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What is the Current and Potential Future Role of Imaging in the Investigation of Patients with Suspected Meniere's Disease?

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Abstract

Meniere's disease (MD) represents a diagnostic challenge. It's symptoms of episodic vertigo, hearing loss, tinnitus, and a sense of aural fullness share overlap with multiple other conditions. It remains a diagnosis of exclusion, with imaging only used to rule out other pathologies, notably acoustic neuroma. Technological advance has brought increased resolution to the inner ear. In the future it is likely that MD will become a radiological diagnosis.

Keywords: Hydrops; Imaging; Dizziness; Vertigo

Meniere's Disease

Currently, the primary role of imaging in the investigation of suspected MD is to exclude other pathology. MD can present with asymmetrical hearing loss. In such circumstances it is usual to perform an MRI to exclude an acoustic neuroma or other forms of cerebellopontine angle tumour. The guidance on precisely when a patient requires an MRI to investigate asymmetrical SNHL is less clear. A comparison of eight screening protocols found that the best compromise between sensitivity and specificity was achieved by a criterion requiring either a 20 dB asymmetry at two neighbouring frequencies, or 15 dB asymmetry at two frequencies between 2 and 8 kHz [1]. Similarly, in the vertiginous patient an MRI can be used to exclude, space occupying lesions such as endolymphatic sac tumours, cholesteatoma fistulas, and brain metastasis [2].

In the acute setting, the symptoms of a Meniere's attack can be mimicked by an anterior inferior cerebellar artery (AICA) stroke, which shares the symptoms of vertigo, tinnitus, deafness. The other symptoms of hemiplegia, and facial weakness offer greater distinction. However, all patients presenting with such a first episode of such symptoms should have a Computed Tomography (CT) scan to exclude a central cause [3]. The symptoms of AICA occlusion, display the lack of collateral vessels to the inner ear and its high metabolic rate, an anatomical fact which may have a bearing on the pathophysiology of MD.

The future importance of imaging in the investigation of the suspected MD patient will depend on the other diagnostic modalities with which it is competing. Therefore it is necessary to comparatively discuss their diagnostic accuracy.

A pure tone audiogram (PTA) is a basic step in the Meniere's diagnosis. MD classically produces a fluctuating low frequency hearing loss that becomes permanent as the disease progresses. However, it can cause many other patterns of hearing loss, which may mimic differentials such as otosclerosis, or noise induced losses [4].

Electronystagmography (ENG), is another routine investigation as MD causes a reduced vestibular response in the ear affected. However, it has limited sensitivity as recent work shows only a 50% positive rate in confirmed MD cases [5].

Electrocochleography (ECoG) provides electrophysiological verification. An elevated summating potential/ nerve action potential of greater than 35% is a sign of MD. However, whilst a useful part of the diagnostic battery, specificity and sensitivity remains low particularly when performed in between episodes. A retrospective case review between 1995 and 2003 subcategorized patients by the AAO-HNS guidelines into definite, probably and possible MD. The study failed to show a statistically significant difference in the percentage of raised summation potentials between the definite and non-definite groups. Additionally, over 30% of those with definite MD would not have been classified as having MD based on ECoG results alone [6].

Further concern about the test specificity has been raised by a study showing that superior semicircular dehiscence syndrome (SSCD) gives similarly raised thresholds [7]. SSCD has the ability to mimic MD by causing episodic vertigo, and asymmetrical hearing loss.

The glycerol dehydration test involves the ingestion of 1.5 g of the osmotically active glycerol per kg of bodyweight. This causes a transient improvement in hearing in the early stage of MD, that is absent in other causes of hearing loss [8]. However, the sensitivity of the test for MD is not significantly better than that of ECoG [9].

Vestibular evoked myogenic potential (VEMP) is a neurophysiological test which provides a measure of otolithic function. They have been shown to be abnormal in the majority of acute Meniere's episodes [10]. However, no VEMP test has been able to differentiate between MD and vestibular migraine, a major differential [11]. This maybe because they share a common pathophysiology. Furthermore, VEMPs may be similarly reduced in being paroxysmal positional vertigo (BPPV), vestibular neuronitis, and acoustic neuroma.

