

Ménière's disease

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Key-words. Guidelines; Ménière's disease; inner ear; treatment.

Abstract. *Ménière's disease.* Ménière's disease is an idiopathic inner ear disorder characterized by endolymphatic hydrops (ELH), vertigo attacks, sensorineural hearing loss, tinnitus and pressure sensation in the affected ear. The disorder has to be differentiated from known causes of ELH and from other disorders provoking similar symptoms. Ménière's disease can seriously affect quality of life, and medical support is often necessary on a life long basis. The treatment modalities are counselling, preventive measures and life style adaptation, drug therapy, tinnitus coping, provision of hearing devices, and sometimes surgery. We review the indications for these modalities, and the evidence level of their beneficial effect.

1. Definition

In Ménière's disease, also called idiopathic endolymphatic hydrops (ELH), the endolymphatic compartment of the inner ear expands.¹ The clinical picture comprises recurrent spontaneous episodes of rotatory vertigo, fluctuating sensorineural hearing loss (SNHL), and tinnitus with or without aural fullness on the affected side. Patients suffering from Ménière's disease typically experience the vertigo attacks as the most debilitating symptom due to its unpredictable nature. The vertigo attacks may last from several minutes to several days. During attacks, patients experience neurovegetative symptoms and are usually unable to pursue normal life. After an attack, patients are exhausted and may experience disequilibrium for days. During active disease, most patients have a seriously diminished quality of life and anxiety or depression

often develop. In a prospective study of 243 patients, Havia and Kentala found that the proportion of patients reporting severe or very severe attacks increased with the duration of the disease.²

Patients with long-standing Ménière's disease in which vertigo attacks ceased, also report reduced quality of life because of a significant sensorineural hearing loss and tinnitus.³ A limited number of patients affected with Ménière's disease exhibit sudden falls without loss of consciousness or associated vertigo: the so called 'otolithic crises of Tumarkin' or drop attacks.

Up to 25% of the patients with Ménière's disease may eventually require a surgical procedure to control the vertigo attacks.

2. Epidemiology

Throughout the world, the reported incidence and prevalence of Ménière's disease varies by a fac-

tor of 10. In the US, the prevalence is about 200 per 100.000 persons and the incidence about 15 per 100.000 persons per year. Prevalence estimates obtained from Japanese and Scandinavian populations yielded much lower figures: 20 and 45 per 100.000 persons, respectively. Ménière's disease typically starts between the age of 20 and 50 years with a diagnosis rate proportional to the age of 60. Men and women are probably equally affected. In about 1/2 to 2/3 of the cases, Ménière's disease affects only one ear. In bilateral cases, the second ear is mostly affected within five years after onset of the disease.⁴

3. Pathophysiology

Per definition, the cause of Ménière's disease is unknown. ELH is commonly believed as the result of an abnormal fluid balance in the inner ear that can be

caused by many factors. New insights into normal cell biology will reveal new pathophysiologic pathways leading to ELH.

Dunnebier *et al.*⁵ developed a guinea pig model for Ménière's disease by slight destruction of the endolymphatic sac and stimulation of the endolymph production with aldosterone. Different aspects of cochlear physiology in this animal model have been studied. Warmerdam *et al.*⁶ found an abnormal endocochlear potential but, surprisingly, no increased pressure in the scala media. The same research group found that the resistance for fluid flow through the cochlear aqueduct, which connects the scala tympani with the cerebrospinal fluid space, depends on the position of the round window membrane, which can be manipulated with the Meniett device and aquaporin involvement.⁶

3.1. Classification of ELH

ELH in temporal bones of patients with Ménière's disease was for the first time observed by Hallpike and Cairns in 1938.⁷ Ever since, ELH plays a central role in our hypotheses about the pathophysiology of Ménière's disease: a vertigo attack is believed to result from an acute volume increase of the endolymphatic compartment with rupturing of the membranous labyrinth and release of high amounts of K⁺ in the perilymphatic space. Repeated rupturing of membranes is believed to cause progressive destruction of the labyrinth with eventually a profound deafness and disappearance of the vertigo attacks.

