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Clinical Practice Guideline: Ménière's Disease Executive Summary

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Abstract

Objective. Ménière's disease (MD) is a clinical condition defined by spontaneous vertigo attacks (each lasting 20 minutes to 12 hours) with documented low- to midfrequency sensorineural hearing loss in the affected ear before, during, or after one of the episodes of vertigo. It also presents with fluctuating aural symptoms (hearing loss, tinnitus, or ear fullness) in the affected ear. The underlying etiology of MD is not completely clear, yet it has been associated with inner ear fluid volume increases, culminating in episodic ear symptoms (vertigo, fluctuating hearing loss, tinnitus, and aural fullness). Physical examination findings are often unremarkable, and audiometric testing may or may not show low- to midfrequency sensorineural hearing loss. Imaging, if performed, is also typically normal. The goals of MD treatment are to prevent or reduce vertigo severity and frequency; relieve or prevent hearing loss, tinnitus, and aural fullness; and improve quality of life. Treatment approaches to MD are many, and approaches typically include modifications of lifestyle factors (eg, diet) and medical, surgical, or a combination of therapies.

Purpose. The primary purpose of this clinical practice guideline is to improve the quality of the diagnostic workup and treatment outcomes of MD. To achieve this purpose, the goals of this guideline are to use the best available published scientific and/or clinical evidence to enhance diagnostic accuracy and appropriate therapeutic interventions (medical and surgical) while reducing unindicated diagnostic testing and/or imaging.

Keywords

fluctuating aural symptoms, electrocochleography, endolymphatic hydrops, endolymphatic sac decompression, gentamicin, labyrinthectomy, Meniett device, sensorineural hearing loss, sodium-restricted diet, vestibular testing, quality of life

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Introduction

Ménière's disease (MD) is a clinical syndrome affecting approximately 50 to 200 per 100,000 adults and is most common between the ages of 40 and 60 years.[1] In 1861, Prosper Ménière noted that vertigo, off-balance, and hearing loss symptoms associated with MD reflected a lesion of the inner ear. Strict clinical classification to diagnose MD has been established by the American Academy of Otolaryngology–Head and Neck Surgery Foundation (AAO-HNSF).[2-4] These diagnostic criteria for MD were recently revised by the Classification Committee of the Barany Society in cooperation with several national and international organizations and were later approved by the AAO-HNSF Equilibrium Committee.[5,6] These revisions include 2 categories:

Definite MD:

- Two or more spontaneous attacks of vertigo, each lasting 20 minutes to 12 hours
- Audiometrically documented fluctuating low- to midfrequency sensorineural hearing loss in the affected ear on at least 1 occasion before, during, or after 1 of the episodes of vertigo
- Fluctuating aural symptoms (hearing loss, tinnitus, or fullness) in the affected ear

- Other causes excluded by other tests

Probable MD:

- At least 2 episodes of vertigo or dizziness lasting 20 minutes to 24 hours
- Fluctuating aural symptoms (hearing loss, tinnitus, or fullness) in the affected ear
- Other causes excluded by other tests

The diagnosis of MD is made clinically, as the disease typically presents with unilateral ear symptoms that can last for several decades.[7] MD attacks are typically random and episodic (approximately 6-11 per year), with periods of remission that may last months to years.[1] As such, the diagnosis of MD is typically not made at one point in time; rather, it may take months or even years to fully appreciate the clinical manifestations leading to definitive diagnosis. To maximize treatment, it is important to clinically distinguish MD from other independent causes of vertigo that may mimic MD and that also present with hearing loss, tinnitus, and aural fullness. Diseases such otosyphilis, vestibular neuritis, acute labyrinthitis, and so on respond to different treatments. Due to the variability in clinical presentation in patients with definite and probable MD, it is important to acknowledge that a full and accurate diagnosis may take many months to attain. This is an important consideration since this speaks to the natural history and variable clinical presentation of MD that the panelists on this clinical practice guideline (CPG) felt should be highlighted. This can directly affect clinical decision making and subsequent treatment recommendations.

The underlying etiology of MD is not completely clear, yet it has been associated with anatomic changes in inner ear fluid volumes described by the term *endolymphatic hydrops* (ELH), a hallmark feature of the disease that can be pathologically confirmed postmortem.[8,9] While EHL is not synonymous with MD, endolymph within the inner ear membranous labyrinth is postulated to increase, culminating in episodic ear symptoms, including vertigo, fluctuating hearing loss, tinnitus, and aural fullness. Schuknecht and Gulya[10] postulated the theory of Reissner's membrane rupture secondary to endolymphatic duct distention. These microtears would allow potassium-rich endolymph to bathe cochlear hair cells and the eighth cranial nerve. As such, repeated exposure to toxic levels of potassium-rich perilymph could cause episodic vertigo as well as long-term decline in auditory function (reviewed in Oberman et al[11]). While it has been reported that ELH was found in all patients with MD, not all found to have ELH had concurrent MD.[12] The clinical records and histopathologic slides of all cases of ELH in the otopathology laboratory at the Massachusetts Eye & Ear Infirmary were reviewed (n = 79), which included 35 cases with ''idiopathic hydrops'' and 44 cases having secondary hydrops in addition to some other otologic disease process. Among the idiopathic cases, 26 (74%) had clinical MD symptoms, while 9 (26%) cases did not meet the diagnostic criteria for MD. Because it is understood that ELH may be a final common pathologic pathway for a variety of inner ear insults, it is difficult to draw solid conclusions regarding the symptomatology experienced by the 44 cases of secondary hydrops due to factors such as clinical symptom overlap between hydrops and other otologic diseases or possible vestibular organ damage and deafferentation, which could limit the possibility for affected subjects to experience vertigo.

Disorders that may (eg, autoimmune inner ear disease, temporal bone fracture, otosyphilis, end-stage otosclerosis, endolymphatic sac tumors, acoustic neuromas)[9] or may not (eg, vestibular migraine) be associated with ELH can mimic MD, thereby placing an important emphasis on diagnostic accuracy. This also posits that ELH may cause MD but also suggests that ELH may simply be a by-product of a separate underlying process that leads to MD. Therefore, ELH may be necessary but not sufficient for MD development.

The natural course of MD is typically progressive and fluctuates unpredictably. In the early stages of disease onset, the frequency of acute vertigo attacks increases during the first few years and may eventually decline to near complete cessation of vertigo.[13] The natural progression of vertigo attack periodicity and severity over time in MD patients is not well understood, as others have reported that patients with MD can have severe attacks of vertigo even 20 years after the initial diagnosis.[14] While the patient's hearing may worsen or persist, patients with MD may also have hearing that stabilizes over time. While vertigo attacks may or may not improve over time, hearing loss will typically worsen and persist. In fact, a 20-year longitudinal study demonstrated that 82% of MD patients experienced moderate to

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Table 1. Key Definitions for Ménière's Disease (MD) Guideline.[72,a]

Vertigo	Sensation of self-motion (rotary spinning) or movement of the environment when neither is occurring or the sensation of distorted self-motion (rotation or spinning) during an otherwise normal head movement
Imbalance	Sense of unsteadiness, or instability; discrete from vertigo; may be ongoing and not episodic
Acute MD attack	Vertigo episode that lasts for 20 min to 12 hours and aural symptoms (timing impacted by treatment onset)
Active MD	Describes periods during which episodic acute attacks of MD occur with some regularity
Definitive MD	See above definitions in body of text
Drop attacks (Tumarkin's Otolithic Crisis)	Sudden fall associated with discrete MD attacks with no warning; the patient does not lose consciousness. Drop attacks may be experienced during later stages of MD and they are not present in every patient
Usable hearing	Levels of adequate hearing perception often defined by the patient; may be audiometrically defined based on level of hearing loss (HL), pure tone average (PTA) and word recognition/discrimination scores (WRS) from vestibular schwannoma literature: AAO-HNSF Scale: Class A: Discrimination 70-100%; PTA <30 dB Class B: Discrimination 50-69%; PTA 31-50 dB Class C: Discrimination 50-69%; PTA >50 dB Class D: Discrimination <50%; any PTA Most clinicians consider Class A and B/C to be useable or serviceable hearing; Class D not considered serviceable hearing.
Probable MD	See criteria within body of CPG
PTA	Pure Tone Average measured by audiometry
Hearing loss in MD	Often fluctuates from low- to mid-frequency but over time may involve all frequencies

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severe hearing loss (mean pure tone hearing loss >50 dB).[15] Given the episodic nature of MD attacks, it is challenging to distinguish between asymptomatic periods when the disease is quiescent in between attacks and the positive effects of treatment versus alternative diagnoses that may mimic MD (eg, vestibular migraine). Moreover, in the elderly patient or in the patient with long-standing MD that no longer manifests significant vestibular disturbance, there may not be typical MD-like temporal patterns. These patients may manifest episodes of severe imbalance or ''vague'' dizziness. Some vertigo control (up to 60%) has been documented in the placebo groups of published randomized controlled trials (RCTs)[16-19] with commensurate improvements in symptoms other than hearing loss irrespective of treatment.[20] These features pose challenges for formalized clinical trials to study MD, as the power of the studies is nearly impossible to achieve given the low incidence and natural fluctuations of MD.

