Cochleovestibular Nerve Compression Syndrome. I. Clinical Features and Audiovestibular Findings

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Cochleovestibular nerve compression syndrome (CNCS) is the term used to describe a group of audiovestibular symptoms thought to be due to a vessel compressing the cochleovestibular nerve. These symptoms include recurrent vertigo, continuous disequilibrium and acquired motion intolerance. Recently, Moller reported that CNCS can be diagnosed based on abnormalities in the auditory brainstem response (ABR).

After specifically excluding all other vestibular disorders, 63 patients with symptoms suggestive of CNCS were identified. These patients were systematically evaluated with a standard neurotologic test battery, and the results were reviewed retrospectively. Hearing loss was found in 51 (81%) of 63 cases, including 33 cases of unilateral high-frequency loss and 14 cases of middle-frequency loss. ABR data were interpreted with respect to Moller's criteria, and abnormal studies were found in 42 (75%) of 56 cases. Abnormal electronystagmograms were found in 57 (93%) of 61 cases. Thirteen of the patients subsequently underwent a posterior fossa procedure for vertigo and, vessels were found in contact with the cochleovestibular nerve in 11 of 13 cases.

The results of this study suggest that the majority of CNCS patients have neurotologic test findings that suggest an abnormality of the cochleovestibular nerve. The results and their implications are discussed.

INTRODUCTION

Cochleovestibular nerve compression by a vascular loop causes a symptom complex that includes recurrent vertigo spells, continuous disequilibrium and unsteadiness, severe motion intolerance, and visually induced nausea and instability.[1] Most of the patients with these symptoms have audiovestibular test abnormalities, and some are found to have a vessel in contact with the cochleovestibular nerve, either on air-contrasted computed tomography (CT) scans or during surgical exploration of the posterior fossa. The symptoms and test abnormalities in these cases theoretically occur as a consequence of the vessel-nerve contact.

Various criteria have been used to diagnose cases of cochleovestibular nerve compression syndrome (CNCS), including the symptom of acquired motion intolerance, the presence of a vascular loop on imaging studies, and ENG abnormalities.[1] Recently, Moller[2] reported that the auditory brainstem response (ABR) is delayed on the affected side in CNCS. Specifically, Moller found an interwave latency delay between waves I and III in most CNCS patients, which she considered indicative of compression of the cochleovestibular nerve. Based on Moller's criteria, microvascular decompression of the cochleovestibular nerve has been performed. However, these criteria for diagnosing CNCS have never been independently studied or confirmed.

Over a 3-year period from July 1987 to June 1990, 63 patients with symptoms suggestive of CNCS were evaluated at The Vanderbilt Clinic. This evaluation included a complete neurotologic examination, audiovestibular diagnostic tests, and imaging studies. Thirteen of the patients underwent posterior fossa exploration with either vestibular neurectomy or microvascular decompression of the cochleovestibular nerve.

The purpose of this study was to review our experience with a series of suspected CNCS patients, specifically to determine whether these cases can be identified on the basis of symptoms or audiovestibular test abnormalities. Moller's reported criteria for diagnosing CNCS have been applied to this series of suspected cases, and have correlated these results with operative findings. The results of the study are discussed, including the significance of audiovestibular test abnormalities in suspected CNCS patients.

LITERATURE REVIEW

A review of the literature pertaining to CNCS identified numerous reports associating the presence of a vascular loop with various audiovestibular findings. These reports can be divided into three major categories, including: *1.* case studies in which a vascu-

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TABLE I.
Vascular Loop Cases Presenting as Acoustic Neuroma or Discovered at Vestibular Surgery.

Investigator	No. of Cases	Symptoms	Auditory	Auditory Brainstem Response	Electronystagmography	Imaging
Applebaum and Valvassori[5] (1985)	15	Progressive HL	HFSNHL	WNL	SN 14/15 RVR 5/15	Air CT
Brookler and Hoffman[3] (1979)	5	Progressive HL	SNHL	ABN 1/5	RVR 5/5	PFM
McCabe[9] and Harker[7] (1983)	8	Disequilibrium motion intolerance/vertigo	SNHL	N/A	SN 7/8	WNL PFM/CT
Wiet, *et al.*[8] (1989)	6	Vertigo disequilibrium hearing loss	SNHL	ABN 3/5	N/A	N/A
Bertrand, *et al.*[6] (1977)	5	Vertigo disequilibrium HFS or TN	WNL	N/A	RVR 5/5	Pneumoencephalogram

HFSNHL = high-frequency sensorineural hearing loss; SNHL = sensorineural hearing loss; SN = spontaneous nystagmus; RVR = reduced vestibular response; HFS = hemifacial spasm; TN = trigeminal neuralgia; WNL = within normal limits; ABN = abnormal; N/A = not available; PFM = posterior fossa myelogram.

lar loop was found in the process of working up a patient for an acoustic tumor, 2. case studies in which a vascular loop was found during posterior fossa surgery, usually for Meniere's syndrome, and 3. case studies in which CNCS patients were preoperatively identified with disequilibrium, and with either an audiovestibular test or an imaging abnormality. In the first two categories, vascular loop cases were retrospectively reviewed to determine if there was a pattern in the audiovestibular or imaging studies that predicted the presence of the vascular loop. With respect to the third category, most of these patients subsequently underwent surgery to relieve the symptoms. The incidence of finding a vessel and the success rate of the surgery are the focus of these reports. The literature review is summarized in Tables I and II, and the more significant studies are briefly discussed below.

