A Clinical and Histopathologic Study of Jugular Bulb Abnormalities

David R. Friedmann, MD; Jan Eubig, MD; Leon S. Winata, BA; Bidyut K. Pramanik, MD; Saumil N. Merchant, MD; Anil K. Lalwani, MD

Objective: To further define the spectrum of clinical presentation and explore the histologic sequelae of jugular bulb abnormalities (JBAs).

Design: Retrospective review.

Setting: Academic medical center.

Patients: Thirty patients with radiologic evidence of inner ear dehiscence by JBA.

Main Outcome Measure: Thirty patients with radiologic inner ear dehiscence by JBA and 1579 temporal bone specimens were evaluated for consequences from JBA.

Results: We found that JBA-associated inner ear dehiscence could be identified on computed tomography of the temporal bone but not on magnetic resonance imaging scan. Jugular bulb abnormalities eroded the vestibular aqueduct most often (in 25 patients), followed by the facial nerve (5 patients) and the posterior semicircular canal (4 patients). Half of the patients (15) were asymptomatic. Results from vestibular evoked myogenic potential (VEMP) tests were positive in 8 of 12 patients with inner ear dehiscence. Histologically, only 2 of 41 temporal bones with dehiscence of the vestibular aqueduct demonstrated endolymphatic hydrops.

Conclusions: Jugular bulb abnormalities can erode into the vestibular aqueduct, facial nerve, and the posterior semicircular canal. While symptoms may include pulsatile tinnitus, vertigo, or conductive hearing loss, in contrast to earlier reports, half of the patients were asymptomatic. Dehiscence of vestibular aqueduct rarely leads to clinical or histlogic hydrops. The VEMP testing was useful in confirming the presence of inner ear dehiscence due to JBAs. Because the natural history of JBAs is unknown, these patients should be followed closely to evaluate for progression of the JBA or development of symptoms.

of inner ear dehiscence due to JBAs should provide greater
insight into this fascinating disease.

METHODS

PATIENTS

Over 2 years, 30 patients with radiologic evidence of JB-
associated dehiscence involving inner ear structures on high-
resolution computed tomography (HRCT) were identified; the
dehiscence was confirmed by an experienced neuroradiolo-
gist in consultation with a neurotologist (B.K.P. and A.K.L., re-
spectively). The most commonly involved structure was the ves-
tibular aqueduct (25 patients), followed by the facial nerve (5
patients) and posterior semicircular canal (SCC) (4 patients).
Each patient had a detailed otolaryngologic history and exami-
nation, audiologic evaluation, and radiologic imaging. The study
was approved by the institutional review board of New York
University School of Medicine (protocol No. 08-861).

EVALUATION WITH HRCT

All of the patients in this series had undergone HRCT of the
temporal bone. It was performed on 64 multidetector-row com-
tomographic (CT) scanners (Somatom Sensation 64 and
Somatom Definition; Siemens, Erlangen, Germany) using a stan-
dard temporal bone protocol. The acquisition parameters were
120 kV and 250 to 400 mA. Contiguous 0.6-mm scans of the
temporal bone were acquired in the axial plane and reformatted
coronally with a 0.6-mm slice thickness with 0.8-mm incremen-
ts.

AUDIOLOGIC EVALUATION

Audiometry testing was performed in a soundproof booth (In-
dustrial Acoustics Co, Bronx, New York) with the patient wear-
ing TDH 50 headphones (Telephonics Corp, Huntington, New
York) or ER-3A insert earphones (Etymotic Research Inc, Elk
Grove Village, Illinois). A GSI model 16 or 61 Audiometer (Gra-
son-Stadler Inc, Madison, Wisconsin) was used to test air con-
duction (250-8000 Hz) and bone conduction (250-4000 Hz)
thresholds, with appropriate masking as needed. Immittance
testing, including tympanometry and acoustic reflexes (Tymp-
star; Grason-Stadler Inc) as well as Distortion Product Oto-
acoustic Emission (DPOAE) (60 DPOAEs IBM; Grason-
Stadler Inc) testing, was performed.

