

Understanding Autoimmune Inner Ear Disease (AIED)

A patient guide — understanding your condition and your treatment plan.

Your clinician has told you that you have, or may have, autoimmune inner ear disease (AIED). This leaflet explains what it is, how it is diagnosed and treated, and what you can do to protect your hearing and balance.

What is autoimmune inner ear disease?

AIED — when the body's immune system affects the inner ear.

Your immune system normally protects you by fighting infection. In autoimmune inner ear disease it mistakenly affects the inner ear — the organ that controls both hearing and balance. This causes hearing loss, and sometimes dizziness, that usually affects both ears and tends to come on gradually over weeks to months. It is nobody's fault, and unlike many causes of hearing loss, this one can often be treated — especially when it is found early.

Key idea: AIED is an uncommon but treatable cause of progressive hearing loss. The earlier it is recognised and treated, the better the chance of saving your hearing.

What are the symptoms?

The main symptom is hearing loss in both ears that gets worse over weeks or months. It may fluctuate — better some days, worse others. Many people also notice:

- Ringing or buzzing in the ears (tinnitus).
- A feeling of fullness or pressure in the ears.
- Dizziness, unsteadiness, or a sense that the world moves when you move your head.

Some people also have a related autoimmune condition elsewhere in the body, so your doctor may ask about eye redness or pain, joint pains, rashes, or other symptoms.

What causes it?

AIED happens when the immune system loses its normal tolerance and targets the delicate structures of the inner ear. In about one in three people it is linked to a wider autoimmune condition (such as lupus, a form of vasculitis, or Cogan's syndrome); in others it occurs on its own. A viral illness, injury, or other trigger sometimes seems to set it off.

How is it diagnosed?

There is no single test for AIED. Your clinician will build the picture from several steps: hearing tests (repeated over time to track changes), balance tests, blood tests to look for autoimmune markers, and usually an MRI scan to rule out other causes. A key step is a trial of steroid tablets — if your hearing improves or stabilises, this strongly supports the diagnosis.

How is it treated?

Treatment is built up step by step, and the aim is to calm the immune system quickly to protect your hearing.

- Step 1 — Steroid tablets: a course of corticosteroids, started as early as possible. It is important to take the full course and not stop suddenly.
- Step 2 — Steroid-sparing medicines: if the disease returns when steroids are reduced, other immune-calming medicines can be added.
- Step 3 — Newer (biologic) treatments: for disease that does not respond, targeted medicines may be used.
- Step 4 — Hearing and balance support: hearing aids, and for severe loss cochlear implants (which work very well), plus balance (vestibular) rehabilitation if needed.

Living with AIED

AIED is a long-term condition, but it is treatable and not life-threatening. Early treatment gives the best chance of saving hearing. Keep your follow-up appointments and hearing tests, take your medicines as prescribed, and never stop steroids suddenly. Support for hearing and balance makes a real difference to everyday life, and anxiety about hearing changes is common and treatable — please tell us.

Please contact us if: your hearing drops suddenly, your dizziness worsens, or you develop new symptoms such as eye redness or

pain, joint pains, or a rash — these can signal active disease that needs early treatment.