A new test has emerged for evaluating MD, cochlear hydrops analysis masking procedures (CHAMPs). This is an auditory brainstem response (ABR) test that is masked with high-pass noise. MD patients display a shortened latency in Wave V. A Californian group has been able to show with 100% sensitivity and 100% specificity a difference between controls and active MD patients displaying at least three of the four core symptoms [12]. However, later studies have failed to repeat such impressive results. A Belgian group performed an observational retrospective study, of 45 patients, 28 of whom had MD. 49% of the CHAMP results achieved were uninterpretable. Overall, the results yielded a sensitivity of 31% and specificity of 28%. Logistic regression showed that the CHAMP results did not contribute to the differentiation between EH, and normal ears [13].

Given the limitations of the diagnostic modalities discussed, there is opportunity for MRI to become the gold standard diagnostic modality if technological advance can lead to an improved specificity and sensitivity in detecting MD.

The cost of MD, both directly and indirectly, to the UK economy have been shown to be in excess of £500 million per annum [14]. Any improvements in diagnostics will save the healthcare system, and the economy money. Even in austere times, primary care trusts commissioning MRI to exclude other pathologies should not be challenged. At the very minimum it will continue to perform this role in the suspected MD case. The challenge of the future will be consolidate MRIs role in ruling out other diagnoses and to enhance it as a primary diagnostic tool.

Any radiological diagnoses rely on contrast between physiological and pathophysiological states. Most obviously this can represent changes in tissue size and structure, although recent advances have meant it can be subtle changes in blood flow or neural activity.

To understand whether or not MD could theoretically be imaged, requires a discussion of the pathophysiology. The cause of MD has been the source of much debate, both historically and in the present day. In 1938 it was suggested that the symptoms resulted from impaired blood flow, secondary to increases in the endolymphatic pressure that impaired circulation to the inner ear [15]. In the 1950s it was hypothesised that Reissner's membrane rupture caused contamination of perilymph with potassium rich endolymph, which inhibited hair cell transduction mechanism during Meniere's attacks. The authors speculated that the symptoms settled when the pressure reduced and the membrane healed [16]. However, the theory was refuted as the membrane could not heal within the time frame of an attack.

Since then a central theory of MD has developed in which swelling of the endolymph compartment, occurs either as a primary phenomenon or secondary to different insults. Once established the swelling of this compartment, 'endolymphatic hydrops (EH)' causes the symptoms observed, although the mechanism by which this occurs remains a source of scientific debate.

Studies have shown that delayed EH can occur in the contralateral temporal bone, to an inner ear, which appears to have been affected by a mumps or measles labyrinthitis. Thus they may represent a delayed sequela of inner ear damage in childhood [17]. An association also exists with allergy. Animal studies have shown that a provoked immune response can cause a reaction in the inner ear, and that prophylactic medication with leukotriene antagonists can prevent the development of EH [18].

However, some temporal bone studies have shown that EH have been found in asymptomatic patients [19]. This has led to speculation that EH may simply represent an epiphenomenon that occurs post-mortem. Given that EH can be clinically silent some have questioned the importance of being able to image them.

A comprehensive review of articles discussing MD and EH argues against this. The collective data included 541 hydropic temporal bones. It concluded that a person meeting the 1995 criteria for MD had a 'near certain' chance of having at least unilateral EH [20]. The development of EH follows a sequential and repeatable pattern even in asymptomatic cases. A meta - analysis of temporal bone MRI reports on 184 specimens found that the lesion distribution was orderly and cochleocentric. Disease always begins in the cochlear apex, and then progresses sequentially to involve the saccule, utricle, ampullae and canal system [21]. A study correlating MRI confirmed EH, with other tests of audiovestibular functional tests demonstrated a clear correlation of EH of the cochlea and saccule with signal voids from these organs [22]. Again this is suggestive of EH causing MD. The creation of EH animal models by dissecting the distal portion of the endolymphatic sac in guinea pigs, reduced the endocochlear potential [23]. Similarly, the size of the EH in guinea pig models correlated with severity of hearing loss [24]. All this data is suggestive of a direct causative link with the EH, and not an incidental phenomena.

Whatever the underlying mechanism of development, the association of EH with MD is indisputable, and the ability to confidently identify EH by imaging alone would represent a significant progress in MD diagnostics.

Early attempts to achieve this looked to do so indirectly by looking at the width of the vestibular aqueduct, in which the endolymphatic sac runs, reasoning that its narrowing may lead to obstruction and consequently EH. Since the 1980s, work has compared the MRI findings of temporal bone specimens of Meniere's sufferers and controls. One such study found statistically significantly smaller vestibular aqueducts in patients with EH [25].

Similarly CT studies have shown differences between the temporal bones of live MD and non-MD patients. The

retrolabyrinthine width was found to be narrower in MD (3.8 mm vs 5.8 mm). More significantly there was significantly reduced visualisation of the vestibular aqueduct in the MD group [26]. Recent experiments with more advanced MRI resolution showed that the distance from the posterior semicircular canal to the posterior temporal border was bilaterally reduced in MD [27]. However, the authors of even the more recent experiments concede that these subtle differences between the hard tissues of the temporal bone are too crude and inconsistent for consistent diagnostics.

Another possible radiological clue is the height of the jugular bulb. Theoretically the swelling of the endolymphatic compartment should push the jugular bulb superiorly which could be detected on imaging studies. However, in a recent study of the prevalence of a high riding jugular bulb was only slightly more frequently in patients with EH than controls 9.0%, versus 4.5%. The authors concluded that a HRJB does not play a major role in EH [28].

In the 1990s improved MRI resolution led the soft tissue of the endolymphatic duct being the direct target of imaging studies. Submillimeter resolution showed endolymphatic duct and sac with greater resolution. Visualisation of the endolymphatic duct was found to be less frequent in the MD patient than in controls [29,30].

Further progress has been made by injecting contrast into the inner ear. It was first demonstrated that when gadolinium based contrast agents (GBCA), were applied to the round window in guinea pigs improved the resolution of the endolymphatic and perilymphatic spaces [31].

Soon after this was trialled in humans in experiments in Japan. GBCA, can be injected intratympanically through the TM using needle into the middle ear space. After a period to allow uptake into the perilymphatic space. Patients with EH could be identified indirectly by showing that the perilymphatic space surrounding the endolymph was small or non-existent [32]. Using this technique concentrations of 0.1 mmol/L can be achieved in the perilymphatic space. Other investigators have applied contrast through the Eustachian Tube in an effort to avoid any of the complications of tympanic perforation, achieving similarly impressive results.

Improved resolution have been made by using IV contrast materials along with 3D fluid attenuation inversion recovery (3D-FLAIR) MRI images. Using such 3D maximum intensity projections a Japanese group have been able to show that 3D maximum intensity projections of EH size, were strongly correlated with severity of hearing impairment [33].

Further experiments on a cohort group of 41 patients who fulfilled the criteria for definite unilateral MD, by the AAO-HNS guidelines showed strong correlation between the size of the EH detected by the MRI scans and the degree of canal paresis detected by caloric tests. This shows that not only does MRI have the ability to correctly identify EH, but it is accurate enough to predict symptomatology [34].

Given the recent progress which has been made in MRI imaging it would seem natural to compare its accuracy to that of other diagnostic tests for EH. The results of these comparisons have been mixed.

Several direct comparative study has attempted to compare the sensitivity of EcoG and MRI to detection of EH. Tone burst EcoG positively detected EH in 25 cases (83%), whereas gadolinium MRI was positive in (47%). The authors conclude that EcoG was a more sensitive test [35].

Another compared the diagnostic value of 3T MRI after intratympanic injection of gadolinium based contrast agent (GBCA), with electrocochleography. The MRI was able to identify EH in 95% of the cases, compared 60% of ECoG, and 55% in the case of the glycerol dehydration test [36].

