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ORIGINAL ARTICLE

Screening for Menière's disease in the general population – the needle in the haystack

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Abstract

Conclusion. Based on clinical history alone, 98.4% of the population with vestibular vertigo do not qualify for a diagnosis of Menière's disease (MD). Although frequent in dizziness clinics, MD is rare in the general population. *Objective.* To narrow down the prevalence of MD in the general population. *Subjects and methods.* A representative sample adult population sample ($n = 4869$) was screened for moderate or severe dizziness/vertigo. Subsequently, 1003 participants completed a validated neurotologic telephone interview on vestibular vertigo (VV). Prevalence of MD was determined by stepwise application of clinical criteria according to the AAO (1995): (1) at least two vertigo attacks of ≥ 20 min duration, (2) unilateral hearing loss, and (3) accompanying cochlear symptoms. *Results.* Lifetime prevalence of VV was 7.4%. Of 243 participants with VV, 51 (21%) had recurrent vertigo lasting ≥ 20 min. Of these, nine reported unilateral hearing loss, and four had accompanying cochlear symptoms (1.6% of VV patients, population prevalence 0.12%).

Keywords: *Menière's disease, epidemiology, vertigo*

Introduction

Vertigo and cochlear symptoms are common complaints in clinical practice and when patients present with both, Menière's disease (MD) is often suspected. However, while MD is frequently diagnosed in specialized settings, accounting for 4–13% of cases referred to dizziness clinics [1–3], its prevalence at the population level is presumably rather low. Reported prevalence rates of MD vary considerably, ranging from 17/100 000 in Japan to 513/100 000 in Finland [4–7]. The differing rates may be related to methodological limitations of these studies such as a retrospective design, variable diagnostic criteria for MD, and study populations other than the general population. More importantly, however, most of these studies have been based on patient registers and not on screening of populations and may thus underestimate the true prevalence of MD.

This study aimed to narrow down the prevalence of MD in a large sample of community-dwelling adults by systematic screening and stepwise application of clinical diagnostic criteria based on the recommendations of the American Acadamy of Otolaryngology (AAO, 1995) [8].

Subjects and methods

This study was approved by the Institutional Review Board of the Charité, Humboldt University Berlin. Details on the study design of this neurotologic survey have been published previously [9]. In brief, we conducted a cross-sectional neurotologic study on the epidemiology of vestibular disorders in the general adult population living in Germany in 2003. This study combined a population-based and a neurotological approach. A random sample of the general population was contacted by telephone in the German National Health Interview Survey 2003 (GNT-HIS, $n = 8318$, modified random digit sampling, response rate 52%) and screened for the occurrence of dizziness with the question: 'Did you ever experience moderate or

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severe dizziness or vertigo?'. Subsequently, out of a simple random sample of 4869 GNT-HIS participants, 1157 fulfilled the inclusion criteria for the neurotologic survey (consent for further interviews, a history of moderate or severe dizziness/vertigo, and still valid telephone number), and 1003 (response rate 87%) completed the neurotologic interview.

The neurotologic interview was developed by the authors through piloting and validation in a specialized dizziness clinic. It contained both open-ended and standardized questions and allowed for interactive questioning similar to a clinical situation. The interview was designed to differentiate vestibular from non-vestibular vertigo on the basis of explicit diagnostic criteria and to identify specific neurotologic disorders (benign paroxysmal positional vertigo, migrainous vertigo, and MD).

The interviews were conducted by two medical student interviewers (F.L., M.F.) who were extensively trained in a neurological dizziness clinic over a period of 1 year. They discussed each interview with one of four experienced neurotologists (H.N., M.v.B., A.R., T.L.), who also supervised 10% of all interviews in person. Inconclusive cases were discussed in case conferences with all four neurotologists involved in the study. If necessary, additional information was obtained by a complementary interview.

