

Electrocochleography Evaluation in the Diagnosis of Meniere's Disease Meniere

Meniere Hastalığı Tanısında Elektrokokleografi Değerlendirmesi Meniere

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ABSTRACT

Meniere's disease is a heterogeneous disease of the audiovestibular system, etiologically caused by endolymphatic hydrops, accompanied by both vestibular and auditory symptoms. Pure tone audiometry, otoacoustic emission, Electrocochleography (EcochG), vestibular evoked myogenic potentials and magnetic resonance imaging can be used in the diagnosis of the disease. Electrocochleography is a testing method that allows the assessment of cochlear electrical potentials. The primary condition diagnosed by ECochG and available for neuromonitoring intraoperatively is Meniere's disease. In Meniere's disease, the magnitude of the summation potential (SP) increases with the impairment of the transduction process, while the action potential (AP) will remain constant. This condition causes an increased SP/AP ratio and is pathophysiologically associated with endolymphatic hydrops. The aim of our study is to investigate the role and worth of EcochG in the clinical diagnosis of Meniere's disease according to the studies conducted in the current literature. Pubmed and Web of Science database was used for our review study. The search words used to examine the studies in between 2014-2024 were determined as "ECochG, Meniere's disease". In this context, a total of 126 studies were reached. 9 research articles and 4 systematic reviews were included, including the role of Electrocochleography in the diagnosis of Meniere's disease and its evaluation results. As a result of the study, it is thought that the EcochG test method alone will not be sufficient in the diagnosis of Meniere's disease and that examining it together with other evaluation methods will lead clinicians to a more accurate and reliable diagnosis.

Keywords: Electrocochleography, Endolymphatic hydrops, Hearing loss, Meniere's disease, Otoacoustic emissions,

ÖZ

Meniere hastalığı, hem vestibüler hem işitsel semptomların eşlik ettiği, etiyolojik olarak endolenfatik hidropsun sebep olduğu odyovestibüler sistemin heterojen bir hastalığıdır. Hastalığın tanısında saf ses odyometrisi, otoakustik emisyon, elektrokokleografi (EcochG), vestibüler uyarılmış miyogenik potansiyeller ve manyetik rezonans görüntüleme gibi yöntemler kullanılabilir. Elektrokokleografi, koklear elektriksel potansiyelleri değerlendirmeyi sağlayan bir test yöntemidir. ECochG tarafından teşhis edilen ve intraoperatif olarak nöromonitörizasyon için kullanılabilen birincil durum meniere hastalığıdır. Meniere hastalığında, sumasyon potansiyeli (SP) büyüklüğü transdüksiyon sürecindeki bozulma ile artarken, aksiyon potansiyeli (AP) sabit kalacaktır. Bu durum artmış SP/AP oranına sebebiyet vermektedir ve patofizyolojik olarak endolenfatik hidrops ile ilişkilendirilmektedir. Çalışmamızın amacı, EcochG'nin meniere hastalığı klinik tanısında güncel literatürde yapılmış olan araştırmalarca rolü ve değerinin incelenmesidir. Derleme çalışmamız için Pubmed ve Web of Science veritabanı kullanıldı. Çalışmamız kapsamında 2014-2024 yılları arasındaki çalışmaları incelemek amacıyla kullanılan arama sözcükleri "ECochG, meniere disease" şeklinde belirlendi. Bu kapsamda toplam 126 çalışmaya ulaşıldı. Meniere hastalığının tanısında Elektrokokleografinin rolü ve değerlendirme sonuçlarını içeren 9 araştırma makalesi ve 4 sistematik derleme dahil edildi. Çalışmanın sonucunda, EcochG test yönteminin meniere hastalığı tanısında tek başına yeterli olmayacağı ve diğer değerlendirme yöntemleri ile birlikte incelenmesinin klinisyenleri daha doğru ve güvenilir tanıya yönlendireceği düşünülmektedir.

