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Hearing Rehabilitation in a Patient with Sudden Sensorineural Hearing Loss in the Only Hearing Ear

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Abstract

A case report is presented of a 62-year-old software product manager who had normal hearing in one ear and a congenital profound hearing loss in the other ear and then sustained a sudden sensorineural hearing loss in the only hearing ear. The approach to amplification decisions, cochlear implant evaluation, and rehabilitation options are discussed. Providing aural rehabilitation and continually updating and providing new amplification options and accessories are described.

Key Words: Directional microphones, FM systems, hearing aids, rehabilitation, sudden sensorineural hearing loss

Abbreviations: CID = Central Institute for the Deaf; CNC = consonant/nucleus/consonant; HINT = Hearing in Noise Test; IOI-HA = International Outcome Inventory for Hearing Aids; SNR = signal-to-noise ratio; SSHL = sudden sensorineural hearing loss

Sumario

Se presenta un reporte de caso de un gerente de productos de software de 62 años de edad quien tenía audición normal en un oído y un sordera congénita profunda en el otro, y quién súbitamente sufrió una sordera sensorineural súbita en el único oído con audición. Se discute el enfoque de decisiones de amplificación, la evaluación para implante coclear, y las opciones de rehabilitación. Se describen las pautas para proveer rehabilitación aural y para actualizar continuamente y aportar nuevas opciones de amplificación.

Palabras Clave: Micrófonos direccionales, sistemas de FM, auxiliares auditivos, rehabilitación, sordera sensorineural súbita

Abreviaturas: CID = Instituto Central del Sordo; CNC = consonante/núcleo/consonante; HINT = Prueba de Audición en Ruido; IOI-HA = Inventario Internacional de Resultados para Auxiliares Auditivos; SNR = tasa señal/ruido; SSHL = sordera sensorineural súbita

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The daily hearing problems of a person with a unilateral hearing loss are well documented (Bess and Tharpe, 1986). They primarily include difficulty hearing from the side of the hearing loss due to the head shadow effect, difficulty with sound localization due to the lack of interaural time and intensity cues, and particular difficulty understanding speech in a noisy environment due to the loss of binaural processing. Although these three limitations can cause significant communication problems, some patients with normal hearing in one ear and a total unilateral hearing loss in the other ear function well and competently in many everyday listening situations. Reliance on the only hearing ear is often sufficient for many of these people to decline amplification options such as a contralateral routing of signals arrangement or a bone-anchored hearing aid.

Although rare, individuals with a preexisting profound hearing loss in one ear can experience a sudden loss of hearing in a remaining normal-hearing ear (Stahl and Cohen, 2006). The method by which this could typically happen would be through a sudden sensorineural hearing loss (SSHL). A SSHL is defined as a drop of at least 30 dB over at least three frequencies in a period not exceeding three days (Zadeh et al, 2003). The typical explanations for the etiology of SSHL include a viral basis, interruption of blood supply to the cochlea, or a break in Reissner's membrane due to strong physical exertion (Merchant et al, 2005; Oghalai, 2005).

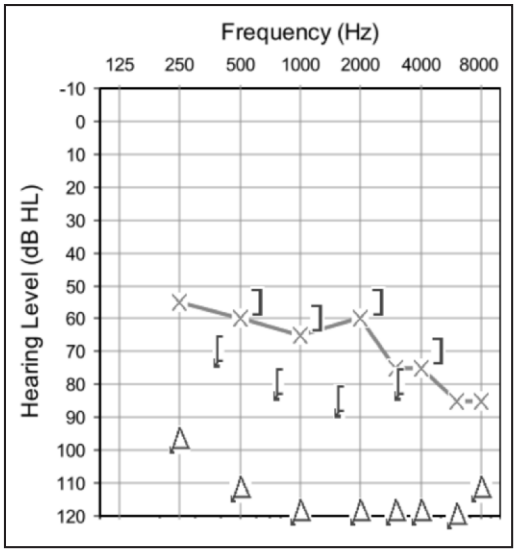

Figure 1. Audiogram of the patient three weeks after the sudden sensorineural hearing loss in the left ear.

