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Derleme Makale / Review Article

**Evaluation of oVEMP and cVEMP in Superior Semicircular Canal Dehiscence Cases**  
*Süperior Semisirküler Kanal Dehissansı Olgularında oVEMP ve cVEMP Değerlendirmesi*

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**ABSTRACT**

Superior semisirküler kanal dehissansı (SSCD) is a disease of the audiovestibular system, which occurs as a result of the lack of bone layer covering the superior semisirküler kanal. The disease forms a third window in the inner ear due to the lack of bone layer. The third window that occurs forms a pseudo conductive type of hearing loss, which has decreased bone thresholds in the audiometry test. Vestibular evoked myogenic potentials are an electrophysiological measurement method in which the responses of the neck or eye muscles to an acoustic stimulus are recorded by means of electrodes. The purpose of use is to evaluate the functions of the utricle and saccule, which are the vestibular end organs. Therefore its primary purpose is to diagnose semisirküler kanal dehissansı. This review is based on research from the last decade; diagnostic results of ocular vestibular evoked myogenic potentials (oVEMP) and cervical vestibular evoked myogenic potentials (cVEMP) tests were evaluated in cases of superior semisirküler kanal dehissansı (SSCD). Studies indicate abnormal decreases in cVEMP test thresholds on the affected side in SSCD cases. Additionally, a significant increase in oVEMP amplitude on the affected side has been reported. Results; It shows that evaluating these two tests together will contribute to the diagnosis of semisirküler kanal dehissansı cases.

**Keywords:** Superior semisirküler kanal dehissansı, Cervical vestibular evoked myogenic potentials, Ocular vestibular evoked myogenic potentials

**ÖZET**

Süperior semisirküler kanal dehissansı, süperior semisirküler kanalın üzerini örten kemik tabakanın eksikliği veya zamanla inceliş erimesi sonucunda oluşan odyovestibüler sistemin bir hastalığıdır. Hastalık, kemik tabakanın olmayışı nedeniyle iç kulakta oval ve yuvarlak pencere dışında üçüncü bir pencere oluşturur. Oluşan üçüncü pencere nedeniyle saf ses odyometri testi sonrası kemik yolu eşiklerini azaltan yalancı iletim tipi işitme kaybı (psödo-konduktif) gözlenir. Vestibüler uyarılmış miyojenik potansiyeller ise akustik bir uyarana, boyun veya göz kaslarının verdiği cevapların elektrotlar aracılığıyla kaydedildiği elektrofizyolojik bir ölçüm yöntemidir. Kullanım amacı, vestibüler son organlar olan urtikül ve sakkülün fonksiyonlarını değerlendirmektir. En patognomonik olduğu hastalık semisirküler kanal dehissansı olmakla birlikte birincil amacı semisirküler kanal dehissansının tanısını koydurmaktadır. Bu derlemede, son on yılın araştırmaları esas alınmış olup; süperior semisirküler kanal dehissansı (SSKD) olgularında, oküler vestibüler uyarılmış miyojenik potansiyeller (oVEMP) ve servikal vestibüler uyarılmış miyojenik potansiyeller (cVEMP) testlerinin tanısasal sonuçları değerlendirilmiştir. Çalışmalar, SSKD olgularında etkilenen tarafta Cvemp testi eşiklerinde anormal düşüşleri ifade etmektedir. Ayrıca oVEMP amplitüdünde, etkilenen tarafta belirgin bir artışta bildirilmiştir. Sonuçlar; bu iki testin birlikte değerlendirmeye alınmasının, semisirküler kanal dehissansı olgularına tanısasal anlamda katkı sunacağını göstermektedir.

