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Case Reports on Disorders of Sound Tolerance: Misophonia and Hyperacusis

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Abstract 1

Introduction: Misophonia is a rare disorder affecting approximately one in 1,500 people, of which, a small percentage are pediatric patients. Misophonia is characterized by the dislike of an explicit sound in a specific context. Treatment for misophonia includes a four-step program that can be modified for pediatrics. Case Presentation: A young female presented to the clinic with a significant distressed reaction when in the presence of specific sounds that included chewing, clicking pens, and loud music bass tones. Discussion: Treatment of misophonia is comparable to a care-plan for both tinnitus and hyperacusis. For this case, a four-step approach was used for treatment that entailed listening to something enjoyable, allowing a trusted adult to control the volume of an enjoyable sound, participating in enjoyable activities, and gradually becoming desensitized to bothersome sounds. Conclusion: Additional misophonia research in young populations is needed in order to develop treatment programs that may be individualized.

Case Presentation 1

Pediatric Misophonia: A Case Report

Introduction

Misophonia is an involuntary negative emotional reaction to a specific sound in a designated context (1). Misophonia is a disorder that affects approximately 15% of adults in the United States and is reported most often by female adults (1). A review of the literature did not detect a published prevalence rate of misophonia in young populations. One survey reported that the onset and severity of misophonia was greatest in individuals under the age of 18 years (1). The audiometric profile of people with misophonia varies, but most individuals have normal hearing (2). Sounds that elicit an emotionally distressed reaction are defined as trigger sounds, and many patients share common trigger sounds. The most common triggers are chewing, breathing, and repetitive sounds (3).

Misophonia is commonly confused with hyperacusis and phonophobia. Hyperacusis is described as that which occurs when a patient experiences discomfort related to the loudness of sound; whereas, phonophobia is related to the anxiety that accompanies anticipation of sound (4). The onset of misophonia commonly occurs in childhood and young adulthood (5). A standard protocol for treating patients with misophonia entails gradual desensitization and increased positive association to sound. This type of approach is person-centered and can be modified for young patients by including friendly instruction and the opportunity for the affected person to gain control (6). Such treatment has demonstrated successful outcomes by decreasing the observed misophonic reaction (5).

Case Presentation

A teenaged student presented to the clinic with a distressed reaction in response to specific sounds. Chewing, repetitive noises, clicking of pens, and low-frequency sound in music were reported by the patient as sounds that triggered distress. The patient reported that her reaction to these sounds were evident for five years but had worsened within the past month. Sequestered in her bedroom, she was avoiding school and had been using a sound generator. Under the care of a therapist, she had been diagnosed with anxiety and depression and was prescribed medication. Employed by a chain restaurant, the patient reported that the ambient noise levels at work allowed her to do her job without difficulty. She frequently wore earbuds at school while listening to white noise.

After an audiological evaluation, the patient completed the Tinnitus Handicap Inventory (THI), obtaining a score of 68, which was indicative of *severe* tinnitus handicap. Audiometric testing revealed that hearing was within normal limits for 250-8000 Hz (Figure 1) with air conduction responses no poorer than 15 dBHL in the right and 10 dBHL in the left ear. Distortion product otoacoustic emission (DPOAE) testing indicated normal cochlear outer hair cell function (Figure 2). Uncomfortable loudness levels (UCLs) demonstrated normal sound level tolerance. After interpretation of the examination data, a diagnosis of misophonia was reached.

Management

Treatment for misophonia included prescription of Oticon OPN 3 miniRITE hearing aids. The Oticon OPN 3 miniRITE hearing instruments were chosen to replace the earbuds that were reported as being used at school by the patient. The devices were equipped with four tinnitus programs, including red noise, shaped noise, white noise, and pink noise, and the patient was instructed to use these noises for all waking hours at a comfortable level. A gradual desensitization plan was administered, and the patient was instructed to listen to noise while being exposed to the bothersome sounds for 15 minutes each day, at least four to five times per week. She was encouraged to gradually decrease the white noise level, then allow a trusted adult to set the intensity of an unpleasant noise to what they believed was a normal volume level and continue to decrease the intensity of the white noise. As that exercise became increasingly less difficult, the patient was instructed to allow a trusted person to eat in the same room, then gradually lower the white noise. Besides gradual desensitization, the patient was instructed to increase positive association to sounds. To do this, we recommended that she listen to something enjoyable without any other activities for 20 minutes each day, at least four to five times per week. Enjoyable sounds that were suggested include music, podcasts, YouTube videos, and other preferred sources of entertainment.

