The Association Between Semicircular Canal Dehiscence and Chiari Type I Malformation

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Objective: To investigate the association between semicircular canal dehiscence (SCD) and Chiari type I malformation (CM-I).

Design: Retrospective case series.

Setting: Military tertiary referral center.

Patients: Adult patients with SCD.

Intervention: Review of records of patients diagnosed as having SCD for the radiologic diagnosis of CM-I and presenting symptoms.

Main Outcome Measures: The prevalence of CM-I among patients with SCD and the presenting symptoms of patients with SCD with and without a coexistent CM-I.

Results: Of 32 patients diagnosed as having SCD, 30 underwent magnetic resonance imaging of the brain. Seven patients (23%; 95% confidence interval [CI], 12%-41%) were found to have a CM-I. Chiari type I malformation was associated with superior SCD in 3 of 26 patients (12%; 95% CI, 3%-30%). Of 10 patients with bilateral superior SCD, 2 (20%; 95% CI, 5%-52%) had a CM-I. Five of 6 patients (83%; 95% CI, 42%-99%) had a CM-I with unilateral or bilateral posterior SCD. Twenty-nine records were reviewed for presenting symptoms, and no significant difference was observed between patients with SCD alone and those with an associated CM-I (P = .09-.64).

Conclusions: Among patients with SCD, the prevalence of CM-I is elevated. This association is especially marked in patients with posterior SCD. This finding suggests a relationship between CM-I and SCD, particularly with posterior SCD.

nal fossa, which results in elevated intracranial pressure, causing characteristic symptoms and effacement of cranial bone.\textsuperscript{13,14} We hypothesize that because of the close proximity of the bony labyrinth to the floor of the cranium, erosive changes over time may lead to SCD. The purpose of this study was to examine the relationship between SCD and CM-I to determine whether the prevalence of CM-I is increased in patients with SCD.

### METHODS

We conducted a retrospective review of the clinical records of all patients diagnosed as having SCD by the senior author (J.J.K.) at Naval Medical Center Portsmouth, Portsmouth, Virginia, during the period from November 2001 to January 2009. The review included neurotology clinical records as well as the clinical records contained in the hospital’s electronic medical record system. Specifically, we noted the location (posterior or superior semicircular canal) and laterality (unilateral or bilateral) of dehiscent semicircular canals as well as the presenting auditory and vestibular symptoms and relevant audiometric findings.

High-resolution CT of the temporal bones was performed on a 64-multidetector-row CT scanner (GE Light Speed VCT scanner; GE Healthcare, Chalfont St Giles, England) using acquisition parameters of 120 kV and 250 to 400 mA. Contiguous direct axial and coronal sectional images were acquired with 0.625-mm collimation. Multiplanar reconstructions were conducted for directly acquired temporal bone CT images that raised suspicion for SCD. These images were reformatted in the plane of the superior semicircular canal (Poschl plane) and perpendicular to the plane of the superior semicircular canal (Stenver plane). Also, we searched for and reviewed MRI findings to identify patients who had been diagnosed radiographically as having a CM-I. In cases of CM-I, the radiographic diagnosis was confirmed by a neuroradiologist based on a finding of tonsillar herniation of at least 5 mm below the foramen magnum. The presence of CM-I was confirmed by a neuroradiologist based on a finding of tonsillar herniation of at least 5 mm below the foramen magnum. In cases of CM-I, the radiographic diagnosis was confirmed by a neuroradiologist based on a finding of tonsillar herniation of at least 5 mm below the foramen magnum.

The relationship between bilateral SCD and CM-I was also explored (Table 2). Of 10 patients with bilateral superior SCD, only 2 (20%; 95% CI, 5%-52%) had a CM-I. One of the 2 had combined bilateral superior and posterior SCD (Figures 1, 2, and 3). Of patients with bilateral posterior SCD, 4 of 4 (100%; 95% CI, 43%-100%) had a CM-I. One of these 4 patients had combined bilateral superior and posterior SCD. Therefore, to accurately compare the prevalence of CM-I in patients with bilateral SCD and one case of unilateral superior and contralateral posterior SCD.

### RESULTS

A total of 32 patients were diagnosed as having SCD. Thirty of the 32 patients had also undergone MRI of the brain. The mean (SD) age of patients with SCD at the time of diagnosis was 41.5 (14.8) years. Sixteen of the patients (50%) were female. Of the 32 patients with SCD, 26 (81%) had unilateral or bilateral superior SCD (Table 1). Likewise, unilateral or bilateral posterior SCD was observed in 4 patients (12%), and combined superior and posterior SCD was seen in 2 patients (6%).

