Scientific Research Publishing

International Journal of Otolaryngology and Head & Neck Surgery, 2022, 11, 82-90
https://www.scirp.org/journal/ijohns
ISSN Online: 2168-5460
ISSN Print: 2168-5452

Bilateral Superior Semicircular Canal Dehiscence without Signs of Vestibular Involvement about a Case

Habib Ould Cheikh El Wely*, Saloua Ouraini, Mohamed Moumni, Jalal Oubenjah, Mohamed Zalagh, Bouchaib Hemmaoui, Nourredine Errami, Fouad Benariba

Otorhinolaryngology and Head & Neck Surgery Department, Mohamed V Military Hospital, Rabat, Morocco
Email: *habibcheikhelwely@gmail.com

How to cite this paper: Ould Cheikh El Wely, H., Ouraini, S., Moumni, M., Oubenjah, J., Zalagh, M., Hemmaoui, B., Errami, N. and Benariba, F. (2022) Bilateral Superior Semicircular Canal Dehiscence without Signs of Vestibular Involvement about a Case. *International Journal of Otolaryngology and Head & Neck Surgery*, **11**, 82-90.
https://doi.org/10.4236/ijohns.2022.112009

Received: February 17, 2022
Accepted: March 19, 2022
Published: March 22, 2022

Abstract

Superior semicircular canal dehiscence (SSCD) is a rare entity recently described whose typical clinical symptomatology is represented by dizziness triggered by a variation of pressure. We reported a case of SSCD which was diagnosed thanks to computed tomography (CT) scan of the petrous bone conducted systematically in front of mixed deafness with normal eardrum. The SSCD was bilateral and was revealed by mixed deafness on the left side and perception deafness on the right with a normal eardrum without the notion of vertigo. The cervical vestibular evoked myogenic potential (cVEMP) and an ultra-high resolution CT scan of the petrous bones in coronal and sagittal sections allowed the diagnosis. The SSCD should be considered in the presence of any conductive or mixed hearing loss with a normal eardrum. The CT scan in coronal and sagittal submillimetric sections allows the diagnosis.

Keywords

Superior Semicircular Canal, Dehiscence, Deafness

1. Introduction

The superior semicircular canal dehiscence (SSCD) is a malformation of the petrous bone which consists of an absence of cortex of the anterior semicircular canal (SCC), more rarely of the posterior SCC; This lack of bone coverage has been suggested in the face of vestibular symptoms induced by a sound stimulation or a pressure variations, generally associated with conductive or mixed hearing loss with a normal eardrum [1] [2].

When it comes to anterior SCC, we talk about Minor's syndrome.

The first description of the syndrome is recent and dates to 1998 in Baltimore, by Minor and his team [3].

The most specific symptom is Tullio's phenomenon, associated with a particular vertigo, nystagmus, a conductive deafness that can complete the picture. It can also manifest as hearing impairment only with no vestibular symptoms.

The high-resolution CT scan of the petrous bone is the examination of choice for the paraclinical diagnosis, with adapted reconstructions and very fine sections to make the diagnosis of the superior semicircular canal dehiscence (SSCD).

Dehiscence can be uni- or bilateral. The rate of bilaterality is on average 25% in the literature, but can reach 50% in certain series [4].

The vestibular evoked myogenic potential (VEMP) test can be a very useful screening tool to assess SSCD, to differentiate between middle and inner ear pathologies [5]. A VEMP response is a measure of the inferior vestibular nerve function [6]. There are two types of VEMP tests: cervical (cVEMP) and ocular (oVEMP). Cervical VEMP is an inhibitory electromyographic signal measured on contracted sternocleidomastoid muscle ipsilateral to the sound-stimulated ear, a consequence of saccular activation. Patients with SSCD have lower than normal threshold to trigger cVEMP responses.

The Video Head Impulse Test (VHIT) in the assessment of superior semicircular canal dehiscence is therefore unlikely to add disease-specific information to understand the severity of symptoms. However, it can aid in decision-making by showing abnormal responses on the opposite side prior to any intervention (which may prevent bilateral vestibular failure) and supplies more information during the informed consent process.

The objective of our work is to report a rare case of bilateral dehiscence of the superior semicircular canal without vestibular signs, but presenting a cochlear symptomatology with in particular a severe mixed deafness on the left side and a deep perception deafness on the right side, and to underline the interest and effectiveness of cochlear implantation (CI) in this rare pathology.

