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Sequential Bilateral Cochlear Implantation in a Patient with Bilateral Meniere's Disease

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Abstract

This case study describes a 45 year old female with bilateral, profound sensorineural hearing loss due to Meniere's disease. She received her first cochlear implant in the right ear in 2008 and the second cochlear implant in the left ear in 2010. The case study examines the enhancement to speech recognition, particularly in noise, provided by bilateral cochlear implants.

Speech recognition tests were administered prior to obtaining the second implant and at a number of test intervals following activation of the second device. Speech recognition in quiet and noise as well as localization abilities were assessed in several conditions to determine bilateral benefit and performance differences between ears. The results of the speech recognition testing indicated a substantial improvement in the patient's ability to understand speech in noise and her ability to localize sound when using bilateral cochlear implants compared to using a unilateral implant or an implant and a hearing aid. In addition, the patient reported considerable improvement in her ability to communicate in daily life when using bilateral implants versus a unilateral implant.

This case suggests that cochlear implantation is a viable option for patients who have lost their hearing to Meniere's disease even when a number of medical treatments and surgical interventions have been performed to control vertigo. In the case presented, bilateral cochlear implantation was necessary for this patient to communicate successfully at home and at work.

Keywords

Cochlear implant; hearing aid; Meniere's disease; sensorineural hearing loss; speech recognition; vertigo

INTRODUCTION

Effective communication is necessary for adults to maintain employment, raise a family and enjoy life. Cochlear implantation improves the face-to-face communication of nearly all postlinguistically deafened adults and allows most to regain use of the telephone. For a young working mother, communication is of utmost importance; however, with debilitating vertigo due to Meniere's disease, effective communication may be secondary to functioning in daily life. The physical symptoms of Meniere's disease, vertigo, fluctuating hearing loss, and tinnitus, can be incapacitating and can lead to depression and anxiety (Anderson and Harris, 2001). The prevalence of Meniere's disease in the United States is estimated to be 190 per 100,000 with a higher prevalence for women and older adults (Harris and Alexander, 2010). For patients whose vertigo is not controlled through diet, vestibular

rehabilitation, and/or medication, a number of surgical options exist; however, a risk of decreased hearing may come with surgical treatment (Lustig et al, 2003; Colletti et al, 2007).

Bilateral severe-to-profound hearing loss secondary to bilateral Meniere's disease is relatively rare, but should it occur, cochlear implantation is a viable option (Lustig et al, 2003). Lustig et al (2003) examined the speech recognition abilities of a group of nine cochlear implant (CI) recipients with Meniere's disease; seven of the nine patients had bilateral Meniere's disease. All study patients were implanted unilaterally in the poorer hearing ear, and all had significant improvement in open-set speech recognition scores at six months post initial activation of the CI compared to pre-operative scores with the exception of one patient whose device failed. The speech recognition abilities of the Meniere's group were compared to a group of postlingual CI recipients without Meniere's disease. No significant difference in scores was found between the two groups except for the CID sentence test (Davis and Silverman, 1978) presented in quiet for which the Meniere's group scored significantly higher. The results indicate that cochlear implantation is an option for patients with severe-to-profound hearing loss secondary to Meniere's disease.

The case study presented in this paper concerns a young woman diagnosed with bilateral Meniere's disease that resulted in profound hearing loss, bilaterally. The multiple procedures used to manage her dizziness will be discussed. Hearing rehabilitation included sequential, bilateral cochlear implantation. The results presented illustrate the advantages of bilateral CI use compared to unilateral CI use including improved speech understanding in noise, improved localization skills, and enhanced sound quality.

HISTORY

The patient was first seen at Washington University in St. Louis School of Medicine (WUSM) Department of Otolaryngology in 2007 at the age of 42. She reported having her first episode of dizziness, and fluctuating hearing loss in the right ear in 2000. Several months after this first attack, she began experiencing episodic vertigo lasting several minutes to several hours. After suffering with vertigo and fluctuating hearing loss for a year, she had a right endolymphatic shunt decompression operation performed at another center. The patient reported that the surgery along with medication provided some relief from the dizziness for several years. In 2004, the episodic vertigo began again and became worse during her pregnancy that same year. By 2006, she reported having severe vertigo two to three times per day. She was breast-feeding at that time and was unable to take medication. She stopped breast-feeding in early 2007 and began taking a number of medications daily which decreased the severity of her dizziness; however, she reported having mini episodes of vertigo daily and major episodes at least three times a month.

The patient obtained a hearing aid for the right ear in 2000 when she was first identified with hearing loss in that ear. She was identified with hearing loss in the left ear during a school hearing screening when she was eight years old. The hearing in the left ear had been stable until 2003 when she noticed a decrease in hearing, and at that time, obtained a hearing aid. The etiology of the hearing loss in the left ear was reported to be genetic as her father also had hearing loss as a child. She reported experiencing tinnitus in the right ear in 2000 when her Meniere's symptoms began, and then shortly after, she noticed tinnitus in her left ear as well. The tinnitus was masked by use of the hearing aids and was not bothersome. Her hearing loss in the right ear progressively worsened since being diagnosed with Meniere's disease. The hearing in the left ear also continued to deteriorate. A hearing test performed at WUSM in 2007 revealed a severe-to-profound sensorineural hearing loss in the both ears with 32% and 36% word recognition scores (WRS) on the NU-6 word test (Tillman and

Carhart, 1966) for the right and left ears, respectively. Even though the patient had very poor hearing, her main concern was eliminating her vertigo.