Discussion

Misophonia is distinguished from other abnormal auditory perceptions by the intense physical and emotional reaction to specific trigger sounds (1). An effective method for differentiating misophonia is a comprehensive diagnostic audiologic evaluation. This approach should include pure-tone air and bone conduction thresholds, DPOAEs, tympanometry, UCLs, and speech audiometry. Audiometric testing should provide validation of the patient's reported concerns, but the most important information for diagnosis of misophonia is the patient's case history and its documentation. The application of UCL testing should allow the clinician to rule out hyperacusis.

There are several comorbidities that accompany misophonia, which often include anxiety and depression. These psychological conditions may occur simultaneously and, when indicated, the patient should be referred to a mental health or related specialist (1). In addition, it has been reported that Cognitive Behavioral Therapy (CBT) is effective for treating misophonia (1). Misophonia is primarily diagnosed based on the patient's reports of trigger sounds and the emotional reaction that accompanies those sounds; therefore, a short questionnaire for both children and adults might facilitate a more prompt and accurate diagnosis.

Conclusion

This case of misophonia in a young adult included treatment that focused on decreasing the misophonic reaction with gradual desensitization and increased positive association to sound. Nevertheless, further research is needed to improve diagnosis and treatment outcomes for future cases of misophonia. Improvement in diagnosis and treatment of misophonia should include standardized testing that contains otoacoustic emissions, ultrahigh frequency air conduction testing, and measurement of UCL. Additionally, for children and adults, there may be a need for the development of an increased number of evidence-based treatment plans.

Abstract 2

Introduction: Hyperacusis affects approximately 1 in 50,000 individuals, so it is classified as a rare disorder. Hyperacusis is clinically indicated by a sensitivity to moderate sounds that can lead to physical pain or discomfort, and it co-occurs commonly with tinnitus. Case Presentation: An older adult presented to the clinic with intolerance to loud sound that included music played in her church, the voices of children, and booming laughter. She frequently donned circumaural headphones in noisy listening environments to self-prevent falling into a pattern of avoidance behavior. Discussion: Treatment of hyperacusis has a strong foundation in counseling and may be accompanied by a four-step approach that is comparable to treatment for tinnitus and misophonia. The goal of this four-step approach is to target sounds that are perceived as too loud and train the affected individual to become more tolerant. Conclusion: Testing and treatment for hyperacusis is comparable to that of individuals affected by tinnitus, as it has been hypothesized that cochlear cell damage may be responsible for these disorders. Nevertheless, there is a need for a test battery specific to hyperacusis in order to differentiate other disorders of sound tolerance.

Case Presentation 2

Hyperacusis: A Case Report

Introduction

Hyperacusis occurs when an affected individual perceives sounds that are typically tolerable as too loud (7). It is classified as a rare disorder, affecting one in 50,000 people, including children (8). Individuals suffering from hyperacusis commonly complain of tinnitus as well (8). The audiometric thresholds of patients with hyperacusis are typically within normal limits, although hearing loss may be present in some cases (8).

Hyperacusis has been associated with several causes, which include Bell's palsy, Lyme disease, overexposure to hazardous levels of noise, and autism, and avoidance behavior is a frequent consequence of the condition (9). Those suffering from hyperacusis often use earmuffs when exposed to the moderate sound intensity of daily life. Use of earmuffs can produce a compounding effect of further intolerance to moderate sound (9). Overall, individuals afflicted with hyperacusis may experience significant disruption to their quality of life.

It has been hypothesized that the biological cause of hyperacusis is at the structure of the auditory nerve (10). Animal studies have suggested that hyperactivity is not the cause of the hyperacusis, but, conversely, the nature of sound intolerance is that which stems from the cochlea (10). The idea of hyperacusis originating from a peripheral auditory component, such as the cochlea, provides a reasonably plausible connection between hyperacusis, presbycusis, and tinnitus (10). This theory purports that hyperacusis, presbycusis, and tinnitus each derive from damaged cochlear-nerve cells.

Treatment for hyperacusis has the same structure as treatment for tinnitus and other disorders of sound intolerance. When audiologists attempt to quantify the impact of the hyperacusis, they may collect loudness tolerance measurements such as uncomfortable loudness levels (UCL), accompanied by sound perception inventories (11). Similar to individuals suffering from tinnitus, sound therapy may be administered in conjunction with cognitive behavioral therapy (CBT) to increase sound tolerance (11). This procedure has demonstrated that most individuals with hyperacusis demonstrate improvement within six to twelve months (11).

