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SFORL Guidelines

Diagnostic and therapeutic strategy in Menière's disease. Guidelines of the French Otorhinolaryngology-Head and Neck Surgery Society (SFORL)

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ABSTRACT

Objectives: The authors present the guidelines of the French Otorhinolaryngology-Head and Neck Surgery Society (*Société française d'oto-rhino-laryngologie et de chirurgie de la face et du cou*: SFORL) for diagnostic and therapeutic strategy in Menière's disease.
Methods: A work group was entrusted with a review of the scientific literature on the above topic. Guidelines were drawn up, then read over by an editorial group independent of the work group. The guidelines were graded according to the literature analysis and recommendations grading guide published by the French National Agency for Accreditation and Evaluation in Health (January 2000).
Results: Menière's disease is diagnosed in the presence of the association of four classical clinical items and after eliminating differential diagnoses on MRI. In case of partial presentation, objective audiovestibular tests are recommended. Therapy comprises medical treatment and surgery, either conservative or sacrificing vestibular function. Medical treatment is based on lifestyle improvement, betahistine, diuretics or transtympanic injection of corticosteroids or gentamicin. The main surgical treatments, in order of increasing aggressiveness, are endolymphatic sac surgery, vestibular neurotomy and labyrinthectomy.

1. Introduction

The etiology of Menière's disease remains unknown and diagnosis, which is basically clinical, tends to be over-inclusive, without respecting established criteria. Criteria were first laid out more than 20 years ago, by the AAO-HNS [1] (level of evidence 4), and were modified in 2015 by an extended consensus between international scientific societies [2] (level of evidence 4). To help practitioners in this difficult diagnosis and in assessing severity, complementary examinations, mainly screening for underlying hydrops (which is frequent), have been developed. Treatment is varied, and efficacy hard to assess. Treatment may be medical, or surgical, either conservative or sacrificing vestibular function with risk, in some cases, for cochlear function.

The present guidelines are intended for all ENT physicians liable find themselves managing a patient with Menière's disease. The objectives are three-fold: making the diagnosis, selecting appropriate tests to confirm diagnosis and monitor treatment efficacy, and selecting the treatment option best suited to the patient. We considered it particularly important:

- to clarify the diagnostic criteria for the disease so as to prevent over-diagnosis in patients with heterogeneous and incomplete

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presentations (level of evidence 4), and also to rule out differential diagnoses (level of evidence 4);
- to inform practitioners in selecting instrumental tests that will be useful and contributive or essential to paraclinical work-up. Such tests should meet two objectives:
  - screen for hydrops [3–6] (level of evidence 2), which is usually associated and requires specific treatment,
  - assess cochlear and vestibular involvement at a given time-point in disease progression, bearing in mind the wide fluctuations that are characteristic [2] (level of evidence 4);
- to specify the respective roles of the various treatment modalities, both destructive and conservative [7,8] (level of evidence 2), medical and surgical, according to disease stage and possible bilateral involvement; this should include possible physiotherapy or psychotherapy [9] (level of evidence 1).

A national work group was entrusted with drawing up guidelines for Diagnostic and Therapeutic Strategy in Menière's Disease. The French Health Authority (HAS) formalized expert consensus methodology for good practice guidelines was used (http://www.has-sante.fr). A pilot group organized the logistics of the consensus conference, and choice of members of the editorial group performing the literature analysis by means of the PubMed database. Each retrieved article was graded A, B, C or "expert opinion" according to decreasing level of evidence, in line with the guide to literature analysis and recommendations grading of the French National Health Evaluation and Accreditation Agency (ANAES). Based on a written rationale, an initial series of guidelines was drawn up and then assessed by the editorial group and modified in line with the results and comments received.

2. Results

2.1. 1 Diagnosis of Menière's disease

Diagnosis of Menière's disease is clinical, based on simple criteria associated to presumed presence of endolymphatic hydrops. It is first of all essential to distinguish between "definite" and "probable" Menière's disease. In the opinion of the experts, vertigo crises associated with hearing loss are preconditions for any diagnosis of Menière's disease.

