Spring 3-13-2023

Alternative Techniques for Amplification: A Capstone Case Series

Toryn Davis
trdavi1@ilstu.edu

Antony Joseph
Illinois State University, arjosep@ilstu.edu

Toryn R. Davis, B.S.

For Fulfillment of the Doctor of Audiology Degree

Illinois State University, Normal, Illinois

Alternative Techniques for Amplification: A Capstone Case Series

March 2023

Advisor: Antony Joseph, MA, Au.D., Ph.D., ABAC, CCC-A, CPS/A, F-NAP

Table of contents

Title Page ……. 1

Table of Contents ……. 2

Abstract 1 ……. 3

Case Presentation 1: Cochlear implantation candidacy in a patient with sensorineural hearing loss and Charcot-Marie-Tooth disease ……. 4

Abstract 2 ……. 8

Case Presentation 2: Cochlear implantation candidacy in a pediatric patient with congenital single-sided deafness ……. 9

References ……. 12

Table/Figure (Case 1) ……. 14

Table/Figure (Case 1) ……. 15

Table/Figure (Case 2) ……. 16

Table/Figure (Case 2) ……. 17

Cochlear implantation candidacy in a patient with sensorineural hearing loss and Charcot-Marie-Tooth disease: A Case Report

Abstract 1

Introduction: Charcot-Marie-Tooth (CMT) disease is a rare and chronic hereditary disorder that causes motor and sensory damage to the peripheral nervous system. As the disease progresses, hearing sensitivity may worsen.

Case Presentation: A young adult-aged patient presented to the clinic with congenital hearing loss and difficulty with conversational speech in quiet and noisy conditions.

Discussion: When encountering a patient with CMT, clinicians should be cognoscente of amplification options, especially if nerve pathways begin to deteriorate. When audiologic testing reveals a progression of hearing loss, alternative amplification may be indicated because conventional hearing aids may not provide enough benefit for CMT cases.

Conclusion: Cochlear implantation is a valid option for individuals with CMT if they meet the candidacy requirements and no longer perceive a benefit from conventional hearing aids.

Keywords: Charcot-Marie-Tooth disease, CMT, Cochlear implantation, amplification, sensorineural, hearing loss

Cochlear implantation candidacy in a patient with sensorineural hearing loss and Charcot-Marie-Tooth disease: A Case Report

Introduction

Charcot-Marie-Tooth (CMT) disease is classified in a heterogenous group of genetic disorders that weaken the peripheral nervous system. The disease is progressive and deteriorates the neurological pathways of the motor and sensory nerves, leading to demyelination of the pathways and axon firing dysfunction (Szigeti & Lupski, 2009). The etiology of hearing loss in patients with CMT is unknown, but, depending on the genetic mutation, the disease can lead to sensorineural hearing loss (SNHL) and very poor speech recognition ability (Anzalone et al., 2018). It has been hypothesized that hearing loss in patients with CMT is caused by the auditory signal failing to reach the brainstem due to weakened neurological pathways and cochlear nerve dysfunction (Goswamy et al., 2012). Additionally, this disease has several sub-classifications, although the two most common categories are CMT1 and CMT2. A diagnosis of CMT1 is primarily associated with demyelination of the nerve pathways, while CMT2 presents with axonal dysfunction accompanied by normal nerve conduction velocities at less than 25 m/s (Giuliani et al., 2019). The basis of this condition suggests that amplification options, such as cochlear implants, should be considered in the care plan for those with CMT.

Case Presentation

A young adult woman was seen for a comprehensive audiological evaluation and hearing aid selection appointment following a chief complaint of poor speech perception and extreme difficulty hearing. Significant medical history included a diagnosis of CMT at the age of three years. Her father, paternal grandmother, and paternal aunt were diagnosed with CMT and had cochlear implants. Secondary symptoms of her CMT diagnosis included gait and balance difficulties, fatigue, anxiety, depression, problems with vision, and SNHL. She presented to the

clinic with a documented bilateral SNHL that was diagnosed at age ten years and was gradually worsening.

An audiological evaluation from a different facility determined that her right hearing aid had reached its highest obtainable low-frequency gain. Due to loudness discomfort and poor clarity, she refrained from using her hearing aids for a few years. Assessment results suggested severe, rising to moderately-severe sensorineural hearing loss in the right ear and mild to moderately-severe sensorineural hearing loss in the left ear (Figure 1). Using full visual cues, speech recognition thresholds were obtained at 85 dBHL for the right ear and 50 dBHL for the left. With contralateral masking present, word recognition performance was *very poor* at elevated intensity levels in both ears (16% at 86 dBHL and 28% at 75 dBHL).