Case Presentation

An older adult female presented to the clinic with decreased sound tolerance. The environment in which she was reportedly most affected was at church services, where she relied on the use of circumaural headphones. Other environments that presented discomfort for her were restaurants and exercise classes, especially when there was loud laughing, children screaming, or high-intensity music being played. She had been suffering with this condition for six to twelve months and explained that she had not avoided any of her typical activities due to the availability of headphones. A history of frequent migraines accompanied her hyperacusis.

Before audiological testing was conducted, the patient responded to the Modified Khalfa Hyperacusis Questionnaire (MKHQ) and a Patient Health Questionnaire (PHQ-9). She obtained a score of 58 on the MKHQ, which is indicative of a *moderate* handicap. A score of 2 on the PHQ-9 was evidence of no concerns for depression. Audiometric testing conducted prior to her appointment with the clinic revealed hearing within normal limits through 2000 Hz, with a moderately-severe sloping sensorineural hearing loss, bilaterally (Figure 3). During the examination, she experienced aversion (sound intolerance) at threshold intensity levels during high-frequency pure-tone testing. The aversion reported was observed as an involuntary pained facial expression. Audiometric testing completed three months later revealed similar findings, bilaterally (Figure 4). Uncomfortable loudness levels for pure tones from 250-8000 Hz were obtained at intensity levels that exceeded typical conversational speech intensities (e.g., 50 dBHL), but below 90 dBHL, indicative of decreased sound level tolerance. According to Goldstein and Schulman, a UCL recorded below 90 dBHL at any test frequency is a marker for hyperacusis (12). After interpretation of the examination data obtained, the patient was given a diagnosis of hyperacusis.

Discussion

A test battery to reliably diagnose hyperacusis should focus on calculating a dynamic range (DR). An individual's DR is calculated by the difference between pure tone audiometric threshold and UCL (12). A DR consistent with hyperacusis is 60 dBHL or less (12). Calculating dynamic range can also provide information regarding the severity of the hyperacusis. The DR and severity of hyperacusis have an inverse relationship, in that, a decrease in DR is typically associated with increased severity (12).

Outer hair cell (OHC) damage typically begins in the high frequencies. This can be caused by aging, noise exposure, and the ingestion of toxic medications. Damage to OHCs has also been theorized to cause tinnitus and hyperacusis. The damage to high frequency OHCs could cause heightened sensitivity to lower frequencies which provide intensity to speech sounds (10). This research might explain why tinnitus and hyperacusis are common comorbidities. An emotional response to hyperacusis and tinnitus may cause a cycle creating a greater handicap.

Treatment for hyperacusis included a four-step approach similar to treatment of other disorders of sound tolerance. The four steps included, (1) reduce reliance on noise-cancelling headphones, (2) avoid silence, (3) increase positive association to sound, and (4) gradually desensitize to problematic sounds. Musician's earplugs, with a 10 dBHL attenuation filter, were recommended to reduce reliance on noise-cancelling headphones. To increase positive association to sound, we recommended that she stream music, podcasts, or other sources of enjoyment for 20 minutes, several days each week. A gradual desensitization plan was indicated targeting moderately intense sounds such as restaurant noise, children screaming, and moderately loud music. She understood that this would entail spending time listening to sounds slightly below the level that she identified as intolerable.

Our patient was satisfied with recommendations made by the clinical team. To assess outcomes related to the treatment plan, a Tinnitus Functional Index (TFI) questionnaire may be administered, comparing differences for targeted items at follow-up visits. Although the TFI was designed to monitor the impact of tinnitus on individuals, it may be applied for the purpose of identifying the clinical implications of hyperacusis as well.

Conclusion

This case of hyperacusis in an older adult included treatment that focused on decreasing reliance on noise-cancelling headphones, increasing positive association to sound, and gradual desensitization. Although we believe this approach was effective for our patient, more research is needed to understand the mechanisms behind hyperacusis as well as create a reliable method for differential diagnosis.

