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Power Reflectance as a Screening Tool for the Diagnosis of Superior Semicircular Canal Dehiscence

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Abstract

Hypothesis—Power reflectance (PR) measurements in ears with Superior Canal Dehiscence (SCD) have a characteristic pattern, whose detection can assist in diagnosis.

Background—The aim of this study is to determine if PR coupled with a novel detection algorithm can perform well as a fast, non-invasive, and easy screening test for SCD. The screening test is to determine if patients with various vestibular and/or auditory symptom(s) should be further considered for more expensive and invasive tests that better define the diagnosis of SCD (and other third-window lesions).

Methods—PR was measured in patients diagnosed with SCD by high-resolution CT. The study included 40 ears from 32 patients with varying symptoms (e.g., with and without conductive hearing loss, vestibular symptoms, and abnormal auditory sensations).

Results—PR results were compared to previously published norms, and showed that SCD is commonly associated with a PR notch near 1 kHz. An analysis algorithm was designed to detect such notches and to quantify their incidence in affected and normal ears. Various notch detection thresholds yielded sensitivities of 80%–93%, specificities of 69%–72%, negative predictive value of 84%–93% and positive predictive value of 67%.

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Conclusion—This study shows evidence that PR measurements together with the proposed notch-detecting algorithm can be used to quickly and effectively screen patients for third-window lesions such as SCD in the early stages of a diagnostic workup.

Keywords

superior semicircular canal dehiscence; SCD; power reflectance; reflectance; energy reflectance; wideband acoustic immittance; admittance

Introduction

Superior canal dehiscence (SCD) is considered rare, but since its initial description by Minor in 1998 (1), identification of patients with SCD syndrome (SCDS) continues to increase with awareness of this condition and improvements in diagnostic methods. The clinical diagnosis of SCDS is generally suspected in the subset of patients with signs and symptoms of: a) a vestibular nature such as dizziness and vertigo induced by noise (Tullio's) or pressure (Hennebert's), and/or b) an auditory nature such as low-frequency conductive hearing loss with normal tympanometry and stapedial reflexes and supranormal bone conduction on pure tone audiometry as reflected by hypersensitivity to bone-conducted sounds (e.g., hearing eye movements or footfalls). However, patients with SCDS may present with other common symptoms that mimic a number of diseases frequently encountered by otolaryngologists. These symptoms include non-specific intermittent dizziness, unsteadiness, aural fullness, or autophony (2). Wrong diagnoses have resulted in the delay of proper treatment, consultation of multiple specialists, and unnecessary surgery. These diagnoses include: psychiatric disease, migraine, Ménière's disease, Eustachian tube dysfunction (leading to tympanostomy tube placement), and middle ear disease (resulting in middle ear exploration or stapedectomy).

Various tests are used today to help confirm the diagnosis of SCD in patients who have radiologic imaging evidence of a bony defect, including cervical vestibular evoked myogenic potential (cVEMP) (3). Zuniga et al. (4) reported high sensitivity and specificity for cVEMP to diagnose SCD. However, cVEMP presently falls short due to: a) lack of standardization, b) lack of appropriate signal processing schemes (to account for differences in background muscle activity, muscle mass, and fatigue), c) lack of artifact rejection schemes for detection in noise, and d) inconsistencies across institutions and subjects (5). Although cVEMP testing has not yet received FDA approval, many institutions perform cVEMP in the workup of a patient in whom SCDS is suspected. Ocular vestibular evoked myogenic potential (oVEMP) has been recently shown to be useful for diagnosing SCDS (4); however, few institutions perform this test. In the context of conductive hearing loss, stapedial reflex testing is useful, as the reflex is often present in SCDS and absent in other conductive pathologies (2). Nevertheless, the stapedial reflex may be absent in SCD ears due to other factors. Another characteristic in SCDS patients is hypersensitive low-frequency bone conduction (2, 6), but the prevalence of this marker is not well known. From the series of patients used in Niesten et al. (7), we find that 57% (73/129 ears) have bone conduction thresholds better than 0 dB at one or more frequencies (unpublished data).

