Ménière's Disease

Ménière's disease (pronounced "many-ears") is infrequent problem of the inner ear.

It is a repeating problem of the inner ear in which there is a build-up of fluid in the membranes of the inner ear giving a feeling of pressure or fullness in the ears. These membranes then suddenly burst making you feel very dizzy.

It is characterized by 2 or more attacks of vertigo* lasing 20-min to 12 hours with fluctuating hearing loss and a feeling of fullness in affected ear.

The classical "sequential" presentation occurs in about a 1/3rd of patients.

The attack starts with a "fullness" sensation in the ear with a low frequency buzzing followed by hearing loss and then vertigo with nausea, vomiting and sweating. Gradually the hearing recovers. The patient often has a period of imbalance lasting for days.

A measure of severity is the record of how many of the episodes were associated with vomiting and those without vomiting

However, in about 2/3rd of patient there is a more simultaneous pattern where there is no warning the attack with all the symptoms of vertigo etc. occurring at the same time. Again, the hearing gradually recovers.

The vertigo can occur at any time of day and night and can be triggered by the head being in various positions. Patients may also complain that loud sounds can induce vertigo (the Tullio Phenomenon).

The clinical picture is spells of multiple irregular attacks and then spells without attacks. The disease may be controlled by medication, resolve or progresses with fluctuating but diminishing hearing loss. Initially 1/7th of cases involve both ears, but by 10 years 1/3rd of patients have both ears involved and by 10 years it increases to ½ of patients.

In the long run the balance system can so damage such that the spinning vertigo is replaced by a constant feeling of unsteadiness** because the inner ear balance system is failing (vestibular failure).

The disease has well described variants: -
i)Tumarkin's Otolithic Crises (Vestibular Drop Attacks): This occurs in 7% of patients in which the patient suddenly falls to the ground without loss of

consciousness. There is complete recovery within seconds 10 minutes. Patients describe a sensation as if they are suddenly in “free fall”, they desperately try to right themselves, and end up on the floor with injuries being very common. Patients feel as if they have been "shoved" or “pushed” or “flipped”.

ii) Vestibular Meniere’s where there is dizziness without hearing complaints or evidence of hearing problems
iii) Cochlear Meniere’s where there is hearing problems without balance complaints or evidence of balance problems
iv) Delayed Meniere’s where it starts with hearing loss with a later development of Meniere’s symptoms.
v) Lermoyez syndrome. In this form the hearing loss very rapidly recovers after the attack of vertigo so that the patient may describe the vertigo as “restoring the hearing”.

The certainty of the diagnosis is either definite or probable: -

In definite disease the patient has to experience two or more vertigo attacks coming “out of the blue” which last between 20 minutes but less than 24 hours. These attacks must be associated with at least one hearing test showing low to mid frequency hearing loss in one ear. In probable disease the patient is the same but without a recorded hearing test.

Salt
Labyrinth is a sodium osmolality change detecting organ – in other words it is a “salt meter”. Any sudden increase in salt intake result in spikes in the plasma concentration which are detected (not the absolute level) by the membranous labyrinth. Patients who eat a salty meal may get a return of their symptoms.

Examination
In an acute attack there are 2 phases: -

In the initial excitatory phase patients have abnormal eye movements: unidirectional horizontal jerk nystagmus towards the affected ear, and a tendency to fall to the side of the unaffected ear

In the subsequent inhibitory phase patients, the abnormal eye movements change to unidirectional horizontal jerk nystagmus towards the unaffected ear and a tendency to fall to the side of the affected ear.

The patient’s response to tuning fork test are Rinne positive in both ears with the Weber to the unaffected ear consistent with a damaged inner ear hearing damage. Patients may also have a condition called “diplacusis” where they

describe the pitch of the tuning fork as being higher in the affected ear (256Hz). In addition, patients may exhibit a positive “Henneberts Sign” where pressure on the ear canal induces eye movement.

It is worth considering four-hour delayed contrast MRI to predict contralateral Meniere’s before clinical signs become apparent.

Most importantly of all the patient does not have abnormal brain function.

Hearing test abnormalities

There is a low tone neural deafness which fluctuates but declines over time. The accepted definition of is a reduction of the hearing levels at two contiguous frequencies on the pure tone audiogram at the time of spell of ≥30dB at 250Hz/500Hz/1KHz.

High tone loss does occur and should not be dismissed

Imaging

Intravenous Gadolinium 4 hour 3D FLAIR MRI (3 Tesla scanner) of the inner ear shows black cochlear endolymphatic lacunae. It not used in definite cases but it can be used to exclude other possible causes. This form of scan does not help in tracking disease progress or response to treatment. The scan may also pick up imaging changes which are associated with disease in the other ear.