VESTIBULAR EVOKE

MYOGENIC POTENTIALS

The Navigator Pro Bio-Logic system (Mundelein, Illinois) was
used to record vestibular evoked myogenic potential (VEMP)
responses. Electrode montage was set up on the patient for a
1-channel recording (surface electrodes on the right and left
sternocleidomastoid muscles [SCMs], and the ground elec-
trode was placed on the sternum). Patients assumed a supine
position and were asked to raise and turn their heads as far as
possible toward the contralateral side of the stimulated ear
to activate and contract the ipsilateral SCM muscles.

The vestibulocollic reflex was evoked by air conduction
stimulation via an insert earphone. The stimulus parameters
were as follows: a rarefaction 500-Hz tone burst (rise/fall time=4
milliseconds; no plateau; stimulation rate=4.30 milliseconds).
The analysis time following the tone was 100 milliseconds;
no plateau; stimulation rate=4.30 milliseconds. The signal was bandpass filtered from 30 Hz to 1500 Hz.
First, 2 runs at 95-dB normal hearing level to attempt to abolish the VEMP response. P1 and N1
were marked on the averaged waveform at the higher inten-
sity level. Approximately 200 sweeps were averaged. The VEMP
thresholds were categorized as normal if thresholds were present
only at 95 dB and as low if thresholds were present at 70 dB.
The same procedure was then followed for the affected side.
Analysis included assessing for symmetry between the right and
left sides in addition to measuring thresholds, latencies, and
amplitudes.

TEMPORAL BONE HISTOPATHOLOGIC
CHARACTERISTICS

Because dehiscence of the vestibular aqueduct was most com-
mon radiologically, a study of temporal bone specimens was
undertaken to determine the histopathologic consequences of
vestibular aqueduct dehiscence caused by a JBA. The study ma-
terial consisted of 1805 temporal bones in the Massachusetts
Eye and Ear Infirmary collection that had been sectioned in the
axial (horizontal) plane. The temporal bones were processed
for light microscopy using the standard method consisting of
fixation in formalin, decalcification using trichloroacetic acid
or EDTA, embedding in celloidin, serial sectioning in the hori-
zontal plane at a section thickness of 20 μm, and staining of
every 10th section using hematoxylin-eosin.7 Of the 1805 tem-
poral bones available, a total of 1579 ears were studied; 226
specimens were excluded because of artifact, pathologic le-
sions distorting the bony anatomy, or sections that did not reach
the inferior limit of the IAC. Temporal bone sections were ex-
amined using the light microscope. An HRJB was defined as
one that reached the level of the IAC. For every ear with an
HRJB and vestibular aqueduct dehiscence, all available sec-
tions were examined to look for endolymphatic hydrops. Be-
cause the clinical series had several patients with both JBA-
associated vestibular aqueduct dehiscence and otosclerosis, the
presence of otosclerosis in patients with and without JBA was
also noted. Clinical information was obtained from the medi-
cal records.

RESULTS

RADIOLOGY OF INNER EAR DEHISCENCE

High-resolution computed tomography was more sen-
sitive than magnetic resonance imaging (MRI) scan in
identifying inner ear dehiscence due to JBAs. In this se-
ries of 30 patients, HRCT demonstrated that JBAs most
commonly eroded into the vestibular aqueduct, fol-
lowed by the facial nerve and posterior semicircular ca-
nal (Figures 1, 2, and 3, respectively). An HRJB was
present in almost all cases of dehiscence, while in 16 of
these cases a diverticulum or distinct outpouching from
the JB was also noted. The dehiscence was most often on
the right side (in 20 cases [67%]), whereas bilateral de-
hiscence was noted in 4 cases (13%). Ten patients (33%)
had also undergone MRI scans. Two patients were noted
to have a large JB, whereas inner ear dehiscence was not
suspected or identified on any of the MRI studies.

