


Original Article

An International Survey of the Diagnosis and Management of Ménière's Disease Amongst Otolaryngology Consultants

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BACKGROUND: Ménière's Disease (MD) is a disease that may be difficult to diagnose and manage. Our UK survey showed variability in the practice of UK Otolaryngology consultants. We hence surveyed Otolaryngology consultants internationally, to assess their confidence levels in diagnosing MD, their use of the AAO-HNS guidelines and current diagnostic and treatment modalities.

METHODS: An online questionnaire was distributed internationally over four weeks. The questionnaire asked respondents to anonymously rank their confidence in diagnosing MD, identify the minimum investigations required to make a diagnosis, describe their use of the AAO-HNS criteria, share their preferred treatment modalities for acute attacks, and state their 1st and 2nd-line preventative treatment options.

RESULTS: A total of 173 responses were collected with 77% of respondents reporting high levels of confidence in diagnosing MD. Most respondents stated the minimum tests required were "History, Otoscopy, Clinical Vestibular testing, and Pure Tone Audiometry" although some chose as few as 1 test. Regarding the use of the AAO-HNS criteria, responses ranged from "always" (20.2%) to "never" (22.5%). Cinnarizine was the first-line treatment for acute attacks followed by betahistine. Betahistine (30.1%) and dietary restrictions (28.3%) were recommended almost equally as first-line preventative measures. The most popular second-line measure was intratympanic steroids injection (30.1%).

CONCLUSION: Our survey revealed disparities in the diagnosis of MD and its management, like the results of our previously conducted UK survey. This suggests the need for an international consensus regarding the diagnosis and subsequent management strategies for this disease.

KEYWORDS: Diagnosis, guidelines, inner ear, Ménière's disease, vertigo

INTRODUCTION

Ménière's Disease (MD) is a rare condition¹ that results in spontaneous, severe bouts of rotatory vertigo, along with sensorineural hearing loss and tinnitus in the affected ear. Most commonly, MD is unilateral but can become bilateral in 30% of cases.² The exact

cause of MD is still unknown, with genetic and environmental factors likely contributing to both pathogenesis and progression of this disease. Histological and radiological studies have demonstrated the expansion of the endolymphatic compartment within the inner ear^{3,4} but the cause of this remains unclear.

Diagnosing MD can be challenging since episodes are intermittent, attacks can occur weeks or months apart, and clinical signs are usually limited to the duration of the acute attacks.⁵ Moreover, patients may not be able to access medical attention during the attacks, resulting in delayed diagnosis and treatment.

To aid in the diagnosis of MD, the American Academy of Otorhinolaryngology-Head and Neck Surgeons (AAO-HNS) developed guidelines for the diagnosis and treatment of MD in 1972 and revised them in 1985 and 1995.⁶ Published literature on MD has largely adhered to these criteria, but accurate diagnosis and hence exact treatment remains a challenge. Guidelines were updated in 2015 with a supplement published in 2020 aiming to “improve the quality of the diagnostic workup and treatment outcomes of MD”.⁷ Furthermore, anecdotal evidence suggests that Otolaryngology Consultant practice in the UK varies significantly.

This research group performed two surveys to assess current practice, including the use of the AAO-HNS guidelines for diagnosis and management of MD. A UK survey⁸ indicated significant variability in national practice in both diagnosis and treatment of MD, as well as low adherence to the recent AAO-HNS guidance.

As a result of the controversies surrounding the diagnosis and treatment of MD, and disparities seen in the UK Otolaryngology Consultant population, we conducted an international survey to assess current practice among Otolaryngology Consultants worldwide, aiming to potentially improve the management of this condition.

METHODS

An anonymous online questionnaire, approved by the Research and Innovation Department of Medway Maritime Hospital, Gillingham, Kent, was generated via Google Forms and distributed electronically

between the dates of April 24, 2023 to May 2, 2023, via otolaryngology European Board Examiners (Appendix 1). This was similar to the survey that was distributed to the UK Otolaryngology Consultant population earlier the same year. The contributors were Otolaryngology specialists currently working and voluntarily consented to participate in the study. No ethics approval was required for this type of study.

