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Audiologic Management of a Patient with a Sudden Hearing Loss and Vestibular Schwannoma in the Contralateral Ear

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Abstract

The case of a 59-year-old male with a sudden-onset sensorineural hearing loss in one ear and an incidental finding of an intracanalicular vestibular schwannoma in the contralateral, normally hearing ear is reported. The patient was successfully fitted with a hearing aid in the ear with the sudden hearing loss, which notably had very poor word recognition. The questionable value of word-recognition scores in determining hearing aid candidacy is discussed. The importance of considering nonaudiologic factors in determining hearing aid candidacy is also highlighted.

Key Words: Aural rehabilitation, hearing aids, sudden sensorineural hearing loss, vestibular schwannoma

Abbreviations: CID = Central Institute for the Deaf; IOI-HA = International Outcome Inventory for Hearing Aids; NAL–NL1 = National Acoustic Laboratories, Non-linear Version 1; SADL = Satisfaction with Amplification in Daily Living; SSHL = sudden sensorineural hearing loss

Sumario

Se reporta el caso de una mujer de 59 años de edad con una hipoacusia sensorineural de inicio súbito en un oído, y el hallazgo incidental de un schwanoma vestibular intracanalicular en el oído contralateral con audición normal. Al paciente se le adaptó exitosamente un auxiliar auditivo en el oído con la hipoacusia súbita, que notablemente tenía un reconocimiento de palabras muy pobre. Se discute el cuestionable valor de los puntajes de reconocimiento de palabras para determinar la candidatura a un auxiliar auditivo. También se destaca la importancia de considerar los factores no audiológicos a la hora de determinar la candidatura para un auxiliar auditivo.

Palabras Clave: Rehabilitación aural, auxiliares auditivos, hipoacusia sensorineural súbita, schwanoma vestibular

Abreviaturas: CID = Instituto Central del Sordo; IOI-HA = Inventario Internacional de Resultados con Auxiliares Auditivos; NAL-NL1 = Laboratorios Nacionales de Acústica, Versión No Lineal 1; SADL = Satisfacción con la Amplificación en la Vida Diaria; SSHL = Hipoacusia sensorineural súbita

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Although audiologists often see patients with vestibular schwannomas or sudden sensorineural hearing loss (SSHL), both pathologies are relatively rare in the general population. The incidence of vestibular schwannomas is 1 in 80,000 people (Ramsden et al, 2005), and SSHL accounts for only 1 percent of sensorineural hearing losses (Hughes et al, 1996). It is even more unusual for both of these auditory pathologies to occur in the same patient, though such cases have been documented (Driscoll et al, 2000).

A common finding with both vestibular schwannomas and SSHL is poor word-recognition ability (Fetterman et al, 1996; Harner et al, 2000). Reports of successful hearing aid fittings in individuals with poor word-recognition scores are not commonly seen in the audiologic literature. This could be in part due to the belief that an individual with poor word-recognition scores may not derive significant benefit from hearing aids.

This article documents the audiologic management of a patient with SSHL in one ear and a vestibular schwannoma in the normal-hearing contralateral ear. This same patient was successfully fitted with a hearing aid in the ear with the SSHL, which had very poor word recognition. It is an interesting and unusual case from both a diagnostic and a rehabilitative perspective and may challenge the way some dispensing audiologists approach ears with poor word recognition.

CASE REPORT

Approximately three months prior to arriving at Mayo Clinic Florida, a 59-year-old male accountant awoke one morning with sudden hearing loss and tinnitus in his left ear. He immediately sought care from his local family physician, who placed him on a corticosteroid and ordered a magnetic resonance imaging (MRI) scan of the internal auditory canals to rule out retrocochlear pathology. The scan was unremarkable on the left side. However, the scan did reveal an incidental finding of an intracanalicular lesion (10 × 8 × 4 mm) on the right side, consistent with a right vestibular schwannoma.

The patient presented to Mayo Clinic Florida seeking assistance in determining what treatment he should pursue, if any, for the right vestibular schwannoma. In addition, he requested an opinion as to whether anything further could be done to improve the hearing in his left ear. He described significant auditory-based communication difficulties in everyday life as a result of the SSHL. He also mentioned that he was quite bothered by the tinnitus that he continued to experience in his left ear.

Audiometric results for the right ear with the vestibular schwannoma are shown in Figure 1. Pure-tone sensitivity was within normal limits. Word recognition was 92 percent at 50 dB HL (Central Institute for the Deaf [CID] W-22 list), and ipsilateral and contralateral acoustic reflex thresholds were obtained at normal levels. There was no acoustic reflex decay at 500 or 1000 Hz at 10 dB sensation level. The left ear with the SSHL presented a mild sloping to profound sensorineural hearing loss, with poor word-recognition scores (20% at 80 dB HL and 4% at 90 dB HL, CID W-22 lists) and elevated or absent ipsilateral and contralateral acoustic reflex thresholds.

