FACT SHEET

Hair Cell Regeneration and Hearing Loss

NATIONAL INSTITUTES OF HEALTH

Our ability to hear relies on hair cells, small sensory cells in the inner ear. Hair cells are named for microscopic hair-like extensions, called stereocilia, projecting from their tops in bundles. These "hair bundles" convert sound vibrations into electrical signals, which travel to the brain by way of the auditory, or hearing, nerve. When hair cells are damaged—by disease, injury, or aging—a person experiences hearing loss, sometimes profound. Although fish, amphibians, and birds are able to grow new hair cells to replace damaged ones, mammals cannot regenerate hair cells on their own. Through gene therapy and stem cell research, however, scientists have been able to grow new hair cells in laboratory animals, in some cases restoring some hearing to deafened mammals. Such promising results have led researchers to wonder if we might be able to regenerate hair cells in people one day.

Yesterday

- More than 90 percent of hearing loss occurs when either hair cells or auditory nerve cells are destroyed. Scientists believed that hair cells in mammals could never be replaced if they were injured or destroyed.
- Early studies suggested that when a hair cell develops, it inhibits its neighbor from becoming a hair cell which, instead, becomes a supporting cell. Scientists wondered if supporting cells had the ability to become hair cells if neighboring hair cells were injured or destroyed.
- To test this theory, NIH-supported scientists destroyed hair cells in a mouse inner ear. Time lapse photography showed neighboring supporting cells migrating to the hair cell region and growing hair bundles on their surfaces. Although the process did not result in restored hearing in the mice, researchers began to investigate whether experimental augmentation of the process might improve hearing loss.

Today

- Hearing loss affects individuals from birth to adulthood, and there is a compelling need for research to address the problem. For example, according to 2007 data from the Centers for Disease Control and Prevention (CDC), approximately 36 million American adults report some degree of hearing loss. But hearing loss also affects young people. The CDC estimates that two to three out of 1,000 babies born in the United States each year have a detectable hearing loss, which can affect their speech, language, social, and cognitive development.
- Scientists have made a number of critical discoveries about hair cell growth and development that are contributing to new ideas about the possibility of regenerating hair cells in people one day.
- Knowing that supporting cells have the ability to become hair cells under certain conditions, scientists focused on identifying specific molecules that might be involved in a hair cell developing from a supporting cell.
- Studying experimental animals, scientists have identified genes that are necessary for hair cell formation and function. In some cases, they found that these genes were similar even though they were from different animals. For example, a gene in mice, called Atoh1, is necessary to make hair cells and a similar gene is required for hearing in fruit flies.
- NIH-supported scientists treated deafened guinea pig ears with a harmless virus carrying the gene Atoh1. The Atoh1 gene caused supporting cells to become hair cells in the deafened guinea pigs and, importantly, the treated animals were able to regain some of their hearing. This was the first demonstration of gene therapy that improved hearing in formerly deaf animals.
- NIH-supported scientists have identified a gene in mice, called Rb1, that shuts down the growth of new hair cells early in development. Mice bred to be missing the gene were able to grow more hair cells than mice possessing the gene. In addition, mature

hair cells growing in culture dishes were able to regenerate when the Rb1 gene was deleted.

- NIH-supported scientists have demonstrated that mouse embryonic stem cells can develop into functional, immature hair cells in the laboratory. The ability to re-grow hair cells will not restore hearing without properly reconnected nerve endings. NIH-supported scientists found that newly formed hair cells and nerve cells successfully reestablish connections in an organized way, although the reconnected nerve endings are simpler than those generated during normal development. This and other research will help reveal how nerve cells form connections with newly generated hair cells.

Tomorrow

The NIH is poised to make major discoveries in the study of hair cells and hearing loss. NIH-supported scientists are:

- Conducting studies to coax immature hair cells generated from stem cells to develop into a more mature state by mimicking the environment that developing hair cells would normally experience.
- Exploring how to encourage newly-generated hair cells to send appropriate signals to the hearing centers in the brain.
- Using their knowledge of hearing genetics to generate new hair cells.
- Gaining a better understanding of the cellular and molecular mechanisms the body uses to protect auditory hair cells against damage, with the hope of developing methods to enhance survival of hair cells following trauma or disease.

Contact: NIDCD Office of Health Communication and Public Liaison, nidcdinfo@nidcd.nih.gov ; 301-496-7243.

Visit the National Institute on Deafness and Other Communication Disorders (NIDCD) website at: http://www.nidcd.nih.gov/