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Management and Complications of Enlarged Vestibular Aqueduct and Meniere's Disease

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Abstract 1

Introduction: Vestibular aqueducts follow a route through the inner ear that terminates within the skull and, in most circumstances, aqueducts are narrow and bony. An enlarged vestibular aqueduct (EVA) is most commonly caused by a mutation in the SLC26A4 gene. There are other unidentified environmental and genetic causes that produce EVA as well. At the time this case was reported, no surgeries or treatments existed that could repair an EVA. Case presentation: A bilingual male child presented with congenital bilateral EVAs. His EVAs were accompanied by sensorineural hearing loss and language delay. Discussion: Enlarged vestibular aqueducts can affect language learning during the early acquisition stages. Most notably, some cultural groups may not readily accept amplification for treatment. Conclusion: It is important to provide children who have EVAs with access to developmental language opportunities. Families of children with hearing loss may need additional language support and therapy if amplification is not adopted.

Case Presentation 1

Enlarged Vestibular Aqueducts and Cultural Values

Introduction

The vestibular aqueduct contains an endolymphatic duct and endolymphatic sac and can become enlarged, causing medical complications (1). An enlarged vestibular aqueduct (EVA) can be caused by a mutation in the SLC26A4 gene or unidentified environmental and genetic factors (2). In order to be classified as enlarged, the vestibular aqueduct must be greater than one millimeter in diameter. An EVA is often accompanied by sensorineural hearing loss (2); however, the hearing loss may instead be conductive or mixed in nature (1).

Literature Review

Enlarged vestibular aqueducts are congenital and often result from problems during early childhood development (1). The pediatric population accounts for 5 to 15% of the EVAs (1). The vestibular aqueduct begins to change shape and narrow by the fifth week of life (1). The specific purpose of the endolymphatic sac is unclear; however, there is speculation that it preserves the endolymph's accurate ionic composition and volume to ensure proper transmission of nerve signals for the auditory and vestibular systems (1). While there are currently no surgical options that prove useful in correcting EVA, there is emerging research on the use of steroid therapies to reduce the severity of hearing loss (3). In addition, the avoidance of contact sports or activities that could cause head trauma or barotrauma has been recommended (3). Pendred Syndrome is a symptom associated with EVA. Pendred Syndrome is autosomal recessive, and often related to an enlarged portion of the thyroid gland, known as a goiter, and with Mondini's malformation. The goiter does not usually inhibit thyroid function (4). Pendred Syndrome is caused by two mutations on the SLC26A4 gene (2) and occurs in about 8% of all congenital hearing losses (4).

Case Presentation

This male patient was born full term by way of Cesarean section, and his mother was diagnosed with gestational diabetes. He had no family history of early childhood hearing loss. No other complications with the dyad were evident. The patient was referred as a result of his newborn hearing screening and was re-screened up through eight months of age, with results that continued to indicate referral. Subsequently, he was enrolled in early intervention services.

Around age eight months, a diagnostic auditory brainstem response test was completed, which identified a moderate sensorineural hearing loss in the right ear and severe sensorineural hearing loss in the left. At nine months of age, he was prescribed binaural hearing aids and two hours of monthly audiologic rehabilitation.

At 12 months, conventional behavioral audiometry was administered, which revealed a moderate, sloping to moderately-severe, sensorineural hearing loss in the right ear and a moderate, sloping to profound, sensorineural hearing loss in the left ear. His hearing impairment progressed to moderately-severe in the right ear and profound in the left ear.

At 18 months, the child's high frequency thresholds suggested a mild degree of hearing loss in both ears. The otolaryngologist ordered a computed tomography (CT) scan and genetic testing. At 20 months, the high frequencies worsened to a moderately-severe, sloping to severe, sensorineural hearing loss in both ears. At 24 months, CT scans and genetic tests were conducted, which provided evidence of EVAs. At that time, the patient was negative for Pendred Syndrome.

Results of audiologic testing were discovered and thresholds were stable over a four-year period (Figure 1 and 2). The most recent audiologic evaluation (Figure 2) showed mild sloping to severe rising to moderate suspected sensorineural hearing loss in the right ear and severe sloping

to profound hearing loss in the left ear. Tympanometry provided evidence of normal middle ear function in both ears. Word recognition performance was 68% in the right but was unreported in the left ear. It is unclear why word recognition testing was not attempted in the left ear, but it was likely due to the excessive degree of hearing loss, as well as patient fatigue during the encounter.

