



# Rapid **Attack**

## Solving the Puzzle of Autoimmune Inner Ear Disease

BY KAREN WADA

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**T**he patient, a businessman in his 50s, had always had normal hearing. But a couple of months ago, both ears started going bad and he began experiencing dizzy spells and tinnitus with episodes of severe imbalance. He had come to the House Clinic for answers.

Was it a tumor? A mystery infection? Too much loud music? “No,” Dr. M. Jennifer Derebery, told him. He was suffering from a rare disorder called autoimmune inner ear disease (AIED).

AIED is a complex of symptoms that can cause rapidly progressing sensorineural hearing loss as well as tinnitus, vertigo and dizziness. It ultimately affects both ears, although it can start on one side. Problems tend to worsen over weeks or months as opposed to the sudden hearing loss associated with a virus or the years of progressive loss that come with aging.

“The key thing to watch for is the hearing level rapidly changing,” says Dr. Derebery, an associate of the House Ear Clinic and Institute and a clinical professor of otolaryngology at USC’s Keck School of Medicine. While all House doctors treat AIED, Dr. Derebery – who also serves as director of the

clinic’s allergy department – sees the largest number of cases.

Diagnosis can be tricky. “You first have to rule out other causes,” she says. This requires obtaining or reviewing a patient’s history (generally, the only things that produce similar symptoms are ototoxic drugs). Doctors also order tests such as an MRI or blood work looking for evidence of markers of other known autoimmune diseases, or a test for a specific protein that is positive in about 60 percent of patients called the Western Blot (heat shock protein 70).

“About 80 percent of the time we don’t find any other known autoimmune disorder in these patients,” Dr. Derebery says, “because blood testing for autoimmune diseases can wax and wane, even for diseases we know about.”

AIED is believed to be related to inappropriate autoimmune responses that can damage inner ear tissues. A few theories about how this can occur have been advanced, based on knowledge of other autoimmune diseases. For instance, a virus that contains DNA similar to a patient’s DNA may settle in the inner ear and fool the body into attacking itself. Damage also may be caused by what Dr. Derebery calls “a primary immune attack,” involving a



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reaction against inner ear tissue caused by triggers we have not yet identified.

“And there is a group that have environmental or food allergies, which are technically autoimmune,” she adds. “The inner ear is the target organ in the same way that what an allergic person eats or inhales can target their skin or nose.”

Some see ties to Ménière’s disease, a syndrome that can cause vertigo and fluctuating sensorineural hearing loss as well as tinnitus and a feeling of fullness in the ear. While the effects of Ménière’s usually are one-sided, about a quarter of patients are affected bilaterally. Forty percent of this group have allergies. “There are patients who are more difficult to treat and who exhibit an overly active immune response,” says Dr. Derebery. “That’s why we suspect a connection.”

Autoimmune inner ear disease, which was first recognized in 1979, affects less than one percent of patients with hearing loss. In the past five years, House, which probably sees a disproportionate number of cases, has made a primary diagnosis of AIED 117 times.

There is no known cure. The standard treatment involves prescribing the steroid prednisone, which can stabilize or improve symptoms. “The initial thought was that if you didn’t treat aggressively, patients would go deaf,” says Dr. Derebery. “However, the thinking has

changed. The last time we analyzed our patients only six percent went deaf and required cochlear implants. Most sustained some hearing loss, which we could stabilize. Ninety-four percent could hear well with hearing aids or on their own.”

Prednisone has serious side effects so, says Dr. Derebery, “the goal of therapy is to use just enough to induce serviceable hearing and long periods of remission.” Steroids are taken orally or can be injected through the eardrum, which reduces systemic impact and places a higher concentration of the drug in the inner ear.

House is working with rheumatologists to find alternatives to steroids. So far, no clear winner has emerged. Dr. Derebery was co-principal investigator in a 2003 National Institutes of Health study of methotrexate, which is used to treat cancer and autoimmune diseases. “We found it doesn’t work in the inner ear,” she says.

Recently, she has been prescribing drugs such as Enbrel, Imuran and Plaquenil. “We also have a significant percentage of patients who are undergoing allergy treatments.” She notes that some patients prefer to see if the disease will burn out on its own – without the use of powerful medications.

The businessman in his 50s responded well to four weeks of a high-dose (60 milligrams-a-day) prednisone regimen. As soon as it ended, however,

his hearing worsened. The high-dose regimen was resumed and, after five months of tapering, he was able to go steroid-free. “He’s been stable for most of the past eight years and does well with hearing aids,” says Dr. Derebery.

A trickier case involves a graduate student who came to see Dr. Derebery four years ago because of rapid bilateral hearing loss. High doses of prednisone improved her condition, but she developed steroid-related weight gain and high blood sugar. Attempts to wean her off the drug failed. Now she is taking Plaquenil, an anti-malaria agent that is used to treat autoimmune diseases. She is trying to reduce her prednisone intake to very low levels that do not give her significant side effects and hopes to discontinue usage entirely.

The search for better ways to treat AIED continues. In April, House hosted a small conference of experts in hopes of designing a clinical trial to look for alternatives to steroid therapy.

“I tell patients that contrary to what they see on the Internet and in older ENT literature only a small percentage will lose their hearing,” says Dr. Derebery. “There are things that we can do.” ♦

