

Clinical and Audiovestibular Profile of Meniere's Disease in Remote Tertiary Center in India

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Abstract: ***Aim:** To determine the frequency of patients presenting with Meniere's Disease (MD), using the American Academy of Otolaryngology-Head and Neck Surgery (AAO) diagnostic criteria, and to describe the clinical and audio vestibular profiles of these patients. **Method:** Prospective case series design in the settings of a tertiary referral hospital. The study included all consecutive patients aged between 5 and 75 years presenting with the history of hearing loss, vertigo, tinnitus and or aural fullness as participants, satisfying inclusion and exclusion criteria for MD (AAO 1995) followed for a 12 months period. Main outcome measures comprised the evaluation of epidemiological profile, clinical features, and results of audio vestibular investigations like Pure Tone Audiometry with and without glycerol, Impedance Audiometry, Electrocochleography (ECOG), Distortion Product Otoacoustic Emission and Electronystagmography (ENG). **Result:** The frequency of MD was 15.6%, being commoner in males than females (2.6:1) and occurring more in the age group 40–49 years among males and 30–39 years among females. Low frequency hearing loss and high frequency tinnitus are more common. ECOG shows 76% positive predictive value. **Conclusion:** There is very high chance of missing those cases until we will not kept in mind of AAO 1995 diagnostic criteria.*

1. Introduction

- Meniere's disease (MD) is a clinical disorder first described by Prosper Meniere in 1861, characterized by fluctuating hearing loss, recurrent spontaneous episodic vertigo, fluctuating low pitch tinnitus and aural fullness. Endolymphatic hydrops is the basic pathology in MD wherein there is dilatation of endolymphatic spaces of membranous labyrinth due to excessive endolymphatic fluid. Currently, the diagnosis of MD is based on the diagnostic criteria set by the Committee on Hearing and Equilibrium, adopted in 1995 by the American Academy of Otolaryngology-Head and Neck Surgery (AAO). Accordingly, MD is classified into certain, definitive, probable and possible Meniere's disease.
- A literature search showed there were no studies that described the clinical and audio vestibular profile of MD in the Indian settings. Therefore this study was done to establish the frequency, clinical and audio vestibular profile of MD in a specialized tertiary care hospital in rural India using the 1995 criteria of the Committee on Hearing and Equilibrium.
- It is a very frequent complaints for ENT consultation. It mainly seen in unilateral ear but bilateral may involve.
- There may be immunological and autonomic over reactivity that leads to headache, pain abdomen & diarrhea etc.
- It is due to either excessive production of endolymph by dark cell of macula or by stria vascularis or defective absorption due to perisaccular ischaemia. Later one possess stronger basis.
- It is very distressing if not addressed early that can lead to functional level 5 – unable to find meaningful job or functional level 6- not able to do any work or handle family.

2. Material & Methods

All patients fulfilling the AAO criteria over a 12 month period (2018–19) attending the out-patient dept. of Anugrah Narayan Magadh Medical College & Hospital Gaya was considered for the study. Inclusion criteria were patients with true episodic vertigo or disequilibrium, fluctuating hearing loss and tinnitus/or aural fullness. Patients with chronic suppurative otitis media of tubotympanic type or atticofurcal type, history of head injury, autoimmune disorders, CNS abnormality on oto-neurological examination and investigation, typical benign positional paroxysmal vertigo, vestibular neuronitis, exposure to ototoxic drugs, mental retardation and pregnant or lactating mothers were excluded. The latter group was excluded due to the practical difficulties in performing the battery of audiological tests in this subset of patients.

All patients included were evaluated by an ENT specialist using a structured vertigo evaluation form that included a detailed history, complete ENT and oto-neurological examination. Screening tests were done to rule out systemic diseases like dyslipidaemia, diabetes mellitus, thyroid dysfunction and autoimmune disorders. The patients were also subject to the following audio vestibular tests: pure tone audiometry (PTA), PTA with glycerol if hearing loss was present, impedance audiometry, electrocochleography (ECochG), distortion product oto-acoustic emission (DPOAE), electronystagmography (ENG).

