

Hearing Loss and Chiari

Hearing loss, whether sudden or gradual, is not the most common ear related Chiari symptom but it can be extremely troubling. Of the nearly 2,000 patients in the Chiari1000 database, overall, 22% reported some level of hearing loss at the time of diagnosis, while more than half reported ringing, fullness, or popping in the ears. Patients who do experience hearing loss often must face it without knowing whether their hearing will ever return to normal.

Unfortunately, a recent literature review failed to shed much light on the issue. A group of researchers searched the medical databases for English language studies published between 2000-2025 in which the level of hearing was quantitatively assessed through testing. Unfortunately, they only found eight publications which met these criteria and of the eight, three were single patient case reports.

The studies represented 139 Chiari patients ranging in age from 10-44 years. The most common type of hearing loss was sensorineural which involves damage to either the auditory nerve or the inner ear hairs. Since the auditory nerve is a cranial nerve which originates in the brainstem, theoretically it is likely the source of the problem in Chiari patients. Two-thirds of the patients had hearing loss in one ear (unilateral) and one third in both ears (bilateral). A moderate level of hearing loss was the most common but there were some patients with either mild or severe levels of loss. Interestingly, some patients only experienced hearing loss intermittently, usually during a headache. In these cases, the testing for sensorineural hearing loss was usually negative.

There were only three reports on the effects of decompression surgery on hearing loss, with all three being single subject case studies. In these cases, which were all pediatric, hearing function returned to normal levels a few months after Chiari surgery. The specific mechanism which leads to hearing loss in Chiari patients is not well understood and attempts to further define it have so far produced mixed results. Given that sensorineural hearing loss can be permanent, this is an important topic that deserves more attention from the Chiari research community.

Source: Migliorelli A, Manuelli M, Bianchini C, et al. Hearing Loss and Chiari Malformation Type I: A Scoping Review. Diseases. 2025;13(10):315. Published 2025 Sep 25. doi:10.3390/diseases13100315

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