J Am Acad Audiol 29:187–191 (2018)

Case Report

Cervical Vestibular Evoked Myogenic Potential in Hypoglossal Nerve Schwannoma: A Case Report

DOI: 10.3766/jaaa.16012

Aravind Kumar Rajasekaran*
Amey Rajan Savardekar†
Nagaraja Rao Shivashankar*

Abstract

Background: Schwannoma of the hypoglossal nerve is rare. This case report documents an atypical abnormality of the cervical vestibular evoked myogenic potential (cVEMP) in a patient with schwannoma of the hypoglossal nerve. The observed abnormality was attributed to the proximity of the hypoglossal nerve to the spinal accessory nerve in the medullary cistern and base of the skull.

Purpose: To report cVEMP abnormality in a patient with hypoglossal nerve schwannoma and provide an anatomical correlation for this abnormality.

Research Design: Case report.

Study Sample: A 44-yr-old woman.

Data Collection: Pure-tone and speech audiometry, tympanometry, acoustic stapedial reflex, auditory brainstem response, and cVEMP testing were performed.

Results: The audiological test results were normal except for the absence of cVEMP on the lesion side (right).

Conclusions: A cVEMP abnormality indicating a compromised spinal accessory nerve was observed in a patient with hypoglossal nerve schwannoma. This case report highlights the importance of recording cVEMP in relevant neurological conditions and provides clinical proof for the involvement of the spinal accessory nerve in the vestibulocollic reflex pathway.

Key Words: cVEMP, hypoglossal schwannoma, spinal accessory nerve, vestibular evoked myogenic potential, vestibulocollic reflex

Abbreviations: ABR = auditory brainstem responses; CNXI = cranial nerve XI; CNXII = cranial nerve XII; cVEMP = cervical vestibular evoked myogenic potential; HC = hypoglossal canal; JF = jugular fossa; SCM = sternocleidomastoid muscle; VCR = vestibulocollic reflex

INTRODUCTION

The hypoglossal nerve is cranial nerve XII (CNXII), which innervates all muscles of the tongue except the palatoglossus (Standring and Berkovitz, 2005). CNXII controls tongue movements during swallowing and speech. Schwannoma of CNXII is rare, constituting only 5% of all nonvestibular intracranial schwannomas (Boban et al, 2007; Osborn, 2013). CNXII schwannomas can compromise the spinal accessory and other lower cranial nerves (Sato et al, 1996; Hoshi et al, 2000; Awobuluyi, 2013).

The cervical vestibular evoked myogenic potential (cVEMP) is an inhibitory potential recorded from the sternocleidomastoid muscle (SCM) (Colebatch, 2001). It

*Department of Speech Pathology and Audiology, National Institute of Mental Health and Neurosciences, Bangalore, India; †Department of Neurosurgery, National Institute of Mental Health and Neurosciences, Bangalore, India

Corresponding author: Aravind Kumar Rajasekaran, Department of Speech Pathology and Audiology, National Institute of Mental Health and Neurosciences, Bangalore, India; E-mail: raravindk@gmail.com

reflects the integrity of the vestibulocollic reflex (VCR) pathway. Insights into the VCR pathway have primarily been gained through animal research. The otolith organs, vestibular nerve, vestibulospinal tract, cervical segment of the spinal cord, nucleus of the spinal accessory nerve, and SCM constitute the VCR pathway (Shinoda et al, 1989; Sato et al, 1997; Uchino et al, 1997; Tsubota et al, 2007; Sheykholeslami et al, 2009; Curthoys et al, 2016). Various neurological disorders in humans present with cVEMP abnormalities (Colebatch and Halmagyi, 1992; Colebatch et al, 1994; Murofushi et al, 2001; Sartucci and Logi, 2002; Deftereos et al, 2008; Felipe et al, 2009; Pollak et al, 2009; Kim et al, 2010; Güven et al, 2014; Magnano et al, 2014; Ribeiro et al, 2015). These patients had lesions at various levels of the VCR pathway; however, none had a cranial nerve XI (CNXI) lesion. Thus, CNXI involvement in the VCR pathway lacks clinical evidence in humans.

The present case study reports cVEMP abnormality in a patient with CNXI schwannoma. Furthermore, anatomical correlation provides clinical evidence for CNXI involvement in the VCR pathway.