Vascular Loops Associated With Suspected Acoustic Tumors

Brookler and Hoffman[3] reported 5 cases of vascular loop in the internal auditory canal. All 5 patients complained of either vertigo or disequilibrium, and 4 of 5 had a caloric weakness on electronystagmography (ENG) testing. In addition, all 5 had a unilateral sensorineural hearing loss with cochlear findings on site-of-lesion testing.

Applebaum and Valvassori[4,5] concluded that a patient with spontaneous nystagmus, a normal caloric test, and a cochlear hearing loss should be suspected of having a vascular loop in the internal auditory canal. These patients may have a widened internal auditory canal, and may also have a questionable mass in the cerebellopontine angle on routine-contrasted computed tomography (CT) scan.

Vascular Loops Discovered at Surgery for Vertigo

Bertrand, *et al.*[6] reported five cases with either hemifacial spasm or trigeminal neuralgia, in addition to vestibular symptoms. Bertrand stated that, like other cranial nerves, the cochleovestibular nerve can be cross-compressed by a vascular loop, and, furthermore, that the finding of caloric weakness with spontaneous nystagmus should make the clinician suspicious of this entity.

McCabe and Harker[7] stated that vascular loop cases resembled vestibular Meniere's disease, although the true vertigo spells usually lasted only 2 to 3 minutes. The predominant symptom seen in this this group of patients was acquired motion intolerance, especially to motions that required head turning. In contrast to patients with Meniere's disease, these patients had continuous disequilibrium and motion intolerance between vertigo spells. McCabe concluded that patients with acquired motion intolerance, other symptoms suggestive of vestibular Meniere's disease, spontaneous nystagmus, and sway on the sharpened Romberg test should be evaluated for a vascular loop.

In 1989, Wiet, *et al.*[8] concluded that patients with vascular loops may have disequilibrium and motion intolerance, may have varying degrees of sensorineural hearing loss, and may have occasional abnormalities on special audiometric testing.

Cochleovestibular Nerve Compression Syndromes: Preoperative Diagnosis Based on Disequilibrium and Either Audiovestibular Test or Imaging Abnormality

In 1980, Leclercq, *et al.*[9] concluded that a patient with a slowly progressive unilateral sensorineural loss with good speech discrimination, a decreased caloric response, and disabling symptoms should undergo exploration and decompression of the cochleovestibular nerve.

McCabe and Gantz[1] stated that a vascular loop should be considered in patients with frequent brief spells of vertigo, disabling motion intolerance, and the failure of medical and surgical regimens. McCabe concluded that the clinical symptomatology, vestibu-

TABLE II.
Vascular Loop Compression Syndrome: Diagnosis Based on Symptoms and Tests.

Investigator	No. of Cases	Symptoms	Test Abnormality	Vessels Found	Results of MVD
LeClerq, *et al.*[9] (1980)	10	Vertigo	Hearing loss caloric weakness	100% (surgery)	100% cured 0% failed
McCabe and Gantz[1] (1989)	34	Acquired motor sickness Vertigo Disequilibrium	Air-contrast CT	100% (surgery)	79% cured 17% improved 4% failed
Moller-DPV[2] (1990)	41	Disequilibrium Vertigo	ABR (Wave I–III prolongation)	100% (surgery)	50% cured 23% improved 27% failed
Ter Bruggen *et al.*[10] (1987)	10	Vertigo	ABR	70% (arteriogram)	66% cured 33% failed

MVD = microvascular decompression.

lar test abnormalities, and the presence of vessels on air-contrasted CT scans should be used as the criteria for surgery.

Ter Bruggen, *et al.*[10] reported findings for a series of 10 cases of "octavus nervous compression syndrome" (ONCS). This diagnosis was based on the presence of recurrent vertigo spells, vertigo exacerbated by changing head positions, and abnormalities on ABR. ABR abnormalities used as criteria for diagnosis included wave I absolute latency delays, wave I–III latency interval delays, and wave I–V latency interval delays.

Meyerhoff and Mickey[11] reported two cases in which patients had tinnitus and high-frequency hearing loss without vertigo or disequilibrium. These patients had retrocochlear findings on audiometric evaluation and normal magnetic resonance imaging (MRI) scans of the cerebellopontine angle. At surgery, a vascular loop was found compressing the cochlear nerve.

"Disabling Positional Vertigo"

M. Moller, A. Moller, and Jannetta have published extensively regarding CNCS, a condition they have termed "disabling positional vertigo" (DPV).[2,12–15] The onset in DPV patients was characterized by sudden, intense whirling vertigo, nausea, and vomiting—an onset that is indistinguishable from acute vestibular neuritis or viral labyrinthitis. However, as the acute symptoms abated, the patient then experienced a constant feeling of unsteadiness and disequilibrium. Nausea and vomiting were also persistent symptoms. Symptoms were worsened by ambulation and improved by bedrest.

Auditory symptoms were characterized by a slowly progressive hearing loss, usually accompanied by tinnitus. The following patterns of hearing loss were found in a series of 41 cases[2]: unilateral high-frequency loss (7), middle-frequency "notch" loss (8), low-frequency loss (2), and severe flat hearing loss (5). Moller, *et al.*[15] also found ENG abnormalities in 25 of 41 cases. These abnormalities consisted of the following: reduced vestibular response (5), reduced vestibular response with spontaneous nystagmus (7), spontaneous nystagmus alone (6), directional preponderance (4), and direction fixed or changing positional nystagmus (3). No cases of hyperactive calorics were reported.