A diagnosis of vestibular vertigo required one of the following criteria: (1) rotational vertigo, (2) positional vertigo, or (3) recurrent dizziness with nausea and either oscillopsia or imbalance. Rotational vertigo was defined as an illusion of self-motion or object motion, and positional vertigo was defined as vertigo or dizziness precipitated by changes of head position, such as lying down or turning in bed. As reported previously [9], vestibular vertigo was detected by our interview with a specificity of 94% and a sensitivity of 84% in a concurrent validation study using neurotology clinic diagnoses as an accepted standard ($n = 61$).

The prevalence of MD among those with vestibular vertigo was narrowed down by stepwise application of the clinical criteria of the AAO (1995) [8]: (i) occurrence of at least two vertigo attacks of at least 20 min duration, (ii) unilateral hearing loss, and (iii) at least one cochlear symptom such as tinnitus, hearing loss or aural fullness during at least two vertigo attacks. The fourth AAO criterion, namely exclusion of other causes, could not be fully applied as this would have required personal examination and neurotological testing. However, migrainous vertigo, benign paroxysmal positional vertigo, and vestibular neuritis were excluded by interview and the results have been reported [10,11]. To exclude a CNS origin of vertigo, patients were asked about additional central neurological symptoms such as weakness, double vision, sensory or speech problems.

The prevalence of vestibular vertigo and of MD was calculated taking into account the two-stage sampling design by multiplying the proportion of vestibular vertigo in the neurotologic survey with the proportion of dizziness/vertigo in the GNT-HIS participants as reported in a previous paper [9]. Thereby, non-responders and those lost to follow-up between the two sampling stages were assumed to have the same probability of vestibular vertigo as participants of the neurotologic survey. The confidence intervals (CI) for the estimated prevalences were calculated using the conservative Wilson method and taking into account the loss of power through non-response and loss to follow-up between the GNT-HIS and the neurotological survey. The frequency of MD among those with vestibular vertigo was then estimated by stepwise application of the clinical AAO criteria.

Results

Of the 1003 interviewed participants of the neurotological survey, 243 had experienced vestibular vertigo. This corresponds to a prevalence of vestibular

Table I. Stepwise estimation of the prevalence of Menière's disease in a representative sample of the adult general population in Germany ($n = 4869$).

Reported symptoms	*n*	Percentage of participants with vestibular vertigo	Prevalence in the general population (%)
Vestibular vertigo	243		7.4
Recurrent vestibular vertigo	216	89	6.5
Recurrent attacks of vertigo >20 min	51	21	1.5
Self-reported hearing loss	15	6.2	0.44
Unilateral hearing loss	9	3.7	0.27
Recurrent cochlear symptoms* during vertigo	4	1.6	0.12

*Cochlear symptoms: hearing loss, tinnitus or aural fullness.

Table II. Characteristics of four participants with accompanying cochlear symptoms during two or more attacks of vestibular vertigo and unilateral hearing loss.

No.	Sex	Age (years)	Interview diagnosis	Unilateral hearing loss	Accompanying cochlear symptoms	Other accompanying symptoms	Self-reported cause of hearing loss
1	F	60	MD	Yes	Tinnitus	No	None
2	M	76	MV	Yes	Aural pressure Hearing loss	Migrainous headaches	Sudden hearing loss (fully recovered)
3	F	32	Other*	Yes	Tinnitus Aural pressure Hearing loss	No	None
4	F	68	Other	Yes	Aural pressure Hearing loss	No	Otoslcerosis

MD, Menière's disease; MV, migrainous vertigo.
*Other is classified as 'other vestibular vertigo'.

vertigo in the general population of 7.4% [9]. Recurrent attacks of vertigo that lasted more than 20 min as required by the AAO criteria were present in 51 participants, representing a frequency of 21% of participants with vestibular vertigo and a prevalence of 1.5% in the general population (Table I). Of these, nine reported unilateral hearing loss, corresponding to 3.7% of vertigo patients and 0.27% of the general population. According to our stepwise approach, only four subjects additionally reported recurrent cochlear symptoms with their vertigo, thus narrowing down the prevalence of MD to 1.6% of vertigo patients and 0.12% of the general population.