The patient's family was not amenable to cochlear implants, but a listening and spoken language program was initiated in school. Only Spanish was spoken at home. Because of an apparent articulation disorder, a program of two hours of weekly speech-language therapy was started, but no improvement was observed. To improve his communication skills, a bilingual speech language pathologist (SLP) began delivering 30 minutes of therapy weekly, yet no improvement to expressive or receptive speech-language ability was reported. The child was transitioned to a total communication environment that included sign language, which, subsequently, resulted in significant advancement.

The patient received total communication, speech and language therapy and audiologic rehabilitation services. His family began learning sign language. Preferential classroom seating, a frequency modulation (FM) amplification system that coupled to his hearing aids, checks for understanding, and notes in written and verbal form were administered. Caution during contact sports was recommended by his physician.

Discussion

The family expressed discomfort with the appearance of the hearing aids, so alternative colors were offered, and a more suitable color was selected. A child's disability may cause humiliation for the family, including a need to conceal and protect their child from external sources of judgment (5). A perceived need to conceal the hearing loss may have driven the family's opposition to cochlear implant surgery. Hence, during counseling, clinicians should be

mindful that empathetic understanding, cultural sensitivity, and family support may be key during the adjustment of the child and their family members.

This strong line of support from medical and allied health professionals is a crucial component in any diagnosis; however, it may be especially important in cases of rare pediatric disorders, such as EVA, wherein language development may be at risk. Enlarged vestibular aqueducts are irreversible and could present with other health problems such as hearing loss and Pendred Syndrome. Genetic studies provide ample information about the presence of this disorder and other comorbidities (2).

Conclusion

Enlarged vestibular aqueducts cause progressive hearing loss that can lead to delayed speech and language acquisition. Bilingualism may offer challenges with speech and language development. Thus, the use of spoken language coupled with manual communication may be beneficial for children who present with EVA and bilingualism. Pediatric patients should receive audiometric monitoring, school-based audiologic rehabilitation, and a total communication program. A medical advocate should be considered to assist the family with future decisions. Cochlear implant candidacy should be discussed when the patient is old enough to make independent medical decisions. Cultural differences may affect the manner in which families view hearing loss and treatment options, so clinicians must be insightful and competent during these circumstances.

Abstract 2

Introduction: The presence of Meniere's disease is associated with an increased volume of inner-ear endolymphatic fluid and distention of the endolymphatic space. The root cause of change in endolymph, also known as cochlear hydrops, is indefinite. There is no surgical procedure or treatment that completely eliminates effects of Meniere's disease. Case presentation: An adult male presented to the clinic with Meniere's disease, accompanied by fluctuating hearing loss that affected his social, occupational and personal life. The patient presented to the clinic for a cochlear implant candidacy evaluation. It was determined that he was unqualified for cochlear implant surgery, so hearing aid selection was conducted. Discussion: Amplification for patients with Meniere's disease may be challenging due to fluctuating hearing loss. Audiologic rehabilitation is an important step in the dispensing process in order to address all aspects of the fluctuating nature of Meniere's disease.

Case Presentation 2

Meniere's Disease and quality of life

Introduction

It has been estimated that approximately 200 of every 100,000 individuals suffer from Meniere's disease (6), which is commonly an inner-ear unilateral condition (7). Hallmark symptoms of Meniere's disease include bothersome tinnitus, fluctuating hearing loss, aural fullness and a sensation of imbalance or spinning (7). Meniere's disease is usually associated with sensorineural hearing loss (6). Increased endolymph in the inner ear leads to the associated symptoms, but a specific reason for buildup of endolymph has not been identified (7). At the time of this report, a procedure or treatment to eliminate Meniere's disease was not found (7). Common treatment options for alleviation of symptoms include the use of steroids, amplification devices, surgery and medication (7). Hearing loss associated with Meniere's disease, and many other disorders, can produce communication problems. Quality of life can be even severely affected if steps are not taken to treat the hearing loss, especially in the elderly (8).