CLINICAL SYMPTOMS

The clinical presentation, symptoms, and the results of
diagnostic tests are summarized in Table 1. The aver-
Age at presentation was 47 years, with a minimum age of 5 years and a maximum age of 82 years. Of the 30 patients, 18 (60%) were female, and 12 (40%) were male. Presenting symptoms attributable to the inner ear dehiscence included CHL (8 patients), pulsatile tinnitus (5 patients), and vertigo (4 patients). Two patients in our series had histories consistent with endolymphatic hydrops ipsilateral to vestibular aqueduct dehiscence by the JB aqueduct. In 15 patients, the inner ear dehiscence was clinically silent or thought to represent an incidental finding because their presenting symptoms were better explained by other diagnoses. In some of these cases, it was difficult to determine if the symptoms could be attributed in part to the inner ear dehiscence. For example, 3 patients with dehiscence and CHL had bilateral otosclerosis; in these patients, the CHL was attributed to otosclerosis. However, the possibility that the inner ear dehiscence was also contributing to the CHL could not be completely excluded.

Acoustic reflexes were present in all patients with the exception of those with otosclerosis or bilateral profound sensorineural hearing loss. The VEMP findings of reduced thresholds consistent with inner ear dehiscence were present in 8 of the 12 patients who underwent this study.

**Temporal Bone Histopathologic Characteristics**

Of the 1579 temporal bones evaluated, 41 ears showed a dehiscence of the vestibular aqueduct due to an HRJB (Figure 4). However, endolymphatic hydrops was an infrequent consequence of HRJB eroding into the vestibular aqueduct. Only 2 of these 41 cases demonstrated endolymphatic hydrops; 1 of the 2 cases had a clinical history of Ménière’s disease. In comparison, in the 89 ears with an HRJB but without dehiscence of the vestibular aqueduct, 9 ears had hydrops (Table 2), and in 7 of these cases, the patients had Ménière’s disease. The χ² analysis suggests that endolymphatic hydrops is not more common in the presence of JB-mediated dehiscence of the vestibular aqueduct (Fisher exact test; 2-tailed P = .50).

Histologically, otosclerosis was not more common in patients with HRJB (Fisher exact test, 2-tailed P = .62). Of the 130 ears that had an HRJB histopathologically, 19 (14.6%) had otosclerosis. This compares with 242 ears (16.7%) that had otosclerosis of the remaining 1449 ears that did not have an HRJB (Table 3).
Until recently, inner ear dehiscence by JBAs was underappreciated and underreported in both the radiology and otolaryngology literature. To our knowledge, in this report, we present the largest series of patients with inner ear dehiscence due to JBAs identified radiologically and the largest histopathologic study to explore the consequences of the JB impinging on the vestibular aqueduct. In contrast to our previous report in which all patients were symptomatic, we found, in this larger study, that most patients with JBA-associated inner ear dehiscence are asymptomatic. Jugular bulb abnormalities most commonly eroded into the vestibular aqueduct, followed by facial nerve and posterior semicircular canal as detected on HRCT. While CT scan reliably identified the JB-related dehiscence of inner ear structures, MRI did not do so in any of the cases. Two-thirds of the dehiscence was on the right side—the side that normally has the dominant venous system draining the head. This suggests that blood flow dynamics may be important in the etiology of JBA and its associated complications. We also explored the possible association of otosclerosis with JBA. In our clinical series, otosclerosis seemed to be present in a greater than expected number of patients (17%), suggesting that the underlying pathophysiologic mechanisms responsible for otosclerosis may also predispose patients to JBA. However, this hypothesis was not supported by the histopathologic study of temporal bones: there was an equal incidence of otosclerosis in specimens with and without HRJB, suggesting that there is no relationship between the 2.

