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Case Reports on Auditory Deprivation and Audiologic Management

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Abstract 1

Speech Perception Outcomes in Long-term Cochlear Implant Users

Introduction: Long-term follow-up appointments are beneficial for cochlear implant (CI) users. Several factors can influence individual auditory performance, even after years of experience with amplification use. Maintaining routine follow-up appointments with an audiologist can provide verification measures, auditory monitoring, and ongoing counseling to achieve optimal performance with CI technology. Case Presentation: An elderly, female patient with a monaural CI presented to the clinic with a significant decline in speech perception after years of use. Discussion: The goal of CI programming is to effectively convert acoustic input into a usable electric signal for each stimulated electrode to produce adequate auditory exposure and avoid the deleterious effects of auditory deprivation. Conclusion: Commitment to annual programming and follow-up is critical to maintaining the stability of auditory performance and speech perception abilities. The need for additional research on the stability of speech perception abilities in the older adult CI population was evident.

Case Presentation 1

Speech Perception Outcomes in Long-term Cochlear Implant Users

Introduction

The multiple-channel cochlear implant (CI) is the most successful sensory prosthetic device in the history of medicine (1). Cochlear implants provide significant improvement in speech perception abilities for those with moderate to profound sensorineural hearing loss who receive little to no benefit from conventional amplification. They do so by circumventing damaged sensory hair cells of the cochlea and directly stimulating the auditory nerve, delivering coded signal information to the brain (1). Follow-up programming appointments are critical for setting the appropriate parameters in the processors for the individual user to achieve the best auditory performance (2).

Generally, post-lingually deafened adults experience significant improvements in speech perception within the first six months after implantation (3). A study of individuals that underwent cochlear implantation during their adult years revealed meaningful improvement in speech understanding and quality of life (4). Processors must be appropriately individualized by programming ideal settings through adjusting parameters. This ensures electrical stimulation patterns generated by the device in response to sound that yield optimal speech intelligibility (2).

Unfortunately, the stability of speech perception performance varies after years of listening experience with a CI (2). Half of the variability can be attributed to factors related to the duration of auditory deprivation, age-relatedness, cognitive factors, differences in sensitivity to temporal and spectral cues, and positioning of the electrode array in the internal implant component (1). Inconsistencies in performance may be attributed to programming adjustments resulting from the user adjustment period, experience listening with a CI, and physiological changes (5). Most factors that influence performance can be addressed with a program of routine lifetime follow-up appointments. However, after the first year following the initial activation of the implant, only recommendations and as-needed methods are typically employed (2). Annual or routine follow-up care for CI users would safeguard optimal performance through verification of device functions, monitoring of auditory status, and delivery of educational counseling and support.

Case Presentation

An elderly female was seen for a CI mapping appointment with the chief complaint of inability to understand speech without reading lips. She presented to the clinic with bilateral sensorineural hearing loss that had reportedly progressed through school-aged years. She had used a hearing aid on her right ear until she received a cochlear implant. A slim straight internal electrode component was implanted on her right side when she no longer demonstrated benefit from conventional hearing aids. At the time of her appointment, she had a unilateral Nucleus 22 electrode array with a CP290 processor on the right with no amplification on the left. Reportedly, amplification had never been used on the left ear.

During her first clinical encounter, no audiogram was available for review. Years before, speech perception testing was performed that revealed a score of 0% on the Hearing in Noise Test (HINT) and 8% on the Consonant-Vowel Nucleus-Consonant (CNC) test. Unaided and aided thresholds were obtained from 125-8000 Hz (Figure 1). Unaided thresholds were impaired profoundly from 125-500 Hz, bilaterally. No response was detected at the limits of the audiometer for frequencies above 500 Hz for air and bone conduction in both ears. Aided thresholds for the right CI were obtained in the sound-field with the external processor in place. Hearing thresholds were in the moderate severity range at 250 Hz, severe at 500 Hz, mild at

1000 Hz, and moderate from 2000-8000 Hz. Due to the patient's extreme difficulty comprehending speech, aided word recognition testing was not conducted.