Based on clinical case reports and temporal bone studies, Schuknecht and Gulya proposed a

classification of ELH according to aetiology. They distinguished embryopathic, idiopathic and acquired ELH. Embryopathic ELH results from a prenatal developmental disorder, and is associated with morphological anomalies like Mondini's malformation or a decreased space between the posterior semicircular canal and the subarachnoid space. Acquired ELH may occur after trauma of the labyrinth, infection, inflammation, auto-immune processes, and genetically mediated involution. ELH provoked by food substances or allergens could be classified as acquired.⁸

3.2. Endolymphatic Hydrops and Ménière's disease

ELH can explain a great deal of Ménière's disease, but many questions remain. An alternative view is that Ménière's disease reflects a neuroganglionitis and that ELH is merely an epiphenomenon.⁹

Improved imaging of the inner ear and fundamental research will be necessary to better understand Ménière's disease.

3.2.1. Evidence for ELH

Histological studies of temporal bones of patients with Ménière's disease provide strongest evidence for ELH. These studies consistently find that ELH is more pronounced in the cochlea and sacculus than in the utricle and semicircular canals.⁹⁻¹⁰

Other data in support of ELH is based on electrocochleography (ECoG). Patients with Ménière's disease often (50-70%) have a large summing potential that can be reduced after administration of hyperosmotic substances.¹¹ In animal models, blockage of the endolymphatic canal or foreign protein deposition in the peri-

lymph causes an ELH that is associated with hearing loss at low frequencies.

Morphological temporal bone features that limit extension of the endolymphatic sac or circulation of fluid are predisposing for ELH. These features include a decreased distance between the posterior semicircular canal and the subarachnoid space, a lack of visualisation of the endolymphatic duct on MRI, and an apical and middle cochlear segment displacement related to a reduced endolymphatic sac volume.¹²

3.2.2. Evidence against ELH

The absence of ELH in temporal bones of some Ménière's disease subjects, and the presence of ELH in some subjects who did not have Ménière's disease, place the significance of ELH for Ménière's disease in doubt.¹⁰ Histopathologic findings other than ELH are found in temporal bones of patients with Ménière's disease: perilymphatic fibrosis, loss of spiral ganglion cells innervating the apical cochlea, and axonal degenerations. ELH is present in animal models of Ménière's disease, but these animals do not exhibit vestibular symptoms, and perilymphatic fibrosis is not found in their temporal bones.⁹ In addition, few patients with Ménière's disease who received a cochlear implant, showed fluctuations of hearing performance. Moreover, hyperosmotic or diuretic agents, which are supposed to reduce hydrops, do not improve the subjective and objective symptoms in all patients with Ménière's disease.¹¹

3.3. Other pathophysiological factors

Many factors can trigger a crisis of Ménière's disease symptoms. It

has been suggested that food and air-born allergens can trigger a crisis, but up until now, corresponding antibodies have not been detected. For some patients intake of caffeine, alcohol, or chocolate increases the frequency of attacks. Sometimes, a middle ear infection triggers an attack, and even weather changes can trigger an attack.

Auto-immune disorders of the inner ear can provoke ELH with rapidly progressive bilateral symptoms that respond to immuno-suppressive treatment. Ménière's symptoms are observed in some genetic cochleovestibular disorders, especially DFNA9.¹³ Above all, psychosomatic factors play an important role in Ménière's disease: increased stress can trigger a crisis and disturbances in the personal or professional life are associated with a reactivation of the disease.

4. Diagnostic management

4.1. Diagnostic criteria for Ménière's disease (recommendation A)

The committee on Hearing and Equilibrium of the American Academy of Otolaryngology-Head and Neck Surgery suggested guidelines for reporting and evaluation of therapy in Ménière's disease (Committee AAO-HNS 1972, 1985, and 1995).¹ Nowadays, these guidelines are widely accepted and provide a solid framework that is applicable in the clinic. According to the guidelines, diagnosis of Ménière's disease is based on clinical symptoms and exclusion of identifiable "other causes". Four degrees of diagnostic certainty are defined: certain, definite, probable and possible Ménière's disease