A medical consultation with a Mayo Clinic otologist included a physical examination and review of an MRI scan that the patient brought with him to the evaluation. The MRI scan is shown in Figure 2 with an arrow directed toward the vestibular schwannoma. The diagnosis by the otologist was SSHL in the left ear, likely secondary to a viral idiopathic etiology, and an incidental finding of an intracanalicular

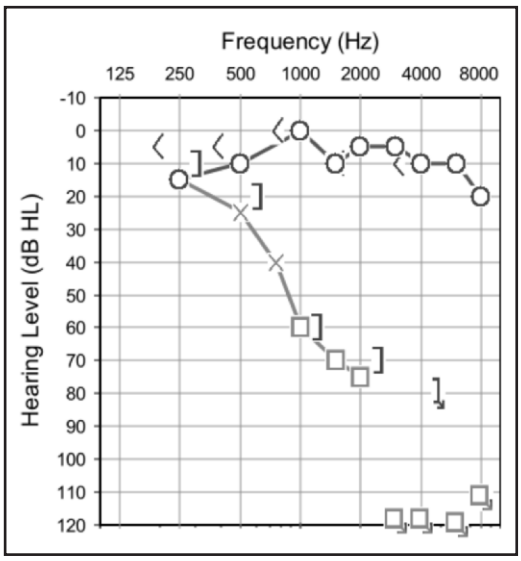

Figure 1. Pure-tone audiogram approximately three months after the onset of sudden hearing loss in the left ear.

vestibular schwannoma on the right. The otologist acknowledged the difficult situation that the patient faced in deciding whether to treat a vestibular schwannoma in his normal-hearing right ear. The options available to treat the vestibular schwannoma, including stereotactic radiosurgery and surgical excision, were reviewed with the patient, while acknowledging that either option could precipitate hearing loss in the right ear, thus potentially resulting in very poor hearing bilaterally. Given the risk of hearing loss and the fact that the vestibular schwannoma was small and asymptomatic, the otologist recommended that the patient defer treatment of the vestibular schwannoma and instead return every six months for MRI scans to monitor tumor growth. The patient was counseled that should he lose his hearing in his right ear, he would then likely become an excellent candidate for a cochlear implant in his left ear, especially because the left ear had received normal auditory input for most of his lifetime. Given the significant difficulties the patient reported with his hearing, he was referred to the Mayo Hearing Aid Clinic for a discussion of amplification treatment options.

At his hearing aid consultation, the patient reiterated the significant difficulties he had experienced since the SSHL in the left ear. He described difficulty hearing from the affected side, increased difficulty hearing in background noise, and problems with localization. He was also troubled by the sudden "lack of auditory balance" he experienced between ears and persistent tinnitus in the left ear.

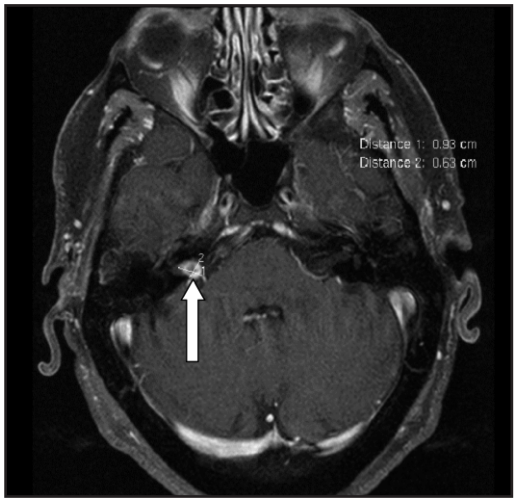

Figure 2. Magnetic resonance imaging scan of the posterior fossa showing a right-sided intracanalicular vestibular schwannoma. The arrow shows the location of the vestibular schwannoma.

Several amplification options were discussed with the patient, including a bone-anchored hearing aid, contralateral routing of signal hearing aid system, and a traditional monaural hearing aid for the left ear. In spite of the poor word recognition in the left ear, it was decided to first try a monaural hearing aid in the left ear, as it was the only option of the three that could potentially reduce the perception of tinnitus and lack of auditory balance. An additional factor in this decision was the fact that we have encountered a number of patients at our clinic with poor word recognition who reported significant benefit from hearing aids.

In selecting hearing aid circuitry for the left ear, emphasis was placed on finding a hearing aid with sufficient bands for good frequency shaping and feedback cancellation to address the patient's steeply sloping audiogram and lack of usable hearing beyond 2000 Hz. In order to provide listening options for both quiet and noisy environments, the presence of multiple settings with both an omnidirectional and a directional microphone was also considered important in the selection process. The patient indicated that cost was not an important variable to consider in the selection of a hearing aid. As such, a high-end digital half-shell hearing aid with 20 bands, multiple memories, and an adaptive directional microphone was selected for the left ear. The patient was counseled extensively regarding potential benefits and limitations that he could expect from the use of a hearing aid in the left ear.