The patient's CI mapping summary may be found in Table 1. Upon initiating CI programming, electrodes 18, 16, and 14 were noticeably flagged by the software, and electrodes 18-14 were disabled. All electrodes were unflagged and enabled for accurate measurement of T- and C-levels. T-levels were obtained via hand-raising methods when tones were heard in her right ear. C-levels were obtained in "live" mode, with the patient indicating when sounds were *loud but comfortable*. Dynamic ranges were made appropriate across all enabled electrodes. Sweeps of 4 electrodes were performed and C-levels were decreased on electrodes 14 and 4 for patient comfort. Her T- and C-levels and the dynamic range of electrodes were recorded: electrodes 22, 21, 18-15, 5, and 3 were disabled when no response was found when T- and C-Levels were collected, and electrodes 15 and 17 were disabled due to facial nerve stimulation (Table 1).

Ling Sounds were evaluated through right speaker sound-field presentation (0-degree azimuth, 3-foot distance) by indicating when the stimuli were heard. Ling sounds were used, given the patient's history of difficulty with the HINT and CNC. Although she encountered problems identifying the correct sound card associated with the presented stimulus, the patient was able to detect sounds between 30- and 75-dBHL. The goal is to detect these sounds at levels around 20 to 25 dBHL (Table 2). Overwhelmed, the patient was unable to recognize speech. Nevertheless, she was encouraged to continue wearing her processor and speech-read to allow her brain to associate sounds with meaning and apply additional strategies for practice. These strategies included listening to audiobooks and television with closed captioning.

Discussion

Life-long CI programming and care are necessary to avoid the detrimental effects a user may experience related to auditory performance and quality of life (2). Initial encounters between a clinician and a patient establish a commitment to ongoing care and support. This case was experiencing adversity with her CI and needed ongoing support to improve communication. Cochlear implant success may depend on various issues, including the quality of the programming experience (2). To provide benefit, the parameters of soft and loud sounds for individual electrodes should be adjusted. To provide sufficient auditory nerve stimulation, this adjustment should be administered to accommodate the changes happening in a person's life (5).

Long-term follow-up appointments should allow the audiologist to monitor speech perception and auditory changes, make device adjustments, check and troubleshoot equipment, and identify the need for replacement instrumentation or technology upgrades (5). Routine programming and ongoing support should ensure that the device is being used effectively during waking hours while insufficient auditory exposure is avoided (1). Due to the risk of decreased speech perception in long-term CI users, there is a need for standard protocols to monitor patients after their first year of implantation. This should be done to ensure improved outcomes and lifelong benefits for every person.

Conclusion

This case demonstrates the need for annual follow-up appointments to address CI performance for achieving life-long benefits. When the brain has been deprived of sound and receives insufficient auditory information, the ability of patients to process information correctly becomes degraded, which results in a severe impact on speech perception and intelligibility. Life-long follow-up and consistent use of amplification are necessary to ensure appropriate audibility of speech and environmental sounds. Hence, this case illustrates the need for further research to examine the older, adult CI population, the stability of speech perception in long-term CI users, and to establish best practices for ongoing support to gain lifelong benefit from this classification of surgically implanted devices.

Abstract 2

Outcomes of Longstanding Untreated Hearing Loss in Adults

Introduction: Auditory deprivation occurs when the auditory system is deprived of sound, resulting in an inability to process auditory information over time. Individuals with untreated hearing loss are susceptible to the negative consequences associated with auditory deprivation, regardless of the type and degree of hearing loss. Presentation: A young adult female patient with asymmetrical hearing loss since childhood and significant hearing health history presented to the clinic. She had a history of unsuccessful hearing aid use and had been without any form of amplification in either ear for approximately 11 years. Discussion: The implications of hearing loss can go unnoticed for years, especially when hearing-impaired people implement strategies to compensate for their hearing loss. Hearing aids will provide ample access to auditory signals to avoid late-onset auditory deprivation. Conclusion: Sufficient and consistent auditory stimulation is essential to avoid the implications of auditory deprivation.