**Anahtar Kelimeler:** SSCD, cVEMP, oVEMP, Psödo-konduktif işitme kaybı



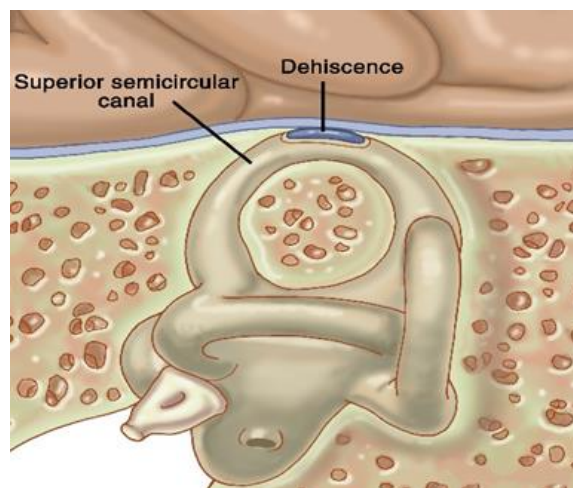
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## INTRODUCTION

Superior semicircular canal dehiscence (SSCD) is one of the different diseases of the audiovestibular system, which was first described in the late 1990s. (Goplen et al., 2022). This disorder arises from the absence or thinning of the bony layer covering the superior semicircular canal, creating an opening. The formed opening is considered a third window in the inner ear, in addition to the oval and round windows. Due to the lower impedance of the oval window, the energy transmitted to the bone is increased, while there is a decrease in energy transmitted through the air. While the bone path hearing thresholds are much better than normal, the drop in airway hearing thresholds causes “false conductive type (pseudo-conductive) hearing loss” (Merchant and Rosowski, 2008; Janky et al., 2015; Göçer and Dinç, 2019).

With a prevalence of 0.7%, it is not a very common disease. The likelihood of observing dehiscence increases with age, typically manifesting symptoms in patients between the ages of 40 and 60. However, cases of SSCD in children have also been reported, affecting both genders similarly (Nielsen et al., 2013; Sood et al., 2017).

The etiology of this disease is not exactly known, but the most widely accepted theory is “Third Window Theory”. According to this theory, the presence of a dehiscence in the upper part of the superior semicircular canal (SSC) leads to changes in pressure in the intracranial cavity or from the middle ear to the inner ear (Grieser, 2015). This event causes the neural excitation rates to change in the vestibular system. In addition, the inner ear, which leads to the decrease of acoustic energy, causes changes in fluid dynamics. The third window in SSCD is connected to the scale vestibular. Thus, stapes movements induced by perileaf pressure are channeled into this pathway with low impedance. This event leads to less energy being passed to the basillary membrane. The resulting fluid flow results in the movement of the coupling in the SSC. This movement leads to inappropriate stimulation of vestibular structures with acoustic energy (Göçer and Dinç, 2019). On the other hand, environmental factors such as traumatic situations (such as head trauma) or Valsalva may also occur SSCD (Carey et al., 2000; Minor, 2005).



**Figure 1. Physiopathological View of SSCD** (Kohan, 2015)

While some patients may have predominantly vestibular symptoms, others may experience both vestibular and auditory symptoms, and some may have only hearing problems (Göçer and Dinç, 2019).

Among the vestibular symptoms of the disease:

- "Hennebert sign," defined as pressure-induced vertigo,
- "Tullio phenomenon," known as sound-induced vertigo,
- Torsional nystagmus,
- "Oscillopsia," described as the up-and-down movement of objects in the field of vision while walking (Crane et al., 2010; Ward et al., 2017).

Among the auditory symptoms of the disease:

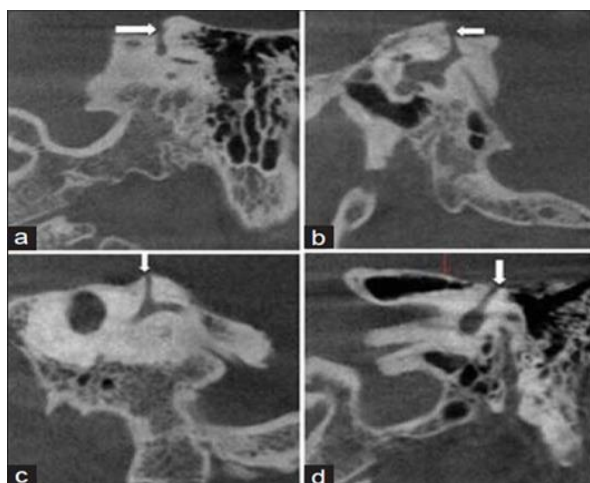
- Autophony or hyperacusis conditions, where patients report hearing their own heartbeats, intestinal sounds, footsteps, and eye movements,
- Pulsatile tinnitus,
- Pseudo-conductive (pseudoconductive) hearing loss due to an osseous defect in SSCD (Janky et al., 2015; Minor et al., 2001).

