

Hearing loss caused by transport disorders in the inner ear

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The composition of the endolymphatic fluid that bathes the sensory hair cells of the cochlea in the inner ear is strictly regulated to enable the transduction of sound into neural signals. Abnormalities in volume or composition of this fluid will impair the hearing process.

After basic information on inner ear anatomy and physiology, the topic of hearing loss caused by mutations in transporters for chloride (ClC-K), protons (H⁺-ATPase) and bicarbonate (Cl⁻/HCO₃⁻-exchanger) and their involvement in acid/base homeostasis of fluids in the inner ear as well as kidney will be discussed.

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