

Insight gained into age-related hearing loss

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Researchers at the University of Minnesota Medical School have gained insight into how different types of age-related hearing loss may occur in humans. The discovery could eventually help physicians develop drugs to combat progressive hearing loss. Their paper is published on October 14 in the open-access journal *PLoS Genetics*.

James Ervasti, Ph.D., and colleague Ben Perrin, Ph.D., studied how two very closely related genes contribute to hearing function in mice. Mutations in the same genes are associated with deafness in humans. The duo discovered two key cellular processes that are required to maintain auditory function.

The genes encode proteins called β -actin and γ -actin. In humans, deafness-causing mutations have been linked to both proteins. β - and γ -actin comprise the primary structural elements of stereocilia (hair-like fibers in the ear), which convert mechanical sound energy into the nerve signals that allow humans to hear.

The two proteins are 99 percent identical; however, their slight differences have been exactly conserved through evolution from birds to mammals, suggesting that each protein may have important and distinct functions. Ervasti and Perrin tested the idea that two closely linked proteins have separate, but important, roles in hearing by knocking out each gene in mouse auditory hair cells.

They found that β -actin and γ -actin do have different maintenance functions that together keep the hair-like fibers -- that allow mice to hear



-- healthy. Both knockout mice had normal hearing at young ages, but developed specific types of progressive <u>hearing loss</u> and stereocilia pathology that differed depending on which protein was lost.

"These separate maintenance pathways are likely important for maintaining auditory function during aging and may contribute to future understanding of common forms of age-related hearing loss in humans," Perrin said.

More information: Perrin BJ, Sonnemann KJ, Ervasti JM (2010) b-Actin and c-Actin Are Each Dispensable for Auditory Hair Cell Development But Required for Stereocilia Maintenance. PLoS Genet 6(10): e1001158. doi:10.1371/journal.pgen.1001158

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