Case Presentation 2

Outcomes of Longstanding Untreated Hearing Loss in Adults

Introduction

Hearing loss is one of the most underestimated sensory dysfunctions and one of the more common chronic health conditions (6). The prevalence of hearing impairment increases with age, but the uptake of hearing aid use in the U.S. is less than 25% (7). Hearing-impaired people are often not aware of the sounds they are missing, and the implications presented by hearing loss (6). Research has suggested that most adults do not pursue amplification because they believe that their hearing loss is not bad enough or performs adequately (6). Unfortunately, if hearing loss is left untreated, affected individuals may be at risk for auditory deprivation and associated negative consequences (8).

Auditory deprivation occurs when the hearing mechanism is deprived of sufficient auditory stimulation, which results in the atrophy of the auditory nerves and speech perception regions of the brain (9). Importantly, auditory deprivation gives rise to cross-modal plasticity, which refers to a phenomenon of the cerebral ability to reorganize and compensate for a sensory deficit (9). The brain does this by recruiting auditory cortical areas to function differently due to a lack of auditory stimulation from hearing loss (9). For example, studies have proposed that individuals with untreated hearing loss that rely on lip-reading show auditory brain functions being recruited for visual functioning instead (9). This places hearing-impaired individuals at risk for auditory deprivation and a decrease in social, behavioral, emotional, and cognitive functions, which places strain on other sensory modalities (6).

Regrettably, the effects of auditory deprivation are fast-acting. Even mild hearing loss can cause subsequent brain changes (9). Research performed on an adult with bilateral suddenonset sensorineural hearing loss demonstrated cross-modal plasticity three months after the hearing loss, secondary to auditory deprivation (9). Fortunately, with audiologic rehabilitation and intervention, the effects of auditory deprivation may be reversed (7). Audiologic intervention relies on the principles of neuroplasticity and the brain's ability to adapt to new or restored auditory input (8). Case studies have shown that individuals fitted with hearing aids demonstrate a reversal of cross-modal effects and this coincides with an improvement in speech perception and cognitive abilities (9). It must be highlighted that long periods of deprivation can limit potential treatment effects with amplification (8). With this, early and effective treatment with hearing assistive technology is necessary to offer the best opportunity for improved outcomes (8).

Case Presentation

A young woman presented to the clinic for an audiologic evaluation to assess her hearing status. Asymmetrical hearing loss was identified, with her left ear being worse than the right. Her hearing loss was initially identified in the left ear during the first few years of her life. No family history of early childhood hearing loss was indicated. At the age of four, she was diagnosed with Kawasaki's disease, which causes coronary arteries to become inflamed, restricting blood flow to the heart. A history of chronic otitis media and bilateral tympanosclerosis was discovered as well. She underwent ossicular chain reconstructive surgery on her left side during her teenage years, and a Wehrs incus prosthetic bone was placed. As a young adult, she experienced bilateral tympanoplasties and used conventional amplification with some benefits, which was used in school only. Amplification was not used for approximately 11 years before her clinical evaluation.

The patient's most recent audiological evaluation was several years prior, which revealed a mild sensorineural hearing loss through 1000 Hz, rising to normal hearing sensitivity in the right ear, and a moderately-severe mixed hearing loss in the left ear (Figure 2). Speech testing demonstrated excellent word recognition ability, bilaterally. Middle ear evaluations provided evidence of normal middle ear mobility in the right ear and reduced mobility in the left.

During her clinical encounter, audiometric test data revealed a mild to moderate sensorineural hearing loss through 1000 Hz, rising to mild through 8000 Hz (Figure 3). The left ear data demonstrated moderate to moderately-severe mixed hearing loss through 8000 Hz with a moderate sensorineural notch at 2000 Hz (Figure 3). Tympanometry indicated poor middle ear mobility in both ears. Acoustic reflex thresholds for 500-4000 Hz showed absent responses in ipsilateral and contralateral conditions, although responses in the right ipsilateral condition from 500-2000 Hz were elevated. Distortion product otoacoustic emission was performed using a 12-frequency protocol from 1500-12000 Hz to assess cochlear outer hair cell integrity. Emissions were present in the right ear and absent in the left ear across all frequencies, except for a present emission found in the left ear at 3000 Hz. Speech recognition performance was excellent in both ears. Binaural hearing aids and routine audiological monitoring were recommended. The consequences of auditory deprivation and the importance of stimulating the auditory system to maintain auditory abilities and speech perception were discussed during counseling.