Anahtar Kelimeler: Elektrokokleografi, Endolenfatik hidrops, İşitme kaybı, Meniere hastalığı, Otoakustik emisyon

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INTRODUCTION

Meniere's disease (MD) is a complex, heterogeneous inner ear disorder in which multiple underlying factors interact, including dysfunction of the labyrinth structures responsible for balance and hearing, anatomical variations in the temporal bone, genetic predisposition, autoimmunity, migraine, allergy, cellular and molecular mechanisms.¹ Meniere's disease, first named after the French physician Prosper Ménière in 1861, manifests itself as recurrent and severe vertigo attacks accompanied by symptoms such as fluctuating hearing loss, tinnitus and a feeling of fullness in the affected ear.² Its prevalence is known to affect approximately 50 to 200 in 100,000 adults, and it is 1.3 times more prevalent in women than in men.³ Although the presence of MD has also been documented in children, it is most commonly seen between the ages of 40-60.⁴ The etiology of this disease is not fully known, but the leading theory is that the disease is caused by excess endolymph fluid in the membranous labyrinth of the inner ear. This condition leads to increased pressure in the endolymphatic sac and is called endolymphatic hydrops (EH).⁵ EH disrupts both the auditory and vestibular systems by disrupting the normal function of sensory hair cells and nerve fibers.⁶ Hydrops in the cochlear canal causes hearing loss and tinnitus, while hydrops in the vestibular canal causes dizziness attacks.⁴ Although the exact cause of endolymph accumulation is not known, it is thought to be impaired fluid homeostasis. However, the relationship between EH and MD is still unclear.⁷

In Meniere's disease, the disruption of the fluid balance in the labyrinth leads to damage to the vestibular system. In this malfunction, in addition to episodic vertigo attacks, symptoms such as imbalance, spatial disorientation, cold sweat, feeling faint, nausea and vomiting are accompanied.⁵ Dizziness occurs in attacks and there is no loss of consciousness during these attacks. Meniere's disease is excluded in vertigo attacks lasting more than a day.⁴ The sensorineural type of hearing loss that occurs initially affects low frequency tones.

However, in later stages, high frequencies are also affected, so it turns into a bowl-like configuration. In later stages, it affects all frequencies. Therefore, the hearing loss is of the fluctuating type.⁸ Tinnitus is a buzzing sound and is present in some patients. There is a fullness in the ear felt from the head and neck.⁴ Treatments range from lifestyle changes, dietary modifications, vestibular rehabilitation and medication to more invasive options such as surgery.⁵⁻⁹

Electrocochleography and Other Audiological Assessment Methods

Electrocochleography (ECochG) is a test method that allows the evaluation of cochlear electrical potentials.¹⁰ ECochG has clinical applications such as determining hearing thresholds and assessing auditory neuropathy, intraoperative neuromonitoring in cochlear implantations.¹¹ In addition, the primary condition diagnosed by ECochG and used for intraoperative neuromonitoring is MD.¹²

ECochG consists of 3 potentials: cochlear microphonic (CM), action potential (AP), and summation potential (SP).¹³⁻¹⁴ SP represents the electrical responses of the hair cells located in the inner ear. It is particularly generated due to the movement of the basilar membrane and the asymmetry of this movement. More technically, it is a direct current shift that results from the temporal summation of the alternating current responses known as CM. In Meniere's disease, due to an increase in endolymphatic fluid (endolymphatic hydrops), this potential may be higher than normal. Both the amplitude and latency of SP tend to increase in Meniere's disease, which highlights its diagnostic importance. AP is the electrical response generated by the auditory nerve. In the ECochG test, AP is measured as the first major electrical response of this nerve following an acoustic stimulus delivered to the ear.¹⁵

In Meniere's disease, SP value increases with impairment of the transduction process, while AP remains relatively constant. This results in an increased SP/AP ratio that is

pathognomonic for MD. Therefore, if the SP/AP ratio of patients evaluated with TT ECoChG is greater than 0.45, it is considered abnormal. However, since the electrodes are placed more peripherally in the measurement with ET ECoChG, the ratio is expected to be higher in order to be considered abnormal.¹⁶

