Outcomes following cochlear implantation for patients with single-sided deafness, including those with recalcitrant Ménière's disease

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Abstract

Objective—Compare pre and postoperative performance in patients undergoing cochlear implantation (CI) for unilateral severe-to-profound sensorineural hearing loss (single-sided deafness, SSD).

Study design—IRB-approved, prospective

Setting—Tertiary center

Patients—Twenty-nine patients have undergone CI for SSD. SSD was due to Ménière's disease (MD) in 10 subjects; these also suffered from recalcitrant vertigo spells and in these 10 patients along with 2 others the CI was placed simultaneous with a labyrinthectomy.

Intervention(s)—CI with or without labyrinthectomy.

Main outcome measure(s)—CNC word and AzBio sentences in quiet were administered to the implanted ear. A multiple-loudspeaker sound localization test was administered in the bilateral listening condition. All data were collected pre-operatively and 3, 6, 12-months post-operatively with post-operative data available for 19 subjects. Additionally, a tinnitus handicap questionnaire is administered pre- and 12-months post-operatively.

Results—CNC word and AzBio sentence scores showed improvement in the implanted ear. Sound localization appeared to improve in an experience dependent fashion in some patients. Most patients reported diminished tinnitus following cochlear implantation. All patients undergoing labyrinthectomy experienced resolution of vertigo attacks.

Conclusions—CI restores auditory function to the deafened ear. Additionally, the binaural input appears to improve sound localization for most patients. In patients with severe hearing loss and recalcitrant vertigo attacks due to MD, simultaneous labyrinthectomy and CI effectively relieves vertigo attacks and improves auditory function.

Keywords

cochlear implant; unilateral hearing loss; tinnitus; vertigo; endolymphatic hydrops

Introduction

Binaural hearing provides several advantages over monaural hearing including binaural summation, the binaural squelch effect, and the ability to localize sounds (1). Binaural

summation occurs when the same stimulus is available to both two ears. Higher order processing of the redundant information received by the two ears provides a 2-6 dB improvement in signal detection threshold and leads to improvement in speech perception in quiet and in noise. The binaural squelch effect represents the ability to combine the noise from the ear with the poorer signal-to-noise (S/N) ratio with the noise from the ear with the more favorable S/N ratio allowing increased speech perception in background noise (2). Finally, the ability to localize sounds depends on the comparison of interaural timing and intensity differences between two independent ears (3).

An additional advantage of hearing with two ears relates to the head shadow effect. This occurs when the head acts as an acoustic barrier, which creates a spectral difference between the two ears and, in turn, a greater S/N ratio at one ear (4). Although two ears are not necessary to benefit from the head shadow, they enable the listener to attend to the ear with a better S/N ratio. Due to the loss of these binaural advantages, patients with severe-to-profound unilateral sensorineural hearing loss (SNHL, single-sided deafness, SSD) experience difficulties in hearing and understanding speech presented to the deaf side, and sound localization.

Standard rehabilitation options for SSD include contralateral routing of sound (CROS) and osseointegrated implants (OI) (5-9). Both help overcome the head shadow effect and restore auditory perception to the side with severe-to-profound SNHL. However, they do not restore binaural hearing and fail to fully rehabilitate the deficits of SSD. In particular, they provide little improvement in sound localization and modest improvement in speech perception in noisy environments (5-10). Electrical stimulation with a cochlear implant (CI) remains the only method to restore auditory perception to a deafened ear. Recent reports suggest that CIs provide improved ability to localize sound and speech perception in some situations with background noise compared to CROS and OI devices (11). Suppression of tinnitus represents another potential advantage of electrical stimulation over CROS and OI devices (12,13). Given the potential advantages and the initial encouraging results we have begun to offer CIs as an off-label indication to patients with SSD.

