International Tinnitus Journal, Vol. 13, No. 1, 41–44 (2007)

Applications of the Marchbanks Transcranial–Cerebral Sonography Technique in Neurootology: Preliminary Report

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Abstract: Transcranial-cerebral sonography (TCCS) is a noninvasive technique that allows clinicians to detect nanoliter (billionths of a liter) displacements of the tympanic membrane. This technique was developed to assess cerebrospinal fluid (CSF) pressure in cases of shunted hydrocephalus; it takes advantage of the CSF connection to the inner ear through the cochlear aqueduct. The movements of the tympanic membrane that are observed in TCCS are those evoked by the acoustic stapedius reflex and those spontaneous movements generated by intracranial arterial, venous, and respiratory pulses transmitted through the inner ear to the stapes and thence to the tympanic membrane. Analysis of the amplitude and direction of these displacements has enabled neurosurgeons and neurologists to estimate CSF pressures accurately in patients evaluated by TCCS. TCCS allows for applications in neurootology, particularly in those patients who present with symptoms of pulsating tinnitus, dizziness and imbalance, or hearing loss. This preliminary report describes the test and its application in a series of patients whose diagnoses included pulsating tinnitus, idiopathic intracranial hypertension, Ménière's disease, perilymphatic fistula, perilymphatic hypertension, arterial stenosis, and Arnold-Chiari syndrome. We conclude that TCCS is a valuable addition to the armamentarium of neurootologists.

Key Words: cerebrospinal fluid pressure; dizziness; hearing loss; imbalance; pulsating tinnitus; transcranial-cerebral sonography

Marchbanks et al. [1–3] developed the tympanic membrane displacement (TMD) test to determine cerebrospinal fluid (CSF) pressures noninvasively in a population of hydrocephalics. These authors accomplished the measurement of CSF pressure by the detection of nanoliter displacements of the tympanic membrane resulting from eliciting the acoustic stapedial reflex (ASR). They measured the direction and amplitude of the displacement.

The position of the tympanic membrane and its displacement during the contraction of the stapedius muscle are determined by the position of the stapes in the oval window. That position is determined by the pressure of perilymph in the vestibule. An increase or decrease in CSF pressure will be mirrored in the perilymph pressure when the cochlear aqueduct is patent. The cochlear aqueduct is usually patent in children, but the percentage of patent cochlear aqueducts decreases with age.

An increase in the perilymphatic pressure will cause the resting position of the stapes footplate to occur in a more outward (lateral) position than is normal so that the ASR results in inward movement of the tympanic membrane. Conversely, the tympanic membrane moves outward when the stapes footplate rotates into the oval window in conditions in which perilymph pressure is decreased [4].

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Presented at the Twenty-Fourth International Tinnitus Forum, Toronto, Canada, September 16, 2006.

The Marchbanks TMD test has been successfully used for many years to detect CSF pressure increases in cases of hydrocephalic children with ventricular shunts. Marchbanks next turned his attention to the inner ear to determine whether his technique could detect inner-ear disorders related to inner-ear pressure changes [5]. Such conditions as pulsating tinnitus secondary to increased CSF pressures have been known to exist, as described by Sismanis [6]. Although the pulse and respiratory pressure test has always been part of the TMD protocol, the significance of these measurements was not realized until Marchbanks et al. [7] hypothesized in 1987 that intracranial pressure waves can be measured intra-aurally. These authors' hypothesis was proved by Doumoto et al. [8] in 1994, who concluded that intra-aural pressure waves correlate with intracranial pressure in humans.

The TMD test has since been renamed *transcranial-cerebral sonography* (TCCS). The pulse and respiratory pressures are measured by analysis of "spontaneous" TMD, as opposed to the "evoked" TMD test, which measures displacements secondary to the ASR. The determination of CSF pressures in both cases is based on clinical observations and correlations established by direct measurement of CSF pressures at the time of the TMD measurements [1].

Our group embarked on a study of TCCS in patients with various inner-ear disorders. We used equipment and techniques as reported by Marchbanks [1–5,7].

Our research group of 72 patients included patients with certain inner-ear disorders and various central disorders that could affect the inner ear. These disorders included perilymphatic fistula (PLF), perilymphatic hypertension and Ménière's disease. The central disorders included Arnold-Chiari malformation, idiopathic intracranial hypertension (IIH; previously called *pseudotumor cerebri* or *benign intracranial hypertension*), and cerebral arterial stenosis. We also tested patients having nonpulsating tinnitus, headache, a variety of vestibular problems, and hearing loss. Our preliminary report describes our experiences with the specific conditions described. They are presented as case reports with an overview.

IDIOPATHIC INTRACRANIAL HYPERTENSION AND PULSATING TINNITUS

TCCS has been valuable in treating IIH and pulsating tinnitus. The positive finding has been an increase in pulse pressure. Magnetic resonance imaging, magnetic resonance arteriography, and magnetic resonance venography are used to evaluate affected patients for abnormalities of the brain or its vasculature. If these are ruled out, a diagnosis of IIH can be established.