Vestibular Evoked Myogenic Potentials (VEMPs) are an electrophysiological measurement method that records reflex responses resulting from the stimulation of vestibular afferents by acoustic stimuli. While VEMPs are commonly elicited with acoustic stimuli, other stimuli such as vibration, electrical stimulation (galvanic), and certain physiological motion stimuli can also be used (Belgin and Şahlı, 2017). In clinical settings, the most common stimuli can also be used (Belgin and Şahlı, 2017). The most common stimulus to

obtain VEMP responses is a 500 Hz toneburst (Zuniga et al., 2014). There are two types of VEMP methods: Cervical VEMP (cVEMP) and Ocular VEMP (oVEMP) (Zuniga et al., 2013).

There is no gold standard test for the diagnosis of SSCD (Ho et al., 2017). SSCD can be diagnosed through audiological evaluation and various imaging methods (Janky et al., 2015). High-resolution computed tomography (CT) helps in diagnosing SSCD by revealing bone defects in the superior semicircular canal. Çeliker et al. (2018) in a study, demonstrated that MRI has high sensitivity and specificity in diagnosing SSCD in patients with neurotological symptoms (Çeliker et al., 2018; Noij and Rauch, 2020). Cadaveric studies have shown that in 0.5%, the superior semicircular canal is separated, and in 1.4%, it is near separation ( $\leq 0.1$  mm bone thickness) (Carey et al., 2000). Computed tomography imaging studies of the temporal bone have also reported radiological separation rates between 3.0% and 9.0% (Berning et al., 2019).

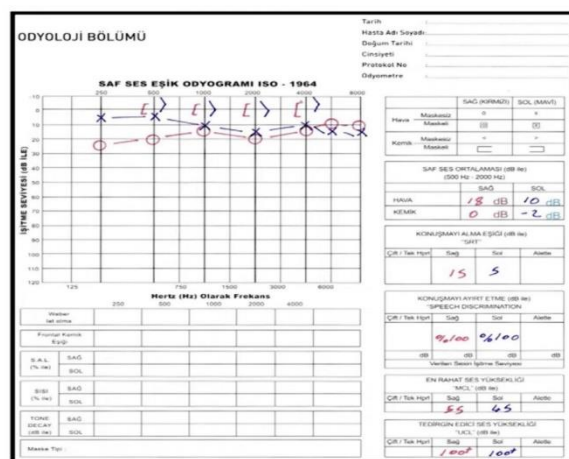
With cone beam computed tomography, the temporal bone can be examined under various planes. In these images, it occurs if there is a deficiency in the superior semicircular canal (Sepúlveda et al., 2014). In Figure 2., a male patient with left superior semicircular canal dehiscence was examined by cone beam computed tomography.



**Figure 2. Male Patient Diagnosed with Left Superior Semicircular Canal Dehiscence. Cone Beam Computed Tomography of The Temporal Bone, (a and d) Coronal, (b) Sagittal, (c) Oblique Sagittal Images Show a Separation in The Left Superior Semicircular Canal (White Arrow) (Sepúlveda et al., 2014)**

In audiological evaluation, pure tone audiometry, conductive hearing loss with negative bone thresholds and usually affects low frequencies, but sometimes sensorineural or mixed hearing loss can also be seen. However, sensorineural or mixed-type hearing loss may also occur. Uncomfortable sound frequencies can range from 250 to 3000 Hz, and nystagmus usually occurs at levels of 100-110 dB. (Weber, 2008). While a single tone might be effective in some cases, usually, a range of frequency levels triggers symptoms. Tympanometric measurements show a Type A pattern, consistent with inner ear pathology. Acoustic reflexes are also obtained. Electrocochleography reveals an increase in the ratio of summation potential to action potential ( $>0.25$ ) (Adams et al., 2011; Arts et al., 2009). Electronystagmography (ENG) can be used to determine the direction and type of nystagmus, but ENG and other vestibular assessment methods like rotational chairs are not believed to provide reliable and accurate results in diagnosing SSCD (Ward et al., 2017; Göçer & Dinç, 2019).