The goals of MD treatment are to prevent or at least reduce the severity and frequency of vertigo attacks. In addition, treatment approaches aim to relieve or prevent hearing loss, tinnitus, and aural fullness and to improve overall quality of life (QOL). Treatment approaches to MD are many and typically include modifications of lifestyle factors (eg, diet) and mental health treatment or are medical and/or surgical. A separate goal is to enhance patient preferences and preference-centered care to minimize the adverse effects of therapies in both scope and frequency. Because the etiology of MD is not clearly known, inherent limitations about the efficacy of proposed treatments exist. Moreover, the variable or variables that cause symptoms in the setting of ELH are not clearly understood. As a result, the literature reports many MD studies that are poorly designed and often underpowered with inadequate controls, which can lead to inconclusive results. This can lead to the belief by many clinicians in specific unsubstantiated therapeutic approaches, resulting in tremendous practice pattern variation and subjective treatment regimens and reporting of MD control.

Some of the traditional treatment approaches for MD include dietary/lifestyle and/or trigger management approaches[21,22]; medical, surgical, complementary/alternative, allergy, immunomodulatory, vestibular, and aural therapy; and oral[21,22] or intratympanic medications—all with variable results.[23,24] For those MD patients with persistent and disabling attacks after several months of conservative therapy, other more invasive or involved treatments can be considered.[25,26] One main consideration about the choice of treatment is the hearing status and whether it is usable or not. In those patients with usable hearing (based on vestibular schwannoma literature; see definitions in **Table 1**), nonablative procedures have been advocated. These interventions include those designed to affect the natural history of MD with conservation of inner ear auditory function by suppressing vestibular function or endolymph production. Conversely, in those patients with no meaningful/useful hearing, surgical or chemical inner ear ablative treatments are often implemented.[27] The rationale for ablative approaches is to attempt to convert a dynamic fluctuating inner ear lesion (active MD) to a static state through destruction of the inner ear. In doing so, most therapies are designed to control vertigo rather than other MD-associated

symptoms (eg, hearing loss, ear fullness, tinnitus) even though they are also quite vexing to patients.

The purpose of this CPG is to evaluate the many possible therapies for MD and to use evidence-based data from published literature to report on their efficacy in controlling MD symptoms, keeping in mind that MD may affect both ears in 10% to 25% of cases over time.[28] The only existing guideline to assist health care providers in the diagnosis and management of MD patients to date is a consensus statement that is >2 decades old. This updated CPG uses current evidence-based data and a multidisciplinary approach to improve timely, accurate MD diagnosis for optimal symptom control and patient outcomes. Key definitions used within this guideline can be found in **Table 1**.

Guideline Purpose

The primary purpose of this CPG is to improve the quality of the diagnostic workup and treatment outcomes of MD. To achieve this purpose, the goals of this CPG are to use the best available published scientific and/or clinical evidence to enhance diagnostic accuracy and appropriate therapeutic interventions (medical and surgical), while reducing unindicated diagnostic testing and/or imaging. The CPG is intended for all health care providers (eg, emergency medicine, primary care, otolaryngology, neurology, audiology, physical/vestibular therapy), in any setting, who are likely to encounter, diagnose, treat, and/or monitor patients with suspected MD. The target patient for the CPG is ≥18 years old with suspected diagnosis of definite or probable MD. The CPG makes specific recommendations about the history and physical examination of potential MD patients, the appropriate diagnostic workup, and effective treatment options that may include medical and/or surgical intervention. The CPG focuses only on MD, recognizing that MD may arise in conjunction with or separate from other conditions presenting with vertigo, hearing loss, and/or tinnitus. This CPG does not discuss the specific management of those conditions that may mimic MD. This CPG is not intended for comprehensive management of MD.

In 1995, the AAO-HNSF published a consensus statement on the diagnosis of MD.[2] These criteria were reviewed in 2015 by the Equilibrium Committee, yet >2 decades have elapsed since the original publication. Therefore, this current multidisciplinary group was convened to review the most recent and updated published scientific and clinical evidence available to craft an updated version of the MD consensus statement as a formal CPG. By using a published transparent CPG process, the primary goal was to create actionable statements (key action statements [KASs]) that reflect current evidence-based advances in knowledge with respect to MD.

Main considerations in this CPG are to increase rates of accurate diagnosis, improve symptom control with appropriate treatments, and reduce inappropriate use of medications, procedures, or testing. It is also intended to reduce adverse events associated with undiagnosed or untreated MD. Other CPG considerations include increasing patient-provider shared decision making, minimizing diagnostic and treatment costs, reducing unnecessary return physician visits, and maximizing the health-related QOL of individuals afflicted with MD. This CPG is also designed to clarify the term ''vertigo.'' Because many ''dizzy'' patients present with some form of subjective movement hallucination (eg, rocking side to side, listing, imbalance, light-headedness), it is the sensation of spinning that is characteristic of acute inner ear disorders and MD. Typically, among those who experience them, spinning attacks of vertigo with MD abate over time, and movement symptoms become vague. It is important to note that MD should have spinning vertigo at some point in its presentation. Currently, the public and the medical community in general have great confusion and disagreement about the term ''vertigo,'' and one goal of this CPG is to clarify that terminology as it relates to the diagnosis and management of MD.[29]

Health Care Burden

Epidemiology

Accurate estimation of the incidence and prevalence of MD has proved to be challenging, due to methodological limitations and the rarity of the condition. Prevalence estimates as low as 3.5 per 100,000 and as high as 513 per 100,000 have been reported from studies worldwide.[30] These estimates may reflect geographic and demographic variation but are also likely influenced by differences in case definitions over time (eg, 1972 American Academy of Ophthalmology and Otolaryngology criteria[3] vs 1995 AAO-HNSF criteria[2]), settings (hospital vs outpatient), duration, and methods of case capture (survey, records, or insurance claims).[31] One of the most rigorous studies involved reviewing the health records of 103,797 inhabitants of an Italian community between 1973 and 1985.[32] Using the 1972 guidelines of the American Academy of Ophthalmology and Otolaryngology,[3] the authors arrived at an incidence of 8.2 per 100,000, from which they calculated a prevalence of 205 of 100,000. The largest cohort assessed was drawn from insurance claims from 60 million commercially insured Americans, yielding an estimated prevalence of 190 per 100,000.[30] Thus far, no epidemiologic study has employed the most recent Barany Society diagnostic criteria.[5]

MD is almost exclusively reported in adults, with <3% of cases estimated to occur at age <18 years.[33-36] The disease is most prevalent between 40 to 60 years, with peak onset in the 40 to 50s.[37-42] In a large US claims–based study, the prevalence increased with age, ranging from 61 per 100,000 in patients 18 to 34 years old to 440 per 100,000 for patients aged >65 years.[30] Despite differences, most studies cite either an equal prevalence between males and females or a slightly higher prevalence of MD in women than in men,[14,35,38,41,42] with a reported female:male ratio in the United States of 1.89:1.[30] Data on the prevalence of bilateral MD yield variable estimates. Simultaneous presentation with bilateral MD appears to be exceptionally rare, whereas bilateral involvement may affect a significant number of patients within 2 decades of disease onset.[43] In

many MD patients, the most detrimental decline in hearing and balance function occurs within the first decade of diagnosis,[43] yet patients continue to have long-standing deficits that render MD a chronic disease.[44]