Literature Review

Meniere's disease is diagnosed by an evaluation of hearing and assessments of balance (7). Medical investigators speculate that the symptoms associated with Meniere's disease could be due to viral infection, genetics, obstruction, individual deformity or reduced immune activity (7). Hearing loss associated with Meniere's ranges in severity. Hearing thresholds can worsen or even improve with progression of the disease (6), and the hearing loss may eventually become permanent (7). Treatment options include medication to reduce nausea associated with motion, amplification devices, injectable steroids, and surgery (7). The use of amplification devices often requires a range of programming options in order to address the possibility of fluctuating hearing

loss (9). Hearing loss associated with audiologic disorders, such as Meniere's disease, may produce anxiety and depression (6). According to the American Academy of Audiology, elderly patients with hearing loss reported increased rates of social isolation due to negative interactions with others (8).

Besides amplification, there are several medical treatment options. Injectable steroids, for example, may be introduced into the middle ear to reduce vestibular symptoms (7). In addition, surgical alternatives include severing of the vestibular nerve, deflation of the endolymphatic sac, or extraction of the inner ear balance structure (7).

Case Presentation

Upon referral by a neurologist, the patient presented to the clinic for a cochlear implant evaluation. He reported debilitating left sided tinnitus, anxiety, panic, depression, nausea, visual disturbances, hearing loss, ataxive dizziness, shortness of breath, and difficulty concentrating. The patient expressed concerns about his occupational safety due to his hearing loss. While conducting roof work, he reported having trouble with sound localization. He described his experiences with social isolation due to his vestibular symptoms and hearing loss. Prior to the surgeon's referral, audiologic testing showed sloping moderately severe sensorineural loss in the right ear. The patient had a moderately-severe sloping to severe hearing loss in the left ear (Figure 3). Word recognition performance with monosyllables in quiet was found to be 94% in the right ear and 16% in the left ear, revealing a large disparity.

During the cochlear implant candidacy appointment, audiologic testing demonstrated sloping moderate sensorineural loss in the right ear and a moderately-severe rising to moderate sensorineural loss in the left ear (Figure 4). Distortion product otoacoustic emissions (DPOAEs) were conducted due to the change in hearing and improved word recognition score. Distortion

product otoacoustic emissions (DPOAEs) were grossly absent, bilaterally. Cochlear implant candidacy testing was not conducted due to the patient's fluctuating audiologic test results. Instead, a hearing aid selection protocol was administered.

Discussion

Prescribing amplification devices for a patient with fluctuating hearing thresholds may prove to be a challenging task. In the case of hearing aids, the patient may benefit from multiple memory programs to address the occurrence of fluctuations in hearing sensitivity (9). Patients with unilateral hearing loss may benefit from the use of assistive listening devices (ALDs), a contralateral routing of signal (CROS) or bilateral CROS (BiCROS) system, or a bone anchored hearing aid (BAHA) (9). In the case of cochlear implants, patients may require follow-up appointments after hearing has improved or stabilized (10). The hearing loss for this patient fluctuated in such a way that he was no longer classified as a candidate for surgically placed cochlear implants, although it has been reported that Meniere's patients can benefit from cochlear implantation (10).

This patient had personal and occupational concerns that could be improved through audiologic rehabilitation. Fluctuating hearing loss and bothersome symptoms, such as tinnitus and aural fullness, can lead to stress and subsequent avoidance of social situations. According to the American Speech-Language-Hearing Association, audiologic rehabilitation assists patients in the hearing aid adaptation process and helps them learn new communication and advocate strategies (11). Audiologic rehabilitation can be delivered in the form of individual and group support services (11). Patients with Meniere's may find a greater degree of benefit from these services due to their complex amplification needs. When patients obtain amplification devices, rehabilitation services that include empathetic counseling and outcome measures are important

for establishing attainable goals and expectations (9), aimed at improving communication and quality of life.

Conclusion

Meniere's disease can be accompanied by debilitating symptoms such as hearing threshold shift, roaring tinnitus, bothersome aural fullness and imbalance or dizziness. This array of associated symptoms can make amplification an ongoing exercise of adjustment. Audiologic rehabilitation is a necessary part of the treatment plan for patients with Meniere's disease as it can help them overcome difficulties associated with amplification acceptance, everyday communication and social isolation.

