larynx 27(3):**201-5, 2000
28. Yamakami I, Uchino Y, Kobayashi E, Yamaura A, Oka N. Removal of large acoustic neurinomas (vestibular schwannomas) by the retrosigmoid approach with no mortality and minimal morbidity. **J Neurol Neurosurg Psychiatry 75(3):**453-8, 2004