After seeing an otologist/neurotologist (JGN) at WUSM, a sensorineural hearing loss was confirmed in the left ear and active cochleo-vestibular Meniere's disease was confirmed in the right ear. All blood work was normal and an MRI revealed no abnormalities. Binocular video-oculography results revealed intact central ocular motor function. Bithermal caloric responses were strong and symmetric. Computerized Dynamic Posturography revealed low scores on the Sensory Organization Test (SOT) and one fall on SOT 6. This is not consistent with a known pattern but suggests a visual preference which was consistent with the patient's reported history. It was recommended that the patient continue her low-salt diet, begin vestibular exercises, and slowly decrease her medications. The patient, however, continued to have multiple episodes of vertigo daily and reported that she was about to lose her job due to absences related to the Meniere's symptoms.

Over the course of the next several months, the patient had a number of intratympanic gentamicin injections, an endolymphatic sac operation (revision) with shunt and finally a vestibular nerve section in the right ear. One month after the vestibular nerve section, the patient reported that she had not had a vertiginous attack since the surgery although she reported feeling unbalanced. In addition, she reported that four days after surgery she could no longer hear sound when using her right hearing aid. The vestibular nerve section specimen was reviewed by the otologist/neurotologist and only vestibular nerve was found. An audiogram revealed a profound hearing loss in the right ear and a 0% WRS. No change in hearing was seen for the left ear. She was referred to the Adult CI Program at WUSM and began the pre-operative process to obtain an implant for the right ear. The patient continued to report no dizziness since the vestibular nerve section. A hearing test prior to CI surgery indicated slight improvement in hearing for the right ear (severe-to-profound loss, 8% WRS). In April 2008, she was implanted in the right ear with the Nucleus Freedom CI system.

The patient wore the Freedom speech processor on the right ear and a Starkey Destiny 400 behind-the-ear hearing aid (HA) on the left ear and reported doing well in most communication situations. Speech recognition results will be discussed later in this paper. In October 2008, the patient described having mini attacks of vertigo and one major attack which lasted two to three hours. She also reported that her hearing in her left ear was different and somewhat distorted. A hearing test in November 2008 revealed a decrease in hearing for the left ear. The dizziness continued and the patient stated that she could not live through dizziness like she had experienced in the past; therefore, a tympanostomy grommet tube was placed in the left tympanic membrane, and the patient received four injections of dexamethasone through the tube over a one month period. An improvement in the dizziness was reported with only minor attacks of dizziness since the injections. An infection around the tube in January 2009 precipitated its removal. The patient continued to report minor vertiginous episodes daily with a major episode occurring every three to four days. In deference to the patient's request, a left endolymphatic sac operation with shunt was performed in March 2009 after which her vertiginous episodes markedly improved, but the hearing in her left ear decreased to a profound sensorineural loss. Her hearing did not recover, and she discontinued the use of the hearing aid in that ear.

The patient reported to her CI audiologist that she struggled to understand speech at home and at work since she had been using the CI without the HA. At home she needed to communicate with her husband, her teenager, and her three year old. Her work environment included a large office with a number of people and office equipment. She was expected to use the telephone. She reported that speech sounded distorted and that her understanding in

any type of background noise was very poor. An evaluation of her speech recognition scores with the CI revealed no change in performance since discontinuing the use of the HA and substantial open-set speech recognition. The possibility of a CI in the left ear was discussed.

In June 2009, the patient discontinued her medications due to dry mouth and was taking meclizine for dizziness only as needed. She saw the otologist/neurotologist several months later and reported mini episodes of vertigo daily, mainly in the mornings. At that time, she was given a prescription for valium. By March 2010, the patient was considering cochlear implantation for her left ear. She believed that the valium had helped to control her dizziness but continued to report great difficulty communicating especially at work where the background noise was detrimental to her speech understanding. In May 2010, two years after her first CI, the Nucleus 512 CI device was implanted in her left ear. Table 1 provides a general time line of the patient's symptoms and treatments. [TABLE 1]

FINDINGS

After the first CI (right), the patient participated in a longitudinal study examining monosyllabic word recognition over time (Skinner et al, 2007a). Figure 1 shows her consonant-vowel nucleus-consonant (CNC) monosyllabic word scores (Peterson and Lehiste, 1962), using the right CI only, at 12 test intervals from two weeks to two years post initial activation. [FIGURE 1] At two weeks, her word score was 44% and by six months, 87%. Her score, then, reached a plateau and was maintained at that level through the two year test interval. With a score of 87% on CNC words, the patient would be considered a "star performer". She was scoring well above the average score of 62% for a group of 110 postlinguistically deafened adult CI users who participated in the same longitudinal study (Finley et al, 2010). The patient reported that she was pleased with her communication abilities when wearing the CI and HA together; however, about a year after cochlear implantation, she stopped wearing the HA due to a decrease in hearing. When using the CI only, she was no longer satisfied with her ability to communicate in daily life. As can be seen in Figure 1, her CI performance at one year was stable and above average for CNC words. Her difficulties understanding with just the CI were not related to a decrease in performance with the implant. The patient reported that she was comfortable with the CI during face-to-face communication in quiet environments, but when listening in background noise, she found herself struggling to understand. In fact, it was noted at her CI evaluations that her scores in noise were considerably lower than her scores in quiet. At her one year CI evaluation, her score on the TIMIT sentences (Lamil et al, 1986) presented in the sound-field at 60 dB SPL in quiet using the implant only (no lip-reading) was 76%. When the TIMIT sentences were presented at the same level in four-talker babble (Auditec of St. Louis; +8 signal-to-noise [SNR]), she scored 34% using the CI only, a decrease of 42% from listening in quiet to listening in noise. The TIMIT sentences are difficult due to the composition of the talkers, using both male and female speakers, different regional dialects, and varied speaking rates. Furthermore, the language is complex. For example, "His name became synonymous with cold-blooded cruelty" and "Vietnamese cuisine is exquisite" are two TIMIT sentences. Because of the variability of the talkers and complexity of the sentences, the TIMIT sentences are thought to be more representative of real-life listening. It appeared that, indeed, the patient did fairly well with the CI listening in quiet (76%) but as reported, much poorer in noise (34%). Difficulty understanding speech in noise is a common report by CI users (Donaldson et al, 2009). Additionally, research has shown that CI users have a significant decrease in sentence scores at SNRs that would not affect those with normal hearing (Spahr et al, 2007). Because of the communication difficulties she encountered in daily life, the patient expressed a strong interest in obtaining a second CI.

