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Researchers identify protein needed to develop auditory neurons

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Loss of spiral ganglion neurons or hair cells in the inner ear is the leading cause of congenital and acquired hearing impairment. Researchers at the University of California, San Diego, School of Medicine and the National Institutes of Health found that Sox2, a protein that regulates stem cell formation, is involved in spiral ganglion neuron development. The study was published in the January 13 issue of the *Journal of Neuroscience*.

"These findings may provide the first step toward regenerating spiral ganglion neurons, the nerve cells that send sound representations to the brain," said Alain Dabdoub, PhD, co-investigator and assistant professor of surgery with the division of otolaryngology at the UC San Diego School of Medicine. "This has significant implications for advances in cochlear implant technology and biological treatments for hearing loss."

In the cochlea, auditory neurons transmit sound vibrations conveyed by hair cells. These vibrations are then converted to nerve impulses that communicate with the brain. If the neurons are lost or damaged, hearing loss occurs. Existing therapies for hearing loss are based on either increasing hair cell stimulation with hearing aids or introducing an electronic substitute for the hair cells with cochlear implants. In either case, the presence of functional spiral ganglion neurons is required for a successful outcome.

Prior research shows that as few as 10 percent of the normal number of spiral ganglion neurons is sufficient for the success of cochlear implants.

"The identification of factors that induce functional neurons has important implications for hearing restoration," said Chandrakala Puligilla, PhD, a research fellow at the National Institutes of Health. "The ability to induce even a small number of cells with gene-based therapy could be enormously beneficial."

Sox2 is part of the SoxB1 family of proteins, which plays a significant role in neural development in the spinal cord and elsewhere. The study demonstrates a novel role for Sox2 in ear development showing that Sox2 is critical for the production of auditory neurons and that generating new neurons is possible. Understanding the molecular signaling pathways responsible for the development of spiral ganglion neurons could be applicable to other neurons, especially those in the central nervous system.

Provided by University of California - San Diego