The reference for the diagnosis of anatomic SCD is high-resolution computed tomography (CT). However, if only common non-specific vestibular or auditory symptoms are present, SCDS is not suspected during the initial workup, and a high-resolution temporal bone CT scan is not performed. A simple, inexpensive non-invasive SCD diagnostic screening test that would aid a general otolaryngologist in the initial consultation would be of value to determine if further testing (CT and VEMP) should be performed. Additionally, very small dehiscences (<0.5 mm diameters) that cannot be detected by CT might result in significant symptoms [e.g., small dehiscences have been shown to sometimes produce larger changes in mid-frequency intracochlear pressure than large dehiscences (8)]. Consistent with this finding, patients with thin bone over the superior semicircular canal with SCDS had reduced symptoms by surgical plugging (9). Furthermore, the bony wall of the semicircular canal can be uneven and ‘scalloped’ (10). This structure may result in multiple micro-openings when the bone is thinned, leading to similar effects like very small dehiscences (8).

In the past we have demonstrated that *in cases of conductive hearing loss*, non-invasive measurements of sound-induced umbo velocity using laser-Doppler vibrometry or power reflectance (PR) can reliably differentiate between SCD and other conductive lesions (11, 12). Reflectance (R) is the complex ratio between the reflected pressure wave and the incident pressure wave propagating in the ear canal. PR is calculated as the square of the magnitude of the reflectance, $PR=|R|^2$ where PR generally ranges between 0 and 1 (where 1 indicates all energy is reflected and 0 indicates all energy is absorbed).

In this study we determine if PR measurements (an inexpensive FDA-approved test that is easily performed), coupled with a new detection algorithm sensitive to specific SCD features, can act as a simple non-invasive SCD screening test for patients *with varying symptoms (vestibular and/or hearing related)*. Such a screening test could be helpful at primary centers to provide an indication of SCD with high sensitivity and reasonable specificity, helping to determine whether more expensive or invasive diagnostic procedures are warranted.

Materials and Methods

This work was approved by the institutional review board of the Massachusetts Eye and Ear Infirmary (MEEI). We recruited 50 patients with 59 ears that were diagnosed with SCD by high-resolution CT. We utilized specialized methods in conjunction with CT measurements for determining the size of the dehiscence as described below and in more detail in Niesten et al. (7). A total of 32 patients (17 females and 15 males) met the following inclusion criteria: a) presence of SCD on CT scan, b) absence of any middle ear disease such as cholesteatomas or tympanic membrane (TM) lesions, and c) absence of previous ear surgery except for placement of tympanostomy tubes more than two years before measurement. The mean age was 48.1 years, ranging from 25 to 69 years old. Eight patients had bilateral SCD, resulting in the inclusion of a total of 40 ears with SCD. Of these 40 ears, 27 were on the left side, and 13 on the right side. They were referred for PR measurements from the Otologic Clinic at the Massachusetts Eye and Ear Infirmary between January 2010 and July 2012.

All patients included in this study reported at least one sign or symptom such as autophony, fullness of the ear, hyperacusis (including the sensation of hearing one's eye motion, pulse or footsteps), tinnitus, hearing loss, and/or various forms of dizziness or unsteadiness. The TM appeared normal on microscopic observation in all patients. Patients underwent audiometric air conduction and bone conduction threshold testing, and most underwent stapedial reflex testing, tympanometry, and cVEMP testing. A summary of all of the testing results in our population can be found in Table 1.

All audiologic testing was performed by audiologists at the Massachusetts Eye and Ear Infirmary using standard techniques (12, 13). Conductive hearing loss was defined as mean air-bone gap (ABG) between 250 and 1000 Hz of 10 dB or more. Seventeen out of the 40 SCD ears (42.5%) fulfilled this criterion. Bone conduction thresholds were evaluated at levels as low as 10 dB HL in all patients, and hypersensitive bone conduction (less than 0 dB HL at one or more frequencies) was noted in 19/40 patients with SCD. Acoustic reflex was performed in 16/17 ears with conductive hearing loss and was present (reflex elicited for <95 dB HL) in 14/16 (87.5%) ears. Reflex testing was performed in 13/23 ears without conductive hearing loss and found present in 11/13 (84.6%). Thirty-seven ears underwent 226 Hz tympanometry with normal results in 35/37 ears (2 had sharp peaks consistent with increased mobility).

