

Autoimmune Inner Ear Disease

WHAT IS AIED?

Autoimmune inner ear disease (AIED) is an inflammatory condition of the inner ear. It occurs when the body's immune system attacks cells in the inner ear that are mistaken for a virus or bacteria. AIED is a rare disease occurring in less than one percent of the 28 million Americans with a hearing loss.

HOW DOES THE HEALTHY EAR WORK?

The ear has three main parts: the outer, middle and inner ear. The outer ear (the part you can see) opens into the ear canal. The eardrum separates the ear canal from the middle ear. Small bones in the middle ear help transfer sound to the inner ear. The inner ear contains the auditory (hearing) nerve, which leads to the brain.

Any source of sound sends vibrations or sound waves into the air. These funnel through the ear opening, down the ear canal, and strike your eardrum, causing it to vibrate. The vibrations are passed to the small bones of the middle ear, which transmit them to the hearing nerve in the inner ear. Here, the vibrations become nerve impulses and go directly to the brain, which interprets the impulses as sound (music, voice, a car horn, etc.)

SYMPTOMS OF AIED

The symptoms of AIED are sudden hearing loss in one ear progressing rapidly to the second ear. The hearing loss can progress over weeks or months. Patients may feel fullness in the ear and experience vertigo. In addition, a ringing, hissing, or roaring sound in the ear may be experienced. Diagnosis of AIED is difficult and is often mistaken for otitis media until the patient develops a loss in the second ear. One diagnostic test that is promising is the Western blot immunoassay.

TREATMENT FOR AIED

Most patients with AIED respond to the initial treatment of steroids, prednisone, and methotrexate, a chemotherapy agent. Some patients may benefit from the use of hearing aids. If patients are unresponsive to drug therapy and hearing loss persists, a cochlear implant may be considered.

HISTORY OF AIED

Until recently it was thought that the inner ear could not be attacked by the immune system. Studies have shown that the perisacular tissue surrounding the endolymphatic sac contains the necessary components for an immunological reaction. The inner ear is also capable of producing

an autoimmune response to sensitized cells that can enter the cochlea through the circulatory system.

AIED RESEARCH

A multi-institutional clinical study, Otolaryngology Clinical Trial Cooperative Group (OCTCG) co-sponsored by the NIH and the American Academy of Otolaryngology-Head and Neck Surgery Foundation, is being conducted to measure the benefits and risks of treating AIED with two different immunosuppressive drugs: prednisone and methotrexate, a chemotherapy drug.