2. Observation

A 60-year-old patient, with no significant pathological history, in particular no notion of trauma, consulted us with the onset of progressively worsening bilateral hypoacusis evolving for 7 years, associated with bilateral tinnitus without any notion of vertigo induced by pressure variations, nor to high intensity sounds. On otoscopy, the eardrums were complete and normal. Neurovestibular examination was normal. Tonal and vocal audiometry was performed showing severe mixed hearing loss on the left side with an average Rinne of 30 dB, deep perception hearing loss on the right and almost zero intelligibility. On impedancemetry, the tympanogram was central with a stapedial reflex present on both sides. Vestibular myogenic evoked potentials (VEMP) (**Figure 1**): cVEMP performed, shows in our patient on the left side a wave present up to 70 dB which is in favor of the dehiscence of the upper left semicircular canal. On the right side we noticed

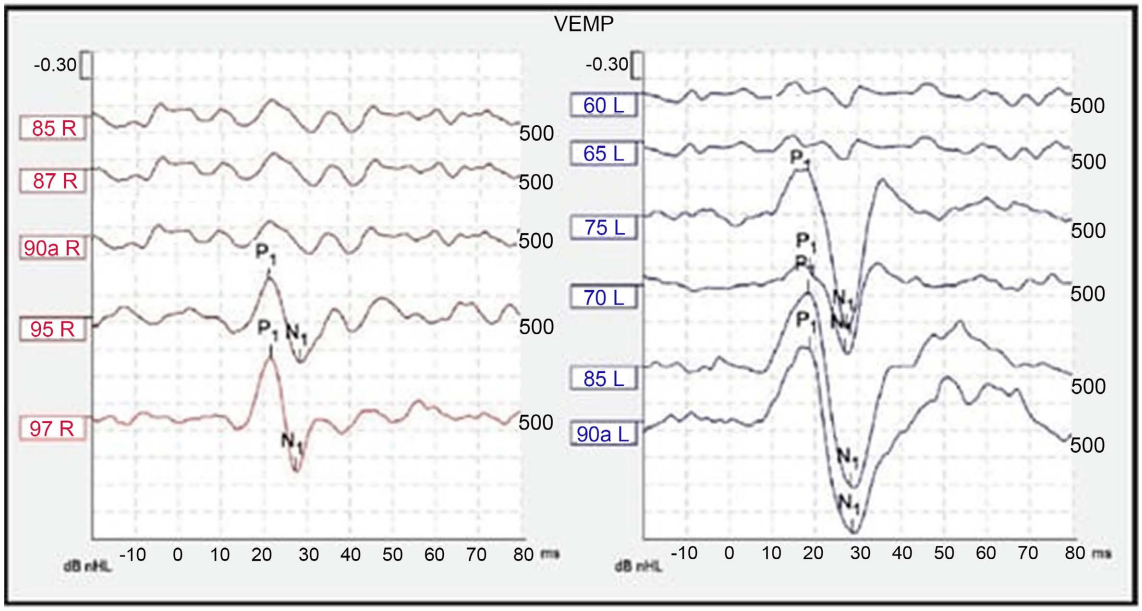


Figure 1. c.VEMP HMIMV Flap: Left side: wave present up to 70 dB. Right side: wave absent below 95DB.

the absence of the wave below 95DB which may be due to a profound right deafness. Video Head Impulse Test (VHIT) (**Figure 2**): showed pathological gains at 0.67 at the left superior CSC and at 0.65 left and 0.62 rights of the homo and contralateral lateral canals. This is consistent with the cVEMP which objectified a decrease in thresholds in the left ear.