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Figure 1 (Case 1)

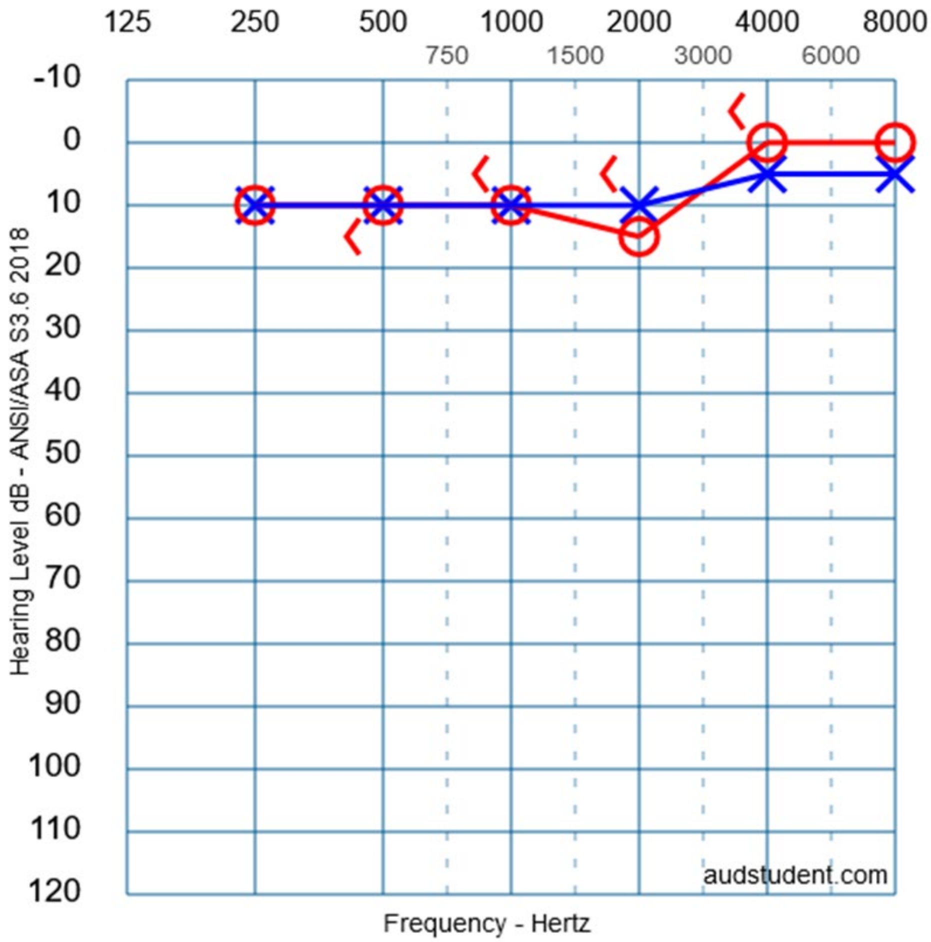

Figure 1. Audiometric data for left and right ear, including air and bone conduction thresholds.

Figure 2 (Case 1)

