

Rapid hearing loss may be a symptom of rare Creutzfeldt-Jakob Disease

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Rapid hearing loss in both ears may be a symptom of the rare but always-fatal Creutzfeldt-Jakob Disease and should be considered a reason for clinicians to test for the disorder.

That was the conclusion of Henry Ford Hospital researchers after encountering a 67-year-old patient who had been progressively losing hearing in both ears for two months and was eventually diagnosed with the disease.

Creutzfeldt-Jakob Disease, or CJD, is often confused with so-called "[mad cow disease](#)," and though they are in the same family of disorders, are not the same.

However, both are always fatal and share such symptoms as impaired thinking, jerky body movements, [memory loss](#) and dementia. Once infected with CJD, the brain develops holes, resulting in tissue which resembles a sponge.

The report will be presented March 19 during the annual scientific meeting of the American Academy of Neurology in San Diego.

According to Ahmad Riad Ramadan, M.D., a Henry Ford [neurologist](#) and lead author, when the patient sought treatment he had no significant [medical history](#) and was complaining only of a continuing, rapid loss of hearing in both ears, and tinnitus – a "ringing in the ears" – that is a common side effect of [hearing loss](#).

"This was followed by the kind of [cognitive decline](#) that is typical of CJD," Ramadan said. "During the patient's hospital stay, he also showed signs of ataxia – a lack of coordination – and myoclonus – a spastic muscle twitch."

Testing found the presence of a telltale protein, and other conditions, that led to a diagnosis of CJD. Researchers noted that the patient's hearing never improved and he died a month after seeking treatment.

Ramadan said the researchers' findings were only the fourth time, based on available literature, that hearing loss such as that found in their patient was recognized as the first symptom of CJD.

This "sensorineural hearing loss," also called "nerve deafness," is the most common cause of permanent impairment; it is hearing loss which results from involvement of the inner ear, auditory nerve, or central auditory pathways in the brain/brainstem.

As the first, or presenting, symptom of their patient, the researchers concluded that testing for CJD in those with fast-progressing hearing impairment should be considered by treating physicians.

Provided by Henry Ford Health System

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