Because of variability across institutions, we included in our analysis only the 33/40 SCD ears with cVEMP tests performed at the MEEI. The cVEMP thresholds at 250, 500, and 1000 Hz were compared to the 95% confidence interval of established thresholds for normal subjects (14). If two or more of the three frequencies had thresholds below the normal confidence limits (peak sound pressure of 105 dB at 250 Hz, 95 dB at 500 Hz, 100 dB at 1000 Hz), the test was considered suggestive of SCDS (14). Sixteen of the 33 tested ears (48.5%) showed measurements consistent with SCDS.

In the 38 patients with high-resolution CT scans performed at MEEI, the length of the SCD was determined using the analysis methods described in Niesten et al. (7). High-density temporal bone CTs were analyzed by viewing the oblique multiplanar reformatted images while making a curved planar reconstruction of the superior semicircular canal. From the reconstruction, 0.2 mm thick radial sections were analyzed for radiodensity of the bone overlying the superior semicircular canal. An optical density of <300 Hounsfield Units was defined as absence of bone.

PR was measured with an FDA-approved commercially-available Mimosa Acoustic HearID system (Champaign, IL USA). PR measurements used wideband (0.2–6 kHz) chirp stimuli of 60 dB SPL intensity and three-second duration with individually calibrated acoustic systems as previously described (12, 13).

Results

Power Reflectance (PR)

Three representative PR measurements from ears with SCD are plotted in Fig. 1A, along with $\pm$ one standard deviation around the mean (gray region) determined from 58 normal

ears (13). Most of the measurements from SCD ears had a prominent notch-like local minimum centered between 0.6 and 1.8 kHz that was more than 1.5 standard deviations below the normal mean, followed by a local maximum at a higher frequency. Figure 1B plots the mean (solid black line) of 40 SCD ears and $\pm$ one standard deviation (dashed lines). The normal ears are plotted as $\pm$ 1 standard deviation (gray region) around the mean (gray line). Averaging across ears smoothes out the notches that occur at different frequencies in the different SCD ears, but the SCD mean near 1 kHz is still about one standard deviation lower than normal mean.

Notch-detection algorithm for PR measurement to diagnose SCD

As shown in Fig. 1A, ears with SCD generally exhibited a notch near 1 kHz in the PR curves. To determine if this feature in the PR can be used as a diagnostic indicator for SCD, we developed a ‘notch-detection’ algorithm (implemented in MATLAB) to identify their occurrence in individual measurements.

The simple notch-detection algorithm relies on three parameters that allow for variations in the notch shape and frequency range. Details of the algorithm are described in the Supplemental Digital Content available with this manuscript. Figure 2 is a representative PR difference curve (the difference between a PR response from an individual with SCD and the mean PR of normal ears). Three parameters can be adjusted in the algorithm: notch frequency range, minimum notch depth, and minimum notch size. This algorithm determines the presence of a V-shaped notch, within the parameters that determine the range of shape and size, in a particular frequency range. Figure 2 shows an example where the algorithm has sensed the existence of a notch.

We were able to adjust the three parameters in the algorithm (*notch frequency range*, *minimum notch depth*, *minimum notch size*) to distinguish between SCD and normal ears with high sensitivity and moderately high specificity. Detection thresholds of the above parameters were originally determined based on optimization around a subset of the data (26/40 ears) together with the normal population of 58 ears. Details of how we determined optimized parameters are described in the Supplemental Digital Content. Later, the same method of determining the best parameter values were repeated for the 40 SCD ears with respect to the 58 normal ears, resulting in the same optimized parameter set, which demonstrated the validity of the selected parameter values. Fig. 3 A & B show receiver operating characteristic (ROC) curves calculated for the 40 SCD ears and 58 normal ears for parameter variations. In Figure 3A, we compare the use of *notch size* as a decision variable after first applying two different *minimum notch depths*. In Figure 3B we look at the use of *notch depth* as a decision variable after first applying a *minimum notch size*. A *minimum notch size* on the order 0.1 and a *minimum notch depth* between 0.05 and 0.1 were found to separate most SCD ears from normals.