When a significant SSHL does occur in an only remaining hearing ear, it is cause for immediate diagnostic and treatment efforts by the audiologist and otologist. For the patient it is a psychosocial-vocational traumatic event that deserves immediate attention. In this article, a case of this type is presented. The audiologic rehabilitative approaches that ensued are discussed.

CASE REPORT

A 62-year-old software product manager came to Mayo Clinic Florida at the request of a local otolaryngologist to be evaluated for a cochlear implant for his left ear. Three weeks earlier the patient experienced a SSHL in his previously normal-hearing left ear upon waking up one morning. His right ear had a profound sensorineural hearing loss since childhood. The local otolaryngologist had prescribed a course of oral steroids, which was now finished. The audiogram that the patient brought from the local otolaryngology office showed a gently sloping 60–80 dB HL sensorineural hearing loss in the left ear and a profound sensorineural hearing loss (no responses at any frequency) in the right ear. It was reported by the audiologist that the patient could repeat only limited spondaic words in the left ear at 85 dB HL, and word recognition could not be tested as the patient could not repeat any monosyllabic words (level unspecified).

The patient reported that the congenital profound hearing loss in his right ear had not posed significant communication problems prior to the SSHL in the left ear: "I grew up with no hearing in the right ear, and I never felt handicapped by it. I learned to hear that way." As a result of the SSHL in the left ear, the patient reported extreme difficulty hearing at work where he was heavily involved in meetings throughout the day, at home, and in social situations. He was highly agitated and very emotional as he talked about the problems he was experiencing. Audiometric findings from our clinic for the patient at three weeks after the onset of the SSHL are shown in Figure 1 and confirmed the previous findings of a moderate to severe sloping sensorineural hearing loss in the left ear and a profound sensorineural hearing loss in the right ear. In contrast to the results obtained at the local otolaryngology office, some word recognition was

now evident in the left ear, with a score of 12 percent correct on a Central Institute for the Deaf (CID) W-22 50-word list presented at 95 dB HL. Poor word recognition is a common finding in patients with SSHL. For example, Banerjee and Parnes (2005) found a mean word-recognition score of 26 percent for 26 patients with SSHL having average pure-tone thresholds of 80 dB HL, and Slattery and colleagues (2005) report a mean word-recognition score of 8 percent (range of 0–52%) for 20 patients having average pure-tone thresholds of 83 dB HL.

The otologic consultation included a detailed history from the patient, otoneurological evaluation, examination of the audiologic report and findings, and a computed tomography scan (magnetic resonance imaging could not be accomplished due to an artificial heart valve). The resulting diagnosis was idiopathic (undetermined cause) SSHL. Given the wide variability in potential recovery of hearing and the time course of any recovery from SSHL, the otologist recommended postponing for one month an evaluation to determine cochlear implant candidacy and referred the patient to the Mayo Hearing Aid Clinic to begin immediate rehabilitative management. This extra month would make the patient approximately two months post-SSHL before the cochlear implant candidacy evaluation would be considered. Yeo and colleagues (2007) report that of patients who recovered from SSHL, 78 percent did so within one month; 6 percent, within one to two months; 13 percent, within two to three months; and 4 percent, in longer than three months.

At the hearing aid–selection appointment the patient indicated a desire to obtain an inexpensive hearing aid for the left ear, as he was sure that he "would end up getting a cochlear implant" and did not want to spend money on a expensive hearing aid that he assumed he would be using for only a few months. This was deemed a reasonable approach, as the patient would be a potential cochlear implant candidate if the pure-tone thresholds and poor word understanding did not show improvement in the near future. As a result, the audiologist recommended an entry-level high-gain flexible six-band digital behind-the-ear hearing aid with a strong telecoil and direct audio input capability. The patient agreed with the recommendation, and the hearing aid and an earmold for the left ear were rush ordered and fit within five days. Future periodic hearing-monitoring appointments were arranged to assess any possible changes in hearing.

