

Hearing What the CELLS AND Tell U

HEI'S Cell and Molecular Scientists Expl

Beginning in the early 1960s, William F. House, M.D., brother of House Ear Institute (HEI) Founder Howard P. House, M.D., developed the first single-channel cochlear implant at the Institute. Today there are more than 50 thousand people worldwide who have had their hearing improved with cochlear implants. This is just one of many scientific advances made through cutting-edge otologic research at HEI.

The development of the cochlear implant at HEI exponentially expanded the international reputation of the Institute. But as remarkable as it is, the implant is an assistive device and not a cure. The search for a cure lies in our ability to understand and ultimately correct damage to hearing and balance organs at the cellular level.

Established in 1996, the Gonda (Goldschmied) Department of Cell and Molecular Biology is one of the fastest growing research areas at HEI. Scientists are working within three main labs – Developmental Biology, Cell Biology and Molecular Otolaryngology. Their studies focus on the genes involved in the formation of

the ear including how cells differentiate to form the intricate components of the inner ear and the entire hearing system. Exploring the mechanisms that affect the development and function of the auditory system will help our scientists understand the molecular machinery involved in the creation and maintenance of the cells inside the hearing and balance organs, which may eventually lead to the development of a cure.

John W. House, M.D., President of the Institute, believes that it will be possible to restore the damaged or missing sensory cells associated with sensorineural hearing loss through the regeneration of hair cells within the inner ear. "Through our cell and molecular biology research, we are exploring ways to duplicate or re-grow the sensory cells in the cochlea," says Dr. House. "When we reach that goal, there won't be a need for some of the treatments currently being used to restore hearing, such as cochlear implant surgery. I could never have predicted some of the things I've witnessed during my 25 years in otology practice, and the next 25 years at the Institute promise to be just as remarkable."

A high-magnification electron micrograph of the Organ of Corti, showing the intricate cellular structure of the cochlea. The image features a prominent, dark, layered structure with a granular texture, likely representing the stereocilia and supporting cells. The background is a lighter, more uniform color, possibly representing the surrounding tissue or fluid.

MOLECULES

Their Research

Organ of Corti


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T racking the Pathways of Middle Ear Infections

Second only to the common cold, recurring middle ear infection, (otitis media), is the most prevalent infectious disease affecting young children. Antibiotics are currently the common treatment for otitis media. The problem is that strains of bacteria are becoming resistant to these drugs. The research of David Lim, M.D., chief scientist of Molecular Otolaryngology and executive vice president of research at HEI, focuses on how this troublesome infection begins and is transmitted at the cell and molecular level in the middle ear. His laboratory is also studying the way the body's innate immune system and the secreted molecules in the ear may function as natural antibiotics. Dr. Lim's research team hopes to develop new strategies in the form of a vaccine or medication that can boost the inherent power of the body in fighting this disease.



F ollowing the Signals of Middle Ear Infections

The laboratory of Jian-Dong Li, M.D., Ph.D. chief scientist of the Section on Signal Transduction, is pursuing a different approach to the study of otitis media with mucoid effusion (sticky mucus). This common childhood ailment leads to conductive hearing loss, which can result in poor school performance.

The current goal of this lab is to reduce middle ear infections in children by investigating the overproduction of mucus in the middle ear. In a bacterial infection, cell receptors set off signaling mechanisms to help the body fight off harmful bacteria and enhance its protection. This produces an inflammatory response, (overproduction of mucus).

Through studying this response mechanism, Dr. Jian-Dong Li and his research team hope to develop new therapies specifically targeted to the bacterial components that are responsible for the overproduction of mucus. Once scientists identify the different bacterial proteins and their pathological responses that play a role in otitis media, the information gained may be used to develop a drug to reduce the inflammatory response and the number of days associated with the illness.