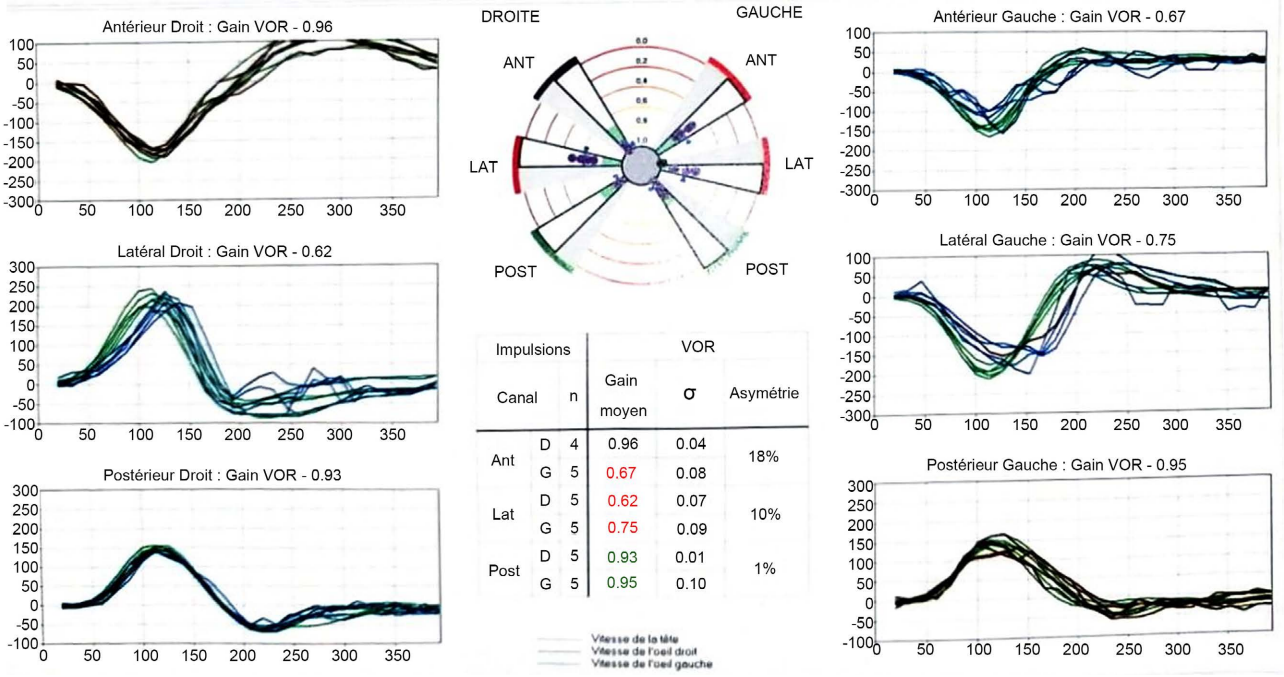
The patient benefited from a scanner of the rocks (**Figure 3**, **Figure 4**) which highlighted the dehiscence of the superior semicircular canal bilaterally, predominant on the left side: the dehiscence is extended over 5.3 mm on the left against 2.2 mm on the right.

The therapeutic abstention for the superior semicircular canals dehiscence was decided in view of the absence of disabling a vertiginous symptomatology. However, she benefited from a cochlear implantation on the right side.

The patient tolerated the operation well. The postoperative course was simple with no complications. She reported a disappearance of tinnitus on the implanted side. The first adjustment took place 20 days after the implantation, she was able to understand a few words immediately, she is currently in her second session of speech therapy, she adapts more and more better; the perception of speech is better at home and by putting the loudspeaker on the telephone. She manages to understand the majority of the words; however in a noisy environment; she still has a problem discerning speech. Her prognosis is good as she is on the 40th day after cochlear implantation and she is already beginning to adapt and report increasingly a better hearing comfort as in patients implanted without SSCD and she still has no vestibular symptoms.

3. Discussion

Superior semicircular canal dehiscence (SSCD) is a rare entity that has recently



Impulsions			VOR		
Canal		n	Gain moyen	σ	Asymétrie
Ant	D	4	0.96	0.04	18%
	G	5	0.67	0.08	
Lat	D	5	0.62	0.07	10%
	G	5	0.75	0.09	
Post	D	5	0.93	0.01	1%
	G	5	0.95	0.10	

Figure 2. VHIT HMIMV Flap: pathological gain in the superior left canal and the homo and contralateral lateral canals.

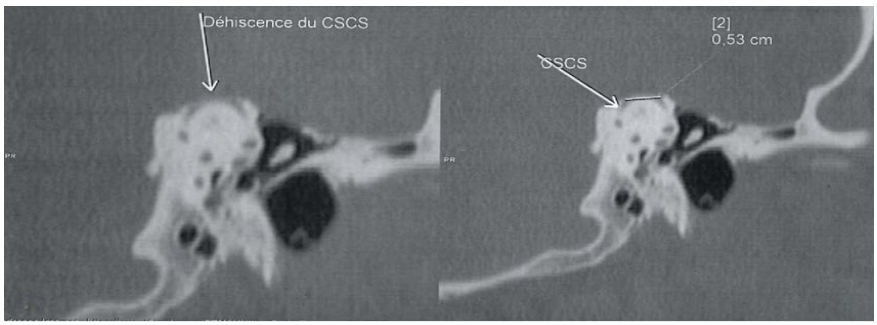


Figure 3. CT of the left petrous bone HMIMV Rabat: dehiscence of the superior semicircular canal extended over 5.3 mm.