At the time of the hearing aid fitting, the patient and his wife were enrolled in a group adult aural rehabilitation program that met weekly for four 1¼ hour sessions. The purposes of these groups have been described by Hawkins (2006); they are intended to develop insight into hearing loss and communication strategies and better emotional acceptance of hearing loss for both the person with the hearing loss and a family member. The group that this patient and his wife attended had 12 other participants (seven hearing-impaired adults and five spouses). The group was conducted in its normal fashion, with no special modifications due to the presence of a patient with the SSHL. Briefly, approximately half of each session is devoted to providing information to the patients, such as how the ear works, causes of hearing loss, understanding their audiograms, speechreading, and so on, and half is spent in group discussions about problems that are encountered, their reactions to these problems, and various strategies for both the patient and family member to better deal with difficult listening situations. At the conclusion of the course both the patient and his spouse reported that the sessions were very beneficial and gave them methods of dealing better with the SSHL. In addition to the group setting during this first month of hearing aid use, approximately two hours of individual discussions and counseling took place during routine hearing aid follow-up visits.

After a month of successful hearing aid use and the adult aural rehabilitation group, the patient completed the International Outcome Inventory for Hearing Aids (IOI-HA [Cox et al, 2000]), the results of which are shown in Figure 2. The patient responses are plotted with the normative values from Cox, Alexander, and Beyer (2003) in Figure 3. The shaded area in Figure 3 represents the middle 50 percent of responses for each item for the group used to create norms for the IOI-HA. (The normative group consisted of 156 hearing aid users from private practice settings with binaural in-the-ear hearing aids that had been used for 6–12 months prior to completing the IOI-HA.) Higher numbers indicate more positive results in each dimension. As

1. Think about how much you used your present hearing aid(s) over the past two weeks. On an average day, how many hours did you use the hearing aid(s)?

None	Less than 1 hour a day	1 to 4 hours a day	4 to 8 hours a day	More than 8 hours a day
☐	☐	☐	☐	☒

2. Think about the situation where you most wanted to hear better, before you got your present hearing aid(s). Over the past two weeks, how much has the hearing aid helped in those situations?

Helped not at all	Helped slightly	Helped moderately	Helped quite a lot	Helped very much
☐	☐	☐	☒	☐

3. Think again about the situation where you most wanted to hear better. When you use your present hearing aid(s), how much difficulty do you STILL have in that situation?

Very much difficulty	Quite a lot of difficulty	Moderate difficulty	Slight difficulty	No difficulty
☐	☐	☒	☐	☐

4. Considering everything, do you think your present hearing aid(s) is worth the trouble?

Not at all worth it	Slightly worth it	Moderately worth it	Quite a lot worth it	Very much worth it
☐	☐	☐	☐	☒

5. Over the past two weeks, with your present hearing aid(s), how much have your hearing difficulties affected the things you can do?

Affected very much	Affected quite a lot	Affected moderately	Affected slightly	Affected not at all
☐	☐	☒	☐	☐

6. Over the past two weeks, with your present hearing aid(s), how much do you think other people were bothered by your hearing difficulties?

Bothered very much	Bothered quite a lot	Bothered moderately	Bothered slightly	Bothered not at all
☐	☐	☐	☒	☐

7. Considering everything, how much has your present hearing aid(s) changed your enjoyment of life?

Worse	No change	Slightly	Quite a lot better	Very much better
☐	☐	☐	☒	☐

8. How much hearing difficulty do you have when you are not wearing a hearing aid?

Severe	Moderately severe	Moderate	Mild	None
☒	☐	☐	☐	☐

Figure 2. International Outcome Inventory for Hearing Aids responses from the patient.

expected with this degree of hearing loss, even with the reduced word-recognition score, the hearing aid was extremely helpful, because without a hearing aid, very little, if any, normal conversational speech was audible. Benefit and quality of life were at the 75th percentile, with the patient reporting that the hearing aid "helped quite a bit" and enjoyment of life was "quite a bit better." Use and satisfaction were given the highest absolute rating, with a reported use of more than eight hours/day and an indication that the hearing aid was "very much worth it." However, probably due to the limited unaided word understanding in the left ear (12% at the most recent test) and the complete loss in the right ear, residual handicaps and restrictions remained. For example, the patient reports on the IOI-HA that he still experienced "moderate difficulty" in situations where he most wanted to hear better and with his hearing aid his hearing difficulties had "affected moderately" the things that he could do.