CASE REPORT

A 44-yr-old woman presented with articulation difficulty and deviation of the tongue to the right side (on protrusion) lasting 3 mo. No other significant otological history such as hearing loss or vestibular symptoms was present. The patient underwent a neurological examination. The tongue showed fasciculations and left side atrophy. These findings indicated right lower motor neuron hypoglossal palsy. The rest of the cranial nerve examination was normal. Magnetic resonance imaging revealed a $2.6 \times 2.14 \times 2.2$ cm^3 lesion (schwannoma) arising from the right CNXII at the entrance of the right hypoglossal canal (HC). The lesion occupied the right jugular fossa (JF) and had a mild protrusion into the premedullary cisternal space (Figure 1).

The patient underwent audiological evaluation as part of routine presurgical documentation, and the following tests were performed: pure-tone and speech audiometry (speech recognition threshold and word recognition score), tympanometry and acoustic stapedial reflex (ipsilateral and contralateral), distortion product otoacoustic emissions, and auditory brainstem response (ABR). Words from standardized lists were presented in monitored live-voice mode to establish the speech recognition threshold (Padmaja, 1987) and word recognition score (Mayadevi, 1978). All audiological tests were carried out with calibrated equipment in sound-treated rooms.

cVEMP was recorded using IHS equipment (Intelligent Hearing Systems Inc., Miami, FL) to evaluate the integrity of the VCR pathway. Both the positive and negative electrodes were placed on the test side. The ground electrode was placed on the forehead.

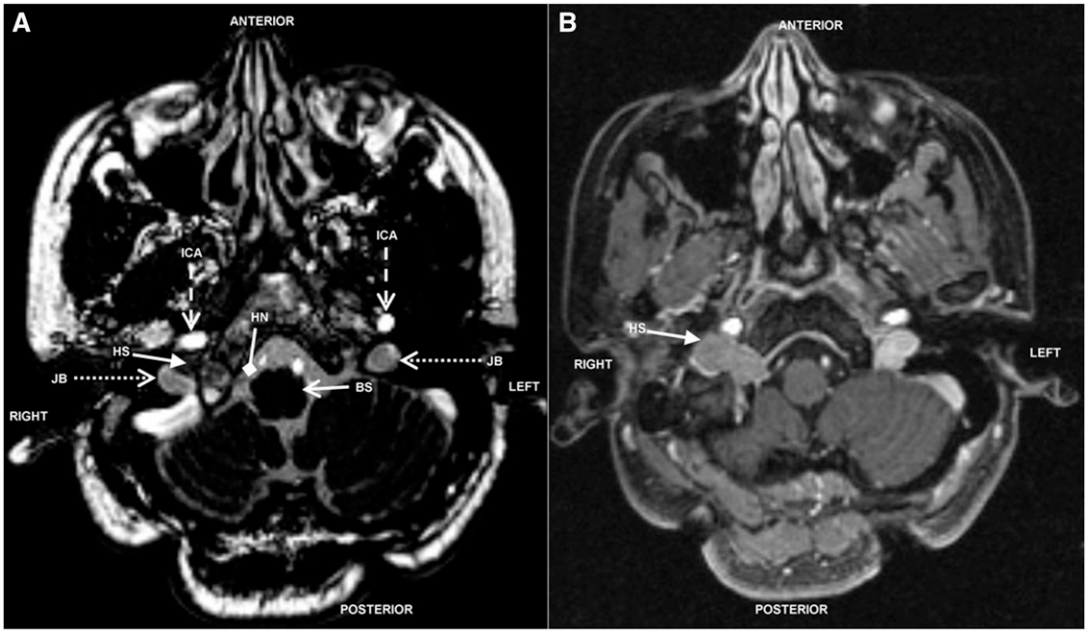

Figure 1. (A) T2-weighted axial magnetic resonance (MR) image of the patient showing the posterior fossa structures, with "open arrow" pointing at the brainstem (BS), and "diamond arrow" showing the hypoglossal nerve (HN) exiting the brainstem between the pyramid and the inferior olive, and exiting the skull base through the hypoglossal foramen. The "open arrows with dashed line" show the bilateral internal carotid arteries (ICA) in the petrous bone. The "open arrows on dotted line" show the bilateral jugular bulbs (JB). The "closed arrow" points at the hypoglossal schwannoma (HS) arising from the hypoglossal nerve (HN) and occupying the JF between the right ICA anteriorly and right JB posteriorly. It is at this location that the tumor is compressing upon the rootlets of the spinal accessory nerve (CNXI). (B) Contrast-enhanced T1-weighted axial MR image of the patient showing the HS depicted by the "closed arrow."