3. Result

A total of 447 new patients had attended the ENT OPD during the study period among whom 70 (15.6%) satisfied the AAO criteria. Among the 70 patients, 53 were classified as definite MD, 3 had probable and 14 had possible MD as per the AAO criteria (Fig. 1). Patients ranged from 19 to 70 years, the largest group seen in the 41 to 55 age group (Table 1). The mean age for males was 45 and 39 years for

females. Among the 70 patients, 52 (74%) were males and 18 (26%) were females (M:F 2.6:1).

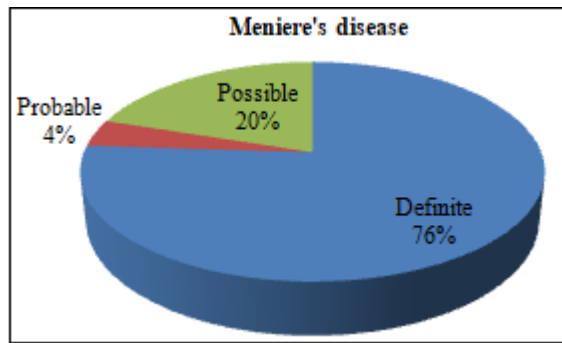


Figure 1: Distribution of meniere's disease as per AAO criteria

Table 1: Age distribution of pt recruited

Age groups in years	Numbers	Percent
10-25	7	10
26-40	16	23
41-55	33	47
56-70	13	19
>70	1	1

- Majority of the patients had symptoms of Meniere's disease lasting for more than a year. Of the 70 patients, 55 (79%) had typical surrounding rotatory type of vertigo (Fig. 2) and 15 (21%) had head rotating vertigo. Of the 15 with head rotating vertigo without the illusion of movement of surrounding, 9 (17%) had definitive MD and 6 had possible MD. 51 patients (71%) had vertigo for more than a year whereas 19 (27%) had these attacks for less than a year

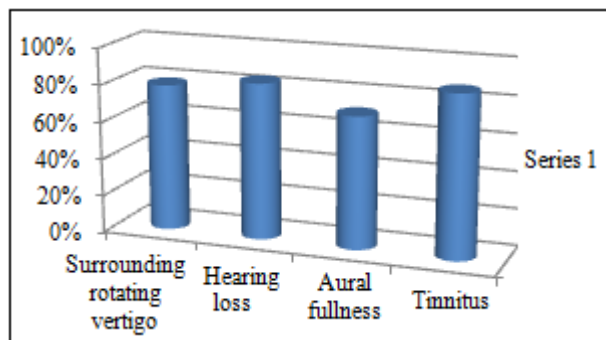
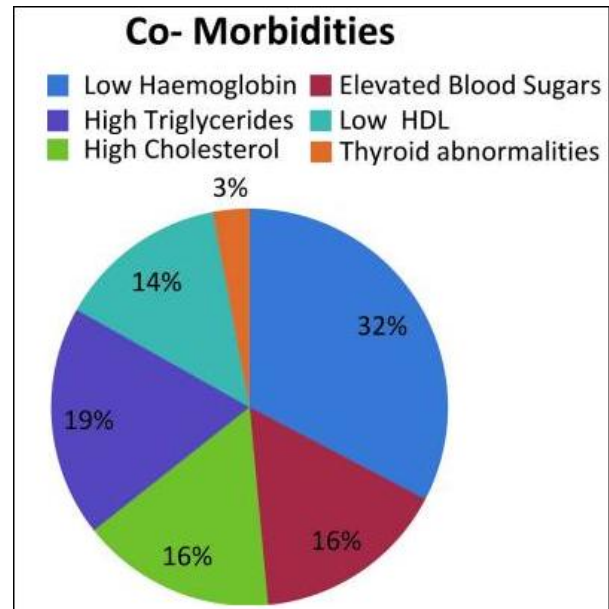