According to Moller,[2] the most important study to diagnose DPV is the auditory brainstem response (ABR) because it represents "definitive" evidence of involvement of the cochleovestibular nerve itself. Moller's criteria for abnormal ABRs are listed in Table III. In her most recent report regarding DPV,[2] she reported that 35 (85%) of 41 cases had abnormal ABRs, as defined by these criteria. Most of these cases had prolonged wave I–III intervals. Acoustic reflex abnormalities were also found in 16 (39%) of the 41 cases.

In an earlier report, Jannetta, *et al.*[14] correlated the location of the vessel with the specific symptoms. He reported that in cases of vertigo and disequilibrium, the vessel crossed the vestibular nerve and in cases of tinnitus, the vessel crossed the cochlear nerve. When both symptom complexes were present, the entire nerve root was cross-compressed or multiple vessels were present.

MATERIALS AND METHODS

Patient Selection

History. The period of study was from July 1, 1987 to

TABLE III.
Criteria for Abnormal BAEPs in Disabling Positional Vertigo (from M. Moller[2]).

Wave I–III interval differences ≥ 0.2 msec
Wave I–III interval difference ≥ 0.16 msec if low or absent wave II
Wave II amplitude < 33% contralateral side
Contralateral wave III–V interval difference ≥ 0.2 msec
Contralateral wave III–V interval difference ≥ 0.16 msec if low or absent wave II
Ipsilateral wave I–III absolute interval > 2.3 msec
Contralateral wave III–V absolute interval > 2.2 msec

June 30, 1990. All of the patients were seen and evaluated by the authors at The Vanderbilt Clinic. During the initial evaluation, a vertigo history was obtained, including the onset, duration, and frequency of vertigo spells. Vertigo that had not occurred within the last year was noted, as was the presence of positional vertigo. Each patient was specifically questioned regarding the presence of continuous unsteadiness and disequilibrium. Patients with intermittent vertigo but no unsteadiness were excluded from the study. Specifically, patients with symptoms suggestive of acute vestibular neuritis, perilymph fistula, traumatic or suppurative labyrinthitis, benign paroxysmal positional vertigo, classic Meniere's disease, or a known acoustic neuroma were specifically excluded from the study. Other vertigo symptoms specifically sought were the presence of motion intolerance and optokinetic-induced vertigo.

An auditory history was also obtained, including the presence of a hearing loss, and whether it had progressively worsened. Other auditory symptoms specifically sought were tinnitus, ear fullness, and fluctuation of hearing. Patients noting any of these three symptoms were carefully evaluated to determine if they had endolymphatic hydrops. This evaluation included electrocochleographic (ECochG) responses, autoimmune and syphilis blood tests and, in some cases, glycerol testing. Patients with a history of ear drainage, stapes surgery, or a known temporal bone fracture were specifically excluded from the study group. A medical history was also obtained, including prior medical illness, vestibular diagnoses, and vestibular surgery.

Physical Examination. The physical examination included a complete otolaryngological/head and neck examination and a neurotologic evaluation. The head and neck examination specifically included auscultation of the carotid arteries, and those with a distinct carotid bruit were excluded. The otologic examination included either microscopic or otoscopic inspection of the tympanic membrane, and cases with obvious middle ear disease were also excluded from the study. The neurotologic examination included the following: *1.* an evaluation for spontaneous nystagmus and gaze nystagmus, *2.* a standard Romberg test in addition to a sharpened Romberg, performed with one foot in front of the other, *3.* an observation of the gait, *4.* dysmetria testing, *5.* pneumatic otoscopy to detect a positive Hennebert's sign. Patients with findings suggestive of cerebellar or cortical abnormalities were excluded from the study.

Audiometric Evaluation. Each patient had a basic audiometric evaluation with impedance audiometry. The basic audiometric evaluation included standard air- and bone-conducted threshholds for pure tones and speech reception, and speech discrimination testing using a standard list of phonetically balanced (PB) words. When indicated, speech discrimination testing was performed at multiple intensities to derive a speech function. Impedance audiometry included tympanometry, acoustic reflex threshholds, and acoustic reflex decay testing. The audiometric data were interpreted at the time of the initial evaluation. Patients with a low-frequency sensorineural hearing loss or a markedly asymmetric flat sensorineural loss were specifically excluded from the study. Patients with a low-frequency trend to their hearing, but with threshholds within the normal range were not excluded.

Electrophysiological Testing. The audiovestibular tests were performed by several audiologists, but each test was reviewed and interpreted by one of the authors. Patients with a history or findings remotely suggestive of Meniere's disease underwent electrocochleography evaluation to determine if there was evidence of enlargement of the summating potential. ECochG was recorded using either external canal or transtympanic electrode recording sites. A click stimulus was used, at intensities of 90 to 95 dB nHL. Clicks were presented at a rate of 7.1/s. Three averages of 1000 stimulus repetitions each were recorded using a standard, commercial evoked-potential system. Responses were filtered with a bandpass of 3 to 1500 Hz. The action potential (AP) was identified and its amplitude measured from the baseline; the summating potential (SP) was also identified (occasionally requiring a rapid rate for identification) and its amplitude was measured from the same baseline. An SP:AP amplitude ratio was calculated, and was judged to be increased if the ratio was greater than 0.30 when recorded transtympanically and 0.50 when recorded using a canal electrode. Patients with an increased SP:AP ratio were excluded from the study.