Figure 3. shows the pure tone audiogram of a patient with semicircular canal dehiscence whose bone conduction hearing thresholds reached very low limits.



**Figure 3. Pure-Tone Audiogram of an SSCD Patient (Evren, 2021)**

This study aims to evaluate the diagnostic results of cVEMP and oVEMP tests in SSCD cases. It is important to distinguish which test may be more diagnostically valid and reliable in SSCD cases, as it contributes to the literature.

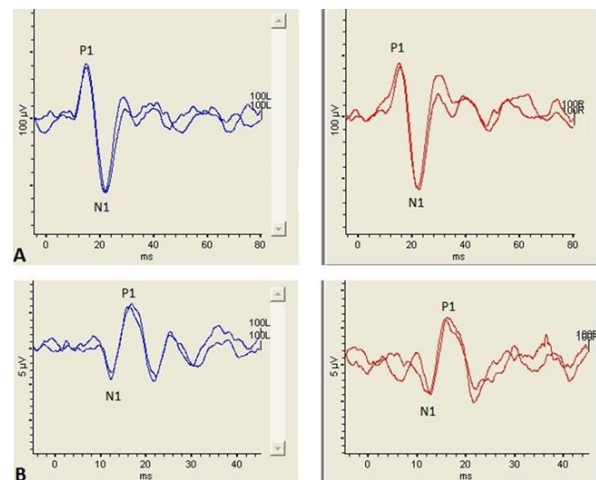
### SSCD Evaluation with cVEMP and oVEMP

In the SSCD diagnosis of VEMP research, its sensitivity was shown to be between 80-100%; and its specificity was shown to be between 90-100%. For this reason, the disease in which VEMP responses are most pathognomonic is SSCD (Rosengren et al., 2019; Ward et al., 2021). The presence of the third window created by SSCD activates the vestibulo-ocular and vestibulo-colic pathways, leading to an increase in VEMP amplitude and a decrease in the threshold. There are two types of VEMP methods: cVEMP and oVEMP. The primary function of cVEMP is to assess saccular function and the inferior vestibular nerve. It involves an inhibitory neural reflex pathway from the saccule to the ipsilateral sternocleidomastoid muscle. Electrodes are placed on the neck muscles (sternocleidomastoid) to measure the cVEMP reflex response. The response consists of a first positive peak of 23 ms, approximately 13 ms after the acoustic stimulus began, followed by a negative peak of about 23 ms (Zuniga et al., 2013). On the other hand, oVEMP is a more recently discovered method that assesses utricular function and the superior vestibular nerve. It involves an excitatory neural reflex pathway from the utricle to the bilateral inferior oblique muscle (Ward et al., 2017). Electrodes are placed over the extraocular muscles to measure the oVEMP reflex response. The response consists of a first negative peak of about 10 ms followed by a positive peak of 16 ms (Todd et al., 2007).

According to Figure 4; the left panel shows cVEMP (top) and oVEMP (bottom) recorded from the left ear, while the right panel displays cVEMP (top) and oVEMP (bottom) recorded from the right ear. (A): cVEMP consists of an initial positive peak (P1) occurring at 13 ms and followed by a negative peak (N1) at approximately 23 ms. (B): In contrast to cVEMP, oVEMP comprises an initial negative peak (N1) at 10 ms, followed by a positive peak (P1) at approximately 15 ms. There is no scaling difference between cVEMP and oVEMP waveforms. cVEMP is significantly larger than oVEMP because it is thought to be due to the fact that the sternocleidomastoid muscle is a much larger muscle than the lower oblique (extraocular) muscle (Piker, 2014).