The questionnaire first asked respondents to rank their confidence in diagnosing Meniere’s disease on a 1-5 Likert scale. Respondents were also asked to identify the minimum required investigations for a diagnosis of MD from a list of 11 items that ranged from clinical assessment to radiological imaging and formal vestibular tests. Additionally, the survey assessed the use of the AAO-HNS guidelines in diagnosing MD and the recommended treatment of acute attacks of the disease. Finally, respondents were asked their initial first-line treatment options to prevent acute attacks from a list of 16 items (with an option to include treatments not listed) and their second-line option should this fail.

RESULTS

A total of 173 responses were recorded. On asking consultants about their confidence in diagnosing MD, the majority of respondents reported high levels of confidence. On a scale of 1 (not confident) to 5 (very confident), 46.8% (n=81) of respondents scored a 4 and 30.6% (n=53) of respondents scored a 5 (Figure 1). Looking at the data per country (Figure 2), only respondents from Bulgaria (n=1), Saudi Arabia (n=1), and the Czech Republic (n=1) rated their confidence below an average of 3/5 per country, with the average response in the rest of the countries being above that.

The second question asked clinicians about the minimum tests required to make a diagnosis of MD. Respondents were asked to choose 1 or more responses from an 11-item list that included history, clinical vestibular testing, vestibular function testing, and radiological imaging. The results varied from “History” alone (n=4, 2.3%) to “History, Otoscopy, Clinical Vestibular Testing, Pure Tone Audiometry (PTA), Caloric Testing and Video Head Impulse Testing (vHIT), MRI” (n=6, 3.4%). A significant proportion of respondents stated, “History, Otoscopy, Clinical Vestibular Testing, and Pure Tone

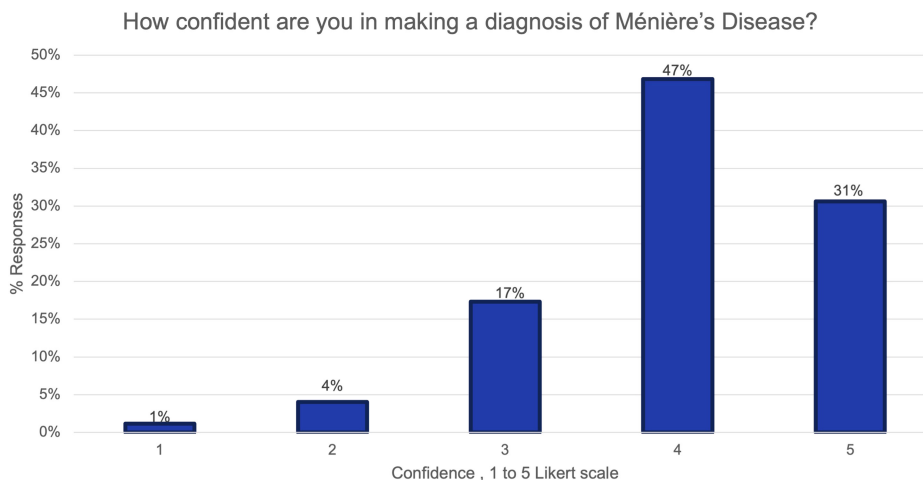


Figure 1. Otolaryngology consultant answer distribution to survey question “How confident are you in making a diagnosis of Ménière’s Disease?”, 1 to 5 Likert scale (1 being not confident and 5 being very confident).