Of the 30 patients with SCD who underwent MRI of the brain, 7 (23%; 95% CI, 12%-41%) were found to have radiographic evidence of a CM-I (Table 2). When the cases were stratified by specific semicircular canal involvement, CM-I was seen most often in association with posterior SCD (with or without concomitant superior SCD). Four of the 4 patients (100%) with SCD involving only the posterior canal and 1 of 2 patients (50%) with combined posterior and superior semicircular canal involvement were found to have a CM-I. Therefore, the proportion of patients with a CM-I among those with any posterior SCD was 83% (95% CI, 42%-99%). With regard to patients with superior SCD, 3 of 26 (12%; 95% CI, 3%-30%) were found to have a CM-I. To accurately compare the prevalence of CM-I in patients with superior SCD with those with posterior SCD, cases of combined superior and posterior SCD were excluded from analysis. This exclusion revealed that CM-I is associated with posterior SCD significantly more often than with superior SCD (100% vs 8%; P = .02).

We found that
a CM-I occurred in 100% of the bilateral posterior SCD cases compared with 11% of the bilateral superior SCD cases; however, the difference was not found to be significant ($P = .12$).

As noted herein, 30 patients with SCD also underwent MRI. Of this group, 29 (97%) had clinical records available for review. The records were reviewed for audiometric findings and presenting symptoms. Audiometric findings of interest included the presence of conductive hearing loss, elevated bone conduction thresholds, and vestibular evoked myogenic potential (VEMP) testing. Results of audiometric testing demonstrated variability in the expected audiometric manifestations of SCD. For example, while conductive hearing loss was seen in 14 of 30 patients (47%), elevated bone conduction thresholds were seen in only 2 of the 30 patients (6%). In these cases, audiometric testing was specifically performed to detect thresholds below 0 dB hearing level. Because VEMP testing was not available at our institution before 2007, it was performed in only 14 patients. Still, VEMP testing in 8 of 14 patients (57%) demonstrated reduced thresholds (<70 dB), while 5 patients did not have a recordable VEMP. This outcome was attributed to early use and inexperience with VEMP testing. **Table 3** compares presenting symptoms among patients with SCD with and without a CM-I. No significant difference was found between the 2 groups with respect to symptoms.

<table>
<thead>
<tr>
<th>Variable</th>
<th>SCD Only, 22 Records Reviewed</th>
<th>SCD With CM-I, 7 Records Reviewed</th>
<th>$P$ Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pressure-related vertigo</td>
<td>7 (32)</td>
<td>1 (14)</td>
<td>.64</td>
</tr>
<tr>
<td>Head motion vertigo</td>
<td>17 (77)</td>
<td>6 (86)</td>
<td>.63</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>10 (46)</td>
<td>5 (71)</td>
<td>.39</td>
</tr>
<tr>
<td>Pulsatile tinnitus</td>
<td>4 (18)</td>
<td>0</td>
<td>.56</td>
</tr>
<tr>
<td>Aural fullness</td>
<td>7 (32)</td>
<td>5 (71)</td>
<td>.09</td>
</tr>
<tr>
<td>Autophony</td>
<td>2 (9)</td>
<td>2 (28)</td>
<td>.24</td>
</tr>
<tr>
<td>Tullio phenomenon</td>
<td>7 (32)</td>
<td>2 (28)</td>
<td>.09</td>
</tr>
<tr>
<td>Hypersensitivity to sound</td>
<td>3 (14)</td>
<td>3 (43)</td>
<td>.13</td>
</tr>
</tbody>
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**Table 3. Presenting Symptoms in 29 Patients With Semicircular Canal Dehiscence (SCD) With and Without Chiari Type I Malformation (CM-I)**

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Figure 1. Sagittal T1-weighted magnetic resonance image of the brain. Herniation of cerebellar tonsil to 17 mm below the level of foramen magnum (arrow).

Figure 2. Axial high-resolution computed tomogram of the right temporal bone demonstrating superior semicircular canal dehiscence (arrow).