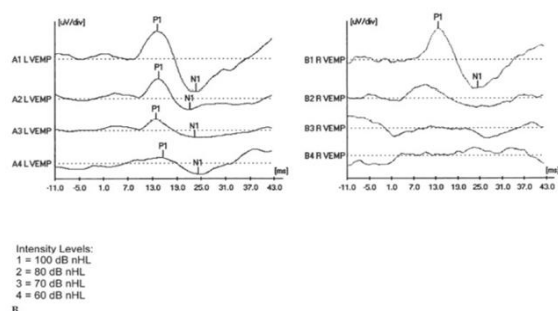
The most powerful aspect of the cVEMP and oVEMP tests is that it is more advantageous than videonistagmography and rotational tests in that it can measure a different part of the vestibular

system (i.e., the otoliths end organs). VEMP tests also enhance their utility by separately evaluating the left and right labyrinths, aiding in localizing VEMPs to the lesioned side. Another advantage is that both tests are relatively fast and well-tolerated by patients. However, a significant limitation of VEMP tests is the reduced response rate in elderly patients. As individuals age, the absence of bilateral VEMP waveforms increases even in healthy controls. Despite these limitations, one of the most valuable applications of the VEMP test is its ability to assist in the diagnosis of superior SSCD (Piker, 2014; Su et al., 2004).



**Figure 4. cVEMP and oVEMP Waveforms Recorded with a 500 Hz Toneburst Stimulus at 100 dB nHL from a Healthy Adult (Piker, 2014)**

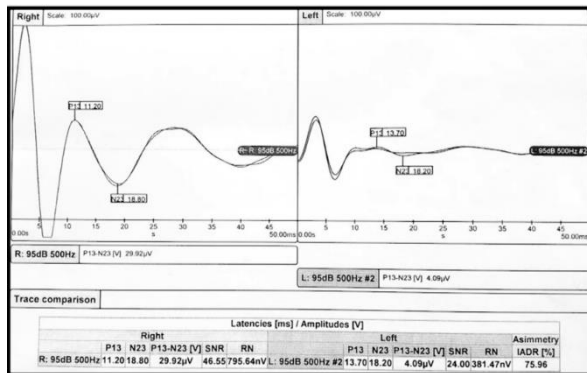
In Figure 5., cVEMP waves were looked at in both ears by changing the stimulus intensity given to the right SSCD phenomenon. According to the severity of the stimulus, cVEMP waves between the ear with and without SSCD were examined (Musiek et al., 2020).



**Figure 5. cVEMP Thresholds Appearance in the Case of Right SSCD (Musiek et al., 2020)**

In Figure 6, the stimulus intensity and frequency were left constant and the cVEMP waveform in

both ears was examined. Latency/amplitude ratio and asymmetry ratio between the two ears were examined (Evren, 2021).



**Figure 6. cVEMP Test of a Right SSCD Patient (Evren, 2021)**

### Research on the Subject

In a prospective study by Zuniga et al. (2014), 29 patients diagnosed with Semicircular Canal Dehiscence and a matched control group of 25 individuals were included. The results showed that cVEMP thresholds exhibited sensitivity and specificity between 80-100% for diagnosing SSCD, while oVEMP amplitudes demonstrated over 90% sensitivity and specificity. Consequently, the study concluded that oVEMP amplitudes were more dominant in diagnosing SSCD compared to cVEMP thresholds (Zuniga et al., 2014).

Govender et al. (2016), included 13 patients diagnosed with SSCD in their study to compare cVEMP and oVEMP threshold and amplitude characteristics. Despite cVEMP showing higher amplitudes, reflex gradients for air and bone-conducted measurements were significantly lower in SSCD compared to normals. However, there was no significant difference in reflex gradients for air and bone-conducted oVEMP. Both cVEMPs and oVEMPs often show amplitude and threshold abnormalities in SSCD compared to normal subjects, but the sensitivities did not differ between measurements (Govender et al., 2016)

Hunter et al. (2016), in a retrospective study of 53 patients with SSCD (average age 52.7 years) were analyzed. oVEMP amplitudes ( $r = 0.61$ ,  $p < 0.0001$ ), cVEMP amplitudes ( $r = 0.62$ , etc,  $p < 0.0001$ ), air conduction thresholds at 250 Hz ( $r = 0.25$ ,  $p = 0.043$ ) and air-bone range at 500 Hz ( $r = 0.27$ ,  $p = 0.01$ ) has been reported to be positively related to the increased separation rate in the superior channel. cVEMP thresholds ( $r = -$

0,56, an inverse relationship was observed between  $P < 0,0001$ ) and the surface area of separation in the superior semicircular canal (Hunter et al., 2016).