Geographical Heatmap of International Survey Responses

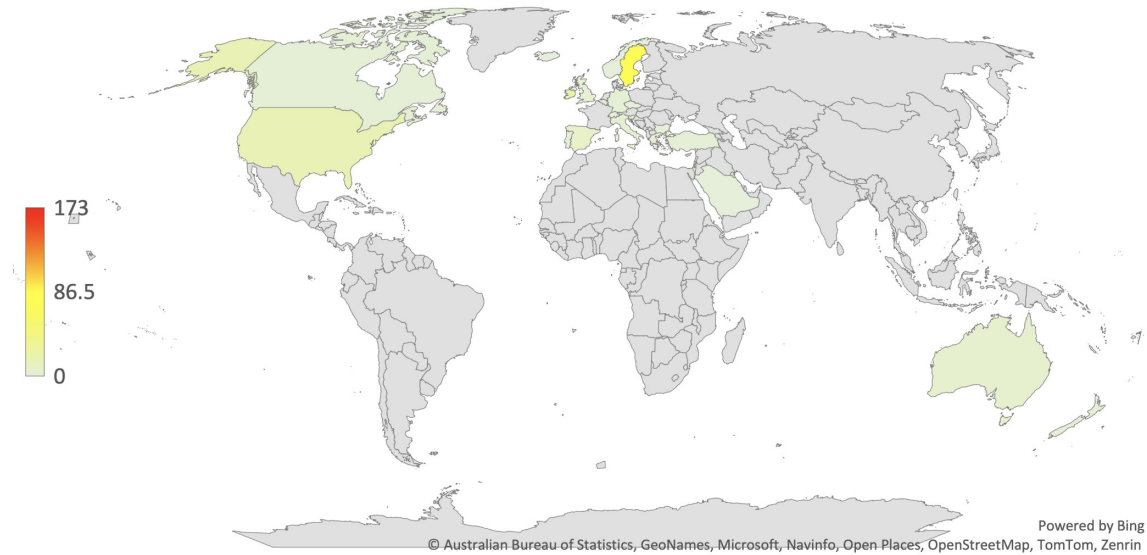


Figure 2. Geographical heatmap of international survey responses.

Audiometry” (n=20, 11.6%) would be the minimum tests required (Figure 3). History (n=169, 97.7%), PTA (n=162, 93.6%) and Otoscopy (n=108, 62.4%) were the three most chosen diagnostic modalities.

Respondents varied significantly when asked if they used the most recent AAO-HNS criteria for diagnosing MD. Answers ranged from “always” (20.2%, n=35) to “never” (22.5%, n=39). Thirty point one percent of respondents (n=52) stated they used the criteria “often”, 16.2% (n=28) “sometimes” and 11% (n=19) “rarely” (Figure 4).

The first line treatment option of acute attacks also varied, with cinnarizine being chosen by 32.9% (n=56) of respondents (52 of whom were from Sweden). This was followed by betahistine (18.5%, n=32) and prochlorperazine (12.7%, n=22). A minority of respondents recommended benzodiazepines (12.1%, n=21), dietary modifications (7.5%, n=13), vestibular rehabilitation (2.3%, n=4), promethazine (6.4%, n=11), and diuretics (6.9%, n=12) (Figure 5).

With regards to the first-line preventative measures recommended by Otolaryngology Consultants, the majority suggested the use of either betahistine (30.1%, n=52) or “dietary restrictions (e.g., salt-limiting diet)” (28.3%, n=50). The remaining minority were recommended diuretics (17.3%, n=30), grommet insertion (8.27%, n=15), intratympanic steroids (4.05%, n=7), vestibular rehabilitation (2.89%, n=5), and oral steroids (1.2%, n=2) (Figure 6).

