Revision Cochlear Implantation for Facial Nerve Stimulation in Otosclerosis

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Objective: To find if patients experiencing postsurgical facial nerve stimulation caused by underlying disease process (ie, otosclerosis) can improve their hearing performance with their cochlear implant by reimplantation and by an optimal programming strategy.

Design: Retrospective analysis.

Setting: Academic tertiary referral center.

Patients: Two cochlear otosclerosis patients with resistant facial nerve stimulation (FNS). Both patients were initially implanted with Nucleus 22 devices (Cochlear Corporation, Englewood, Colo) and they developed FNS after a period of use. Owing to the decreasing number of active electrodes, concurrent decreases in speech understanding occurred.

Interventions: Various programming approaches were used to address the FNS. Both subjects ultimately received Nucleus 24 devices. One was reimplanted in the same ear, and the other was implanted in the opposite ear. Both have been followed up for 8 months following the reimplantation.

Main Outcome Measures: Cochlear implant programming levels, cochlear implant performance, and facial nerve stimulation.

Results: The FNS was managed for more than 3 years through optimized programming. However, the FNS progressed until performance dropped below acceptable levels. Reimplantation was believed to be the only option for improvement. After reimplantation and programming, both subjects showed immediate improvement in speech discrimination. One user increased his consonant-nucleus-consonant word score from 12% preoperatively to 42%, and the other's performance increased from 0% to 86%.

Conclusions: Our results suggest that having more programming options with newer devices is critical in otosclerotic or ossified users who experience FNS. Also, reimplantation may be a useful tool to improve performance.

unsuccesful are presented. The objective of this study was to find if patients experiencing postsurgical FNS caused by otosclerosis can improve their performance with their cochlear implant by reimplantation and by an optimal programming strategy.

**METHODS**

This was a retrospective study approved by the appropriate institutional review board at the University of Miami, Miami, Fla. The procedures followed were in accordance with the ethical standards of the Helsinki Declaration.

**RESULTS**

**CASE 1**

A 60-year-old male patient was referred to our clinic with bilateral profound hearing loss in December 1994. He was using a hearing aid on the right side. He had a history of left middle ear exploration with stapes mobilization in 1989 at another institution. His speech awareness and speech reception thresholds were 85/95 dB hearing level (HL) for the right and 80/95 dB HL for the left ear, respectively. His pure tone average was higher than 105 dB HL for the right side and higher than 107 dB HL on the left. His discrimination Northwestern University 6 (NU-6) word test scores were 30% for the right ear and 10% for the left. A computed tomographic (CT) scan of the temporal bones revealed indistinct left cochlear walls, consistent with cochlear otosclerosis. The right cochlea appeared normal. With the intent of improving the patient’s hearing without the need for cochlear implantation, a left stapedectomy was planned. During that operation, it was noted that the stapes was indeed mobile and no stapedectomy was performed. Because the patient had some residual hearing on the right and had some benefit from his hearing aid on that side, it was decided to implant a device in his left ear. He received a Nucleus 22 cochlear implant (Cochlear Corporation, Englewood, Colo) on the left side in March 1995. During the procedure, it was observed that there was an extensive amount of new bone formation in the cochlea, and during cochleostomy, a distance of 8 mm had to be drilled out to reach the scala tympani. All 22 electrode rings and 4 stiffening rings were inserted through the cochleostomy; electrode 22 is the most apical electrode. Immediately after the surgery, the patient moved to a new location, and the programming of his device was followed in another center. During the first 5 years, the patient’s implant was programmed very sporadically. His performance with the cochlear implant never reached a level of understanding comparable to his hearing aid on the right side. Thus, the patient almost immediately began to use a powerful hearing aid on this right side in conjunction with the Nucleus 22 cochlear implant on the left side. In December 2000, 3 years after the last programming, the patient returned to the programming audiologist, stating that over the past few months he had begun to notice nonauditory sensations while his cochlear implant was on and recently his left eye had begun to twitch in the presence of loud surrounding sounds. Eye twitching was caused by 2 electrodes. Soon after the reprogramming, the eye twitching returned, and the patient was referred to our clinic. Figure 1 illustrates the progression of his FNS. Note that the stimulus level and bipolar (BP) +1 mode stimulating mode were used. If the FNS occurred below the behaviorally judged maximum comfortable loudness level or behavioral threshold, the measurement of maximum comfortable loudness level or behavioral threshold was discontinued. Prior to December 2000, electrodes 1 through 7 were removed for reasons other than FNS that were unknown to the user and the audiologist (Figure 1A). An integrity test showed normal function of these electrodes, and electrodes 2 through 7 were reactivated.