Impairments

MD is associated with substantial functional disability, although the level of handicap varies across individuals.[45] As the clinical diagnostic criteria state, most patients with MD have some level of hearing loss, tinnitus, ear fullness, or balance disturbance, with nearly one-third afflicted by severe symptoms in one of these categories.[46] Sensory loss and unpredictable episodic attacks often further restrict participation during work, domestic, and leisure activities.[47,48] While most patients are able to perform activities of daily living between attacks, during acute MD episodes they are likely to become entirely or partially dependent on the assistance of others.[45] Individuals with MD are also at increased risk of falling. Among the UK Biobank sample (n = 1376), MD patients were more than twice as likely to have experienced $\geq$2 falls in a year (13.7% vs 6.6%, $P < .001$).[39] Major injuries, including hip fractures, occur more frequently when falls are experienced by individuals with vertigo than by those without and may result in nursing home placement and further loss of independence.[49,50]

Quality of Life

Based on validated metrics, overall QOL of MD patients appears to be similar to that of patients with other chronic illnesses.[51,52] As they face a chronic battle with fluctuating balance and auditory dysfunction, MD patients also experience a heavy emotional burden. Health-related QOL has been assessed in patients with MD by the SF-36 (Short Form–36), a validated instrument that consists of 8 subscales that reflect different aspects of QOL (eg, general and mental health, physical functioning, role limitations) and 2 summary scores for physical and mental components of QOL.[53] On the SF-36, MD ranks closer to minor medical problems in physical handicap scores but closer to major medical problems in emotional handicap.[46] Vertigo is more closely associated with the physical aspects of QOL instruments, whereas hearing loss and tinnitus have greater impact on psychological aspects.[54] When the intrusiveness of chronic conditions is compared, MD ranks higher than end-stage renal disease and laryngeal cancer.[55] Notably, during acute MD attacks, ratings of the quality of well-being fall between those of noninstitutionalized patients with Alzheimer's disease and patients with end-stage cancer or AIDS, making acute MD attacks one of the most debilitating conditions that do not require institutionalization.[51] As such, anxiety and depression are common in MD patients,[56] with 33% of men and 41% of women affected with MD carrying diagnoses of depression.[55]

Health Care Costs

The diagnosis and management of MD produces significant direct health care costs. The symptom of dizziness is one of the most common reasons for ambulatory care visits in the United States and often leads to high utilization of diagnostic services (ie, imaging, audiovestibular, and cardiac testing) as well as consultation with various clinical specialists.[57,58] In one series, patients had undergone a mean 3.2 diagnostic tests, including magnetic resonance imaging (MRI; 78%), computed tomography (CT) or x-rays (52%), electro- or videonystagmography (64%), electrocardiography (51%), and electroencephalography (36%), before receiving the diagnosis of MD.[59] Some patients with classic MD symptoms experience lengthy diagnostic delays, potentially driving greater health care utilization. In a Finnish sample, 20% of patients experienced a delay in MD diagnosis of $\geq$5 years following the onset of hearing loss and vertigo.[37] Additional costs are incurred if patients first receive an incorrect diagnosis.

As it is a chronic clinical condition with occasional acute episodes, MD patients require health care resources for decades, including additional clinical encounters and devices for auditory rehabilitation.[60] Patients in the UK Ménière's Society reported needing $\geq$5 visits to their general practitioners per year.[60] Among practices in the US-based CHEER network (Creating Healthcare Excellence through Education and Research), MD patients had an average of 3.2 otolaryngology clinic visits per year, with intratympanic injections of steroids or gentamicin being the most common procedure performed (90%), followed by endolymphatic sac decompression (8%), transmastoid labyrinthectomy (2%), and vestibular nerve section (0.4%).[61] Thus far, one study in the United Kingdom has characterized the economic burden of MD, and the total direct costs were estimated to be £61.3 million (US $81.1 million) annually.[60] Similar analyses have not been carried out in the United States.

Indirect Costs

The direct costs of MD are surpassed by the indirect costs estimated to result from reductions in work productivity, increased sick leave, and lost earnings.[60] Patients report that work performance is most affected by vertigo, followed by hearing loss and the unpredictability of acute MD attacks.[45] Among patients presenting to a US academic medical center, 86% reported that their job performance had suffered as a result of their symptoms, 70% had to modify their jobs to be able to perform them, and 35% changed jobs.[45] Similarly, in the 3 months prior to presenting for care in clinics in Europe, Asia, and Africa, 70% of patients with MD lost working days, 72% required a reduced workload, 9% changed jobs, and 9% quit their jobs altogether.[62] Consequently, patients with MD have lower average household incomes and are more likely to receive disability benefits.[60,63] The long-term financial effects may be particularly severe, as the disease typically strikes during work-productive midlife. The annual cost of lost earnings from MD in the United Kingdom was estimated at £442.7 million (US $585.9 million). Altogether, indirect costs composed 88% of the total cost estimate for MD. Notably, the per-person average total annual cost was estimated to be

between £3341 (US $4421.65) and £3757 (US $4972.21), which is greater than estimates for asthma and migraine.[60]

Methods

General Methods

This CPG was developed with an explicit and transparent a priori protocol for creating actionable statements (KASs) based on supporting evidence and the associated balance of benefit and harm as outlined in the ''Clinical Practice Guideline Development Manual, Third Edition: A Quality-Driven Approach for Translating Evidence into Action.''[64] The Guideline Development Group (GDG) consisted of 21 panel members representing experts in advanced practice nursing, audiology, consumer advocacy, emergency medicine, family medicine, otolaryngology, otology and neurotology, otolaryngic allergy, neuroradiology, and neurology.

Literature Search

An information specialist conducted 2 systematic literature searches using a validated filter strategy to identify CPGs, systematic reviews (SRs), and RCTs. The following search terms were used:

> ''meniere disease''[MeSH Terms] OR meniere*[tiab] OR ''endolymphatic hydrops''[MeSH Terms] OR (endolymphatic[tiab] AND hydrops[tiab]) OR (cochle*[tiab] AND hydrops[tiab]) OR (vestibular[tiab] AND hydrops[tiab]) OR (morbus[tiab] AND meniere*[tiab]) OR tumarkin[tiab] OR (Vestibulocochlear[tiab] AND hydrops[tiab]) OR ''drop attack''[tiab] OR ''episodic vertigo''[tiab] OR ''periodic vertigo''[tiab] OR ''fluctuating vertigo''[tiab].

The English language searches were performed from February to March 2018 in multiple databases, including PubMed (MEDLINE), Excerpta Medica database (Embase), Cumulative Index to Nursing and Allied Health, Cochrane Central Register of Controlled Trials, National Guideline Clearinghouse, National Institutes for Health and Care Excellence (United Kingdom), SIGN (Scotland), New Zealand Guidelines Group, Australian National Health and Medical Research Council, TRIP Database, Guideline International Network, Canadian Medical Association Database, NHS Evidence (United Kingdom), Australian National Health and Medical Research Council, Guideline Internal Network, Cochrane Database of Systematic Reviews, Web of Science, the Allied and Complementary Medicine Database, CAB Abstracts, Agency for Healthcare Research and Quality, and Health Services/Technology Assessment Texts.

1. The initial search for CPGs identified 64 guidelines. After removal of duplicates and references that did not meet the inclusion criteria, a total of 18 guidelines were distributed to the panel for review. Quality criteria for including guidelines were (a) an explicit scope and purpose, (b) multidisciplinary stakeholder involvement, (c) systematic literature review, (d) explicit system for ranking evidence, and (e) explicit system for linking evidence to recommendations. The final data set retained 6 guidelines that met inclusion criteria.
2. The initial search for SRs identified 424 SRs or meta-analyses. After removal of duplicates and irrelevant references, a total of 96 SRs were distributed to the panel for review. Quality criteria for including reviews were (a) relevance to the guideline topic, (b) clear objective and methodology, (c) explicit search strategy, and (d) valid data extraction methods.[64] The final data set retained was 55 SRs or meta-analyses that met inclusion criteria.
3. The initial search for RCTs identified 558 RCTs. After removal of duplicates and irrelevant references, a total of 77 RCTs were distributed to the panel for review. Quality criteria for including RCTs were (a) relevance to the guideline topic, (b) publication in a peer-reviewed journal, and (c) clear methodology with randomized allocation to treatment groups. The total final data set retained 27 RCTs that met inclusion criteria.