Further recent studies have been even more promising. An Italian group set out to evaluate the reliability of MRI after intratympanic gadolinium. They compared 26 patients with definite MD with 12 controls who had different unilateral non-MD inner ear disorders. 100% of the MD cohort showed reduced perilymphatic enhancement on the scans of the affected ears. All the unaffected contralateral ears failed to show similar changes. Similarly in 11 of the 12 controls, there was no change in the perilymphatic enhancement [37].

The same group have produced work showing that the changes in perilymphatic enhancement in these experiments is proportional to the length of time the patient has suffered with MD. This suggests that EHs is a progressive phenomenon [38].

Having established that imaging has the potential to identify EH further questions arise. Could MRI ever reach a point when it could say that the EH identified was quiescent or responsible for the patients symptoms? A recent hypothesis by Foster may give an insight into how this might be achieved. They have suggested that a Meniere's attack may represent an ischaemic episode for the inner ear, which resolves when the ear is reperfused. According to the theory EH develop and act as a Starling resistor. Initially they are asymptomatic, as oxygen levels remain adequate. However, when combined with cerebrovascular disease in later life, the attacks start. The sensitivity of the stria to ischaemia explains why tinnitus, and hearing loss are typical symptoms of an attack [39]. By combining the improved resolution of MRI with an intravenous contrast agent, inner ear perfusion could be monitored in the ear with EH to study disease progression, and guide further management. However, this is in the realms of scientific speculation.

In conclusion, MDs transition to becoming a radiological diagnosis would represent a 'paradigm shift'. Vertiginous patients are notoriously difficult to manage, with recent data showing that 20% do not receive a definitive diagnosis [40]. They often fall between the specialties of otolaryngology, neurology and audiovestibular medicine, all of whom would welcome this advance. Once established reliance on less reliable tests such as electrocohleography would be reduced and MD could be increasingly diagnosed and managed in primary care. For patients it would mean appropriate management could be facilitated rapidly, improving long term prognosis. The advances in imaging will further advance understanding of the pathophysiology, and may eventually help predict which patients

will respond best to the medical and surgical interventions currently available [41-48].

References

1. Gimsing S (2010) Vestibular schwannoma: when to look for it? The Journal of laryngology and otology 124: 258 -264.
2. Weissman JL (1997) Imaging of Meniere's disease. Otolaryngologic Clinics of North America 30: 1105-1116.
3. Park JH, Kim H, Han, HJ (2008) Recurrent audiovestibular disturbance initially mimicking Ménière's disease in a patient with anterior inferior cerebellar infarction. Neurological sciences 29: 359-362.
4. Enander A, Stahle J (1967) Hearing in Meniere's disease: a study of pure-tone audiograms in 334 patientsActa oto-laryngologica 64: 543-556.
5. Wu Z, Zhang S, Zhou N, Yi F, Chen A, et al. (2006) Significance of some otologic function tests in diagnosis of Meniere's disease. Journal of clinical otorhinolaryngology 20: 433-435.
6. Kim HH, Kumar A, Battista RA, Wiet RJ (2005) Electrocochleography in patients with Meniere's disease. American journal of otolaryngology 26: 128-131.
7. Adams ME, Kileny PR, Telian SA, El-Kashlan HK, Heidenreich KD, et al. (2011) Electrocochleography as a diagnostic and intraoperative adjunct in superior semicircular canal dehiscence syndrome. Otol Neurotol 32: 1506-1512.
8. Klockhoff I, Lindblom U (1967) Glycerol test in Meniere's disease. Acta oto-laryngologica 63: 449-451.
9. Lu JZ, Zhang JG, Lai H (2000) The relationship between ECochG and glycerol test in vertigo patients (report of 112 cases). Journal of clinical otorhinolaryngology 14: 510-511.
10. Kuo SW, Yang TH, Young YH (2005) Changes in vestibular evoked myogenic potentials after Meniere attacks. Annals of Otology, Rhinology & Laryngology 114: 717-721.
11. Zuniga MG, Janky KL, Schubert MC, Carey JP (2012) Can vestibular-evoked myogenic potentials help differentiate Meniere disease from vestibular migraine? Otolaryngol Head Neck Surg 146: 788-796.
12. Don M, Kwong B, Tanaka C (2005). A diagnostic test for Ménière's disease and cochlear hydrops: impaired high-pass noise masking of auditory brainstem responses. Otology & Neurotology 26: 711-722.
13. De Valck CF, Claes GM, Wuyts FL, Van de Heyning PH (2007) Lack of diagnostic value of high-pass noise masking of auditory brainstem responses in Ménière's disease. Otol Neurotol 28: 700-707.
14. Tyrrell J, Whinney DJ, Taylor T (2016) The Cost of Meniere's Disease: A Novel Multisource Approach. Ear Hear 37: e202-e209.
15. Hallpike CS, Cairns H (1938) Observations on the pathology of Meniere's syndrome. The Journal of Laryngology & Otology 53: 625-655.
16. Lawrence M, McCabe BF (1959) Inner-ear mechanics and deafness: special consideration of Ménière's syndrome. Journal of the American Medical Association 171: 1927-1932.
17. Schuknecht HF, Suzuka Y, Zimmermann C (1990) Delayed endolymphatic hydrops and its relationship to Meniere's disease. Annals of Otology, Rhinology & Laryngology 99: 843-853.
18. Weinreich HM, Agrawal Y (2014) The link between allergy and Menière's disease. Curr Opin Otolaryngol Head Neck Surg 22: 227.
19. Merchant SN, Adams JC, Nadol Jr JB (2005) Pathophysiology of Meniere's syndrome: are symptoms caused by endolymphatic hydrops? Otology & Neurotology 26: 74-81.
20. Foster CA, Breeze RE (2013) Endolymphatic hydrops in Ménière's disease: cause, consequence, or epiphenomenon? Otology & Neurotology 34: 1210-1214.
21. Pender DJ (2014) Endolymphatic hydrops and Ménière's disease: a lesion meta-analysis. The Journal of Laryngology & Otology 128: 859-865.
22. Seo YJ, Kim J, Choi JY, Lee WS (2013) Visualization of endolymphatic hydrops and correlation with audio-vestibular functional testing in patients with definite Meniere's disease. Auris Nasus Larynx 40: 167-172.
23. Warmerdam TJ, Schröder FHHJ, Wit HP, Albers FWJ (2001) Perilymphatic and endolymphatic pressure in the guinea pig after distal dissection of the endolymphatic sac. Otology & Neurotology 22: 373-376.
24. Hott ME, Graham M, Bonassar LJ, Megerian CA (2003) Correlation between hearing loss and scala media area in guinea pigs with long-standing endolymphatic hydrops. Otology & neurotology 24: 64-72.
25. Sando I, Ikeda M (1984) The vestibular aqueduct in patients with Meniere's disease: a temporal bone histopathological investigation. Acta oto-laryngologica 97: 558-570.
26. Nidecker A, Pfaltz CR, Matefi L, Benz UF (1985) Computed tomographic findings in Meniere's disease. ORL 47: 66-75.
27. Lorenzi MC (2000) Magnetic resonance imaging of the temporal bone in patients with Meniere's disease. Acta oto-laryngologica 120: 615-619.
28. Brook CD, Buch K, Kaufmann M, Sakai O, Devaiah AK (2015) The Prevalence of High-Riding Jugular Bulb in Patients with Suspected Endolymphatic Hydrops. J Neurol Surg B Skull Base 76: 471-474.
29. Welling DB, Clarkson MW, Miles BA, Schmalbrock P, Williams PM, et al. (1996) Submillimeter magnetic resonance imaging of the temporal bone in Meniere's disease. The Laryngoscope 106: 1359-1364.
30. Tanioka H, Machida T, Sasaki Y, Zusho H, Shirakawa T (1992) High-resolution MR imaging of the inner ear: findings in Menière's disease. Eur J Radiol 15: 83-88.
31. Zou J, Pyykkö I, Bjelke BOR, Dastidar P, Toppila E (2005) Communication between the perilymphatic scalae and spiral ligament visualized by in vivo MRI. Audiology and Neurotology 10: 145-152.
32. Nakashima T, Naganawa S, Sugiura M, Teranishi M, Sone M, et al. (2007) Visualization of endolymphatic hydrops in patients with Meniere's disease. The Laryngoscope 117: 415-420.
33. Sepahdari AR, Ishiyama G, Vorasubin N, Peng KA, Linetsky M, Ishiyama A (2015) Delayed intravenous contrast-enhanced 3D FLAIR MRI in Meniere's disease: correlation of quantitative measures of endolymphatic hydrops with hearing. Clinical imaging 39: 26-31.
34. Gürkov R, Flatz W, Louza J, Strupp M, Ertl-Wagner B, et al. (2012) In vivo visualized endolymphatic hydrops and inner ear functions in patients with electrocochleographically confirmed Ménière's disease. Otology & Neurotology 33: 1040-1045.