"Definite" Menière's disease is to be diagnosed in the absence of other identified cause and presence of an association of the following 4 clinical signs (Expert opinion):

- vestibular signs: at least 2 rotational vertigo episodes lasting between more than 2 minutes and 12 hours, or Tumarkin's otolithic crises (drop attacks without initial loss of consciousness);
- auditory signs: low frequency (< 2 kHz) hearing loss on two contiguous frequencies, of at least 30 dB in case of normal contralateral hearing or at least 35 dB in case of bilateral hearing loss, on an audiogram performed during or after a crisis; these signs may occur several months or years before onset of vertigo;
- other otologic signs: tinnitus or aural fullness;
- fluctuating otologic signs.

It is important to bear in mind that endolymphatic hydrops may be primary, in which case the term Menière's disease is fully appropriate, or may be secondary to a potentially severe pathology requiring specific treatment.

Menière's disease should be diagnosed only after ruling out differential diagnoses of tumor (cerebellopontine ngle or endolymphatic sac tumor), deformity (Chiari malformation) or degenerative inflammatory pathology (multiple sclerosis) on MRI of the posterior fossa and cervico-occipital hinge on axial and sagittal slices with and without contrast enhancement, and including high-resolution T2-weighted 3D acquisitions (Expert opinion).

In "probable" Menière's disease, incomplete symptomatology leaves diagnosis uncertain, and certain objective paraclinical examinations that have shown specificity in definite forms are contributive.

In case of suggestive but incomplete clinical presentation ("probable" or merely hypothetical Menière's disease), before screening for hydrops or intralabyrinthine pressure or volume defect, some or all of the following objective audiovestibular explorations should be performed (Grade B): multifrequency admittancemetry, and/or acoustic phase-shift test using transient otoacoustic emissions (TOAEs) and/or distortion product OAEs (DPOAEs), and/or electrocochleography to assess the summating potential/action potential ratio (SP/AP). Positive findings demonstrate intralabyrinthine pressure regulation disorder.

Before informing the patient of the diagnosis and setting up adapted treatment, it is essential to check progression status and especially the size of any lesions already present in the vestibule or cochlea, even if measured deficits are fluctuating. Instrumental assessment should be backed up by functional and quality-of-life assessment, going beyond simply counting the number of crises. The 1995 AAO-HNS Functional Level Scale (Appendix 1) assesses severity in 6 levels [1] (Expert opinion), and a French version has been validated [2] (Expert opinion).

When definite Menière's disease is diagnosed, pre-treatment instrumental assessment should be performed, including at least complete pure-tone and speech audiometry and videonystagmography with calibrated caloric test (Expert opinion); preferably, vestibular evoked myogenic potentials (VEMP) and a video head impulse test (VHIT) should be associated, if available. Disability should be assessed on the validated Functional Level Scale to quantify disease impact on quality of life, before treatment and to monitor efficacy [1].

2.2. Treatments for Menière's disease

Drawing up therapeutic guidelines was hampered by the low level of evidence of studies in the international literature. The authors therefore chose to detail pros and cons for each treatment, before recommending indications. No fixed decision-tree, however, could be drawn up. We can only recommend determining the patient's complaints as clearly as possible and assessing global health status so as to guide optimal treatment, which should initially comprise the most functionally conservative interventions.

2.3. Medical treatments for Menière's disease

There are many kinds of maintenance therapy in Menière's disease, and they can be classified into main families. Initial management should include lifestyle counselling. Next come the various types of conservative therapy; here we shall discuss betahistine, diuretics, and general route or intratympanic corticosteroids; then transtympanic gentamicin, a destructive treatment, will be dealt with.

Adjuvant treatments, such as physiotherapy and psychotherapy are important and will be dealt with independently. Finally, it is essential to compensate hearing loss, but this is a challenge for all those involved in managing a patient with Menière's disease: ENT specialist, hearing-aid specialist and speech therapist.

It is important that the strategy should be adapted not only to symptom severity but also to global health status (Expert opinion). It is essential that first-line treatment should conserve vestibular function, keeping destructive treatments as a last resort [7,8]

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