Hearing Rehabilitation in Vestibular Schwannoma

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The most common complaint among patients with vestibular schwannoma (VS) is hearing loss. This significantly affects the quality of life before, during, and after treatment for patients with VS. Untreated hearing loss in VS patients may even lead to depression and feelings of social isolation. A variety of devices are available for hearing rehabilitation for patients with vestibular schwannoma.

Keywords: vestibular schwannoma ; CROS ; BiCROS ; BAHS ; ABI ; cochlear implant

1. Introduction

Vestibular schwannomas (VS) are benign neoplasms that arise from Schwann cells surrounding the superior or inferior vestibular (eighth cranial) nerve [1][2]. Tumors can be present in a number of intracranial or extracranial locations, thus making clinical symptoms at the time of presentation vary significantly. Symptoms at presentation may include any combination of the following: sudden sensorineural hearing loss, progressive hearing loss, central vestibular weakness, peripheral vestibular weakness, tinnitus, facial numbness, or facial weakness. In a study of more than 1000 patients, 85.8% had hearing loss, 48.9% had paresthesia of the face, there was a gait imbalance in 48.6%, and 40.1% had tinnitus [3]. The Characterization of these functional deficits at the time of initial work is imperative, not only in treatment planning but also in the determination of the most effective rehabilitative options as well [4]. There are not only many different treatment options for VS, but also many different options for subsequent or simultaneous auditory rehabilitation as well. New technologies are being developed and improved upon daily to better assist VS patients before, during, and after their treatment. The functionality of many of these implantable devices depends on the integrity of the cochlear nerve during and after treatment, which has remained a challenge to assure as a result of the nature of this disease process.

2. Nonsurgical Options for Auditory Rehabilitation: Conventional Hearing Aids, Contralateral Routing of Signals (CROS), and Bilateral Contralateral Routing of Signal (BiCROS) Hearing Aids

Patients with unilateral vestibular schwannoma typically present with asymmetrical sensorineural hearing loss in the affected ear with normal hearing thresholds in the unaffected ear, poor speech discrimination, and tinnitus [5]. Conventional hearing aids are offered to patients with vestibular schwannoma if they have mild to moderately severe sensorineural hearing loss. However, conventional hearing aids offer little to no benefit to patients with severe to profound unilateral hearing loss. For these patients, CROS hearing aids are better options for hearing rehabilitation.

CROS hearing aids are used to route sound from the worse-hearing ear to the better-hearing ear and are typical for patients with single-sided deafness. Two different internal devices within the hearing aids themselves are used to execute this contralateral routing process. The first device is inside the hearing aid, which is placed in the worse-hearing ear. It has a microphone that detects sound from that worse-hearing side; however, instead of routing that signal into the worse-hearing ear, similar to what a traditional hearing aid would do, the signal is routed to the opposite (the better-hearing) ear using a transmitter. Thus, sounds and information from the side of the worse-hearing ear are processed through the better-hearing ear.

BiCROS is recommended for patients with mild-to-moderate hearing loss in the contralateral ear, as compared to a normal-hearing contralateral ear for CROS. Historically, only a small percentage of patients with single-sided deafness (SSD) reported improvement in their quality of life with the use of CROS or BiCROS [6]. This may be due to the inherent limitations of sound/information transfer by air conduction as compared to bone conduction systems or electrical signal conduction systems, both of which have a greater capacity to send a stronger signal into the auditory centers for interpretation.

Advances in CROS hearing aid system technology have addressed previous limitations, such as short battery life, poor sound quality, discomfort, and aesthetics due to their large size. The addition of wireless streaming allows the sound signal to be transmitted to the better-hearing ear without delays or audible interference [7]. Advanced algorithms have allowed for improved noise reduction, and directional microphones with automatic adaptive directionality have improved the signal-to-noise ratio for users [8][9]. This has led to patients with VS accepting and increasing their usage of CROS and BiCROS devices as compared to previous adaptations of these devices. However, CROS and BiCROS do not provide binaural hearing and can even disrupt the individual's use of monaural spectral cues [10][11]. Surveys of vestibular schwannoma patients have reported that 30–32% of patients used conventional hearing aids, 23–30% used CROS devices, and 21.7% used BAHS [12][13].