Some patients with late stage Ménière's disease (MD) experience debilitating vertigo and/or Tumarkin drop attacks that are unresponsive to medical therapy and/or non-ablative surgery (14,15). Many of these patients also suffer severe-to-profound SNHL in the affected ear. Chemical or surgical labyrinthectomy offers a high rate of vertigo control in these patients; however patients still suffer the consequences of SSD (16-18). In an effort to alleviate vertigo spells and rehabilitate the effects of SSD, we have recently offered simultaneous surgical labyrinthectomy and cochlear implantation to patients with MD, poor unilateral hearing and incapacitating vertigo.

Here we provide preliminary speech perception and sound localization outcomes on a cohort of 26 patients that have received a CI as an off-label indication to rehabilitate SSD. This report also includes 12 patients who underwent simultaneous labyrinthectomy and cochlear implantation.

Materials and Methods

Subjects

The Institutional Review Board of the University of Iowa approved the study protocol. From 2012- 2013, 29 patients with good to normal hearing in one ear and profound hearing loss in the opposite ear have received a CI at our institution. Of those patients, pre-operative data are available from 25 subjects and post-operative data are available from 19 subjects who have had at least three months of CI experience. Table 1 presents the demographic data for

the patients implanted at the University of Iowa. Sixteen of the patients received Nucleus devices and 13 patients received Advanced Bionics devices. The mean age at implantation was 55.2±1.69 (mean±standard error, SE) years (range 31 years to 75 years) and the mean duration of severe to profound hearing loss in the implanted ear was 3.5±0.76 years. Females comprised 52% (15/29) of the patients. The mean pre-operative pure-tone average (PTA) calculated at 0.5, 1, 2, 4 kHz was 76.6±2.71 (SE) for the implanted ear and 22.1±2.31 for the better hearing ear. One patient had tried a CROS hearing aid prior to implantation (patient C1) as part of a separate study and one patient utilized an OI for 5 years prior to implantation (patient 28). All patients were counseled heavily either with a CI audiologist, physician, and/or hearing aid audiologist on their rehabilitative options prior to making the decision of a cochlear implant.

Idiopathic sudden sensorineural hearing loss (ISSNHL) represented the most common etiology for the hearing loss (14/29, 48.2%). One patient with ISSNHL presented with poly cranial neuropathy including facial paralysis, vertigo, and profound SNHL. Another patient with ISSNHL had a vestibular schwannoma in the contralateral, better hearing ear.

Ménière's disease was the second most common etiology, accounting for the unilateral hearing loss in 10 patients (38.5%). All patients with MD had stage IV disease based on the modified system of the AAOHNS Committee on Hearing and Equilibrium (19). All 10 patients with MD underwent simultaneous labyrinthectomy for management of intractable vertigo. In another patient a labyrinthectomy was performed for management persistent dizziness following 3 doses of intratympanic gentamicin at another institution; however the patient did not fulfill diagnostic criteria for definite MD and was not including in the MD cohort. One other patient underwent simultaneous labyrinthectomy and CI during a translabyrinthine approach to an internal auditory canal lipoma with pre-existing profound unilateral HL and intractable vertigo. In these 12 cases the primary indication for surgical intervention was control of dizziness; cochlear implantation to improve hearing was performed simultaneously. One case each of unilateral HL resulted from middle ear surgery, head trauma, and an ipsilateral facial nerve schwannoma involving the internal auditory canal.

Speech perception

Speech perception was measured in quiet using recorded Consonant-Nucleus-Consonant monosyllabic words (CNC) (20) and AzBio sentences (21). CNC scoring was based on percent-correct performance at both the word and the phoneme levels and the AzBio sentences were scored by dividing the total number of words correctly identified by the total number of words possible. Two lists of CNC words and AzBio sentences were presented to each subject. All speech perception lists were randomized between subjects and no subject received two of the same lists during any test session.

Pre-operatively, the CNC words and AzBio sentences were presented in the sound field, in a $10' \times 9.3' \times 6.5'$ sound-treated booth at 60 dB A. In order to isolate the poorer ear, masking, using speech-shaped noise, was presented to the better ear via an insert ear phone. If hearing loss allowed, a hearing aid was verified and fit in the poorer ear. Post-operatively, direct electrical connection to the cochlear implant sound processor was used to present CNC words allowing us to test the ear with the CI in isolation.

Sound localization

A localization test was administered using everyday sounds presented at 60 dB A from (22) an array of eight loudspeakers spanning a horizontal arc of 108°. Loudspeaker one and loudspeaker eight were placed 54° to the left and to the right of the straight-ahead (0°)

position. Each of the 16 different sound items (23) were repeated six times randomly from one of the eight loudspeakers for a total of 96 presentations for the entire test. The subject was told to identify the loudspeaker number from which the sound originated, but not to identify the sound itself. Subjects were not given feedback as to a right or wrong answer. Localization performance was determined by calculating the average Root Mean Square (RMS)-error in degrees. All presentations of the sounds were used to calculate the Average RMS-error in degrees.

Results

All patients except one (patient 2) wear their devices throughout the waking day and all report satisfaction with their devices. Patient 2 wears his device daily, but intermittently.

Speech perception outcomes

Figures 1 and 2 present preoperative and 3, 6, 12, and 24-month postoperative speech perception scores in the implanted ear (CNC words and AzBio sentences, respectively) for the patients for which the data were available. For both tests, the data indicate a high degree of individual variability, as is commonly seen in regards to CI performance. Thirteen subjects have experienced some level of benefit (most recent post minus pre score) on word scores (average benefit 32% ± 5.7 SE; range 9% to 64%) and 14 subjects have experienced some level of benefit on sentence scores (average benefit 39% ± 7.7 SE; range 4% to 92%). Average scores for the cohort demonstrate improved hearing in the implanted ear following implantation and continued improvement with experience. Overall, the average improvement on CNC word scores for the 19 subjects with pre- and most recent post-operative data was 28% ± 5.1 (mean ± SE). This improvement ranged from 64% for one subject to a decrement of 26%. Overall, the average improvement on CNC word scores for the 19 subjects with pre- and post-operative data was 28% ± 5.1 (mean ± SE). This improvement ranged from 64% for one subject to a decrement of 26%. The differences in preoperative CNC word scores compared to most recent postoperative scores was statistically different (p<.05, Wilcoxon Signed Ranks test). On AzBio sentences, average improvement was 40% ± 6.6 with a range from 92% improvement for one subject to a decrement of 57% for another subject. The differences in preoperative AzBio word scores compared to most recent postoperative scores was statistically different (p<.05, Wilcoxon Signed Ranks test).

Sound localization

Figure 3 presents preoperative and 3, 6, and 12-month postoperative individual and group average sound localization results. Performance on this task improved post-operatively in most, but not all, individuals. Further, as demonstrated by the average data for the group, performance improved from 3 to 12-months postoperatively suggesting that sound localization is experience dependent.

Vertigo

All patients with MD undergoing labyrinthectomy reported complete resolution of their vertigo attacks at their latest follow-up. Four patients with less than 6 months of follow-up from the labyrinthectomy noted mild dysequilibrium as they undergo vestibular compensation. Most patients do not yet have 18-24-month follow-up data so it is not possible to classify them according to the AAOHNS Committee on Hearing and Equilibrium guidelines, yet assuming continued control they would all be regarded as class A (19). The other two patients with disabling dizziness that underwent labyrinthectomy likewise experienced a resolution of the dizziness.

Tinnitus

In addition to improved speech perception in the implanted ear, most patients reported subjective improvement in tinnitus. We are actively collecting prospective pre and post-implantation tinnitus questionnaires to quantify changes in tinnitus perception over time; however at this time we do not have sufficient 12-month follow-up data to report.

Discussion

SSD significantly impacts auditory perception and performance, which in turn impacts function in educational, social and employment settings (24-26). For patients who seek rehabilitation, CROS and OIs systems remain the standard management options with proven efficacy (5-9). Cochlear implantation has recently emerged as another off-label option for the rehabilitation of SSD (11,27,28). Here we report our preliminary results with cochlear implantation in a series of patients with SSD and good hearing in the contralateral ear.