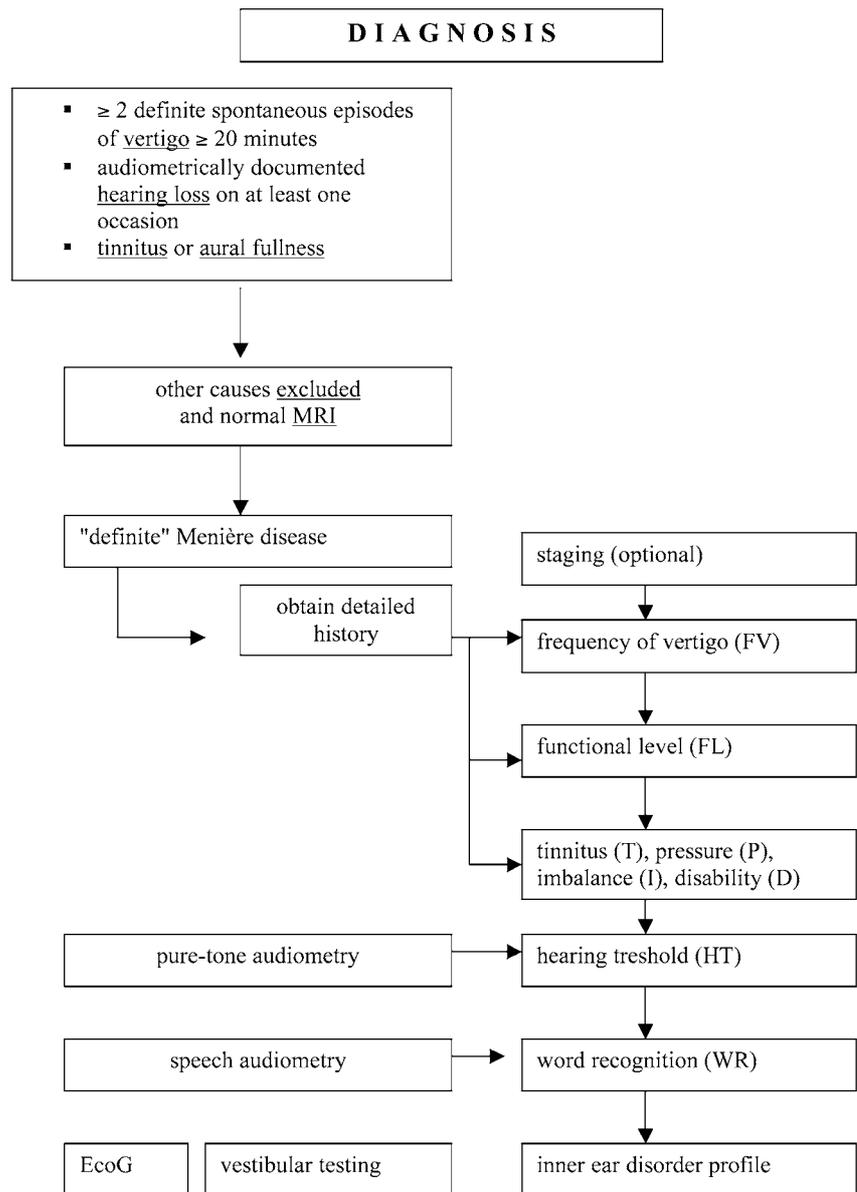


Figure 1 Diagnostic algorithm

(Table 1). Additional evidence for ELH can be provided with electrocochleography, auditory and vestibular testing.

Table 2 lists "other causes" of Ménière-like diseases that must be excluded using any diagnostic technique available (e.g. microscopic otoscopy, medical imaging including contrast enhanced MRI and high definition CT scan, virtu-

al endoscopy, hematological, serological, biochemical and genetic tests). Taking into account the numerous reports of pathological findings in Ménière's disease, it is the opinion of the authors that at least once during the course of the disease a MRI scan with gadolinium enhancement of the posterior fossa should be performed (recommendation B). Associated

Table 1
Diagnostic criteria of Ménière's disease

(Following Committee AAO-HNS 1995)
Certain Ménière's disease Definite criteria and histopathologic evidence of ELH (E.g., postmortem analysis).
Definite Ménière's disease ≥ 2 definite spontaneous episodes of vertigo lasting longer than 20 minutes. Audiometrically documented hearing loss on at least one occasion. Tinnitus or aural fullness.
Probable Ménière's disease Idem but only one occasion.
Possible Ménière's disease Episodic vertigo without documented hearing loss. Sensorineural hearing loss (fluctuating or fixed) with disequilibrium.
For all classes other causes excluded

Table 2
Differential diagnosis for Ménière's disease

Inner ear disorders
Congenital disorders
Semicircular canal dehiscence
Large Vestibular Aqueduct Syndrome
Infections
Bacterial and viral labyrinthitis
Vestibular neuritis
Inflammations
Auto-immune inner ear disorder
E.g. syphilis
Cogan syndrome
Benign Positional Paroxysmal Vertigo
Vascular
Genetic and metabolic
DFNA9
Otosclerosis
Post-traumatic and post-surgery
Perilymphatic fistula
Central Nervous System pathology
Cerebellopontine angle tumours
Vestibular Schwannoma
Petroclival meningioma
Endolymphatic sac tumours
Epidermoid cysts
Other tumours
Neurovascular conflict with N VIII
Demyelinating disorders
E.g., multiple sclerosis
Vertebrobasilar insufficiency
Migraine and vestibular migraine
Side effects of drugs and toxic pollutants

pathology needs to be assessed e.g. thyroid disease or auto-immune pathology.