3. Bone Anchored Hearing System (BAHS)

Bone-anchored hearing systems are surgical solutions used to mitigate unilateral hearing loss in patients with VS. It consists of a titanium screw or plate fixed to the bone behind the ear, and it connects either percutaneously or transcutaneously to an external bone oscillator. The microphone, which receives the sound signal, is integrated into the bone oscillator. The sound processor transforms the signal into an oscillatory signal, which is transmitted through the plate/screw, which in turn causes the bone to vibrate at a certain frequency and send a signal into the cochlea on the better hearing side. Unlike the CROS hearing aid, which uses air conduction, the BAHS uses bone conduction for sound transmission.

BAHS works best for patients with VS who have good contralateral hearing. Additionally, similar to CROS hearing aids, BAHS is limited by its inability to provide binaural hearing, which is important for sound localization and improving speech understanding in noise [6]. However, of note, due to the nature of bone conduction over air conduction, the high-power BAHS devices are able to provide power signal input through bone conduction. Surgery is overall safe and confers less risk than the implantation of electrical devices, such as CI or ABI, as no inner ear structures are opened during BAHS surgery. Additionally, there are surgical and nonsurgical treatment options for BAHS, which are interesting treatment options for all sorts of VS patients.

The wide variety of BAHS devices may be categorized into percutaneous or transcutaneous, and within the transcutaneous category, there are surgical and nonsurgical options. The variety of BAHS devices that are available, make for many different rehabilitative options for both pediatric and adult VS patients, as well as those who are unable to undergo surgery.

4. Auditory Brain Stem Implant (ABI)

ABI is a surgically implanted device that bypasses the cochlear nerve to electrically stimulate the neurons in the brain stem dorsal cochlear nucleus. For a long time, ABI was the only treatment for hearing rehabilitation for patients with hearing loss caused by VS. The ABI electrodes are embedded in a silastic paddle and placed in the fourth ventricle. The device has an external battery, a microphone, a speech processor, a transmitter coil, and a magnet that is worn behind the ear. ABI has been approved by the US FDA for use in patients age 12 and older with NF2 for hearing rehabilitation. Patients with ABI typically have sound awareness, but results with ABI have been modest, and open-set speech (conversational speech in daily life) after ABI is an exception [14][15]. Less than 5% of NF2 patients have open-set speech. It can assist lip reading, enable the identification of environmental sounds, and can drastically improve the patient's quality of life [16][17].

ABI is a great treatment option when the cochlear nerve has been rendered absent or nonfunctional; however, determining the status of the cochlear nerve remains a challenge. We must establish a standardized method of testing the cochlear nerve for functional integrity during and/or after observation or treatment for VS. This will allow a better treatment algorithm to be established and help us determine exactly who might benefit from CI over ABI. Since ABI surgery is more invasive than CI surgery, it should only be considered in appropriate patients who would not benefit from CI. Currently, though, our ability to make that determination on a consistent basis remains somewhat limited.

5. Cochlear Implant (CI)

Cueva et al. in 1992 reported data from six patients with grossly intact cochlear nerves and anacusis after resection of VS who were able to perceive sound following the electrical promontory stimulation. They postulated that these patients could potentially benefit from cochlear implantation [18]. Subsequently, Hoffman et al. reported a case of a young NF2 patient

who lost his hearing despite hearing preservation with retrosigmoid resection of the VS with sparing of the cochlear nerve. The patient underwent cochlear implantation three months after VS resection and achieved open-set speech [19].

Since then, several studies have demonstrated CI to be an effective means for auditory rehabilitation in patients with sporadic unilateral VS as well as NF2 patients after microsurgical resection or radiation therapy [4][6][19][20][21][22][23]. There are also studies that demonstrate that CI is an effective option for hearing rehabilitation in patients with small/stable VS who are being followed with serial imaging [12][24][25].

The CI electrically stimulates the cochlear nerve in the inner ear to enable identification, interpretation, and associating meaning with the sound (auditory perception). This is unlike the ABI, which stimulates the neurons of the dorsal cochlear nucleus in the brain stem. CI is a device consisting of a surgically implanted electrode array with a receiver-stimulator coupled with an external sound processor. The microphone on the external processor receives sound and converts it to an electrical signal, which is carried to the spiral ganglion cells of the cochlear nerve via a multielectrode array. Cochlear implant electrode arrays utilize the tonotopic organization of the cochlea to maximize sound intelligibility by covering almost the entire spectrum of perceivable auditory frequencies. For a cochlear implant to be effective, cochlear nerve integrity is essential and may play a role in hearing outcomes after CI in VS patients. This means that any therapeutic modality offered to treat VS must also be able to preserve cochlear nerve integrity. Even if the cochlear nerve is preserved during surgical resection, it has been shown that a majority of those patients can go on to lose their hearing [14]. Several factors, such as vascular compromise, trauma to the nerve, and cochlear fibrosis, can influence conduction along the cochlear nerve after surgical resection and the ability of the patient to benefit from a cochlear implant [14].