Figure 2: Distribution of various symptoms in MD

- A history of hearing loss was present in 59 (84%) of the 70 patients (Fig. 2), of which 27 (45%) had a history of fluctuating hearing loss. Half of the 53 with definite MD had fluctuating hearing loss and among these, 37 (53%) had worsening of hearing in the course of their disease and in 13 (19%) the hearing had remained the same since its onset.
- Among the 70 patients, 50 (71%) had aural fullness, among whom 20 patients (40%) had typical ear fullness or pressure at the time of vertigo attack. Seventeen of the 53 with definite MD, (32%) had intermittent aural fullness.
- Of the 70 patients, 61 (87%) had tinnitus. 51 (96%) patients with definite MD had tinnitus of whom 12 (21%) had bilateral tinnitus. Tinnitus lasted for more than

3 years in 41 (67%) of the 61 patients. In more than half (59%) it was high pitched, in 25 (41%) it was low pitched and in 23 (38%) it increased during an attack of vertigo.

- Routine blood investigations showed that a number of patients had associated co-morbidities like low hemoglobin, elevated blood sugars, high cholesterol, high triglycerides and low HDL levels. One patient had high TSH level and one had low TSH level (Fig. 3). None of the patients had positive serology test for syphilis



Pure Tone Audiometry showed 12 (17%) patients with hearing loss of the right ear, 16 (23%) patients with hearing loss of the left ear and 29 (41%) patients with bilateral hearing loss. 13 (19%) patients did not have any hearing loss. The commonest pattern of audiogram was the flat type. Among the 57 patients with documented hearing loss, all had glycerol pure tone audiometry test. Positive glycerol test was noted in 25 cases (44%) (Table 2).

Table 2: Tests done on MD and its results

Tests done	Total tested	Abnormal test	Comments
PTA	70	Hearing loss- 57(81%)	Pattern of PTA: Flat (47%), down sloping (4%), upsloping (11%)
Glycerol PTA	57	Positive- 25(47%)	17 Positive among 53 definite MD
Tympanometry	70	Type C-4(5.6%)	
Acoustic reflex		Absent reflex (15%)	
DPOAE	70	Absent 42 (60%)	
ECohG	52	Unilateral- 21(30%), Bilateral 39(56%)	Positive predictive value- 76%
ENG and bithermal caloric test	70	43(61%)	Right canal paresis-30%, Left canal paresis-11%, Bilateral canal paresis-20%

- Among the 53 definite MD patients, 25 patients (44%) had positive glycerol test. Among the 17 without definite Meniere's (3 probable + 14 possible), only two had glycerol test (as the others did not have hearing loss); of which one had a positive glycerol test. On impedance audiometry, 66 had type A and 4 had C type of tympanogram. 85% patients had normal reflexes and 15% had absent reflex. 60% ($n = 42$) had absent responses on DPOAE while remaining had normal DPOAE.
- Among the 70 patients, 21 had (30%) unilateral abnormal ECoG, 39 (56%) bilaterally abnormal ECoG and 9 (13%) had normal ECoG. Eight patients with possible MD also showed bilateral abnormal ECoG while 9 had normal ECoG. Among the 53 definite MD patients, 52 patients had ECoG test (not done in one). Of the 52, 45 (87%) had a positive ECoG test, 7 (13%) had negative ECoG (Table 2); 14 (82%) out of 17 probable and possible Meniere's had a positive test. The positive predictive value calculated was 76% and the negative predictive value was 30%
- Of the 70 recruited, 67 (95.7%) had normal smooth pursuit system, 21 (30%) had right and 8 (11%) had left canal paresis and 14 (20%) had bilateral canal paresis. Among 14 bilateral canal paresis only 9 had definite MD; 16 had right directional preponderance (DP), 15 had left DP and 39 had no DP.
- Among the 53 definite MD cases 31 patients (57%) were in stage 3 and 8 (15%) were in stage 4. 27 (39%) were in functional level scale 3 and 23 (32%) were in level 4.