Her acoustic reflex pattern was consistent with bilateral severe sensory hearing loss. Pure tone air and bone conduction audiometry revealed severe to profound mixed and sensorineural hearing loss through 3000 Hz, rising to severe loss from 6000-8000 Hz in the right ear. In the left ear, precipitous moderate to severe sensorineural hearing loss rising to moderate at 4000 Hz with a severe loss from 6000-8000 Hz was observed (Figure 2). Speech recognition thresholds with visual cues were obtained at 75 dBHL in the right ear and 70 dBHL in the left. Speech awareness thresholds were obtained via monitored live voice presentation of */ba/* at 60 dBHL for the right ear and 40 dBHL for the left ear. Word recognition scores were attained using recorded Phonetically Balanced Kindergarten (PBK) word lists and were *extremely poor* with 4% in the right ear at 95 dBHL and 4% in the left ear at 90 dBHL.

When compared to previous test results, a significant worsening of air and bone conduction thresholds was evident, as well as a worsening of word recognition scores in both ears. The Speech, Spatial, and Qualities of Hearing Scale (SSQ) was administered, and her

responses were indicative of extreme difficulty understanding speech and localizing to sound. The patient also reported dissatisfaction with the unnatural and distorted quality of her hearing. She expressed interest in cochlear implants due to the minimal benefit of hearing aids and was scheduled for a full cochlear implantation candidacy evaluation.

Discussion

Awareness of alternative amplification options when hearing aids no longer provide benefits, data becomes a critical component of care. When speech recognition performance was reviewed, it was evident that this patient could no longer benefit from hearing aids. Thus, counseling on cochlear implants as an amplification option was administered,

In the United States, CMT is characterized as a prevalent neurogenetic disease affecting about 150,000 individuals, and there are few studies published on how to manage hearing loss in individuals with this genetic mutation (U.S. National Library of Medicine, 2014). Only three case reports describing the use of cochlear implants for individuals with CMT were discovered in the literature.

Postelmans and Stokroos (2006) described word recognition performance for a 53-year-old female with CMT1 six months after the initial right ear cochlear implantation surgery to be 59%. Goswamy et al. (2012) reported the use of open-set BKB sentences during the pre-implantation phase, and at one week, three months, nine months, and twenty-one months after the surgical procedure. This study revealed zero-percent word recognition before implantation and 53% at post-surgical follow-up (Goswamy et al., 2012). Anzalone et al. (2018) described similar findings, reporting improvements in speech recognition with the AzBio sentences and CNC phoneme testing three weeks post-surgery.

Conclusion

Cochlear implants are a viable option for the care of individuals with hearing loss who no longer perceive benefit from conventional amplification. Audiologists should consider alternative options when hearing aids are no longer powerful enough to provide sufficient audibility. Furthermore, additional research is indicated to determine the effectiveness and long-term benefits of cochlear implantation for those with CMT.

Cochlear implantation candidacy in a pediatric patient with congenital single-sided deafness (SSD): A Case Report

Abstract 2

Introduction: Congenital or acquired, single-sided deafness (SSD) may be defined as the inability to hear in one ear, resulting in unilateral hearing loss. Research data supports cochlear implantation for the management of SSD to preserve residual hearing in the affected ear.

Case Presentation: A young pediatric-aged patient presented with a congenital SSD. Her father reported minimal benefit from the prescribed hearing aid.

Discussion: Amplification alternatives were recommended based on current data because the use of a single conventional hearing aid might not provide enough benefit for patients with SSD.

Conclusion: Cochlear implantation is a viable option for individuals with SSD, as long as they meet the new candidacy requirements.

Keywords: Single-sided deafness, SSD, cochlear implantation, amplification, congenital

Cochlear implantation candidacy in a pediatric patient with congenital single-sided deafness (SSD): A Case Report

Introduction

Unilateral hearing loss affects an estimated 2.5-6% of school-aged children, resulting in a higher risk for many secondary problems (e.g., speech, language, behavior, and cognition) affecting development and success in school (Park et al., 2022). A more severe type of unilateral hearing loss may be classified as single-sided deafness (SSD). Those with SSD typically experience frustration with speech understanding in the presence of noise, difficulty with sound localization, debilitating tinnitus, and reduced perception of quality of life (Galvin et al., 2019). In the past, SSD was typically treated with contralateral routing of signal (CROS) amplification or a bone-anchored hearing aid. Instead, cochlear implants (CI) may be considered, for individuals diagnosed with SSD; however, cochlear implantation has just recently been approved by the federal Food and Drug Administration (FDA) for such cases (Tokita et al., 2014). In 2022, two CI manufacturers received FDA approval for CI procedures for individuals with SSD aged 5 years and older (Park et al., 2022). This case report aims to discuss cochlear implantation as a form of treatment for a patient diagnosed with SSD.