Patient 1

A 45-year-old, overweight woman with a sensorineural hearing loss and a tilt toward the low frequencies presented with pulsating tinnitus. TCCS revealed increased pressure on pulse-testing bilaterally, with normal ASR responses. We placed the patient on topiramate (Topamax), which has an effect similar to that of acetazolamide (Diamox) in lowering CSF pressure. Therapy for 6 weeks resulted in some weight loss (a side effect of the drug), a decrease in the tinnitus symptoms, and a decrease in pulse pressures. Ten weeks of therapy resulted in a 7-kg weight loss, further decrease in the pulsating tinnitus, and bilateral decrease in the hearing loss in the low- and middle-frequency ranges and in the discrimination scores.

Patient 2

A 40-year-old, overweight woman presented with pulsating tinnitus and normal hearing. TCCS revealed normal ASR responses with increased pulse pressure in the left ear. The patient declined treatment.

Patient 3

A 34-year-old, normal-weight woman developed vestibular symptoms associated with a finding of imbalance 6 months after successful repair of a left PLF. Lasix test results were positive for a PLF, in that they revealed secondary endolymphatic hydrops. TCCS revealed increased pulse pressures, more so on the right side. ASR responses were normal on the repaired left side, with a decrease in pressure noted on supine testing on the right side, a finding consistent with a right-sided PLF. We instituted Topamax therapy to decrease the CSF and inner-ear pressures. After 10 weeks of Topamax therapy, the imbalance had lessened. The pulse pressure on the right had returned to normal, but the pressure on the left was still increased. The ASR response was normal on the left, with a finding of PLF on the right. The patient experienced anorexia with a 5% loss of body weight. She experienced a need to increase the dose of Topamax to treat continued vestibular symptoms.

ARTERIAL STENOSIS

A 62-year-old woman presented with an 8-month history of bilateral pulsating tinnitus. TCCS revealed bilaterally increased pulse pressures with a normal ASR response. Magnetic resonance arteriography revealed an 80% stenosis of the proximal left middle cerebral artery. We are now treating the patient with acetylsalicylic acid.

PERILYMPHATIC HYPERTENSION

Perilymphatic hypertension, as an inner-ear phenomenon, has been diagnosed by improvement or resolution of abnormal inner-ear findings during a 2-hour period after intravenous administration of 500 mg of Diamox. Primary perilymphatic hypertension should show an increased pulse pressure with a closed cochlear aqueduct on ASR response. However, in our cases, we have found the cochlear aqueduct to be open, thus indicating that perilymphatic hypertension is secondary to increased CSF pressure and is thus the expression of inner-ear symptoms and findings in IIH.

A 48-year-old woman with posttraumatic dizziness and imbalance showed improvement in findings of imbalance on the Diamox test. We found her pulse pressures to be increased and found the cochlear aqueduct to be open, with a normal increase of pressure noted in the supine position on the ASR response test. On Topamax therapy, the patient showed improvement and, finally, resolution of her symptoms of dizziness and imbalance. Topamax, like Diamox, is a carbonic anhydrase inhibitor and, like Diamox, decreases CSF pressure.

PERILYMPHATIC FISTULA

We expected PLF to be demonstrated on the ASR response portion of TCCS by an absence of increase, or even a decrease, of pressure when we test a patient in the supine position. The normal increase in CSF and perilymph pressures induced in the supine position would be dissipated by an increase in the leakage of perilymph when the inner-ear pressure is increased. This has been noted in surgical cases in which the leakage is encouraged by pressure on the ipsilateral internal jugular vein or by positioning a patient in the head-down position. We have seen this finding on ASR response testing in nine patients with proven PLFs.

Another pattern could be a large outward displacement of the tympanic membrane during ASR response testing. This would indicate that the stapes is displaced into the vestibule secondary to decreased inner-ear pressure. We have seen this in one patient.

We have also seen three patients with normal ASR responses but with increased pulse pressures. This could indicate the absence of a free flow of perilymph at the time of the testing, so that the increase in perilymph pressure was not dissipated in the supine position.

MÉNIÈRE'S DISEASE

Great interest is being shown in the potential utility of TCCS in treating Ménière's disease. The first patient whom we tested had a worsening of his condition, apparently in response to the effect of the intensity of the ASR stimulus on his inner ear. We have proceeded with caution after that experience. We did test one patient who had increased pulse pressure in the affected ear. He volunteered to undergo ASR testing and was found to have normal responses. Rosingh et al. [9] reported finding no changes in perilymph pressures in Ménière's disease patients who were evaluated by the ASR response test.

ARNOLD-CHIARI MALFORMATION TYPE I

A 34-year-old woman with Arnold-Chiari malformation type I presented with a long history of imbalance and dizziness. She reported dizziness provoked by visual inputs. TCCS revealed bilaterally increased pulse pressures with normal ASR responses. We are considering neurosurgical treatment.

CONCLUSIONS

TCCS is a valuable addition to the armamentarium of neurootologists. We have found it quite useful in identifying IIH in cases of pulsating tinnitus. IIH can result in a variety of cochlear and vestibular symptom complexes, as Weider et al. [10] have found and as we described earlier. TCCS can provide evidence of increased CSF pressure, which can be crucial in the successful management of inner-ear disorders, because some of these disorders and symptom complexes may be secondary to an increased CSF pressure. TCCS can also assist clinicians in tracking inner-ear symptoms and findings back to a central source, such as in Arnold-Chiari malformation cases.

ACKNOWLEDGMENT

The authors express their appreciation to Dr. Robert Marchbanks for his invaluable assistance and invariable good humor, both of which were essential to this study.

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