Patients who have a symmetrical brackets one-sided hearing loss) should have an MRI to exclude an acoustic neuroma.

Other diagnoses

In a patient with Meniere’s disease 40% also have migraine, 20% get BPPV and many have a secondary Persistent Postural Perceptual Dizziness (PPPD) response.

1 in 200 patients have an acoustic neuroma (and 1 in 20 acoustics neuroma patients have Meniere’s disease.

For the symptomatic relief of the acute vertigo and nausea.
See - Advice and Medication for Acute Vertigo and Nausea for all conditions other than BPPV. In addition, see below for the specific management of a severe episode of Ménière's disease.

For the specific preventative treatment of the Ménière's disease
This is used when there is a significant reduction in the quality of life of the patient. It is clearly needed in patients with severe frequent attacks and not in patients with mild infrequent attacks.

What preventative treatment?

Treatment should be 3 times as long as typical interval between attacks to see an effect.

Patients should be initially recommended

i) avoidance of high salt peaks in the diet
ii) Low/no caffeine diet
iii) histamine analogue
iv) Diuretics

The evidence for the efficacy of diuretics is controversial and many specialists in the UK and Germany do not usually prescribe.

In the USA Dyazide® which is co-triamterzide (triamterene 37.5mg with hydrochlorothiazide 25mg o.d.) Triamterene is a potassium-sparing diuretic often used in combination with a thiazide diuretic e.g.hydrochlorothiazide (See Risks of medication below)

In the UK bendroflumethaside 2.5mg o.d, in the morning, especially if related to menstruation.
(See Risks of medication below)

Betahistine – Histamine Analogue
This has been popular for many years and may help, at present, subgroups but studies suggest that it may be no more effective than placebo. It is a low-risk medication and may be worth considering.
The recommended dose :
48mg orally four times daily should be given for:-
3-6 months if the patient has had 2 attacks in the last 6 months
6-12 months if the patient has had 1 attack or more a month in the last 6 months

In individual cases, this dose can be increased up to 480 mg daily if there is insufficient symptom alleviation after 3 months of treatment on 48mg four times daily.

Recent work from Strupp M et al from the Department of Neurology and German Centre for Vertigo and Balance Disorders, University of Munich Hospital, Germany, has documented the use of approximately 1000mg betahistine per day in a group of patients which was subsequently reduced to 200mg per day with the concomitant use of MAO-B inhibitor selegiline (5mg/day) - https://www.ncbi.nlm.nih.gov/pubmed/29532287
Other medical evidence which supports its use
https://www.ncbi.nlm.nih.gov/pubmed/29282702

However, The BEMED trail (https://pubmed.ncbi.nlm.nih.gov/26797774/ BMJ. 2016 Jan 21;352) did not demonstrate a therapeutic effect above the placebo effect.

Intratympanic injections of steroids

This treatment has a strong scientific basis and should be very much considered (Lancet. 2016 Dec 3;388(10061):2753-2762).I do not offer this but would refer to Mr John Harcourt for this therapy.

The indications for its use in this paper were 2-3 attacks a month for at least 6 months.

Drug management of a very severe spell/episode of Ménière's disease (Status Hydrops).

Prednisolone* EC in the morning
Day 1- 80mg
Day 2- 80mg
Day 3- 60mg
Day 4- 60mg
Day 5- 40mg
Day 6- 40mg
Day 7- 20mg
Day 8- 20mg
Day 9- 10mg
Day 10- 10mg
To be taken with a PPI – please be aware of side effects and contraindications - https://bnf.nice.org.uk/drug/prednisolone.html)

AND
Betahistine 48mg, three times a day for 6 months.

Other Issues: Vestibular Migraine

Ménière's disease and vestibular migraine often coexist and thus if there is a reasonable suspicion that a contributory element to the patient's dizziness is caused by vestibular migraine then I treat with the following migraine prophylaxis:-

Medication

Propranolol*** 10mg bd escalating gradually to 240-320mg day . It is especially useful in the anxious patient. Please note it can be associated with weight gain.

*** propranolol should not be used in patients with asthma, heart and diabetes problems.

OR

Nortriptyline 10mg o.d then 25mg o.d increasing to 50 mg (75 mg for males) by 3 weeks. It is particularly useful in patients who do not sleep well.
The main side effect is a dry mouth and palpitations, but weight gain and sedation are less than amitriptyline. It is precluded in pregnancy.
https://bnf.nice.org.uk/drug/nortriptyline.html

OR

Topiramate 25mg od increasing to 25mg bd after 2-4 weeks, then increasing in 25mg steps to 50mg bd then up to 100mg bd. Patient should ensure that they remain well hydrated.

Possible side-effects of this medication that should be mentioned are weight loss, the needles in your hands and feet, kidney stones, a feeling of slight fogginess of your thinking and possibly even slurred speech. It is also associated with low mood.

In particular women and girls of childbearing potential need to be advised that topiramate is associated with a risk of fetal malformations and can impair the effectiveness of hormonal contraceptives.
https://bnf.nice.org.uk/drug/topiramate.html

Prescribing issues

Dyazide®: There is a risk of increased skin cancer* with dyazide is related to the length of drug use, amount of sun exposure and especially a risk in susceptible populations (i.e. fair-haired, blue-eyed Scandinavians). Essentially the evidence suggests a significant increased risk of non-melanoma skin cance if the medication has been used for 6 years+. I don't think this warrants discontinuation of the drug for Meniere's disease if it seems to be working but I would advise patients to use adequate sunscreen when exposed.

*Hydrochlorothiazide use and risk of nonmelanoma skin cancer: A nationwide case-control study from Denmark. J Am Acad Dermatol 2018;78:673-81.
and https://www.gov.uk/drug-safety-update/hydrochlorothiazide-risk-of-non-melanoma-skin-cancer-particularly-in-long-term-use

Bendroflumethaside
https://bnf.nice.org.uk/drug/bendroflumethiazide.html

Betahistine is contraindicated in patients with phaeochromocytoma.
Caution is advised in the treatment of patients with peptic ulcer or a history of peptic ulceration, because of the occasional dyspepsia encountered in patients on betahistine.

Clinical intolerance to Betahistine may occur in bronchial asthma patients. These patients should therefore be monitored carefully during the treatment with betahistine.

Caution is advised in prescribing betahistine to patients with either urticaria, rashes or allergic rhinitis, because of the possibility of aggravating these symptoms.

Caution is advised in patients with severe hypotension.

Patients with rare hereditary problems of galactose intolerance, the Lapp lactase deficiency or glucose-galactose malabsorption should not take this medicine.

In vitro data indicate an inhibition of betahistine metabolism by drugs that inhibit monoamino-oxidase (MAO) including MAO subtype B (e.g. selegiline).

Caution is recommended when using betahistine and MAO inhibitors (including MAO-B selective) concomitantly.

It is preferable to avoid the use of Betahistine during pregnancy.

Breast feeding: The importance of taking the medicine by the mother must be weighed against the benefits of breastfeeding and the potential risk for the child.

Fertility issues: Animal studies show no influence on fertility in rats.

The following undesirable effects have been experienced with the below indicated frequencies in betahistine-treated patients in placebo-controlled clinical trials and in post-marketing reports: very common (1/10); common (1/100 to <1/10); uncommon (1/1,000 to <1/100); rare (1/10,000 to <1/1,000); very rare (<1/10,000); and not known (frequency cannot be estimated from the available data).

Immune system disorders:

Not known: hypersensitivity reactions, e.g. anaphylaxis.

Nervous system disorders:

Common: headache, occasional drowsiness

Cardiac disorders

Not known: palpitations

Respiratory disorders

Not known: Bronchospasms may occur in patients with bronchial asthma (see section 4.4)

Gastrointestinal disorders:

Common: dyspepsia *, nausea

Skin and subcutaneous tissue disorders

Not known: cutaneous and subcutaneous hypersensitivity reactions, in particular angioneurotic oedema, urticarial, rash, and pruritus

*Mild gastric complaints (e.g. vomiting, gastrointestinal pain, abdominal distension and bloating) have been observed. These can normally be dealt with by taking the dose during meals or by lowering the dose."

A few overdose cases have been reported. Some patients experienced mild to moderate symptoms with doses up to 640 mg (e.g. nausea, somnolence, abdominal pain).Other symptoms of betahistine overdose are vomiting, dyspepsia, ataxia and seizures. More serious complications (convulsion,

pulmonary or cardiac complications) were observed in cases of intentional overdose of betahistine especially in combination with other overdosed drugs. No specific antidote. Gastric lavage and symptomatic treatment are recommended within one hour after intake.

Surgical Intervention

Semi-circular canal occlusion surgery, saccus decompression and labyrinthectomy are available. The evidence for their use is uncertain

Other issues : Driving

The DVLA requires that the patients who suffer from sudden severely disabling vertiginous/dizziness attacks, which come on without warning, should report their condition to the DVLA.

In addition, current ENT opinion is that a patient suffering from sudden severely disabling vertiginous/dizziness disorder should not drive for a six-week period after a significant attack.

*vertigo is the sensation of spinning or the world moving: "I am spinning" "the room is spinning" or "I feel like I'm falling backward" "the room is rocking like on a boat"
**non-spinning imbalance: "Unsteady on walking", "Unsteady on feet", "cannot feel the ground properly" "Like being drunk"

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