4. Discussion

- MD is a condition which is mainly diagnosed from a detailed history and examination. According to AAO it is a diagnosis by exclusion. A literature search showed that there were no data published in peer reviewed journals showing the incidence or prevalence of MD in a tertiary hospital in a developing country like India. Hence the need to have a comprehensive insight into the various clinical presentations of a not so rare condition formed the basis of this study.
- In world literature, the frequency of MD varies widely, with incidence ranging from 10 to 157 per 100,000 in developed countries. The present study found that the frequency of MD in a tertiary level hospital is about 15.6%. However this hospital being a tertiary centre does not reflect the extent of the problem among the general public.
- In our study of 70 patients, 53 (75.7%) had definitive MD, 3 (4.3%) had probable Meniere's disease and 14 (20%) had possible MD. Their age ranged from 14–70 years, the mean age for males being 45.84 years while the mean age for females was 39 years. Mizukosh et al. showed that the age distribution peaked at 40–49 years for males, while the peak for females was around 30–39 years. While the age of our patients was similar to elsewhere, we observed a male preponderance which is different from the equal male: female ratio shown in the world literature. However a few reports have also shown a female preponderance. Our findings could be because the male/female attendance in our ENT OPD also has a

male preponderance (2:1) and more men in India seek medical attention than women.

- Paparella and Mancini described vertigo in about 91% of Meniere's patients. The typical Meniere's type of vertigo attack seen in our study was 79% ($n = 55$) while the remaining 15 (21%) presented with head rotatory vertigo without illusion of movement of surrounding. It is important to remember that Meniere's patients could present with both typical 'surrounding vertigo' and just 'head rotatory vertigo'
- Literature reports stabilization of hearing loss after a year of deterioration, while our experience has been progressively worsening in half the patients although they had been suffering from the disease for many years. Deterioration of hearing levels are seen initially in the low frequencies especially in patients with unilateral disease. Paparella showed aural fullness in MD is 74.1%. In our study 71% patients had aural fullness with worsening symptoms during the attacks in 42%.
- The tinnitus in MD is typically described as low pitched. However in our study high pitch tinnitus was more common than low pitched tinnitus. A third of our patient had low hemoglobin, elevated sugars, dyslipidaemia and thyroid dysfunction. This may be due to the high incidence of these conditions in the Indian population. Haid et al. have described these as co-morbid factors related to age. Regular screening for thyroid dysfunction is required especially in the elderly age group
- Mayerhof et al. revealed the audiogram pattern was flat in 42%, rising in 7%, peaked in 32%, and sloping downward in the remaining 19%. This observation is similar to what we observed that the flat type was the most common (49%); however the peak type of audiometric pattern was not seen in our patients.
- Akioka et al., found that 47% patients had positive glycerol test in their series. Our study showed 44% of patients had positive glycerol test. This is well correlated with the other studies. This test is time consuming and uncomfortable for the patient and not sensitive enough as a screening test for MD.
- In this study 60% of patients had absence of DPOAE which was similar to the study by Cianfrone et al. The usefulness of this test is very limited in this condition. Electrocochleography has an important role in MD. Gibson et al. found enlarged SP:AP ratio in 62–78% of patients with Meniere's disease. Another study by Chung et al. found sensitivity and specificity of extra tympanic electrocochleography in the diagnosis of MD to be 71 and 96%, respectively. The positive predictive value for ECoG in our study was 76% but the low negative predictive precludes this test from being used to screen out those who do not have MD. In a tertiary care and specialist clinic setting, 76% of ECoG positive patients will have disease. This makes it a good screening tool when combined with history.
- Patients are likely to be referred late in the illness to us since 57% presented in stage 3 with 41–70 dB hearing loss and 15% in stage 4 in definite Menier's disease. The functional levels of most patients were level 3 (39%) and level 4 (32%), both of which would have influenced daily activity. Early diagnosis and treatment could have provided near normal life and may have prevented further progression of the condition