Figure 4. CT of the right petrous bone: dehiscence of the superior semicircular canal extended over 2.2 mm.

been described [2] [3]. The prevalence, according to an autooptic study, is 0.67% [3] [4] [5] [6] [7].

SSCD has been described in patients aged 13 - 78 years and appears to be

more common in men [2] [8] [9].

In the literature, many cases have been described, the clinic of which varied from one patient to another.

The dehiscence of the CSCS is sometimes of a posttraumatic origin, the trauma inaugurating the symptomatology [2] [8].

It could be secondary to an anomaly of bone development in the first weeks of life and would become symptomatic following a triggering event, either minimal trauma or pressure variation [9].

SSCD can be unilateral or rarely bilateral. The bilaterally rate varies according to the series between 23.5% and 37.5% [9] [10].

The topography of the dehiscence is variable: the top of the SSC, the posterior part of the SSC at the level of the superior petrosal sinus, the anterior part of the SSC [4] [11]. In our patient, the dehiscence was found at the level of the top of the two superior semicircular canals.

The clinical symptomatology is variable and would depend on the size of the dehiscence and its topography [12] [13]. Dizziness and oscillopsia induced by pressure variations or loud sounds are the most typical clinical symptoms [14] [15] [16]. The clinical examination should look for vertical torsional nystagmus, induced by variations in pressure: Valsalva maneuver, effort with closed glottis, tympanometry inducing a variation in pressure in the external auditory canal.

We must also look for this nystagmus following the exposure to sound with an intensity of 100 to 110 Decibels and a frequency of 500 to 2000 HZ.

Clinical symptoms may consist of conductive or mixed hearing loss [2] [3] [7]. The decrease in bone conduction may only concern the high frequencies or appear from the low frequencies.

The stapedial reflexes are generally present as in the case of our patient. However, in Mikulec's series [8], among eight patients with CSCS dehiscence, three had an abolition of stapedial reflexes. Pulse-synchronous tinnitus is sometimes noted.

However, this SSCD may be asymptomatic [17].

However, it is essential to emphasize that there are people with hearing loss who only have hearing problems and no vestibular symptoms, such as the case of our patient who presented with bilateral sensorineural hearing loss, without any other associated signs.

The conductive hearing loss in the dehiscence of the upper semicircular canal would be explained by the creation of a third window at the level of the inner ear, which will lead to a loss of acoustic energy, hence an increase in threshold air conduction that would lead in parallel to a lowering of the bone conduction threshold by increasing the impedance difference between the round and oval window [15] [18].

This third window mechanism is also involved in other inner ear abnormalities such as wide vestibular aqueduct syndrome and labyrinthine fistulas [15] [18].

Tullio's phenomenon is based on the physiological response of the vestibule to sound stimuli. According to experiments in chinchillas, SSC fenestration lowers the threshold for acoustic stimulation of the vestibule and increases the amplitude of responses [15]. It has also been proved that it engenders hyperexcitability during pressure variations.

The otolith evoked potential tests the vestibulospinal reflex, in case of SSCD, an increase in the amplitude of the P13 and N23 waves is observed [10].

The cervical vestibular evoked myogenic potential (VEMP) test is an inhibitory electromyographic signal measured on contracted sternocleidomastoid muscle ipsilateral to the sound-stimulated ear; a consequence of the saccular activation, in case of SSCD, we observe a lower than normal threshold to trigger cVEMP responses as the case of our patient.

High resolution (1 mm sections) or ultra-high resolution (0.5 or 0.6 mm sections) computed tomography is the key diagnostic examination.

The CT scan of our patient found the existence of a dehiscence of the superior semicircular canal bilaterally; largely predominant on the left side (the dehiscence is extended over 5.3 mm on the left against 2.2 mm on the right). She does not present vestibular symptoms.

However, one must be wary of false images of SSCD linked to the thinness of the roof [12] [13].

Magnetic resonance imaging (MRI) is not indicated for diagnosis and is in the majority of cases interpreted as normal [13].

Surgical treatment is indicated in case of disabling vestibular pathology.

The intervention is a dehiscence patching surgery. It consists of closing the fenestration from an approach to the middle cerebral fossa because it is only way to objectify the dehiscence. A transmastoid approach has been proposed by several authors [4] [11] [18].

