

Meniere's Disease (A Clinicoaudiological Prospective Study)

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Abstract

Background: Meniere's disease (MD) is a disease of the inner ear that causes vertigo, fluctuating hearing loss, tinnitus and aural fullness. The etiology is multifactorial. A characteristic sign of MD is endolymphatic hydrops, in which excessive endolymph accumulates in the inner ear and causes damage to the ganglion cells. The diagnosis of MD is based on clinical symptoms and complemented with functional inner ear tests, including audiometry, vestibular-evoked myogenic potential testing. MRI has been optimized to directly visualize EH in the cochlea, vestibule and semicircular canals, and its use is shifting from the research setting to the clinic. The management of MD is mainly aimed at the relief of acute attacks of vertigo and the prevention of recurrent attacks. Therapeutic options are based on empirical evidence and include the management of risk factors and a conservative approach as the first line of treatment. When medical treatment is unable to suppress vertigo attacks, intratympanic gentamicin therapy or endolymphatic sac decompression surgery is usually considered.

Objectives: To throw some light on Meniere's disease in Basra and to determine the diagnostic value of cVEMP and PTA in patients with Meniere's disease and their relation to the clinical features.

Patients and method: It is a clinical, descriptive, prospective cross sectional study carried out from October 2019 to February 2021 at Basra otolaryngology center in Basra teaching hospital. The study included 70 patients diagnosed with unilateral Definite MD according to the clinical criteria proposed by Barany society and International classification of Vestibular Disorder (ICVD) 2015, of whom 36 were females and 34 were male, female to male ratio 1.05:1, aged between 16 years and 63 years with a mean of 43 years. A full Clinical history, General physical examination, otoneurological examinations, and audiological tests including pure tone audiometry and cVEMP was done. Data was collected and included in a data based system and analyzed by (SPSS v.23). Data were presented using descriptive statistics which expressed as frequencies and percentages, P-value less than 0.05 considered significant.

Results: The majority of the affected patient within the age group of 41-50 years (36%) while 4% in 11-20 years, 11% in 21-30 years, 22% in 31-40 years, 20% in 51-60 years, and 7% of more than 60 years. With no significant sex predisposition. Regarding the affected side, right side involved in 51% of cases, while left side involved in 49% of them. Regarding the duration of the disease it's found that 36% of the studied patients had less than 1 year, while 37% of them had (1-5 years duration) and 27% of them had more than 5 years duration. Positive family history of same illness in first and second degree relatives is present in 71% of patients. All of the patients having a low frequency SNHL of variable severity, its mild in 24.3%, moderate in 48.6%, sever in 21.4%, and profound in 5.7% of cases. cVEMP amplitude ratio is abnormal in 87% of patients (61 patients), and this indicate that the cVEMP have a good diagnostic value in patient with MD. There is no significant relation between the result of cVEMP and the severity of low frequency SNHL (P value 0.5). Also there is no significant relation between the severity of low frequency SNHL and the duration of the disease (P value 0.08).

Conclusion: Meniere's disease affect different age group but the age group (40-50 years) being more predominant. With No significant sex predilection. Its diagnosis is mainly clinical and augmented by using simple, readily available clinical tests of hearing and vestibular function (PTA, cVEMP). cVEMP have a good diagnostic value in patients with unilateral definite Meniere's disease.

Keywords: Meniere's disease, Endolymphatic hydrops, Sensorineural hearing loss, cVEMP, Pure-tone audiometry.

1. Introduction

Meniere's disease, with a symptoms triad of vertigo, tinnitus and hearing disorders, was first reported as a disease of the inner ear by Prosper Meniere in 1861 (1). MD is a multifactorial disorder where the onset of disease is determined by the combined effects of genetic factors and environmental factors (2). It's affecting approximately 50 to 200 per 100,000 and most common between the ages of 40 and 60 years

with no significant sex predominance (3). The classical clinical symptoms of an attack of MD include episodic rotatory vertigo with spontaneous nystagmus, nausea and or vomiting, and ear symptoms such as, hearing loss, tinnitus and aural fullness (4). The course of the disease is generally variable, and the development of cochlear and vestibular symptoms may take years, in individual patients Cochlear symptoms may occur in between episodes of vertigo. The episodes of vertigo are

usually more common in the first few years of illness. also Hearing loss and vestibular hypo function show a great variability among patients (5). Clinical classification to diagnose MD developed by the American Academy of Otolaryngology Head and Neck Surgery Foundation (AAO-HNSF) (6). These diagnostic criteria for MD recently revised by the Classification Committee of the Barany Society in cooperation with a lot of national and international organizations and were later approved by the AAO-HNSF Equilibrium Committee (2,7).The diagnosis made clinically, as the disease typically presents with unilateral ear symptoms that can last for several decades (8). A detailed study of inner ear anatomy and physiology is important to understand the pathophysiology and the course of the disease.

2. Methods

This is a clinical, prospective cross sectional study carried out from October 2019 to February 2021 .This study included 70 patients diagnosed with unilateral Definite MD according to the clinical criteria proposed by Barany society and International classification of Vestibular Disorder (ICVD) 2015, of whom 36 were females and 34 were male, aged between 16 years and 63 years with a mean of 43 years.

The exclusion criteria include

1. MD + Middle ear pathology.
2. MD+ Ear surgery.
3. MD + central abnormality.
4. MD + vestibular migraine.
5. MD + BPPV.
6. Bilateral MD.
7. MD + high freq. SNHL.
8. Probable MD

A full Clinical history was taken (According to a pre-designed questionnaire) to inquire about information related to age, sex, residence, symptoms, durations, and family history of same illness. General physical examination was done, a full otoneurological examinations, and audiological tests including pure tone audiometry and cVEMP amplitude ratio (at 500 HZ and 1000 HZ) was done. The differential diagnosis was excluded and the clinical diagnosis of definitive MD according to the (ICVD) 2015 criteria was confirmed.

Analysis

Data was collected and included in a data based system and analyzed by statistical package of social sciences (SPSS v.23). Parametric data continuous variables were presented using descriptive statistics which expressed as frequencies and percentages, P-value less than 0.05 considered significant.

3. Results

Seventy cases were include in this study , the studied patient’s age was between 16 years and 63 years with a mean of 43 years. It is found that the majority of studied patients; 25 patients (36 %) ; belong to

age group (41 – 50) years , while only 3 patients (4 %) belong to age group (11 – 20) years ,The female to male ratio was 1.05:1. Table 1 show Demographic characteristics . It is found that the right side affected in 51% of the studied patients, while the left side affected in 49% of the patients included.

It is found that 37% of the studied patients (26 patients) had the duration between 1 and 5 years duration, and 36% (25 patients) of them had less than one year duration ,and 27% of them (19 patients) had more than 5 years duration.

Table 1: Demographic characteristics

		Sex				Total	
		Male	Freq.	Female	Freq.	No.	Freq.
Age	11-20	1	3%	2	5%	3	4%
	21-30	3	9%	5	14%	8	11%
	31-40	4	12%	11	31%	15	22%
	41-50	15	44%	10	28%	25	36%
	51-60	7	20%	7	19%	14	20%
	>60	4	12%	1	3%	5	7%
Total		34	100%	36	100%	70	100%

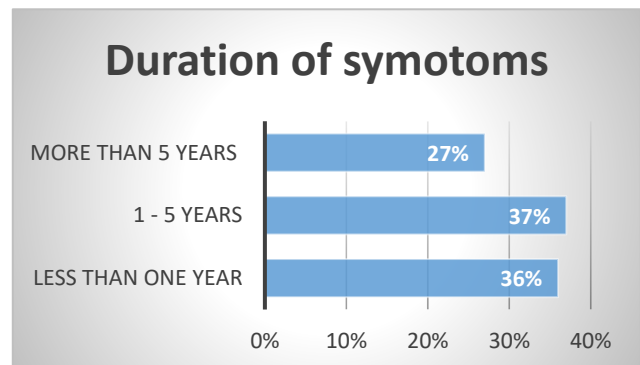


Figure 1: The duration of symptoms in the involved patients

Seventy one percent of the studied patients (50 patients) gave a positive family history of same illness in their first and second degree relatives. While 29% of the studied patients (20 patients) had no family history of same symptoms. it’s found that 48.6% of patients had moderate low frequency SNHL , and 24.3% of them had mild low frequency SNHL , and 21.4% of them had sever low frequency SNHL, while 5.7% of them had profound low frequency SNHL as shown below:

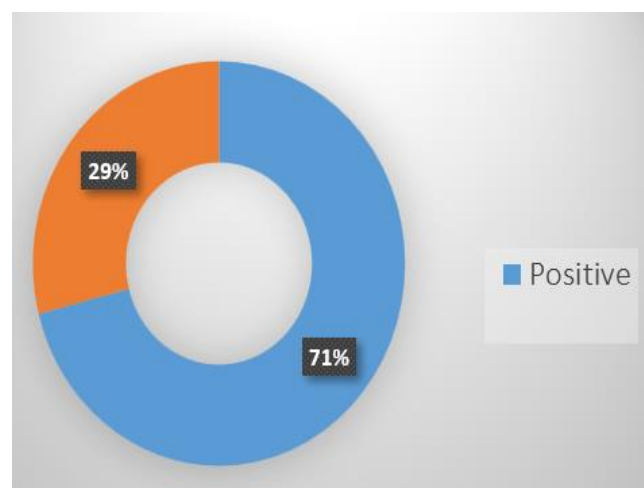


Figure 2: Percentage of family history of same illness

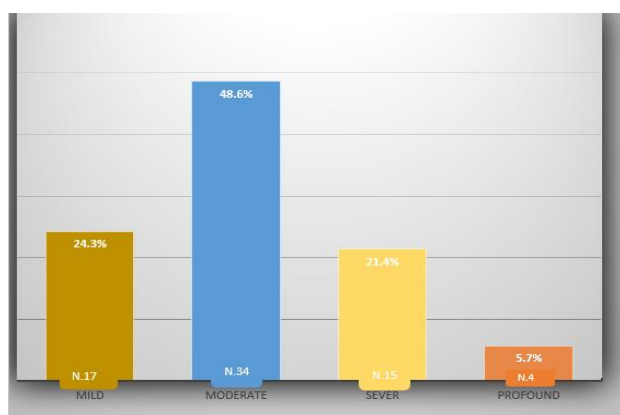


Figure 3: The severity of low frequency SNHL

The result of cVEMP amplitude ratio, in which it is abnormal in 87% of the studied patients (61 patients), while 13% of the patients (9 patients) showed normal findings . it is found that in patients with disease duration of less than 1 year the low frequency SNHL is mild in 32% of them, moderate in 60% of them, severe in 4% of them ,and profound in 4% of them. While in patients with disease duration of 1-5 years it’s found that the low frequency SNHL

is mild in 23% of them, moderate in 27% of them ,sever in 38% , and profound in 12% of them. In patients with disease duration of more than 5 years it is found that the low frequency SNHL is mild in 16% of them, moderate in 63% of them, severe in 21% of them and there was no profound low frequency SNHL in them (P value 0.08).

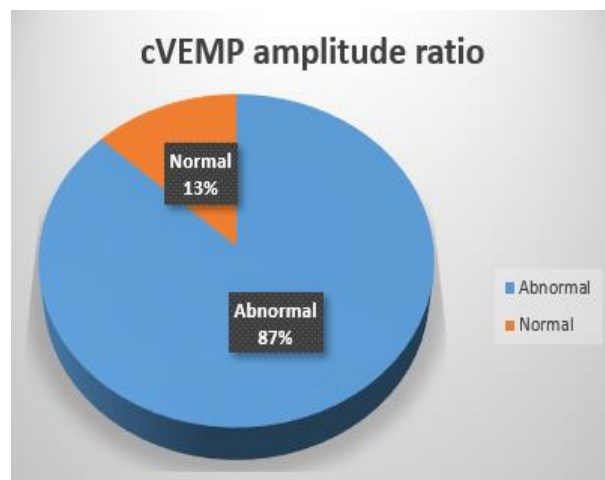


Figure 4: Percentage of abnormal cVEMP amplitude ratio

	Duration of symptoms								Total		
			<1 year		1-5 years		> 5 years		no	%	
	no	%	no	%	no	%	no	%	no	%	
Severity of low freq. SNHL	Mild	8	32%	6	23%	3	16%	17	24%		
	Moderate	15	60%	7	27%	12	63%	34	49%		
	Sever	1	4%	10	38%	4	21%	15	21%		
	Profound	1	4%	3	12%	0	0%	4	6%		
Total				25	100%	26	100%	19	100%	70	100%

(P value 0.08)

We found that in patient with mild low frequency SNHL, cVEMP is abnormal in 82.3% of them, and in patients with moderate low frequency SNHL, cVEMP is abnormal in 85.3% of them, and in patients with

severe low frequency SNHL, cVEMP is abnormal in 93.3% of them, while in patients with profound low frequency SNHL cVEMP is abnormal in 100% of them, and this is clearly displayed n table 3.3. (P value 0.5)

		Severity of low frequency SNHL								Total	
		Mild		Moderate		Sever		Profound		no.	%
		no.	%	no.	%	no.	%	no.	%	no.	%
cVEMP ratio	abnormal	14	82.3%	29	85.3%	14	93.3%	4	100%	61	87%
	normal	3	17.7%	5	14.7%	1	6.7%	0	0%	9	13%
Total		17	100%	34	100%	15	100%	4	100%	70	100%

(P value 0.5)

4. 4. Discussion

Meniere's disease, with a symptoms triad of vertigo, tinnitus and hearing disorders, was first reported as a disease of the inner ear by Prosper Meniere in 1861 (1).

The diagnosis of MD is mostly based on historical information and the results of the audiogram. Although vestibular tests have been used for many years, audiometry remains the most useful test to support the diagnosis of MD in patients presenting

with episodic vertigo. More recently, cVEMP has been introduced as diagnostic tools for superior semicircular canal dehiscence and for macular vestibulopathies . However, the diagnostic value of cVEMP in MD has not been fully determined (9).

The demographic distribution of this study (presented in table 3.1) showed that the study included 70 patients with unilateral definite MD , males were 34 (49%) , female were 36 (51%), female to male ratio 1.05:1, so there is no significant sex predilection in our studied patients. this agreed with

Brookler et al (10) and Mizukoshi, et al (11) and Maxwell et al (12) and Angeli et al (13) who found that there is no significant sex predilection in MD.

The age of the studied patients between 16 years and 63 years with a mean of 43 years. The majority of the affected patient within the age group of 41-50 years (36%) while 4% in 11-20 years, 11% in 21-30 years, 22% in 31-40 years, 20% in 51-60 years, and 7% of more than 60 years.

this is compatible with Singh, Kumar, et al (14) Mizukoshi, et al (11) and Teggi, et al (15) in that the disease is more commonly affect the age group of (40-50) years but differ from Angeli et al (13) and Shojaku et al (16) who report a mean of 53 years and its affecting 40-60 years, probably due to inclusion of patients older than 80 years old and a large sample size.

Only patients with unilateral definite MD included in the study (51% right side affected and 49% left side affected) this is nearly similar to Maxwell et al (12) and Singh, Kumar, et al (14).

Regarding disease duration, 37% of cases (26 patients) between 1- and 5-years duration and 36% of cases (25 patients) less than one year, while 27% of cases (19 patients) gave history of more than 5 years duration.

The present study shows no significant difference in the duration of illness probably due to involvement of only definite MD in a cross-section study.

Positive family history of similar illness is seen in 71% of cases and this may indicate that definite MD may have a strong genetic association, and this is nearly compatible with Morrison et al (17), Koyama S et al (18) and Arweiler et al (19) who stated that positive family history is present in more than 60% in of cases, while Paparella et al (19) stated that the family history is less than 20%, probably due to involvement of all patients with MD and not only definite MD.

The degree of unilateral low frequency SNHL in patients included in this study vary from mild to profound. it was mild in 24.3% of cases (17 patients) , moderate in 48.6% (34 patients), sever in 21.4% (15 patient)and profound in only 5.7% (4 patient) of cases. The present study shows that there is a varying degree in the severity of low frequency SNHL in patient with MD. and this is agreed with Noguchi et al (20) and Meyerhoff et al (21)

The evaluation of the relation between the severity of low frequency SNHL and the duration of the illness showing that there is no significant relation between them , and this is compatible with Mateijsen et al (22). Who concluded that the severity of low frequency SNHL in MD does not depend on duration of the disease.

cVEMP amplitude ratio is abnormal in 87% of patients(61 patients) with unilateral definite MD involved in this study(normal cVEMP ratio 0.36) , and this is compatible with Maxwell et al (12), Angeli, Simon et al (13) who concluded that The cVEMP amplitude ratio is abnormal in patients with MD. Salviz et al (9), Sandhu et al (23), Zhang, Sulin, et al (24), Rauch, Steven D., et al (25) all of them stated

that the cVEMP amplitude ratio is abnormal in patients with unilateral MD.

Evaluation of the relation between the abnormal cVEMP ratio and the severity of low frequency SNHL showed that there is no significant relation between them despite that there is slight increase in percentage of abnormal cVEMP with the increasing in the severity of low frequency SNHL (abnormal cVEMP ratio is 82.3% in mild ,85.3% in moderate,93.3% in severe and 100% in profound low frequency SNHL),and this is agree with Simon et al (13) who found that there is no association between the cVEMP amplitude ratio and audiometric result. Probably this is due to the fact that since cVEMP assesses saccular function, the VEMP results may not correlate with ipsilateral audiometric thresholds because the saccule may be significantly affected but the still the hearing is mildly affected.

5. Conclusion

Meniere's disease affect different age group but the age group (40-50 years) being more predominant. With no significant sex predilection.

Its diagnosis is mainly clinical and augmented by using simple, readily available clinical tests of hearing and vestibular function (PTA , cVEMP) cVEMP have a good diagnostic value in patient with unilateral definite Meniere's disease

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