Determining the functional integrity of the cochlear nerve has always presented a challenge. The electrical promontory stimulation (EPS) test is a test that has been used traditionally to determine the functionality of the cochlear nerve. It is performed by placing a transtympanic needle electrode on the surface of the cochlea, and the cochlea nerve is electrically stimulated through the monitoring of subjective and electrically evoked auditory brainstem responses. A loss of neural integrity at the spiral ganglion or cochlear nerve level is indicated by an absence of response to stimulation. On the other hand, the presence of a response is indicative of vascular compromise of the cochlea with the sparing of the spiral ganglion and the cochlear nerve. This test has its drawbacks, as some patients with good EPS do not benefit from a CI, while some with poor EPS have achieved open-set speech with a CI [14][18][20].

In a large systematic review, 65% of patients with NF2 who underwent surgical resection with ipsilateral cochlear implantation were found to have developed open-set speech with the CI [23]. Interestingly, no differences in outcomes after CI were observed when comparing simultaneous to sequential tumor resection and implantation [23]. In another systematic review performed by Wick et al., CI performance was not found to be influenced by the timing of cochlear implantation, whether during or after tumor resection [14]. In an ongoing FDA-approved clinical trial, a device monitoring the auditory nerve during VS resection called the auditory nerve test system (ANTS) is helping the surgeon-investigators preserve the nerve during VS resection and rehabilitate patients with simultaneous cochlear implants [26].

References

1. Tos, M.; Stangerup, S.-E.; Cayé-Thomasen, P.; Tos, T.; Thomsen, J. What Is the Real Incidence of Vestibular Schwannoma? Arch. Otolaryngol. Head Neck Surg. 2004, 130, 216–220.
2. Marinelli, J.P.; Lohse, C.M.; Carlson, M.L. Incidence of Vestibular Schwannoma over the Past Half-Century: A Population-Based Study of Olmsted County, Minnesota. Otolaryngol.–Head Neck Surg. 2018, 159, 717–723.
3. Huang, X.; Xu, J.; Xu, M.; Zhou, L.-F.; Zhang, R.; Lang, L.; Xu, Q.; Zhong, P.; Chen, M.; Wang, Y.; et al. Clinical features of intracranial vestibular schwannomas. Oncol. Lett. 2013, 5, 57–62.
4. Tian, L.; West, N.; Caye-Thomasen, P. Cochlear Implantation After Radiotherapy of Vestibular Schwannomas. J. Int. Adv. Otol. 2021, 17, 452–460.
5. Von Kirschbaum, C.; Gürkov, R. Audiovestibular Function Deficits in Vestibular Schwannoma. Biomed. Res. Int. 2016, 2016, 4980562.
6. Patel, N.S.; Carlson, M.L.; Link, M.J.; Neff, B.A.; van Gompel, J.J.; Driscoll, C.L.W. Cochlear implantation after radiosurgery for vestibular schwannoma. J. Neurosurg. 2020, 135, 126–135.
7. Evans, D.R. Neurofibromatosis type 2 (NF2): A clinical and molecular review. Orphanet. J. Rare Dis. 2009, 4, 16.
8. Fanous, A.A.; Prasad, D.; Mathieu, D.; Fabiano, A.J. Intracranial stereotactic radiosurgery. J. Neurosurg. Sci. 2018, 63, 61–82.