In a series of conference calls, the GDG defined the scope and objectives of the proposed guideline. During the 18 months devoted to guideline development, the GDG met twice, with in-person meetings following the format previously described.[64] Electronic decision support software (BRIDGE-Wiz; Yale Center for Medical Informatics, New Haven, Connecticut) was used to facilitate creating actionable recommendations and evidence profiles.[65] Internal electronic review and feedback on each guideline draft were used to ensure accuracy of content and consistency with standardized criteria for reporting CPGs.[66]

AAO-HNSF staff used the Guideline Implementability Appraisal and Extractor to appraise adherence of the draft guideline to methodological standards, to improve clarity of recommendations, and to predict potential obstacles to implementation.[66] Guideline panel members received summary appraisals and modified an advanced draft of the guideline based on the appraisal. The final draft of the CPG was revised per the comments received during multidisciplinary peer review, open public comment, and journal editorial peer review. A scheduled review process will occur at 5 years from publication or sooner if new compelling evidence warrants earlier consideration.

Classification of Evidence-Based Statements

Guidelines are intended to reduce inappropriate variations in clinical care, to produce optimal health outcomes for patients, and to minimize harm. The evidence-based approach to guideline development requires that the evidence supporting a policy be identified, appraised, and summarized and that an explicit link between evidence and statements be defined. Evidence-based statements reflect

Table 2. Strength of Action Terms in Guideline Statements and Implied Levels of Obligation.[a]

Strength	Definition	Implied Obligation
Strong recommendation	A strong recommendation means the benefits of the recommended approach clearly exceed the harms (or, in the case of a strong negative recommendation, that the harms clearly exceed the benefits) and that the quality of the supporting evidence is high (grade A or B).[b] In some clearly identified circumstances, strong recommendations may be based on lesser evidence when high-quality evidence is impossible to obtain, and the anticipated benefits strongly outweigh the harms.	Clinicians should follow a strong recommendation unless a clear and compelling rationale for an alternative approach is present.
Recommendation	A recommendation means that the benefits exceed the harms (or, in the case of a negative recommendation, that the harms exceed the benefits), but the quality of evidence is not as high (grade B or C).[b] In some clearly identified circumstances, recommendations may be based on lesser evidence when high-quality evidence is impossible to obtain, and the anticipated benefits outweigh the harms.	Clinicians should also generally follow a recommendation but should remain alert to new information and sensitive to patient preferences.
Option	An option means that either the quality of evidence is suspect (grade D)[b] or well-done studies (grade A, B, or C)[b] show little clear advantage to one approach versus another.	Clinicians should be flexible in their decision making regarding appropriate practice, although they may set bounds on alternatives; patient preference should have a substantial influencing role.

[a]Adapted from American Academy of Pediatrics classification scheme.[73]
[b]See Table 3 for definitions of evidence grades.

both the *quality of evidence* and the *balance of benefit and harm* that is anticipated when the statement is followed. The definitions for evidence-based statements are listed in **Tables 2** and **3**.

Guidelines are never intended to supersede professional judgment; rather, they may be viewed as a relative constraint on individual clinician discretion in a specific clinical circumstance. Less frequent practice variation is expected for a strong recommendation than what might be expected with a recommendation. Options offer the most opportunity for practice variability.[67] Clinicians should always act and decide in a way that they believe will best serve their individual patients' interests and needs, regardless of guideline recommendations. Guidelines represent the best judgment of a team of experienced clinicians and methodologists addressing the scientific evidence for a specific topic.[68]

Making recommendations about health practices involves value judgments on the desirability of various outcomes associated with management options. Values applied by the GDG sought to minimize harm and diminish unnecessary and inappropriate therapy. A major goal of the panel was to be transparent and explicit about how values were applied and to document the process.

Financial Disclosure and Conflicts of Interest

The cost of developing this CPG, including travel expenses of all panel members, was covered in full by the AAO-HNSF. Potential conflicts of interest for all panel members in the past 5 years were compiled and distributed before the first conference call and were updated at each subsequent call and in-person meeting. After review and discussion of these disclosures,[69] the panel concluded that individuals with potential conflicts could remain on the panel if they (1) reminded the panel of potential conflicts before any related discussion, (2) recused themselves from a related discussion if asked by the panel, and (3) agreed not to discuss any aspect of the CPG with industry before publication. Last, panelists were reminded that conflicts of interest extend beyond financial relationships and may include personal experiences, how a participant earns a living, and the participant's previously established ''stake'' in an issue.[70]

Guideline Key Action Statements

Each evidence-based statement is organized in a similar fashion: a KAS in bold, followed by the strength of the recommendation in italics. Each KAS is followed by an ''action statement profile'' that explicitly states the quality improvement opportunity, aggregate evidence quality, level of confidence in evidence (high, medium, low), benefit, harms, risks, costs, and a benefits-harm assessment. Additionally, there are statements of any value judgments, the role of patient preferences, clarification of any intentional vagueness by the panel, exceptions to the statement, any differences of opinion, and a repeat statement of the strength of the recommendation. Several paragraphs subsequently discuss the published evidence supporting the statement. An overview of each evidence-based KAS in this guideline can be found in **Table 4**; for an algorithm based on the CPG and KASs, see **Figure 1**.

Table 3. Aggregate Grades of Evidence by Question Type.[a]

Grade	CEBM Level	Treatment	Harm	Diagnosis	Prognosis
A	1	Systematic review[b] of randomized trials	Systematic review[b] of randomized trials, nested case-control studies, or observational studies with dramatic effect	Systematic review[b] of cross-sectional studies with consistently applied reference standard and blinding	Systematic review[b] of inception cohort studies[c]
B	2	Randomized trials or observational studies with dramatic effects or highly consistent evidence	Randomized trials or observational studies with dramatic effects or highly consistent evidence	Cross-sectional studies with consistently applied reference standard and blinding	Inception cohort studies[c]
C	3-4	Nonrandomized or historically controlled studies, including case-control and observational studies	Nonrandomized controlled cohort or follow-up study (postmarketing surveillance) with sufficient numbers to rule out a common harm; case series, case-control, or historically controlled studies	Nonconsecutive studies; case-control studies; or studies with poor, nonindependent, or inconsistently applied reference standards	Cohort study, control arm of a randomized trial, case series, or case-control studies; poor-quality prognostic cohort study
D	5	Case reports, mechanism-based reasoning, or reasoning from first principles			
X	N/A	Exceptional situations where validating studies cannot be performed and there is a clear preponderance of benefit over harm			

Abbreviation: CEBM, Oxford Centre for Evidence-Based Medicine.

[a]Adapted from Howick and colleagues (Oxford Centre for Evidence-Based Medicine Work Group).[74]

[b]A systematic review may be downgraded to level B because of study limitations, heterogeneity, or imprecision.

[c]A group of individuals identified for subsequent study at an early, uniform point in the course of the specified health condition or before the condition develops.

Table 4. Summary of Guideline Key Action Statements.