In 2003, he was left with only 12 active electrodes (electrodes 2-13; Figure 1C). An integrity test of his device showed 4 electrodes (17, 19, 21, and 22) to be malfunctioning. Integrity testing was performed using the Crystal Integrity Test system developed by Cochlear Corporation. The system uses surface potentials to identify any possible malfunctions. The test did not show any open or shorted electrodes; however, for these electrodes the stimuli artifact saturation level occurred for very low current levels, suggesting very high impedances for these electrodes. The patient stated that he was no longer able to use the telephone and was seeking a solution to improve his hearing by possible reimplantation. Although
his residual hearing on the right side had diminished over that period, he was still using a powerful hearing aid on his right side and was extremely reluctant to give up his hearing on that side. At evaluation, the patient’s Hearing in Noise Test (HINT), City University of New York (CUNY) sentences, and consonant-nucleus-consonant (CNC) word scores were 37, 87, and 12%, respectively, with the cochlear implant and hearing aid together and 8, 25, and 12%, respectively, with the cochlear implant only. The preoperative pure tone average was higher than 115 dB HL on the left side. Because of the resistant FNS and the decrease in implant performance due to inactivation of the electrodes, it was decided to explant the current device and reimplant the same side. In February 2004, cochlear reimplantation with a Nucleus 24 straight array was performed. A CT scan confirmed approximately the same electrode insertion (22 electrodes and 4 stiffening rings) as it was during the first surgery. 

Figure 2 shows FNS 1 week after the initial stimulation with his new cochlear implant. Note that current level and BP + 1 stimulating mode with the pulse duration of 100 microseconds (µs) for each electrode were used. For comparison of both cochlear implants, we attempted to use the same stimulating parameters with the new Nucleus 24 as those the subject was programmed with for his Nucleus 22. The R126 Nucleus 24 programming software does not allow exactly the same stimulating parameters as those available in the Nucleus 22 cochlear implant (ie, interphase gap was decreased from 45 µs to 8 µs). Stimulus level mode is not available for Nucleus 24. Instead, a current level for different pulse durations is used. The rest of the programming parameters were kept the same. The visible FNS with the Nucleus 24 occurred for the same electrodes and at approximately the same stimulating charge levels as with the Nucleus 22. Similarly, the most comfortable levels occurred at approximately the same levels. The most noticeable change occurred for the thresholds of the apical electrodes.

The Nucleus 24 uses a wide variety of programming parameters. Using the new features allowed all 22 electrodes to remain activated. Figure 3 shows the programming map without any FNS (ie, the FNS occurred above every comfortable loudness level). The parameters used were advanced combination encoders with the stimulating rate of 900 Hz. The subject was stimulated in the combined monopolar (MP) 1 + 2 mode with the pulse duration of 37 µs except for the electrodes 21 (100 µs) and 20 and 19 (50 µs).

The subject attended all programming appointments and followed all the instructions discussed during the appointments. He was advised to stop wearing his hearing aid on the right side for a period, and 7 months after the initial stimulation of his new cochlear implant system, the subject’s HINT, CUNY sentences, and CNC word scores were 72, 96, and 42%, respectively, with the cochlear implant only. The patient is now considering cochlear implant surgery on the opposite side.