9. Carlson, M.L.; Jacob, J.T.; Pollock, B.E.; Neff, B.A.; Tombers, N.M.; Driscoll, C.L.; Link, M.J. Long-term hearing outcomes following stereotactic radiosurgery for vestibular schwannoma: Patterns of hearing loss and variables influencing audiometric decline. J. Neurosurg. 2013, 118, 579–587.
10. Snapp, H. Nonsurgical Management of Single-Sided Deafness: Contralateral Routing of Signal. J. Neurol. Surg. B Skull Base 2019, 80, 132–138.
11. Pedley, A.J.; Kitterick, P.T. Contralateral routing of signals disrupts monaural level and spectral cues to sound localisation on the horizontal plane. Hear. Res. 2017, 353, 104–111.
12. Drusin, M.A.; Lubor, B.; Losenegger, T.; Selesnick, S. Trends in hearing rehabilitation use among vestibular schwannoma patients. Laryngoscope 2020, 130, 1558–1564.
13. Macielak, R.J.; Marinelli, J.P.; Spear, S.A.; Hahs-Vaughn, D.L.; Link, M.J.; Nye, C.; Carlson, M.L. Hearing Status and Aural Rehabilitative Profile of 878 Patients with Sporadic Vestibular Schwannoma. Laryngoscope 2021, 131, 1378–1381.
14. Wick, C.C.; Butler, M.J.; Yeager, L.H.; Kallogjeri, D.; Durakovic, N.; McJunkin, J.L.; Shew, M.A.; Herzog, J.A.; Buchman, C.A. Cochlear Implant Outcomes Following Vestibular Schwannoma Resection: Systematic Review. Otol. Neurotol. 2020, 41, 1190–1197.
15. Deep, N.; Choudhury, B.; Roland, J. Auditory Brainstem Implantation: An Overview. J. Neurol. Surg. B Skull Base 2019, 80, 203–208.
16. Fernandes, N.F.; Goffi-Gomez, M.V.S.; Magalhães, A.T.D.M.; Tsuji, R.K.; de Brito, R.V.; Bento, R.F. Satisfação e qualidade de vida em usuários de implante auditivo de tronco cerebral. Codas 2017, 29, e20160059.
17. Lundin, K.; Stillesjö, F.; Nyberg, G.; Rask-Andersen, H. Self-reported benefit, sound perception, and quality-of-life in patients with auditory brainstem implants (ABIs). Acta Otolaryngol. 2016, 136, 62–67.
18. Cueva, R.A.; Thedinger, B.A.; Harris, J.P.; Glasscock, M.E. Electrical Promontory Stimulation in Patients with Intact Cochlear Nerve and Anacusis Following Acoustic Neuroma Surgery. Laryngoscope 1992, 102, 1220–1224.
19. Hoffman, R.A.; Kohan, D.; Cohen, N.L. Cochlear implants in the management of bilateral acoustic neuromas. Am. J. Otol. 1992, 13, 525–528.
20. Neff, B.A.; Wiet, R.M.; Lasak, J.M.; Cohen, N.L.; Pillsbury, H.C.; Ramsden, R.T.; Welling, D.B. Cochlear Implantation in the Neurofibromatosis Type 2 Patient: Long-Term Follow-up. Laryngoscope 2007, 117, 1069–1072.
21. Laborai, A.; Ghiselli, S.; Cuda, D. Cochlear Implant in Patients with Intralabyrinthine Schwannoma without Tumor Removal. Audiol. Res. 2022, 12, 33–41.
22. Häussler, S.M.; Szczepek, A.J.; Gräbel, S.; Olze, H. Challenges of Cochlear Implantation in Intralabyrinthine Schwannoma Patients: Surgical Procedures and Auditory Outcome. J. Clin. Med. 2021, 10, 3899.
23. Carlson, M.L.; Breen, J.T.; Driscoll, C.L.; Link, M.J.; Neff, B.A.; Gifford, R.H.; Beatty, C.W. Cochlear Implantation in Patients with Neurofibromatosis Type 2. Otol. Neurotol. 2012, 33, 853–862.
24. Bartindale, M.R.; Tadokoro, K.S.; Kircher, M.L. Cochlear Implantation in Sporadic Vestibular Schwannoma: A Systematic Literature Review. J. Neurol. Surg. B Skull Base 2019, 80, 632–639.
25. Longino, E.S.; Manzoor, N.F.; Cass, N.D.; Tawfik, K.O.; Bennett, M.L.; O'malley, M.R.; Haynes, D.S.; Perkins, E.L. Cochlear Implantation Outcomes in Observed Vestibular Schwannoma: A Preliminary Report. Otolaryngol.–Head Neck Surg. 2022, 167, 149–154.
26. "ANTS". Available online: https://hearinghealthmatters.org/hearingnewswatch/2020/acoustic-neuroma-surgery-hearing/ (accessed on 1 November 2022).