There is no single gold standard test for the diagnosis of MD. In pure tone audiometry, sensorineural hearing loss is present and initially has a configuration that decreases towards low frequencies. However, with the progression of the disease, a configuration that decreases also at high frequencies may occur. In the late stages, a hearing loss that decreases at all frequencies may occur. This is why hearing loss shows fluctuation.⁸⁻¹⁷ In these patients, the glycerol test is quite useful and a 15 dB improvement in one frequency, 10 dB improvement in 3 consecutive frequencies or a 12% increase in speech

discrimination scores (SDS) is considered in favor of Meniere's disease. Although OAE test results vary, improvements in Distortion Product Otoacoustic Emission (DPOAE) values are very significant after the glycerol test. If there is no additional pathology in the tympanogram, Type A is seen. Initially, VEMP (Vestibular Evoked Myogenic Potentials) responses are normal, but as the disease progresses, the amplitudes of the responses decrease.⁴ Magnetic resonance imaging (MRI) technique can be used to visualize the increase in fluid pressure in the inner ear. Contrast-enhanced MRI is usually used to detect this condition.¹⁸ This study aims to comprehensively examine the diagnostic effectiveness, clinical relevance, and limitations of ECoChG in the assessment of MD, by analyzing and synthesizing findings from research articles and systematic reviews published between 2014 and 2024.

MATERIALS AND METHOD

Stage One: Determining the Research Question

Three research questions were determined to guide our review study.

1. How has the literature on Meniere's disease and Electrocochleography evolved over the past few years?
2. What evidence exists in the field of Meniere's disease and Electrocochleography?
3. What are the most important elements to consider to facilitate understanding of the current literature on Meniere's disease and Electrocochleography?

Second Stage: The analysis and results of the Search Strategy should follow well-established and recent approaches.

Pubmed and Web of Science database was used for our review study. The search words used to examine the studies between 2014-2024 within the scope of our study were determined as "ECoChG, Meniere's disease".

Study Selection

The PRISMA flow diagram used for study selection is shown in Figure 1.¹⁹

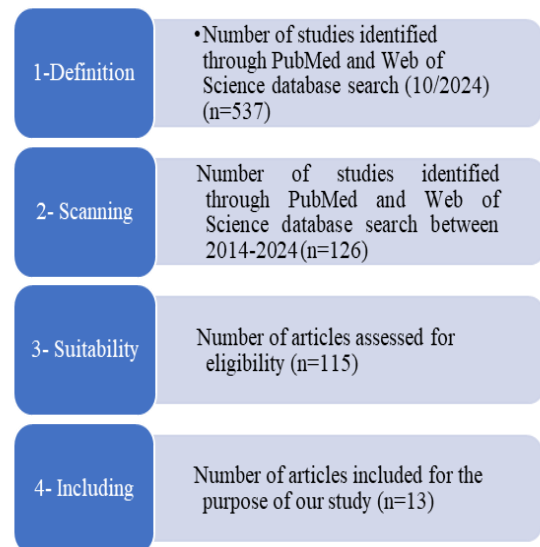


Figure 1. PRISMA flow diagram

According to the PRISMA flow diagram, a total of 126 studies were identified with the search words “ Meniere's disease and electrocochleography” from 2014 to 2024. Among these studies, 111 studies were research articles, 3 studies were systematic

reviews, 4 studies were clinical research, 7 studies were reviews and 1 study was a book and document study. In our study, 111 research articles and 4 clinical studies were examined for study eligibility. 9 research articles and 4 systematic reviews were included, including the role of electrocochleography in the diagnosis of Meniere's disease and its evaluation results.

Tabulation of studies

All authors were tasked with summarizing and tabulating the included sections in accordance with the purpose of the study.

Collection, Summarization, and Reporting of Results

The results of our review study are summarized and discussed, and the role of electrocochleography in the diagnosis of Meniere's disease and the evaluation results are given.

STUDIES ON THE SUBJECT

Lamounier et al. (2014) included a total of 19 articles in their review and examined the use of ECochG in the diagnosis of Ménière's disease. As a result of the study, the researchers reported that ECochG is a noninvasive, easy-to-use procedure that helps clinicians in the treatment of Ménière's disease and is a valuable tool in the diagnosis of hydrops.²⁰

Oh et al. (2014) investigated the diagnostic value of ECochG in the early symptomatic period of Meniere's disease in their study involving 60 Meniere's patients and 30 healthy individuals. Extratympanic ECochG was used in the evaluation. SP/AP area and amplitude ratios of the patient and control groups were calculated and compared. The results showed that there was no statistically significant difference in SP/AP area and amplitude ratios between the patient group and the control group. However, the researchers reported that ECochG has limited value in the diagnosis of Meniere's disease in the early symptomatic period.²¹

Hornibrook et al. (2015) included 102 patients, 57 of whom had Meniere's disease, in their study and performed MRI, click and tone burst EcochG evaluations on the patients. As a result of the study, it was stated that tone burst EcochG was more sensitive than MRI evaluation in the diagnosis of Meniere's disease.²²

Gerenton et al. (2015) included 73 individuals with definite Meniere's disease in

their study. The diagnostic specificity and sensitivity of distortion product otoacoustic emission (DPOAE) and ECochG in Meniere's disease were evaluated. The study firstly based on SP/AP ratio alone and then examined the combination of DPOAE and SP/AP ratio. The study result showed that SP/AP ratio alone has poor sensitivity to hydrops. The evaluation consisting of the combination of DPOAE and EcochG was recommended for the sensitive detection of active hydrops.²³

Ziylan et al. (2016) applied intratympanic gadolinium to patients with definite Meniere's disease. They prepared a systematic review on this subject to analyze the sensitivity, specificity, positive predictive value and negative predictive value of comparing MRI with electrocochleography. As a result of this systematic review based on a small number of patients, it was reported that click-stimulus EcochG had a relatively low sensitivity and negative predictive value in the diagnosis of definite Meniere's disease compared to MRI.²⁴

Lamounier et al. (2016) performed a neuro-otological examination including pure tone and speech audiometry, VEMP and extratympanic ECochG on 12 patients with definite Meniere's disease and 12 healthy volunteers included in their study. The sensitivities and specificities of VEMP and ECochG tests for the diagnosis of the disease were calculated. As a result of the study, a weak correlation was determined between

these two tests because VEMP and EcochG tests had similar specificity and sensitivity rates in the diagnosis of the disease. However, although both tests had high specificity, it was reported that the sensitivity of VEMP test was higher than ECochG.²⁵

Mammerella et al. (2017) included 34 patients with Meniere's disease in their study and retrospectively examined the SP/AP ratio in peritympanic electrocochleography. The results reported that the SP/AP ratio showed high predictability in the diagnosis of Meniere's. This EcochG study showed that it can be used for early diagnosis of Meniere's disease.²⁶

Ciorba et al. (2017) included a total of 87 studies in their research examining VEMP, ECochG and otoacoustic emissions (OAE), which are tools used in the evaluation of Meniere's disease. As a result of the study, they reported that OAE was useful in determining cochlear damage in Meniere's disease. In addition, it was concluded that ECochG and VEMP tests may be helpful in clinical monitoring in Meniere's disease, but they are not sufficient for diagnosis alone.²⁷

Quatre et al. (2019) prospectively included 41 patients with Meniere's disease in their study and performed MRI, pure tone audiometry, EcochG, DPOAE and cVEMP evaluations on the patients. The results showed that MRI evaluation showed hydrops in the inner ear when hearing loss was over 35 dB. It was stated that DPOAEs could provide fast and easy measurements when hearing loss was less than 60 dB. It was reported that EcochG could make the diagnosis regardless of the hearing loss and when used together with DPOAE, it would increase the chance of a more reliable diagnosis.²⁸

Ayub et al. (2019) analyzed and compared the sensitivity and specificity of EcochG (315 individuals) and combination audiological diagnostic measurements (113 individuals) in Meniere's disease in five articles they included in their study. In their results, it was concluded that the SP/AP ratio was lower than the

sensitivity and specificity of the combination measurement results when used alone for diagnosis. However, they also stated that more studies were needed in the future due to the small sample size.²⁹

