
AUTOIMMUNE INNER EAR DISEASE

Autoimmune inner ear disease (AIED) is an inflammatory condition caused by an uncontrolled immune system response that attacks the inner ear causing progressive sensorineural hearing loss (SNHL) that usually starts in one ear and then affects the other ear. The body thinks a part of the inner ear should not be there and makes antibodies and immune, also called antigen-antibody, complexes that attack the inner ear. This causes blood vessel inflammation, inner ear tissue damage, and hearing loss. Antigens are the molecules that cause an immune response and antibodies are proteins that protect the body against these antigens.

AIED is rare and diagnosed only when all other causes have been ruled out. The estimated prevalence of AIED is about 15 out of 100,000 people. AIED is felt to be responsible for less than one percent of all SNHL cases, realizing many AIED cases might not be diagnosed due to lack of specific tests.

AIED is considered “primary” when the inner ear is the only organ affected. However, in 15 to 30 percent of cases, AIED is “secondary” when it occurs as part of a larger autoimmune disorder that affects the whole body, such as rheumatoid arthritis, lupus, scleroderma, ulcerative colitis, or Sjogren’s syndrome. AIED is more common among middle-aged women.

Diagnosing AIED is challenging because there is no definite blood or imaging criteria to measure. Timing the progression of hearing loss over weeks to months is an important diagnostic clue for AIED. If AIED is suspected, early corticosteroid and immunosuppressive treatment may prevent irreversible hearing loss. A multidisciplinary team approach between an ENT (ear, nose, and throat) specialist, or otolaryngologist, an audiologist, and a rheumatologist is recommended to manage the condition.

WHAT ARE THE SYMPTOMS OF AIED?

Common symptoms of AIED can include:

- Progressive SNHL in both ears that occurs over weeks to months that is not always the same in both ears
- Fluctuating hearing
- Dizziness or imbalance (approximately 50 percent of AIED cases)
- Ringing in the ears, or tinnitus
- Ear fullness (approximately 25 to 50 percent of AIED cases)
- Conductive hearing loss may be present due to Eustachian tube obstruction from inflamed middle ear lining and/or if AIED is because of systemic autoimmune diseases
- Symptoms of systemic autoimmune diseases, such as fatigue, achy muscles, swelling and redness, low-grade fever, and more

WHAT CAUSES AIED?

The causes of AIED can include:

- The body’s uncontrolled immune system attacks the inner ear protein, forming immune complexes and antibodies and causing progressive hearing loss in both ears.
- Cochlin is a protein located in the inner ear that is attacked by the immune system.
- Endolymphatic sac, a structure of the inner ear, can become dilated as the immune response of the inner ear.

AUTOIMMUNE INNER EAR DISEASE

WHAT ARE THE TREATMENT OPTIONS?

If you are having hearing loss that is getting worse in both ears over weeks to months, you should see an ENT specialist who can make a diagnosis after reviewing your hearing tests and imaging scans. If your doctor suspects that you may have AIED, you may respond well to medical therapy—steroid and immunosuppressive medication—if started early.

Corticosteroid is the main treatment to suppress the immune response to reduce inner ear swelling and inflammation. Early detection of AIED and prompt steroid treatment may help reverse your SNHL. If you cannot tolerate steroid treatment, there are alternative medications, such as cyclophosphamide, methotrexate, azathioprine, and rituximab.

Your doctor may also evaluate you for a hearing aid or other assistive listening devices. If hearing loss is substantial, your doctor may recommend a hearing device called a cochlear implant. Speak with your doctor about other specific treatment options.

WHAT QUESTIONS SHOULD I ASK MY DOCTOR?

1. Do I have a systemic autoimmune disorder?
2. What tests should I have performed to evaluate my hearing loss?
3. Do I need to get an MRI to rule out other inner ear disorders?
4. What are my treatment options for AIED?
5. What medications are available to me?
6. What other healthcare professionals should I see for AIED?