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# **Audiological Assessment in Meniere's Disease**

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## **Abstract**

Meniere's disease is a progressive disorder characterized by recurrent episodes of spontaneous vertigo, sensorineural hearing loss and tinnitus, often with a feeling of fullness in the ear. The exact etiology is not known. In 1972, a diagnostic criterion for Meniere's disease was proposed by American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS), and till date, it has been revised twice in the years 1985 and 1995. The principal audiological investigation is pure tone audiometry combined with a glycerol test. Speech audiometry and otoacoustic emissions also play a limited role. The value of electrocochleography is limited.

**Keywords:** pure tone audiometry, glycerol test, speech audiometry, OAE, electrocochleography

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## **1. Introduction**

Meniere's disease named after an Italian scientist, Prosper Meniere, is a progressive disorder characterized by recurrent episodes of spontaneous vertigo, sensorineural hearing loss and tinnitus, often with a feeling of fullness in the ear [1]. The characteristic of the disease is that it is an unpredictable, fluctuating illness with noteworthy hidden disability [2].

The precise cause of Meniere's disease is still being investigated. It is believed to be associated with endolymphatic hydrops, that is, raised endolymph pressure in the membranous labyrinth of the inner ear, which gets dilated like a balloon when pressure increases and drainage is blocked. This results in swelling of the endolymphatic sac and other tissues in the vestibular system (responsible for the body's sense of balance), creating an acute vestibular imbalance, resulting in vertigo and fluctuating hearing loss [3].

Another suggested etiology is the autoimmune nature of the disease. The idea of autoimmunity was brought forward when improvement in bilateral progressive sensorineural hearing

loss was recorded following immunosuppressive therapy. The studies of the human endolymphatic sac have also suggested that it is the primary immunocompetent structure in the inner ear, which is capable of processing antigen, synthesizing antibodies and raising a cellular immune response [4].

No single test that makes the diagnosis of Meniere's disease with conformity has been established. Complete history, including a detailed description of the pattern of disease presentation supported by quantitative testing, helps in arriving at a diagnosis [5].

A number of international interdisciplinary organizations in a consensus paper have drawn diagnostic criteria for Meniere's disease. The paper suggests two categories of the Meniere's disease, definite and probable. The definition of definite variety incorporates a clinical criteria and observation of episodes of vertigo associated with audiometric findings of sensorineural hearing loss involving low and middle frequencies and triad of symptoms that include fluctuant hearing loss, tinnitus and/or fullness of the involved ear. The criteria limit the duration of vertigo from 20 min to 12 h. The definition of probable variety of Meniere's disease encompasses vertigo or dizziness and extends to episodic ear-related symptoms, which may occur for a variable period time between 20 min and 24 h [2].

The natural history of Meniere's disease is inconstant but usually progressive. The classical triad of tinnitus or aural fullness with episodic vertigo and hearing impairment is often not seen at the beginning of the disease. The disease starts as a single symptom entity, and only cochlear symptoms occur at the first stage. The period between the primary symptoms and the manifestation of other symptoms varies from months to several years with an estimated average of 6–18 months. After this period of variable duration, the complete triad of symptoms will appear [6].

Episodic attacks of vertigo (so-called Meniere's attack) are the most troublesome of the symptoms to the patient, and it is usually the symptom that causes the patient to seek medical treatment. The vertigo patient perceives either that the world is spinning around them or that they themselves are spinning. Typically, it occurs in the form of a series of attacks over a period of weeks or months, interspersed by periods of remission of variable duration [7].

When the patient experiences a feeling of rotation, the sign is nystagmus, which has been described as a condition of involuntary movements of eyeball. This is accompanied by other symptoms such as giddiness and sweating [8].

On most occasions, patients experience a heaviness or fullness of the involved ear, which is accompanied by impairment of hearing and ringing sensation. Often the beginning of symptoms is precipitous, which reaches its zenith within a span of minutes to hours. The entire episode persists for an hour so before it wanes. The patient may experience unsteadiness for a couple of hours or days after the attack subsides. In between episodes of the disease, subjects may suffer from positional vertigo [9].

Vertigo is the most disabling one among the cardinal symptoms of the disease. It adversely affects almost every aspect of life disturbing the normal lifestyle of the patient. The vertigo is made worse especially when movement is involved. Patient's ability to lead a normal way

life is hampered by risks of fall. The chances of such events are made worse by small head movements, which make the patient subjectively very "ill." Vertigo can completely undermine the individual. This leads the patients to confine themselves to bed until the symptoms improve [10].