Case Presentation

A young female, elementary-school age, was seen for a comprehensive audiological evaluation appointment with a chief complaint of poor perceived benefit when using a single conventional hearing aid. Born by Cesarean section at 38 weeks gestation, her auditory brainstem response examination confirmed a significant hearing loss in the left ear, with no known family history of congenital hearing loss. From birth to age three, she received speech and language early intervention services.

An audiological evaluation documented severe to profound unilateral sensorineural hearing loss affecting the left ear (Figure 3). A diagnostic distortion product otoacoustic emissions (DPOAE) test was performed at a follow-up appointment. For the right ear, DPOAEs were present and robust, suggesting normal or near-normal cochlear function at the level of the outer hair cells for the tested frequencies. For the left ear, DPOAEs were absent, which is consistent with the definitive severity of the hearing loss. Pure tone air- and bone-conduction audiometry was performed from 125-8000 Hz. Responses revealed hearing within normal limits in the right ear and severe to profound sensorineural hearing loss in the left ear (Figure 4).

For the right ear, the speech reception threshold was 10 dBHL and, for the left ear, there was no response at the limits of the audiometer. Thus, for the left ear, sound awareness thresholds were attained via monitored live voice presentation of */ba/* at 75 dBHL with contralateral masking. Word recognition scores, using recorded Phonetically Balanced Kindergarten (PBK) word lists, were *excellent* in the right ear at 45 dBHL and *extremely poor* (0%) in the left ear at 110 dBHL. Her behind-the-ear hearing aid was programmed, and she was encouraged to wear the device while using an FM system at school.

Discussion

While CI treatment for adults with SSD has been supported, CI use for pediatric cases with SSD requires efficacy research. Studies support the use of pediatric auditory rehabilitation for SSD, however, Arndt et al. (2015) reported improvement of binaural hearing ability in quiet and noise, and for sound localization in children with SSD who received a CI. Some might argue that a child with SSD would not accept a CI due to good audibility in the better ear. Ganek et al. (2020), researched the acceptance of cochlear implants for 23 children with SSD. Through data logging, the authors discovered that subjects wore the CI for about 6.2 hours per day and an

analysis of their listening environments revealed that children with SSD frequently communicated in quiet sound environments under more audible speech conditions. Quieter settings are considered more favorable for speech and language acquisition than noisy environments. To achieve language, listening, learning, and neural development for children with SSD, CIs are a viable care plan consideration (Ganek et al., 2020).

Park et al. (2022) presented the characteristics for CI candidacy and included medical and audiologic considerations, age and duration of deafness, experience with conventional technology, family counseling and readiness, and candidacy determination. The most important technical recommendations for children with SSD were discussed: (a) a high-resolution 3D MRI of the inner ear; (b) for children with a diagnosis of unilateral hearing loss, an aided and unaided CI evaluation should be conducted; (c) trial amplification with signal re-routing devices (e.g., contralateral routing of signal devices) are not suggested for families that prioritize bilateral input; (d) a spatial hearing assessment should be conducted; and (e) children should complete individualized listening therapy sessions (Park et al., 2022).

Conclusion

To achieve optimized listening and language development, behavioral development, and binaural auditory access, CIs are a viable option for children with SSD. Clinicians should consider alternative amplification, as opposed to only conventional devices, for young patients with SSD. Furthermore, future research should be conducted to determine the effectiveness and long-term benefits of cochlear implantation with the younger SSD population.