As mentioned above, two years after obtaining the Nucleus Freedom CI for the right ear, the patient was implanted with the Nucleus 512 CI in the left ear. She was programmed using the Advanced Combination Encoder (ACE; Skinner et al, 2002) speech processing strategy (pulse width = 25- μ s/phase, monopolar stimulation) and 1800 pulses per second /channel (pps/ch) stimulation rate in the first implanted ear. At the initial activation of the second implant, a speech processor program or map was created using similar parameters to those used with the first CI (ACE strategy, 1800 pps/ch stimulation rate). The patient was familiar with the procedures used to program the speech processor. No programming issues were encountered and the maps created appeared typical. The patient scored 100% when simple sentences were presented live voice using the second CI only. Three days after initial activation of the second CI, the patient reported having a “full blown” vertiginous attack that lasted all day. She had not reported having an attack of vertigo since her left endolymphatic sac operation with shunt was performed several months prior to the second CI surgery. For the next month, the patient reported feeling unsteady to the point of having to remove the second CI speech processor every couple of days to recover.

The patient wore the first CI speech processor at a volume setting of 9 (range: 1–9) and sensitivity setting of 12 (range: 1–20), but in order to prevent feeling unsteady, the patient wore the second CI speech processor at a softer level, volume of 4 and sensitivity of 10. She reported that the first CI was louder than the second CI; however, she noted an improvement in speech understanding when the two were worn together. Furthermore, her CNC word score using the second CI only was 75% at her one month evaluation. Five weeks post-initial activation of the second CI, the patient could consistently wear both processors but still needed to keep the volume and sensitivity of the second CI speech processor at lower settings. In an attempt to equalize loudness between the two ears without causing further dizziness, the stimulation rate of the second CI was changed from 1800 pps/ch to 500 pps/ch. The stimulation rate used with the first CI remained at 1800 pps/ch. After using the 500 pps/ch map for two weeks, the patient reported that the unsteadiness was not occurring as frequently and that she was able to increase the volume to 5. She reported that the 500 pps/ch map sounded louder than the previous 1800 pps/ch map; moreover, she appreciated the longer battery life that the 500 pps/ch map provided. She stated that when she first used the 500 pps/ch map with the second CI and the 1800 pps/ch map with the first CI, there was a difference in sound quality between the ears. After several weeks of continued use, she no longer noticed a difference in sound quality when wearing both the right and left speech processors together. Table 2 shows the speech processor parameters that are currently used by the patient for each ear. [TABLE 2]

Figure 2 shows the placement of the electrode array within the cochlea for the first and second CI. [FIGURE 2] CT scans were obtained both pre-operatively and post-operatively to create these images using a technique described by Skinner et al, 2007b. The rectangular black and white marks within the cochlea represent the 22 electrodes of the Nucleus internal device (Nucleus Freedom with Contour Advance Electrode, first CI; Nucleus CI512 cochlear implant with Contour Advance Electrode, second CI). Ideally, all electrodes should be in scala tympani. The 3D reconstruction of the CT scans, which is based on a cochlear atlas model (Teymouri et al, 2010), indicate that for the first CI (right), thirteen electrodes are located in scala tympani, and nine electrodes are involved in a transition from scala tympani through scala media to scala vestibuli with the two most apical electrodes in scala vestibuli. All electrodes remain in scala tympani for the second CI (left). Note that the electrodes are closer to the modiolar wall of the cochlea for the second CI compared to first CI; consequently, electrodes may be in closer proximity to spiral ganglion cells with the second CI compared to the first CI.

When creating a CI speech processor program or map for the Nucleus device, two levels need to be set for each of the active electrodes, a minimum and a maximum electrical stimulation level. For the Nucleus device, the minimum electrical stimulation levels are commonly referred to as T-levels and maximum electrical stimulation levels are commonly referred to as C-levels. T-levels and C-levels are set based on the each patient's perception of loudness. The technique used to set T- and C-levels can vary between CI centers, but in general, T-levels are set at a percept of "very soft", and C-levels are set at a percept of "loud but comfortable". Both levels can be adjusted to allow soft speech and sound to be audible and conversational speech comfortably loud in daily life. T- and C-levels are different for each patient and can vary between ears as well as across the array within a single ear for the same patient. The T- and C-levels used in the patient's map for each ear (1800 pps/ch stimulation rate) were averaged across electrodes. For the first CI, averaged T- and C-levels were 178 and 209 (range: 1–255) current levels¹, respectively. For the second CI, averaged T- and C-levels were 85 and 128 current levels, respectively. This is a substantial difference in T- and C-levels between ears when using the same stimulation rate and may be related to the difference in the placement of the electrode arrays within the cochlea for the two implants.

