



CC neuroradiologist and lead study author Dr. John Butman.

CC imaging study focuses on von Hippel-Lindau patients' hearing

The results of an imaging study conducted at the Clinical Center could help doctors protect and preserve the hearing of people with the rare genetic disorder von Hippel-Lindau disease (VHL).

VHL causes the abnormal growth of benign and cancerous tumors in numerous parts of the body. This imaging study focused on endolymphatic sac tumors (ELSTs). The connection between ELSTs and hearing loss has been a curiosity because the endolymphatic sac is on the temporal bone that contains the ear but is not part of the inner ear associated with hearing. Some cases of hearing loss in VHL can be attributed to large tumors on the sac that invade the hearing apparatus. But CC neuroradiologist and lead study author Dr. John Butman noticed, and set out to explain, why some patients lost hearing when they had only a very small tumor outside the sensory part of the ear.

He says he could see blood flowing from the endolymphatic sac into the inner ear in about half of the small tumors he imaged. Researchers then talked to patients to correlate the images with their symptoms. "It turned out virtually all patients who had evidence of bleeding had a sudden hearing loss," Butman explains. "For example, they were talking on the phone and suddenly their hearing went out, which is characteristic of a hemorrhage."

Other patients with small tumors experienced gradual hearing loss, but no bleeding. Butman says in those cases the ELST was disrupting the flow of fluids in the inner ear and causing a condition known as endolymphatic hydrops, which creates pressure, ringing in the ears, vertigo, and gradual hearing loss.

NINDS neurosurgeon Dr. Russell Lonser, senior author on the study, says the findings prompted a change in thinking about how to manage these tumors. Previous practice was a wait-and-see approach. Given that the tumors were benign, slow-growing, and their natural history was unknown, doctors monitored their growth and did not remove them until they became big enough to threaten hearing.

"It was believed these tumors probably would be best observed," Lonser says, "Now we believe they probably should be operated on very early on, as soon as they are identified on imaging." Butman used CT imaging to study the bone and MR technology for the soft tissue and inner ear. He studied the inner ear with an imaging technique that is typically used for the brain to detect some of the cases of hemorrhage.

"Part of radiology is solving puzzles," Butman says. "Being able to figure out most of the story from imaging findings alone is very satisfying. Then having the clinical findings correlate so well with the imaging gives a sense of the solution to the puzzle. The fact that this has affected the thinking on how to manage patients is very nice."