Some sufferers experience "drop attacks," which are sudden, severe unexplained falls without loss of consciousness or associated vertigo. These drop attacks are due to acute utriculosaccular dysfunction and are triggered by changes in inner ear pressure affecting otolith function [11].

Another unusual pattern of clinical presentation has been described, known as Lermoyez attacks. As opposed to typical spells in which tinnitus and hearing loss precede and worsen with the onset of vertigo, in Lermoyez attacks increased tinnitus and hearing loss precede the vertiginous episode and dramatically resolve with onset of vertigo [12].

Tinnitus experienced by Meniere's patients is continual and does not abate with time, although its intensity may vary. In addition, it may be heard more as a loud roaring or buzzing sensation, rather than a whistling, and is most commonly non-pulsatile and of the low-frequency type. The pitch tends to be related to the region of the most severe hearing loss and the magnitude of tinnitus roughly proportional to the severity of hearing loss [13].

A sensation of aural fullness that may precede a definite vertiginous spell, is considered a symptom alternative to tinnitus in the criteria of AAO-HNS (1985, 1995) and is experienced by 74.1% of the patients [14].

The hearing loss usually affects one ear, which typically loses sensitivity to low-frequency sounds and is of sensorineural type. As the hearing thresholds rise, dynamic range decreases, the sounds are typically described as "tinny" by the patient, the quality of sounds becomes poor, and loudness of loud sounds rises rapidly due to a phenomenon known as recruitment. The patients become intolerant to such loud sounds. During the early days of the disease, the hearing loss tends to return to within normal thresholds, and however, later in course of disease, hearing loss persists and even deteriorates over the course of following episodes. Even in terms of frequency involvement, the hearing loss spreads to involve all the frequencies showing a flat line on the audiogram. The sensorineural hearing loss in Meniere's disease involves low frequencies giving a flat audiometric pattern, but sometimes we get peak audiograms that are nearly normal hearing at around 2 kHz and decreased sensorineural hearing at lower and higher frequencies. This type of pattern is considered to be diagnostic of Meniere's disease and is more commonly seen in patients with disease of short duration. Over time, the hearing loss flattens and becomes less variable [15].

Patients become profoundly deaf rarely in 1–2% of severely affected patients [2].

Additional features are diplacusis that is unusual sensitivity to noises, sounds can seem tinny or distorted known as dysacusis, a difference in the perception of pitch between the ears (43.6%) and recruitment (56%) [16].

## 2. Investigations used to support the diagnosis of Meniere's disease

As of now, no single test can claim to make a reliable diagnosis of Meniere's disease; rather it is based on a complete history with a detailed description of the pattern of disease presentation, supported by quantitative testing.

In 1972, a diagnostic criterion for Meniere's disease was proposed by American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS), and till date, it has been revised twice in the years 1985 and 1995 [5].

The latest criteria used for diagnosis of Meniere's disease are the one proposed by AAO-HNS in the year 1995 [5].

AAO-HNS (1995) also introduced a staging system for cases of definite Meniere's disease. Staging is based solely on hearing, which is the most readily measurable variable and most closely related to the natural history of the disease [17].

Stages 1 and 2 are considered representative of early reversible disease that is susceptible to remission, whereas stages 3 and 4 are considered fixed or not reversible.

A good classic history with the criteria as outlined by AAO-HNS is adequate for diagnosing a case of Meniere's disease.

### 2.1. Pure tone audiometry (PTA).

PTA is the elementary investigation for reaching at a diagnosis and following up the patient during the course of treatment. A four-tone average of 0.5, 1, 2 and 3 kHz has been adopted in the guidelines of AAO-HNS (1985, 1995), and a change of 10 dB or more in PTA or a greater than 15% change in word recognition score is considered a clinically significant change during diagnosis and treatment [5].

Pure tone audiometry is the primary tool applied by the clinicians to ascertain hearing thresholds of the patient. It is used to document the degree and type of auditory impairment. These elements along with shape of the audiogram lay the ground for designing and implementing a line of treatment. This investigation is principally used in adults and grown-up children as PTA is dependent on patient's cooperation and his/her response to the pure tone signals [7].