Prior to the second CI surgery, the patient agreed to participate in a longitudinal study evaluating performance after sequential, bilateral cochlear implantation. Subjects in this study are tested before the second CI surgery (pre-operatively) and at one, three, six, nine, twelve, fifteen and eighteen months post-initial activation of the second CI (post-operatively). Pre-operative testing is carried out with the patient using a CI in one ear and a HA in the other ear. Post-operative testing is completed with the patient using an implant in each ear. Prior to speech recognition testing, each patient's external equipment is checked and frequency modulated (FM) tone, sound-field threshold levels from 250–6000 Hz are obtained. If a patient reports changes in speech understanding or sound-field threshold levels are inconsistent with those previously obtained, a check of the map is performed with reprogramming as necessary. Testing through six months has been completed for this patient. FM tone, sound-field threshold levels obtained at the six-month test interval are shown in Figure 3 for each ear. [FIGURE 3] The patient's sound-field threshold levels with each CI are 20 dB HL across the frequency range ensuring the audibility of soft speech and sound (Holden et al, 2007, Davidson et al, 2009).

The test materials used to evaluate speech recognition performance for this study are difficult. Many of the tests are presented in noise or at soft presentation levels in order to replicate difficult listening situations that CI users encounter in daily life. Since the patient reported experiencing difficulty understanding speech in noise when using her first CI alone, the test results will predominately focus on her performance in noise. Two lists of each test were presented in a pseudo-randomized order at each test interval. All testing was performed in the sound-field. The majority of tests were performed with the patient seated at 0° azimuth approximately 1.5 meters from a loudspeaker. For the R-SPACE™ test environment (Revit et al, 2002; 2007) and the localization test (Potts et al, 2009), the loudspeaker configuration was different and will be described below.

Figure 4 shows CNC monosyllabic word scores for each ear separately at each study test interval (Pre-operatively: CI and HA; Post-operatively: first CI and second CI). [FIGURE 4] Pre-operatively, the patient scored 91% with the first CI and 0% with the HA. Post-operatively, scores with the second CI progress quickly and are higher than scores with the first CI at the three and six month test intervals.

¹Current level represents the amount of electrical current delivered to electrodes and is expressed in clinical programming units. For a more detailed description, see Cochlear™ Clinical Guidance Document (February, 2010) page 4.

The next several figures show results for each test interval in three conditions. Pre-operative scores are shown with the patient using the first CI only, the HA only, and then both together (bilateral condition). It is important to remember that at the pre-operative test interval, the patient had been using the first CI (right) for approximately two years. Post-operative scores are shown for the first and second CI separately and bilaterally. Scores for TIMIT sentences presented at 60 dB SPL in noise (four-talker babble, +8 dB SNR) are shown in Figure 5. [FIGURE 5] Note the improvement in the bilateral scores from the pre-operative to the six-month test interval. In addition, the patient's scores in noise are higher with the second CI than with the first CI at all test intervals. It is interesting to note an improvement in the noise score with the first CI at the six-month test interval compared to the pre-operative and three-month test intervals. At the six-month test interval, the patient had been using the first CI for approximately 2.5 years.

The Bamford-Kowal-Bench Speech in Noise Test (BKB-SIN; Killion et al, 2004) is a sentence test in noise where the SNR varies throughout the test. The first sentence is presented at an SNR of +21 dB, and for each consecutive sentence, the SNR decreases by 3 dB. An SNR-50 score is obtained; that is, an SNR for which the subject can correctly repeat the key words within the sentences 50% of the time. The BKB-SIN was presented with the speech and noise (four-talker babble) coming from the same loudspeaker located in front of the patient. In addition, the noise was presented through a loudspeaker located 90° to the left of the patient as well as 90° to the right of patient with the speech always presented through the front speaker. Scores obtained when speech and noise were presented through the front speaker are shown in Figure 6 for the pre-operative, three and six-month test intervals. [FIGURE 6] The score is an SNR; therefore, a lower number represents a decrease in SNR and a better result. In other words, a low SNR means that the listener can understand speech in a greater amount of noise. Recall that pre-operatively, scores were obtained with the first CI, a HA, and both together (bilaterally), whereas post-operatively, scores were obtained with each CI separately and both together (bilaterally). The patient could not do the task when using the HA only. The scores improved considerably, especially in the bilateral condition, by the six-month test interval. At each postoperative test interval, the SNR is lower with the second CI than with the first CI. Again, note the improvement in SNR with the first CI at the three-month and six-month test interval following implantation of the second ear.

Figure 7 illustrates the benefits of bilateral cochlear implantation when noise was presented to the sides of the patient. [FIGURE 7] The left side of Figure 7 shows the SNR obtained when noise was presented 90° to the right of the patient. Pre-operatively, the patient was listening with the first CI (right) only and with the first CI and HA. Post-operatively, the patient was listening with the first CI only and with both implants. The right side of Figure 7 reveals the SNR obtained when noise was presented 90° to the left of the patient. Pre-operatively, the patient was listening with the HA in the left ear and then with the HA and first CI together. Post-operatively, the patient was listening with the second CI (left) only and with both implants. A substantial decrease in SNR was seen in the bilateral condition from the pre-operative to the six-month test interval regardless of the direction of the noise. When the noise was presented on the right side, an improvement in SNR of 12 dB was obtained by the addition of the second CI (left). When noise was presented to the left of the patient, an improvement in SNR of 9 dB was obtained when using two implants. When the noise was presented to the left, the SNR with the second implant (left) was lower, at each post-operative test interval, than the SNR with the first implant (right) when noise was presented to the right.