Most of the study patients then underwent ABR testing. Each test was administered in a quiet room, with the patient reclining on a stretcher. Recording sites were the earlobe (inverting electrode) and the high forehead (noninverting electrode), with the nasion used as a ground. Either ear inserts (ER-3A) or headphones (TDH-49) were used during the course of the study. A click stimulus, and in some cases, a 2000-Hz tone burst stimulus were delivered at 90 to 95 dB nHL. Responses were recorded with a high-pass filter setting at 30 Hz and a low-pass filter setting at 3000 Hz. Responses were analyzed for wave latencies, interwave intervals, and for abnormal waveform morphology. Wave II amplitudes were specifically measured bilaterally and recorded. The specific data tabulated for the study are further described later.

Most patients also underwent electronystagmography testing (ENG). ENGs were obtained using conventional electrode sites, and were plotted on a two-channel strip chart recorder. Standard saccadic testing included a measure of spontaneous nystagmus in the supine position with the eyes closed, optokinetic testing, gaze nystagmus, and light-tracking tests. Positional nystagmus was elicited using the various head positions, including Dix-Hallpike maneuvers. Caloric excitability was performed using an open irrigating system, delivering water at 30°C and 44°C, for 45 seconds per irrigation. Caloric excitability and directional preponderance were calculated in the usual fashion. A caloric weakness greater than 20% or a directional preponderance greater than 25% were specifically reported. Hyperactive caloric responses were also reported, and were defined as either a single response greater than 50 degrees per second, or a total response for 4 caloric tests of greater than 150 degrees per second.

Imaging Studies. During the course of the study, three different types of images were obtained: routine-contrasted CT scans, air-contrasted CT scans, and magnetic resonance imaging (MRI) of the head and internal auditory canals. Routine-contrasted CT scans were obtained using Omnipaque®; high-resolution bone and soft-tissue enhancement programs were used in some cases. In these cases, 1.0-mm sections through the region of the internal auditory canal were obtained. The presence of enlargement (a 1.5-mm difference or greater) of the internal auditory canal or of vascular enhancement were specifically noted.

Air-contrasted CT scans were performed by the radiologist. Air-contrasted CT scans were obtained by first performing a lumbar puncture and removing 7 to 10 mL of cerebrospinal fluid. The spinal fluid was sent for protein, glucose, and cell analysis. After removal of the spinal fluid, 7 to 10 mL of air was injected into the subarachnoid space. The patient was placed in the scanner in the lateral position. Simultaneously, intravenous Omnipaque® was administered. Thin-section studies of both internal auditory canals were obtained and interpreted for the presence of a vascular loop.

Magnetic resonance imaging scans were obtained using both T1- and T2-weighted sequences, with 3-mm sections through the posterior fossa, to visualize the cochleovestibular and facial nerve bundle. The special sections were made using axial and coronal planes. The majority of cases had gadolinium-DTPA (Magnavist®) contrast enhancement, although some did not because the studies were obtained prior to Food and Drug Administration approval of the agent. Each scan was magnified to enable close inspection of the cochleovestibular nerve. An artery on this study is represented by a signal void, created by the rapid flow of blood through the vessel. To be considered positive for a vascular loop, the radiologist had to clearly see a vessel, not just the indentation of the cochleovestibular nerve. Cases that did not clearly show a vessel were read as negative.

Treatment. After completing the electrophysiologic testing and imaging studies, most patients were treated symptomatically. The more severe cases were counseled regarding surgery. Medically managed patients were treated with either Valium® or Xanax®, in addition to Cawthorne habituating exercises. Patients treated medically were followed periodically, as the severity of symptoms warranted. At each visit, patients were questioned regarding their symptoms, adjustments were made in medicines as necessary, and the information was recorded. Persistent disabling vertigo and disequilibrium despite medical treatment were symptoms that prompted the consideration of surgery.

Surgical therapy was offered to patients with severe, uncontrolled unsteadiness and disequilibrium, and, in most cases, recurring vertigo. The side of the lesion was based on the history and the presence of either a hearing loss, caloric weakness, or ABR abnormality. In some cases, the side of the lesion was determined by the presence of a vessel on air-contrasted CT scan. Most surgical patients were treated with vestibular nerve section, although some were treated with microvascular decompression.

Surgical Techniques. Surgical procedures were performed by the senior author and a neurosurgical colleague. The initial setup and exposure were the same for both vestibular nerve section and microvascular decompression. After performing a suboccipital craniotomy and opening the dura, the cerebellum was gently retracted posteriorly, with particular attention to the inferior portion of the cerebellopontine angle. The basilar cistern was opened first, and the cerebrospinal fluid (CSF) allowed to egress. The cerebellum was retracted by a neurosurgical Layla retractor. The various adhesions surrounding the cochleovestibular and facial nerves were then dissected, and the flocculus of the cerebellum was retracted to further visualize the root entry zone of the nerves. At this point, a drawing or photograph was made to demonstrate the relationship of the cochleovestibular nerve and the nearby vessels.

TABLE IV.
Demographic Data and Symptoms Tabulated.

A. Demographic Data
   1. Total number of patients seen during study period
   2. Total number of vertigo patient seen during study period
   3. Age at initial evaluation
   4. Sex
   5. Duration of symptoms
   6. Other head and neck dysfunctions
   7. Prior vestibular surgery
   8. Associated medical illness

B. Audiovestibular Symptoms
   1. Vertigo spells within previous year
   2. Vertigo duration: minutes to hours or brief positional
   3. Presence of maldebarquement or severe disequilibrium
   4. Tinnitus: present or absent
   5. Optokinetic induced symptoms
   6. Motion intolerance

In cases where a vestibular nerve section was performed, the cleavage plane between the cochlear and vestibular nerves was first dissected with a blunt instrument. Using microsurgical scissors, the vestibular nerve was transected. In cases where a nerve specimen was obtained, the vestibular nerve was sectioned in two spots, near the porus acousticus and near the brain stem. The vestibular nerve specimen was immediately placed in a vial containing 2% glutaraldehyde.