CASE 2

A 64-year-old male patient presented to our clinic in 2001 with the complaint of FNS manifested by severe eye twitching of 2 years’ duration. His medical history was positive for otosclerosis. He had a right stapedectomy performed in 1962 and a left stapedectomy in 1967. Revision right stapedectomy surgery in 1972 resulted in severe hearing loss, and given the diagnosis of cochlear otosclerosis, the patient was started on fluoride treatment to prevent further loss in his hearing. The patient began to experience fluctuations in his hearing starting in 1984, which seemed to respond to diuretic treatment. In 1988 and 1992, he experienced severe drops in hearing that failed to respond to steroids and diuretics. He also had minor imbalance, without true vertigo. By the year 1992, he had profound sensorineural hearing loss in both ears necessitating a cochlear implant. His preoperative speech awareness was 85 dB HL for the right and 80 dB HL for the left ear. His pure tone average was 95 dB HL for the right and 85 dB HL for the left side. His discrimination NU-6 word test scores were 24% for the right ear and 0% for the left ear. The cochlear implant surgery was performed on the right side with a Nucleus 22 device at another institution. During that surgery, the round window was obliterated with new bone formation, and extensive drilling had to be performed to gain access to the scala tympani. All 22 electrode rings, including 9 stiffening rings, were inserted through the cochleostomy. In 1994, when switched from multipeak (MPEAK) to spec-
central peak (SPEAK) speech coding strategy, the patient showed a great improvement and was able to use his cochlear implant to a limited degree while on the telephone. His NU-6 word test and Central Institute for the Deaf (CID) sentences scores were 38% and 95%, respectively. At the same time, the patient started to have the first signs of FNS. At this point, although the FNS was limited to electrode 12, it occurred during stimulation around the most comfortable level. The maximum stimulating level for the electrode 12 was lowered, and all the electrodes were kept active. The patient reported that his performance began to worsen around 1996. In 1998, the FNS started to occur even for very soft sounds and the first electrodes, specifically electrodes 13 and 14, were turned off.

Figure 4 depicts the development of his FNS. Note that stimulus level mode was used. The BP $+ 1$ stimulating mode was used in maps A through C, and BP $+ 2$ stimulating mode was used in map D. Early in 2001, when the patient was referred to our center, he had severe FNS. The patient was very satisfied with the programming session results; however, electrodes 11 through 17 were turned off (Figure 4B). In the end of 2002, 2 additional electrodes, 9 and 10, were turned off (Figure 4C). Also, for some active electrodes a saturation of the stimulating current source was reached, and the patient was switched to the BP $+ 2$ stimulating mode (Figure 4D).

Until 1998, the patient was able to understand speech in a quiet setting in a 1-on-1 situation. The gradual deactivation of electrodes resolved the problem of FNS; however, the patient continued to gradually lose his speech understanding. This was reflected in large changes of his subjective thresholds for his active electrodes, especially for the most apical electrodes. Integrity testing was performed using the Crystal Integrity Test system. Although the test showed that each electrode was functioning normally, abnormal parameters (in certain stimulating modes, ie, common ground or pseudomonopolar mode, stimuli artifact levels for medial and apical electrodes are relatively small compared with the basal electrode responses or they change the polarity) consistent with this type of cause, especially in the area for electrodes 9 though 22, were observed, which could possibly explain the patient’s progressive drop in his speech understanding. In January 2003, the subject scored 0% on CUNY sentences with his cochlear implant alone. A CT scan of the patient showed hypolucency in the pericochlear areas on both sides, consistent with otosclerosis (Figure 5).

The patient received a Nucleus 24 contour implant on the left side in May 2003. The preoperative pure-tone threshold was 90 dB HL on the left side, and 15 months after the initial stimulation, the subject’s HINT, CUNY sentences, and CNC word scores were 96%, 95%, and 86%, respectively, with the cochlear implant only. Since the surgery, the subject has not experienced FNS with the cochlear implant system on the left side. Figure 6 shows the programming map parameters 15 months after the initial stimulation. The parameters used were ACE with the stimulating rate 900 Hz. The subject was stimulated in the MP1 $+ 2$ mode with the pulse duration of 25 µs.

After the reimplantation and map programming, both subjects showed an immediate improvement in speech discrimination. The first subject increased his CNC word score from 12% preoperatively (with cochlear implant only) to 42% (with reimplanted cochlear implant only) and has discontinued relying on the hearing aid in the contralateral ear. The speech tests were performed 7 months after the initial stimulation of his reimplanted device. Based on prior experiences and the subject’s impression, it can be expected that further improvement in his speech discrimination will occur. The second subject showed an even more dramatic change. Prior to the reimplantation, his speech discrimination was 0%, and 15 months after the initial stimulation of his reim-
planted device, he scored 86% on the CNC word test and became an exceptional cochlear implant user.