The recommended second-line preventative measure if the first-line option were to fail also demonstrated significant variability. The most popular choice was intratympanic steroids (ITS) (30%, n=52) followed by diuretics (16%, n=28), betahistine (14%, n=25), and grommet insertion (11.5%, n=20). A minority selected grommet insertion with intratympanic steroid injection (6%, n=10), cinnarizine (5%, n=8), intratympanic gentamicin (6%, n=10), vestibular rehabilitation (3%, n=6), promethazine (2%, n=3), and saccus decompression (2%, n=3) (Figure 7). One response was given for each of lifestyle change,

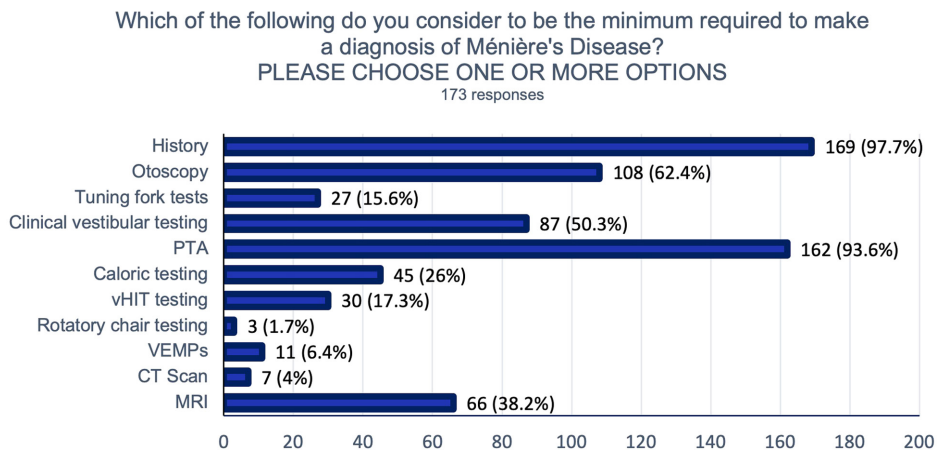


Figure 3. Otolaryngology consultant answer distribution to survey question “Which of the following do you consider to be the minimum required to make a diagnosis of Ménière’s Disease?”

Do you use the most recent American Academy of Otolaryngology - Head and Neck Surgery Criteria (AAO-HNS) when diagnosing Ménière's Disease?

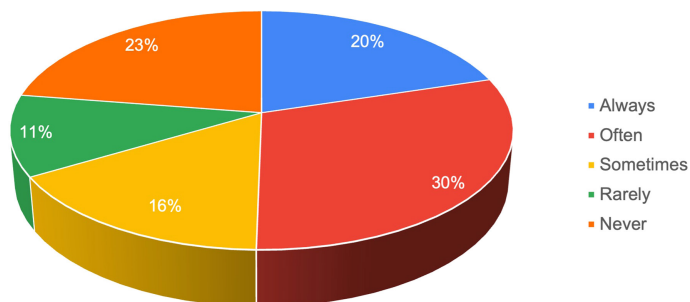


Figure 4. Otolaryngology consultant answer distribution to survey question "Do you use the most recent American Academy of Otolaryngology—Head and Neck Surgery Criteria (AAO-HNS) when diagnosing Ménière's Disease?"

dietary restrictions, and anti-migraine medication (0.6%, n = 1) each). Four respondents used the free-text insertion to state that each case is different (0.6%, n = 1) and that they use methods not mentioned such as SPC flakes (0.6%, n = 1) or CGRP- antagonists (0.6 %, n = 1).

DISCUSSION

MD is a rare disease with a prevalence ranging from 3.5 to 513 per 100,000 of the population.⁹ The variability in prevalence has been partly attributed to different populations and partly to frequent changes in the definition and diagnostic criteria for MD. The diagnosis of MD is relatively straightforward when the presentation fits the criteria, but many patients do not present with a classical history and are either too early in the disease process or have multiple co-existent pathologies. Diagnosis is complicated further by the complexity of differentiating MD from other causes of vertigo such as vestibular migraine, BPPV, vestibular hypofunction, and emotional overlay.¹⁰ This diagnostic complexity has been thoroughly discussed, and multiple attempts have been made to clarify the diagnostic criteria, with the most recent one being published by AAO-HNS in 2015. Unfortunately, despite the recent efforts, the results of our previous survey⁸ show that there is significant variability in standard practice and many clinicians do not regularly use the AAO-HNS guidelines. The lack of a universally used and agreed-upon framework and a common definition for the diagnosis and treatment of MD fuels the problem further.

