

# The Value of Transcranial Cerebral Sonography in Diagnosing Neurootological Disorders

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**Abstract:** Transcranial cerebral sonography (TCCS) is a noninvasive technique that allows the clinician to detect abnormal intracranial–inner-ear fluid interactions in terms of nanoliter tympanic membrane displacements. The displacements recorded in TCCS are evoked either by the acoustic stapedius reflex or spontaneous movements generated by intracranial cardiovascular or by respiratory pressure waves 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 cerebrospinal fluid pressures in patients evaluated by TCCS. This procedure allows for applications in neurootology, particularly in those patients who present with symptoms of pulsating tinnitus, dizziness and imbalance, or hearing loss. This study describes the application of TCCS tests in a series of patients whose diagnoses included perilymphatic fistula and a variety of neurological conditions such as idiopathic intracranial hypertension, type I Arnold-Chiari malformation, sigmoid sinus thrombosis, hydrocephalus, and cerebrovascular malformations. We conclude that both raised intracranial pressure and abnormal intracranial pressure waves are associated with common neurootological symptoms, including tinnitus, dizziness, and hearing dysfunction. Furthermore, TCCS is a valuable addition to neurootologists' test batteries.

**Key Words:** cerebrospinal fluid pressures; inner ear pressures

Transcranial cerebral sonography (TCCS), also termed the *tympanic membrane displacement technique*, has been described and reported by Marchbanks [1]. The test measures the direction and amount of the displacement of the tympanic membrane caused by the acoustic stapedial reflex [2]. It is used in the evaluation of cerebrospinal fluid (CSF) pressures. The analysis of intra-aural pressure waves was found to be of value and was added to the test protocol [3].

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Analysis of inner-ear pressure waves caused by transmission of intracranial pulsations from arterial, venous, or respiratory sources revealed that these pressure waves were a reflection of intracranial pressures (ICPs). This function of the TCCS has been called the *spontaneous test*. The capture of responses generated by the use of the acoustic reflex is called the *evoked* or *reflex test*.

The test relies upon the connection of the CSF to the perilymph of the inner ear through the cochlear aqueduct. Neurootologists, particularly Dudley Weider, had noted increased CSF pressures in patients with perilymphatic fistulae (PLF). Weider became interested in the application of TCCS to inner-ear disorders that were caused by distortions of the inner-ear fluid system that in turn were caused by abnormalities in the CSF pressure [4]. TCCS has been used in treating patients presenting to neurootologists with symptoms of inner-ear disorders. The presenting symptoms included tinnitus, particularly of the pulsating type, vestibular complaints, and hearing loss.

The following cases are described to represent some of our experiences with TCCS and to support our conclusion that TCCS can provide valuable diagnostic information for neurootologists. We hope that other centers will use this test to gain more experience with TCCS. We also hope that, as more experience is gained, advances in technique and interpretation will result.

### **INTRACRANIAL HYPERTENSION FROM SIGMOID SINUS THROMBOSIS**

A 50-year-old woman presented with severe headache of 1-month's duration accompanied by bilateral pulsating tinnitus and blurry vision in the right eye. Eye examination revealed right papilledema. We found the woman to have a partially blocked right sigmoid sinus on magnetic resonance imaging (MRI). A computed tomography scan of the mastoids revealed only a few opacified mastoid cells, with no evidence of bony breakdown. TCCS revealed increased pressure waves on the spontaneous test.

We treated the patient with anticoagulation and 500 mg of acetazolamide (Diamox) b.i.d. for 7 weeks. The papilledema improved, and the pulsating tinnitus resolved. TCCS showed a decrease in the elevated pressures.

This patient could be classified as having had increased CSF pressure secondary to obstruction in a venous sinus. The pressure increase was transmitted to the eye, causing blurry vision, after which we detected the papilledema. The increased pressure was also transmitted to the inner ears, which caused the pulsating tinnitus. TCCS detected the increased inner-ear pressure.