The pathophysiologic characteristics of hearing loss in patients with otosclerosis is considerably different from hearing loss from other causes. The pathologic condition is focused on the lateral wall of the cochlea, resulting in degeneration of the spiral ligament and stria vascularis, with only secondary involvement of the organ of Corti. The site of the lesion, in theory may account for the good postimplantation performance seen in our patients.

Bony changes within the cochlea due to otosclerosis are well known. In the early stages of the disease, there is resorption of the endochondral bone. Later in the disease process, there is deposition of new, basophilic, immature bone, which with time undergoes maturation into acidophilic bone. These processes of osteolysis and osteogenesis go on simultaneously, and the extent ofotic capsule destruction shows variability among patients with otosclerosis. The associated cochlear hearing loss is proposed to result from toxic metabolic changes in the cochlear fluids.

Demineralization of the otic capsule can be demonstrated on CT scans as varying degrees of radiolucency, which may involve only a small portion of the cochlea or may involve a widespread osteolysis of the temporal bone. The finding of pericochlear lucencies has been reported to be highly specific for cochlear otosclerosis. This is usually referred to as a “ring” or “double halo.”

Cochlear implants generate electrical fields that allow some current to spread outside the cochlea. Current flow within the cochlea depends on a variety of factors, including the cochlear anatomy, stimulus parameters, position and geometry of the electrode array, and local impedance. It has been proposed that the remodeled, softer bone in otosclerosis is proposed provides a pathway with decreased resistance through which the current disperses easily, acting like a shunt away from the modiolus to stimulate the facial nerve. Thinning of the bone between the facial nerve and cochlea during the otosclerotic process has also been implicated as a causative factor.

The prevalence of FNS in the overall cochlear implant population is reported to vary among different studies, from less than 1% up to 14.9%. It is rarely seen in the pediatric population. In this group, meningitis-induced otic capsule bony changes are primarily responsible for this complication as well as cochlear anomalies. Posttraumatic hearing loss is another potential cause of FNS after implantation. Endolymphatic hydrops, by unknown mechanisms, may also contribute to FNS in cochlear implant recipients.

Facial nerve stimulation may have its onset as soon as initial stimulation of the device or it may be delayed.

Figure 5. Computed tomographic scans showing pericochlear lucencies. A, Left side; B, right side.

Figure 6. A programming map of case 2, 15 months after the initial stimulation of the new Nucleus 24 cochlear implant system (Cochlear Corporation, Englewood, Colo) (September 2004). The y-axis shows the stimulation level in charge units (nC). The x-axis shows electrodes from base to apex. C indicates maximum comfortable loudness; T, behavioral threshold.
Facial nerve stimulation has also been noted to migrate to adjacent electrodes. The reason for this is unknown; however, it has been postulated that electric current generated by the cochlear implant can result in thinning of the bone between the cochlea and the facial nerve, or the underlying disease process (eg, otosclerosis) may soften or thin the cochlear bone. However, the effect of electrical fields on long bones is new bone formation, not osteolysis.4

Several studies involving temporal bone dissection and radiological measurements have been conducted to define the anatomical relationship of the cochlea and the facial nerve segments.4,8 One of the aims of these studies was to find if the electrodes within the Nucleus 22 implant that are closest to facial nerve segments are really the offending electrodes in the clinical setting. According to Kelsall et al,8 the thinnest bone measured was between the labyrinthine segment of the facial nerve, 1.00 mm from the geniculate ganglion and the basal turn of the cochlea, with an average thickness of 0.52 mm. The electrodes closest to the labyrinthine segment of the facial nerve were 12 through 16 (average, 14.8). In their study, 8 of 11 patients with FNS had good correlation between the clinically offending electrodes and the electrodes measured to be closest to the labyrinthine and geniculate segments of the facial nerve on CT scans.8 This correlation was especially strong in patients with cochlear otosclerosis. The findings in the 2 patients described herein support this. In the first case, before reimplantation, FNS occurred for electrodes 20 through 14 (BP + 1 mode). In the second case, before reimplantation, FNS occurred for electrodes 19 through 9 (BP + 2 mode). Considering the insertion depth (case 1: 22 electrodes plus 4 stiffening electrodes; case 2: 22 electrodes plus 9 stiffening electrodes) and postoperative CT scan in both cases, FNS occurred at approximately 340° to 210° (0° is round window).