The R-SPACE™ test environment is shown in Figure 8. [FIGURE 8] The patient was seated in the center of eight loudspeakers through which restaurant noise was presented. The

speech stimuli were paired HINT sentence (Nilsson et al, 1994) lists presented from the loudspeaker directly in front of the patient. The noise was fixed at 60 dB SPL and the sentence presentation level was varied depending upon the patient's response. An adaptive procedure in which the sentence presentation level was decreased for each correct response and increased for each incorrect response was utilized. For sentences 1–4, a 4 dB step size was used and then for sentences 5–20, a 2 dB step size was used. An SNR at which the patient could correctly repeat 50% of the sentences was determined by averaging sentence presentation levels for sentences 5–20 and a calculated presentation level for sentence 21. Figure 9 shows the considerable decrease (improvement) in SNR from the pre-operative to the six-month test interval for all three listening conditions. [FIGURE 9] A +6 dB SNR was obtained in the pre-operative bilateral test condition (HA and CI), whereas a -2 dB SNR was obtained in the bilateral test condition (both CIs) at six months. Note that for each post-operative test interval, a lower SNR is obtained with the second CI than with the first CI. Additionally, note the improvement in SNR, with the first CI at both the three-month and six-month test intervals compared to the pre-operative SNR.

In addition to difficulties understanding speech in noise, many unilateral CI users report difficulty localizing sound. To evaluate localization skills, testing was performed pre-operatively using the CI and HA and post-operatively using both implants. The test environment consisted of a 140° loudspeaker array with 15 loudspeakers, 10° apart. Figure 10 is a schematic of the test configuration. [FIGURE 10] The localization task required the patient to state the perceived location of the stimulus (i.e. loudspeaker number 1–15). The patient faced the front (0° azimuth) prior to initiation of each trial, but was permitted to turn her head (i.e., turn toward the loudspeaker from which the word was perceived) during the trial. An equal number of words were presented from each of 10 selected positions; five of the visible loudspeakers were inactive ($\pm 60^\circ$, $\pm 40^\circ$ and 0°), but the patient was not aware of this. The words were presented at a roving level of 60 dB SPL (± 3 dB SPL). The scores are presented in terms of degree of RMS error. The RMS error is the mean deviation of the patient's response from the actual location of the presentation, and it is not related to the direction of the deviation (Potts et al, 2009). Pre-operatively, the patient obtained an RMS error of 59° , and at the six-month test interval, an RMS error of 26° degrees was obtained. An RMS error of 0° represents 100% accurate localization; therefore, the patient's localization ability improved using two implants compared to using a HA and CI.

Even though this patient was scoring near 90% on the CNC word test with her first implant, she stated that she struggled to communicate in daily life when using only one CI. The results of the testing described above revealed that this patient obtains considerable benefit from bilateral cochlear implantation; however, these speech tests were all performed in a sound booth. No matter how difficult and varied the testing, the multitude of listening environments encountered in daily life can not be replicated in a sound booth. To better understand the benefits of bilateral implantation in different listening environments in daily life, the Speech, Spatial and Qualities of Hearing Scale (SSQ; Gatehouse and Noble, 2004) was given at each test session. The test consists of three scales. The Speech scale questions the patient's abilities to understand speech in a variety of difficult listening situations. A question from the Speech scale is as follows: "You are talking with one other person and there is a TV on in the same room. Without turning the TV down, can you follow what the person you're talking to says?" The Spatial scale concerns spatial hearing or the ability to tell the direction and judge the distance of the sound. A question from the Spatial scale is as follows: "You are outside. A dog barks loudly. Can you tell immediately where it is without having to look?" The Sound Qualities scale of the SSQ questions a patient's ability to segregate sounds. That is, can a patient recognize each of two sounds that are going on at once, for example water running and a radio playing? Additionally, this third scale examines the "naturalness" and "clarity" of sound and determines the amount of effort the patient

exerts to listen (“Do you have to put in a lot of effort to hear what is being said in conversations with others?”). For each item on the SSQ, the patient provides a rating on a continuum from 0–10 with a higher number indicating greater ability. The patient was able to see her ratings from the previous test interval while completing each SSQ. Figure 11 indicates the average rating across items for each of the three scales. [FIGURE 11] An improvement from the pre-operative test interval, where the patient was using one CI, to the six-month test interval (bilateral CIs) was seen for all scales with the greatest improvement seen for the Spatial scale.

DISCUSSION

Fortunately, the procedures used to treat this patient’s Meniere’s disease successfully alleviated her symptoms. She reported experiencing vertigo several days after the initial activation of the second CI and continued to report imbalance for the next five weeks. Since that time, she reports no dizziness or imbalance. She is able to work full time as well as take care of her family; furthermore, she is satisfied with her ability to communicate at work and at home when using both implants.