- MD in Indian population seems to have a profile similar to that in Western literature with minor variations. In spite of the established quadrad of symptoms for MD, patients present with varied clinical scenarios making the diagnosis equally confusing. A detailed structured questionnaire has been found to be very useful to identify the particular type of vertigo, detect various associated aggravating factors and to rule out other causes of vertigo. Majority of the patients had a functional level of 3 and 4, making them quite incapacitated to perform their daily routine activities. Early diagnosis, with a keen focus on a proper detailed history, and prompt treatment may prevent further episodes of vertigo, thus enabling them to maintain a better functional quality of life. Thus the role of history taking plays a pivotal role, and having a structured proforma will help the clinician in establishing the cause of vertigo as investigatory tests is at best only complementary in making a diagnosis.
- It is familial and follow autosomal dominant inheritance. It is more commoner in white race.
- Here one thing is clear that due to higher positive predictive value and greater positive cases for SP/AP complex or amplitude ratio makes ECochG/ECOG gold standard test for MD.
- Again only pure tone audiometry shows 81% hearing loss, if combined with dehydrating agent eg. Glycerol (others are Urea, Furosemide) in this study shows 47% positive cases which is very much comparable to ECOG. But again if combined with speech audiometry which shows >15 % improvement along with >10dB in PTA during vertigo spell it become best tool to diagnose MD according to AAO-HNS (1995) criteria.
- Pt. Can be managed well with medication, rest and labyrinthine adaptation exercise eg. Cooksey-Cowthorne.

5. Conclusion

Patients with MD constitutes a major group in an otolaryngology. In view of the varied clinical picture a detailed and structured history based on a preformed questionnaire is important to elicit the required diagnosis. In this study incidence rate is 15.6%. Definite category came larger 76%. Non invasive test ie Glycerol dehyd & PTA with speech audiometry are better screening tool. Early diagnosis and prompt medical treatment may ensure that the patients maintain a significant period of disease free intervals, thus achieving better functional levels of one or two. Attaining an improved functional level at the earliest would thus enable the patients to maintain a better quality of life.