In other words, based on history alone and before taking into account audiometric criteria and differential diagnoses, 98.4% of general population vertigo sufferers do not appear to have MD and the order of magnitude of the prevalence of MD in the general adult population is under 0.2%. One of the remaining four patients appeared to have migrainous vertigo and not MD (he fulfilled the diagnostic criteria for migrainous vertigo and had auditory symptoms which fully recovered) and one reported otosclerosis as the cause of her hearing loss (Table II).

Discussion

Our cross-sectional population-based neurotological survey showed that MD is a very rare condition at the population level. While the prevalence of vestibular vertigo was high in our study sample with an estimated 7.4% of the general population, only a small fraction of participants with vestibular vertigo qualified for a diagnosis of MD with stepwise application of the AAO criteria. Only one-fifth of participants with vestibular vertigo reported recurrent attacks lasting more than the required 20 min. A combination of vertigo attacks typical of MD and unilateral hearing loss occurred in only 3.7% of vertigo sufferers, corresponding to a prevalence of 0.27% of the general population. Additional accompanying cochlear symptoms reduced this number even further to an estimated population prevalence of MD of under 0.2%. This means that based on history-taking alone, the great majority of patients with vestibular vertigo did not qualify for the diagnosis of MD according to the AAO criteria.

There are some considerations as regards the strengths and limitations of this study. This survey provides a population-based estimate of the order of magnitude of the prevalence of MD. The results are likely to be highly representative of the general population, since the response rate of 87% of the neurotologic survey was high and the initially contacted sample of GNT-HIS participants is largely representative of the general adult population in Germany, as shown by a previously reported detailed analysis [9]. In addition, diagnostic criteria for MD were based on the 1995 AAO criteria. However, since the prevalence of MD has been found to be very low, a limitation of this study is its lack of power to reliably determine the exact magnitude of MD prevalence.

Uncertainty additionally arises from a possible misclassification of MD, since diagnoses were based on clinical history obtained by telephone interview only. The AAO criteria require audiometrically documented hearing loss, which could not be assessed by interview. However, a recent study showed that diagnosis of MD by means of a structured interview at the patient's first visit is reliable, with a specificity of 97% and sensitivity of 80% when compared with re-evaluation according to the AAO guidelines by an ENT specialist at follow-up. The best predictor for the presence of MD was self-reported hearing loss (sensitivity 100%, specificity 85% [12]).

Clinically, MD patients with audiometric evidence of hearing loss have usually experienced subjective hearing loss as well. Therefore, it seems unlikely that

we would have missed many MD patients with audiometric but not clinical evidence of hearing loss. On the contrary, among participants with subjective hearing loss, there may be some with normal audiometry. This would reduce the actual prevalence of MD even further.

Our estimated lifetime prevalences of MD provide a conservative estimate for other reasons. Firstly, although not specified in the AAO criteria, our diagnostic criteria required the occurrence of at least two attacks of vestibular vertigo with accompanying cochlear symptoms in order to minimize the bias of a chance coincidence. When developing the interview we also took into consideration that other conditions may mimick MD. To minimize bias by recurrent vertigo of central origin, which may rarely be accompanied by unilateral hearing loss (e.g. transitory ischemic attacks of the anterior inferior cerebellar artery), we incorporated a set of screening questions for evidence of central neurological involvement. None of the 51 participants with the required 2 or more attacks of vestibular vertigo of more than 20 min duration reported additional neurological deficits. Another important differential diagnosis of MD is migrainous vertigo (MV), which may present with vertigo attacks similar to those in MD and may equally be accompanied by cochlear symptoms including transient hearing disturbances [13]. Unlike MD, however, MV does not usually progress to permanent and profound hearing loss [14]. In our study, we diagnosed recurrent MV according to previously published clinical criteria, which include a history of migraine and accompanying migrainous symptoms during at least two attacks of vestibular vertigo [15]. Prevalence and clinical characteristics of MV in our study sample have been reported separately [11]. In this study, only one participant who qualified for MD according to clinical AAO criteria also fulfilled diagnostic criteria for MV.

