Description for the general public.

Human fascination with the world, the drive to understand it and transform, stems from our innate ability to perceive what surrounds us in great detail, using diverse sensory modalities. We use our senses constantly in everyday life, often subconsciously. They inform us about the physical world we interact with, are vital for survival and allow communication with others. Their exquisite capabilities may become apparent when we plan or perform a complicated task, one that requires speed and precision, or when we contemplate a work of art. Sadly, we are also sometimes forced to face the limitations of our perception. Be it a result of an accident, an illness or a genetic condition, when one or more of our senses fail life becomes more challenging.

If our senses are what allows us to perceive the world, than it is only natural to ask questions about the way they work. The domain of sensory biology has a long history and is currently developing rapidly, taking advantage of the unprecedented progress in molecular and genetic research, the availability of ever improving research models and new imaging techniques. Identification of new genes that play a role in the sensory organs and elucidating their exact function has been one of the driving forces behind this development.

Our team is focused on discovering new molecular mechanisms of hearing. The perception of sound is a fascinating subject to study, still guarding mysteries that provoke curiosity. Hearing happens in a highly specialized organ that resides in the inner ear. In it lie the sensory hair cells, a peculiar type of epithelial cells, in many ways similar to the cells that line the inside of the airways or the digestive tract. Yet, hair cells are special. The cells got their name because of hair-like protrusions, called stereocilia, present on their apical surface. Unlike real hair, these protrusions are rigid and can pivot around their base when exposed to the vibrations of sound. Specialized proteins at the stereociliary tips form miniature channels that can open in response to this motion, leading to a sudden change of the electrical potential of the hair cell. The electrical signal can then travel to the neurons that connect with the hair cell and relay the information about the received sound to the appropriate centers in the brain.

Hair cells needed to develop many adaptations in order to function as sound receptors. This is reflected by the presence of cell type specific proteins. We have recently identified one such protein that we have evidence to be important for the development of the hair cell ability to perceive mechanical stimuli. Our preliminary data indicate that this protein is located in the cell body of the hair cell, within the intracellular membranes called the endoplasmic reticulum. This structure is vital for controlling the protein synthesis, the sorting of different proteins to the appropriate parts of the cell and many metabolic processes. This project's aim is to discover the exact biological function of the new hair cell protein and expand our understanding of hearing.

In many cases hair cell specific proteins are encoded by genes that can cause a hearing disorder when mutated. Sometimes deafness is the only manifestation of a mutation, other times it can be accompanied by even more severe symptoms. Roughly 10% of the world population will experience a form of hearing impairment at some stage of their life. Genetic factors play a big role in many of the most severe cases. For this reason, apart from a purely scientific interest in how hearing works, we are motivated by the thought that our research has a potential to improve understanding of deafness, provide new targets for molecular diagnostic and tools and perhaps ultimately inspire a successful therapeutic approach to treating certain forms of hearing loss.