# **Tinnitus in Hereditary Menière's Syndrome**

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Abstract: In 1997, the authors described a large family with classic Menière's syndrome and migraine inherited as an autosomal dominant trait. Using a special questionnaire applicable to all the affected members of this family, the majority of respondents convincingly demonstrated true migraine preceding or after the Menière's attacks. Now we have used another questionnaire designed to describe the natural history of tinnitus and have posed the questions therein to all members of this sibship affected by migraine, Menière's syndrome, or both. In this way, we obtained a clear description of tinnitus regarding volume, intensity, and degree of annoyance; time of onset and duration; variations with time; and so on. The relations of tinnitus with the other Menière's symptoms and with the migraine also were explored. Only headache appears more often than tinnitus in these patients, and the latter is the only symptom constantly present, with little variation during migraine and Menière's attacks. These findings are discussed.

Keywords: hereditary Menière's syndrome, migraine, tinnitus

I n 1941 [1] and 1949 [2], Brown reported the familial incidence of Menière's syndrome. The mechanism of genetic transmission was autosomal dominant in all the families he described. One family, however, included three affected siblings whose parents were first-degree cousins. This finding raised the possibility of an autosomal recessive mode of genetic transmission. In 1965, Bernstein [3] reported several families with Menière's syndrome transmitted in an autosomal dominant mode. In 1992, Oliveira and Braga [4] reported one family having Menière's syndrome also transmitted in an autosomal dominant mode.

All the sibships cited here had headache either preceding or after the Menière's symptoms, and the headache was reported as "migrainelike." However the authors failed to clarify whether the headache indeed was migraine. In 1997, Oliveira et al. [5] described a large family with Menière's syndrome and migraine transmitted in an autosomal dominant mode. This time, however, they submitted a special questionnaire to all affected members of this family so as to answer the

<u>Reprint requests</u>: Carlos A. Oliveira, M.D., Ph.D., SHIS QL22, Conjunto 4, Casa 9, Brasília - D.F. - 71650-145, Brazil. Fax: 55 61 3463772; e-mail: Oliv@abordo.com.br This study was presented at Twenty-Fifth Ordinary Scientific Congress of the Neurootological and Equilibriometric Society, March 19–22, 1998, Bad Kissingen, Germany. question: Is the headache true migraine? In more than 80% of the cases, true migraine was present.

In 1979, Dolowitz [6] studied 125 patients with idiopathic (sporadic, nonhereditary) Menière's syndrome (Menière's disease) and found headache associated with the Menière's attacks in more than 80%. However, he did not characterize the headache as true migraine. In his statistical analysis of the symptoms, he found tinnitus to be a relatively independent symptom and to have a positive association with headache when positional vertigo was present. He speculated that tinnitus and headache could be central symptoms in Menière's disease.

From Dolowitz's work [6], we know that headache is very prevalent also in idiopathic (nonhereditary) Menière's syndrome. Therefore, it is possible that migraine is associated also with Menière's disease.

We have submitted a special questionnaire, designed to describe the natural history of tinnitus and its relationship with the Menière's and migraine attacks, to all affected members of the family that we described in 1997. Here, we describe the findings gathered from the answers to this questionnaire, and we offer our interpretation of these results.

Our goal was to describe the natural history of tinnitus in hereditary Menière's syndrome and migraine patients and to see what could be learned from its relations with other symptoms: migraine (central) and vertiginous attacks (peripheral).

#### **MATERIAL AND METHODS**

A standard questionnaire focusing on the symptom of tinnitus was submitted to 31 members of the family whose pedigree is shown in Figure 1. We excluded those members of this family who did not have any of the following symptoms: migraine headache, tinnitus, or vertiginous attacks. All family members who had any of the foregoing symptoms and were available answered the questionnaire.

#### RESULTS

A total of 31 members of the family answered the questionnaire. Tinnitus and vertiginous attacks were present in 19 and were absent in 12. Headache was present in 26 and was absent in 5. Of headache cases, 80% were true migraine. Tinnitus was present for more than 5 years in 13 patients, for 2-5 years in 3, for 1 year in 2, and for less than 1 year in 1. On a scale of 1-10, tinnitus intensity was 4 or more in 13 patients and was less than 4 in 6 cases. Vertiginous episodes preceded the appearance of tinnitus in eight patients, and tinnitus came first in four. Tinnitus and vertiginous attacks started together in two patients, and two patients could not remember the order of appearance. In nine patients, migraine was present before tinnitus started; tinnitus preceded migraine in two patients; and another two could not remember which symptom came first. Tinnitus intensity did not change during acute episodes of vertigo in 10 patients, increased during the crisis in 5, and had a variable behavior during the vertiginous attack in 1. During the migraine crisis, tinnitus was unchanged in 10 patients and worsened in 3 patients. In 14 cases, tinnitus did not change over time; in 4 it wors-



**Figure 1.** Pedigree of the described family. Some patients who had only tinnitus or headache appear in the figure only as open circles and squares but did answer the questionnaire. ( $\bigcirc$ , unaffected female;  $\blacksquare$ , affected female;  $\square$ , unaffected male;  $\blacksquare$ , affected male;  $\square$ , deceased female;  $\square$ , deceased male;  $\rightarrow$ , proband.)

ened; and it disappeared altogether in only 1. Hearing loss was present in 13 tinnitus patients and was absent in 6. Tinnitus worsened with alcohol intake in four cases, with stress in three, with noise in one, and with exercise in one. A total of 10 patients did not identify any intercurrent events that would worsen tinnitus. Finally, 15 patients described their tinnitus as a noise close to white (radio static, waterfall, whistle; Table 1).

### DISCUSSION

Headache was the most frequent symptom found in this family, being present in 26 of 31 patients who answered the questionnaire and, in more than 80% of these patients, it could be characterized as true migraine. Furthermore, migraine crisis appeared before vertiginous attacks and tinnitus in 76% of the patients who had all these symptoms.

Tinnitus and vertiginous attacks were present in 19 of 31 patients who answered the questionnaire. Tinnitus was present for more than 5 years in 68% of the patients and, in 16%, it was 2–5 years old. Of the tinnitus patients, 68% rated the intensity of the symptom as greater than 4 on a scale of 1–10.

Vertiginous attacks preceded tinnitus in 50% of the patients who had both symptoms, and tinnitus preceded the vertiginous crisis or came at the same time in 50% of the patients. Somewhat surprisingly, tinnitus did not change during the vertiginous attacks in 62% of patients, became more intense during the vertiginous episodes in 31%, and had a variable behavior during the vertiginous attacks in 7%. Regarding the migraine crisis, tinnitus did not change during the crisis in 77% of the patients who had both symptoms, and it worsened in 22%.

When asked about tinnitus behavior over time, 74% of the tinnitus patients said that the symptom did not change, 21% said that the symptom worsened over time, and only 5% said it disappeared altogether. Hearing loss was present in 68% of the patients with tinni-

Table 1.	Patient's	Description	of the	Tinnitus	Symptom
(N = 19)					

Description	No. of patients
Radio static	5
Ringing	1
Pulsation	1
Waterfall	3
Motor	2
Whistle	7

Note: Radio static + waterfall + whistle = white noise = 15.

tus, and 53% of them could not point out any stimulus that influenced their tinnitus behavior. Finally, 79% of the tinnitus patients described the symptom as a white noise (radio static, waterfall, whistle; see Table 1).

## CONCLUSIONS

In 76% of patients who have migraine, tinnitus, and vertiginous attacks, migraine starts before tinnitus and vertiginous crisis. As migraine is a central symptom, this progression would suggest that the pathological process begins in the central nervous system.

Tinnitus and vertiginous attacks are present with the same frequency (19 of 31 patients interviewed) and appear after migraine. This finding suggests that both are peripheral symptoms and that the pathological process that started in the central nervous system reaches the inner ear later.

In hereditary Menière's syndrome, tinnitus and vertiginous attacks seem to be caused by a permanent change in the inner ear. We do not know why tinnitus is constant and vertiginous attacks are episodic. However, migraine crisis often precedes vertiginous attacks, and this finding suggests a central trigger for both. Wellknown and generally accepted is that emotional crisis can provoke both migraine and Menière's attacks. Fluctuant hearing loss is the cochlear counterpart of the vestibular crisis and is absent in 22% of the tinnitus patients in our family.

Of the tinnitus patients we interviewed, 79% described their symptom as something close to a white noise. This incidence points to a diffuse rather than a localized process in the cochlea. This process probably involves the homeostasis of the inner ear fluids and causes both Menière's syndrome and endolymphatic hydrops.

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