Understanding congenital unilateral sensorineural hearing loss: A PhD project

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Background: Congenital unilateral sensorineural hearing loss (uSNHL) accounts for approximately 25% of sensorineural hearing losses identified in newborn hearing screening programs, presenting a significant yet under-researched challenge in pediatric audiology.

Aim: The study aimed to elucidate the etiology and mechanisms of congenital uSNHL and assess the impact of intervention strategies.

Method: In three studies, we examine two main themes: 1) the effects of hearing aid (HA) amplification in children with congenital uSNHL, and 2) the etiology and auditory mechanisms in infants with the condition, including anatomical and genetic analyses.

Result: The HA amplification study showed mixed results, indicating improvements in one-on-one communication but disadvantages in sound localization accuracy. A high incidence of anatomical malformations and a notable rate of genetic diagnoses were observed among the subjects. Additionally, the studies highlighted an absence of loudness recruitment in auditory processing.

Discussion: These findings emphasize the unique aspects of congenital uSNHL compared to bilateral SNHL. The research supports the implementation of routine imaging and selective genetic testing in cases suspected of being syndromic. It also sheds light on the complex effects of late-fitted HAs, indicating potential benefits from earlier fitting and HA adjustments tailored to individual auditory needs. By presenting these three published studies, we aim to communicate a better understanding of congenital uSNHL and to develop more effective intervention strategies.