In cases where microvascular decompression was performed, the various vessels and arterioles were elevated from the surface of the cochleovestibular nerve. This usually required the division of arachnoidal adhesions as well as the blunt dissection of the vessels. Small pieces of cardiac felt were then insinuated between the vessels and the nerve to prevent reattachment. The cochleovestibular nerve was decompressed from the brain stem to the porus acousticus.

Data Collection

At the end of the study period, the author reviewed the medical records of the patients meeting the selection criteria, and tabulated the data. The information that was tabulated is summarized in Tables IV through VII, and includes demographic data, audiovestibular symptoms, audiometric data, acoustic reflex data, ABR data, ENG data, and imaging data.

Audiometric data are summarized in Table V. The specific patterns of sensorineural hearing loss were determined from the basic audiogram, and the patterns were categorized as either a middle-frequency sensorineural loss (1000 to 4000 Hz), a high-frequency sensorineural loss (4000 to 8000 Hz), a severe loss (>60 dB HL), or a low-frequency tendency (250 to 1000 Hz) but with a normal pure-tone average. To be included in one of these categories, a 15-dB loss within the specified frequency range must have been present; these cases might not have a 15-dB difference in pure-tone average. Cases with a 15-dB difference in pure-tone average or a 15% difference in speech discrimination were also specifically tabulated.

The ABR and ENG data were copied directly from the report form. The ABR data tabulated were based on Moller's reported criteria for the diagnosis of disabling positional vertigo as listed in Table III. Also, a difference between ears

TABLE V.
Audiometric Data Tabulated.

A. Audiometric Data
   1. Presence or absence of sensorineural hearing loss
   2. Patterns of hearing loss (≥ 15 dB in specific range)
      a. Middle-frequency loss (1,000–4,000 Hz)
      b. High-frequency loss (4,000–8,000 Hz)
      c. Severe hearing loss (≥ 60 dB HL)
      d. Low-frequency tendency (250–1,000 Hz) (with normal pure-tone average)
   3. Presence of a 15-dB difference in pure-tone average
   4. Presence of 15% difference in speech discrimination
B. Acoustic Reflex Data
   1. Presence or absence of acoustic reflex
   2. Reflexes with elevated threshold ≥ 95 dB HL in the presence of normal pure-tone thresholds
   3. Presence of reflex decay
   4. Reflexes abnormal unilateral or bilateral

TABLE VI.
Electrophysiologic Data Tabulated.

A. Auditory Brainstem Response
   1. Wave I–III interval difference ≥ .20 msec
   2. Contralateral wave III–V interval differences ≥ .20 msec
   3. Wave II amplitude < .3 of contralateral value
   4. Wave I–III absolute interval > 2.3 msec
   5. Contralateral wave III-V absolute interval > 2.2 msec
   6. Wave I–V interval difference ≥ 0.4 msec
   7. Wave I–V absolute interval ≥ 4.6 msec

(Interval difference means "as compared to opposite side")

B. Electronystagmography data
   1. Normal or abnormal ENG
   2. Presence of caloric weakness ≥ 20%
   3. Presence of directional preponderance ≥ 25%
   4. Presence of spontaneous nystagmus
      a. Toward ear with hearing loss
      b. Away from ear with hearing loss
      c. Could not determine
   5. Presence of hyperactive caloric responses (either single value ≥ 50°/sec or total value ≥ 150°/sec)

in the wave I–V interval of 0.4 msec or greater, or an absolute wave I–V interval greater than 4.6 msec was specifically recorded. ABR data are listed in Table VI.

The record of patients treated medically were reviewed to determine if the vertigo and disequilibrium had improved or resolved over time. Only cases with a follow-up of at least 6 months were categorized. Four categories were used: no symptoms, improved symptoms, unchanged symptoms, and worse symptoms. The medical records of patients undergoing surgery for suspected CNCS were reviewed, and categorized into cases of vestibular nerve section and into cases of microvascular decompression. The results of each procedure were tabulated into the following groups: vertiginous spells, no vertiginous spells, disequilibrium, and no disequilibrium. Also, the presence of absence of vessels at surgery was tabulated, as well as the presence of the anterior inferior cerebellar artery (AICA) in the internal auditory canal, the presence of AICA crossing the nerve in the cerebellopontine angle, and the presence of arterioles and veins crossing the cochleovestibular nerve.

RESULTS

During the course of the 3-year study, 68 patients with symptoms suggestive of CNCS were evaluated. During this same time period, the author evaluated 2859 patients with primarily otologic complaints, of which 986 patients had vertigo and balance disorders. Patients with symptoms suggestive of CNCS made up 2.4% of all otologic patients and 6.9% of vertigo patients. Of the 68 patients identified with symptoms, 5 records were incomplete. The remaining 63 cases were retrospectively reviewed in this study.