Our estimated prevalence of MD lies within the lower range of previously published prevalences. Due to methodological restrictions, however, the reported rates also vary considerably and, unlike our data, do not always reflect population-based estimates.

The lowest prevalence of MD of 17/100 000 has been reported from nationwide surveys based on clinical reports from several medical centers in Japan [4]. However, the identified cases of MD were restricted to patients who presented to one of the hospitals affiliated with the research committee. Thus, these data are not population-based and the prevalence in the general population is likely to be higher. Furthermore, their diagnostic criteria for MD differed from the AAO criteria in that the duration of vertigo attacks and evidence of hearing loss were not considered, which on the contrary, may have resulted in an overestimation of the actual prevalence.

A retrospective study with cases of MD recruited from patient registers of seven hospitals in Finland reported a prevalence of 43/100 000 for MD according to the AAO criteria [5]. The true prevalence is presumably higher, since MD patients who did not seek medical advice or were managed by general practitioners or ENT specialists outside these hospitals are not reflected in this study. A considerably higher prevalence of 218/100 000 was reported from a population-based survey by means of a chart review of a centralized diagnostic index in Rochester, MN, USA [6]. Here, the high prevalence may in part be explained by the diagnostic criteria applied in this study, which were based on an older version of the AAO criteria [16]. These criteria still comprised the subclassification of 'vestibular MD,' delineating a syndrome of recurrent vertigo attacks without accompanying cochlear symptoms. Accordingly, the Rochester study included cases of isolated vestibular vertigo that would no longer be classified as MD according to the revised AAO criteria.

Surprisingly, a recent population-based questionnaire study reported a prevalence of MD of 513/100 000 in Southern Finland, which is considerably higher than all previously reported rates [7]. In this study – similar to our study – a population sample was first screened for vertigo. However, the prevalence of (presumably vestibular) vertigo as a prerequisite for the diagnosis of MD of 29% was almost four times higher than in our neurotological survey (8% vestibular vertigo, 21% non-vestibular vertigo [9]) and equals the prevalence of unspecified vertigo and dizziness reported at 20–30% in previous population-based studies [17,18]. According to the published questionnaire of the Finnish group, classification of 'vertigo' was based on a single question asking about experience of 'vertigo together with a moving sensation.' This question may not have been sufficiently discriminative in differentiating vestibular from non-vestibular vertigo. Secondly, from the report of Havia et al. [7] it remains unclear if the duration of vertigo attacks of a minimum of 20 min – as required by the AAO – was fulfilled, since their questionnaire provides only one category of vertigo duration (vertigo lasting 5 min to 4 h). Similarly, audiometrically documented hearing loss was apparently not available for all cases defined as definite MD. Due to these modifications of the diagnostic criteria for MD, this study may have overestimated the prevalence of MD.

The greatly varying prevalence estimates of these previous investigations as well as of our own study

underline the notion that MD, in fact, is a very rare disease at the population level. Determining its true prevalence therefore resembles searching for 'a needle in a haystack' and requires a very large study sample with enough power to reliably detect the few existing cases of MD according to strictly applied clinical criteria.

Previous studies have shown that both vertigo and cochlear symptoms are frequent in the general population: in our neurotological survey, vertigo was prevalent in 7.4% of the adult population [9]. In a nationwide survey on ear, nose, and throat problems in Scotland, dizziness characterized as a spinning sensation was experienced by 21% of the adult population, recurrent tinnitus was reported by 17%, any difficulty with hearing by 18% of participants [19]. This implies that vestibular and cochlear symptoms will often coincide by chance alone. In contrast, our study shows that MD is very rare in the general population and that, therefore, a wide differential diagnosis should be considered in patients presenting with combined vestibular and cochlear symptoms.

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