What is your first-line treatment option for the treatment of acute attacks of Ménière's Disease?

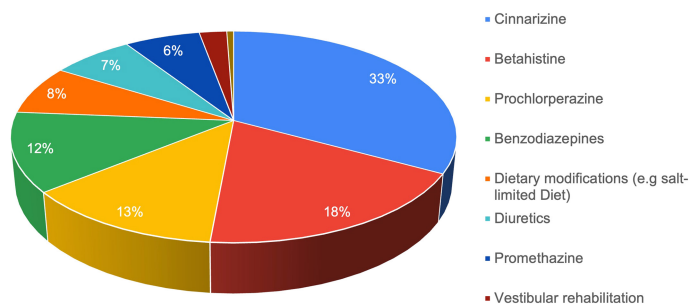


Figure 5. Otolaryngology consultant answer distribution to survey question "What is your first-line treatment option for the treatment of acute attacks of Ménière's Disease?"

What is your first line preventative treatment option for Ménière's disease?

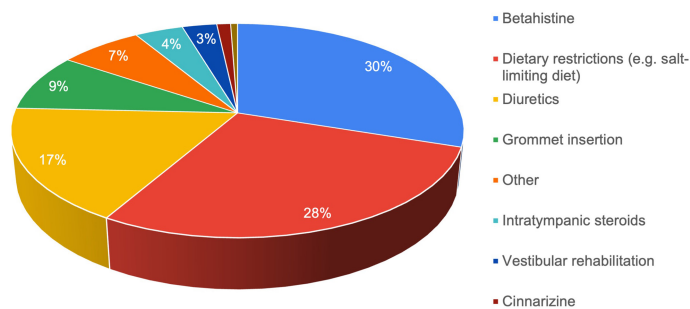


Figure 6. Otolaryngology consultant answer distribution to survey question "What is your first-line preventative treatment option for Ménière's Disease?"

Compared to the UK population surveyed, internationally more clinicians selected the need for specialist investigations such as vestibular testing, MRI, and PTA to diagnose MD. In this survey, the majority of clinicians considered taking a patient history, performing an otoscopic examination, PTA, and clinical vestibular testing to be the minimum tests required to make a diagnosis of MD. Interestingly, only 38% of respondents chose an MRI scan, and less than a third chose instrumental vestibular testing as part of the minimum tests required to make a diagnosis of MD. This finding might reflect the difference in availability, cost, or use of those resources internationally. There is recent evidence that gadolinium-enhanced MRI can help differentiate between MD and other menieriform diseases by detecting the degree of endolymphatic hydrops.¹¹ Given the complexity and cost of this investigation, though, it remains to be seen if it will be used internationally. It is important to set guidelines that both provide good diagnostic accuracy and acknowledge the differences in resource availability internationally.

Variability was also seen in the treatment options chosen for both prevention and acute MD treatment. MD has a relapsing and remitting course; this makes it very difficult to assess and analyze with adequate statistical power the effect of medical treatments. Most studies looking at medication are hence underpowered, and clinicians seem to choose medication more empirically rather than research-based.

If your first line option were to fail (i.e. acute attacks continue), what is your second line preventative treatment option for Ménière's disease?

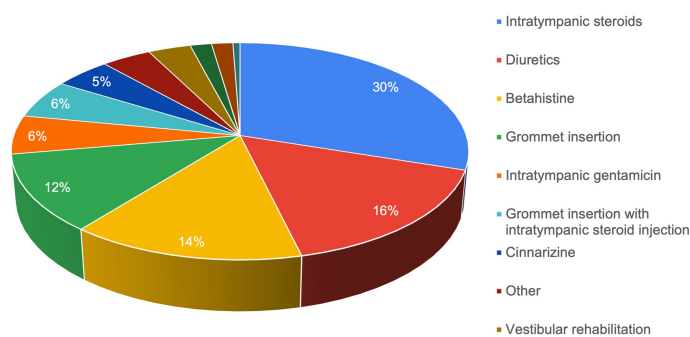


Figure 7. Otolaryngology consultant answer distribution to survey question "If your first-line option were to fail (i.e., acute attacks continue), what is your second-line preventative treatment option for Ménière's Disease?"