References

Case 1

Anzalone, C. L., Nuhanovic, S., Olund, A. P., & Carlson, M. L. (2018). Cochlear Implantation in Charcot-Marie-Tooth Disease: Case Report and Review of the Literature. *Case Reports in Medicine*, 1–3. https://doi.org/10.1155/2018/1760978

Giuliani, N., Holte, L., Shy, M., & Grider, T. (2019). The audiologic profile of patients with Charcot-Marie Tooth neuropathy can be characterised by both cochlear and neural deficits. *International Journal of Audiology*, *58*(12), 902–912. https://doi.org/10.1080/14992027.2019.1633022

Goswamy J, Bruce IA, Green KM, & O'Driscoll MP. (2012). Cochlear implantation in a patient with sensori-neural deafness secondary to Charcot-Marie-Tooth disease. *Cochlear Implants International: An Interdisciplinary Journal*, *13*(3), 184–187. https://doi-org.libproxy.lib.ilstu.edu/10.1179/1754762811Y.0000000021

Postelmans JT, & Stokroos RJ. (2006). Cochlear implantation in a patient with deafness induced by Charcot-Marie-Tooth disease (hereditary motor and sensory neuropathies). *Journal of Laryngology & Otology*, *120*(6), 508–510. https://doi-org.libproxy.lib.ilstu.edu/10.1017/s0022215106000727

Szigeti, K., & Lupski, J. R. (2009). Charcot–Marie–Tooth disease. *European Journal of Human Genetics*, *17*(6), 703–710. https://doi.org/10.1038/ejhg.2009.31

U.S. National Library of Medicine. (2014). Charcot-Marie-Tooth disease. *MedlinePlus Genetics*. Retrieved March 13, 2021, from http://ghr.nlm.nih.gov/condition/charcot-marie-tooth-disease

Case 2

Arndt, S., Prosse, S., Laszig, R., Wesarg, T., Aschendorff, A., & Hassepass, F. (2015). Cochlear implantation in children with single-sided deafness: does aetiology and duration of deafness matter? Audiology & Neuro-Otology, 20, 21–30. https://doi.org/10.1159/000380744

Galvin III, J. J., Fu, Q.-J., Wilkinson, E. P., Mills, D., Hagan, S. C., Lupo, J. E., Padilla, M., & Shannon, R. V. (2019). Benefits of Cochlear Implantation for Single-Sided Deafness: Data From the House Clinic-University of Southern California-University of California, Los Angeles Clinical Trial. Ear & Hearing (01960202), 40(4), 766–781. https://doi.org/10.1097

Ganek, H. V., Cushing, S. L., Papsin, B. C., & Gordon, K. A. (2020). Cochlear Implant Use Remains Consistent Over Time in Children With Single-Sided Deafness. Ear & Hearing (01960202), 41(3), 678–685. https://doi.org/10.1097/AUD.0000000000000797

Park, L. R., Griffin, A. M., Sladen, D. P., Neumann, S., & Young, N. M. (2022). American Cochlear Implant Alliance Task Force Guidelines for Clinical Assessment and Management

of Cochlear Implantation in Children With Single-Sided Deafness. Ear & Hearing (01960202), 43(2), 255–267. https://doi.org/10.1097/AUD.0000000000001204

Tokita, J., Dunn, C., & Hansen, M. R. (2014). Cochlear implantation and single-sided deafness. Current Opinion in Otolaryngology & Head & Neck Surgery, 22(5), 353–358. https://doi.org/10.1097/MOO.0000000000000080

Figure (Case 1)