Table 1. Stimulus and Recording Parameters for ABR and cVEMP

Parameters		ABR	cVEMP
Stimulus	Type	Click: 100 μsec	Tone burst: 500 Hz; 1-0-1 cycle
	Rate	11.1/sec	4.1/sec
	Polarity	Rarefaction	Rarefaction
	Presentation	Monoaural	Monoaural
Recording	Montage		
	Positive	Vertex	Upper half of ipsilateral SCM
	Negative	Ipsilateral ear lobe	Inspectional base of ipsilateral SCM at sternoclavicular junction
	Ground	Contralateral ear lobe	Forehead
	Filter	100–3000 Hz	10–1500 Hz
	Amplification	100,000	5,000
	Rejection voltage		
	EEG	30 μV	Open window (100 μV)
	EMG	Not applicable	<50 and >500 μV
Averaging	Poststimulus window	12 msec	50 msec
	Prestimulus window	13 msec	10 msec
	Number of sweeps	1,024	100

Note: EEG = electroencephalogram; EMG = electromyogram.

The stimulus and recording parameters are given in Table 1. The stimulus was presented through an insert earphone (ER-3A; Etymotic Research, Elk Grove Village, IL). The patient was seated in a comfortable reclining chair facing the equipment monitor and turned her head to the right or left side as instructed. Biofeedback was given to achieve appropriate and steady SCM tension. The appearance of a green bar and a smiling face on the monitor as well as a green light on the feedback indicator box confirmed appropriate muscle tension. A red bar and a frowning face on the monitor along with a red light on the indicator box implied inappropriate muscle tension. Electromyographic waveforms with amplitudes <50 μV or >500 μV were automatically discarded, and excessive muscle tension was monitored during testing.

RESULTS

The audiological tests revealed normal findings bilaterally except for cVEMP. Pure-tone averages for the right and the left ears were 15 and 13.3 dB HL, respectively (Figure 2). Speech recognition thresholds were 10 dB HL, and the word recognition score was 100% for both ears. Distortion product otoacoustic emissions were present bilaterally at all frequencies from 1 to 8 KHz.

Tympanometry revealed no abnormalities in either ear (Table 2). Acoustic stapedial reflexes were present at 500 Hz, 1 KHz, and 2 KHz bilaterally on ipsilateral and contralateral stimulation (Table 2). The ABR recording demonstrated good wave morphology and replicability, and the absolute and the interpeak ABR latencies were normal (Table 2). cVEMP was absent on the lesion side (right) and present on the nonlesion side (left). The cVEMP latency and amplitude values are given in Table 2 and shown in Figure 3.

DISCUSSION

The audiological tests (pure-tone audiometry, speech audiometry, tympanometry, acoustic reflex, and ABR) revealed normal bilateral findings in the patient. The only abnormality noted was the absence of cVEMP on the lesion side (right). Studies have reported cVEMP abnormalities in patients with unilateral brainstem tumors, vestibular schwannoma (Welgampola et al, 2013; Chiarovano et al, 2014), cerebellopontine angle schwannoma and meningioma (Hu et al, 2009; Su et al, 2013), and cerebellopontine angle epidermoid (Chu et al, 2006). To our knowledge, this clinical report is the first to describe a cVEMP abnormality in a patient with hypoglossal schwannoma. Based on their review, Curthoys (2010) posited that cVEMP recorded at SCM

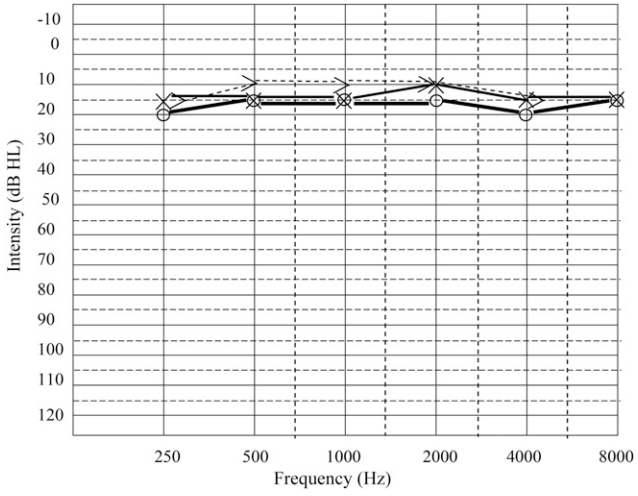

Figure 2. Pure-tone audiogram depicting the hearing thresholds. ○, right-ear air conduction; ×, left-ear air conduction; and >, left-ear bone conduction.

