

Study finds new role for protein in hearing

August 15 2011

University of Iowa scientists have discovered a new role for a protein that is mutated in Usher syndrome, one of the most common forms of deaf-blindness in humans. The findings, which were published Aug. 8 in *Nature Neuroscience*, may help explain why this mutation causes the most severe form of the condition.

The study suggests that the protein called harmonin, which is known to be involved in sound sensing in the inner ear, may also play a role in the transmission of sound information to the brain.

Hearing starts with the transmission of sound by inner hair cells in the ear. Sound waves cause movement of special structures called stereocilia on the tips of the hair cells. Harmonin is thought to mediate this movement, which then activates the cells and initiates transmission of sound information as electrical and [chemical signals](#) to the brain.

"Most of the research until now has concentrated on the input end of the inner hair cells where the [sound waves](#) produce motion of the stereocilia," said Amy Lee, Ph.D., senior study author and UI associate professor in the Departments of [Molecular Physiology](#) and Biophysics, Otolaryngology-Head and Neck Surgery, and Neurology. "Now we have found a new role for harmonin at the opposite end of these sound-sensing inner hair cells where it appears to control the signal output of the cell."

Lee and colleagues, including UI postdoctoral fellows Frederick Gregory, Ph.D., and Keith Bryan, Ph.D., found that harmonin is

important for regulating the number of [calcium channels](#) present at the sound-transmitting synapse of inner [hair cells](#).

Studies from other labs have shown that too few or too many calcium channels at the hair cell synapse cause deafness in mice. This means factors that control how many channels are available are likely to be important for normal hearing.

"Harmonin appears to precisely control how many channels are available," Lee said. "What we think is happening in [Usher syndrome](#) where the harmonin protein is mutated is that there are too many calcium channels available, which causes abnormal signaling at the synapses.

"We are most excited about the idea that this mutation could contribute to the disease process of Usher syndrome in a way that was not imagined before," Lee added. "It may eventually be possible to alter this interaction between harmonin and the calcium channels in a way that might be useful as a therapy for patients with this form of Usher syndrome."

Harmonin is also expressed in the retina -- the light-sensitive tissue in the eye -- which is affected in Usher syndrome, and there are calcium channels in the photoreceptor cells of the retina. It is not known how the harmonin mutation affects the retina and how it might contribute to blindness in Usher syndrome, but that is another area of research Lee's team hopes to investigate.

Provided by University of Iowa Health Care

Citation: Study finds new role for protein in hearing (2011, August 15) retrieved 19 April 2024 from <https://medicalxpress.com/news/2011-08-role-protein.html>

This document is subject to copyright. Apart from any fair dealing for the purpose of private study or research, no part may be reproduced without the written permission. The content is provided for information purposes only.