Statement	Action	Strength
Statement 1. Diagnosis of Ménière's disease	Clinicians should diagnose definite or probable Ménière's disease in patients presenting with 2 or more episodes of vertigo lasting 20 minutes to 12 hours (definite) or up to 24 hours (probable) and fluctuating or nonfluctuating sensorineural hearing loss, tinnitus, or pressure in the affected ear, when these symptoms are not better accounted for by another disorder.	Recommendation
Statement 2. Assessing for vestibular migraine	Clinicians should determine if patients meet diagnostic criteria for vestibular migraine when assessing for Ménière's disease.	Recommendation
Statement 3. Audiometric testing	Clinicians should obtain an audiogram when assessing a patient for the diagnosis of Ménière's disease.	Strong recommendation
Statement 4. Utility of imaging	Clinicians may offer magnetic resonance imaging of the internal auditory canal and posterior fossa in patients with possible Ménière's disease and audiometrically verified asymmetric sensorineural hearing loss.	Option
Statement 5. Vestibular or electrophysiologic testing	Clinicians should not routinely order vestibular function testing or electrocochleography to establish the diagnosis of Ménière's disease.	Recommendation against
Statement 6. Patient education	Clinicians should educate patients with Ménière's disease about the natural history, measures for symptom control, treatment options, and outcomes.	Recommendation
Statement 7. Symptomatic management of vertigo	Clinicians should offer a limited course of vestibular suppressants to patients with Ménière's disease for management of vertigo only during Ménière's disease attacks.	Recommendation
Statement 8. Symptom reduction and prevention	Clinicians should educate patients with Ménière's disease on dietary and lifestyle modifications that may reduce or prevent symptoms.	Recommendation
Statement 9. Oral pharmacotherapy for maintenance	Clinicians may offer diuretics and/or betahistine for maintenance therapy to reduce symptoms or prevent Ménière's disease attacks.	Option
Statement 10. Positive pressure therapy	Clinicians should not prescribe positive pressure therapy for patients with Ménière's disease.	Recommendation against
Statement 11. Intratympanic steroid therapy	Clinicians may offer, or refer to a clinician who can offer, intratympanic steroids to patients with active Ménière's disease not responsive to noninvasive treatment.	Option
Statement 12. Intratympanic gentamicin therapy	Clinicians should offer, or refer to a clinician who can offer, intratympanic gentamicin to patients with active Ménière's disease not responsive to nonablative therapy.	Recommendation
Statement 13. Surgical ablative therapy	Clinicians may offer, or refer to a clinician who may offer, labyrinthectomy in patients with active Ménière's disease who have failed less definitive therapy and have nonusable hearing.	Recommendation
Statement 14a. Role of vestibular therapy for chronic imbalance	Clinicians should offer vestibular rehabilitation/physical therapy for Ménière's disease patients with chronic imbalance.	Recommendation
Statement 14b. Role of vestibular therapy for acute vertigo	Clinicians should not recommend vestibular rehabilitation/physical therapy for managing acute vertigo attacks in patients with Ménière's disease.	Recommendation against
Statement 15. Counseling for amplification and hearing assistive technology	Clinicians should counsel patients, or refer to a clinician who can counsel patients, with Ménière's disease and hearing loss on the use of amplification and hearing assistive technology.	Recommendation
Statement 16. Patient outcomes	Clinicians should document resolution, improvement, or worsening of vertigo, tinnitus, and hearing loss and any change in quality of life in patients with Ménière's disease after treatment.	Recommendation

The role of patient preferences in making decisions deserves further clarification. For some statements, where the evidence base demonstrates clear benefit, the role of patient preference for a range of treatments may be less relevant (as with intraoperative decision making). Clinicians should provide patients with clear and comprehensible information on the benefits to facilitate patient understanding and shared decision making, which in turn leads to better patient adherence and outcomes. In cases where the supporting evidence is weak or the benefits are unclear, shared decision making employing a collaborative effort between the clinician and an informed patient is extremely useful. Factors related to patient preference include, but are not limited to, absolute benefits (number needed to treat), adverse effects (number needed to harm), cost of drugs or procedures, and frequency and duration of treatment, as well as less tangible personal factors (eg, religious and/or cultural beliefs or personal levels of desire for intervention).

STATEMENT 1. DIAGNOSIS OF MENIÈRE'S DISEASE: Clinicians should diagnose definite or probable Ménière's disease in patients presenting with 2 or more episodes of vertigo lasting 20 minutes to 12 hours (definite) or up to 24 hours (probable) and fluctuating or nonfluctuating sensorineural hearing loss, tinnitus, or pressure in the affected ear, when these symptoms are not better accounted for by another disorder. *Recommendation based on observational studies with consistently applied reference standard and a preponderance of benefit over harms.*

Action Statement Profile: 1

- Quality improvement opportunity: Improving accuracy of diagnosis and increasing awareness of proper diagnosis for MD. National Quality Strategy domain: Effective Communication and Care Coordination
- Aggregate evidence quality: Grade C, based on observational studies with consistently applied reference standard
- Level of confidence in evidence: High
- Benefits: Improved accuracy and efficiency of diagnosis, appropriately directed treatment, reduced misdiagnosis, appropriately directed diagnostic testing, educating clinicians about accurate diagnosis, appropriate referrals, reduced use of inappropriate testing, reduced cost, improved patient QOL
- Risk, harm, cost: Provider time for making diagnosis
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: The group preferred to be more inclusive in the initial clinical diagnosis to capture more patients who prove to have MD, with the understanding that some patients with other diagnoses may initially be included.
- Intentional vagueness: Use of *definite* versus *probable*. Also, the presence of a documented/audiometrically objectified hearing loss may not be present at the time of testing.
- Role of patient preferences: small
- Exclusions: None
- Policy level: Recommendation
- Differences of opinion: There was disagreement among the panel regarding whether to include fluctuation as part of the criteria.

STATEMENT 2. ASSESSING FOR VESTIBULAR MIGRAINE: Clinicians should determine if patients meet diagnostic criteria for vestibular migraine when assessing for Ménière's disease. *Recommendation based on nonconsecutive studies, case-control studies, or studies with poor, nonindependent, or inconsistently applied reference standards with a preponderance of benefit over harm.*

Action Statement Profile: 2

- Quality improvement opportunity: Vestibular migraine is a common cause of dizziness that can closely mimic MD. Appropriate assessment for vestibular migraine could lead to more appropriate treatment. National Quality Strategy domains: Prevention and Treatment of Leading Causes of Morbidity and Mortality, Effective Communication and Care Coordination
- Aggregate evidence quality: Grade C, based on case-control studies or studies with poor, nonindependent, or inconsistently applied reference standards
- Level of confidence in evidence: Low, studies were done in specialty populations and may not be generalizable to more primary care populations
- Benefits: Accuracy of diagnosis, avoid unnecessary treatments or testing, potential for more appropriate treatment, patient education, promotes multidisciplinary care
- Risk, harm, cost: Extra time for assessment. Referral to other specialists.
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: None
- Role of patient preferences: Small
- Exclusions: None
- Policy level: Recommendation
- Differences of opinion: None

STATEMENT 3. AUDIOMETRIC TESTING: Clinicians should obtain an audiogram when assessing a patient for the diagnosis of Ménière's disease. *Strong recommendation based on SRs of cross-sectional studies with consistently applied reference standard and blinding for diagnostic testing with a preponderance of benefit over harms.*

Action Statement Profile: 3

- Quality improvement opportunity: Determining both pure tone thresholds and measures of speech recognition will lead to more accurate diagnosis and appropriate and timely referrals for aural

rehabilitation, hearing aids, and/or cochlear implants and may have significant implications for treatment options. National Quality Strategy domain: Effective Communication and Care Coordination
- Aggregate evidence quality: Grade A, based on SRs of cross-sectional studies with consistently applied reference standard and blinding for diagnostic testing
- Level of confidence in evidence: High
- Benefits: Improving diagnostic accuracy, identifying deficits in contralateral ear (question of bilateral disease), improving treatment planning, establishing baseline of hearing prior to treatment, directing treatment options based on degree of residual hearing (ablative vs nonablative), and identifying opportunities for aural rehabilitation
- Risk, harm, cost: Cost of testing, time of testing, patient distress at unrecognized hearing loss, discrimination based on hearing impairment (vocation, access to disability benefits)
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: An audiogram is essential to make the diagnosis of definite MD.
- Intentional vagueness: The presence of documented/audiometrically objectified hearing loss may not be present at the time of testing.
- Role of patient preferences: Small. Some patients may elect not to get an audiogram for various reasons.
- Exclusions: None
- Policy level: Strong recommendation
- Differences of opinion: There was a minority of the group who felt that patients with probable MD can be treated without an audiogram, but the majority felt that the audiogram is key to confirming the diagnosis and all subsequent management. One committee member noted that there are no studies in MD that assess outcomes in those receiving an audiogram as compared with those who do not. The audiogram is required to move from a diagnosis of possible MD to definite MD. Some patients and providers may elect to proceed with noninvasive management without an audiogram.

