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## Gangliosides and Hearing

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### ABSTRACT

Severe auditory impairment observed in GM3 synthase-deficient mice and humans indicates that glycosphingolipids, especially sialic-acid containing gangliosides, are indispensable for hearing. Gangliosides associate with glycoproteins to form membrane microdomains, the composition of which plays a special role in maintaining the structural and functional integrity of hair cells. These microdomains, also called lipid rafts, connect with intracellular signaling and cytoskeletal systems to link cellular responses to environmental cues. During development, ganglioside species are expressed in distinctive spatial and temporal patterns throughout the cochlea. In both mice and humans, blocking particular steps of ganglioside metabolism produces distinctive neurological and auditory phenotypes. Thus each ganglioside species may have specific, non-overlapping functions within the cochlea, central auditory network, and brain.

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