Speech perception

Recent reports have demonstrated improved speech perception for patients with SSD following cochlear implantation in the implanted ear (11,27,28). They also demonstrated that cochlear implantation enhanced speech perception in noise, particularly when the noise is presented to the implant side (11). Our results are largely consistent with this experience, with the majority of patients demonstrating significant improvement in speech perception in the deafened ear following CI. Three subjects demonstrated decreased CNC word scores 3-6 months post-operatively compared to the preoperative data. We do not yet have longer-term data on these two subjects and will continue to monitor their progress over the next 24 months. In one case (patient 15), poorer performance may be related to device issues. This patient required deactivation of 8 electrodes and demonstrated rising impedances. We are continuing to monitor the integrity and function of this device. Another patient (patient 16), underwent a simultaneous labyrinthectomy so the preoperative hearing does not reflect the actual hearing status of the ear without a CI. For this individual along with the other patients undergoing simultaneous labyrinthectomy, we are beginning to test these individuals postoperatively with or without the use of the CI to better compare the benefit of CI for speech perception with their current hearing status. Most patients in this study do not yet have >6 months experience with their CI so we have not been able to obtain speech perception in noise testing in patients with significant CI experience. Thus, outcomes for speech perception in noise testing for our patients will require further follow-up.

Our study was limited to adult patients with acquired hearing loss. SSD occurs in children and likely impacts development and performance (29). Whether or not rehabilitation of children with acquired or congenital SSD with CIs is beneficial remains to be shown, however early case reports are encouraging (30,31).

Sound localization

The ability to localize sounds depends on binaural input. It is not clear how well the brain can integrate acoustic signals from one ear with input from a CI in the other ear to localize sounds. The data presented here confirm that patients with SSD perform poorly on sound localization tasks. We only have 12-month postoperative localization data available for six subjects, limiting the conclusions that can be drawn on the ability of a CI to improve sound localization in patients with SSD. Also, some patients demonstrated decreased sound localization ability following CI in early post-operative testing. Nevertheless, the data from the entire cohort viewed collectively appear to support the notion that rehabilitation of SSD with a CI allows for improved sound localization and that this ability improves with time. Apparently with experience the brain is able to integrate acoustic signals from one ear with

those provided by a CI in the opposite ear to help localize the source of a sound. For those patients undergoing simultaneous labyrinthectomy that had some residual hearing prior to labyrinthectomy, preoperative sound localization measures likely overestimate the patients' function without a CI. As with speech perception measures, we are beginning to compare sound localization in these individuals postoperatively with or without the use of the CI to better determine the benefit of CI in sound localization for these subjects. Additional follow-up data in a larger cohort of patients will be necessary to confirm these preliminary findings. Nevertheless, an improved ability to localize sounds represents one likely advantage of a CI over CROS or OI devices for rehabilitation of SSD.

Tinnitus suppression

Several studies suggest that cochlear implantation reduces tinnitus severity. CIs were initially considered in patients with SSD and severe tinnitus as a potential treatment to reduce tinnitus (28,32). Although we do not yet have 12-month post-operative tinnitus survey results to publish, most patients reported tinnitus suppression following activation of the CI. Tinnitus suppression by electrical stimulation may be limited to the duration of the stimulation while in other patients the suppression may persist beyond the period of electrical stimulation (12,13,33)

Simultaneous labyrinthectomy and cochlear implantation in MD

Chemical and surgical labyrinthectomy represent highly effective methods to reduce or eliminate vertigo attacks in patients with MD (16-18). Surgical labyrinthectomy results in profound deafness and is typically reserved those patients with pre-existing severe-to-profound hearing loss in the affected ear. The outcomes reported here mirror those reported by others confirming that surgical labyrinthectomy relieves vertigo spells in nearly all patients with intractable MD. Patients require a period of recuperation following chemical or surgical labyrinthectomy to allow for vestibular compensation. Nevertheless, most patients welcome the relief from unpredictable, severe vertigo spells.

