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The effect of increased intracranial pressure on vestibular evoked myogenic potentials in superior canal dehiscence syndrome

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HIGHLIGHTS

- In normal controls and patients with superior canal dehiscence syndrome (SCDS), ocular VEMP amplitudes significantly decreased in response to inversion.
- There was not a differential change in VEMP amplitude with inversion between SCDS and normal subjects.
- Consistent with previous findings, subjects with SCDS demonstrated significantly higher ocular VEMP amplitudes than control subjects.

ABSTRACT

Objective: To determine if vestibular evoked myogenic potential (VEMP) responses change during inversion in patients with superior canal dehiscence syndrome (SCDS) compared to controls.
Methods: Sixteen subjects with SCDS (mean: 43, range 30–57 years) and 15 age-matched, healthy subjects (mean: 41, range 22–57 years) completed cervical VEMP (cVEMP) in response to air conduction click stimuli and ocular VEMP (oVEMP) in response to air conduction 500 Hz tone burst stimuli and midline tap stimulation. All VEMP testing was completed in semi-recumbent and inverted conditions.
Results: SCDS ears demonstrated significantly larger oVEMP peak-to-peak amplitudes in comparison to normal ears in semi-recumbency. While corrected cVEMP peak-to-peak amplitudes were larger in SCDS ears; this did not reach significance in our sample. Overall, there was not a differential change in o- or cVEMP amplitude with inversion between SCDS and normal subjects.
Conclusions: Postural-induced changes in o- and cVEMP responses were measured in the steady state regardless of whether the labyrinth was intact or dehiscent.
Significance: VEMP responses are blunted during inversion. Although steady-state measurements of VEMPs during inversion do not increase diagnostic accuracy for SCDS, the findings suggest that inversion may provide more general insights into the equilibration of pressures between intracranial and intralabyrinthine fluids.

1. Introduction

Superior canal dehiscence syndrome (SCDS) consists of an opening in the bony roof over the superior semicircular canal (Minor et al., 1998). This dehiscence is considered a pathologic third window in the labyrinth and is thought to produce inner ear conductive hearing loss by shunting air-conducted energy through the dehiscence while enhancing bone conducted energy due to the lower impedance of scala vestibuli (Merchant and Rosowski, 2008; Minor et al., 2003). The dehiscence also serves as a conduit for energy transmission to and from the cranial cavity, resulting in other unique patient symptoms such as increased

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sensitivity to bone conducted physiological sounds (i.e., hearing their eyes move, digestive sounds, etc.) and an increased sensation in the loudness of one's own voice (autophony) (Crane et al., 2010). Further evidence of this pressure transduction through the dehiscence is the presence of eye movements in the plane of the superior canal in response to loud sounds (Tullio phenomenon) or pressure changes (Hennebert sign) (Minor et al., 1998, 2001). These symptoms usually improve or resolve upon canal plugging (Crane et al., 2010; Minor et al., 2003; Minor, 2005).

SCDS is typically diagnosed using a combination of ocular and cervical vestibular evoked myogenic potential (VEMP) testing, audiometric testing and computed tomography (CT). The VEMP measures muscle potential changes at either the inferior oblique muscle for the ocular VEMP (oVEMP), or the sternocleidomastoid muscle for the cervical VEMP (cVEMP) (Colebatch et al., 1994; Weber et al., 2012). Considerable work has been published regarding the clinical utility of these responses; however, the origin and response patterns remain debated. While the cVEMP has been regarded as an inhibitory response, primarily saccular in origin and the oVEMP regarded as an excitatory response, primarily utricular in origin (Colebatch et al., 1994; Curthoys et al., 2011; Manzari et al., 2010a, 2010b), there is debate that the origin and response patterns are not this concrete (Todd, 2014). Evidence exists suggesting that the utricle may have both inhibitory and excitatory contributions to the cVEMP (Rosengren et al., 2009) and the saccule extends projections centrally via both the inferior and superior vestibular nerves (Terasaka et al., 2000; Todd, 2014). Nonetheless, SCDS results in hypersensitivity of these responses, which is demonstrated by lower thresholds and higher amplitudes on both c- and oVEMP testing (Brantberg et al., 1999; Janky et al., 2012; Manzari et al., 2012; Rosengren et al., 2008; Streubel et al., 2001; Welgampola et al., 2008; Zuniga et al., 2012). In addition, patients with SCDS may also demonstrate conductive hearing loss that is not of middle ear origin and visible dehiscence on CT.

The current clinical challenge in SCDS diagnosis is determining when sufficient energy transfer is occurring through the dehiscence to warrant surgical intervention. In cases with obvious nystagmus induced by loud sounds or changes in intracranial or middle ear pressure, there is little question that the dehiscence is responsible for abnormal cupular stimulation from fluid motion within the labyrinth as a response to these stimuli, especially in light of the fact that evoked nystagmus and vertigo resolve with surgical correction of the dehiscence (Crane et al., 2008). However, when patients present with a CT scan showing apparent dehiscence, but lack validating clinical findings (i.e., inducible nystagmus) of a patent dehiscence, the decision to initiate surgical intervention is difficult. In some of these patients, the isolated finding of bony conductive hyperacusis can indicate a patent dehiscence. In this cohort, low-frequency air-bone gaps and negative bone conduction thresholds may provide diagnostic evidence to justify surgery as patients with hyperacusis indeed benefit from SCDS surgery (Crane et al., 2010). The patients who particularly present a dilemma for surgery candidacy are those having apparent dehiscence on CT with subjective symptoms of imbalance, sound sensitivity, and autophony, but without definitive evidence of SCDS on secondary vestibular and audiometric tests. For this group of patients, there is a need for more objective measures of actual pressure transmission through their apparent dehiscence prior to surgical candidacy.

The present study seeks to fill this gap with a novel measure that might reflect pressure transmission through a dehiscence: postural changes in VEMPs. We sought first to build upon our recent finding that cases of surgically-proven SCDS usually have abnormal preoperative VEMP responses. Findings indicate that cVEMP thresholds and suprathreshold oVEMP amplitudes provide excellent sensitivity and specificity when obtained in response to air conducted sound (ACS) stimulation (500 Hz toneburts or click stimuli) compared to an age-matched population (Janky et al., 2012; Zuniga et al., 2012). These VEMP responses are believed to be enhanced in SCDS due to lower overall input impedance for ACS to enter the otic capsule (Merchant and Rosowski, 2008). We reasoned that pressure transmission across the dehiscence could be modulated by changing intracranial pressure (ICP) with postural changes because increasing ICP should increase the overall input impedance, bringing it closer to the impedance of the labyrinth without SCDS. For this reason, we sought to determine whether we could document a change in the VEMP response going from a semi-recumbent to inverted position, a measure that should reflect the degree of pressure transmission through the dehiscence. Specifically, the goal of this study was to determine if changes in VEMP responses during a postural change (inversion, causing increased ICP) would provide further information regarding the functional status of a radiographically apparent dehiscence.

2. Methods

2.1. Subjects

Sixteen subjects diagnosed with SCDS (mean 43 years, range 20–57 years, 9 females) and 15 age-matched control subjects with no history of balance or dizziness complaints (mean 41 years, range: 22–57 years, 11 females) participated in the study. HRCT was the gold standard for diagnosis of SCDS, combined secondarily with other auditory and vestibular signs and symptoms (audiometry, semi-recumbent VEMP, and bedside assessment consisting of head impulse test, tullio test, Hennebert sign, and 512 Hz tuning fork localization). All subjects diagnosed with SCDS had HRCT demonstrating apparent dehiscence as well as at least one physiologic sign (either auditory or vestibular) of a third mobile window in the affected labyrinth. Six subjects were diagnosed with unilateral SCDS and 10 with bilateral SCDS. Of the 10 subjects diagnosed with bilateral SCDS, 3 subjects had already undergone unilateral canal plugging and were being seen for signs and symptoms referable to the contralateral ear; 6 subjects had signs and symptoms primarily referable to 1 ear; and 1 subject had signs and symptoms referable to both ears, which were included as independent SCDS ears. Thus, a total of 17 ears with SCDS were used for analysis. Additionally, only the left ears ($n = 15$) of normal subjects were used for analysis.

2.2. Vestibular evoked myogenic potential (VEMP) testing

VEMP stimuli and recording techniques have been previously described (Nguyen et al., 2010). In short, a commercial electromyographic (EMG) system (Medelec Synergy, Care Fusion, software version 14.1, Dublin, OH) was used for VEMP testing. ACS stimuli were delivered monaurally via intra-auricular speakers from VIASYS Healthcare (Madison, WI) with foam eartips (Aearo Company Auditory Systems, Indianapolis, IN). Two types of ACS stimuli were delivered: (1) 0.1-ms, 105 dB nHL (140 dB peak SPL) clicks of positive polarity at a repetition rate of 5 per second; and (2) 500 Hz, 125 dB SPL tone bursts of positive polarity, with a linear envelope (1 ms rise/fall time, 2 ms plateau), at a repetition rate of 5 per second. One hundred sweeps were averaged for each ACS test. Two types of midline vibration stimulation (i.e., midline taps) were delivered at Fz (in the midline at the hairline, 30% of the distance between the inion and nasion): (1) manual taps delivered with the VIASYS system's reflex hammer fitted with an inertial microswitch trigger; and (2) "mini taps," as described by Iwasaki et al. (2007) were delivered with a *Brüel* and *Kjær* Mini-Shaker Type 4810 (1-ms clicks of positive polarity, with a repetition rate of 5