Obeidat and Bell (2019) examined the sensitivity and specificity of the ECochG SP/AP amplitude ratio and the 95% normative range of the 500 Hz cVEMP threshold in 19 healthy individuals and 15 individuals diagnosed with bilateral definite Meniere's disease. As a result of the research, they concluded that the specificity and sensitivity of the ECochG SP/AP ratio in the diagnosis of Meniere's disease was higher than that of the cVEMP threshold, but it was not perfect for diagnosis.³⁰

Martines et al. (2020) evaluated 21 patients with vestibular migraine and 21 patients who met definite Meniere's criteria using vHIT and ECochG tests and compared the results. As a result of the study, it was stated that 11 patients with definite Meniere's disease and 3 patients with vestibular migraine with suspected endolymphatic hydrops had high SP/AP ratio (> 0.4). However, it has also been reported that higher abnormal ECochG responses are obtained in Meniere's disease compared to vestibular migraine.³¹

Bawazeer et al. (2024) included 95 patients with Meniere's disease in their study examining the specificity and sensitivity of the EcochG method for the diagnosis of Meniere's disease. In this study, the SP/AP ratio was found using click stimuli and compared with the SP amplitudes found using tone burst stimuli. The study results showed that the results obtained using click stimuli had higher specificity and sensitivity values than the results obtained using tone burst stimuli. It was also reported that the SP/AP ratio (sensitivity: 88.5%) was more diagnostically significant than the SP/AP amplitude ratio (sensitivity: 51.7%).³²

The articles included in our study were examined and summarized in Table 1.

Table 1. Included Studies and Their Result

Year	Researchers	Type of Study	Number of People/Studies Included	Test Method and Findings	Conclusions/Recommendations
2014	Lamounier et al.	Systematic Review	n _s =19	ECochG	SP/AP ratio is a valuable and sufficient evaluation method in the diagnosis of Meniere's.
2014	Oh et al.	Original Article	n _p =60, n _c =30	ECochG	EcochG is not sensitive in the early symptomatic phase.
2015	Hornibrook et al.	Original Article	n _p =57, n _c =45	MRI, Click EcochG, Tone Burst EcochG	Tone burst EcochG is more sensitive than MRI in the diagnosis of hydrops.
2015	Gerenton et al.	Original Article	n _p =73	DPOAE, EcochG	DPOAE and EcochG increase diagnostic sensitivity when used together.
2016	Ziylan et al.	Systematic Review	n _s =4	Click EcochG, MRI	Click-stimulated EcochG showed lower diagnostic sensitivity than MRI.
2016	Lamounier et al.	Original Article	n _p =12, n _c =12	PTA, EcochG, VEMP	The diagnostic sensitivity of the VEMP test was higher than that of EcochG.
2017	Mamerella et al.	Original Article	n _p =34	ECochG	EcochG SP/AP ratio is a useful tool in the diagnosis of Meniere.
2017	Ciorba et al.	Original Article	n _s =19	VEMP, EcochG, OAE	Diagnostic sensitivity of OAE was higher than EcochG.
2019	Quatre et al.	Original Article	n _p =41	VEMP, EcochG, DPOAE, PTA, MRI	HL >35 dB MRI high sensitivity in diagnosis, HL <60 dB DPOAE high sensitivity in diagnosis, EcochG showed high sensitivity in diagnosis in all degrees of hearing loss.
2019	Ayub et al.	Systematic Review	n _s =5	EcochG	The diagnostic sensitivity of the SP/AP ratio alone is not high.
2019	Obeidat et al.	Original Article	n _p =15, n _c =19	EcochG, cVEMP	SP/AP ratio showed higher sensitivity in diagnosis than cVEMP.
2020	Martines et al.	Original Article	n _p =42	vHIT, EcochG	Compared with VM patients, the SP/AP ratio showed higher sensitivity in the diagnosis of Meniere's disease.
2024	Bawazeer et al.	Original Article	n _p =95	Click EcochG (SP amplitude), Tone burst EcochG (SP/AP area ratio)	SP/AP area ratio showed higher sensitivity in diagnosis than SP/AP amplitude ratio.