Discussion

Individuals and their families are often unaware of the consequences of untreated hearing loss. This may be associated with the gradual progression of the hearing loss, unknown existence of inaudible sounds, acclimation to hearing loss, compensation by use of communication strategies, and working memory (6). The patient described experiencing adequate communication performance and speech perception abilities without the use of hearing aids and was not aware of the negative outcomes associated with untreated hearing loss. Although significant signs had not yet been presented, the patient was at risk for implications related to auditory deprivation, including cross-modal plasticity, reduced speech perception abilities, and cognitive decline (8).

The detection of an auditory signal at the ear level does not guarantee its perception. Perceptual events depend on the integrity and transduction of sound from an electrical signal in the cochlea to a neural signal in the brain and, eventually, the auditory cortex (7). Amplification and audiological rehabilitation are essential to avert the deleterious effects of auditory deprivation (8). Research has demonstrated that clinical intervention with properly prescribed amplification can restore cortical organization, improve speech perception, and increase cognitive benefit (8). From this, it is evident that services should include routine audiometric monitoring and a clinical evaluation of cognitive reserve for favorable long-term outcomes. Also, treatment with hearing aids is necessary to ensure that sufficient auditory stimulation is achieved and that the detrimental effects of auditory deprivation are avoided.

Conclusion

Auditory deprivation can affect individuals with various degrees of hearing loss. This case was included to demonstrate that treatment for hearing loss is essential to avoid the harmful effects of auditory deprivation. When the brain ceases to receive sufficient auditory stimulation, it reallocates its cognitive abilities to other task centers. Thus, its ability to process sounds becomes lost over time, which impacts auditory ability, cognitive function, and quality of life. Although hearing-impaired populations may exhibit satisfactory communication performance without amplification, subtle signs of auditory deprivation from untreated hearing loss can arise later in life. Routine audiologic monitoring, treatment, and counseling are necessary to ensure appropriate stimulation of the auditory system to maintain quality of life. Further research is needed to examine longstanding hearing loss in the young adult population and the prognosis of cognitive abilities during early stages of auditory deprivation for optimal outcomes from intervention.

References

1. Wolfe, J. (2020). Cochlear Implants: Audiological Management and Considerations for Implantable Hearing Devices. *Plural Publishing.*

2. Vaerenberg, B., Smits, C., Ceulaer, G. D., Zir, E., Harman, S., Jaspers, N., Tam, Y., Dillon, M., Wesarg, T., Martin-Bonnito, M., Gärtner, L., Cozma, S., Kosaner, J., Prentiss, S., Sasidharan, P., Briaire, J. J., Bradley, J., Debruyne, J., Hollow, R., Govaerts, P. (2014). Cochlear Implant Programming: A Global Survey on the State of the Art. *Scientific World Journal, Article ID: 501738;*12. http://dx.doi.org/10.1155/2014/501738

3. Dillon, M. T., Buss, E., Adunka, M. C., King, R. E., Pillsbury, H. C., Adunka, O. F., & Buchman, C. A. (2013). Long-term Speech Perception in Elderly Cochlear Implant Users. *JAMA Otolaryngology Head Neck Surgery*, *139*(3); 279-283.

4. Wick, C. C., Kallogjeri, D., McJunkin. J. L., et al. (2020). Hearing and Quality-of-Life Outcomes After Cochlear Implantation in Adult Hearing Aid Users 65 Years or Older: A Secondary Analysis of a Nonrandomized Clinical Trial. *JAMA Otolaryngol Head Neck Surg.* doi:10.1001/jamaoto.2020.1585

5. Cullington, H., Kitterick, P., DeBold, L., Weal, M., Clarke, N., Newberry, E., & Aubert, L. (2016). Personalized long-term follow-up of cochlear implant patients using remote care, compared with those on the standard care pathway: study protocol for a feasibility randomized controlled trial. *BMJ Open,* 6:e011342. doi:10.1136/bmjopen-2016-011342

6. Arlinger, S. (2003). Negative consequences of uncorrected hearing loss- a review. *International Journal of Audiology*, 42:2 S17-2 S20. Doi: 10.3109/14992020309074639

7. Lin, F. R. & Albert, M. (2014). Hearing Loss and Dementia – Who's Listening? *Aging Mental Health*, 18(6): 671–673. doi:10.1080/13607863.2014.915924.