Pure tone audiometry has assumed an established role in diagnosis of hearing impairment. Pre-calibrated pure tones are presented to the patient in sound-treated chambers. Circumaural headphones, insert phones and bone oscillators are used to deliver these tones to patient to determine air-conduction and bone-conduction thresholds across a frequency range from 250 to 8000 Hz. The thresholds are recorded in terms of decibel HL. Specific algorithms are available to determine need for masking the non-test ear when asymmetrical hearing presents the risk of cross hearing. Procedures and formulae are available to calculate the initial masking levels needed both for air-conduction and for masking levels to determine masked thresholds. The thresholds so detected are plotted on an audiogram using standardized symbols. Air-conduction thresholds tell about the degree, and bone-conduction thresholds convey type of hearing loss.

Apart from the degree of hearing loss, the pattern of audiogram tells about the type of hearing loss, whether it is conductive, sensorineural or mixed. Pure tone audiometry is regarded by some as initial screening test of choice for audiological dysfunction [18].

Staging of Definite Meniere's disease may be done in all the patients on the basis of degree of hearing loss.

Stage	Four-tone average (dB) (0.5, 1, 2 and 3 kHz)
1	<25
2	26–40
3	41–70
4	>70

The hearing loss in Meniere's disease is unilateral in about 70–85% of cases. However, the incidence of bilaterality increases with the duration of the disease, reaching about 40% after 15 years [19].

Audiograms are performed at different points during the course or progression of disease, and the hearing loss shows a fluctuating pattern. During the initial stages of the disease, if the PTA is performed after an episode has subsided, the audiogram may look to be normal. As the patient continues to experience more and more attacks, some degree of hearing loss tends to persist in between the attacks. However, in the later stages of the disease, the hearing loss establishes as non-fluctuating and permanent [20].

In the early stages of Meniere's disease, the characteristic audiometric configuration is a rising curve, that is, as the frequency increases, the hearing loss decreases. This contour has also been called a "reverse slope" audiogram, since high-frequency losses are by far the most frequent pattern in the hearing-impaired population. Word recognition (discrimination) has been reported in some studies as poor as 32%. Although mid- and high-frequency sensitivity tends to be good in the early stages, as the disease progresses, these frequencies become involved, leaving the patient with a "flat" audiometric configuration. The hearing loss may progress to a profoundly impaired degree as the disease process continues. However, the degree of hearing loss seldom exceeds a 70 dB average [21].

There is no classical audiogram pattern that can be used to identify Meniere's disease. However, there are certain features that have been commonly observed. These include:

- A varying degree of sensorineural hearing loss.
- This hearing loss is usually low frequency.
- The audiogram may be "flat," upward sloping or downward sloping.

In a study of 211 consecutive patients with classic Meniere's disease, the audiological pattern was flat in 42%, peaked type in 32%, downward sloping in 19% and rising in 7% patients [22].

In a study conducted in our department, 31 patients of either sex between the age group of 18 and 65 years presenting with history of attacks of vertigo, accompanied with tinnitus,



sensorineural hearing loss and aural fullness were selected randomly; 64% of these patients had either a flat audiometric graph or a low-frequency hearing loss as evidenced by downward sloping audiometric pattern. In this study, in 18 patients, the duration of symptoms was less than 12 months. In these 18 patients, 29 ears were affected; of these, 45% ears had rising or peak type of audiograms. Further analysis of the audiometric pattern in this study revealed that in patients with sudden onset of symptoms, 50% ears showed either a rising or a peak type of graph. On the contrary, among those patients with gradual onset of symptoms, 76% ears showed a flat or sloping type of graph. This correlates with the findings of a study which concluded that rising or peak audiograms appear more commonly in patients with disease of short duration [23].

In a study comprising of 111 patients with Meniere's disease, pure tone and speech audiometry was performed. The affected ears showed reduced hearing in both the modalities. An objective classification method used to determine audiogram shape indicated that affected ears more frequently show "low" or "low + high" hearing losses. The study concluded that shape of hearing loss does not depend on duration of affection of disease [24].

## **2.2. Short increment sensitivity index test (SISI)**

It is a useful test in distinguishing between cochlear and retro-cochlear lesion. It determines the capacity of a patient to detect a brief 1 dB increment at a 20 dB supra-threshold tone (called carrier tone) in various frequencies (preferably at 1000 and 4000 Hz). If SISI score is above 70%, it is considered as positive SISI and pathology lies in the cochlea. If SISI score is <30%, it is negative SISI and here pathology lies elsewhere than inner ear [18].

On the basis of PTA report, all the patients suffering from sensorineural hearing loss may further be subjected to glycerol test.