It is good practice to challenge the diagnosis of Ménière's disease

repeatedly during the course of the disease. Differential diagnosis with migraine associated vertigo, auto-immune inner ear disorders, neuro-vascular conflict in the

cerebello-pontine angle and verte-bro-basilar insufficiency may be difficult.

4.2. Guidelines for a multidimensional description

4.2.1. Basic reporting (recommendation A)

The standard guidelines of the AAO-HNS imply a multidimensional description of Ménière's disease. The basic elements are pure-tone sensitivity, word recognition, number of vertigo attacks lasting at least 20 min, and functional level rating. Researchers are of course free to quantify other aspects of the disease such as tinnitus, aural fullness, quality of life, OAEs, ECoG. Reporting raw data prior to statistical processing is recommended. Characterising patients with Ménière's disease using the basic elements is not only useful for research purposes but also helps in making the best treatment choice.

4.2.1.1. Hearing threshold (HT)

Hearing loss is an important qualifier of Ménière's disease. The AAO-HNS guidelines advise the use of the arithmetic mean of the pure-tone thresholds at 0.5, 1, 2, 3 kHz. If 3 kHz is not available, it can be replaced by the mean of 2 and 4 kHz.

Based on hearing loss, patients can be categorized into four stages (Stage 1: HT ≤ 25 dB; Stage 2: HT between 26-40 dB; Stage 3: HT between 41-70 dB; Stage 4: HT > 70 dB). The frequency profile of the hearing loss is not taken into account for diagnosis. A SNHL that predominates low frequencies is often found in patients with Ménière's disease, but is insufficient to label as possible Ménière's disease because only a

minority of the patients with low frequency hearing loss without vertigo attacks develop definite Ménière's disease.

4.2.1.2. Word recognition (WR)

Concerning reporting the speech discrimination performance for monosyllabic words, maximum phoneme score expressed in percent is the metric of choice.

4.2.1.3. Frequency of vertigo (FV)

When considering the frequency of the vertigo attacks, the patient's complaints should be evaluated over a period of 6 months. The average number of rotatory spells lasting at least 20 minutes is commonly used as the index for reporting FV.

4.2.1.4. Functional level (FL)

In any patient-centered treatment, quality of life is a central concern. The AAO-HNS guidelines provide a 6 grade functional level scale relevant to Ménière's disease: the patient is asked to mark the description which fits best with his/her condition (Table 3).

4.3. *Reporting additional characteristics (recommendation B)*

Apart from the basic elements of the AAO-HNS guideline, additional metrics that quantify other symptoms of the disease may be reported: semi-quantitative data on tinnitus (T), pressure (P), imbalance (I), disability (D), indexes of ECoG, vestibulo-ocular reflex, postural stability, and quality of life questionnaires for patients suffering from vertigo.

4.3.1. Tinnitus, pressure and imbalance-dizziness

Initial guidelines of the International Prosper Ménière's Society suggested grading of tinnitus, pressure, and imbalance according to a 7 grade scale: the inner ear disorder profile.

Two reporting instruments for complaints that are difficult to quantify prevail: the visual analogue scale (VAS) and validated quality of life (QOL) questionnaires. With the VAS, the patient indicates a value between 0 (no symptom awareness) and 10 (maximal imaginable intensity).

In Ménière's disease, the VAS is mainly used for tinnitus and ear pressure. Disease specific QOL questionnaires are available for dizziness and for tinnitus.

Recently, Kato *et al.*¹⁴ used an 18-item questionnaire for evaluating the outcome of endolymphatic sac decompression in patients with Ménière's disease.