STATEMENT 4. UTILITY OF IMAGING: Clinicians may offer magnetic resonance imaging (MRI) of the internal auditory canal and posterior fossa in patients with possible Ménière's disease and audiometrically verified asymmetric sensorineural hearing loss. *Option based on observational and case studies with a preponderance of benefit over harm.*

Action Statement Profile: 4

- Quality improvement opportunity: To reduce variations of care and unnecessary expense as well as potential adverse effects from radiation (if CT is used) and/or contrast (CT/MRI) exposure. National Quality Strategy domain: Making Quality Care More Affordable
- Aggregate evidence quality: Grade D, based on observational and case studies
- Level of confidence in evidence: Medium
- Benefits: Avoid unnecessary testing; minimize cost and adverse events; maximize the diagnostic yield of MRI when indicated; avoid radiation; patient reassurance
- Risk, harm, cost: Cost of the MRI scan, potential risks of contrast agents, potential for risk of injury in MRI scanner (eg, heating of metallic wires and implants or subsequent malfunction of implants with magnetic components), physical discomfort of the imaging procedure (noise, claustrophobia), psychological distress of incidental findings (and further workup necessitated by those findings), and potential for delayed/missed diagnosis[71]
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: None
- Role of patient preferences: Moderate
- Exclusions: Patients unable or unwilling to have MRI
- Policy level: Option
- Differences of opinion: The group was divided regarding the benefit of MRI. Specifically, many clinicians were uncomfortable treating MD without ruling out inner ear or retrocochlear lesions in either unilateral hearing loss or subsequent second-side loss in the setting of possible bilateral MD. Others felt comfortable using nonablative therapies without MRI.

STATEMENT 5. VESTIBULAR OR ELECTROPHYSIOLOGIC TESTING: Clinicians should not routinely order vestibular function testing or electrocochleography to establish the diagnosis of Ménière's disease. *Recommendation against based on systematic reviews of cross-sectional studies and observational electrocochleography studies.*

Action Statement Profile: 5

- Quality improvement opportunity: Avoidance of unnecessary testing. National Quality Strategy domains: Patient Safety, Prevention and Treatment of Leading Causes of Morbidity and Mortality
- Aggregate evidence quality: Grade B, based on SRs of cross-sectional studies and observational electrocochleography studies
- Level of confidence in evidence: Medium, based on difficulty in assessing the quality of the SRs, the meta-analyses, and the subgroups within the cohort

- Benefits: Avoidance of unnecessary testing, decreased cost, improved efficiency of diagnosis, reduced patient burden of unpleasant testing
- Risk, harm, cost: Missed or delayed diagnosis of comorbid conditions
- Benefit-harm assessment: Preponderance of benefit over harms
- Value judgments: While some of these tests may have a role in individualized patients, Ménière's disease requires a clinical and audiometric diagnosis.
- Intentional vagueness: The word *routine* is used to allow for individualized use of these testing modalities in some of the settings specified in the supporting text.
- Role of patient preferences: Small
- Exclusions: None
- Policy level: Recommendation against
- Differences of opinion: None

STATEMENT 6. PATIENT EDUCATION: Clinicians should educate patients with Ménière's disease about the natural history, measures for symptom control, treatment options, and outcomes. *Recommendation based on an RCT on patients educating themselves and shared decision-making literature and a preponderance of benefit over harm.*

Action Statement Profile: 6

- Quality improvement opportunity: Informing patients about their disease to participate in shared decision making. National Quality Strategy domain: Effective Communication and Care Coordination
- Aggregate evidence quality: Grade C, single RCT evaluating a patient education booklet and the considerable literature on shared decision making
- Level of confidence in evidence: High
- Benefits: Patient engagement, patient satisfaction, improved adherence to treatment, avoidance of unnecessary treatments, more optimal use of health care resources, improved symptom control, improved shared decision making
- Risk, harm, cost: Time for education, patient distress, diagnosis uncertainty
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: Education allows for improved shared decision making. This assumes that the patient is not already appropriately educated.
- Intentional vagueness: None
- Role of patient preferences: Small but patients may express preference for optimal method of education.
- Exclusions: None
- Policy level: Recommendation
- Differences of opinion: None

STATEMENT 7. SYMPTOMATIC MANAGEMENT OF VERTIGO: Clinicians should offer a limited course of vestibular suppressants to patients with Ménière's disease for management of vertigo only during Ménière's disease attacks. *Recommendation based on nonrandomized or historically controlled studies, including case-control and observational studies, and a preponderance of benefit over harm.*

Action Statement Profile: 7

- Quality improvement opportunity: Communication with clinicians and their patients about how and when to use vestibular suppressants to control vertigo. National Quality Strategy domains: Effective Communication and Care Coordination, Person and Family Centered Care
- Aggregate evidence quality: Grade C, nonrandomized or historically controlled studies, including case-control and observational studies
- Level of confidence in evidence: Medium due to grade C evidence.
- Benefits: Better symptom control, improved QOL
- Risk, harm, cost: Cost, side effects—urinary retention, dry mouth, visual changes, sedation, addiction. Impaired vestibular compensation
- Benefit-harm assessment: preponderance of benefit over harm
- Value judgments: Vertigo can have a detrimental impact on QOL, and patients tend to feel better when vertigo symptoms are alleviated.
- Intentional vagueness: None
- Role of patient preferences: Large depending on severity of symptoms
- Exclusions: None
- Policy level: Recommendation
- Differences of opinion: None

STATEMENT 8. SYMPTOM REDUCTION AND PREVENTION: Clinicians should educate patients with Ménière's disease on dietary and lifestyle modifications that may reduce or prevent symptoms. *Recommendation based on RCTs, observation studies, and cohort studies with indeterminate benefit, with a preponderance of benefit over harms.*

Action Statement Profile: 8

- Quality improvement opportunity: Identification of MD triggers may reduce symptoms in some patients. Allergies have been shown to contribute to symptoms of Ménière's disease in up to 30% of patients. National Quality Strategy domains: Effective Communication and Care Coordination, Person and Family Centered Care, Prevention and Treatment of Leading Causes of Morbidity and Mortality

- Aggregate evidence quality: Grade C, based on a dearth of RCTs regarding dietary modifications (1 small RCT on sodium restriction, negative for effectiveness but with study limitations; 1 relatively strong observational/survey study showing advantage to both low sodium and caffeine restriction), 1 RCT regarding decreasing stress hormone vasopressin and 1 RCT of booklet-based symptom control through relaxation and cognitive-behavior strategies to reduce anxiety, 1 RCT regarding an acupressure technique for treatment of dizziness and 2 SRs regarding acupuncture, 3 RCTs regarding antisecretory therapy (2 positive, 1 negative for effectiveness), and a number of observational studies and a strong literature review (human and animal) regarding the role of treatment of allergy symptoms in reducing symptoms of MD in allergic patients. There is a Cochrane SR currently underway for dietary modifications.
- Level of confidence in evidence: High.
- Benefits: May improve symptom control, avoid unnecessary lifestyle modifications, improved QOL, patient empowerment, potential avoidance of more invasive/higher-risk therapy
- Risk, harm, cost: Time of counseling, burden of potentially ineffective lifestyle modifications on the patient/family, potential risk of hyponatremia, increased cost of Ménière's diet
- Benefit-harm assessment: Preponderance of benefit over harms
- Value judgments: While the evidence of benefit of dietary and lifestyle modifications is limited, individual patients may have identifiable triggers, the identification of which may improve symptom control.
- Intentional vagueness: None
- Role of patient preferences: Small regarding the provision of education but large with regard to the choice to adopt lifestyle or dietary changes or not
- Exclusions: None
- Policy level: Recommendation
- Differences of opinion: A small group of panel members felt that there was a limited role and expressed concern regarding possible negative effects of sodium restriction, specifically hyponatremia, although this has not been reported in any of the studies and could be minimized as a risk with use of appropriate nutritional counseling.