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Figure 1 (Case 1)

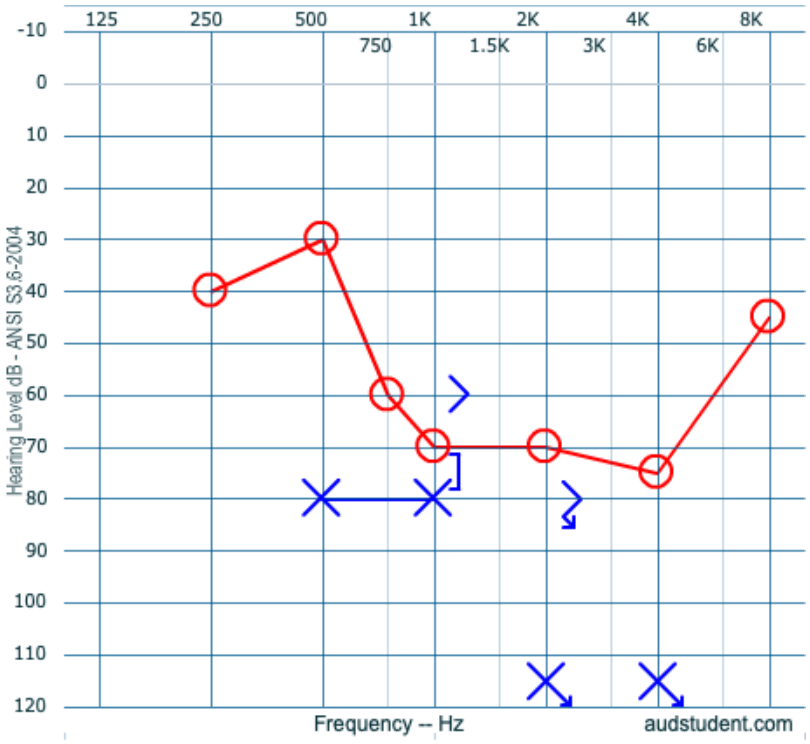


Figure 1. Initial available audiometric data for right and left ear, including air and bone conduction thresholds. An arrow pointing downward from the symbol denotes no response to the stimulus presented at that level.

Figure 2 (Case 1)

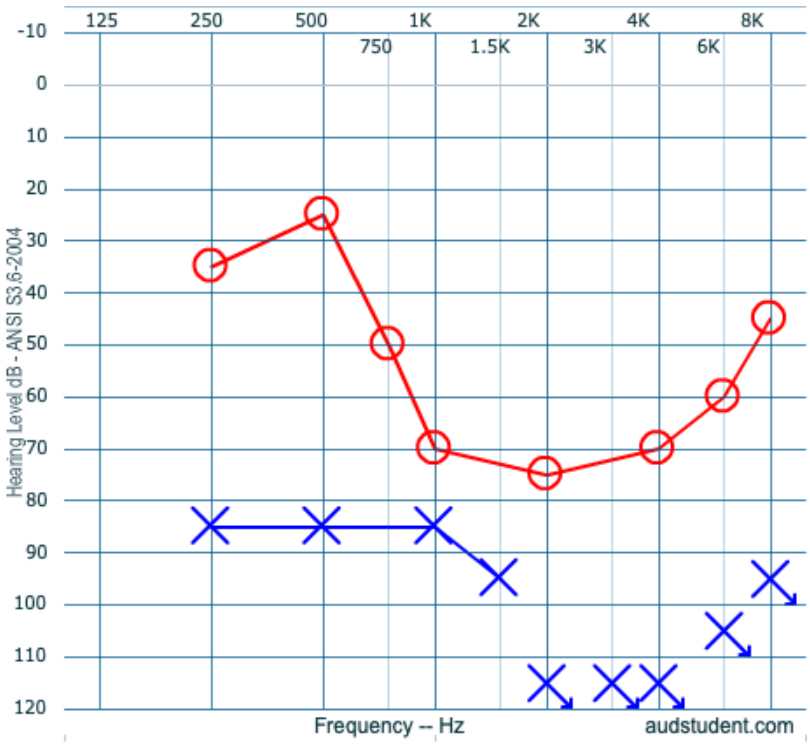


Figure 2. Most recent audiometric data for right and left ear, including air and bone conduction thresholds four years after initial testing. An arrow pointing downward from the symbol denotes no response to the stimulus presented at that level.

