Pediatric Neurology Part III: Chapter 158. Hearing loss and deafness in the pediatric population: causes, diagnosis, and rehabilitation (Handbook of Clinical Neurology)

Paul Deltenre, Lionel Van Maldergem

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With prevalence figures close to 0.2% at birth and rising to 0.35% during adolescence, hearing loss is the most frequent sensory impairment in childhood. This silent handicap has to be actively sought for without delay as it will seriously interfere with the development of speech, language, cognitive and socio-emotional behavior. Objective physiological techniques (evoked potentials, oto-acoustic emissions, tympanometry) combined according to the cross-check principle allow early diagnosis. Objective testing yields invaluable information about the mechanism of the loss and the contribution of disruption of the neural code to the handicap. Among the acquired causes, cytomegalovirus (CMV) infections plays a major role and may take elusive forms. Aminoglycoside ototoxicity has a genetic determinant. Meningitis can lead to rapid endocochlear ossification prompting for rapid cochlear implantation. Genetic causes account for more than 60% of congenital hearing loss, new genetic causes being discovered at an amazing rate. The high number of genetic entities and their huge heterogeneity among them requires guidelines for requesting genetic testing when desirable. Several syndromes prone to request neuropediatricians' attention as an early diagnosis followed by specific treatment can considerably limit the ensuing handicap. Whatever the type of assistive device fitted (amplifying hearing aid or cochlear implant) and the importance of associated handicaps, a multidisciplinary rehabilitation combined with educated parental commitment is necessary for optimal results.

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