4.3.2. Electrocochleography

During ECoG, sound evoked electrical activity near the cochlea is recorded. This technique is not widely available and therefore not included as a basic element of the AAO-HNS guidelines. Nevertheless, ECoG can ascertain the presence of ELH (recommendation B). From the recorded signal, two metrics are extracted: the amplitude of the action potential (AP) and the amplitude of the summing potential (SP). The SP/AP-ratio is most useful for the diagnosis of ELH but normative data differ with stimulus parameters and equipment. With click stimuli, following values are indicative for hydrops:

- Extratympanic ECoG (ET-ECoG) (click stimulation): SP/AP ratio > 0.42.
- Transtympanic ECoG (TT-ECoG) (click stimulation): SP/AP ratio > 0.35.

With pure tone bursts stimulation and TT-ECoG, an SP < -2 μV is suggestive for ELH.¹⁵

Some authors claim that the diagnostic value of ECoG can be enhanced by administration of dehydrating substances prior to the recordings. For most of the patients with fluctuating hearing loss, ECoG is crucial in establishing the diagnosis of ELH.

Middle ear infection, effusion, liquorrhea, and former surgery

Table 3
Functional level according to AAO-HNS

<p>Regarding your current state of overall functioning, not just during attacks, check the ONE that best applies:</p> <ol style="list-style-type: none"> 1. My dizziness has no effect on my activities at all. 2. When I am dizzy, I have to stop what I am doing for a while, but it soon passes and I can resume activities. I continue to work, drive and engage in any activity I choose without restriction. I have not changed any plans or activities to accommodate my dizziness. 3. When I am dizzy, I have to stop what I am doing for a while, but it does pass and I can resume activities. I continue to work, drive and engage in most activities I choose, but I have had to change some plans and make some allowance for my dizziness. 4. I am able to work, drive, travel, take care of a family or engage in most essential activities, but I must exert a great deal of effort to do so. I must constantly make adjustments in my activities and budget my energies. I am barely making it. 5. I am unable to work, drive or take care of a family. I am unable to do most of the active things that I used to. Even essential activities must be limited. I am disabled. 6. I have been disabled for one year or longer and/or I receive compensation (money) because of my dizziness or balance problem.

may constitute a contra-indication for TT-ECoG. In case of a contralateral deaf ear, use of ECoG should be conservative.

4.3.3. Vestibular testing

As with all other single disease parameters, vestibular testing cannot establish the diagnosis itself, but is useful for differential diagnosis, and for determining the functional status of the labyrinths. Dynamic posturography, gait and balance tests assess the vestibulo-spinal reflexes. Vestibular findings in patients with Ménière's disease are highly variable and well documented.¹⁶

5. Therapeutic management

Nowadays, no cure for Ménière's disease has been proven. Nevertheless, different treatment modalities can significantly improve the patient's quality of life. Management of Ménière's disease is focussed on the vertigo attacks: reducing the symptoms during an attack, and reducing the chance for new attacks.

5.1. Treatment of attacks

In case of an attack, treatment aims at reducing the rotational sensation and neurovegetative symptoms. A number of drugs can be given (recommendation B): anti-emetic drugs (e.g. domperidone), vestibulosedative anti-histamines (e.g. meclizine), and central sedative drugs with vestibulo-suppressive and anti-emetic effect (e.g. diazepam, sulphiride, dihydrobenzperidol, and phenothiazine) (evidence based medicine (EBM) Level II). Because of nausea and vomiting, intra-rectal or parenteral administration is often necessary. No studies demonstrate superiority

