Mice shed new light on causes of childhood deafness

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Deafness is the most common disorder of the senses. Tragically, it commonly strikes in early childhood, severely damaging an affected child's ability to learn speech and language. In many cases, children gradually lose their hearing to become profoundly deaf over a long period of months to years, but scientists know very little about how this progressive loss happens, making prospects for prevention and cure very slim. Over half the cases of childhood deafness are estimated to be due to defects in just one gene passed from either the mother or father, and many of these deafness genes have been identified. However, as the way we hear is so complicated, it has been really difficult to work out exactly how these genes cause such wholesale effects.

Dr John Oghalai, of Baylor College of Medicine in Texas, has been wrestling with this problem for his whole career. His work as a clinician, directing a busy team performing cochlear implants and corrective surgery on the ear and cranium, has armed him with crucial clinical insights which inform his laboratory's research into the causes and treatment of deafness. Together with a team spearheaded by postdoctoral fellow Anping Xia, he has now created mice which carry a mutation in one of the genes, called alpha tectorin, known to cause progressive childhood deafness.

Remarkably, the mice develop the same hearing problems as children. Oghalai's team are now able to look in detail at exactly what is going wrong in the development of the inner ears of their mice. New insights have already been gained into how and why the mutation affects

processing of soundwaves entering the ear, and, in an ironic twist to the story, there are clues that the inner ear's attempts to compensate for the hearing loss may cause ongoing damage leading eventually to profound deafness.

One day, Oghalai hopes that the mice will help in developing new therapies to slow or even halt hearing loss, not only in children, but also in elderly people, tackling a major cause of isolation and depression.

More information: The study is published in the research journal, Disease Models & Mechanisms (DMM), .

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