OBJECTIVE: To characterize the audiometric natural progression in patient-ears with small volume (<1,000 mm), treatment-naïve cochleovestibular schwannomas (CVSs) in Neurofibromatosis Type 2 (NF2).

STUDY DESIGN: Prospective, longitudinal cohort study.

SETTING: Quaternary medical research institute.

PATIENTS: One hundred eleven ears in 71 NF2 patients with small, treatment-naïve CVSs observed from July 2006 to July 2016.

INTERVENTION: Serial audiometric testing, including pure tone audiometry, speech audiometry, and magnetic resonance imaging (MRI).

OUTCOME MEASURES: Four-frequency pure tone average (4f-PTA) of 0.5, 1, 2, and 4 kHz and word recognition score (WRS) were recorded. Their changes were compared with MRI changes in CVS volume over time. Times to significant hearing loss (10 dB loss in 4f-PTA) and WRS based on 95% critical difference were measured.

RESULTS: Linear regression analysis showed a significant correlation with baseline hearing level (4f-PTA) and internal auditory canal (IAC) tumor volume to annual hearing decrease rate (AHDR) (p = 0.003, p = 0.0004). Hearing level at baseline and tumor volume correlate with AHDR while tumor volume growth rate does not. Two-way analysis of variance found significant differences in AHDR, risk of significant hearing loss, and risk of critical difference in WRS based on baseline hearing level (abnormal or normal) and IAC tumor volume (greater or less than 200 mm).

CONCLUSION: Subjects with normal baseline hearing and small IAC tumor component had a low AHDR and low risk of significant hearing loss and may warrant conservative management while the presence of baseline hearing loss and large IAC volume resulted in higher ADHR and greater risk for further hearing loss and may benefit from early treatment interventions.

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