Figure 3 (Case 2)

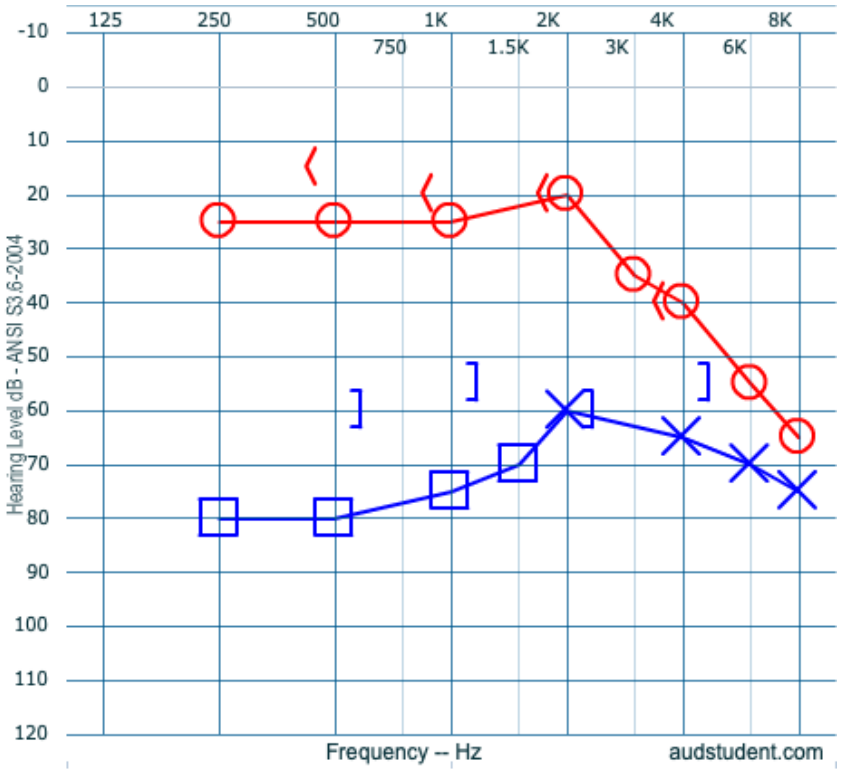


Figure 3. Initial available audiometric data for right and left ear, including air and bone conduction thresholds.

Figure 4 (Case 2)

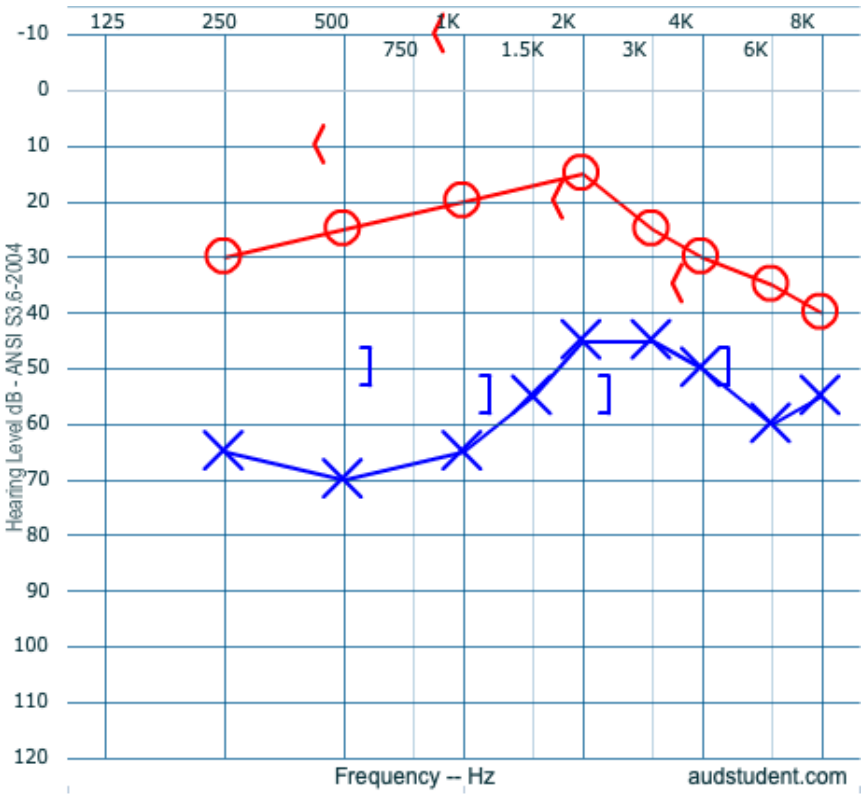
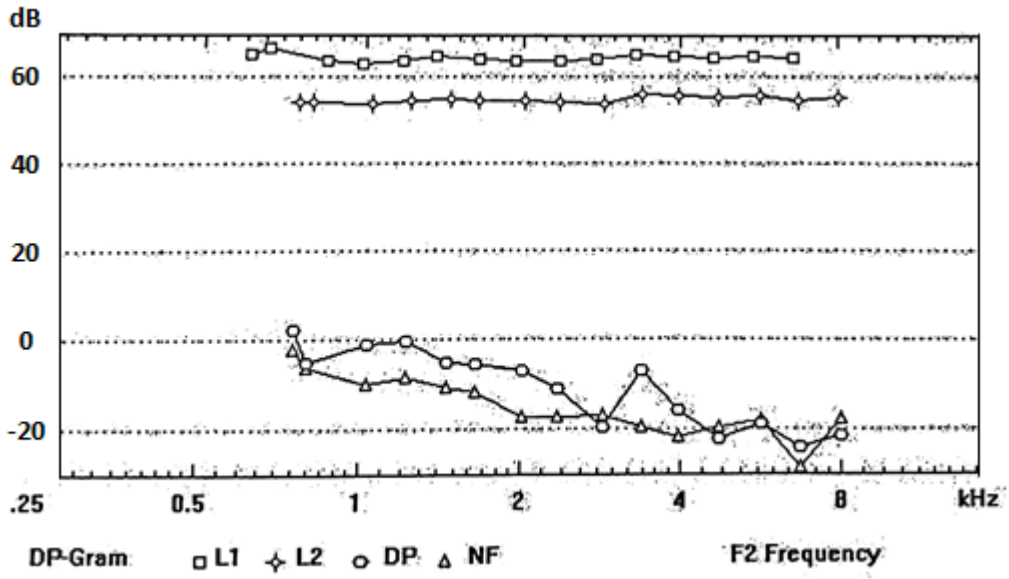