The vestibular aqueduct was the most common structure made dehiscent by the JB radiologically. Few of these patients displayed a consistent symptom complex. Only 2 of these patients had endolymphatic hydrops clinically. Our clinical findings are similar to the experience of Hourani et al, who found dehiscence between JB and vestibular aqueduct in 11.5% of cases, predominantly on the right side, but did not find a significant correlation between the incidence of dizziness, hearing loss, and the dehiscence. The temporal bone histologic findings in specimens demonstrating dehiscence of the vestibular aqueduct from JBA were consistent with the relative absence of symptoms clinically. Only 2 of 41 temporal bones harboring a dehiscence between the JB and vestibular aqueduct showed histologic evidence of endolymphatic hydrops. This suggests that impingement on the vestibul-

### Table 1. Demographic, Clinical, Auditory, and Radiologic Findings in the 30 Patients in This Study

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</table>

**Abbreviations:** AR, Acoustic reflexes; Asym, asymptomatic; B, bilateral; CHL, conductive hearing loss; FND, facial nerve dehiscence; HL, hearing loss; JB, jugular bulb; JBD, jugular bulb diverticulum; lat, laterality; L, left; nI, normal; MRI, magnetic resonance imaging; nl, normal; NT, not tested; Otos, otosclerosis; PSCD, posterior semicircular canal dehiscence; PT, pulsatile tinnitus; R, right; SNHL, sensorineural hearing loss; V, vertigo; VAD, vestibular aqueduct dehiscence; VEMP, vestibular evoked myogenic potentials; x, present.
The vertical portion of the facial nerve was the second most common structure to be dehiscent from an HRJB. None of the patients with the radiologic finding of facial nerve dehiscence were symptomatic. Vascular compression, pulsation, and irritation of neural structures were well known to cause symptoms such as trigeminal neuralgia or tinnitus. Absence of facial nerve symptoms on its contact with the JB may be due to the lower venous pressure and less intense pulsation within the venous system, suggesting that unlike arterial compression in the cerebellopontine angle, venous compression of this motor nerve is well tolerated.

The posterior SCC was the least common structure to be dehiscent but was most frequently symptomatic. All 4 cases occurred on the right side. The dehiscence of posterior SCC is analogous to superior SCC dehiscence syndrome and thus would be expected to cause a low-frequency CHL (seen in 2 of 4 patients) and a positive VEMP test (in 2 of 2 patients tested). In a recent series of 7 patients with posterior SCC dehiscence due to the JB, 6 occurred on the right side, and all had CHL. In contrast to our earlier study, nearly half of the patients with inner ear dehiscence were asymptomatic. This is not an entirely unexpected finding because a significant number of patients with radiologic and VEMP evidence of superior SCC dehiscence syndrome are similarly clinically asymptomatic. In JBA-associated inner ear dehiscence, the absence of clinical symptoms may reflect the lower pressure within the venous system. Alternatively, dehiscence may be “overcalled” radiologically based on the resolution of cross-sectional imaging. Previously, studies have questioned the specificity of radiologic imaging alone in making the diagnosis of bony dehiscence because CT is insensitive to the presence of thin bone owing to volume averaging. For example, error resulting from volume averaging most likely explains the finding of a higher rate of superior SCC dehiscence radiologically than observed histopathologically. This potential error may be minimized by interpreting dehiscence only when an absence of intervening bone is seen in at least 2 consecutive images and in multiple planes—a strategy used in the current study.

Nonetheless, even with a conservative radiologic analysis strategy, radiologic evidence of inner ear dehiscence should prompt additional testing to provide corroborative evidence for dehiscence. Specifically, in patients with an appropriate clinical history and CT findings, VEMP testing should be used to support the diagnosis of inner ear dehiscence. In our series, the finding of reduced VEMP thresholds consistent with inner ear dehiscence was most useful in attributing patients’ clinical symptoms to the JB finding.