### **IDIOPATHIC INTRACRANIAL HYPERTENSION PRESENTING WITH PULSATILE TINNITUS AND SENSORINEURAL HEARING LOSS**

A 45-year-old obese woman presented with bilateral pulsating tinnitus and a bilateral low-tone sensorineural hearing loss. TCCS revealed normal reflex responses and increased pressures on the pulse wave (spontaneous) test. We made a diagnosis of idiopathic intracranial hypertension on the basis of TCCS, with symptoms and findings in the inner ears being caused by transmission of the increased CSF pressure into the inner ear. We treated the patient with Topamax (topiramate), which, like Diamox, is a carbonic anhydrase inhibitor that decreases CSF pressure. Topamax therapy is also associated with decreased appetite and weight loss. Ten weeks of Topamax therapy resulted in a 7-kg weight loss, a significant decrease in the pulsating tinnitus, and improved hearing in the low frequencies and an improved discrim-

ination score on audiometric evaluation. The patient experienced resolution of her vestibular symptoms, obtained improvement in her hearing, and lost 7 kg of weight. Our conclusion was that she suffered from idiopathic intracranial hypertension affecting the inner ear.

### **ARNOLD-CHIARI TYPE I PRESENTING WITH IMBALANCE**

A 48-year-old woman was referred by her neuroophthalmologist with a known type I Arnold-Chiari malformation (ACM) and symptoms of chronic dizziness. We found the patient to have imbalance. She reported being dizzy in stores and while reading, symptoms that we interpreted as being consistent with a vestibular-visual system disorder. A previous Epley maneuver was unhelpful. Suprathreshold stapedial reflex testing revealed evidence of impaired transmission in the brainstem. We treated the woman with Periactin (cyproheptadine hydrochloride) for its antiserotonin effects, with no relief. TCCS revealed increased pressures on the spontaneous test with normal reflex responses. We interpreted this as evidence of increased CSF pressure secondary to an obstruction caused by the ACM. MRI revealed compression of the brainstem. We referred the patient to a clinic specializing in management of ACM.

### **PERILYMPHATIC FISTULAE**

#### **PLF Patient 1**

A 59-year-old woman presented with left-sided headache and dizziness and imbalance that had lasted for 1 year. She had been treated with Topamax for the headache, with some improvement, but her balance became worse on the Topamax. On examination, we noted imbalance and bilaterally positive fistula test results (worse on the left). TCCS revealed increased pressures on both sides on the spontaneous test. The reflex test result was normal on the right. We noted a decreased pressure in the supine position (rather than the expected increase) on left reflex testing and explored the left ear, finding PLF of the oval and round windows. We repaired the fistulae in August 2008, and the patient has had a successful outcome.

#### **PLF Patient 2**

A 55-year-old overweight woman presented in October 2006 with a 1-month history of dizziness and imbalance. She had undergone a successful repair of a left-sided PLF in 1979. She had recently suffered a fractured left elbow in a fall in which she had also struck her head. Our examination revealed imbalance and bilateral

positional nystagmus. We could not perform an Epley maneuver because of the elbow fracture (the arm was in a sling). The patient became very dizzy that evening, and we saw her the next day with findings of a left vestibular neuritis. She was placed on prednisone and improved. We performed an Epley maneuver for right-sided benign positional vertigo (BPV) 1 month later. Three months later, she suffered a recurrence of the right BPV, and we performed another Epley maneuver. The prednisone treatment improved her balance, but it did not return to normal and ultimately became worse, with a finding of falling on the Romberg test in the fourth month of her illness. The BPV had resolved at that time. We performed TCCS in December 2007, the spontaneous test showing normal pressures. The normal increase in pressure in the supine position on the reflex test was absent bilaterally. Results from a Lasix (furosemide) test for PLF were positive in January 2008. We placed the patient on oral Lasix, but she still experienced dizziness and complained of dehydration, so we explored the left ear in March 2008 and repaired a recurrent PLF. The patient has done well since that surgery, her last visit occurring in December 2008.

### PLF Patient 3

A 32-year-old woman suffered head trauma in an auto accident in January 2006. We first saw her in March 2006 presenting with symptoms of dizziness and imbalance. Our examination revealed imbalance and positive results from bilateral fistula testing in both ears. Results from a glycerin test that we performed in May 2006 were positive for PLF. TCCS performed in October 2007 revealed normal responses on the spontaneous test. Right reflex test results were normal. The left reflex test showed a decreased pressure on testing in the supine position. We performed left ear surgery in October 2008, during which we found and repaired PLFs. The patient experienced improved balance after the surgery.

