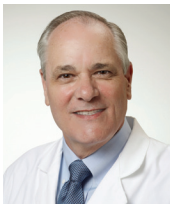
Autoimmune Inner Ear Disorders: A Case Study

By Dennis Colucci, AuD, MA

Autoimmune inner ear diseases (AIEDs) occur in less than one percent of patients with hearing loss or balance disorders (AFRF, 2012). This inflammatory condition is caused by the body's immune system acting against its own healthy cells and tissues. Taking several days up to three months to develop, AIED is known to come with tinnitus, and half of the patients have also reported experiencing vertigo (*Hearing Journal.* 2017;70[8]:6). AIED affects more men than women. It is not always a standalone disease, but can be a response from a larger disease process such as systemic lupus, Cogan's syndrome, polyarteritis nodosa (artery inflammation), or Reiter's syndrome (reactive arthritis), which is one of a number of multi-focal diseases (*Clin Case Rep.* 2014;2[6]:310). Even Ménière's disease is implicated as an autoimmune-mediated condition because of its relationship with specific genotypes. In some AIED cases, the triggering mechanism can be unknown.

AIEDs also occur because of an immune system response to infection, allergy, or toxic and foreign substances that attack normal tissue. When a foreign substance or microorganism invades the body, it is recognized as an antigen because its molecular structure does not match those of naturally occurring substances or of those recognized by the body. This stimulates various mechanisms in the form of antibodies or immunoglobulin to eliminate the intruder. There are two primary actions: one that binds with antigens to change their chemical make-up (B cells) for destruction, and another wherein antibodies digest the foreign substance directly (phagocytic cells). Once the production of antibodies is stimulated, these protective proteins may remain in the body for several months. What makes this action an autoimmune disorder is what the antibodies attack–healthy cells.

Theories on ear involvement in this disease have focused on the development of antibodies within the inner ear. These antibodies attack the normal tissue, resulting in inflammation that develops into progressive asymmetrical fluctuating hearing loss. The prevailing treatment for AIEDs includes corticosteroids and other anti-inflammatory medications for pain and other conditions. However, many patients respond poorly to medical treatment, so hearing aids are prescribed, as well as cochlear implantation or a bimodal montage for severe cases.



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CASE HISTORY

The patient is a 53-year-old man who was initially evaluated for bothersome bilateral high-frequency tonal tinnitus, which he noticed right after firearms practice six months prior to the evaluation. He reported typically using earmuffs, but at times went to firearms practice without ear protection. His medical history includes a diagnosis of Reiter's syndrome when he was 42, presumed to be secondary to a gastrointestinal infection caused by Giardia, a waterborne parasite. At present, he has some chronic joint pain, primarily on the knees and hips, for which he continues to take medication. He has no history of ear disease, tinnitus or vertigo, environmental ototoxicity, recent infection, familial hearing loss, trauma to the head or neck, barotrauma, or noise exposure, except from firearms. His unaided Abbreviated Profile of Hearing Aid Benefit (APHAB) assessment reveals a moderate degree of hearing difficulty in familiar conversations, reverberant rooms, and with background noise.

At the initial examination, the patient's audiometric test results revealed a mild to moderate, moderately sloping, high-frequency sensorineural hearing loss bilaterally (Figure). Notching at 4,000 Hz was observed in the right ear. Speech discrimination at 50 dB HL is within normal limits in either ear. At a +10 dB signal-to-noise ratio using recorded speech and speech babble, discrimination is in the 40th percentile range bilaterally. After the initial evaluation, the patient was fitted with binaural amplification.