Cochlear implantation is the only available therapy to restore auditory perception to an ear deafened by MD. Assuming the encouraging preliminary hearing results seen in our patients hold up over time, simultaneous labyrinthectomy and cochlear implantation not only provides relief from vertigo attacks, but also provides improved hearing for these patients. Depending on the long-term hearing outcomes following cochlear implantation, it may be reasonable to extend the indication of labyrinthectomy to patients with intractable vertigo and some residual hearing. Another advantage of cochlear implantation with or without labyrinthectomy in ears deafened by MD relates to the fact that over time some patients will develop the disease in the contralateral ear placing the patient at risk for bilateral deafness (34-38). Indeed many adults receiving a cochlear implant for bilateral deafness suffer from MD (39). Having a functioning CI in the initially deafened ear will allow the patient to continue hearing in the event that the hearing in the contralateral eventually deteriorates.

We prefer to perform simultaneous labyrinthectomy and CI in patients with severe-to-profound SNHL and intractable vertigo attacks for several reasons. These include reduced risks associated with another surgical procedure and anesthetic. Also, surgical labyrinthectomy may lead to soft tissue scarring or even ossification in the cochlea which could prohibit later implantation (40), although this is not a uniform response (41,42). Finally, immediate implantation reduces the time of deafness and performance with a CI is correlated with duration of deafness (43). One disadvantage of simultaneous labyrinthectomy and CI is that patients with residual hearing preoperatively will have not experienced the full consequences of SSD and may not fully appreciate the benefit of the CI for rehabilitation of the new deficit.

Conclusions

Patients with SSD experience significant problems with sound localization and speech perception in background noise leading many to seek rehabilitation options. CROS and OI devices provide significant benefit, yet fail to address deficits that require binaural auditory processing. The data reported here, coupled with recent reports, raise the possibility that restoration of binaural auditory perception via cochlear implantation offers advantages not provided by CROS or OI devices. The extent to which the advantages of CI outweigh those provided by CROS or OI technologies requires further comparative studies.