### CEREBROVASCULAR LOOP WITH TINNITUS AND HEARING LOSS

A 58-year-old man presented in Southampton with a blockage sensation in his left ear and an “echoing” that became worse with exercise. He had a left-sided high-frequency hearing loss with tinnitus that he described as “electrical-high pitched” and “popping.” The symptoms were of sudden onset. Although MRI showed a left vascular loop, we did not believe this to be consistent with the otological symptoms. We arranged to investigate an independent condition associated with a hydroptic change or patulous eustachian tube.

TCCS tests showed significant CSF disturbance and larger amplitude cardiovascular pulsations, particularly on the left vascular loop side. These findings were consistent with the left-sided vascular loop as the underlying cause. The sudden onset of the condition could be due either to a “sudden occurrence” of close coupling between intracranial and labyrinth fluids or to the vascular loop’s becoming an active generator of ICP waves.

### ASSUMED ARRESTED HYDROCEPHALUS WITH DIZZINESS

An 11-year-old boy presented in Southampton with incapacitating dizziness episodes that included rotary objective vertigo and “subjective” vertical motion (i.e., dizziness of both peripheral and central origin). He reported “popping and crackling” sounds in his head but no significant headache. The child had been born with hydrocephalus of unknown origin, and this was assumed to be “arrested” (i.e., no longer clinically significant).

TCCS tests showed extremely large spontaneous activity that was more than 10-fold larger than the normal mean value and was synchronous with respiration. We have seen similar pressure waves with aqueductal stenosis or type 1 ACM. MRI revealed enlarged lateral ventricles and periventricular changes consistent with an old hemorrhage.

### AN OBSCURE NEUROLOGICAL DISORDER

A 59-year-old woman presented in Southampton with constant imbalance and “white noise–tinkling” tinnitus that was most pronounced in her left ear. She needed to use a walking stick for support and experienced a buzzing and pulsing sensation mid-spine that “echoed” in her head. Her problems seemed to coincide with an epidural-spinal anesthesia for hysterectomy 9 years earlier.

TCCS tests showed significant CSF disturbance. The pulse amplitude was large in both ears, particularly in the left ear. Also noticeable was a left-sided cardiovascular pulse that appeared to be reverberating. One possible explanation was a restricted CSF flow from the intracranial fluid space to the lumbar sac and—speculatively—restrictions within the spinal CSF pathways that were causing reflected pressure waves. With this information, radiography confirmed multiple spinal canal narrowings due to diagnosed hemangiomas.

### DISCUSSION

An understanding of how the intracranial (cerebral) fluid interacts with fluid within the inner ear is important in

diagnosing neurootological disorders. Even small elevations in ICP may have adverse effects on the ear and cause tinnitus, imbalance, vertigo, and hearing loss. Sustained elevation of ICP can cause permanent inner-ear damage, such as recurrent PLF, as shown by Weider et al. [4,5].

Many clinical studies link abnormalities of the ICF pressure with inner-ear homeostasis disorders. In their study of 364 symptomatic patients with type 1 ACM, Milhorat et al. [6] provide an intriguing insight into this association. Of the 364 patients tested, a total of 269 (74%) experienced neurootological symptoms that included peripheral vestibular dysfunction, tinnitus, and a low-frequency hearing loss.

Our case studies demonstrated a clear association between these symptoms and neurootological disorders that cause increased intra-aural pressure waves as recorded by the TCCS spontaneous test. Therefore, the assertion is that both raised ICP and abnormal ICP waves are clinically important.

The neurootological symptoms of certain neurological conditions can also form the basis of Ménière's-like disorders [7]. Correct differential diagnosis based on symptoms alone is extremely challenging and is complicated further if the patient does not present with the expected headache intensity or papilledema [8,9]. Without papilledema or a significant headache, these cases are likely to present to the otology clinic. An objective measurement such as TCCS is, therefore, an important addition to the clinical test battery.

## CONCLUSIONS: CLINICAL IMPLICATIONS

The TCCS technique can be used to quantify intracranial inner-ear pressure wave interactions. We have demonstrated that ICP waves are entering the inner ear and that these are abnormally large when associated with certain neurological and neurootological disorders. Any neurological condition that changes the homeostasis of the intracranial fluid is likely to change the homeostasis of the ear. Auditory dysfunction, vertigo, tinnitus that is often

pulsatile in nature, and imbalance may occur as a result of ICP waves interacting directly with the hearing and balance organs.

We conclude that TCCS can provide valuable information with regard to the homeostasis of the inner ear and thus assist clinicians in the diagnosis and management of disorders affecting this system.

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