In our series, patients’ symptoms have been managed conservatively, with a plan for observation until more is known about the tendency of these abnormalities to progress. Serial imaging of the patients with JBA-associated dehiscence of vestibular aqueduct and posterior SCC at 3-year intervals is planned to define the natural history of JBA. Surgical approaches have been described for the treatment of JBA to address specific symptoms. One case report describes successful treatment of intractable vertigo by plugging of a dehiscent posterior semicircular canal caused by an HRJB, akin to what has been done in cases of superior SCC dehiscence syndrome. Ligation of the ipsilateral internal jugular vein has also been described for treatment of pulsatile tinnitus secondary to JBA. Clinically significant complications from these interventions have been described, and such treatment must be considered in light of the severity of the patients’ symptoms and only after ruling out other possible etiologies. In our series, patients’ symptoms have

Table 2. Histopathologic Frequency of Endolymphatic Hydrops in Patients With Jugular Bulb (JB) Associated Dehiscence Into the Vestibular Aqueduct*  

<table>
<thead>
<tr>
<th>Presence of Hydrops</th>
<th>JB Dehiscence of Vestibular Aqueduct (41 Ears)</th>
<th>Nondehiscent High-Riding JB (69 Ears)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>2 (4.9)</td>
<td>9 (10.1)</td>
</tr>
<tr>
<td>No</td>
<td>39 (95.1)</td>
<td>80 (89.9)</td>
</tr>
</tbody>
</table>

*In a study of 1579 temporal bones from the Massachusetts Eye and Ear Infirmary collection. Fisher exact test, 2-tailed P = .50.

Table 3. Frequency of Otosclerosis in Patients With Jugular Bulb (JB) Associated Inner Ear Dehiscence Histopathologically*  

<table>
<thead>
<tr>
<th>Presence of Otosclerosis</th>
<th>High-Riding JBs (n = 120)</th>
<th>Normal JBs (n = 1449)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>19 (14.6)</td>
<td>242 (16.7)</td>
</tr>
<tr>
<td>No</td>
<td>111 (85.4)</td>
<td>1207 (83.3)</td>
</tr>
</tbody>
</table>

*In a study of 1579 temporal bones from the Massachusetts Eye and Ear Infirmary collection. Fisher exact test, 2-tailed P = .62.

Figure 4. Histopathologic specimen (hematoxylin-eosin) demonstrating dehiscence between a high-riding jugular bulb and the vestibular aqueduct without evidence of hydrops (arrow). EL sac indicates endolymphatic sac; IAC, internal auditory canal; PSC, posterior semicircular canal; and RM, Reissner membrane.
been managed conservatively, with a plan for observation with serial imaging until more is known about the tendency of these abnormalities to progress.

In conclusion, JBAs may lead to dehiscence of the vestibular aqueduct, facial nerve and posterior SCC that can be detected with CT but not MRI imaging. It can be associated with varying manifestations, including hearing loss, pulsatile tinnitus, and vertigo. Almost half of the patients with inner ear dehiscence demonstrated radiologically were asymptomatic. When identified radiologically, it should be corroborated with a VEMP test. In most patients, symptoms can be managed conservatively with serial imaging.

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Correspondence: Anil K. Lalwani, MD, Department of Otolaryngology, New York University School of Medicine, 550 First Ave, 7Q, New York, NY 10016 (anil.lalwani@nyumc.org).

Author Contributions: Drs Pramanik, Merchant, and Lalwani, the co–senior authors, had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Friedmann, Eubig, Pramanik, and Lalwani. Acquisition of data: Friedmann, Winata, Merchant, and Lalwani. Analysis and interpretation of data: Friedmann, Eubig, Pramanik, Merchant, and Lalwani. Drafting of the manuscript: Friedmann, Winata, Merchant, and Lalwani. Critical revision of the manuscript for important intellectual content: Friedmann, Eubig, Pramanik, and Lalwani. Statistical analysis: Friedmann. Administrative, technical, and material support: Friedmann, Eubig, Winata, Merchant, and Lalwani. Study supervision: Pramanik, Merchant, and Lalwani.

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REFERENCES