Figure 3. Axial high-resolution computed tomogram of the right temporal bone demonstrating posterior semicircular canal dehiscence (arrow) and scalloping along the posterior face of the temporal bone.
The clinical presentation, audiometric and vestibular electrodagnostic findings, and high-resolution CT findings as well as the surgical management of superior SCD are well described. \(^5-24\) Furthermore, variants such as bilateral superior SCD, dehiscence of the posterior SCD, and dehiscences involving multiple canals have also been described in retrospective case series\(^16-20,24\) and case reports. \(^25-29\) However, to our knowledge, the coexistence of a CM-I in patients with SCD has not been previously described in the medical literature. Although several reports have reviewed the neurotologic manifestations of CM, none have identified a specific radiographic abnormality of the semicircular canals. \(^5,8,10-32\)

A CM-I is a disorder of unknown origin that consists of a downward displacement of the medial and inferior aspects of the cerebellar tonsils through the foramen magnum. \(^33\) It may also be associated with minor degrees of elongation of the fourth ventricle and medulla. Cerebrospinal fluid (CSF) outflow obstruction and caviation of the spinal cord (syringomyelia) may occur in association with increased displacement of intracranial structures through the foramen magnum. Chiari type II malformation (CM-II) is also associated with protraction of the cerebellar vermis and medulla and caudal displacement of the fourth ventricle into the vertebral canal. It is also associated with hydrocephalus and meningomyeloceles. Despite the radiographic finding of hydrocephalus in 1 of our patients, other aspects of a CM-II malformation were absent. This patient could therefore be classified as having an extended CM-I malformation. In contrast to other forms of CM, CM-I tends to present in the second and third decades of life and may be referred to as an “adult-type” CM. In contradistinction to a CM-I, a CM-II is typically present at birth, as are the more severe types III and IV.

The results of this review demonstrate that the prevalence of CM-I in patients with SCD occurs at a substantially higher rate than that expected in the general population (23% vs 0.6%-1%). In particular, there appears to be a strong association between posterior semicircular canal involvement and the coexistence of CM-I. Five of 6 patients (83%) with posterior SCD were also found to have a CM-I. Of these 5 patients with a coexistent CM-I, 4 (80%) had bilateral posterior canal involvement. Therefore, given the proximity of the posterior semicircular canals to the posterior cranial fossa, it would appear that some process related to CM-I influences the development of posterior SCD. This same process may also affect the middle cranial fossa and the subtemporal surface of the temporal bone, although to a lesser extent. Our results demonstrate a much lower rate of CM-I among patients with either unilateral or bilateral superior SCD (12%) as compared with patients with any posterior canal involvement (83%). In either case, a much higher rate of CM-I was found in patients with any form of SCD (23%) when compared with the estimated prevalence of CM-I in the general population (0.6%-1%). While it is tempting to compare the elevated prevalence of CM-I in this cohort with that of the estimated population prevalence, we recognize that the small sample size limits any assumptions regarding the significance of this association.

Current evidence suggests that the pathogenesis of CM-I can be attributed to overcrowding of the hindbrain by an underdeveloped posterior cranial fossa, resulting in dynamic changes in CSF flow that may lead to increased intracranial pressure and the formation of syringomyelia. \(^13,14,34\) Phased-contrast cine MRI studies in patients with CM-I have demonstrated CSF flow abnormalities involving pathways through the aqueduct, fourth ventricle and its outlets, foramen magnum, preopticine cistern, around the cerebellar tonsils, and ventral and dorsal to the cervical spinal cord. \(^35,36\) Although most patients with CM-I may demonstrate CSF flow obstruction, fewer than 50% of patients will present with occipital headaches. \(^37\) In our series, only 1 of 7 patients with a CM-I complained of headache. The remaining patients in this group were asymptomatic as it pertains to the typical symptoms attributable to CM-I, such as headaches, cervical pain, motor weakness or sensory changes in the extremities, gait disturbance, and cranial neuropathies. Not only are these considered the most common presenting symptoms in CM-I, but they also tend to predominate and precede the development of auditory symptoms or symptoms that would suggest a peripheral vestibular deficit. \(^3\) In one study, the results of vestibular electrodagnostic testing in a group of 77 patients with a CM-I suggested a functional deficit in the vestibulocerebellum. \(^39\) The authors therefore attributed symptoms of vertigo to a central vestibular disorder. Although we do not present the results of comprehensive vestibular electrodagnostic testing in our patient population, there was no evidence of a central vestibular disorder in 6 of the 7 patients with an associated CM-I. The 1 patient with a history of occipital headaches, whose imaging studies are presented in Figures 1, 2, and 3, demonstrated evidence of a central vestibular disorder on oculomotor testing. He eventually underwent posterior fossa decompression and cervical laminectomy. After surgery, his headaches and neck pain resolved, and a neurologic examination revealed resolution of a lateral gaze-evoked, nontorsional, down-beat nystagmus. His pressure-related (vertigo with cough or sneezing) and sound-evoked symptoms of vertigo were unchanged, suggesting that persistent symptoms of vertigo were due to SCD. Of the remaining patients who were considered to be asymptomatic as it pertains to an associated CM-I, 1 patient had surgical plugging of a unilateral superior SCD with resolution of auditory (autophony) and vestibular (pressure-related and sound-evoked vertigo) symptoms after surgery. The fact that there was no significant difference in symptoms reported by patients with SCD alone compared with those with an associated CM-I may be attributed to the possibility that 6 of 7 patients with an associated CM-I were symptomatic entirely as a result of SCD. Given that we observed a much lower rate of CM-I in association with superior canal dehiscence, it is hypothesized that a pressure differential may exist between the posterior and middle cranial fossae in patients with a CM-I and that there may be other factors to account for the development of superior SCD. In a cadaveric temporal bone survey, Carey et al\(^1\) reported a 0.7% incidence of
superior SCD and a 1.9% incidence of either dehiscence or significant thinning (<1.0 mm) of bone overlying the superior canal. They hypothesized that although this finding may represent an abnormality in postnatal bone development, there may be a second event that fractures the thin bone or destabilizes the dura over a preexistent dehiscence (closed head injury or increased intracranial pressure) or that there may be erosion of abnormally thin bone caused by the weight and pressure of the temporal fossa. These concepts have been forwarded by other studies in the literature.3,28,29