The filling materials used are varied: temporal fascia, sawdust, biological glue, cortical bone graft, hydroxyapatite. The postoperative course is marked by an improvement in vestibular symptoms, while hearing generally remains unchanged. Cases of impaired postoperative bone conduction have been reported justifying abstention in the event of a pathology that is not very disabling [4] [11].

In our patient, the reason was a cochlear complaint, in particular a disabling deep bilateral deafness, responsible for social isolation. At first sight, a hearing aid was used as soon as the auditory symptoms appeared without clinical improvement; Powerful prostheses were then considered but the gain remained almost inexistent and the sensory discomfort was manifest. A cochlear implantation was done. It is a hearing rehabilitation device that will improve the patient's quality of life.

Cochlear implantation in patients with SSCD has been the subject of several studies that compare surgical, vestibular, and audiologic outcomes in CI users with SSCD versus CI users with temporal bone anatomy normal. Matic *et al.* (2020) [19] report comparable CI results between recipients with and without

SCD. Specifically, auditory preservation rate and ability to perceive words in the electrical state only do not appear to be defective by SCD, however Puram *et al.* (2015) [20] reported significantly lower word repetition ability in patients with SSCD compared to those without it. However, the reference group and the SSCD group in their study showed a significant difference regarding the duration of hearing loss (249 versus 494 months), which is a known factor influencing the outcome of CI (van den roek and Dunnebier, 2009) [21]. They therefore accounted for the significance of the difference between the two groups being primarily due to a significantly longer duration of hearing loss in the SSCD group.

In our case, we have not yet noticed a difference in the results of cochlear implantation recipients compared to CI without SSCD performed at the service.

4. Conclusions

Any conductive or mixed hearing loss or even sensorineural hearing loss with a normal eardrum should evoke a dehiscence of the semicircular canal higher, although the most typical symptomatology consists of dizziness on exposure to high intensity sounds or during pressure variations.

The diagnosis is confirmed by a high-resolution CT scan of the rock in coronal and axial sections and by reconstructions in the axis of the superior semicircular canal.

Although the cVEMP test is considered the gold standard for identifying SSCD, it is not always able to detect them. In our patient, only the left-sided SSCD was detected, although the contralateral SSCD was radiologically significant.

Surgical treatment is indicated only in case of incapacitating vertigo.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Banerjee, A., Whyte, A. and Atlas, M.D. (2005) Superior Canal Dehiscence: Review of a New Condition. *Clinical Otolaryngology*, **30**, 9-15. https://doi.org/10.1111/j.1365-2273.2004.00940.x

[2] Minor, L.B. (2005) Clinical Manifestations of Superior Semicircular Canal Dehiscence. *The Laryngoscope*, **115**, 1717-1727. https://doi.org/10.1097/01.mlg.0000178324.55729.b7

[3] Minor, L.B., Solomon, D., Zinreich, J.S. and Zee, D.S. (1998) Sound- and/or Pressure-Induced Vertigo Due to Bone Dehiscence of the Superior Semicircular Canal. *Archives of Otorhinolaryngology—Head & Neck Surgery*, **3**, 249-258. https://doi.org/10.1001/archotol.124.3.249

[4] Minor, L.B. (2000) Superior Canal Dehiscence Syndrome. *American Journal of Otolaryngology*, **21**, 9-19. https://doi.org/10.1016/S0196-0709(00)80068-X

[5] Zhou, G., Poe, D. and Gopen, Q. (2012) Clinical Use of Vestibular Evoked Myogen-

ic Potentials in the Evaluation of Patients with Air-Bone Gaps. *Otology & Neurotology*, **33**, 1368-1374. https://doi.org/10.1097/MAO.0b013e31826a542f

[6] Teixido, M.T., Artz, G.J. and Kung, B.C. (2008) Clinical Experience of Symptomatic Superior Canal Dehiscence in a Single Neurotologic Practice. *Otolaryngology—Head and Neck Surgery*, **139**, 405-413. https://doi.org/10.1016/j.otohns.2008.06.023

[7] Carey, J.P., Minor, L.B. and Nager, G.T. (2000) Dehiscence or Thinning of Bone Overlying the Superior Semicircular Canal in a Temporal Bone Survey. *Archives of Otorhinolaryngology—Head & Neck Surgery*, **126**, 137-147. https://doi.org/10.1001/archotol.126.2.137