Demographic Data and Symptoms

These data are summarized in Table VIII. Of the 63 cases, 40 were women and 23 were men. The age range of the patients was from 24 to 74 years; 54 of 63 patients were between 35 and 55 years of age. The duration of symptoms varied from 0.5 years to 16 years, with an average duration of 3.3 years. Five of 63 patients had a second head and neck abnormality, including cases of hemifacial spasm, trigeminal neuralgia, facial and vocal cord palsies, and blepharospasm. Seven patients had undergone previous surgery that related to their symptoms, including four patients who had a previous endolymphatic shunt, 1 who had a middle ear exploration for a possible perilymph fistula, and 1 patient who had both procedures performed at the same setting.

During the course of the study, unsuspected vestibular disorders were discovered in two patients. One patient had an acoustic neuroma. He was included on the list of study cases because he had continuous disequilibrium, a progressive sensorineural hearing loss, and three previous imaging studies interpreted as negative for acoustic neuroma. These studies included two noncontrasted MRI scans and one high-resolution CT scan of the internal auditory canal region. However, gadolinium-enhanced MRI scans clearly showed a 1.5-cm acoustic neuroma in this case. The second patient had an elevated sedimentation rate, elevated antinuclear antibody (ANA) titers, and a positive syphilis serology. Although she presented initially with continuous disequilibrium, vertiginous spells, and normal hearing, over the 3 years of the study she developed classic syphilitic endolymphatic hydrops.

At the initial evaluation, 53 patients (84%) reported a history of vertiginous spells within the previous year, while 10 (16%) had no vertigo spells within the previous year. Of the 53 patients with recurrent vertigo, 46 described their vertiginous spells as nonpositional, lasting from a few minutes to several hours. Seven patients described vertiginous spells that were more positional in quality, being brief and movement related. All of the patients had continuous disequilibrium, and 15 patients (24%) described the disequilibrium as severe. Two patients described a rocking sensation to their disequilibrium, or a maldebarquement-like syndrome. Other symptoms noted

TABLE VII.
Imaging Data Tabulated.

A. Routine-contrasted computed tomography scans
   1. Number of studies performed
   2. Presence of widened (≥ 1.5-mm difference) internal auditory canal
      a. On side of hearing loss?
   3. Presence of increased vascularity in cerebellopontine angle
      a. On side of hearing loss?
   4. Other findings
   5. Normal studies
B. Air-contrasted computed tomography scans
   1. Number of studies performed
   2. Presence or absence of vessels on study
C. Magnetic resonance imaging scans
   1. Number of studies performed
   2. Presence or absence of vessels on study
   3. Other abnormalities

TABLE VIII.
Demographic Data (n = 63).

Men 23 Women 40
Age Range 24–74 years (mean 47 years)
(54 of 63 cases between ages 35–55)
Duration of illness 0.5–16 years (mean 3.3 years)
Other cranial nerve abnormalities (5 cases)
1 Hemifacial spasm
1 Trigeminal neuralgia
1 Blepharospasm
1 Facial palsy (ipsilateral)
1 Vocal cord palsy (ipsilateral)
Prior surgeries for vestibular disorder (7 cases)
4 Endolymphatic shunt
1 Middle ear exploration
1 E. shunt with middle ear exploration
1 Middle cerebral–temporal artery bypass
Associated illnesses (5 cases)
1 Arnold-Chiari malformation
1 Multiple pulmonary emboli
1 Seizure
2 Muscle diseases

were tinnitus in 36 patients (57%), optokinetic-induced symptoms in 29 patients (46%), and motion intolerance in 36 patients (57%).

Audiometric Data

The results of the audiometric and acoustic reflex studies are summarized in Table IX. Fifty-one patients had a sensorineural hearing loss, whereas 12 were normal. The following patterns were found: high-frequency loss (33), middle-frequency loss (14), severe (0), low-frequency trend but in normal range (4). Among the cases with high-frequency loss, 6 were bilateral and symmetric, 1 had a conductive component of 20 dB, and 3 had a sloping loss, from normal in the low frequencies to severe in the high frequencies. Fifteen patients in this series had a difference in pure-tone average between ears of 15 dB, and 8 had a difference in discrimination of 15%.

Acoustic reflexes were recorded in 53 of 63 cases, and abnormalities were found in 21 of 53 cases. These abnormalities were unilateral in 11 cases and bilateral in 10 cases. In 1 case (the acoustic tumor case) reflex decay was observed.

Electrophysiological Tests

The results of the electrophysiological tests are summarized in Table X. ABRs were recorded in 56 of 63 cases. Applying Moller's criteria as listed in Table III, 42 of 56 cases showed some abnormality suggestive of CNCS. Twenty-eight cases showed asymmetry of the wave I–III interval, and 13 showed asymmetry of the contralateral wave III–V interval. Twenty-four cases showed abnormalities of wave II amplitude, including an absent wave II in 12 and decreased amplitude of wave II in 12. Wave II was bilaterally abnormal in three cases. The ipsilateral absolute wave I–III interval was prolonged in 10 cases, and the contralateral absolute wave III–V interval was prolonged in 1 case.

Applying more conventional criteria to ABR analysis revealed a wave I–V interval difference of 0.4 msec or greater between the two sides in only two cases. One of these cases was the previously mentioned acoustic neuroma. Three cases demonstrated bilateral prolongation of the wave I–V interval greater than 4.6 msec. Of these 3 cases, 1 had trigeminal neuralgia, 1 was an elderly patient with bilateral high-frequency sensorineural hearing loss, and 1 had hyperactive caloric responses on ENG.

