



AUTOIMMUNE INNER EAR DISEASE (AIED)

This handout is intended as a general introduction to the topic. As each person is affected differently, speak with your health care professional for individual advice.



Key points

- Happens when the immune system attacks the inner ear by mistake.
- Most cases are primary and affect only the ear.
- 30% of cases are secondary and are related to an autoimmune disease affecting the whole body.
- Often causes fluctuating (comes and goes) sensorineural hearing loss in one ear, usually getting worse over several days to months.
- Hearing loss spreads to the other ear over time.
- Half of affected people have balance and dizziness problems.
- Usually treated with corticosteroids or other immune-suppressing drugs.
- Vestibular rehabilitation (an exercise-based therapy) can help with balance and dizziness problems.
- Hearing aids and, cochlear implantation later on, may be recommended.

What is AIED?

The immune system includes many different cells and organs that help to protect our bodies from germs, parasites, and other things that can harm us. Part of how the immune system works is by telling the difference between "self" things, which are part of our own bodies, and "non-self" things, which come from outside our bodies. Specialized blood cells called B cells make proteins called antibodies, which attach to "non-self" things and alert other parts of the immune system to attack them.

Sometimes, though, the immune system makes a mistake: it makes antibodies (called autoantibodies or "self-antibodies") that attach to the body's "self" proteins. Normally, the immune system would ignore these self-proteins. But when the autoantibodies label an organ or tissue as "non-self," other parts of the immune system see it as a threat and attack it. This causes autoimmune (literally, "self-immune") disease. There are many different autoimmune diseases, and many different organs and tissues can be affected.

What are the causes?

Researchers are still studying the causes of AIED. It is not clear exactly why and how the immune system identifies the inner ear as non-self and attacks it. But the attack seems to cause inflammation and damage to the cochlea (KAA-lee-uh) the organ of hearing, and the vestibule, the organs of balance.

AIED affects about 15 out of 100,000 people.

About 3 in 10 cases of AIED are secondary AIED. This means they are caused by an autoimmune disease that affects the whole body (systemic autoimmune disease).

Many different systemic autoimmune diseases can cause secondary AIED, including:

- ankylosing spondylitis
- Behcet's disease
- Cogan's disease
- relapsing polychondritis
- rheumatoid arthritis
- scleroderma
- Sjogren's syndrome
- systemic lupus erythematosus (SLE)
- ulcerative colitis
- vitiligo
- Vogt-Koyanagi-Harada syndrome
- Wegener's granulomatosis

What are the symptoms?

AIED causes sensorineural hearing loss that usually happens suddenly and gets worse over 3 days to 3 months. The hearing loss may affect only one ear at first. It often fluctuates and gets worse, and spreads to the other ear over time. Some people also have ringing in the ears (tinnitus) or a feeling of fullness in the ears.

About half of patients also have vestibular problems, which may include:

- a feeling of spinning or movement (vertigo); in some cases, this only happens when the head is in certain positions (positional vertigo), or it may come and go (episodic vertigo)
- unsteadiness or imbalance
- motion intolerance
- problems with muscle control and coordination (ataxia)

How is it diagnosed?

AIED may be diagnosed by a primary care doctor but it is more often diagnosed by a specialist such as an otolaryngologist, an otologist, a neurologist or a rheumatologist.

Your doctor will ask about your symptoms. Try to be as specific as possible about your symptoms and when they get better or worse.

Your doctor will also ask about your medical history, including any medications you are taking or recently stopped taking and any conditions you have been diagnosed with in the past. Your doctor will also do a thorough physical and neurological exam.

There is no specific test for AIED, which means it is not always easy to diagnose. But some tests look for evidence of an immune system problem, such as high levels of certain antibodies or white blood cells. These tests can help to diagnose an autoimmune disorder or rule one out.

You will probably have some of the following diagnostic tests:

- hearing tests
- vestibular function tests
- blood tests
- imaging (MRI scans)

Some of these tests are done to rule out other health problems. Several other conditions can cause the same symptoms as AIED, so your doctor needs to consider all the possibilities before making a diagnosis. The doctor will probably look for a pattern of test results and your case history (how your condition has developed and changed over time).

Often, the doctor will prescribe immune-suppressing medication such as steroids. If your symptoms get better with treatment, this can help to make the diagnosis of AIED. But these medications do not always help.

Proposed clinical criteria for AIED

Researchers are still discussing the definition of AIED and the best way to diagnose it. One group has suggested the following major and minor criteria for AIED. The group suggests that a doctor can diagnose AIED if the patient has at least three of the major criteria OR two major criteria and at least two minor criteria. However, these criteria are not used by all doctors.

Major proposed criteria for AIED:

- bilateral (both sides) hearing loss
- systemic autoimmune disease
- a high level of a type of autoantibodies called antinuclear antibodies (ANA)
- lower-than-normal levels of certain white blood cells called T-naïve lymphocytes
- greater than 80% hearing recovery with treatment

Minor proposed criteria for AIED:

- unilateral (one-sided) hearing loss
- young or middle-aged patient
- female patient
- less than 80% hearing recovery with treatment

How is it treated and managed?

Treating AIED is complicated. You may need to see several different specialists, including an otolaryngologist, an audiologist, and a rheumatologist. Treatment usually involves medication, together with hearing tests to see if the treatment is helping. If medication does not help and your hearing loss becomes severe, your audiologist may recommend a hearing aid and later on a cochlear implant.

Corticosteroids

The doctor will likely start by prescribing a high dose of a steroid such as prednisone (PRED-ni-son) for at least four weeks. Steroids suppress the immune system and reduce inflammation. They also

help to control the amount of fluid in the inner ear. This treatment may help to reverse hearing loss in some people with AIED, but not all.

If the steroid treatment helps, the doctor will probably suggest taking it for a longer time before slowly lowering the dose (tapering) over days or weeks. People with a systemic autoimmune disease will probably need to take steroids for at least a year. But steroids can have side effects if they are taken at high doses for a long time, so your doctor will need to find a balance between treating AIED and managing the side effects.

Steroids may help about 7 in 10 people with AIED at first. Over time, though, steroids help fewer people. Some studies have found that only about 14% of people benefit from steroid treatment in the long run.

Immune-suppressing drugs

If steroids do not work, your doctor may suggest a drug that suppresses the immune system, such as cyclophosphamide (sy-ek-loe-FOSS-fa-mide). Cyclophosphamide can help treat AIED, but it may cause serious side effects, including infections, bladder cancer or infertility.

There are many other immune-suppressing or immune-modulating drugs that are used to treat autoimmune disorders. Some of these are being studied for AIED. But there is not enough scientific evidence yet to say if they work.

Hearing tests

During treatment, you will need to have frequent hearing tests (audiometry) to see if your hearing is getting better or worse. You will need to repeat these tests often until your hearing stabilizes.

Managing dizziness and balance symptoms

If you have any dizziness and/or balance problems, you may have vestibular function tests from time to time for monitoring. Your care team may also recommend manoeuvres for positional vertigo or vestibular rehabilitation if you need them.

What to expect in the future

Some researchers are working on new treatments for AIED, including gene therapy and giving anti-inflammatory drugs such as steroids directly into the inner ear. But it will probably be several years before these treatments are available, and they will need to be tested carefully to make sure they work and are safe.

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