Acknowledgments

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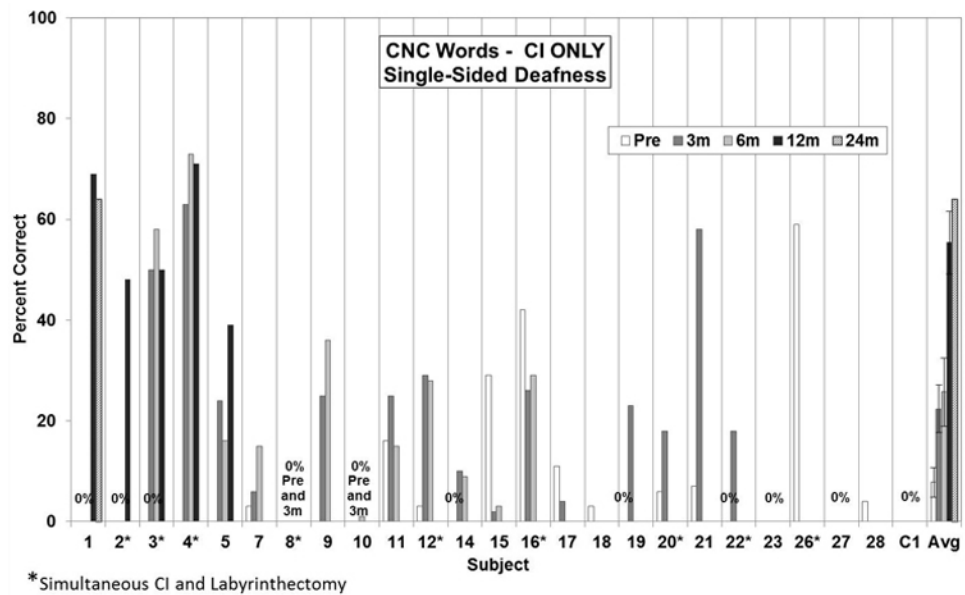
Figure 1.
Pre- and post-operative CNC word scores in the implanted ear. Individual preoperative and 3, 6, and 12-month postoperative scores. The bars at the far right present the group mean. Error bars present standard error. The differences in preoperative scores compared to 6 and 12-month postoperative scores were statistically significant (p<0.05, Kruskall-Wallis ANOVA with post-hoc Dunn's method) whereas the preoperative scores were not statistically different than the 3-month postoperative scores.
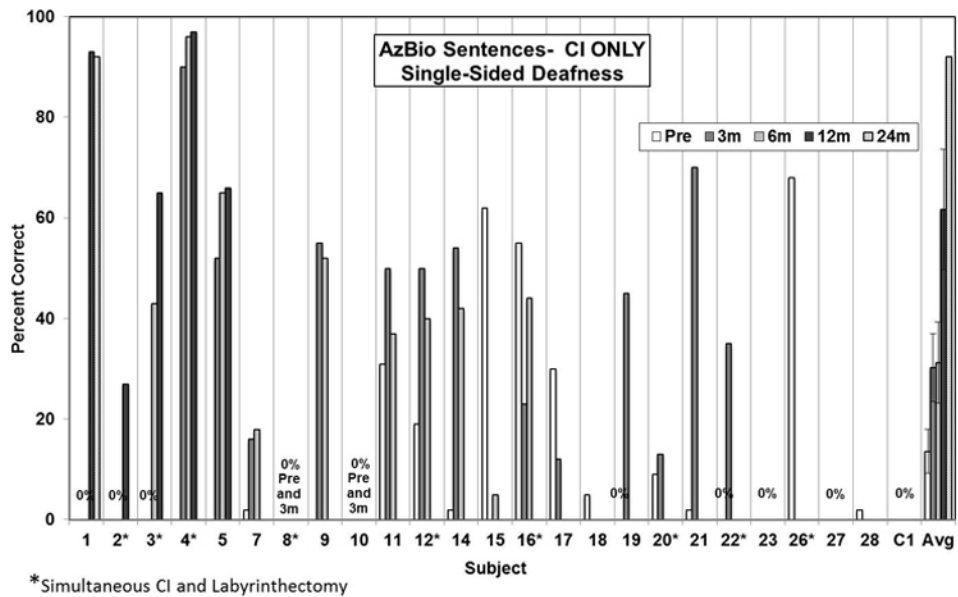
Figure 2.
Pre- and post-operative AzBio sentence scores in the implanted ear. Individual preoperative and 3, 6, and 12-month postoperative scores. The bars at the far right present the group mean. Error bars present standard error. The differences in preoperative scores compared to 3 and 6-month postoperative scores were statistically significant (p=0.020 and p=0.018, respectively, one-way ANOVA with post-hoc Holm-Sidak method) whereas the preoperative scores were not statistically different than the 12-month postoperative scores.
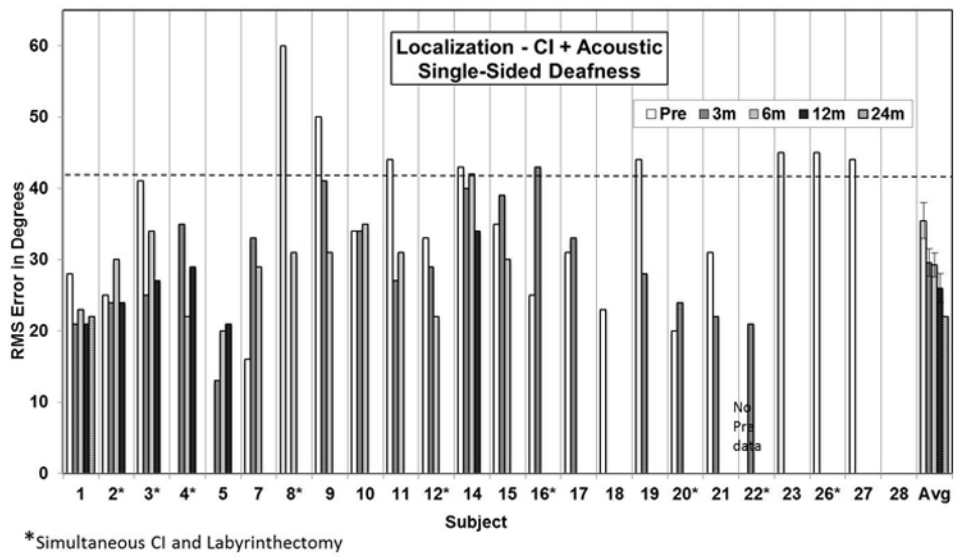
Figure 3.
Pre- and post-operative sound localization results in the bilateral hearing condition. Individual preoperative and 3, 6, and 12-month postoperative root mean square (RMS) error in degrees scores are presented. The bars at the far right present the group mean. Error bars present standard error.