The majority of consultants in this study selected cinnarizine as a first-line treatment option for acute attacks, most originating from Sweden. Excluding Swedish respondents, dietary restrictions and betahistine were the most popular options selected internationally. Limited evidence exists for the use of cinnarizine for the treatment of acute MD attacks. It must be noted that cinnarizine is, however, used in acute vertigo secondary to vestibular migraine or migraine attacks.¹²

Betahistine was a popular option for both UK and international respondents for acute episodes and the preventative treatment of MD. The AAO-HNS guidelines recommend the use of betahistine for prevention of MD attacks in line with previous evidence. A recent systematic review, however, as well as the guideline supplement, notes that the highest quality evidence currently for betahistine indicates no significant difference in reducing frequency of vertigo attacks compared to placebo.¹³ Further pharmacological treatments such as diuretics also lack good quality evidence.¹⁴ The number of patients seen in studies looking at the efficacy of ITS injections is also low and provides a low level of evidence for their use to decrease vertigo attacks.^{15,16} ITS injections were selected as a second preventative treatment option by the majority of Otolaryngology Consultants surveyed. This is also in line with the recommendations of the International consensus (ICON)¹⁷ where ITS are recommended as a second-line treatment after a trial of diuretics. This may indicate that the treatment seems effective to most clinicians despite limited evidence.

Other options included the use of specially processed cereals (SPC) and anti-calcitonin gene-related peptide (CGRP) antibody therapy. SPCs have been shown to increase antisecretory factor (AF) levels intrinsically. AF can modulate water and ion concentrations and is hypothetically able to regulate the endolymphatic pressure in MD. Several studies with low patient numbers have shown promising results using SPC vs. placebo in decreasing vertigo, tinnitus, and improving quality of life^{18,19} but no high-level evidence exists. Anti-CGRP monoclonal antibodies have been used in the treatment of migraine with good results.²⁰ No evidence exists for the use of anti-CGRP in MD. The similarity of MD to vestibular migraine and the difficulty obtaining the correct diagnosis possibly makes such treatments effective in a subpopulation of patients suffering from vestibular migraine and again indicates the need for clear diagnostic criteria to differentiate these two entities.

Patients who are incorrectly diagnosed with MD can receive inappropriate treatment. This may impact a patient's quality of life but also influences research outcomes, resulting in underpowered and inconclusive studies. In the 2015 guidance, MD patients are divided into two categories, "definite" and "probable". For a patient to classify as "probable MD", no specific diagnostic test or examination is required. The authors of this paper believe that the "probable MD" category is too lenient, leading to increased diagnostic uncertainty and possible misdiagnosis.

MD usually has a self-limiting course with severe symptoms resolving in 5-15 years with or without treatment. A study by Silverstein et al. following up with patients with severe MD found that after 2 years, 60% of patients were no longer experiencing vertigo, and after 8 years, 71% no longer had dizziness symptoms.²¹ It should be noted

that despite the arrest of the vertigo episodes, vestibular function and hearing of the affected ear continue to deteriorate with time. A small subgroup of respondents in this survey opted to use ototoxic measures as treatments for MD if first-line treatments failed. The authors believe the use of such measures should be limited as their toxicity on the vestibular system can lead to impaired daily function and a deterioration in the patients' quality of life.²²

A limitation of this study includes limited representation of international practice, as it collected responses from 173 otolaryngology consultants. Furthermore, the spread of responses was not equally distributed between countries, with the majority of the responses (44.5%, n = 77) arising from Swedish clinicians. The survey also failed to capture information on the respondents' levels of experience in managing MD, and it should be noted that not all participants may routinely encounter these patients in their regular clinical practice. Additionally, cultural factors and insurance policies may influence the availability and choice of treatment options in different countries. The survey did not explore the underlying reasons behind each response and therefore could not assess the attitudes of participants toward certain topics such as the AAO-HNS guidelines or acute and preventative interventions.