## **2.3. Glycerol test**

The assumption that an increase in endolymph volume, with its effect on labyrinthine membrane behavior producing in part, the hearing loss and vestibular deficit in Meniere's disease has led to the administration of dehydrating agents like glycerol. The goal is to reduce the volume abnormalities in inner ear and produce a measurable change in response that is improvement in behavioral audiometric test scores.

Glycerol, a potent osmolar agent, elevates osmotic pressure of the liquid in which it is dissolved. In the cochlear context, upon administration to a patient it lowers the volume of membranous labyrinth. Animal studies in which endolymphatic hydrops was induced surgically, responded to glycerol through intracellular and extracellular edema. The increased secretion of glycoproteins following administration of glycerol induces endolymph flow into endolymphatic sac. Thus, glycerol promotes the absorption of endolymph both in radial and in longitudinal directions [25]. This is a simple and rapid method that provides information on the cochlear response to the osmotic changes produced by glycerol in the inner ear.

Glycerol is administered orally to patients to reduce fluid abnormalities in the inner ear. It affects hearing temporarily (for a few hours), the results of which are measured by audiogram.

## 2.4. Method of glycerol test

The patients are advised to report empty stomach on the day of investigation. PTA test is performed before the administration of glycerol and then patient is administered a solution of 86% of glycerol (1.5 mg/kg of body weight) dissolved in equal volume of physiological saline.

Pure tone audiometry is then repeated at 1, 2 and 3 hours of glycerol administration.

The glycerol test is regarded as positive:

- When the hearing threshold is lowered at least 15 dB at minimum three frequencies or
- When there is a total pure tone threshold shift of 25 dB at three consecutive frequencies or
- When there is a 16% improvement in speech discrimination [26, 27].

In the study conducted at our department (whose results have yet to be published), only those patients were included who had definite Meniere's disease based on the AAO-HNS criteria. All these patients were subjected to glycerol testing to determine their suitability for administration of hydrochlorothiazide. In this study, 74% of patients had a positive glycerol test.

Another study has reported its experience of using the glycerol test in 122 patients with combination of sensorineural hearing loss, tinnitus or vestibular symptoms, in which endolymphatic hydrops was considered a possibility. Fifty percent of patients ultimately were found to have endolymphatic hydrops and positive test [28].

In a study of series of 95 patients with Meniere's disease, 47% were found to have a positive glycerol dehydration test. Yet another study reported that 60% of patients with Meniere's disease have positive tests. This study also noted that positive tests were found only in ears with Meniere's disease [29]. In another study, intravenous administration of glycerol was performed instead of oral administration. Positive results were obtained in 50% (15 out of 30) of patients 1 hour after administration. The positive ratio was same as oral glycerol test [30]. Thus, the results of glycerol testing in our study are comparable with other studies. The relatively higher percentage of positive glycerol test in our study could be due to the fact that subjects included in our study met the criteria of "definite Meniere's disease" as laid down by AAO-HNS. In our study, the results of administration of hydrochlorothiazide to patients who tested positive on glycerol testing indicate an across the board improvement in symptoms of Meniere's disease at the end of follow-up. Thus, it may be concluded that glycerol testing in patients with "definite Meniere's disease" as assessed by AAO-HNS criteria is a good idea to select patients who will respond to administration of osmotic diuretics.

Most patients may suffer headache and nausea after drinking the glycerol for post-glycerol audiometric evaluation, which usually subsides after few hours.

## 3. Speech audiometry

The objective of speech audiometry testing is to measure patient's ability to identify speech stimuli, to confirm results of pure tone audiometry and to rule out the presence of nonorganic hearing loss or retro-cochlear pathology.



The battery of speech audiometry tests includes speech detection threshold (SDT), speech recognition threshold (SRT) and word recognition score (WRS).

Word recognition test scores are often plotted on a graph. A point on this graph represents the percentage words correctly repeated by the patient at a specified intensity level from completed standardized list of words. Cochlear pathology tends to demonstrate a “plateau effect,” reaching a ceiling of performance at <100% and no improvement in the score despite a rise in intensity.

#### 4. Otoacoustic emission

An otoacoustic emission (OAE) is a low-level sound emitted by the cochlea either spontaneously or evoked by an auditory stimulus. Specifically, OAEs provide information related to the function of the outer hair cells (OHC) [31].