At the hearing aid fitting appointment, the output of the hearing aid was adjusted to match National Acoustic Laboratories, Non-linear Version 1 (NAL–NL1) targets using real-ear probe microphone measurements with speech signals. Amplified speech was within ±4 dB of NAL–NL1 targets for the frequency range of 250 to 2000 Hz. At a follow-up visit four weeks after the fitting, the patient reported that he was benefiting from the hearing aid and wearing it more than eight hours a day. He indicated that the hearing aid masked "90 percent of the tinnitus" and gave him "[auditory] balance." He stated, "I am better with it than without it," though he noted little to no improvement in speech understanding in background noise.

The International Outcome Inventory for Hearing Aids (IOI-HA [Cox et al, 2000]) and the Satisfaction with Amplification in Daily Life questionnaire (SADL [Cox and Alexander, 1999]) were administered at this time to formally assess patient-perceived hearing aid benefit and satisfaction. The patient's scores on these measures are plotted against the Cox and colleagues' (Cox and Alexander, 2001; Cox et al, 2003) norms in Figures 3 and 4. Higher scores indicate more positive results. As shown in Figure 3, the

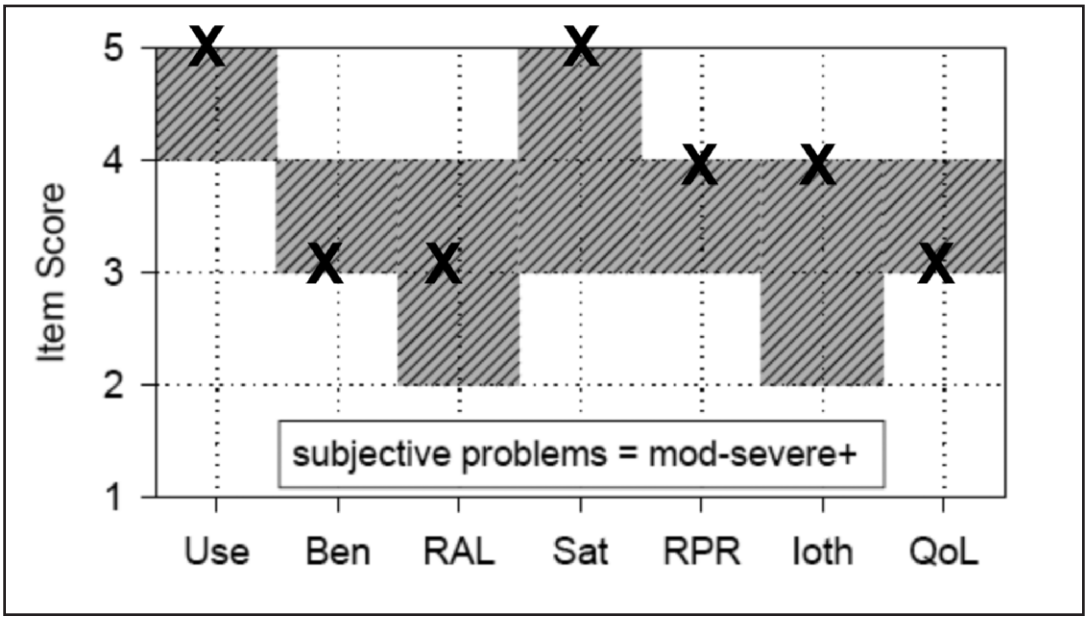

Figure 3. International Outcome Inventory for Hearing Aids (IOI-HA [Cox et al, 2000]) results displayed with the normal values from Cox and colleagues (2003). The IOI-HA was administered four weeks after the hearing aid fitting. The shaded areas represent the middle 50 percent of responses from the normative group, and higher numbers represent more positive results. Use = use time; Ben = benefit; RAL = residual activity limitations; Sat = satisfaction; RPR = residual participation restrictions; Ioth = impact on others; QoL = quality of life.

