

Towards a better understanding of inherited hearing loss

October 15 2013

A team of researchers led by Dr. Michel Cayouette at the IRCM made an important discovery, published online yesterday by the scientific journal *Developmental Cell*, that could better explain some inherited forms of hearing loss in humans. The Montréal scientists identified a group of proteins crucial for shaping the cellular organ responsible for detecting sounds.

For a human to hear, sound-induced vibrations in the inner ear must first be transformed into electrical impulses before they can be relayed to the brain. This transformation is performed by "[hair cells](#)" (or sensory cells) located in the [inner ear](#). On the surface of these cells, microscopic hair-like protrusions known as stereocilia act as specialized sensors to detect vibrations.

"During embryonic development, these stereocilia develop into a characteristic V-shaped brush," says Dr. Cayouette, Director of the Cellular Neurobiology research unit at the IRCM. "In addition, all cells orient their brush with the V pointing in the same direction. This polarized organization is critical for sensory function, but remains poorly understood."

"We studied a group of proteins known to control cell division in the organism and discovered a new role they play in the auditory system," explains Dr. Basile Tarchini, postdoctoral fellow in Dr. Cayouette's laboratory and first author of the study. "We showed that these proteins occupy a specific region at the [cell surface](#) to define the exact placement

of stereocilia and enable the creation of the V-shaped brush."

"Furthermore, we discovered that one of the proteins is also required for coordinating the orientation of the brushes among neighbouring cells, thereby ensuring that the V formed by each brush points in the same direction," adds Dr. Tarchini. "Our results strongly suggest, for the first time, that this group of proteins could be the link between two important molecular mechanisms: the system responsible for the placement of stereocilia into a V-shaped brush at the cell surface, and the system that orients this V-shaped structure in the surrounding tissue."

"Recent studies show that mutations in one of the proteins we studied are associated with inherited forms of [hearing loss](#) in humans," concludes Dr. Cayouette. "By defining a function for this class of proteins in hair [cells](#), our work helps explain the mechanisms that could cause these conditions."

More information: [www.cell.com/developmental-cell ...
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Provided by Institut de recherches cliniques de Montreal

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