8. Glick, H.A. & Sharma, A. (2020). Cortical Neuroplasticity and Cognitive Function in Early-Stage, Mild-Moderate Hearing Loss: Evidence of Neurocognitive Benefit From Hearing Aid Use. *Front. Neurosci*. 14:93. doi: 10.3389/fnins.2020.00093

9. Glick, H.A. & Sharma, A. (2017). Cross Modal Plasticity in Developmental and Age-Related Hearing Loss: Clinical Implications. *Hearing Research*, 343: 191-201. Doi: 10.1016/j.heares.2016.08.012

10. Vannson, N., James, C., Fraysse, B., Strelnikov, K., Barone, P., Dequine, O., & Marx, M. (2015). Quality of Life and Auditory Performance in Adults with Asymmetric Hearing Loss. *Audiology Neurotology*, 20: 38-43. Doi: 10.1159/000380746

Figure 1 (Case 1)

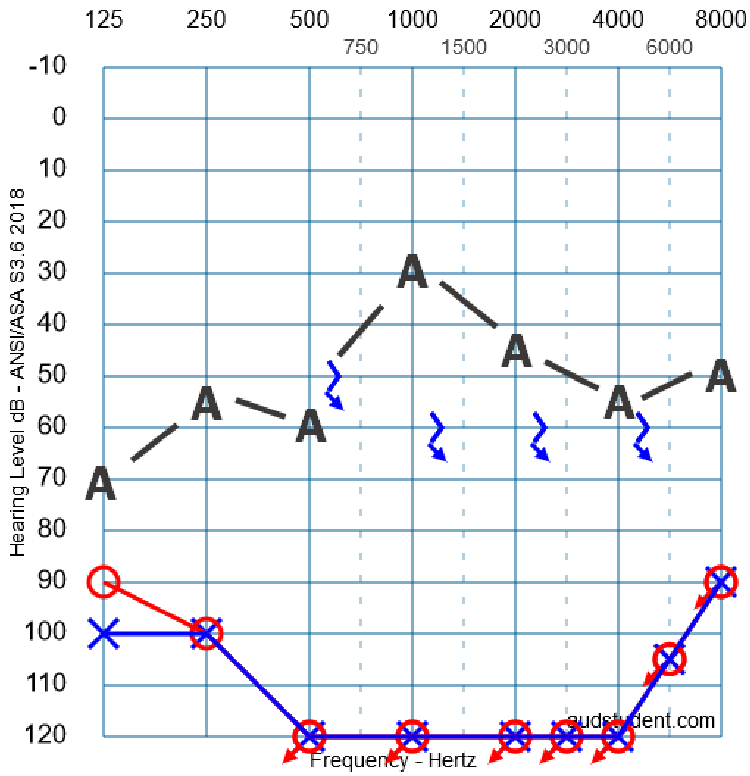

Figure 1. Unaided air and bone conduction threshold for the left and right ears. A down arrow indicates "no response". "A" is a symbol used to represent thresholds obtained under aided conditions in the sound-field.

Table 1 (Case 1)

Electrode #	T-Level	C-Level	Electrode Status
22	X	X	Disabled
21	X	X	Disabled
20	90	130	Active
19	90	130	Active
18	X	X	Disabled
17	X	X	Disabled
16	X	X	Disabled
15	X	X	Disabled
14	98	132	Active
13	120	155	Active
12	130	170	Active
11	100	140	Active
10	100	140	Active
9	110	150	Active
8	100	140	Active
7	120	160	Active
6	120	160	Active
5	X	X	Disabled
4	120	160	Active
3	X	X	Disabled
2	125	165	Active
1	145	185	Active

Table 1. Psychoacoustic Worksheet of Cochlear Implant Mapping.

Table 2 (Case 1)

Ling Sound	Soundfield Detection Level (dBHL)
/ah/	30 dBHL
/oo/	50 dBHL
/ee/	55 dBHL
/sh/	75 dBHL

Table 2. Ling Sound Detection Test Results.