Cervical and ocular VEMP peak amplitudes and thresholds of 39 adult patients over 18 years of age with SSCD were compared with 84 age-matched controls. At the end of the study, using receiver operating characteristic (ROC) curves, cVEMP amplitudes, cVEMP thresholds, and oVEMP amplitudes were found to have areas under the curve of 0.731, 0.912, and 0.856, respectively, all of which were statistically significant ( $P < 0001$ ). As a result of the study, the researchers reported that oVEMP and cVEMP amplitudes, cVEMP thresholds, and air conduction thresholds at 250 Hz were significantly correlated with the surface area of dehiscence among patients with confirmed SSCD (Hunter et al., 2017).

In a study conducted by Lin et al. (2019), SSCD was detected by computed tomography imaging in 48 of 171 patients with symptoms consistent with SCD. A n10 response was revealed in 40 of 48 patients (83.3%) with a high-frequency oVEMP test at 4000 Hz. The presence of the n10 response with 4000 Hz oVEMP, a sensitivity of 0.83, a specificity of 0.93, a positive predictive value of 0.83, and a negative predictive value of 0.93 are consistent with the presence of SSCD (Lin et al., 2019).

Hassannia et al. (2021), in a retrospective study included 26 individuals with normal high-resolution computed tomography and oVEMP thresholds of  $\geq 17$ . The aim of the study was to find that the oVEMPs are more sensitive to an SSCD than cVEMPs, and that the oVEMP test in response to sound transmitted by air conduction, it was done to prove that without computed tomography, it could be sufficient on its own without radiation exposure. However, the study concluded that oVEMPs may provide false-positive results for the diagnosis of SSCD, and that high oVEMP amplitude alone is not sufficient for the diagnosis of SSCD (Hassannia et al., 2021).

Maheu et al. (2021), in a study, CT-confirmed unilateral 16, bilateral 10 SSCD-diagnosed patients were included and patients were evaluated with cVEMP and oVEMP tests. As a result of the study, researchers found that the oVEMP (500 Hz) with amplitudes equal to or higher than 10.8  $\mu V$  had a threshold of equal to or

lower than 77.5 dBnHL, or an amplitude of 3.1  $\mu$ V (4 kHz), They report that the most useful test method for identifying SSCD is oVEMP and its use is supported (Maheu et al., 2021)