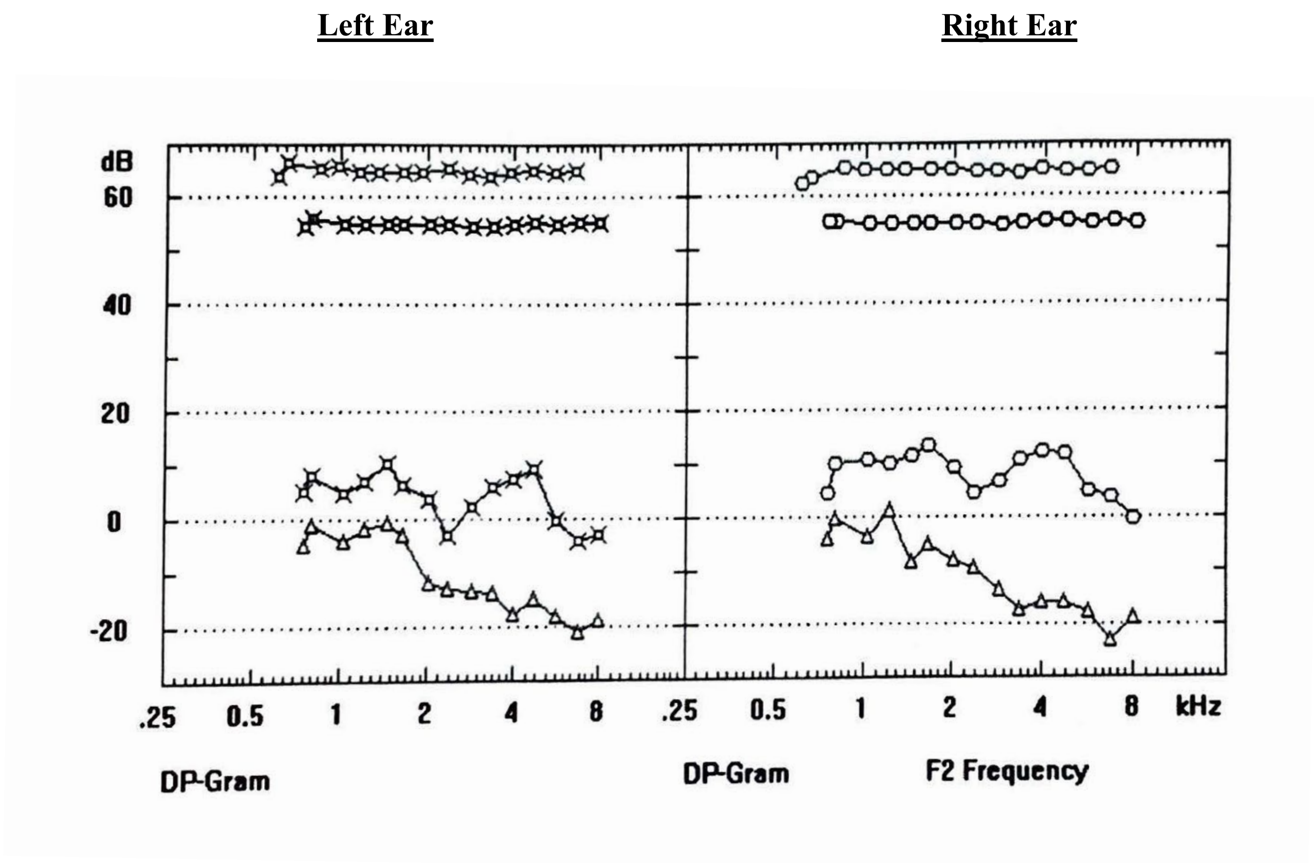

Figure 2. Distortion Product Otoacoustic Emissions (DPOAE) for left and right ear.

Figure 3 (Case 2)