The first follow-up hearing test occurred approximately one month after the hearing aid fitting and five weeks after the initial evaluation. The results are shown in Figure 4 and indicate a 5–10 dB improvement in several pure-tone thresholds. Word recognition for the left ear was 56 percent (CID W-22 word list presented at 80 dB HL) and represented a significant improvement from the 12 percent score obtained five weeks earlier.

In spite of the improvements in pure-tone thresholds and word understanding, the patient requested that he be evaluated for candidacy for a cochlear implant. The patient was tested using his behind-the-ear hearing aid and achieved the following test results on consonant/nucleus/consonant (CNC) words in quiet, CNC phonemes, Hearing in Noise Test (HINT) in quiet, HINT at +10 dB signal-to-noise ratio (SNR), and aided soundfield thresholds.

- CNC Words in Quiet 56 percent
- CNC Phonemes 61 percent
- HINT in Quiet 85 percent
- HINT at +10 dB SNR 21 percent
- Aided Soundfield Thresholds 25–30 dB HL

The presentation level for all speech signals in sound field was 70 dB SPL.

Based on these results, the cochlear implant team decided that the patient was not a cochlear implant candidate. He was informed that his performance on the above tests led the team to believe that he would not experience significant improvement with a cochlear implant over his current amplification arrangement and therefore he was not a candidate for implantation. Given the poor performance in noise on the HINT (21% at a relatively favorable SNR of +10 dB), it was recommended that he consider an FM system to interface with his hearing aid to assist him in work environments

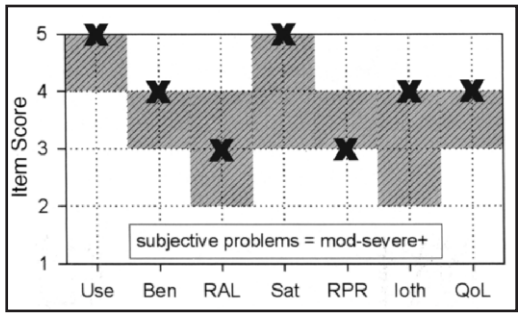

Figure 3. International Outcome Inventory for Hearing Aids results for the patient plotted on the Cox and colleagues (2003) norms. The shaded areas represent the middle 50 percent of responses from the normative group. Higher numbers indicate more positive results on each dimension. Use = use time; Ben = benefit; RAL = residual activity limitations; Sat = satisfaction; RPR = residual participation restrictions; Ioth = impact on others; QoL = quality of life.

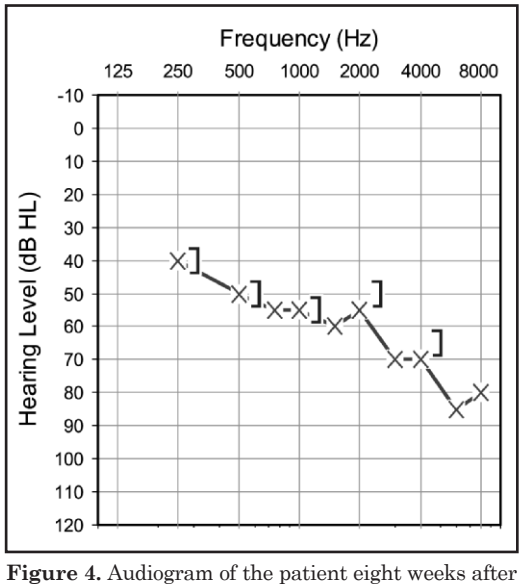

Figure 4. Audiogram of the patient eight weeks after the sudden sensorineural hearing loss in the left ear and four weeks after the hearing aid fitting.

such as meetings and in noisy situations. The patient concurred with these recommendations and returned to the hearing aid clinic to discuss the use of assistive devices to improve his communication abilities.