[8] Mikulec, A.A., McKenna, M.J., Ramsey, M.J., *et al.* (2004) Superior Semicircular Canal Dehiscence Presenting as Conductive Hearing Loss without Vertigo. *Otology & Neurotology*, **25**, 121-129. https://doi.org/10.1097/00129492-200403000-00007

[9] Brantberg, K., Bergenius, J., Mendel, L., Witt, H., Tribukait, A. and Ygge, J. (2001) Symptoms, Findings and Treatment in Patients with Dehiscence of the Superior Semicircular Canal. *Acta Oto-Laryngologica*, **121**, 68-75. https://doi.org/10.1080/000164801300006308

[10] Streubel, S.O., Cremer, P.D., Carey, J.P., Weg, N. and Minor, L.B. (2001) Vestibular-Evoked Myogenic Potentials in the Diagnosis of Superior Canal Dehiscence Syndrome. *Acta Oto-Laryngologica*, **121**, 41-49. https://doi.org/10.1080/000164801750388090

[11] Ramsey, M.J., McKenna, M.J. and Barker, F.G. (2004) Superior Semicircular Canal Dehiscence Syndrome. Case Report. *Journal of Neurosurgery*, **100**, 123-124. https://doi.org/10.3171/jns.2004.100.1.0123

[12] Williamson, R.A., Vrabec, J.T., Coker, N.J. and Sandlin, M. (2003) Coronal Computed Tomography Prevalence of Superior Semicircular Canal Dehiscence. *Otolaryngology—Head and Neck Surgery*, **129**, 481-489.

[13] Barton, F.B., Harrigal, C. and Hirsch, B.E. (2006) Superior Semicircular Canal Dehiscence: Oblique Reformatted CT Images for Diagnosis. *Radiology*, **238**, 938-942. https://doi.org/10.1148/radiol.2382042098

[14] Rosowski, J.J., Songer, J.E., Nakajima, H.H., Brinsko, K.M. and Merchant, S.N. (2004) Clinical, Experimental and Theoretical Investigations of the Effect of Superior Semicircular Canal Dehiscence on Hearing Mechanisms. *Otology & Neurotology*, **25**, 323-332. https://doi.org/10.1097/00129492-200405000-00021

[15] Hirvonen, T.P., Carey, J.P., Liang, C.J. and Minor, L.B. (2001) Superior Canal Dehiscence: Mechanisms of Pressure Sensitivity in a Chinchilla Model. *Archives of Otorhinolaryngology—Head & Neck Surgery*, **127**, 1331-1336. https://doi.org/10.1001/archotol.127.11.1331

[16] Carey, J.P., Hirvonen, T.P., Hullar, T.E. and Minor, L.B. (2004) Acoustic Responses of Vestibular Afferents in a Model of Superior Canal Dehiscence. *Otology & Neurotology*, **25**, 345-352. https://doi.org/10.1097/00129492-200405000-00024

[17] Crovetto, M., Areitio, E., Elexpuru, J. and Aguayo, F. (2008) Transmastoid Approach for Resurfacing of Superior Semicircular Canal Dehiscence. *Auris Nasus Larynx*, **35**, 247-249. https://doi.org/10.1016/j.anl.2007.06.010

[18] Kharrat, S., Dridi, I., Boulakbech, R., El Aoud, C., Tababi, S., Zainine, R., Beltaief, N., Menif, E., Sahtout, S. and Besbes, G. (2010) La déhiscence du canal semi circulaire supérieur. *Journal Tunisien d'ORL et de Chirurgie Cervico-Faciale*, No. 24, 91-93. https://doi.org/10.4314/jtdorl.v24i1.64252

[19] Matica, J., Winklhofer, S., Pfiffner, F., Roosli, C., Veraguth, D., Huber, A. and Dalbert, A. (2020) Influence of Semicircular Canal Dehiscence on Cochlear Implant Outcome. *Audiology and Neurotology*, **26**, 135-139. https://doi.org/10.1159/000508892

[20] Puram, S.V., Roberts, D.S., Niesten, M.E., Dilger, A.E. and Lee, D.J. (2015) Cochlear Implant Outcomes in Patients with Superior Canal Dehiscence. *Cochlear Implants International*, **16**, 213-221. https://doi.org/10.1179/1754762813Y.0000000044

[21] Van den Broek, E. and Dunnebier, E.A. (2009) Cochlear Implantation in Postlingually Hearing-Impaired Adults: Choosing the Most Appropriate Ear. *International Journal of Audiology*, **48**, 618-624. https://doi.org/10.1080/14992020902931566