Table 2. Results of ABR and cVEMP

Test Measures		Right Ear		Left Ear	
Tympanometry	MEP (dapa)	−10		−7	
	ECV (mL)	1.16		1.26	
	SC (cc)	0.78		0.76	
Acoustic reflexes (dB SPL)	I and C	I	C	I	C
	500 Hz	90	95	90	100
	1000 Hz	90	90	90	95
	2000 Hz	90	95	90	90
ABR latencies (msec)	I	1.77		1.75	
	III	3.55		3.45	
	V	5.4		5.42	
	I–III	1.78		1.7	
	III–V	1.85		1.97	
	I–V	3.63		3.67	
cVEMP latencies (msec) and amplitude (μV)	P1 latency	Absent		15.07	
	N1 latency			21.45	
	P1–N1 amplitude			121.39	

Note: C = contralateral; ECV = ear canal volume; I = ipsilateral; MEP = middle ear pressure; N1 = negative peak; P1 = positive peak; SC = compensated static compliance.

is predominantly a VCR output. Thus, VCR pathway dysfunction may be implicated in our patient. A mass lesion of CNXII can compromise CNXI (Sato et al, 1996; Hoshi et al, 2000; Awobuluyi, 2013) because CNXII lies close to CNXI in the cisternal space and at the base of the skull. Within the cranium, CNXI travels in the cerebellomedullary cistern, and CNXII travels in the premedullary cistern. These cisterns lie adjacent to each other. The cerebellomedullary cistern (which lies just in front of CNIX, CNX, and CNXI) forms the lateral border of the premedullary cistern (Rhoton, 2000). In our patient, the lesion extended into the premedullary cistern and therefore could have compromised CNXI. After transiting the cisternal space, CNXI and CNXII leave the cranium at the base of the skull. CNXI exits through the JF, and CNXII exits through the HC (Osborn, 2013). The HC is just 0.75 cm inferomedial to the JF, and they are separated by the jugular tubercle (Roche et al, 2008; Karasu et al, 2009). Furthermore, CNXII lies medial to the JF (Gupta et al, 2014). In our patient, the tumor occupied the right JF and thus could have compromised CNXI.

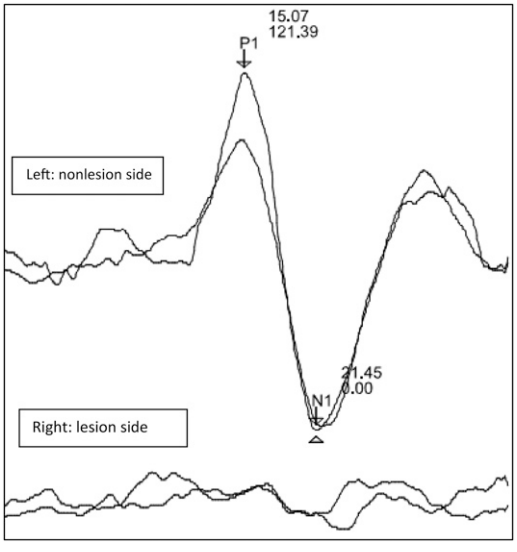

Figure 3. cVEMP waveform of right and left sides with its latencies and amplitudes. P1, positive peak; N1, negative peak; P1 latency, 15.07 msec; N1 latency, 21.45 msec; P1N1 amplitude, 121.39 μV.

The absence of cVEMP could be related to VCR pathway dysfunction owing to CNXI compromise. In animals, cVEMP involves motor neurons of the neck (equivalent to CNXI) (Shinoda et al, 1989; Uchino et al, 1997, Sheykholeslami et al, 2009). Similar findings demonstrating CNXI involvement in cVEMPs are unavailable in humans. CNXI is the primary motor supplier to the SCM (Standring and Berkovitz, 2005), which further strengthens our implication of CNXI compromise. Therefore, this case report provides clinical evidence of CNXI involvement.