CONCLUSION

The results of this survey have provided a snapshot of MD diagnosis and management internationally. Interestingly, the results demonstrated a significant variation in practice both in acute and preventative treatment as well as in the diagnostic methods used to make a diagnosis of MD. This disparity suggests the need for an international consensus for the diagnosis and management of MD in order to improve patient care and permit treatment outcomes to be compared.

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Informed Consent: N/A.

Peer-review: Externally peer-reviewed.

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Appendix 1- Digital survey distributed to Otolaryngology consultants internationally via Google forms

AN INTERNATIONAL SURVEY ON THE DIAGNOSIS AND MANAGEMENT OF MÉNIÈRE'S DISEASE

Ménière's disease has been defined as an inner ear disorder characterized by recurrent spontaneous episodes of vertigo accompanied by sensorineural hearing loss, tinnitus, and aural fullness.

This survey aims to explore expert opinions regarding the diagnosis and management of this disease.

We ask that you kindly take approximately 2 minutes of your time to complete this survey. All responses will be kept anonymous.

*Required

1. How confident are you in making a diagnosis of Ménière's Disease?

- *1. Not Confident
- 2.
- 3.
- 4.
5. Very Confident

2. Which of the following do you consider to be the minimum required to make a diagnosis of Ménière's Disease? *

PLEASE CHOOSE ONE OR MORE OPTIONS

Tick all that apply.

1. History
2. Otoscopy
3. Tuning fork tests
4. Clinical vestibular testing
5. Pure tone audiometry
6. Caloric testing
7. vHIT Testing
8. Rotatory chair testing
9. VEMPs
10. Computerized Tomography (CT) Scan
11. Magnetic Resonance Imaging (MRI)

3. Do you use the most recent American Academy of Otolaryngology—Head and Neck Surgery Criteria (AAO-HNS) when diagnosing Ménière's Disease? *

Never

Rarely

Sometimes

Often

Always

4. What is your first-line treatment option for the treatment of acute attacks of Ménière's Disease? *

PLEASE CHOOSE ONE

1. Dietary modifications (e.g., salt-limited diet)
2. Vestibular rehabilitation
3. Cyclizine
4. Prochlorperazine
5. Cinnarizine
6. Promethazine
7. Betahistine
8. Diuretics
9. Benzodiazepines

5. What is your first line preventative treatment option for Ménière's disease? *

PLEASE CHOOSE ONE

1. Vestibular rehabilitation
2. Dietary restrictions (e.g., salt-limiting diet)
3. Betahistine
4. Diuretics
5. Prochlorperazine
6. Cinnarizine
7. Promethazine
8. Intratympanic steroids
9. Intratympanic gentamicin
10. Grommet insertion
11. Grommet insertion with intratympanic steroid injection
12. Grommet insertion with intratympanic gentamicin injection
13. Saccus decompression
14. Vestibular nerve section
15. Gentamicin ablation surgical labyrinthectomy
16. Other:

6. If your first line option were to fail (i.e., acute attacks continue), what is your second line preventative treatment option for Ménière's disease? *

PLEASE CHOOSE ONE

1. Vestibular rehabilitation
2. Dietary restrictions (e.g., salt-limiting diet)
3. Betahistine
4. Diuretics
5. Prochlorperazine
6. Cinnarizine
7. Promethazine
8. Intratympanic steroids
9. Intratympanic gentamicin
10. Grommet insertion
11. Grommet insertion with intratympanic steroid injection
12. Grommet insertion with intratympanic gentamicin injection
13. Saccus decompression
14. Vestibular nerve section
15. Gentamicin ablation Surgical labyrinthectomy
16. Other: