Zagazig University Medical Journal
www.zumj.journals.ekb.eg

Volume 29, Issue 1, January 2023, Page (9-13) Supplement Issue

Manuscript ID ZUMJ-1907-1297 (R2)
DOI **10.21608/zumj.2020.14235.1297**

TYPE OF THE PAPER (REVIEW ARTICLE)

Idiopathic Endolymphatic Hydrops (Meniere's Disease)

Ebtessam Nada*

Ear, Nose and Throat Department, Faculty of Medicine, Zagazig University, Egypt.

***Corresponding author:**
Name: Ebtessam Nada
Ear, Nose and Throat Department, Faculty of Medicine, Zagazig University, Egypt.
E-mail: e_hossam1999@yahoo.com

Submit Date 2019-10-14
Revise Date 2020-07-22
Accept Date 2020-09-09

ABSTRACT

Background: Meniere's disease is a disorder of the inner ear that is characterized by a collection of episodically occurring symptoms: sensorineural hearing loss (SNHL) "fluctuating in nature", aural fullness, tinnitus "roaring", and spinning vertigo. Incidence was reported to be between 7 to upwards of 150 per 100,000 persons. There is no racial or sex difference. Peak incidence is between 40-60 years old and average age of onset is usually 50 years.
Diagnosis is made by clinical picture of the characteristic tirade which has to consider atypical forms also, objective measures like, Echo G, VNG and VEMP and of course basic audiological evaluation of the degree and type of hearing loss and configuration. Management includes dietary treatment with Meniere's diet, medications like diuretics or steroids, surgical intervention with hearing preservation procedures or even labyrinthectomy or sedatives.
Conclusion: Meniere's disease is a considerable diagnosis to make and for better quality of life all measures of treatments have to be considered.

Keywords: Meniere's disease

INTRODUCTION

Meniere's disease is a disorder of the inner ear that is characterized by a collection of episodically occurring symptoms: sensorineural hearing loss (SNHL) "fluctuating in nature", aural fullness, tinnitus "roaring", and spinning vertigo [1]. The pathological correlate of this inner ear disorder is hydrops of the endolymphatic space [2]. Meniere's disease was named after the French neurologist "Prosper Menière", who discussed in 1861 that episodic vertigo and hearing loss are symptoms that could be caused by a disorder in the inner ear [3]. Although it varies significantly in different countries it has been reported between 7 to upwards of 150 per 100,000 persons. There is no racial or sex difference. Peak incidence is between 40-60 years old and average age of onset is usually 50 years. It is rare in children below 10 years and adults above 60 years in life however; symptoms have been reported occasionally in children 4 years old and in patients > 90 years [4- 6]. Female-to-male distribution is equal 9 or slightly biased toward female patients (1.3:1- 1.9:1 female predominance). Family history is positive for Meniere's in 10- 20 %, genetic transmissions may play a role, inheritance is multifactorial [7, 8]. Right and left ears are equally affected although the disease is usually unilateral and tends to involve the contralateral ear in 30% of cases [9]

Bilateral occurrence was reported in only 5-45% of cases [4- 6]. And 72% of patients who developed bilateral Meniere did so within 5 years of diagnosing the 1st ear. The hydrops can be unilateral or bilateral with affection of both the cochlear duct and saccule in most cases, whereas, the utricular and semicircular ducts are usually spared. In other cases the cochlear duct alone is affected. Another less frequent and debatable form of Ménière's that affects the vestibule only where symptoms are those of vertigo only and no hearing loss. The elastic Reissner's membrane of the hydropic duct bulges variably to reaches the top of the scala vestibuli in the most severe cases and may be in contact with a wide area of cochlear wall. In the apical region it may bulge to fill the helicotrema or even the distended scala media may enter the scala tympani. The saccule is swollen and rises up from its position on the medial wall of the vestibule to touch the vestibular surface of the footplate of the stapes and in some cases it may herniate from the vestibule into the semicircular canals. The utricle may be compressed and less frequently may be distended with small infoldings producing a scalloped appearance [10]. Ruptures are seen in the walls of the distended thin membranes of the hydropic endolymphatic spaces which may be present particularly in Reissner's membrane with curling up of the terminal end [11]. Outpouchings

are seen in which dilatation of part of the wall of the membranous labyrinth occurs with a lining that is thinner than elsewhere. These outpouchings may be healed ruptures, or simply areas of increased distension of parts of the labyrinthine wall, which are originally thinner normally this is supported by their regular features [12].The presence of fibrous tissue external to the endolymphatic space has been described in the scala vestibuli and in the vestibule deep to the footplate of the stapes. These foci of connective tissue in these two situations are reactions to the irritation produced by repeated distension and subsidence of the adjacent coclear duct and saccule respectively [13]. As regards pathophysiology of Meniere's, distortion of the membranous labyrinth which is resulted from increasing accumulation of endolymph is presumed. Endolymphatic hydrops per se can be thought of as a marker of the disease rather than a cause as proved by a study looking at temporal bones which found that all patients with Ménière's disease had hydrops in at least 1 ear but that hydrops was also found in some patients with no signs of the disease [14]. Hydrops is a postmortem finding after various extrinsic mechanisms were thought to contribute to its development including infection like labyrinthitis, mumps or meningitis and otitis media, head trauma, and allergens [15]. The endolymph and perilymph are separated by thin membranes that house the neural apparatus of hearing and balance. Increase in endolymphatic pressure, causes break in this membrane and the resultant chemical mixture bathes the receptors of the vestibular nerve, leading to a depolarization blockade and transient loss of function. The sudden change in the rate of vestibular nerve firing creates an acute vestibular imbalance (vertigo). Also, stressing these nerve-rich membranes by fluctuations in pressure, causes hearing disturbance, tinnitus, and a pressure sensation in the ear.For diagnosis, the American Academy of Otolaryngology–Head and Neck Surgery Foundation (AAO-HNS) Committee on Hearing and Equilibrium" published the clinical diagnosis guidelines of Ménière disease in 1972, 1985, and 1995 [16]. Based on these guidelines, Ménière disease was defined as "recurrent, spontaneous episodic vertigo; hearing loss; tinnitus; and aural fullness. Presence of tinnitus or aural fullness (or both) has to be present on the affected side to make the diagnosis [16].Less than one third of patients present with all the symptoms of the disease at onset having vertigo as the most common initial component and additional components appear after a variable time delay of months to years [17, 18]. Two definitive attacks of vertigo (rotatory) of at least 20 minutes are necessary to make the diagnosis [16]. Onset of the attacks in 50% of cases is preceded by aura of ear fullness and tinnitus in other cases. After the acute attacks, patients may feel tired, unsteady, and nauseated for variable periods of time where in-between the attacks some patients are completely symptom free and others may experience disequilibrium, light-headedness or tilt [17, 18]. Attacks often occur in clusters separated by long remissions with many notice progressive deterioration of hearing and balance function with each successive attack [19]. Associated symptoms include nausea, vomiting, diarrhea and sweating. Attacks are exacerbated by head movement. Consciousness is present with NO neurological manifestation.Atypical forms describe patients who complain of SOME but NOT the classical symptoms of the triad. They involve; Atypical Meniere's (cochlear Meniere's) or (Meniere without vertigo) [20] which is Characterized by fluctuating SNHL and 80% of these cases will develop classical Meniere later on. Similarly, recurrent vestibulapathy (vestibular Meniere's) is another form and describes recurrent attacks of vertigo similar to Meniere but without hearing loss or tinnitus or aural fullness. Only 20 % of these patients may develop Meniere [21, 22]. Lastly, sudden falls with no loss of consciousness "Tumarkin crises" or drop attacks [23] due to acute utriculosaccular dysfunction. (Incidence ranges from less than 10% in some studies up to 72% in other studies) [19]. Diagnosis involves; in the first place clinical examination which reveals in patient with severe vertigo, significant distress and signs of recent vomiting. Patients are sometimes diaphoretic, pale and vital signs may show elevated blood pressure, pulse, and respiration [15]. However, between attacks or during remission, physical examination is completely normal, particularly if the patient is symptom free. Audiologically, low-frequency, fluctuating sensorineural hearing loss is present especially early in the disease. Overtime hearing loss flattens and become less variable but profound deafness is rare, occurs in about 1-2%. If there is coincident non-changing high-frequency loss called peaked or tent-like audiogram and the peak is classically at 2 kHz. Hearing deteriorates essentially within the 1st years of the disease thereafter stabilization occurs. Objectively, Ecoh G measures the ratio of the summating potential (SP) (probably from the movement of the basilar membrane) and the nerve action potential in response to auditory stimuli. Distention of basilar membrane into the scala tympani causes an increase in the normal asymmetry of its vibration and this leads to a larger and more negative SP. Hydrops (elevated pressure) is suggested when this ratio is greater than 35%. The test is most accurate when Ménière disease is active [15].Furthermore, ENG and VNG testing

reveals reduced caloric response in 42- 79% of patients with unilateral Meniere's disease and absent caloric response (100% asymmetry) in 6-11% [24-25]. Increased response secondary to an irritative lesion may be seen and the patient may feel dizzy or nauseated [15]. On the other hand vHIT shows abnormalities in only 13% of cases [24]. Normal angular vestibule-ocular reflex (AVOR) with abnormal caloric response suggests a substantial preservation of semicircular canal function in those patients with normal AVOR [26]. This presents a preferential impairment of the ability of the vestibular system to process low frequency signals [27]. In early stages of the disease, both caloric and AVOR are normal while during acute attack caloric asymmetry is present with increased gain of the AVOR in vHIT test when rotating toward the affected side which indicates that the affected ear may act like a filter "high-pass" that dampens sensitivity to low frequency stimuli and enhances sensitivity to high frequency stimulation [28]. cVEMP or oVEMP both can be used to diagnose Meniere disease but with suspected efficacy. The logic about Meniere's and VEMP's is as follows: Meniere's damage to the ear might involve the input systems for the VEMP (i.e. saccule, utricle, semicircular canals, hearing). Meniere's can dilate the saccule/utricle, and perhaps make them more sensitive than in normal persons to stimulation. Lab Studies usually are not needed to make the diagnosis of Meniere's but they may be needed to delineate other pathologies [15] and to exclude obvious metabolic disturbances, infections, or hormonal imbalances and 2nd endolymphatic Hydrops. For management, the American Academy of Otolaryngology- Head and Neck Surgery (AAO-HNS) [1] has developed formal guidelines for the results in the treatment of Meniere's disease taken into consideration the fact that there is a considerable placebo effect present.

Dietary management aims at stabilizing the body's fluid and electrolyte levels which may result in affection of the amount and composition of the endolymph, leading to hydrops symptoms [29]. Constancy of this regimen is important and involves moderate amount, balanced, regular meals at about the same time every day, without skipping meals.Reduction of salt and sugar intake in diet and avoid eating prepared foods is implemented with adequate fluid intake. Low-sugar sodas, herbal teas, and low-sugar fruit and vegetable juices are required. Caffeine (found in coffee, tea, some herbal teas, colas, chocolate, and some medications) and alcohol may need to be restricted as have strong diuretic properties [29]. Medically, diuretics or the combination of hydrochlorothiazide and triamterene is the most commonly used. Acetazolamide, furosemide, and spironolactone are also used by some. Combining these with sodium restriction has slowed progression of hearing loss [30]. Betahistine has also been used extensively for this condition owing to its vasodilatation effect with inconsistent response [31]. Oral steroids have been used also for treatment to reduce the severity of spells. A study using 18 months of steroid treatment reported a 50% improvement in vertigo spell "frequency" [32]. Lastly, surgical management involves; endolymphatic sac decompression which is both a functional and a hearing preservation procedure that was first described by George Portmann in 1926 [33, 34] and vestibular nerve section (vestibular neurectomy) which represents an intracranial hearing preservation (deafferentative procedure) which was popularized by House in 1961 [35]. The operation of vestibular neurectomy is performed via a middle fossa approach with selective section of the vestibular nerve. In good hands there appears to be a 95 % control of the vertigo. Others have tried different access routes including retrosigmoid and retrolabyrinthine approaches [36-37]. Complications of these operations include CSF leakage, facial palsy, hearing loss, meningitis and subdural haematoma.

Intratympanic therapy is a less invasive method for the deafferentation of vestibular function. Intratympanic steroids have been used as a calming effect on Ménière's ears [38]. The number and severity of spells decreases for up to 2 years after use [39]. Conversely, aminoglycosides such as gentamicin are known to be ototoxic but appear preferentially vestibulotoxic are used as chemical vestibular ablation [40, 41].

In those with incapacitating vertigo and when hearing preservation is not a concern then labyrinthectomy should be considered, a transmastoid total osseous labyrinthectomy has been favored by many authors. Vertigo control is achieved in more than 95% of patients. Finally, Suppressant medications benzodiazepines, meclizine, other antihistamines such as promethazine or diphenhydramine, and transdermal scopolamine patches have all been used to control the sensation of vertigo during acute attacks. Transdermal scopolamine patches require 4 hours to reach a therapeutic blood level and continue to release drug for 72 hours which is a poor match for typical Ménière's attacks of sudden onset and duration of a few hours [42]. Meclizine is often useful to reduce nausea for milder spells, oral benzodiazepines are well tolerated for more severe spells, and promethazine suppositories can be used for the most severe spells with intractable vomiting.

CONCLUSION

Dealing with Meniere's is an important issue to

study, knowing pathogenesis, pathophysiology and clinical picture is important to distinguish and treat.

-Conflict of Interest: None.

-Financial Disclosures: None

REFERENCES

1. The American Acadademy of Otolaryngology Committee on Hearing and Equilibrium and the American Council of Otolaryngology on the medical aspects of noise. Guide for the evaluation of hearing handicap. J Am Med Ass 1979; 241: 2055–59.
2. Committee on Hearing and Equilibrium guidelines for the diagnosis and evaluation of therapy in Meniere's disease: American Academy of Otolaryngology-Head and Neck Foundation, Inc. Otolaryngol Head Neck Surg 1995;113:181–85
3. Baloh RW. Prosper Meniere and his disease. Arch Neurol. 2001; 58(7):1151–6.
4. Perez R, Chen JM and Nedzelski JM. The status of the contralateral ear in established unilateral Meniere's disease. Laryngoscope, 2004; 114: 1373-6.
5. Katsarkas A. hearing loss and vestibular dysfunction Meniere's disease. Acta otolaryngol 1996. 116: 185-8.
6. Green JD, Blum DJ and Harner SG. Longitudinal follow up of patients with Meniere's disease. Otolaryngol Head Neck Surg, 1991; 104: 783- 8.
7. Hamid M and Sismanis A. Peripheral auditory and vestibular disorders. In Medical Otology and Neuriotology: A clinical guide to auditory and vestibular disorders. Thieme medical publisher. 2006. 6: 64-85.
8. Harris JP and Alexander, TH. Current day prevalence of Meniere's syndrome. Audiolo Neurootol, 2010; 15 (5): 318-22.
9. Shojaku H, Watanabe Y, Fujisaka M et al. epidemiologic charecterisitcs of definite Meniere's disease in Japan. A long term survey of Toyama and Nigata perfectures. ORL J Otorhinolaryngol Relat Epec, 2005; 776 (5): 305-9.
10. Thomas K and Harrison MS. Long term follow up of 610 cases of Meniere's disease. Proc R Soc Med, 1071; 64 (8): 853-7.
11. KOHUT RI, LINDSAY JR. Pathologic changes in idiopathic labyrinthine hydrops. Correlations with previous findings. Acta Otolaryngol 1972; 73: 402-12.
12. Schuknecht HF. Pathology of the ear. Cambridge (Mass): Harvard University Press 1974.
13. HALLPIKE CS, CAIRNS H. Observation on the pathology of Meniere's disease. J Laryngol Otol 1938; 53: 625-35.
14. Salvinelli, F. Greco, M. Trivelli, F.H. Linthicum JR. Ménière's disease. Histopathological changes: A post mortem study on temporal bones. European Review for Medical and Pharmacological Sciences. 1999; 3: 189-93.
15. Monsell EM et al. Committee on hearing and equilibrium guidelines for the diagnosis and evaluation of therapy in Meniere's disease. Otolaryngol Head Neck Surg.; (1995); 181–5.
16. Merchant SN, Adams JC, Nadol JB Jr. Pathophysiology of Meniere's syndrome: are symptoms caused by endolymphatic hydrops?. *Otol Neurotol*. 2005 Jan. 26(1):74-81.
17. John C Li. Meniere Disease (Idiopathic Endolymphatic Hydrops) Jun 08, 2018 Chief Editor: Nicholas Lorenzo, MD, https://www.medscape.com/cardiology.
18. Mancini F, Catalani M, Carru M, Monti B. History of Meniere's disease and its clinical presentation. Otolaryngol Clin North Am. 2002 Jun. 35(3):565-80.
19. Paparella MM. Benign paroxysmal positional vertigo and other vestibular symptoms in Ménière disease. *Ear Nose Throat J*. 2008 Oct. 87(10):562.
20. Kentala E, Havia M, Pyykkö I. Short-lasting drop attacks in Meniere's disease. Otolaryngol Head Neck Surg. 2001 May. 124(5):526-30.
21. Park, HJ Migliaccio, AA Della Santina CC Minor L Carey JP. Search-coil head thrus and aloric tests in Meniere's disease. 2005. Acta Otolaryngol, 125; 852-7.
22. Hone SW. Nedzelski, J and Chen J. does intratympanic gentamycin treatment for Meniere's disease cause complete vestibular ablation?. J Otolaryngol.2000; 29 (2): 83-7.
23. Carey JP, Minor LB, Peng GC, Della SatinaCC, Cremer, PD and Haslwanter T. changes in the three-dimensyional angular vestibule-ocular refelex following intratympanic gentamycin injection for Meniere's disease. J Assoc Res Otolaryngol.2002; 3(4): 403-43.
24. VertigoKoichi Yasuda, Atsuko Kitajima. Patients with Meniere's Disease without vertigo. Practica Oto-Rhino-Laryngologica, 1980 Volume 73 Issue 11special Pages 1949-54.
25. Shi Nae Park, Kyoung Ho Park, Dong Jae Im, Jong Hoon Kim, Jun Yop Kim, Sang Won Yeo. Recurrent Vestibulopathy: Clinical Characteristics and Efficacy of Combination Therapy. J Korean Bal Soc.2006; 5(2); 262-8.
26. LeLiever WC, Barber HO: Recurrent vestibulopathy, Laryngoscope 1981; 41:1.
27. Yuri Agrawal and Lioyd B M inor. Meniere's disease and other caused of episodic vertigo. In Bronstein AM "vertigo and imbalance". Oxford University press UK.2013; Ch 22 pp 241-250.
28. Maire, R and van Melle G. vestibule-ocular reflex charectaristics in patients with unilateral Meniere's disease. Otol Neurol.2008; 29 (5): 693-8.
29. Morales-Luckie E, Cornejo-Suarez A, Zaragoza-Contreras MA, Gonzalez-Perez O. Oral administration of prednisone to control refractory vertigo in Ménière's disease: a pilot study. Otol Neurotol. 2005; 26(5):1022–6.
30. Susan Pesznecker, RN. Vestibular Disorders Association https://vestibular.org/secondary-endolymphatic-hydrops-seh
31. Santos PM, Hall RA, Snyder JM, Hughes LF, Dobie RA. Diuretic and diet effect on Ménière's disease evaluated by the 1985 Committee on Hearing and Equilibrium guidelines. Otolaryngol Head Neck Surg. 1993;109(4):680–9

32. Ihler F, Bertlich M, Sharaf K, Strieth S, Strupp M, Canis M. Betahistine exerts a dose-dependent effect on cochlear stria vascularis blood flow in Guinea pigs in vivo. PLoS One. 2012; 7(6):e39086.

33. Portmann G. Vertigo—surgical treatment by opening the saccus endolymphaticus. Arch Otolaryngol 1927; 6: 309–15.

34. Thomsen J and others: Placebo effect in surgery for Meniere's disease, a double blind, placebo-controlled study on endolymphatic sac shunts surgery, Arch Otolaryngol 1981; 107:271.

35. House WF: Surgical exposure of the internal auditory canal and its contents through the middle cranial fossa, Laryngoscope 1961; 71:1363. 20. Silverstein H, Norrell H, Rosenberg S: The resurrection of vestibular neurectomy: a 10-year experience with 115 cases, J Neurosurg 1990; 72:533.

36. Silverstein H, Norrell H, Haberkamp T: A comparison of retrosigmoid IAC, retrolabyrinthine, and middle rossa vestibular neurectomy for treatment of vertigo, Laryngoscope 1987; 97:165.

37. Nedzelski JM and others: Intratympanic gentamicin instillation as treatment of unilateral Meniere's disease: update of an ongoing study, Am J Otol 1993; 14:278.

38. Nedzelski J and others: Chemical labyrinthectomy: local application of gentamicin for the treatment of unilateral Meniere's disease, Am J Otol 1992; 13:18.

39. Densert B, Sass K. Control of symptoms in patients with Meniere's disease using middle ear pressure applications: two year follow-up. Acta Otolaryngol 2001; 121:616- 21.

40. Nedzelski J and others: Chemical labyrinthectomy: local application of gentamicin for the treatment of unilateral Meniere's disease, Am J Otol 1992; 13:18.

41. Gates GA, Green JD, Tucci DL, Telian SA. The effects of transtympanic micropressure treatment in people with unilateral Meniere's disease. Arch Otolaryngol 2004; 130:718-25.

42. Antor MA, Uribe AA, Erminy-Falcon N, et al. The effect of transdermal scopolamine for the prevention of postoperative nausea and vomiting. Front Pharmacol. 2014;5:55

To Cite:

Nada, E., Idiopathic Endolymphatic Hydrops (Meniere's Disease). *Zagazig University Medical Journal*, 2023; (9-13): -.doi: 10.21608/zumj.2020.14235.1297.