

AURICULAR MALFORMATION AND HEARING LOSS IN OCULOauriculoVERTEBRAL SPECTRUM

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Introduction: Oculoauriculovertebral spectrum (OAVS) or hemifacial microsomia is a congenital craniofacial disorder involving structures derivated from first and second pharyngeal archs. Clinical presentation is highly variable and main craniofacial findings involve microtia, mandibular hypoplasia, epibulbar dermoid and vertebral anomalies. External, middle and inner ear can be affected in this condition and microtia with external auditory canal atresia is the most common ear finding. Conductive and mixed hearing loss are commonly described. **Objective:** To describe and correlate auricular malformation and hearing loss in individuals with OAVS. **Methods:** In a retrospective transversal study model, 25 clinical charts of patients with OAVS were reviewed after Research Ethics Committee approval (1.818.192). Data related to genetic diagnosis, microtia classification, hearing antecedents, tonal and vocal audiometry and tympanometry were collected. **Results:** Hearing loss was found in 83% ears with microtia, a statistically significant association. Conductive hearing loss was found in 66% ears with type 1 microtia. Type 2 microtia caused mixed (80%) and conductive (20%) types. Type 3 microtia caused conductive (52%) and mixed (47%) types. Hearing loss degree was mild in most ears with type 1 microtia (66%) and severe in most ears with type 2 and 3 (80%). CT scan of the temporal bones revealed middle ear malformation in types 2 and 3 microtia. **Conclusion:** Microtia is associated with hearing loss in most OAVS cases. Hearing loss type and degree apparently deteriorated as the microtia type increased. Attention should be given to cases with mild ear malformation, since hearing loss may be also present.