After defining useful parameters, the performance of the diagnostic screening test was quantified. The algorithm was used on separate individual PR measurements (40 SCD and 58 normal ears). Optimal sensitivity occurred with a *notch frequency range* of 585–1876 Hz, a *minimum notch size* of 0.097, and a *minimum notch depth* of 0.05. Thirty-seven out of 40 SCD ears and 18/58 normal ears were considered positive, resulting in a sensitivity of 93%,

a specificity of 69%, a positive predictive value (PPV) of 67%, and a negative predictive value (NPV) of 93%. If the *minimum notch depth* was increased to 0.09, then we reached optimum specificity with 32/40 SCD ears and 16/58 normal ears considered positive for SCD, resulting in a sensitivity of 80%, specificity of 73%, a PPV of 67%, and NPV of 84% (see Table 2).

Effect of SCD size and air-bone gap on the PR notch size

The anatomical length of the SCD determined from CT scans (n=38) varied between 1–7.8 mm. Linear regression analysis showed a marginally significant correlation ($p=0.053$) between SCD length and PR *notch size*. A significant correlation ($p=0.008$) was observed between the PR *notch size* and the averaged ABG between 250–1000 Hz (n=39; the ABG could not be computed in one ear due to a profound sensorineural hearing loss).

Discussion

Power reflectance

PR measurements are easy and fast using a relatively inexpensive FDA-approved device. Minimum training is required to insert an ear tip (similar to an earplug) into the ear canal, and to run the computer or control the machine. Two companies (Mimosa Acoustics and Interacoustics) provide FDA approved devices that measure PR. Both companies have various measuring devices that range in price up to \$10,000 USD. To further aid otologic diagnoses, various PR models can also perform otoacoustic emission measurements and simple tympanometry. Both companies work to incorporate research findings into diagnostic paradigms such as those of Nakajima et al. (12). The algorithm we present here could be internalized in their equipment to allow for automatic diagnostic estimates. We have no financial relationship with any company selling these instruments.

We show that an algorithm to sense notches in the PR measurements usually seen in SCDS has promise as a screening procedure for SCD. If SCDS is suspected based on this simple, non-invasive diagnostic test, then more costly, invasive and time-consuming diagnostic procedures (e.g. high-resolution CT scan and cVEMP) can be considered. This is particularly helpful for patients with non-specific auditory and vestibular symptoms that mimic other common pathologies. Furthermore, as shown in Nakajima et al. (12), if a patient has only a conductive hearing loss (without other symptoms), then PR in conjunction with the audiometric data can be used to differentiate between various causes of conductive hearing loss – ossicular fixation, ossicular disarticulation, and SCD. Thus, PR measurements can be utilized early in the assessment of a patient with vestibular and/or audiologic symptoms to reduce misdiagnoses, inappropriate treatment, unnecessary surgery and the need for more costly and invasive diagnostic procedures.

The notch in PR

In most of the ears with CT-confirmed SCD and normal-appearing TM, the PR curves show a notch near 1 kHz. Depending on the parameters used to define the notch, our notch-detecting algorithm was able to provide separate SCD from non SCD ears with sensitivities of 80% to 92%, specificities of 69% to 72%, NPV scores 84% to 93% and a moderate PPV

score of 67%. The high NPV suggests PR with the proposed algorithm can be a useful tool in the initial diagnostic screening of patients with vestibular and/or auditory symptoms by ruling out SCD in patients with normal PRs.

However, there are limitations to PR. PR can be affected by the condition of the TM. An ear with normal audiogram but flaccid TM (sensed by tympanometry) may exhibit a notch similar to that seen in SCD (13). PR can also exhibit a notch in ossicular interruption, although the notch is more prominent and tends to occur at lower frequencies than in SCD (12). This study showed that the PR notches detected by our algorithm can occur in normal ears (16–18 ears out of the 58 normal ears, resulting in specificity of 72% to 69%). Thus, our algorithm is suited as a screening tool due to its moderate specificity.

