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Duchenne muscular dystrophy (DMD), is one of the most prevalent forms of human muscular dystrophy. It is caused by mutations in dystrophin, an important protein in the that links the cellular cytoskeleton to the extracellular matrix. In addition to its well characterized role in muscle, dystrophin is also expressed in other tissues including, the brain, the retina, and cochlear hair cells. This project focuses on the effects of dystrophin loss in cochlear hair cells and subsequent hearing loss. The loss of dystrophin can be compensated for by utrophin, a homologue to dystrophin, which binds to the same complement of proteins. The ultimate goal of this project was to examine hearing loss in mice deficient in utrophin, dystrophin and both utrophin and dystrophin. Mice were tested at different age time points to try to track when a loss of hearing occurred if applicable. By the completion of this project, it should be clear whether or not utrophin can compensate for dystrophin with respect to hearing and vice verse.

The Effects of Utrophin and Dystrophin in the Inner Ear



Poster Number: Session: