# Ménière's:

## why its diagnosis calls for more careful evaluation

### DIFFICULTIES IN DIAGNOSING MÉNIÈRE'S

The triad of vertigo, tinnitus, and deafness is a familiar presentation in a GP surgery, but diagnosis and treatment remain a challenge. *'Difficulty with everything in general practice is spotting the unusual from the commonplace, and continuity of care is really important'*, says Dr Henrietta Hughes, GP and National Guardian for the NHS. *'If a patient comes back to see several different people, they might all try something to fix the immediate problem rather than seeing the whole picture'* (personal communication, January 2017).

There is no definitive test for Ménière's syndrome and it is not at all uncommon for people to be misdiagnosed. The classic symptoms are fluctuating hearing loss, low-pitch tinnitus, fullness in the ear, and episodic spinning vertigo that lasts at least 20 minutes, but typically 2–3 hours. (The whole attack should be over in less than 24 hours.) A number of recent studies examining the experiences of dizzy patients[1–3] have indicated that doctors frequently diagnose it when they see any vertigo they don't understand, and there are patients diagnosed with Ménière's syndrome who simply never get evaluated for anything else. Taking the history of a dizzy patient is essential to differentiate the possible aetiologies of vertigo, and a systematic approach must be used. The mnemonic 'SO STONED'[4] has been proposed, summarising the key factors that allow a first approximation of diagnosis identification: Symptoms, Often (frequency), Since, Trigger, Otology, Neurology, Evolution, and Duration. The most common vestibular diseases have different fingerprints when all these factors are examined.

The most common cause of vertigo is benign paroxysmal positional vertigo (BPPV) and it can be primary or secondary. If it is primary, it is easily treated with canalith repositioning known as Epley manoeuvres. A patient with BPPV may tell a GP they have experienced vertigo 'for days', but a few direct questions often reveal that they have in fact experienced brief episodes of vertigo with a position change for a period of days, but the vertigo actually stopped if they held still. Secondary BPPV is being caused by something, and that something is frequently another inner-ear problem that is being overlooked. Such cases can usually be identified by the fact that the BPPV keeps returning, or that the patient has inner-ear symptoms that cannot be attributed to BPPV, such as hearing loss, tinnitus, or fullness in the ear.

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Patients who complain about fullness in the ear are frequently told it is due to Eustachian tube dysfunction, secondary to allergy or an upper respiratory infection. This is indeed often the case, but for many others it is actually a symptom of an inner-ear problem. The two types of patients can be distinguished by otoscopic examination and a tympanogram. If those are both normal, Eustachian tube dysfunction should essentially be ruled out and the clinician should establish whether other symptoms of an inner-ear disorder can be elicited.

### MÉNIÈRE'S AS SECONDARY DISORDER

When considering Ménière's, it is important to distinguish between Ménière's disease, when the cause cannot be identified (idiopathic in other words), and Ménière's syndrome, when it is secondary to another disorder. There is a long list of possible causes, including hypothyroidism, tumours (most notably acoustic neuromas),[5] infections, metabolic or immune disorders, perilymphatic fistula, genetic mutations,[6] superior semicircular canal dehiscence syndrome (SCDS), and many more. Treating the underlying cause often resolves the Ménière's syndrome with much greater success than the non-specific treatments employed for Ménière's disease. A drug that works for one person does not necessarily work for another because the underlying reasons for the syndrome are so varied and the complex pharmacokinetics of the inner ear make drug delivery to it a tricky business. The diagnosis of Ménière's should come from a specialist with a specific focus on vestibular disorders. If the patient has only had one vertigo attack, it may be because of an inner-ear infection, and a diagnosis of Ménière's disease should not be given unless multiple attacks have occurred.

Distinguishing between central (including cerebrovascular disease and multiple sclerosis) and peripheral causes of vertigo has to be part of the diagnostic process. A sensation of lightheadedness, or being close to fainting, often results from diffuse cerebral ischaemia. The mnemonic 'DANISH' can help identify the clinical signs: Dysdiadochokinesis, Ataxia, Nystagmus, Intention tremor, Scanning dysarthria, and Heel-shin test positivity. If the vertigo can be turned on and off by changing position, BPPV should be considered. If there are no hearing-related symptoms at all, no disturbance in the hearing, and no hearing loss, doctors should consider migraine, particularly if the patient complains of headache or head pressure. Vestibular migraine (also known as MAV, migraine-associated vertigo) typically presents in a patient as unilateral onset of head pain, with throbbing as the pain intensifies. A family history of migraine makes MAV more likely, and it is the pattern of the symptoms that should be examined to reach the diagnosis,

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following the criteria established by the International Headache Society in 2013.[7]

## SUPERIOR SEMICIRCULAR CANAL DEHISCENCE SYNDROME

In recent years it has become apparent that many patients whose Ménière's symptoms have been considered idiopathic are in fact suffering from SCDS, a condition first identified by Dr Lloyd Minor in 1995, and reported in 1998.[8] SCDS involves thinning of the temporal bone over the inner ear resulting in holes that cause dizziness, vertigo, autophony (the hearing of self-generated noises such as heartbeat unusually loudly, or the eyes moving), hearing loss, fullness in the ear, nausea, headache, and fatigue. However, it can occur with thin bone and no actual hole.[9–11] This condition is very varied in its presentation but is often associated with symptoms of anxiety and panic attacks (which can be seen with any vestibular disorder, not just SCDS), as well as cognitive impairments. SCDS has been described as a great otologic mimicker,[12] so a reasonable degree of suspicion is advisable and a high-resolution CT scan is required. Treatment of SCDS by surgical intervention has been shown to be effective in the majority of cases.[13]

Recent studies have indicated that SCDS is much more common than first thought.[14] Dr Timothy Hain, Professor Emeritus at Northwestern University Medical School, stated in October 2016: *'We have diagnosed SCDS in 46 patients (as of 2014). This compares to several thousand patients with BPPV, thus SCDS is much less prevalent than BPPV which affects 2% of the population. As it is known that about 2% of the population has thinning of the bone that can lead to SCDS, our estimate of prevalence of symptomatic SCDS is about 0.1% of the population.'*[15] Dr Gerard Gianoli, at the Ear and Balance Institute in Louisiana, has treated more patients with SCDS than anyone in the world (starting in 1998) and he concurs with this estimate (personal communication, August 2017 ).

## CONCLUSION

When a patient presents with the Ménière's triad of vertigo, tinnitus, and deafness GPs should carefully consider other diagnoses, and SCDS must be among them. What the patient means by the term 'dizziness' needs to be precisely determined, and then a systematic approach employed to evaluate each individual.

**ADDRESS FOR CORRESPONDENCE**

**Philippa Thomson**
4 Kingston Farm Cottages, North Berwick, East Lothian EH39 5LT, UK.

**E-mail: philippathomson0@gmail.com**

**Philippa Thomson,**
Researcher and author, North Berwick, East Lothian.

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## REFERENCES

1. To-Alemanji J, Ryan C, Schubert MC. Experiences engaging healthcare when dizzy. *Otol Neurotol* 2016; **37(8):** 1122–1127. DOI: 10.1097/MAO.0000000000001145.
2. van Leeuwen RB, van der Zaag-Loonen H. Referrals to a specialised dizziness clinic often result in revised diagnoses and new therapeutic advice. *Eur Neurol* 2015; **73(1–2):** 20–22. DOI: 10.1159/000366415. Epub 2014 Nov 1.
3. Jin Z, Zhuang JH, Zhao ZX, *et al.* [Analysis of misdiagnosed cases with BPPV]. [Article in Chinese]. *Zhonghua Yi Xue Za Zhi* 2012; **92(19):** 1346–1348.
4. Wuyts FL, Van Rompaey V, Maes LK. 'SO STONED': common sense approach of the dizzy patient. *Front Surg* 2016: http://dx.doi.org/10.3389/fsurg.2016.00032.
5. Homann G, Fahrendorf D, Niederstadt T, *et al.* HR 3 Tesla MRI for the diagnosis of endolymphatic hydrops and differential diagnosis of inner ear tumors — demonstrated by two cases with similar symptoms. *Rofo* 2014; **186(3):** 225–229. DOI: 10.1055/s-0033-1356221. Epub 2014 Jan 22.
6. Verstreken M, Declau F, Wuyts FL, *et al.* Hereditary otovestibular dysfunction and Meniérè's disease in a large Belgian family is caused by a missense mutation in the COCH gene. *Otol Neurotol* 2001; **22(6):** 874–881.
7. Goto F, Tsutsumi T, Ogawa K. [The clinical features of migraine-associated vertigo]. [Article in Japanese]. *Nihon Jibiinkoka Gakkai Kaiho* 2013; **116(8):** 953–959.
8. Minor LB, Solomon D, Zinreich JS, Zee DS. Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. *Arch Otolaryngol Head Neck Surg* 1998; **124(3):** 249–258.
9. Ward BK, Wenzel A, Ritzl EK, *et al.* Near-dehiscence: clinical findings in patients with thin bone over the superior semicircular canal. *Otol Neurotol* 2013; **34(8):** 1421–1428. DOI: 10.1097/MAO.0b013e318287efe6.
10. Mehta R, Klumpp ML, Spear SA, *et al.* Subjective and objective findings in patients with true dehiscence versus thin bone over the superior semicircular canal. *Otol Neurotol* 2015; **36(2):** 289–294. DOI: 10.1097/MAO.0000000000000654.
11. Wackym PA, Wood SJ, Siker DA, Carter DM. Otic capsule dehiscence syndrome: superior semicircular canal dehiscence syndrome with no radiographically visible dehiscence. *Ear Nose Throat J* 2015; **94(8):** E8–E24.
12. Zhou G, Gopen Q, Poe DS. Clinical and diagnostic characterization of canal dehiscence syndrome: a great otologic mimicker. *Otol Neurotol* 2007; **28(7):** 920–926.
13. Thomeer H, Bonnard D, Castetbon V, *et al.* Long-term results of middle fossa plugging of superior semicircular canal dehiscences: clinically and instrumentally demonstrated efficiency in a retrospective series of 16 ears. *Eur Arch Otorhinolaryngol* 2016; **273(7):** 1689–1696.
14. Thomson P. *A hole in my life: battling chronic dizziness.* Philippa Thomson/Createspace, 2016.
15. Hain TC. Superior canal dehiscence. 2017. www.dizziness-and-balance.com/disorders/unilat/scd.htm (accessed 19 Sep 2017).