Clinical picture of bilateral vestibular schwannomas, sudden bilateral hearing loss, and aviation

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A 17-year-old girl, with no known medical history, woke 4 hours after a transatlantic air flight complaining of bilateral hearing loss, headache, and severe vertigo.

Brain MRI revealed bilateral enhancing cerebellopontine angle masses and brainstem compression (figure). Histology confirmed these masses as schwannomas. Despite surgical treatment, the patient remains unable to hear the human voice and can only appreciate high-pitch noise and deep rumble.

A diagnosis of neurofibromatosis type 2 (NF2) was made by consensus clinical criteria. MRI and brainstem auditory evoked potentials performed on her parents and siblings were normal, indicating a likely de novo mutation in the merlin gene.

This case highlights two important clinical points. First, the inclusion of “at risk” minors in presymptomatic screening programs for NF2 may reduce the risk of long-term sequelae such as this. Second, the temporal and causal relationship between sudden hearing loss, vestibular schwannoma, and aviation needs further exploration.

The bilateral nature of the presentation suggests a causal rather than coincidental relationship with air travel; however, the mechanism is unclear. One hypothesis may be that the reduced cabin pressure resulted in tumor swelling and symptoms.

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Figure. T1-weighted sagittal brain images demonstrating large bilateral vestibular schwannoma and brainstem compression. The axial inset shows these as hyperintense on T2-weighted scans. Histology confirmed benign nerve sheath tissue (schwannoma).

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