In a similar study performed by Bigelow et al,4 it was found that the scala tympani was uniformly closer to the facial nerve than the scala vestibuli. Temporal bone dissection also revealed that electrodes 8 though 13 in the Nucleus 22 implant, located in the superior most aspect of the basal turn, were closest to the nerve. In their clinical series, the electrodes causing FNS ranged from 10 to 18 initially, involving electrodes 7 to 20 over time. This difference between the temporal bone findings and the clinical setting was attributed to variations in the cochlear length, position of the nerve to the cochlea, and to positioning differences and bending of the array on insertion.9

Using an electrical field imaging technique, Vanpoucke and colleagues13 suggested that the facial nerve canal is an important conduction path. Although this study was performed on subjects with a Clarion II device (Advanced Bionics Corporation, Sylmar, Calif) with a HiFocus electrode array, the results may suggest similar outcomes with Nucleus electrode arrays.

Various programming methods can be used to address the problem of FNS in cochlear implants. Turning off the offending electrodes, setting the stimulus or current levels below the threshold for FNS, using a combination of wider stimulation modes for electrodes causing the stimulation, and keeping the nonoffending electrodes in standard modes are among the programming strategies that may be used to overcome this complication.

To avoid or overcome the problem of FNS in cochlear implant recipients, several surgical strategies can be undertaken. One is use of a modiolar hugging electrode array. It is presumed that placing the electrodes as close to the auditory nerve cells as possible would result in less undesired current flow toward the outer rim of the cochlea and hence toward the facial nerve. Also, using a cochlear implant system with a plastic positioner (eg, Clarion systems) can also serve this purpose. The positioner not only acts to push the electrode array toward the modiolus but also acts like an insulator between the electrodes and outer wall of the cochlea, resulting in less current flow to the facial nerve. However, the risk of causing additional trauma to the cochlear structures, which is another potential source for FNS, is higher when inserting this type of array. Scala vestibuli, being further from facial nerve segments, is a potential location for implantation for cases with a high risk of FNS. However, in the study by Bigelow et al,4 1 patient had FNS despite scala vestibuli insertion.

Reimplantation is an option that can be undertaken if programming fails to resolve the FNS. This can be done on the same side, as well as on the contralateral side. The opposite side may be preferred for several reasons. In cases that are resistant to programming, the problem is disease-induced anatomical defects of the cochlea. In most cases, the cochleas on each side are not affected equally. The risk of having the same problem at the exact cochlear site on the other side is low, and usually, the more severely affected ear receives the implant first, leaving a cochlea in a better anatomical condition for reimplantation on the other side, as in case 2. Reimplantation on the same side, although having the disadvantage of the possibility of encountering the same problem in the postoperative period, has the advantage of leaving the other ear a potential source for reimplantation if the problem recurs in the future. One benefit of reimplantation is that, even though a similar electrode array configuration may be used, the more advanced technology used in newer devices allows broader programming possibilities to overcome FNS even if the anatomical problem persists. This is nicely illustrated in case 1. A second possible benefit of reimplantation of the same ear with a new device may be that removing the implant and reinsertion of a new electrode may mechanically eliminate the newly formed spongiotic bone. This may have been reflected in the lower impedances achieved with the new device in case 1.

Sodium fluoride treatment in cochlear implant recipients experiencing FNS is an experimental treatment, and to our knowledge, no studies proving its efficacy have been published.

In conclusion, FNS in otosclerotic cochlear implant patients may be managed through advanced programming techniques in many instances. However, if the FNS becomes too extreme or persistent, reimplantation can be a useful tool to overcome FNS and improve perfor-
mance with the cochlear implant. The results presented in this study suggest that reimplantation with a newer generation device has the advantage of allowing more programming options to overcome resistant FNS.

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