The use of OAEs in the assessment of patients with Meniere's disease has been well documented. According to Van Hufflen et al., patients with Meniere's disease can essentially be divided into four categories. It has been documented that OAEs present in ears with minimal hearing loss and absent in ears where pure tone thresholds exceed 60 dB HL. Both these scenarios are acceptable clinically. Among patients who have hearing thresholds in intermediate range of 30–60 dB HL two scenarios emerge. In the first scenario, OAEs are measurable in large values, while OAEs are not measurable at all. It has been postulated that variable patterns of OAEs in patients with Meniere's disease may be due to more than one precise sites of lesion. It was postulated by Hall that among patients in whom OAEs are present despite a hearing impairment more than 30 dB, the audiometric picture is not representing outer hair cells. He said that in these patients, the outer hair cells may have been spared and data are representative of inner hair cell function [32].

A number of studies have been conducted to determine the utility of OAE and changes in OAE during glycerol test undertaken to diagnose endolymphatic hydrops. In a study, TEOAE and DPOAE were measured before and 3 h after oral administration of glycerol in 22 years of patients with Meniere's disease. The positive result in the glycerol test was observed in 11 of 22 ears. However, of the two OAEs, DPOAE was considered more appropriate than TEOAE for monitoring during the glycerol test because of its high sensitivity in detection of changes in cochlear function. The study concluded that clinical use of OAE, especially DPOAE, as a test complementary to pure tone audiometry during the glycerol test is very useful and will improve the diagnosis of endolymphatic hydrops [33].

The worth of DPOAE to spot minimal inner ear dysfunction which might be due to endolymphatic hydrops and may otherwise go undetected on pure tone testing has been emphasized by a few clinical studies. A study specifically examined a set of patients whose only presenting complaint was fullness of ear/ears. The study used glycerol test with PTA and DPOAE to diagnose subjects at an early stage of Meniere's disease and may have the potential to progress toward the full-blown disease. It was concluded that those patients in whom the only symptom is fullness of ear/ears may potentially be in early stages of the condition [34].

A study was conducted to follow up the dynamics of pure tone threshold and DPOAE amplitude changes induced by glycerol with reference to its activity in inner ear; 38 patients with Meniere's disease and having positive glycerol test were included in the study. It was concluded that audiometry and DPOAE measurements in the glycerol test procedure are the most profitable after the third hour since glycerol administration due to the most significant outcomes of both at this time. Observed changes in pure tone audiometry concern lower frequencies and in DPOAE middle frequencies [25].

## 5. Electrocochleography

"Electrocochleography" (ECoChG) is an investigation employed to measure electrical potentials of cochlea. The components of ECoChG include measurement of cochlear potentials in response to a stimulus and measurement of the whole nerve or compound action potential (AP) of the eighth nerve.

A study conducted to evaluate the role of ECoChG in the diagnosis of Meniere's disease concluded that ECoChG has little role in diagnosing or ruling out Meniere's disease [35].

Although the utility of ECoChG diagnosis continues to be disputed, it has been proposed that in the situation that ECoChG confirms the diagnosis of Meniere's disease, one can be confident in deciding about an invasive therapy. What course be adopted when patient exhibits symptoms that confirm the diagnosis of Meniere's disease, but ECoChG is within normal limits remains a dilemma [36].

## 6. Conclusion

It is a challenging task to arrive at a conclusive diagnosis of Meniere's disease. A detailed history must be elicited from the patients, who should be encouraged to describe the symptoms in their own language. The sequence of events as they appeared and their characteristics help in arriving at a possible diagnosis. A thorough clinical examination is a mandatory component of initial encounter with the patient. Pure tone audiometry is the principal tool to document nature and degree of hearing loss. It is highly useful to have previous audiograms and compare them with the current audiogram. It helps understand fluctuant nature of disease and its progression. Glycerol test is a useful component of audiological assessment and has been combined with PTA and OAE. In author's experience, results of glycerol test in combination with PTA may be used to decide the possible line of treatment. Those patients with a positive glycerol test give better results with diuretics such as hydrochlorothiazide and those with a negative test respond better to oral steroids such as prednisolone. The word recognition scores recorded in speech audiometry demonstrate a typical "plateau effect," reaching a ceiling of performance at <100% and no improvement in the score despite a rise in intensity. OAE, especially DPOAE, has been particularly found useful in diagnosis of Meniere's disease. The role of electrocochleography (ECoChG) in the diagnosis and monitoring of treatment for Meniere's disease remains controversial.

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