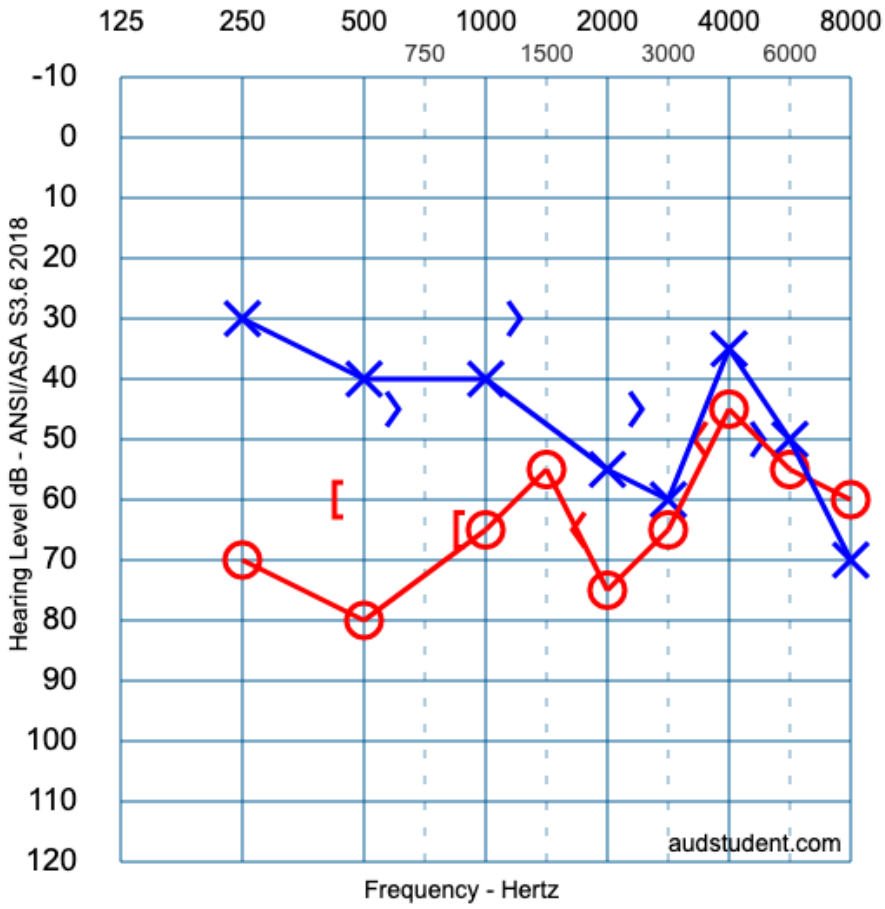

Figure 1: Audiometric data from another clinic, including air and bone conduction thresholds for left and right ears.

Figure (Case 1)

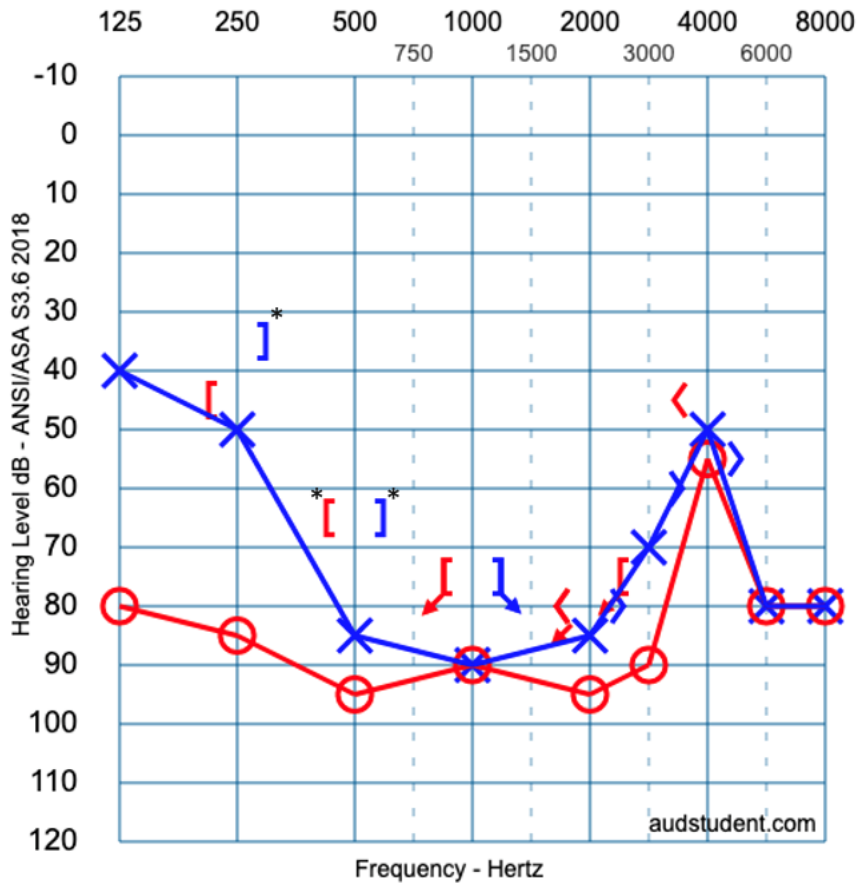

Figure 2: Audiometric data from our clinic, including air and bone conduction thresholds for left and right ears.