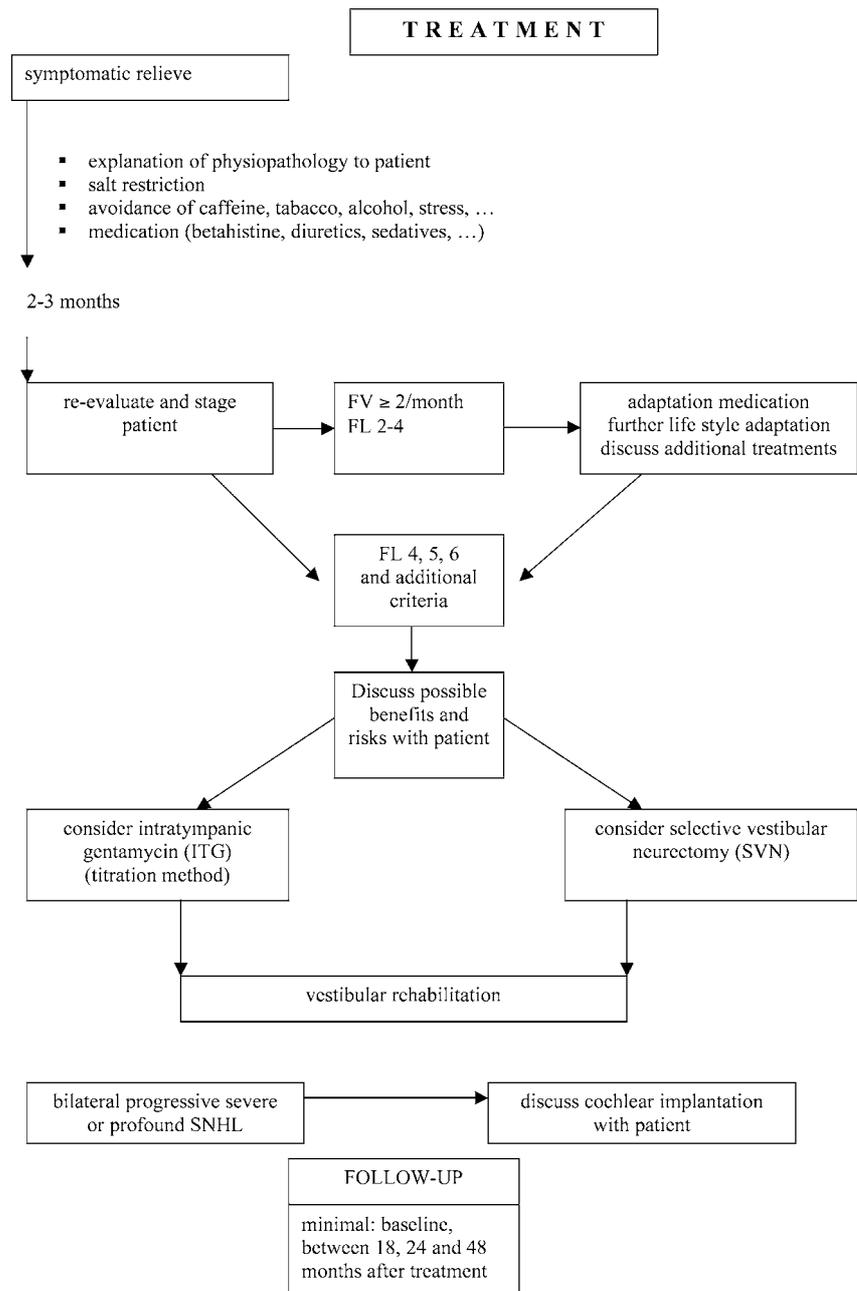


Figure 2
Treatment algorithm

of one of those products for an acute attack.

Fattori *et al.*¹⁷ administered hyperbaric oxygen during the early stages of the disease and found a long term protective effect. His findings, however, are under debate (EBM III; recommendation C). Findings from ani-

mal research also supports the use of hyperbaric oxygen therapy.

5.2. Long term treatment

Apart from treating vertigo attacks, medical support for patients with Ménière's disease can aim at reducing the number of vertigo attacks, compensating the

vestibular deficit, improving the patients coping strategies, and providing hearing aids. Practically, the physician provides information on the disease, suggests life style changes, prescribes medication and rehabilitation, and sometimes performs surgery.

5.2.1. Counselling and life style adaptations (EBM level II; Recommendation B)

During counselling, the physician has to explain gently what the patient may expect from the disease, and from medical support. Patients with basic knowledge about their illness will be able to cope more efficiently with a disease.

The physician should inform that the natural course of the disease is highly variable, that the diagnosis is described in terms of chance, that current knowledge is very limited, and that periodic evaluations are necessary. The coordinates of patient associations can be given.

Life-style adaptations consist of reducing salt, caffeine, alcohol, tobacco, and improving coping styles. Working in shifts should be avoided, and regular sleeping habits should be respected. Many patients can very well designate the event that provoked a vertigo attack. In exceptional cases, psychological support may be indicated.

5.2.2. Chronic drug therapy

Drug therapy plays an important role in the treatment of most patients with Ménière's disease. Since the late 80's, solid comparative studies reported the success of betahistine in reducing the frequency and severity of vertigo attacks without impairing vestibular

compensation (EBM II; recommendation B).

In the same period some studies reported also the benefit of diuretics (EBM level II; recommendation B). However, it is still controversial whether acetazolamide is also a beneficial diuretic (recommendation C).