The pathogenesis of CM has been attributed generally to a neuroectodermal developmental abnormality and more specifically to overcrowding of the hindbrain by an underdeveloped posterior cranial fossa in CM-I.13,14,60 In CM, the CSF space surrounding the cervicomedullary junction is obliterated by the cerebellum, and the arachnoid tissue around the herniated cerebellum is fibrotic. These factors may lead to the development of an associated hydrocephalus. Although hydrocephalus is not typically associated with CM-I, in one series, 5 of 22 (22.2%) asymptomatic or oligosymptomatic patients with a CM-I were found to have hydrocephalus.61 Higher-than-normal CSF pressure can be constant or intermittent.62 It is thought that the partial obstruction of CSF pathways at the foramen magnum blocks the normal efflux and influx of CSF between the head and spine, which compensates for brain expansion and contraction during the cardiac cycle or with Valsalva maneuver.33 This obstruction to the free flow of CSF through the foramen magnum and the exaggerated CSF pressure waves may have a cumulative erosive effect on the surrounding bone. Erosive bony abnormalities of the cranium have been described in CM.43 Scalloping of the skull, also termed craniofacial anomaly, is a common finding in CM-II. In particular, erosion of the posteromedial surface of the temporal bone may occur. This erosion is evident in our patient with an extended CM-I.

Given the low incidence of coexistent hydrocephalus in CM-I, a differential pressure phenomenon within the posterior fossa would not explain the development of posterior SCD in all cases. In fact, it is possible that a developmental bony abnormality involving either the superior or the posterior semicircular canal, or both, may predispose an individual semicircular canal to the eventual development of dehiscence as a result of a second acute (eg, trauma) or chronic (eg, dural pulsation) event. Although, to our knowledge, a causal relationship between CM-I and SCD has not been described in the literature, the fact that neurotologic symptoms tend not to occur until adulthood would favor the assumption that a slow bony erosive process associated with a preexistent developmental bony abnormality may lead to the development of SCD in patients with CM-I.

In conclusion, our study suggests that among patients with SCD, CM-I occurs at a frequency that is substantially higher than the prevalence of CM-I in the general population. This rate of occurrence is especially marked in patients with posterior SCD. While it is not possible to establish a causal relationship, one possible explanation is that altered CSF flow dynamics and increased pressure within the posterior fossa may lead to a slow erosive process. Chronically elevated pressure within the posterior fossa may be at least 1 factor in the pathogenesis of SCD, particularly as it pertains to the posterior semicircular canal. It is also plausible that underlying developmental or acquired factors that lead to CM may also predispose to SCD. Symptoms related to CM and SCD have substantial overlap. Although we did not find a statistically significant difference between the symptoms in patients with SCD alone and those in patients with CM and SCD, it is possible that a larger sample size would reveal such a difference. Despite these findings, it may be premature to recommend that patients with radiographic evidence of SCD undergo MRI of the brain. A larger study may clarify the clinical implications of this association.

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Author Contributions: Dr Kuhn had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Kuhn. Acquisition of data: Kuhn and Clenney. Analysis and interpretation of data: Kuhn and Clenney. Drafting of the manuscript: Kuhn and Clenney. Critical revision of the manuscript for important intellectual content: Kuhn and Clenney. Statistical analysis: Kuhn and Clenney. Administrative, technical, and material support: Kuhn and Clenney. Study supervision: Kuhn.

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REFERENCES