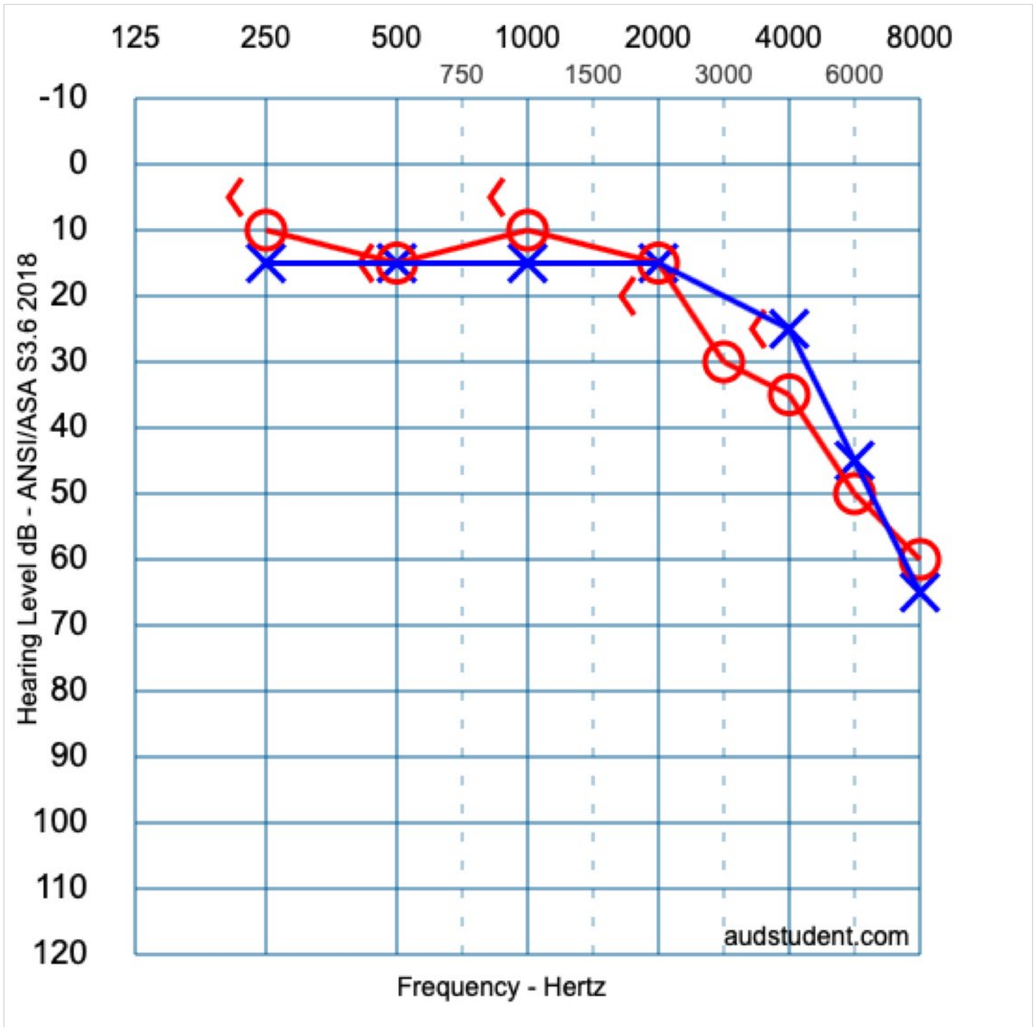

Figure 3. Audiometric data for left and right ear, including air and bone conduction thresholds.

Figure 4 (Case 2)

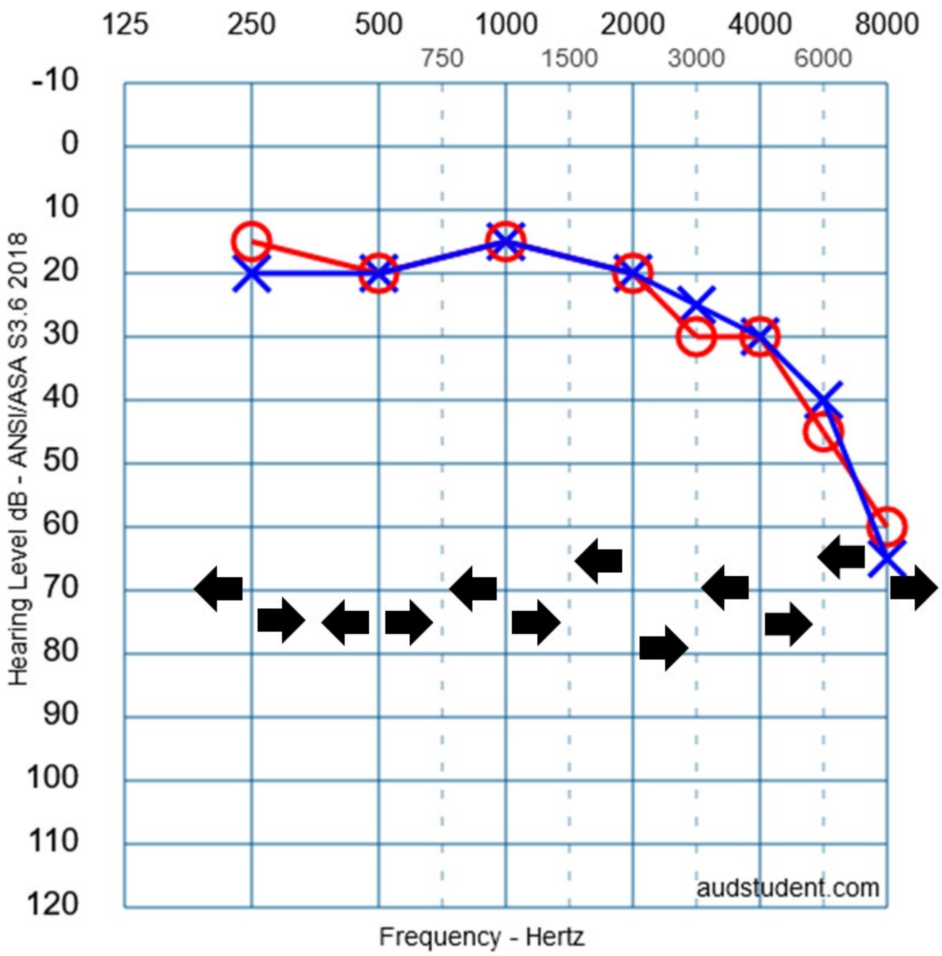

Figure 4. Audiometric data for left and right ear, including air and bone conduction thresholds and UCLs (left facing arrows for the right ear and right-facing arrows for the left).