Help for deaf humans may come on little cat feet

By LEE BOWMAN
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A new study in cats demonstrates how cochlear implants in very young animals allow them to develop normal nerve fibers to transmit sound and restore hearing by reversing damage to the brain's hearing network.

The findings by researchers at the Johns Hopkins University School of Medicine in Baltimore help explain why such implants are about 80 percent successful in restoring hearing to young children who are born deaf, but rarely work in adults who are congenitally deaf.

"What we think this study tells parents of deaf children is that if cochlear implants are being considered, the earlier they're done, the better," said David Ryugo, a professor of otolaryngology and neuroscience at Hopkins' Center for Hearing and Balance and lead investigator for the study, published Friday in the journal Science.

"There is an optimal time window for implants if they are to avoid permanent rewiring of hearing stations in the brain and the long-term effects on language learning that can result."

Cochlear implants are small devices designed to mimic the work of inner-ear structures that react to sound waves and convert tones to electrical nerve signals. Hair cells form a ribbon of vibration sensors along the cochlea, the organ of the inner ear that senses sound. After receiving sonic vibrations through the eardrum and bones of the middle ear, the hair cells convert them to the signals carried to the brain via the auditory nerve.

Unlike hearing aids, which amplify sound through a nerve-to-brain system that may be damaged but still intact, cochlear implants bypass the ear entirely and link right to the nerve cells that interpret sound in the brain.

One part of the device is a microphone located behind the ear that picks up sound and converts it to electrical signals that are transmitted through the scalp to a receiver attached directly to the brain.

More than 10,000 children are born deaf each year in the United States, and an estimated 1.5 million people are believed to be good candidates for cochlear implants. But only a few thousand of the procedures are done each year, with about 10 percent going to children under the age of 3.

For the study, Ryugo and colleagues compared the responses and brain tissue of cats born deaf that got cochlear implants within four months of birth with deaf cats that did not get the implants.

Both groups were exposed to three months of sound stimulation, during which the researchers played music and let the animals roam the lab, with various everyday background noises. Along with three deaf cats with implants and four deaf cats with no implants, they also included three otherwise similar cats with normal hearing as a control group.

To measure the animals' hearing development, all the deaf cats, with or without implants, were subjected to a unique sound, one for each animal, that measured its response to a cue, such as the clapping of hands or ringing of a bell, signifying that there was a food reward nearby.

Within a week, the deaf kittens with implants responded eagerly to the new sound cue, rushing to collect the food, while those without implants did not.

Later, brain-tissue analysis showed that those felines with implants developed regions called synaptic connections between connecting auditory nerve cells that closely resemble those seen in cats with normal hearing. Those connections included large numbers of synaptic vesicles, reservoirs along nerve pathways that store chemicals needed to pass sound signals between nerve cells, and robust membranes that receive the signals.

The deaf cats without implants lacked the reservoirs and the membrane regions were flat and withered in appearance.
Dr. John Niparko, another ear specialist at Hopkins who took part in the study and who has been studying the effects of hearing restoration on children for more than 20 years, said researchers now need to determine what happens between birth and puberty to the auditory system that diminishes the changes of restoring hearing and language skills to the brain, with the aim of finding therapies that could repair or reverse those changes in deaf people.

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(Contact Lee Bowman at BowmanL(at)SHNS.com. Distributed by Scripps Howard News Service, http://www.shns.com)