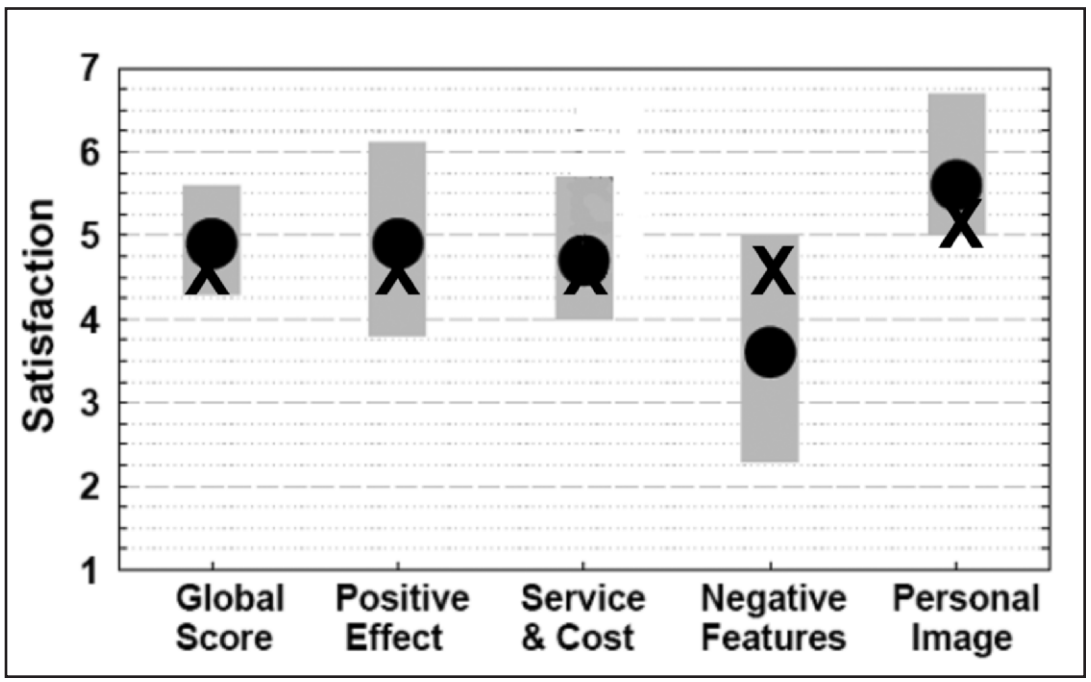

Figure 4. Satisfaction with Amplification in Daily Life (SADL [Cox and Alexander, 1999]) results displayed with the normative values from Cox and Alexander (2001). The SADL was administered four weeks after the hearing aid fitting. The solid circles are the mean values for the normative group, and the gray bars represent the 20th to 80th percentiles. The X symbols are the responses from the patient.

patient's scores on the IOI-HA fell within the middle 50 percent range of Cox and colleagues' norms in all categories. The patient's scores were particularly high in the "use" and "satisfaction" categories and lower in "benefit" and "residual activity limitation." This suggests that even though the patient continues to have difficulty understanding speech, he is highly satisfied with the hearing aid overall. This satisfaction may reflect the improvements he experiences in other areas, such as auditory awareness and the reported reduction of tinnitus and improvement in "balance" between ears.

Figure 4 shows the patient's scores on the SADL. The responses were at or above the Cox and Alexander's mean values in all categories. Thus the patient's perceived satisfaction with his hearing aid was comparable to that of an average hearing aid user, a finding that some might find surprising given the patient's poor word-recognition score in the left ear.

DISCUSSION

Audiologists often begin to suspect a particular pathology based on a certain pattern of test results. For instance, otosclerosis is often suspected in an ear with conductive hearing loss, a normal tympanogram, and absent acoustic reflexes. In the case reported in this article, the patient's audiometric results defied the typical "pattern" one would expect in an ear with a vestibular schwannoma. Normal pure-tone thresholds, word recognition, and ipsilateral and contralateral acoustic reflexes certainly do not lead one to suspect retrocochlear pathology. The surprising and unfortunate finding of a vestibular schwannoma in an audiometrically asymptomatic ear in this case highlights the importance of multidisciplinary collaboration in patients with complex problems.

Also interesting in this case was the level of satisfaction that the patient reported from a hearing aid in an ear with poor word recognition. The patient's satisfaction with the hearing aid may in part be attributed to the sudden onset of his hearing loss. It is possible that he may have felt more handicapped by the hearing loss and more bothered by lack of "auditory balance" due to its sudden onset. As such, he may have perceived greater benefit from the hearing aid than a person with a similar hearing loss of congenital or more gradual onset. The tinnitus relief experienced by the patient with the use of the hearing aid may also have contributed to his overall satisfaction.

Nonetheless, the patient did surprisingly well with a hearing aid in an ear that some audiologists would have considered monaurally "unaidable." This case illustrates that word-recognition scores cannot always be used to predict hearing aid candidacy. According to Dillon and colleagues (1982), word-recognition scores are not a good indicator of hearing aid benefit due to the gain-frequency response of headphones and the reliability of word-recognition tests, among other factors. This has been confirmed by clinical experiences at this facility with other patients. Pure-tone thresholds and word-recognition scores are just two of many factors that one must consider when determining hearing aid candidacy. In this case the degree of self-perceived disability and level of motivation were particularly important factors.

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