**Table 1. Table Summarizing the Results of the Study on the Subject**

Study	N	Diagnostic Tools	cVEMP Evaluation		oVEMP Evaluation
Zuniga et al., 2014	29 (St. Gr.) 25 (Co.Gr.)	Physical examination, Audiometric testing, VEMP testing, CT.	<i>Research Parametre: Threshold</i> <i>Stimulus Type: Click</i> Sensitivity, %87 Specificity, %73		<i>Research Parametre: Amplitude</i> <i>Stimulus Type: 500 Hz TB</i> Sensitivity >%90 Specificity >%90 <i>Stimulus Type: Click</i> Sensitivity >%90 Specificity >%90
Govender et al., 2016	13 patients	Physical examination, VEMP testing, CT.	<i>Amplitude</i> 500 Hz TB: Sensitivity, %100 Specificity, %100	<i>Threshold</i> <%90 NR	<i>Amplitude</i> 500 Hz TB: Sensitivity > %90 Specificity, NR <i>Threshold</i> Sensitivity, %100 Specificity, %92
Hunter et al., 2016	53 patients	Physical examination, Audiometric testing, VEMP testing, CT.	<i>Amplitude</i> High sensitivity with surface area of dehiscence	<i>Threshold</i> NR	<i>Amplitude</i> NR
Hunter et al., 2017	39 (St. Gr.) 84 (Co. Gr.)	Clinical Symptoms, VEMP testing (preoperativel)	<i>Amplitude</i> Sensitivity, %73.7 Specificity,	<i>Threshold</i> %86.5 %80.	<i>Amplitude</i> Sensitivity, %71.4 Specificity, %93.9
Lin et al., 2019	171 patients	VEMP testing, CT.		<i>Threshold</i> 500 Hz TB: Sensitivity, % 63 Specificity, %73	<i>Amplitude</i> 500 Hz: 4kHz: Sensitivity, %62 %83 Specificity, %83 %93
Maheu et al., 2021	36 (St. Gr.) 16 (Co. Gr.)	VEMP testing.	<i>Amplitude</i> Sensitivity, %72.22 Specificity, %70.6	<i>Threshold</i> %69.4 %100	<i>Amplitude</i> 500 Hz: Sensitivity, %83.3 Specificity, %87.5 4kHz: Sensitivity, %47.2 Specificity, %100 <i>Threshold</i> 500 Hz: Sensitivity, %83.3 Specificity, %80

**Abbreviations:** Co. Gr.; Control Group, NR; Not Recorded, St. Gr.; Study Group, TB; Tone Burst Stimuli.

By Zhang et al. (2021), clinical factors (autophony, sensitivity to loud sounds, vertigo caused by loud noises, dizziness, etc, hearing their own heartbeat or other internal organs) and various audiological findings (low-frequency conduction type hearing loss, bone conduction hyperacus, low-cVEMP threshold [ $<80$  db nHL], increased oVEMP [ $>17$ mV] amplitudes) is another study investigating the effects of

determining the surgical treatment option. Of the 38 SSCD patients, 28 were included for the surgical treatment option and CT imaging was performed for patients before surgery. As a result of the study, it was reported that among the most powerful factors, low-frequency conduction type hearing loss and increased oVEMP amplitude were significant determinants of the presence of SSCD from logistic regression and to decide on

the diagnosis and surgical option of the disease (Zhang vd., 2021).

The data of the studies are summarized and shown in Table 1.

## CONCLUSION

SSCD is one of the different diseases of the audiovestibular system originating from the inner ear. There is no single gold standard test used to diagnose. The importance of the VEMP test is great in diagnosing the correct SSCD. Pure tone audiometry test, acoustic reflex responses, high-resolution computed tomography and electrocochleography are other evaluation methods that will increase the diagnostic value in SSCD along with the VEMP test. In our study, research on the subject has been compiled in the last 10 years and SSCD patients evaluated with VEMP test have been examined. The common findings across the majority of the included studies highlight a decrease in cervical VEMP (cVEMP) thresholds (<80 dBnHL) and an increase in ocular VEMP (oVEMP) amplitudes (interpeaks and N1) (>17  $\mu$ V, >8.25  $\mu$ V) on the affected sides of SSCD cases. In addition, in most of the included studies, the sensitivity and specificity of the oVEMP amplitude in the diagnosis of SSCD appears to be higher and more dominant, especially in high frequency measurements. However, in a small number of the studies examined, no significant difference was reported between cVEMP thresholds and amplitudes and the sensitivity and specificity of oVEMP amplitudes. In the diagnosis of SSCD disease, the decrease in cVEMP thresholds, as well as the rise of oVEMP amplitudes, the joint evaluation of the two tests and the examination of their results by comparing them, show the importance of diagnostic. On the other hand, according to these results, it should be taken into consideration that the sensitivity and specificity rates of oVEMP and cVEMP may vary due to anatomical source.

## Author Contributions

Idea/Concept: C.U.; Design: C.U., Y.G.E.; Supervision/Consulting: B.H.; Analysis and/or Interpretation: C.U.; Literature Search: C.U., Y.G.E.; Writing the Article: C.U.; Critical Review: C.U

## Peer-review

Externally peer-reviewed

## Conflict of Interest

The authors have no conflict of interest to declare.

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