STATEMENT 9. ORAL PHARMACOTHERAPY FOR MAINTENANCE: Clinicians may offer diuretics and/or betahistine for maintenance therapy to reduce symptoms or prevent Ménière's disease attacks. *Option based on observational studies and a Cochrane review on betahistine and oral diuretics with a balance of benefits and harms.*

Action Statement Profile: 9

- Quality improvement opportunity: Improved symptom control. National Quality Strategy domains: Prevention and Treatment of Leading Causes of Morbidity and Mortality, Person and Family Centered Care
- Aggregate evidence quality: Grade B, based on observational studies and a Cochrane review on betahistine and oral diuretics
- Level of confidence in evidence: Medium. High risk of bias reported in most studies included in SR
- Benefits: Improved vertigo control, improved QOL
- Risk, harm, cost: Cost of therapy, side effects of medications, promotion of ineffective therapy
- Benefit-harm assessment: Balance of benefits and harm
- Value judgments: There are different practice patterns among treating physicians on the panel. There is no specific preference for one agent over another, and that is why they were grouped for this statement.
- Intentional vagueness: None
- Role of patient preferences: Large
- Exclusions: Patients with comorbid conditions making these medications contraindicated (ie, renal or cardiac disease, asthma). Allergies or sensitivities to these medications
- Policy level: Option
- Differences of opinion: None

STATEMENT 10. POSITIVE PRESSURE THERAPY: Clinicians should not prescribe positive pressure therapy to patients with Ménière's disease. *Recommendation against based on a systematic review and randomized trials showing ineffectiveness of devices like the Meniett devices, with a preponderance of benefit over harm for not using.*

Action Statement Profile: 10

- Quality improvement opportunity: Avoidance of ineffective therapy. National Quality Strategy domain: Prevention and Treatment of Leading Causes of Morbidity and Mortality
- Aggregate evidence quality: Grade B, based on a Cochrane SR and 2 small RCTs on Meniett device showing no effect
- Level of confidence in evidence: High
- Benefits: Avoidance of ineffective therapy
- Risk, harm, cost: Patient or physician concerns at the lack of positive pressure therapy as an option if other noninvasive treatments have failed, with remaining options being destructive and/or invasive procedures
- Benefit-harm assessment: Preponderance of benefit over harms

- Value judgments: While this therapy is generally ineffective, there may be rare patients with limited other options.
- Intentional vagueness: None
- Role of patient preferences: Small
- Exclusions: None
- Policy level: Recommendation against
- Differences of opinion: A small group of panel members felt that some evidence supports the use of the Meniett device and that it could be used in symptomatic patients who have not obtained relief from other nonablative treatments.

STATEMENT 11. INTRATYMPANIC STEROID THERAPY: Clinicians may offer, or refer to a clinician who can offer, intratympanic steroids to patients with active Ménière's disease not responsive to noninvasive treatment. *Option based on a systematic review and a randomized controlled trial with a preponderance of benefit over harm.*

Action Statement Profile: 11

- Quality improvement opportunity: Improved vertigo control. National Quality Strategy domains: Effective Communication and Care Coordination, Prevention and Treatment of Leading Causes of Morbidity and Mortality, Person and Family Centered Care
- Aggregate evidence quality: Grade C, based on 1 Cochrane review that concluded limited efficacy for disability score and 1 small RCT looking at dexamethasone and gentamicin with 90% symptom reduction
- Level of confidence in evidence: High
- Benefits: Improved vertigo control, no risk of hearing loss, less risk of systemic side effects, improved QOL (dizziness handicap), no loss of vestibular function (nonablative therapy)
- Risk, harm, cost: Cost, perforation, possible need for multiple injections, infection, discomfort of the procedure, time for treatment
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: While this is less definitive than gentamicin therapy, the favorable risk-benefit profile makes this a good option for patients.
- Intentional vagueness: The term *noninvasive* refers to medical therapy and lifestyle modification.
- Role of patient preferences: Medium
- Exclusions: None
- Policy level: Option
- Differences of opinion: There was some controversy regarding whether the aggregate evidence strength in favor of this intervention is a grade B or a grade C. Given this, a few panel members felt that this statement should be a recommendation rather than an option.

STATEMENT 12. INTRATYMPANIC GENTAMICIN THERAPY: Clinicians should offer, or refer to a clinician who can offer, intratympanic gentamicin to patients with active Ménière's disease not responsive to nonablative therapy. *Recommendation based on 2 randomized trials and several systematic reviews indicating efficacy in the treatment of vertigo with a preponderance of benefit over harm.*

Action Statement Profile: 12

- Quality improvement opportunity: Improved vertigo control. National Quality Strategy domains: Prevention and Treatment of Leading Causes of Morbidity and Mortality, Person and Family Centered Care
- Aggregate evidence quality: Grade B, based on 2 RCTs and several SRs indicating efficacy in the treatment of vertigo
- Level of confidence in evidence: High
- Benefits: Improved vertigo control, improved QOL, faster return to work, avoidance of general anesthetic, a risk of hearing loss (relative to surgical labyrinthectomy), improved safety
- Risk, harm, cost: Hearing loss, ear drum perforation, persistent imbalance, need for multiple treatments
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: None
- Intentional vagueness: The term *inadequate control* may vary for different patients.
- Role of patient preferences: Large regarding timing and when to initiate therapy
- Exclusions: Patients with contralateral disease or hypofunction. Patients with a known hypersensitivity to aminoglycosides
- Policy level: Recommendation
- Differences of opinion: None

STATEMENT 13. SURGICAL ABLATIVE THERAPY: Clinicians may offer, or refer to a clinician who may offer, labyrinthectomy in patients with active Ménière's disease who have failed less definitive therapy and have nonusable hearing. *Recommendation based on observation studies and case series with a preponderance of benefit over harm.*

Action Statement Profile: 13

- Quality improvement opportunity: Improve awareness of effective therapy. National Quality Strategy domains: Effective Communication and Care Coordination, Prevention and Treatment of Leading Causes of Morbidity and Mortality, Person and Family Centered Care

- Aggregate evidence quality: Grade C, based on observation studies and case series data that show efficacy
- Level of confidence in evidence: High
- Benefits: Definitive vertigo control, expedient treatment (single definitive treatment), ability to stop other less effective therapy (that may have side effects), control of drop attacks
- Risk, harm, cost: Risks of surgery, loss of residual hearing, need for general anesthetic, reduced therapy options in the event that the patient develops bilateral disease, poor compensation after surgery, active tinnitus
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: Labyrinthectomy represents a standard for control of active vertigo in MD.
- Intentional vagueness: Non-usable hearing is not specifically defined and may be determined in conjunction with the patient. Less definitive therapy is also vague as failed nerve section may be considered more invasive but may not have resolved symptoms.
- Role of patient preferences: Large opportunity for shared decision-making
- Exclusions: Bilateral disease or vestibular hypofunction in the other ear.
- Policy level: Recommendation
- Differences of opinion: A minority of panel members felt that offer was too strong a term but that a discussion about this intervention should be undertaken.

STATEMENT 14a. ROLE OF VESTIBULAR THERAPY FOR CHRONIC IMBALANCE: Interictal instability and following ablative therapy: Clinicians should offer vestibular rehabilitation/physical therapy for Ménière's disease patients with chronic imbalance. *Recommendation based on systematic reviews and limited RCTs with a preponderance of benefit over harm.*

Action Statement Profile: 14a

- Quality improvement opportunity: Offer therapy for patients who have chronic imbalance, bilateral MD, and/or following ablative therapy. Promoting effective therapy and increased patient safety. National Quality Strategy domains: Safety, Promoting Effective prevention/treatment
- Aggregate evidence quality: Grade A, based on SRs and limited RCTs
- Level of confidence in evidence: High
- Benefits: Improved symptom control, safety, reduced risk of falls, improved confidence, improved QOL
- Risk, harm, cost: Cost of therapy, time for appointments, potential exacerbation of acute symptoms
- Benefit-harm assessment: Preponderance of benefit over harm
- Value judgments: While ineffective acutely, vestibular rehabilitation therapy has a significant role in the chronic management of MD patients
- Intentional vagueness: Imbalance encompasses multiple varying scenarios, including vestibular dysfunction and chronic balance problems
- Role of patient preferences: Small; however, patients can have a larger role in deciding if they choose to do vestibular rehabilitation.
- Exclusions: Patients in the setting of an acute attack
- Policy level: Recommendation
- Differences of opinion: None

STATEMENT 14b. ROLE OF VESTIBULAR THERAPY FOR ACUTE VERTIGO: Clinicians should not recommend vestibular rehabilitation/physical therapy for managing acute vertigo attacks in patients with Ménière's disease. *Recommendation against based on RCTs studied that evaluated acute vertigo but were not specific to MD and a preponderance of benefit over harms.*

Action Statement Profile: 14b

- Quality improvement opportunity: Avoidance of inappropriate/ineffective therapy. National Quality Strategy domains: Patient Safety, Prevention and Treatment of Leading Causes of Morbidity and Mortality
- Aggregate evidence quality: Grade B, based on subset analysis of RCTs that failed to identify any studies on the topic, as well as expert opinion extrapolated from evidence from a CPG
- Level of confidence in evidence: Medium. The RCTs evaluated acute vertigo but were not specific to MD.
- Benefits: Avoidance of noneffective therapy, preserving coverage for physical therapy at a later stage of disease, avoidance of potential exacerbation of symptoms
- Risk, harm, cost: Delay of treatment in patients with an underlying vestibular hypofunction
- Benefit-harm assessment: Preponderance of benefit over harms
- Value judgments: Avoidance of inappropriate therapy
- Intentional vagueness: None
- Role of patient preferences: None
- Exclusions: None
- Policy level: Recommendation against
- Differences of opinion: None