After further counseling from the audiologist at the hearing aid clinic, the patient did purchase a remote FM microphone and FM receiver and audio shoe for direct input of the FM signal into the behind-the-ear hearing aid. In addition, after discussing his cell phone difficulties, a neck loop with a microphone was provided that could plug into his cell phone and then interface with his hearing aid's t-coil.

The patient returned for follow-up several times in the next year and reported good use of the hearing aid/FM combination, which he used in meetings at work, in restaurants, sometimes with television, and in the car with his wife. He was using the neck loop for both regular and cell phone calls and reported good success.

At approximately 20 months after the SSHL, the patient returned to the hearing aid clinic and stated that although he was doing well and was satisfied with his current hearing aid, FM system, and neck loop, he now wanted to purchase a second hearing aid for the left ear. He indicated that this decision was affected by the realization that he would probably not be obtaining a cochlear implant and he felt the need to have a backup hearing aid in case of the need for a repair or the loss of his current hearing aid. He indicated that he had been reading about advanced hearing aid circuitry on the Internet, and given that price "was not an issue," he expressed a desire to obtain a "high-end digital hearing aid with all available features" to use as his primary hearing aid and intended to relegate his present aid to backup status.

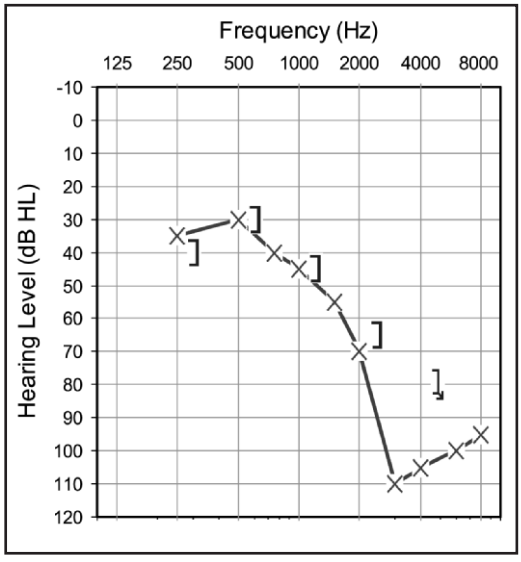

Figure 5. Audiogram of the patient 20 months after the sudden sensorineural hearing loss, showing an improvement in the lower frequencies and a decrease in the higher frequencies.

The patient was counseled that obtaining a second hearing aid as a backup was a good idea but that he should not expect dramatic improvement with a "high-end" hearing aid over his present, less expensive hearing aid. The features that did hold some promise in a new hearing aid would be the utilization of a directional microphone and multiple listening settings. The patient was informed that before a new hearing aid could be obtained, however, a current hearing test was necessary. He insisted that his hearing had not changed and only reluctantly agreed to the test. The new audiogram is shown in Figure 5 and does show a marked change from the previous test obtained 17 months earlier. Better pure-tone thresholds by 5–20 dB were found in the 250–1000 Hz range, and poorer thresholds by 10–40 dB were observed in the 2000–8000 Hz region. Word recognition was unchanged from the previous test and remained at 56 percent (CID W-22 word list presented at 80 dB HL). The pure-tone threshold changes resulted in an otologic referral, after which the patient was cleared to continue with a new hearing aid and was reassured that if further decreases occurred, cochlear implants would be considered if appropriate. The cause of this subsequent improvement in the low frequencies and decrease in the high frequencies is not clear. It is possible that the same mechanism that led to the SSHL was responsible for the decrease in the high frequencies (although this would not explain the low-frequency improvements), as there are reports of further decreases in hearing in 15 percent of patients with SSHL (Monsell et al, 1997).