Figure (Case 2)

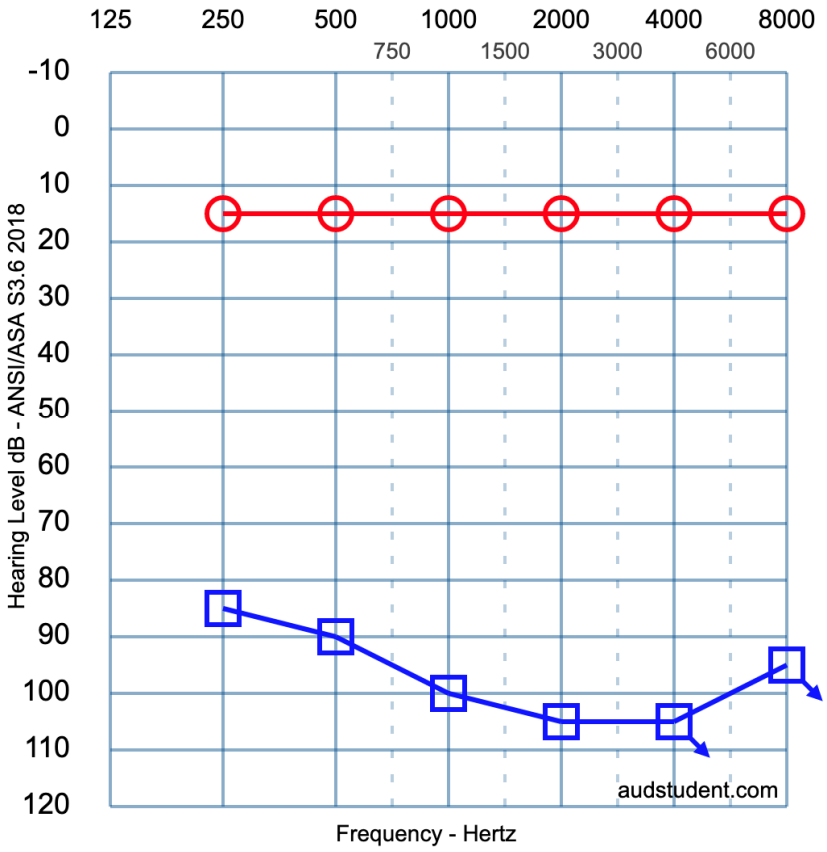

Figure 3: Audiometric data from another clinic, demonstrating air and bone conduction thresholds for the left and right ear.

Figure (Case 2)

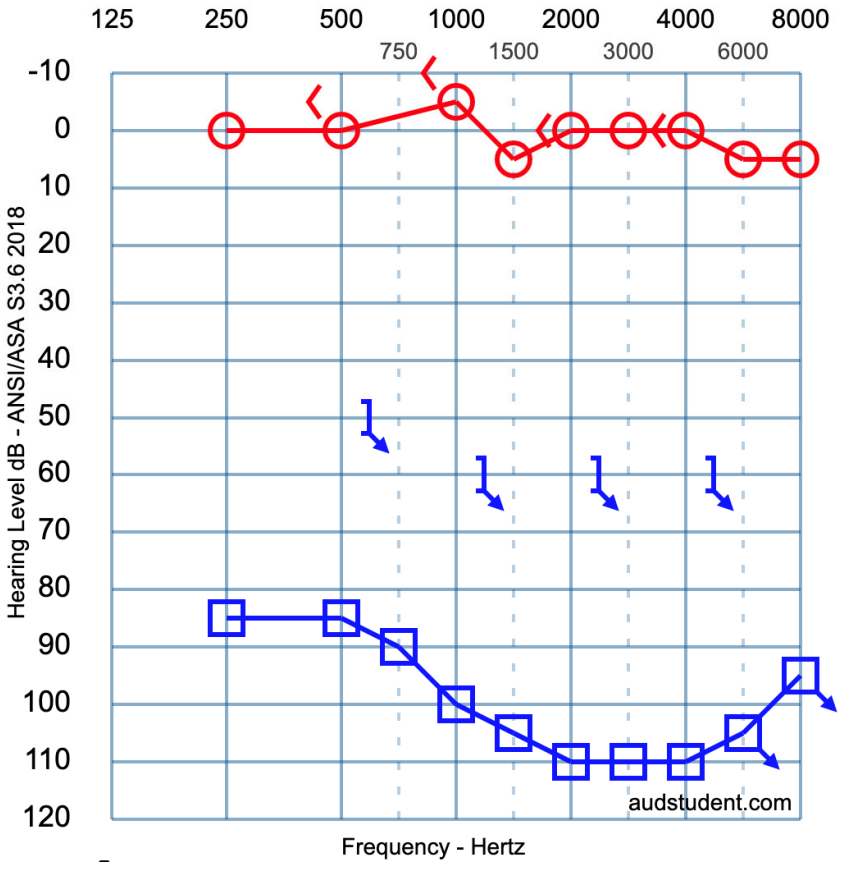

Figure 4: Audiometric data from our clinic demonstrating air and bone conduction thresholds for the left and right ear.