It is important to know that in case of pregnancy or lactation, betahistine cannot be given (class D product) and that antidiuretics are to be avoided (class C product: to be used if unavoidable, no teratogenic effects known). When subsequent to an acute attack, imbalance is a dominant complaint, mild vestibular sedatives (e.g. cinnarizine) may help.¹⁸

In case of bilateral Ménière's disease, inflammatory influences should be suspected, and systemic glucocorticoids may be necessary for a short period. It has recently been proven that glucocorticoids do not only influence inflammation, but also fluid dynamics via interaction with Na⁺-pumps in the semicircular canals.¹⁹ In case of reponse to glucocorticoids, an auto-immune inner ear disorder with ELH has to be suspected.

The presumed beneficial effect of intra-tympanic corticosteroid application is transient, and recently a working group decided to exclude this therapy modality for Ménière's disease.²⁰

If the physician has the impression that anxiety and inefficient coping with stress dominate a patient's complaints, gaba-antagonist (e.g. alprazolam and serenase) can be administered for a short period.

5.2.3. Vestibular rehabilitation (recommendation C)

Vestibular rehabilitation improves central compensation of a peri-

pheral vestibular deficit and its resulting motion intolerance (EBM level I). Ménière's disease patients also benefit from this therapy. When a patient with Ménière's disease develops benign paroxysmal positional vertigo, a canalith repositioning procedure should be attempted (recommendation B). In case of nausea and oscillopsia due to nystagmus, prism spectacles of Utermohlen can be prescribed.

5.2.4. Incapacitating vertigo resistant to drug treatment

If the aforementioned drug treatments fail to control the vertigo attacks (e.g. FV \geq 2/month, FL 2-4), other therapeutic modalities can be proposed. As previously mentioned, treatment effect should be evaluated over a period of at least 6 months.

First we will describe four treatment modalities that are useful for intractable Ménière's disease and that are not based on destruction of the peripheral vestibular system. The effect and most appropriate timing of these modalities are still under investigation, and thus, our recommendations are based on individual experience (no EBM; no recommendation).

1. Transtympanic pressure treatment (Meniett™, Medtronic Inc. USA) with tympanostomy tube initially yielded promising results, but the effects may be limited to reducing the duration of the attacks, and are less effective when administered during end-stage disease.²¹
2. Repetitive administration of hyperbaric oxygen has recently been introduced by Fattori *et al.*¹⁷ who found long term protective effects.

3. Endolymphatic sac decompression has been criticized heavily, but is frequently performed in the USA.¹⁴
4. Franz *et al.*²² revisited the hypothesis that middle ear muscles play a role in the etiology of Ménière's disease and found that tenotomy of the musculus tensor tympani and stapedius had a long term protective effect for patients with Ménière's disease.

Treatment modalities that destroy the peripheral vestibular system gained EBM level I evidence for incapacitating (FL 4, 5, 6) Ménière's disease. Vestibular deafferentation can be achieved with intra-tympanic gentamycin application (ITG), labyrinthectomy, and selective vestibular neurectomy (SVN).

The advantage of ITG is its mildly invasive nature, and high success rate. The dosing and titration regimen determine the outcome and complication rate. The main bottle-neck of ITG is the inability to estimate the permeability of the round window membrane, and hence the optimal gentamycin dosage.

SVN yields maximal control of the vertigo attacks (> 95%), but is a relatively invasive otoneurosurgical procedure. Labyrinthectomy is somewhat less effective than SVN and excludes cochlear implantation if necessary. After an ablative procedure, vestibular rehabilitation is required as it supports central compensation (EBM II).

The pros and cons of ablative procedures must be notified to the patient:

- Persistent, troublesome disequilibrium in 20% of cases after surgical procedures.²³

- A higher risk for SNHL with ITG (15% all procedures) compared to SVN (less than 10%).²⁴

Improvement was observed in half of the patients to such an extent that SVN was not longer necessary 6 weeks after proposing a SVN and explaining the risks and benefits.²⁵ Relative contra-indications for ablative procedures are contralateral Ménière's disease, contralateral vestibular areflexia and any other visual, proprioceptive or CNS pathology that impairs vestibular function.

5.2.5. Hearing loss with Ménière's disease

Up until now, no proven therapy can reduce the loss of cochlear function in Ménière's disease. We, however, believe that preventive measures and life style adaptations offer the best perspectives. For many patients, when vertigo attacks ceased, hearing loss is the major problem. From an audiological point of view, the hearing loss has all characteristics of a SNHL: increased thresholds, recruitment, and reduced frequency selectivity. When compared to SNHL due to age, loudness recruitment associated with Ménière's disease is more pronounced. As a consequence, ampications gains preferred by patients with Ménière's disease differ from those with a common SNHL.