Figure 2 (Case 2)

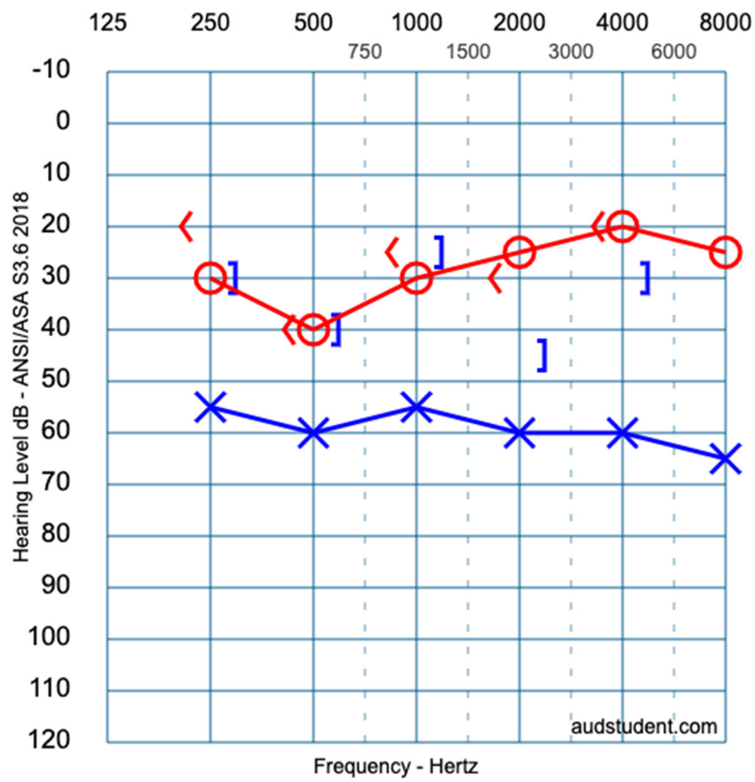

Figure 2. Unaided air and bone conduction thresholds for the left and right ears.

Figure 3 (Case 2)

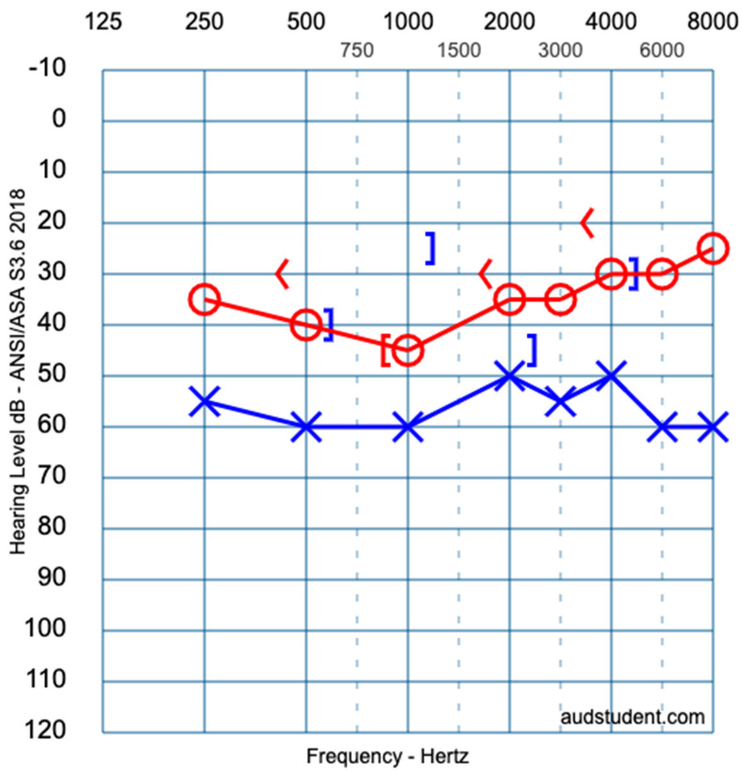

Figure 3. Unaided air and bone conduction thresholds for the left and right ear were obtained seven to eight years following the Figure 2 (Case 2) audiogram.