Abbreviations: n_S; number of studies included, n_P; number of patients, n_C; number of control, EcochG; Electrocochleography, SP; Sumation potential, AP; Action potential, VM; Vestibular migraine, cVEMP; Cervical vestibular evoked myogenic potentials, HL; Hearing loss, DPOAE; Distortion product oto-acoustic emissions, OAE; Oto-acoustic emissions, VEMP; Vestibular evoked myogenic potentials, MRI; Magnetic Resonance Imaging, PTA; Pure tone audiometry.

General Discussion of the Literature

A review of the existing literature reveals that studies investigating the use of ECochG in the diagnosis of Ménière's disease show substantial variability. While some studies emphasize the diagnostic value of the SP/AP ratio with high sensitivity and specificity, others suggest that ECochG offers limited utility, particularly in the early symptomatic stages of the disease. Additionally, several studies indicate that the diagnostic accuracy of ECochG improves when used in conjunction with other assessment tools such as DPOAEs, VEMPs, and MRI. This suggests that ECochG may be more reliable as a complementary diagnostic method rather than as a standalone test.

Nevertheless, differences in stimulus type (click or tone burst), application method (intratympanic or extratympanic), patient populations, and testing protocols make it difficult to draw direct comparisons between studies. Furthermore, the limited sample sizes in certain studies reduce the generalizability of the findings. Despite these limitations, ECochG remains a non-invasive and clinically feasible technique that may contribute meaningfully to the diagnostic process. In this context, the reviewed literature suggests that while ECochG alone may not be sufficient for the definitive diagnosis of Ménière's disease, it holds value as a supportive diagnostic tool.

CONCLUSION AND RECOMMENDATIONS

It is known that EcochG has been used for a long time in the clinical diagnosis of endolymphatic hydrops in Meniere's disease. In our study, the sensitivity and results of this clinical use compared to other diagnostic tests currently used in terms of current technology and literature were examined. When the results of our study were examined, it was seen that EcochG has superior and weak aspects compared to other diagnostic tests.

In literature studies, it has been reported that EcochG has higher sensitivity for all degrees of hearing loss compared to other methods such as MRI and OAE. In addition, studies have determined that EcochG measurements performed using tone burst stimulus have higher sensitivity in diagnosing hydrops compared to MRI findings. However, in some studies, it has been reported that when the click stimulus is evaluated based on the SP/AP area ratio, it has higher sensitivity in diagnosing hydrops in Meniere cases compared to SP amplitude values in the presence of tone burst stimulus.

As a result of some studies, it has been reported that DPOAE and VEMP methods are more reliable than EcochG findings in the diagnosis of hydrops in Meniere's cases and it has been suggested that these methods should be interpreted together in the diagnosis.

It should definitely be kept in mind during the evaluation that EcochG may also show a high SP/AP ratio in some other diseases (such as SSCD). Therefore, using EcochG alone in the diagnosis of Meniere's disease will not provide reliable results to clinicians. Although EcochG has a high sensitivity in diagnosing hydrops in the diagnosis of Meniere's, examining it together with other diagnostic

tests in terms of holistic evaluation will provide clinicians with a more reliable diagnosis.

Study Limitations

This article does not constitute a systematic review; rather, it should be considered a narrative review that summarizes key findings from selected studies in the current literature. Therefore, the scope of this study is limited. In the future, it is recommended that larger-scale, systematically conducted investigations with greater sample sizes be undertaken to provide a more comprehensive and evidence-based understanding of the diagnostic utility of ECochG in Ménière's disease.

Conflicts of interest

This article has no conflict of interest.

Authors' Contributions

The idea and concept of the research belong to Y.G.E. and C.U.

The research design was planned by Y.G.E. and C.U.

The supervision/consultancy phase of the research was carried out by M.P.

The interpretation and analysis process of the data were carried out by Y.G.E., C.U. and M.P.

The literature review in the research was carried out by Y.G.E. and C.U.

The writing of the article was carried out by Y.G.E., C.U. and M.P.

The critical review of the article was carried out by M.P.

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