In many cases, speech recognition scores of aided hearing are disappointingly low. In case of bilateral profound SNHL due to bilateral Ménière's disease, good results were achieved with cochlear implantation. For unilateral profound SNHL, a bone anchored hearing aid (BAHA) may improve the hearing capabilities. With BAHAs, preliminary data demonstrate significant

improvement of severe tinnitus in unilateral profound SNHL due to Ménière's disease.

Different tinnitus treatments are available and can be offered (e.g. tinnitus retraining therapy), but results focussed on patients with Ménière's disease have not been reported.

6. Patient information

6.1. Counselling

Comprehensive information of all aspects of Ménière's disease should be provided and is highly appreciated by all patients with Ménière's disease. A realistic picture of the disease and reassurance has to be given, because many patients experience considerable anxiety in the period prior to diagnosis.

More difficult, but necessary, is dealing with what is known, not known, and uncertain. As medicine progresses, underlying pathologic processes will be uncovered, and reassessments will be necessary. It is also useful to inform close family members, as the occurrence of repeated attacks is sometimes misinterpreted. Patient groups can offer appropriate information and support to individuals with Ménière's disease and their family. The Flemish Ménière's disease patient group has as website: www.meniere.be. The French Ménière's disease patient group is: Entraide Ménière ASBL, 87 rue des Floralies Bt77, 1200 Bruxelles, tel. 02 762 91 83, E-mail entraidemeniere@hotmail.com, website www.entraidemeniere.be.

6.2. Follow-up

The diagnosis of Ménière's disease is based on history, measuring of hearing loss, and

exclusion of "other causes". The history of the complaints and self-appraisal guides the therapy. For this reason, a detailed baseline and semi-quantitative documentation of complaints is instrumental for optimal treatment:

- frequency, duration, severity of vertigo attacks
- imbalance assessed on the dizziness handicap inventory
- hearing difficulties and hyperacusis
- tinnitus disability
- pressure sensation in the ear

Also secondary or associated complaints have to be assessed:

- depression and anxiety
- headache

Aspects concerning work limitation and driving capabilities have to be discussed. So-called "safety functions" are not allowed until the disease is under control. Patients sometimes need some time off work. Especially when severe Tumarkin otolithic crises occur, patients may not drive a car for at least 3 months after the last attack.

Treatment of vertigo comprises three steps

Step 1: counselling and life style adaptation

Step 2: chronic drug therapy and vestibular rehabilitation

Step 3: (partial) destruction of the vestibular epithelium input

Special attention is paid to hearing and tinnitus.

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CME questions

1. The diagnosis of Ménière's disease is based on the clinical triad of symptoms consisting of recurrent vertigo, hearing loss and tinnitus or ear pressure combined with one of the following elements: (please indicate).

A – Fluctuating low-tone sensorineural hearing loss
B – Exclusion of other diagnoses (e.g. with MRI)
C – Positive Electrocochleography
D – Vestibular hypo-reflexia

2. Concerning the underlying pathophysiology of Ménière's disease: which statement is not true?

A – Vertigo attacks can be caused by intra-labyrinthine membrane ruptures
B – In endolymphatic hydrops there is mainly a volume increase of the endolymphatic compartment without measurable pressure increase
C – Ménière's disease is always accompanied with endolymphatic hydrops and vice versa
D – Endolymphatic sac dysfunction is a factor in Ménière's disease

3. Secondary endolymphatic hydrops may arise in following disorders except one: please indicate:

A – Acoustic neuroma
B – Otosclerosis
C – DFNA9 Coch gene mutation
D – Migraine associated dizziness

4. Definite Ménière's disease at first presentation is treated as follows. Indicate what is wrong (one statement).

A – Salt, caffeine and alcohol
B – Stress reduction
C – Intra-tympanic gentamycin therapy
D – Chlorothiazide treatment

5. Several conditions form the indication for ablative vestibular treatment. Indicate what is wrong (one statement).

A – Profound sensorineural hearing loss
B – Vertigo attacks not responding substantially to medical treatment during a 6 month period
C – Comprehensive patient information on the aim and adverse effects of the treatment
D – Moderate to severe degree of handicap (as assessed by the functional level scale)

Answers: 1B; 2C; 3D; 4C; 5A