In conclusion, this case report highlights subclinical VCR pathway compromise in a patient with a CNXII schwannoma. Furthermore, it provides clinical evidence of CNXI involvement in the VCR pathway. This report may also expand the scope of cVEMP test procedures.

REFERENCES

Awobuluyi MT. (2013) Tumors of cranial/spinal nerves. In: Naidich TP, Castillo M, Cha S, Smirniotopoulos JG, eds. *Imaging of the Brain*. Philadelphia, PA: Saunders, 826–836.

Boban M, Brinar VV, Habek M, Rados M. (2007) Isolated hypoglossal nerve palsy: a diagnostic challenge. *Eur Neurol* 58(3):177–181.

Chiarovano E, Darlington C, Vidal PP, Lamas G, de Waele C. (2014) The role of cervical and ocular vestibular evoked myogenic potentials in the assessment of patients with vestibular schwannomas. *PLoS One* 9(8):e105026.

Chu CK, Tseng HM, Young YH. (2006) Clinical presentation of posterior fossa epidermoid cysts. *Eur Arch Otorhinolaryngol* 263(6):548–551.

Colebatch JG, Halmagyi GM. (1992) Vestibular evoked potentials in human neck muscles before and after unilateral vestibular deafferentation. *Neurology* 42(8):1635–1636.

Colebatch JG, Halmagyi GM, Skuse NF. (1994) Myogenic potentials generated by a click-evoked vestibulocollic reflex. *J Neurol Neurosurg Psychiatry* 57(2):190–197.

Colebatch JG. (2001) Vestibular evoked potentials. *Curr Opin Neurol* 14(1):21–26.

Curthoys IS. (2010) A critical review of the neurophysiological evidence underlying clinical vestibular testing using sound, vibration and galvanic stimuli. *Clin Neurophysiol* 121(2):132–144.

Curthoys IS, Vulovic V, Burgess AM, Sokolic L, Goonetilleke SC. (2016) The response of guinea pig primary utricular and saccular irregular neurons to bone-conducted vibration (BCV) and air-conducted sound (ACS). *Hear Res* 331:131–143.

Deftereos SN, Panagopoulos G, Eleftheriadou A, Korres S, Georgonikou D, Kandiloros D, Karageorgiou CE. (2008) Using vestibular evoked myogenic potentials to localise brainstem lesions. A preliminary report. *B-ENT* 4(4):215–219.

Felipe L, Gonçalves DU, Tavares MC, Sousa-Pereira SR, Antunes CM, Lambertucci JR. (2009) Vestibular-evoked myogenic potential (VEMP) in the evaluation of schistosomal myeloradiculopathy. *Am J Trop Med Hyg* 81(4):551–554.

Gupta C, Kurian P, Seva KN, Kalthur SG, D'Souza AS. (2014) A morphological and morphometric study of jugular foramen in dry skulls with its clinical implications. *J Craniovertebr Junction Spine* 5(3):118–121.

Güven H, Bayır O, Aytaç E, Ozdek A, Comoğlu SS, Korkmaz H. (2014) Vestibular-evoked myogenic potentials, clinical evaluation, and imaging findings in multiple sclerosis. *Neurol Sci* 35(2):221–226.

Hoshi M, Yoshida K, Ogawa K, Kawase T. (2000) Hypoglossal neurinoma—two case reports. *Neurol Med Chir (Tokyo)* 40(9):489–493.

Hu YF, Cheng PW, Young YH. (2009) Comparison of vestibular function between large cerebellopontine angle meningioma and schwannoma. *Acta Otolaryngol* 129(2):161–165.

Karasu A, Cansever T, Batay F, Sabanci PA, Al-Mefty O. (2009) The microsurgical anatomy of the hypoglossal canal. *Surg Radiol Anat* 31(5):363–367.

Kim S, Lee HS, Kim JS. (2010) Medial vestibulospinal tract lesions impair sacculo-collic reflexes. *J Neurol* 257(5):825–832.

Magnano I, Pes GM, Pilurzi G, Cabboi MP, Ginatempo F, Giaconi E, Tolu E, Achene A, Salis A, Rothwell JC, Conti M, Deriu F. (2014) Exploring brainstem function in multiple sclerosis by combining brainstem reflexes, evoked potentials, clinical and MRI investigations. *Clin Neurophysiol* 125(11):2286–2296.