Prior to the profound hearing loss in her left ear, the patient was able to use the first CI and a HA to successfully communicate in daily life. When she was no longer able to use the HA, she reported struggling to communicate in all but quiet, face-to-face listening situations. The patient’s report is consistent with the literature describing the importance of symmetric hearing. Individuals with asymmetric hearing have more difficulty localizing sound, segregating sounds and understanding speech in noise than individuals with symmetric hearing (Firszt et al, 2008). Even though the patient did not have complete symmetric hearing with the CI and HA, she was able to make use of the input from both devices to understand speech in a variety of listening situations encountered in her daily life. The loss of the use of the HA, creating greater asymmetry between the ears, proved detrimental to speech understanding in difficult listening environments. The addition of the second CI restored symmetric hearing. The test results reported here reveal substantial improvement in the patient’s speech understanding in noise and in her localization abilities when using bilateral implants compared to using only one implant or an implant and a HA.

The patient’s performance with each implant is excellent with scores of 90% and 98% on the CNC word test with the first and second implants, respectively. Her above average performance is consistent with the findings of Lustig et al (2003). These researchers described significant improvement in speech recognition for a group of nine CI recipients diagnosed with Meniere’s disease. Five of these patients had undergone surgical procedures (endolymphatic shunt and vestibular nerve section) to control vertigo yet had significant open-set speech recognition with the CI. None of the study patients were experiencing vertigo at the time of CI surgery and only one of the nine patients had post-operative vertigo which resolved within several months of surgery. The dizziness was not related to electrical stimulation. Fina et al, (2003) performed a retrospective study examining the incidence of dizziness after cochlear implantation for a group of 75 patients. The researchers found that only one patient experienced vertigo due to electrical stimulation; however, for this patient, the implant was placed in the vestibule due to complete ossification of the cochlea. The results of the study did indicate, however, a higher probability of post-operative dizziness for patients with pre-operative diagnosis of Meniere’s disease. The positive outcomes shown in the study by Lustig et al (2003) as well as the positive outcome for our patient demonstrates that cochlear implantation is a viable option for patients with bilateral Meniere’s disease and bilateral cochlear implantation should be considered. Patients should be counseled on the possibility of post-operative dizziness.

As described above, the patient is doing well with each implant; however, it is interesting to note the rapid progress made in speech recognition with the second implant (left ear) even with the reported long-term hearing loss (identified at age eight), dizziness and unsteadiness presumably caused by electrical stimulation of that ear. Moreover, the patient's speech recognition in noise is better when using the second CI compared to the first CI (right ear). A possible explanation for the rapid progress and better noise scores with the second CI may be the placement of the electrode array within the cochlea with regard to scalar placement and proximity to the modiolar wall. Skinner et al (2007a, b) and Finley et al (2008) reported that higher monosyllabic word recognition scores are correlated with scala tympani placement of the electrode array. For this subject, all electrodes remained in scala tympani with the placement of the second CI, whereas the nine apical-most electrodes transitioned from scala tympani to scala vestibuli during placement of the first CI perhaps damaging cochlear tissues and possibly increasing cross turn stimulation for these electrodes (Finley et al, 2008). The patient did not report aberrant percepts for any electrodes while programming the first CI, and we have found no correlation between aberrant percepts and scala vestibuli placement in our patient population. The electrode array of the second CI is closer to the modiolar wall of the cochlea than the electrode array of the first CI (see Figure 2) which places the electrodes of the second CI nearer spiral ganglion cells and may explain the lower minimum and maximum electrical stimulation levels across electrodes for this device. Closer wrapping of the array around the modiolar wall has also been observed to correlate with better speech recognition (Finley et al, 2010).

A number of procedures were performed to alleviate the patient's dizziness including endolymphatic sac surgery with shunt (right and left ears), intratympanic gentamicin injections (right), intratympanic dexamethasone injections (left) and vestibular nerve section (right). It may be the case that the procedures used caused greater trauma to the peripheral auditory system in the right ear than in the left ear; though, the audiograms for both ears were similar prior to CI surgery. Intratympanic gentamicin injections and vestibular nerve section were used to control the patient's dizziness in the right ear. Both have been shown to cause hearing loss with intratympanic gentamicin injections causing greater degree of hearing loss than vestibular nerve section (Colletti et al, 2007). After a middle fossa complete vestibular nerve section in the right ear, this patient was relieved of the severe episodic vertigo and maintained hearing but did have a considerable reduction in thresholds and in the word recognition score. Later, the patient had a right CI. Intratympanic dexamethasone was used in the left ear to control dizziness. Yilmaz et al (2005) treated 26 patients with intratympanic dexamethasone injections to alleviate tinnitus and found no differences in patient's pure tone averages before or after the injections. Additionally, the study found that outer hair cell function, measured via transient-evoked otoacoustic emission testing, was not affected by dexamethasone treatments. No deterioration in hearing was documented after intratympanic dexamethasone injections for our patient; however, her dizziness continued after treatment. An endolymphatic sac operation with shunt was then performed after which a significant decrease in hearing was documented. The patient obtained a second CI in the left ear about a year after this decrease in hearing. Her vertigo was under control at the time of the second CI surgery.