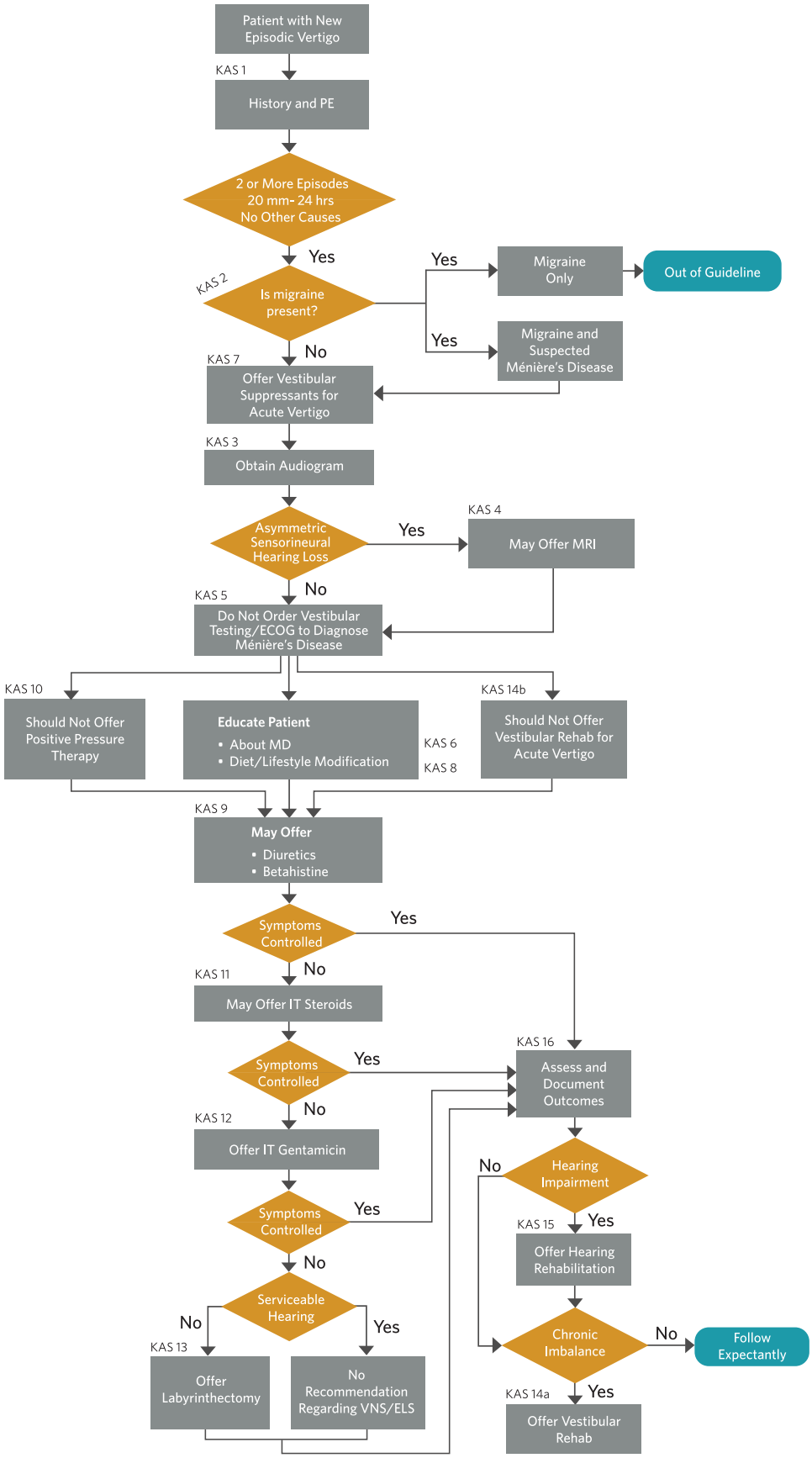

Figure 1. Clinical practice guideline: Ménière's disease algorithm. ECOG, electrocochleogram; ELS, endolymphatic sac; IT, intratympanic; KAS, key action statement; MD, Ménière's disease; MRI, magnetic resonance imaging; PE, physical examination; VNS, vestibular nerve section.

STATEMENT 15. COUNSELING FOR AMPLIFICATION AND HEARING ASSISTIVE TECHNOLOGY: Clinicians should counsel patients, or refer to a clinician who can counsel patients, with Ménière's disease and hearing loss on the use of amplification and hearing assistive technology. *Recommendation based on cohort studies of hearing outcomes in MD and benefits of amplification and cochlear implants with a preponderance of benefit over harms.*

Action Statement Profile: 15

- Quality improvement opportunity: Shared decision-making opportunities between patients and clinicians regarding MD and hearing loss and the use of amplification and other hearing assistive technologies. National Quality Strategy domains: Effective Communication and Care Coordination, Person and Family Centered Care
- Aggregate evidence quality: Grade C, based on cohort studies of hearing outcomes in MD and benefits of amplification and cochlear implants
- Level of confidence in evidence: High
- Benefits: Improved function, improved QOL, improved hearing, less missed work
- Risk, harm, cost: Clinicians and patients' time, creation of unrealistic expectations
- Benefit-harm assessment: Preponderance of benefit over harms
- Value judgments: The handicap of associated hearing loss is underrecognized in MD patients
- Intentional vagueness: None
- Role of patient preferences: Small regarding counseling; large in terms of choice to use these technologies
- Exclusions: None
- Policy level: Recommendation
- Differences of opinion: None

STATEMENT 16. PATIENT OUTCOMES: Clinicians should document resolution, improvement, or worsening of vertigo, tinnitus, and hearing loss and any change in quality of life in patients with Ménière's disease after treatment. *Recommendation based on the controlled arms of RCTs, outcomes from RCTs, cohort studies, and observational studies with a preponderance of benefit over harm.*

Action Statement Profile: 16

- Quality improvement opportunity: Tracking outcomes of therapy provides an opportunity for modification of management to optimize outcomes; to ensure that patients have follow-up until symptoms are under adequate control. National Quality Strategy domain: Effective Communication and Care Coordination
- Aggregate evidence quality: Grade C, based on the controlled arms of RCTs, outcomes from RCTs, cohort studies, and observational studies
- Level of confidence in evidence: Medium due to grade C evidence
- Benefits: Opportunity to adjust for more effective therapy, possibility of more accurate diagnosis, opportunity for hearing rehabilitation, patient engagement
- Risk, harm, cost: Cost and time of visits
- Benefit-harm assessment: Preponderance of benefit over harms
- Value judgments: Not applicable
- Intentional vagueness: The word *symptoms* can refer to vertigo, hearing loss, tinnitus, or pressure depending on what is of most concern to the patient
- Role of patient preferences: Medium. Some patients with subjectively adequate disease control may choose not to follow up.
- Exclusions: None
- Policy level: Recommendation
- Differences of opinion: Several group members wanted to document symptoms before, during, and after treatment, and others wanted to specifically document change in symptoms.

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Disclaimer

This clinical practice guideline is not intended as an exhaustive source of guidance for managing patients with MD. Rather, it is designed to assist clinicians by providing an evidence-based framework for decision-making strategies. The guideline is not intended to replace clinical judgment or establish a protocol for all individuals with this condition and may not provide the only appropriate approach to diagnosing and managing this program of care. As medical knowledge expands and technology advances, clinical indicators and guidelines are promoted as conditional and provisional proposals of what is recommended under specific conditions but are not absolute. Guidelines are not mandates. These do not and should not purport to be a legal standard of care. The responsible physician, based on all circumstances presented by the individual patient, must determine the appropriate treatment. Adherence to these guidelines will not ensure successful patient outcomes in every situation. The AAO-HNSF emphasizes that these clinical guidelines should not be deemed to include all proper treatment decisions or methods of care or to exclude other treatment decisions or methods of care reasonably directed to obtaining the same results.

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