The PR notch near 1 kHz seen in ears with SCD is likely related to the effect of inner-ear dehiscence on cochlear impedance and ossicular motion. Such notches could result from a decrease in cochlear damping, which would exaggerate any TM-ossicular resonances and introduce a notch. Alternatively, a shift in middle ear resonance frequency due to a change in the total stiffness or inertance of the middle and inner ear can produce a peak or notch in the response referenced to normal mean. Future experiments in cadaveric temporal bones and computational models may aid in the understanding of the mechanism behind the significant effect of SCD on PR.

cVEMP Testing

cVEMP has been proposed for diagnosing various vestibular diseases as well as third window lesions such as SCD (15). Third window lesions have been associated with 10–20 dB decreases in low-frequency cVEMP thresholds (2, 5, 16–20). However, patients may have difficulty completing the testing if they have severe Tullio's phenomenon or limited neck motion. Furthermore, cVEMP responses vary with muscle mass, tone, activity, and fatigue and are greatly affected by movement artifacts (5). In this study, we noted that a significant fraction of our SCD ears (17/33) had normal cVEMP responses (95% confidence interval of normal ears).

Effect of SCD size or air-bone gap on PR notch size

In our series we only found a marginally significant correlation of SCD size with respect to PR notch size. On the other hand, significant correlation did exist between PR notch size and ABG. An increase in the PR notch size can be interpreted as a decrease in the impedance that the stapes experiences at the oval window. As the cochlear input impedance decreases due to the third window, the ABG increases due to the decrease in pressure difference across the cochlear partition (21, 8). Somewhat surprisingly, in a larger series the size of the SCD correlated significantly with ABG (7), whereas we found that PR notch depth was significantly correlated with ABG but only marginally with SCD size. The methods for quantifying the ABG were similar in both studies. An increase in the number of PR measurements for SCD may resolve this seeming conflict.

Conclusion

This study provides evidence that PR in conjunction with a new algorithm to detect certain features in PR response can be used to screen patients for SCDS in the early stages of a diagnostic workup. If the PR is consistent with SCD, then more expensive and invasive diagnostic procedures can be considered. In addition, if a patient has a conductive hearing loss, PR can also differentiate between various causes of conductive hearing loss in an intact TM with aerated middle ear (12). PR can aid in the early stages of diagnostic workup to enable earlier diagnosis and prevent unnecessary treatment.

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Figure 1.

(A) Three representative examples of PR recorded from ears with SCD. The gray shaded region represents one standard deviation around the mean of 58 normal ears. Most ears with SCD have a distinctive notch near 1 kHz. (B) PR mean (black solid line) and one standard deviation around the mean (dashed line) for ears with SCD (n=27). PR mean (gray solid line) and one standard deviation around the mean (gray shading) for normal ears.

Figure 2.

The difference in PR between an example SCD and normal mean. The description of this figure, the algorithm to determine whether a notch is found (if so, compute the notch size), is described in the results section and in the Supplemental Digital Content.

Figure 3.

(A) Receiver operating characteristic (ROC) curves constructed as *minimum notch size* is varied with a fixed *notch frequency range* of 585-1876 Hz, and a *minimum notch depth* of either 0.05 or 0.09. As *minimum notch size* decreases, sensitivity increases and specificity decreases.

(B) ROC curve constructed as *minimum notch depth* is varied with a fixed *notch frequency range* of 585-1876 Hz, and a *minimum notch size* of 0.097. As *minimum notch depth* decreases, sensitivity increases and specificity decreases.

Table 1

Summary of audiologic data. CHL is conductive hearing loss.

Summary of Audiologic Data		
Test Procedure/Condition	affected/tested	% ears affected
Conductive Hearing Loss	17/40	42.5%
Hypersensitive Low-Freq Bone Conduction	19/40	47.5%
Acoustic Reflex w/CHL	14/16	87.5%
Acoustic Reflex w/o CHL	11/13	84.6%
Normal Tympanometry	35/37	94.6%
cVEMP Suggestive of SCD	16/33	48.5%

Table 2

Summary of detection performance of the notch detection algorithm.

SCD Detection Performance with a Notch Frequency Range of 585–1876 Hz, a Minimum Notch Size of 0.097 and two different Minimum Notch Depths		
	Minimum Notch Depth 0.05	Minimum Notch Depth 0.09
Sensitivity	92.5%	80%
Specificity	69%	72.4%
Positive Predictive Value	67%	66.6%
Negative Predictive Value	93%	84%
Notch Detected for SCD Ears	37/40	32/40
Notch Detected for Normal Ears	18/58	16/58