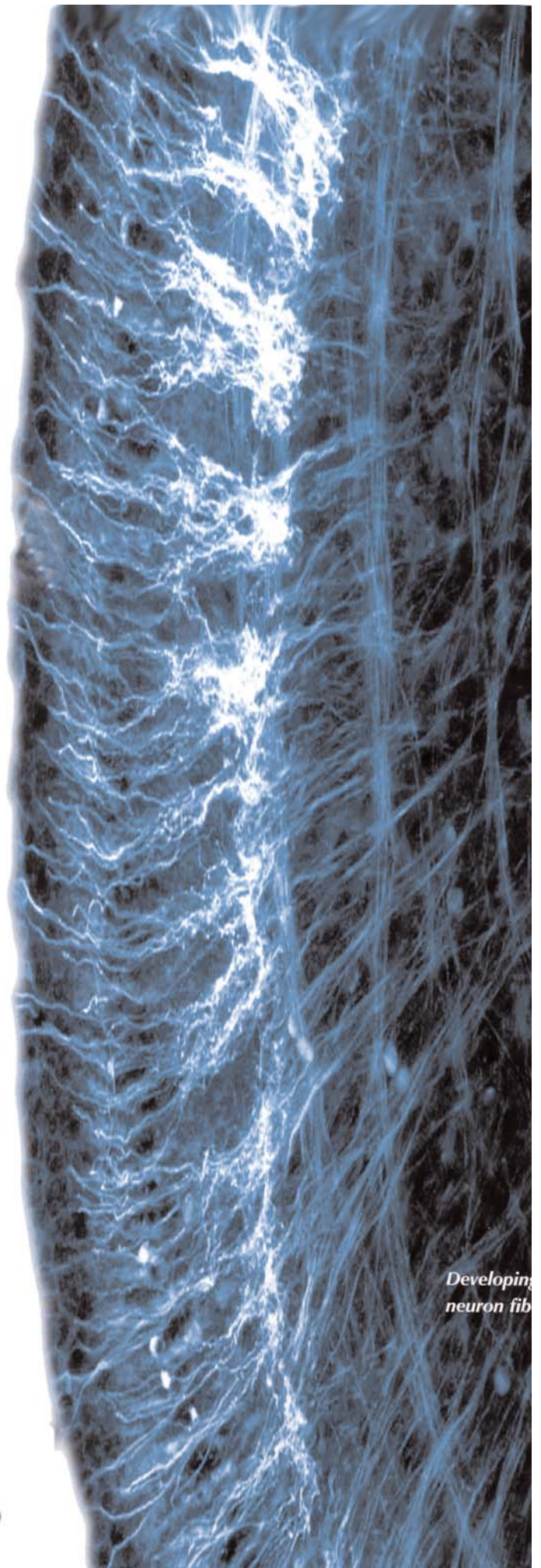
H. Influenza binding on the surface of an epithelial cell

Xi Lin, Ph.D., chief scientist of HEI's Neurobiology Section, examines how cellular mechanisms that ensure the information our inner ear's sensory cells receive are converted into electrical impulses allowing our brain to hear and recognize words and sounds. Dr. Lin's lab is looking at the factors that affect the successful conversion of this information by these sensory cells and neurons.

Cellular Messages from the Ear

Another project of the Neurobiology Section investigates specialized fluids, known as the perilymph and endolymph, that fill the complicated systems of our balance canals in the inner ear. Dr. Xi Lin is collaborating with Dr. David Lim and his research team to explore how inner ear fluids maintain equilibrium and what happens when the molecular mechanisms that regulate these fluids are disrupted. Disorders occur when the chemical composition of each fluid changes in relation to one other. For example, the dizziness that is a symptom of Meniere's disease is a result of an overproduction of endolymph. In another example, findings show that nearly half of hereditary deaf patients in the U.S. and Europe have a common genetic mutation that effects the regulation of potassium levels, leading to the destruction of sensory cells and deafness. Understanding these complex inner ear regulation mechanisms is a first step toward developing strategies in the prevention and cure of these disorders.

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