As with all CI recipients, a number of factors contributed to this patient's speech recognition abilities, and we can only speculate as to why she has somewhat better performance with the second CI than with the first CI. It is noteworthy that a 20% improvement on TIMIT sentences in noise was seen for the first CI at the six-month test interval (see Figure 5). At this test interval, the patient had been using her first implant for approximately 2.5 years. Learning effects may have contributed to the improvement in scores; although learning effects were not evident prior to implantation of the second ear. For example, at her one year evaluation with the first CI, her score on TIMIT sentences in noise was 34% which is

consistent with her scores at the pre-operative and three-month study test intervals (37% and 38%, respectively). Improvements in scores with the first CI after second-side implantation were also observed for the BKB-SIN and the R-SPACE™ measures (see Figures 6 and 9, respectively). Learning effects may have contributed to the improvement as these test measures were not used with the patient prior to this study. Then again, perhaps listening with two implants has helped improve her ability to understand speech in noise with the first implant. If speech cues are more salient in noise with bilateral input, the patient may have been able to learn to use these same cues to improve her understanding in noise in the unilateral condition. Furthermore, given the many crossed and uncrossed auditory pathway connections, conceivably strengthened bilateral projections could influence monaural connections and ultimately function.

Another contributor to the improvement in noise scores with the first CI may be related to the speech processor program or map used at the six-month test interval. The C-Levels on all electrodes with the first CI were high (209 current levels averaged across electrodes), and the patient used the maximum volume setting of 9. C-levels on a number of electrodes were reaching maximum voltage compliance levels allowed by the software. When maximum voltage compliance is reached, the implant is not able to deliver the required current level for electrodes that are at or above the compliance limit. This can result in the absence of loudness growth and distortion of sound (Cochlear™ Clinical Guidance Document). In an effort to equalize the loudness between the ears, the CI audiologist had the patient decrease the volume of the first CI speech processor after the three-month test interval. The patient continued to use the same map with the lower volume setting in daily life and was tested with this map and setting at the six-month test interval. It is suspected that the lower volume setting may have helped to improve her understanding of speech in noise by making sound less distorted. With the Nucleus device, lowering the volume decreases C-levels. Perhaps reaching maximum voltage compliance levels on a number of electrodes caused distortion that may have improved with a lower volume setting and consequently lower C-levels.

The difference in stimulation rates used with the first CI (1800 pps/ch) and second CI (500 pps/ch) is unusual. The majority of bilateral implant patients at WUSM are sequentially implanted and use the same stimulation rate for both implants. At WUSM, a number of stimulation rates are tried with each patient at programming sessions during the first six weeks after initial activation of the CI. After experience with a number of stimulation rates, the patients choose which rate they prefer. A stimulation rate of 1800 pps/ch was chosen by this patient with the first CI as providing the best speech understanding and sound quality in daily life. The same stimulation rate was then used at the initial activation of the second implant; however, the patient experienced dizziness that was controlled by keeping the volume and sensitivity at lower levels than what she felt was needed to provide equal loudness between the two implants. Lowering the stimulation rate to 500 pps/ch with the second CI allowed the patient to raise the volume from 4 to 5 and made speech and sound louder without dizziness. She quickly became accustomed to the sound quality with the two different rates. She declined to try a lower stimulation rate with the first CI reporting that she liked the sound quality when the two implants were worn together. The Nucleus device provides audiologists a number of programming parameters to manipulate in order to improve sound quality and speech understanding. These parameters can and should be utilized in order to maximize CI benefit for each patient.

The test results presented show that the patient is doing well on a number of difficult speech recognition measures presented in quiet and noise, especially in the bilateral condition, yet these tests can not completely replicate the difficult listening environments encountered in daily life. The patient's responses to the items on the SSQ provide a more realistic view of her perceived everyday communication abilities. As seen in Figure 11, there is an

improvement from the pre-operative test interval to the six-month test interval in her averaged ratings across test items for each of the three scales; however, her average rating is only a five for the Speech scale. The Speech scale questions the patient's ability to understand in difficult listening environments. If the patient believed she was able to hear and understand perfectly in every situation on the SSQ, the average rating would be ten. The ratings for both the Spatial and Sound Qualities scales are higher but are still at an average rating of six. The SSQ data show that the patient's communication abilities have improved since being implanted bilaterally and continue to improve over time. It will be interesting to see if the perceived improvement in her communication abilities continues past six months or if it plateaus. Bilateral cochlear implantation has provided this patient with a substantial improvement in her speech understanding compared to unilateral cochlear implantation; however, the results of the SSQ clearly indicate that there are situations in which she has considerable difficulty. Research focusing on improving cochlear implant recipients' speech understanding in difficult listening situations is warranted.

CONCLUSION

This case study focused on a patient with bilateral, profound sensorineural hearing loss secondary to bilateral Meniere's disease. Her debilitating vertigo was controlled after a number of procedures on both the right and left ears including endolymphatic sac surgery with shunt (both ears), intratympanic gentamicin injections (right), intratympanic dexamethasone injections (left), and vestibular nerve section (right). To improve her hearing and speech understanding, the patient obtained a cochlear implant in her right ear and was able to continue to wear a hearing aid in the left ear. She reported to be satisfied with her communication abilities when wearing the CI and HA together. Her hearing deteriorated in the left ear after an endolymphatic sac operation, and she discontinued the use of the HA due to lack of benefit. She reported great difficulty communicating in daily life and especially in noisy environments when using the CI only. A second CI was implanted in her left ear two years after the first CI. Monosyllabic word scores of 90% and 98%, for the first and second implants, respectively, revealed above average speech recognition with each CI. Speech recognition testing in noise revealed a substantial improvement in scores when using two implants compared to scores using either implant alone. Her localization abilities improved when using bilateral implants compared to using a CI and a HA. In addition, the patient's responses to the SSQ indicated that bilateral cochlear implantation improved her ability to communicate in difficult listening situations, improved her localization of speech and sound, enhanced sound quality, and diminished the effort needed to understand speech in daily life. Bilateral cochlear implantation appears to be a viable option for patients with bilateral Meniere's disease whose dizziness has been controlled.