Figure 4. Follow-up audiometric data for right and left ear, including air and bone conduction thresholds 12 months following the initial exam.

Figure 5 (Case 2)

Right Ear:



Left Ear:

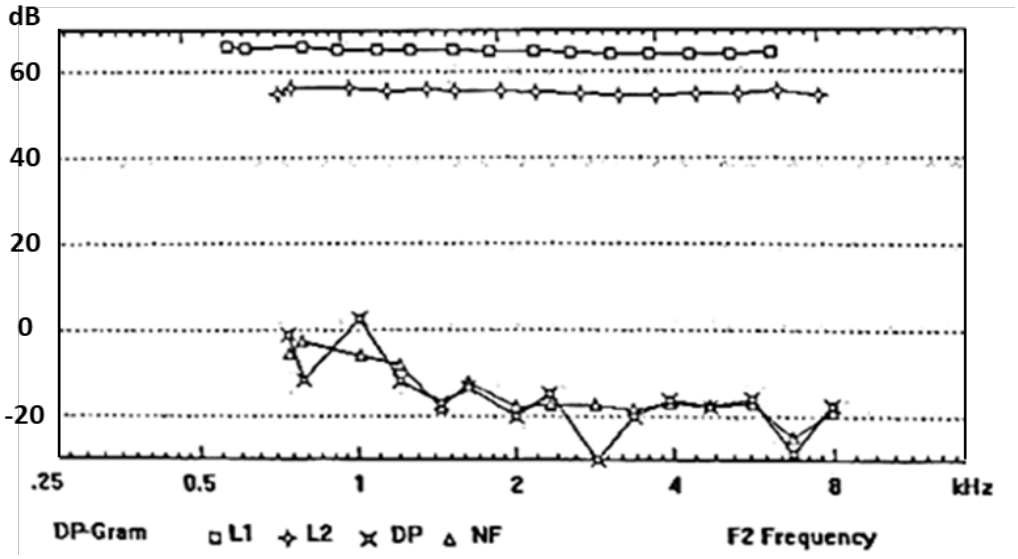


Figure 5: Distortion product otoacoustic emissions (DPOAEs) for right and left ears in follow-up examination. DPOAE appear to be grossly absent, bilaterally.