The results of ENG were recorded in 61 of 63 cases, and abnormalities occurred in 57. The most common abnormality was the finding of spontaneous nystagmus in 48 cases. The spontaneous nystagmus was toward the ear with the greater hearing loss in 29 cases, away from the ear with the greater hearing loss in 12 cases, and, in 7 cases, this determination could not be made. A caloric weakness was recorded in 16 cases, 15 of which also had spontaneous nystagmus. Six cases had hyperactive caloric responses and 1 had caloric reversal, *i.e.*, the nystagmus spontaneously reversed directions during the period after caloric stimulation. Twenty-eight patients had positional nystagmus; in 16 cases, the nystagmus was toward the dependent ear and in 12 cases the nystagmus was away from the dependent ear.

Imaging

Seventeen patients had routine-contrasted CT scans, whereas 7 (41%) were interpreted as suggestive of CNCS. In 5 cases, a slightly widened internal auditory canal was identified. In all 5, the abnormality was on the side with the greater hearing loss. In 2 cases the radiologist interpreted increased vascularity in the cerebellopontine angle and, in both, this was on the side of the hearing loss. One of these two cases had a markedly dolichoectatic basilar ar-

TABLE IX.
Audiometric Data (n = 63).

Sensorineural Hearing Loss	51	Normal Hearing	12
Patterns of Loss			
High-frequency	33		
Middle-frequency	14		
Severe	0		
Low-frequency trend	4		
Pure-tone average difference (≥ 15 dB)			15
Discrimination (15% difference)			8
Acoustic Reflex Data (n = 53).			
Abnormal reflexes			21
Unilateral	11		
Bilateral	10		
Reflex decay	1		
Absent reflexes	1		

TABLE X.
Auditory Brainstem Response Data (n = 56).

Abnormal studies		42 (75%)
Wave I–III interval difference ≥ 0.2 msec		28
Contralateral wave III–V interval difference ≥ 0.2 msec		13
Wave II amplitude abnormalities (< 0.30 of opposite side) (absent 12, decreased 12)		24
Wave I–III absolute interval ≥ 2.3 msec		10
Wave III–V absolute interval ≥ 2.2 msec		1
Electronystagmography Data (n = 61).		
Abnormal studies		57 (93%)
Spontaneous nystagmus		48
toward ear with hearing loss	29	
away from ear with hearing loss	12	
determination not made	7	
Caloric weakness		16
Directional preponderance		6
Positional nystagmus		28
toward dependent ear	16	
away from dependent ear	12	
Hyperactive caloric responses		6

tery. Ten scans were read as normal in the posterior fossa; this group included 1 case with a small, incidental meningioma in the frontal region. Forty-five patients had an MRI scan to evaluate the cerebellopontine angle. In 16 cases (35%), a vessel looping near the cochleovestibular nerve was clearly identified on MRI. In 23 cases, there were no vessels and no other suspicious abnormalities. Six patients had other abnormalities identified. Air-contrasted CT scans were obtained in 9 cases, and a vascular loop was identified in 6 cases. The vascular loops were found either in the internal auditory canal or attached to the cochleovestibular nerve.

Medical and Surgical Management

Twenty-nine medically managed patients that were followed for at least 6 months were identified. Of these 29 patients, 14 stated that they were improved on medical therapy, while 15 stated they were essentially unchanged. In the remainder of cases, follow-up was inadequate to determine the outcome of medical therapy.

Thirteen patients underwent surgery for vertigo and disequilibrium. Nine patients underwent vestibular nerve section. Of these 9 patients, 6 no longer have vertigo, while 3 continue to have some vertiginous spells. Only 2 of the vestibular nerve section patients reported that their disequilibrium was completely resolved, while 7 stated that they still had some disequilibrium. However, 8 of 9 cases are currently working in their previous jobs. Four patients underwent microvascular decompression (MVD) of the cochleovestibular nerve. Three of these 4 have not had vertiginous spells for at least 6 months, while 1 continues to have spells. Two of 4 have no disequilibrium, 1 is improved, and 1 is unchanged after surgery.

Surgical Findings

At surgery, vessels were found in 11 of 13 suspected CNCS cases. In two cases in which the nerve was sectioned, there was no obvious vessel found near the cochleovestibular nerve. Of the 10 surgical patients with ABR abnormalities by Moller's criteria, 8 had vessels in close proximity to the cochleovestibular nerve. The 3 surgical patients with normal ABRs also had vessels in close proximity to the cochleovestibular nerve. The vessels were characterized as follows: anterior inferior cerebellar artery (AICA) in the internal auditory canal (4), AICA crossing the cochleovestibular nerve (4), and arterioles crossing the cochleovestibular nerve (3). In three cases, a vein crossed the cochleovestibular nerve near the brain stem, in addition to finding an artery crossing the nerve.

DISCUSSION

The major objective of this study was to determine if CNCS can be identified on the basis of symptoms and test findings. This study shows that a group of patients with recurrent vertigo, disequilibrium, motion intolerance, and optokinetic-induced symptoms can be identified, after excluding the other common causes of vertigo. These patients represent a very small fraction of the patients seen in a neurotologic practice, and are essentially the same group of patients that might be classified as having uncompensated vestibular neuritis, vestibular Meniere's disease, or recurrent vestibulopathy. Most of the patients in this study had a vertiginous spell within the previous year, and all had a history of vertigo at some time in the past.

This study also shows that, in this series of patients, the audiovestibular test findings suggest an abnormality of the cochleovestibular nerve. These findings include a wave I–III interwave latency delay on ABR, spontaneous nystagmus on ENG testing, and acoustic reflex abnormalities.

Moller's criteria for ABR abnormality in disabling positional vertigo were summarized in Table III. Three fourths of the patients in this study had an ABR abnormality by these criteria. However, these results would not be considered abnormal by most clinicians who use ABR as a screening test for acoustic neuromas or for brainstem disease. In fact, there are no reported studies in which Moller's criteria have been systematically studied in normals or in patients with only a hearing loss and no vestibular symptoms. This is a serious clinical shortcoming, and control studies need to be performed to confirm the validity of these criteria.

The use of ABR to detect diseases of the cochlear nerve other than acoustic neuroma raises two specific issues. The first issue concerns the so-called "false-positives" in all site-of-lesion audiometric tests. Hall[16] reviewed the literature and summarized the diagnostic accuracy of various audiometric procedures, including acoustic reflex, ABR, phonetically balanced (PB) word recognition, and tone decay. Each procedure was analyzed with respect to its accuracy in detecting documented acoustic tumors, as well as its accuracy in correctly predicting the absence of a tumor. In these reports, an audiometric test abnormality in the absence of an acoustic tumor was interpreted as a "false-positive." According to Hall, the reported "false-positive" rate for the various audiometric tests varies considerably: 16% for acoustic reflex, 12% for ABR, 40% for PB word recognition, 9% for tone decay. Hall emphasized the concept that retrocochlear abnormalities on audiometric tests should be viewed as measures of cochlear nerve dysfunction, *i.e.,* a "false-positive" might reflect an intrinsic abnormality in the cochlear nerve that cannot be imaged.

The second issue that is raised by the use of ABR to detect CNCS is the validity of using a measure of cochlear nerve function to predict the presence of vessels contacting the cochleovestibular nerve. Ten of the 13 surgical cases in this study had abnormal ABRs based on Moller's criteria. Two of these 10 cases had no obvious vessel in the proximity of the cochleovestibular nerve. In the 3 surgical cases with normal ABRs, vessels were found in the proximity. These findings suggest that Moller's ABR criteria are not very specific for diagnosing the presence of a vessel compressing the cochleovestibular nerve but, rather, as emphasized by Hall, probably reflect cochlear nerve dysfunction.

Abnormal ENGs were recorded in 93% of the patients in this study and, most patients had either caloric weakness, positional nystagmus, or spontaneous nystagmus. The cases in this study were like McCabe's patients, in that spontaneous nystagmus was the most frequent abnormality on ENG, and the direction of the spontaneous nystagmus was most often toward the ear with the greater hearing loss. With respect to another series of CNCS cases, Applebaum and Valvassori[5] have suggested that spontaneous nystagmus in the absence of a caloric weakness indicates a primary vestibular nerve disorder.

Patients in this study had either a high-frequency loss, a middle-frequency loss, or normal hearing. High-frequency hearing loss has been previously associated with vestibular neuritis,[17] particularly in the early stages of the illness. It is possible that the cases in this study are examples of this relationship. Middle-frequency hearing loss is infrequently encountered in clinical practice and, the finding of 14 cases in this study suggests that a relationship also exists between this pattern and the vestibular symptoms. This study is the first independent confirmation of Moller's previous reports[15] regarding the relationship of middle-frequency hearing loss and CNCS.

The patients in this study were imaged using a variety of scans, including regular-contrast CT scans, air-contrast CT scans, and MRI scans. Regular-contrast CT scans detected a slightly widened internal auditory canal in five cases. In each case, the widening was on the side with the greater hearing loss. MRI scans showed a definite vessel at the root entry zone of the cochleovestibular nerve in 35% of cases, an incidence that is similar to that reported by Parnes, *et al.*[18] in a series of asymptomatic non-CNCS cases. Vessels are nearly always in the vicinity of the cochleovestibular nerve, so that it is not surprising that they are frequently imaged on various scans. A gadolinium-enhanced MRI scan is currently the imaging procedure of choice for evaluating suspected CNCS patients, and the scan should be obtained with special views of the cochleovestibular nerve.

Based on the findings in this study, the author no longer recommends vestibular nerve section for CNCS cases. Although nerve section eliminated the vertiginous spells in two thirds of cases, the procedure failed to control the disequilibrium.

The greatest controversy concerning CNCS is that involving the effects of microvascular decompression on the cochleovestibular nerve. It is not clear if microvascular decompression stops the triggering effect of a vessel, or if the procedure simply traumatizes the nerve, partially denervating it. A third possibility is that microvascular decompression is actually a sham procedure, that depends on the placebo effect for the outcome. It should be specifically stated that postoperative surgical results are not scientific evidence of the validity of this concept.

SUMMARY

Patients with CNCS have a history of current vertigo, disequilibrium, motion intolerance, and visually induced instability. After excluding other vestibular disorders, 63 suspected CNCS patients were evaluated over a 3-year period; these cases represent 2% of the patients seen in a tertiary neurotologic practice. The audiovestibular test findings suggest that suspected CNCS patients have an abnormality of the

cochleovestibular nerve. Suspected CNCS patients have either a high-frequency loss, a middle-frequency loss, or normal hearing. Three fourths of these patients have ABR abnormalities as defined by Moller's criteria. Twenty percent have other retrocochlear audiometric test abnormalities. Most have an ENG abnormality, usually either spontaneous nystagmus, positional nystagmus, or decreased caloric excitablity. Ten percent have markedly increased caloric excitability. Vestibular nerve section eliminates the recurrent vertiginous spells in two thirds of these patients, but does not significantly alter the constant disequilibrium. The effects of microvascular decompression on the cochleovestibular nerve are not known at this time.

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