35. Hornibrook J, Flook E, Greig S, Babbage M, Goh T, et al. (2015) MRI Inner Ear Imaging and Tone Burst Electrocochleography in the Diagnosis of Méniere's Disease. Otology & Neurotology 36: 1109-1114.

36. Fukuoka H, Takumi Y, Tsukada K, Miyagawa M, Oguchi T, et al. (2012) Comparison of the diagnostic value of 3 T MRI after intratympanic injection of GBCA, electrocochleography, and the glycerol test in patients with Meniere's disease. Acta oto-laryngologica, 132: 141-145.

37. Fiorino F, Pizzini FB, Beltramello A, Barbieri F (2011) Progression of endolymphatic hydrops in Ménière's disease as evaluated by magnetic resonance imagingOtology & Neurotology 32: 1152-1157.

38. Fiorino F, Pizzini FB, Beltramello A, Mattellini B, Barbieri F (2011) Reliability of magnetic resonance imaging performed after intratympanic administration of gadolinium in the identification of endolymphatic hydrops in patients with Meniere's disease. Otology & Neurotology 32: 472-477.

39. Foster CA, Breeze, RE (2013) The Meniere attack: An ischemia/reperfusion disorder of inner ear sensory tissues. Medical hypotheses 81: 1108-1115.

40. Post RE, Dickerson LM (2010) Dizziness: a diagnostic approachAm Fam Physician 82: 361-368.

41. Ishiyama G, Lopez IA, Sepahdari AR, Ishiyama A (2015) Meniere's disease: histopathology, cytochemistry, and imaging. Annals of the New York Academy of Sciences 1343: 49-57.

42. Le CH, Truong AQ, Diaz RC (2013) Novel techniques for the diagnosis of Ménière's disease. Current opinion in otolaryngology & head and neck surgery 21: 492-496.

43. Miller ME, Bykowski J (2014) Imaging Analysis of Ménière's Disease. Current Otorhinolaryngology Reports 2: 152-161.

44. Nakashima T, Naganawa S, Katayama N, Teranishi M, Nakata S, et al. (2009) Clinical significance of endolymphatic imaging after intratympanic gadolinium injection. Acta Oto-Laryngologica 129: 9-14.

45. Nakashima T, Naganawa S, Pyykkö I, Gibson WP, Sone M, et al. (2009) Grading of endolymphatic hydrops using magnetic resonance imaging Acta Oto-Laryngologica 129: 5-8.

46. Pyykkö I, Zou J, Poe D, Nakashima T, Naganawa S (2010) Magnetic resonance imaging of the inner ear in Meniere's disease. Otolaryngologic Clinics of North America 43: 1059-1080.

47. Shang YY, Diao WW, Ni DF, Gao ZQ, Li FR (2012) Study of cochlear hydrops analysis masking procedure in patients with Meniere's disease and otologically normal adults. Chinese medical journal 125: 4449-4453.

48. Vassiliou A, Vlastarakos PV, Maragoudakis P, Candiloros D, Nikolopoulos TP (2011) Meniere's disease: Still a mystery disease with difficult differential diagnosis. Ann Indian Acad Neurol 14: 12-18.