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RARE CAUSE OF CONDUCTIVE HEARING LOSS: ABSENCE OF THE SUPERSTRUCTURE OF THE STAPES

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Introduction: Congenital anomalies of the ossicular chain are not a frequent pathology. The absence of the stapes is a very rare anomaly, with a limited number of cases reported in the literature, it appears as either an isolated event or in association with others malformations. Patients have congenital conductive hearing loss. The diagnosis is often made intraoperatively and the treatment depends of the accompanying anomalies.

Material and methods: A 17-year-old female with a history of anxiety and asthma, presented with hearing loss and tinnitus in the left ear of two years of evolution. The examination showed a normal otoscopy, negative Rinne in both ears and centered Weber. Audiometry confirmed a bilateral transmission hearing loss, more severe on the left side, with a gap of 60 decibels and an overall loss of 73%. Tympanometry curves were normal, but the stapedial reflexes were absent in the left side. The temporal bone CT scan was informed as normal, and control audiometry showed no changes. An exploratory tympanotomy was performed in which an agenesis of the stapes superstructure was observed and a reconstruction was made with a TORP (Total Ossicular Replacement Prosthesis). The patient had an uneventful postoperative period and the follow-up audiometry showed improvement in hearing levels.

Discussion: The anomalies of the stapes are very rare among the malformations of the middle ear, the complete absence of the superstructure has been reported in very few occasions in the literature. The most accepted theory is an anomaly in the position of the facial nerve, with an anterior displacement that does not allow the correct contact between the stapes blastema and the otic capsule. The presentation is in early ages as a conductive hearing loss that does not worsen over time. The differential diagnosis are otosclerosis and disruption of the ossicular chain. A CT scan is necessary, but often is challenging to identify this anomaly. In most cases the diagnosis is made by an exploratory tympanotomy. The treatment of choice is the reconstruction of the ossicular chain when possible, or hearing aids, such as bone anchored hearing aid (BAHA) if the reconstruction is not feasible.