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Abbreviations

ACE advanced combination encoder

BKB-SIN	Bamford-Kowal-Bench Speech in Noise Test
CI	cochlear implant
CID	Central Institute for the Deaf
C-Level	maximum electrical stimulation level
CNC	consonant-vowel nucleus-consonant
CT	computed tomography
FM	frequency modulated
HA	hearing aid
HL	hearing loss
LE	left ear
μs/phase	microsecond per phase
NU-6	Northwestern University Auditory Test No. 6
pps/ch	pulses per second per channel
RE	right ear
RMS	root mean square
SM/SV	scala media/scala vestibuli
SNR	signal-to-noise ratio
SOT	sensory organization test
SPHL	severe-to-profound hearing loss
SSQ	Speech, Spatial and Qualities of Hearing Scale
ST	scala tympani
T-Level	minimum electrical stimulation level
WRS	word recognition score
WUSM	Washington University in St. Louis School of Medicine

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Figure 1.
CNC monosyllabic word scores obtained at twelve test intervals from two weeks to two years post-initial activation of the first CI.

Figure 2.

CT-based images of the placement of the Nucleus Contour Advance electrode array within the cochlea for the first and second cochlear implants. The black and white rectangular marks represent each of the 22 electrodes of the Nucleus device. Black marks represent electrodes within scala tympani (ST) and white marks indicate electrodes involved in a transition from scala tympani through scala media (SM) to scala vestibuli (SV).

Figure 3. Frequency-modulated (FM) tone, sound-field threshold levels obtained from 250–6000 Hz at the six-month test interval with the first and second CI.

Figure 4.

CNC word scores obtained at four test intervals. Pre-operatively (prior to the second CI), the patient was tested using the first CI only and the HA only. Post-operatively (1 mo, 3 mos and 6 mos), the patient was tested using each implant separately.

Figure 5.

TIMIT sentences in noise scores obtained at three test intervals. Pre-operatively (prior to the second CI), the patient was tested with the first CI only, the HA only, and the CI and HA together. Post-operatively (3 mos and 6 mos), the patient was tested with each implant separately and together.

BKB-SIN (Speech Front, Noise Front)

Figure 6.

The average SNR obtained for the BKB-SIN test when sentences and noise were presented from a loudspeaker located in front of the patient. Pre-operatively, the patient was tested using the first CI only, the HA only, and the CI and HA together. Post-operatively, the patient was tested using each implant separately and together.

Figure 7.

The average SNR obtained for the BKB-SIN test when sentences were presented from a loudspeaker located in front of the patient and noise was presented 90° to the right and 90° to the left of the patient. Pre-operatively, when noise was presented to the right, the patient was tested with the first CI and with the CI and HA together. When the noise was presented to the right at three and six months, the patient was tested with the first CI and with both implants together. Pre-operatively, when noise was presented to the left, the patient was tested with the HA only and with the HA and first CI together. When noise was presented from left at three and six months, the patient was tested using the second CI only and both implants together.

Figure 8.
Schematic of the R-SPACE™ test environment.

Figure 9.

The average SNR obtained during R-SPACE™ testing. Pre-operatively, the patient was tested with the first CI only, with the HA only, and with both together. At all post-operative test intervals, the patient was tested with each CI separately and together.

Figure 10. Schematic of the localization test environment. The boxes in gray represent the loudspeakers through which no sound was presented.

Figure 11.
The average rating across test items for each of the three scales in the Speech, Spatial and Qualities of Hearing Scale (SSQ).

Table 1

General time line of the patient's symptoms and treatments

Year	Age	Symptoms/Treatments
2000	35	1 st episode of vertigo/fluctuating hearing/tinnitus (RE)
2001	36	Endolymphatic shunt decompression (RE) along with medication provides relief from vertigo
2004	39	Episodic vertigo recurs
2006	41	Severe vertigo 2 to 3 times per day
2007	42	Bilateral SPHL documented
2007–2008	42–43	Intratympanic gentamicin injections, endolymphatic sac operation (revision), vestibular nerve section with subsequent profound HL (RE)
April, 2008	43	Vertigo much improved; CI RE
Oct., 2008	43	Episodic vertigo; Decrease in hearing LE
Nov., 2008	44	Intratympanic dexamethasone injections (LE); Some improvement in vertigo
Jan., 2009	44	Severe vertigo every 3 to 4 days
March, 2009	44	Endolymphatic sac operation; Profound HL (LE) following surgery; Discontinued HA use
June, 2009	44	Improvement in vertigo; Meclizine and valium as needed
March, 2010	45	Vertigo under control; Considering CI LE
May, 2010	45	CI LE

CI = cochlear implant; HA = hearing aid; HL = hearing loss; LE = left ear; RE = right ear; SPHL = severe-to-profound hearing loss; WUSM = Washington University School of Medicine

Table 2

The patient's CI device and speech processor program parameters for each ear.

Ear	Cochlear Implant System	Electrode Array	Speech Processor	Strategy	Stimulation Rate/Channel (pps/ch)	No. of Maxima	Total Stim. Rate (pps)	No. of Active Electrodes
1 st CI Rt Ear	Nucleus Freedom	Contour Advance	Freedom	ACE	1800	8	14,400	19
2 nd CI Lt Ear	Nucleus System 5	Contour Advance	810	ACE	500	8	4,000	20