Two years and four months after the initial evaluation, the patient returned to the clinic with a complaint of asymmetrical hearing loss that developed three months before his visit. This was accompanied by dizziness without vertigo, severe vision loss (he only sees shadows), and increased joint swelling with pain, numbness, and tingling in his legs and feet. Audiometric test results revealed a moderate to severe, mildly sloping sensorineural hearing loss in the right

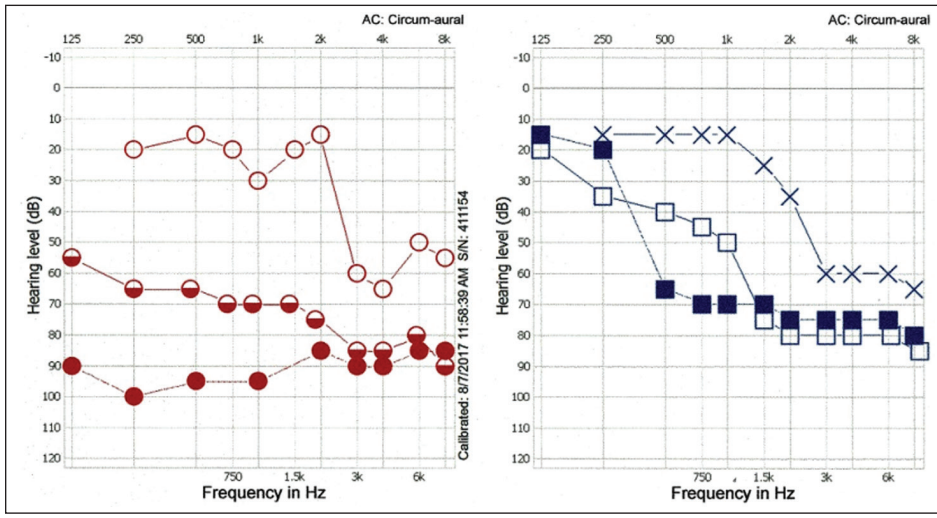

Figure. Audiometric Data: Baseline (O, X), Initial Change (◒, □), and P&S Final (●, ■).

ear and a mild to severe, moderately sloping sensorineural hearing loss in the left ear (Figure). Notably, speech discrimination measured at MCL was 12 percent in the right ear and 52 percent in the left ear. Immittance testing revealed Type A tympanograms with acoustic reflexes only present in the ipsilateral response at 500 Hz in the left ear and 1,000 Hz in the right ear. Auditory brainstem evoked potentials to clicks, electrocochleography, and DPOAEs were absent. ENG test results were consistent with a peripheral vestibular site of lesion exemplified by a 58 percent reduction of vestibular response in the left ear and left beating positional nystagmus with vision denied.

The patient had abnormal findings on saccades, pendular tracking, and optokinetic tests. However, in view of the patient's visual difficulties, the validity of these tests is questionable. Medical treatments included several courses of prednisone in addition to pain and neuropathy medications. Due to the changes in hearing, the patient chose to use amplification in his left ear pending the outcome of his treatments. There was no functional hearing in the right ear. The prevailing diagnosis was complications secondary to Reiter's syndrome.

One year and nine months later, the patient's hearing and other complaints have become stable, with a severe to profound, mildly reversing sensorineural hearing loss in the right ear and a normal low-frequency (125-250 Hz) response with a dramatically sloping to severe sensorineural hearing loss thereafter (Figure). Hearing sensitivity has worsened in his right ear, which remains nonfunctional. Notably, hearing at 250 Hz in the left ear has improved by 15 dB, and speech discrimination also improved by 30 percent since his previous evaluation. Subsequent audiometric testing revealed permanent and stationary hearing sensitivity. The patient got a bimodal hearing solution–a cochlear implant on his right ear and a hearing aid in his left ear. He no longer has dizziness or vertigo and his vision has returned to normal, although he continues to have pain, numbness, and joint swelling, for which he remains on medications.

Audiologists involved in the diagnostic evaluation of AIED patients should perform a variety of tests to define the inner ear and its central effects on hearing and balance, the auditory nerve and brainstem, and functional communication. Most importantly, audiologists must work with a neurotologist and other specialists, provide follow-up testing and reporting, fit hearing aids, and recommend cochlear implantation when appropriate.

These patients need support and assurance that the loss of hearing does not mean lifelong deafness as medical treatments are successful for some of those afflicted and for those with residual hearing loss or non-functional ears, hearing aids and cochlear implants are life-changing options to improve their quality of life. Modern technology affords the clinician many opportunities to help these patients maintain communication and become productive from the onset of this disease process.

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