A high-end digital behind-the-ear hearing aid with extensive frequency shaping (10 bands), advanced directional microphone features (multiband adaptive directional microphone with several pickup pattern options), strong t-coil, and direct audio input capability was then ordered and fit to the patient. An audio shoe allowed connection of the patient's FM receiver to the new

hearing aid and thus allowed continued use of the FM microphone. At a two-week follow-up, he reported that the presence of the directional microphone on the hearing aid had allowed him to perform better in some noisy situations and therefore he did not feel the need to use his FM system as extensively. He appreciated this ability as it allowed him to not "have to carry around as much hardware all the time." In spite of this report of subjective improvement, the IOI-HA administered at four weeks after the fitting of the more advanced hearing aid showed identical results to those obtained earlier with the first hearing aid.

Two months later the patient arrived at the hearing aid clinic with information that he had gathered on the Internet about a Bluetooth device (ELI by Starkey) that would direct input into a hearing aid and allow him to connect to his Bluetooth-enabled cell phone. Given that he used his cell phone extensively, he was interested in such a device. The device was obtained for the patient, and he is now using it successfully and effectively with his cell phone. The patient's hearing is now monitored every six months, and it is emphasized that he should call for testing sooner if he notices any change in his hearing abilities.

DISCUSSION

SSHL in an only hearing ear is obvious cause for immediate action by the otologist and audiologist. The otologist most frequently is interested in placing the patient on oral steroids as soon as possible after the incident occurs. While there is some debate as to the effectiveness of oral steroid therapy from an evidence-based medicine perspective (Ghosh and Jackson, 2005), it is still the primary method of treatment. Some have argued for direct intratympanic injections of steroids (Banerjee and Parnes, 2005), but the data are limited, and a strong recommendation cannot yet be made (Doyle et al, 2004). Regardless of the evidence in the literature, there are probably very few otologists who would not attempt some type of medical therapy. In the case described here, it is not clear that any recovery occurred as a result of the oral steroid therapy, and thus the immediate need for audiologic rehabilitation became critical.

The audiologic rehabilitation needs of a person with SSHL in the only hearing ear are somewhat special, in that immediate emotional support is needed as well as the standard application of amplification. The loss of hearing in an only hearing ear is essentially equivalent to a sudden bilateral hearing loss, and thus the impact on the patient is profound. The emotional support was provided through two main avenues. First, in the initial hearing aid selection and follow-up sessions, ample time was given to the patient to discuss his difficulties and frustrations that were being experienced at work and at home. Approximately two hours of individual discussion and counseling were provided to the patient during this time period. The audiologist is in a unique position to understand these difficulties and frustrations and provide much needed understanding and sympathy in addition to giving suggestions about effective communication strategies. It can also be determined through these discussions whether a recommendation for psychological counseling is warranted. Second, the adult group aural rehabilitation program provided the patient and his spouse access to discussions by other persons with hearing loss about their problems and methods of dealing with the difficulties. Knowing that one is not alone in the struggle for better hearing and adopting better communication strategies can provide much needed comfort and encouragement.

The amplification needs of the SSHL patient can also be somewhat unique. While all of the approaches described with the patient can be considered for any person with hearing impairment, the clear concern about assistive device needs, such as remote FM microphones, neck loops, and Bluetooth devices, is obvious when a patient goes from basically normal everyday functioning to being severely communicatively impaired. Perhaps one lesson from this case is that more patients should be provided information and encouragement to partake of these technological developments that can clearly assist the hearing-impaired person in many aspects of everyday life. Widespread use of such additional equipment may be limited due to the extra cost involved, which fortunately was not an issue with the patient described in this case study. The development of lower-cost assistive devices would be welcome.

A final aspect of this case that is worth

noting is the need for good monitoring strategies for patients with atypical hearing losses. The fluctuations that were observed with this patient, some even without the patient's own perception of a change, were cause for adjustment of the amplification systems and for further medical evaluation.

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