Mayadevi C. (1978) ISHA PB word list-4. In: Kacker SK, Basavaraj V, eds. *Indian Speech, Language and Hearing Tests: The ISHA Battery*. Mysore, India: Indian Speech and Hearing Association.

Murofushi T, Shimizu K, Takegoshi H, Cheng PW. (2001) Diagnostic value of prolonged latencies in the vestibular evoked myogenic potential. *Arch Otolaryngol Head Neck Surg* 127(9):1069–1072.

Osborn AG. (2013) Cranial nerves and nerve sheath tumors. In: Osborn AG, ed. *Osborn's Brain: Imaging, Pathology and Anatomy*. 1st ed. Manitoba, Canada: Amirsys Inc, 613–644.

Padmaja B. (1987) ISHA spondee word list-7. In: Kacker SK, Basavaraj V, eds. *Indian Speech, Language and Hearing Tests: The ISHA Battery*. Mysore, India: Indian Speech and Hearing Association.

Pollak L, Prohorov T, Kushnir M, Rabey M. (2009) Vestibulocervical reflexes in idiopathic Parkinson disease. *Neurophysiol Clin* 39(4–5):235–240.

Rhoton AL, Jr. (2000) The posterior fossa cisterns. *Neurosurgery* 47(Suppl 3):S287–S297.

Ribeiro RS, Pereira MM, Pedroso JL, Braga-Neto P, Barsottini OGP, Manzano GM. (2015) Cervical and ocular vestibular evoked potentials in Machado-Joseph disease: functional involvement of otolith pathways. *J Neurol Sci* 358(1–2):294–298.

Roche PH, Mercier P, Sameshima T, Fournier HD. (2008) Surgical anatomy of the jugular foramen. *Adv Tech Stand Neurosurg* 33: 233–263.

Sartucci F, Logi F. (2002) Vestibular-evoked myogenic potentials: a method to assess vestibulo-spinal conduction in multiple sclerosis patients. *Brain Res Bull* 59(1):59–63.

Sato H, Imagawa M, Isu N, Uchino Y. (1997) Properties of saccular nerve-activated vestibulospinal neurons in cats. *Exp Brain Res* 116(3):381–388.

Sato M, Kanai N, Fukushima Y, Matsumoto S, Tatsumi C, Kitamura K, Ozaki M, Hayakawa T. (1996) Hypoglossal neurinoma extending intra- and extracranially: case report. *Surg Neurol* 45(2): 172–175.

Sheykholeslami K, Megerian CA, Zheng QY. (2009) Vestibular evoked myogenic potentials in normal mice and Phex mice with spontaneous endolymphatic hydrops. *Otol Neurotol* 30(4): 535–544.

Shinoda Y, Ohgaki T, Sugiuchi Y, Futami T. (1989) Comparison of the branching patterns of lateral and medial vestibulospinal tract axons in the cervical spinal cord. *Prog Brain Res* 80:137–147, discussion 127–128.

Standring S, Berkovitz KB. (2005) Neck. In: Standring S, ed. *Gray's Anatomy: The Anatomical Basis of Clinical Practice*. 39th ed. Spain: Elsevier Churchill Livingstone, 531–566.

Su CH, Chen CM, Young YH. (2013) Differentiating cerebellopontine angle meningioma from schwannoma using caloric testing and vestibular-evoked myogenic potentials. *J Neurol Sci* 335 (1–2):155–159.

Tsubota M, Shojaku H, Hori E, Fujisaka M, Hayashi N, Kurimoto M, Hatakeyama N, Yamazaki M, Nishijo H, Ono T, Yamamoto H, Watanabe Y. (2007) Effects of vestibular nerve section on sound-evoked myogenic potentials in the sternocleidomastoid muscle of monkeys. *Clin Neurophysiol* 118(7):1488–1493.

Uchino Y, Sato H, Sasaki M, Imagawa M, Ikegami H, Isu N, Graf W. (1997) Sacculocollic reflex arcs in cats. *J Neurophysiol* 77(6):3003–3012.

Welgampola MS, Ramsay E, Gleeson MJ, Day BL. (2013) Asymmetry of balance responses to monaural galvanic vestibular stimulation in subjects with vestibular schwannoma. *Clin Neurophysiol* 124(9):1835–1839.