Table 1

Subject demographics. Pre-operative pure-tone average (PTA) calculated at 0.5, 1, 2, 4 kHz. NU=Nucleus device; AB=Advanced Bionics device;ISSNHL=idiopathic sudden sensorineural hearing loss; IAC=Internal auditory canal; N/A=have not had post-CI testing; UNK=no data available.

Subject ID	Age at Implant	Duration of Deafness (years)	Etiology	Left PTA	Right PTA	PreOp CI Ear CNC Score	Most Recent Post-Op CNC Score	CI Ear	CI Type
1	58	2	ISSNHL	84	15	0	64	Left	NU CI512
2	50	2	Ménière's/ Labyrinthectomy	NR	16	0	48	Left	NU CI512
3	55	1	Ménière's/ Labyrinthectomy	21	86	0	50	Right	NU Freedom
4	57	2	Ménière's/ Labyrinthectomy	70	28	UNK	71	Left	NU Freedom
5	58	3.9	ISSNHL	64	13	UNK	39	Left	AB 90K
7	48	0.6	ISSNHL	13	71	3	15	Right	AB 90K
8	52	3	poorly compensated vestibulopathy/Labyrinthectomy	NR	15	0	0	Left	NU CI422
9	51	15	ISSNHL	NR	63	0	36	Left	AB 90K
10	52	17	Post-surgical	34	96	0	1	Right	AB 90K
11	55	0.6	ISSNHL	9	68	16	15	Right	AB 90K
12	49	1.3	Ménière's/ Labyrinthectomy	44	75	3	28	Right	AB 90K
13	64	2.8	ISSNHL	6	NR	0	UNK	Right	AB 90K
14	75	0.9	ISSNHL	31	95	0	9	Right	AB 90K
15	56	1	Facial nerve schwannoma	31	85	29	3	Right	NU CI422
16	61	1.6	Ménière's/ Labyrinthectomy	65	30	42	26	Left	NU CI422
17	62	4.7	ISSNHL	83	31	11	4	Left	AB 90K
18	64	3	ISSNHL	75	10	3	N/A	Left	NU CI422
19	53	0.5	ISSNHL	81	15	0	23	Right	AB 90K
20	58	3.4	Ménière's/ Labyrinthectomy	78	33	6	18	Left	NU CI422
21	63	10	Head Trauma	90	19	7	58	Left	NU CI422
22	63	8	Ménière's/ Labyrinthectomy	16	56	0	18	Right	NU CI422
23	64	0.3	ISSNHL	28	NR	0	N/A	Right	AB 90K
26	63	0.8	Ménière's/ Labyrinthectomy	14	55	59	N/A	Right	NU CI422
27	58	1	ISSNHL	93	33	0	N/A	Left	AB 90K
28	36	5	ISSNHL	98	15	0	N/A	Left	NU CI422

Subject ID	Age at Implant	Duration of Deafness (years)	Etiology	Left PTA	Right PTA	PreOp CI Ear CNC Score	Most Recent Post-Op CNC Score	CI Ear	CI Type
CI	59	.8	ISSNHL/Facial nerve paralysis	NR	14	0	N/A	Left	NU Freedom
NC1	42	4.3	Ménière's/ Labyrinthectomy	23	65	UNK	UNK	Right	NU CI422
NC2	45	1.6	Ménière's/ Labyrinthectomy	69	11	UNK	UNK	Left	NU CI422
NC3	31	4	IAC lipoma/Translabyrinthine approach	9	60	UNK	UNK	Right	AB 90K