6. Conflict of Interest

None

References

- [1] Committee on Hearing and Equilibrium Committee on Hearing and Equilibrium guidelines for the diagnosis and evaluation of therapy in Menieres disease. *Otolaryngol Head Neck Surg.* 1995;113:181–185. doi: 10.1016/S0194-5998(95)70102-8. [PubMed] [CrossRef] [Google Scholar]
- [2] Shojaku H, Watanabe Y. The prevalence of definite cases of Meniere's disease in the Hida and Nhkubiki districts of central Japan. A survey of relatively isolated areas of medical care. *Acta Otolaryngol Suppl.* 1997;528:94–96. [PubMed] [Google Scholar]
- [3] Huppert D, Strupp M, Brandt T. Long-term course of Meniere's disease revisited. *Acta Otolaryngol.* 2010;130(6):644–651. doi: 10.3109/00016480903382808. [PubMed] [CrossRef] [Google Scholar]
- [4] Mizukoshi K, Ino H, Ishikawa K, et al. Epidemiological survey of definite cases of Meniere's disease collected by the seventeen members of the Meniere's Disease Research Committee of Japan in 1975–1976. *Adv Otorhinolaryngol.* 1979;25:106–111. [PubMed] [Google Scholar]
- [5] Klockhoff I, Lindblom U. Endolymphatic hydrops revealed by glycerol test preliminary report. *Acta Otolaryngol.* 1966;61(5):459–462. doi: 10.3109/00016486609127084. [PubMed] [CrossRef] [Google Scholar]
- [6] Watanabe Y, Mizukoshi K, Shojaku H, et al. Epidemiological and clinical characteristics of Meniere's disease in Japan. *Acta Otolaryngol Suppl.* 1995;519:206–210. doi: 10.3109/00016489509121906. [PubMed] [CrossRef] [Google Scholar]
- [7] Harris JP, Alexander TH. Current-day prevalence of Ménière's syndrome. *Audiol Neurotol.* 2010;15(5):318–322. doi: 10.1159/000286213. [PubMed] [CrossRef] [Google Scholar]
- [8] Paparella MM, Mancini F, Liston SL. Otosclerosis and Meniere's syndrome: diagnosis and treatment. *Laryngoscope.* 1984;94:1414–1417. doi: 10.1288/00005537-198411000-00003. [PubMed] [CrossRef] [Google Scholar]
- [9] Enander A, Stahle J. Hearing in Meniere's disease. A study of pure-tone audiograms in 334 patients. *Acta Otolaryngol.* 1967;64(5):543–546. doi: 10.3109/00016486709139139. [PubMed] [CrossRef] [Google Scholar]
- [10] Belinchon A, Perez-Garrigues H, Tenias JM, Lopez A. Hearing assessment in Meniere's disease. *Laryngoscope.* 2011;121(3):622–626. doi: 10.1002/lary.21335. [PubMed] [CrossRef] [Google Scholar]
- [11] Paparella MM, Mancini F. Vestibular Meniere's disease. *Otolaryngol Head Neck Surg.* 1985;93(2):148–151. [PubMed] [Google Scholar]
- [12] Herraiz C, Tapia MC, Plaza G. Tinnitus and Meniere's disease: characteristics and prognosis in a tinnitus clinic sample. *Eur Arch Otorhinolaryngol.* 2006;263(6):504–509. doi: 10.1007/s00405-006-0019-9. [PubMed] [CrossRef] [Google Scholar]
- [13] Haid CT, Watermeier D, Wolf SR, et al. Clinical survey of Meniere's disease: 574 cases. *Acta Otolaryngol Suppl.* 1995;520:251–255. doi: 10.3109/00016489509125240. [PubMed] [CrossRef] [Google Scholar]
- [14] Brenner M, Hoistad DL, Hain TC. Prevalence of thyroid dysfunction in patients with Ménière's disease. *Arch Otolaryngol Head Neck Surg.* 2004;130(2):226–228.

- doi: 10.1001/archotol.130.2.226. [PubMed] [CrossRef]
[Google Scholar]
- [15] Meyerhoff WL, Paparella MM, Gudbrandsson FK. Clinical evaluation of Meniere's disease. *Laryngoscope*. 1981;91(10):1663–1668. doi: 10.1288/00005537-198110000-00012. [PubMed] [CrossRef] [Google Scholar]
- [16] Akioka K, Fujita N, Kitaoku Y, et al. A clinical study of the diagnosis of endolymphatic hydrops aspect of Meniere's disease. In: Kitahara M, et al., editors. *Meniere's disease*. Toronto: Springer Verlag; 1990. pp. 125–132. [Google Scholar]
- [17] Cianfrone G, Ralli G, Fabbriatore M, et al. Distortion product otoacoustic emissions in Meniere's disease. *Scand Audiol*. 2000;29(2):111–119. doi: 10.1080/010503900424525. [PubMed] [CrossRef] [Google Scholar]
- [18] Gibson WP, Moffat DA, Ramsden RT. Clinical electrocochleography in the diagnosis and management of Meniere's disorder. *Audiology*. 1977;16(5):389–401. doi: 10.3109/00206097709071852. [PubMed] [CrossRef] [Google Scholar]
- [20] Chung WH, Cho DY, Choi JY, et al. Clinical usefulness of extratympanic electrocochleography in the diagnosis of Meniere's disease. *Otol Neurotol*. 2004;25(2):144–149. doi: 10.1097/00129492-200403000-00011. [PubMed] [CrossRef] [Google Scholar].