Sudden Bilateral Hearing Impairment in Vertebrobasilar Occlusive Disease

Ming-Hung Huang, MD; Chin-Chang Huang, MD; Shan-Jin Ryu, MD; and Nai-Shin Chu, MD, PhD

Background: Bilateral hearing impairment is rare in vertebrobasilar occlusive disease. Summary of Comment: Between 1986 and 1991, we encountered seven patients (four men, three women; median age, 61 years; range, 46–71 years) who had sudden bilateral hearing impairment among 503 patients with vertebrobasilar occlusive disease. The main initial neurological symptoms were sudden bilateral hearing impairment, tinnitus, and vertigo. Acute labyrinthitis or Ménière’s disease was the initial diagnosis until subsequent brain stem or cerebellar signs appeared. Brain stem auditory evoked potentials were abnormal bilaterally in six patients but had unilateral attenuation of the IV-V complex in the remaining one patient. Computed tomographic scans in all six patients showed multiple hypodense lesions in the brain stem and the cerebellum. Cerebral angiography showed complete occlusion on both vertebral arteries in one patient, occlusion on the left with small caliber on the right in another, and severe stenosis on both sides in a third. There was no opacification of internal auditory arteries in these three patients. The remaining patient had arteriosclerotic changes with faint opacification of the bilateral internal auditory arteries. Five patients had a poor prognosis, with locked-in state in four and severe truncal ataxia in one.

Conclusions: We conclude that sudden bilateral hearing impairment in vertebrobasilar occlusive disease is more common than previously recognized and that it may indicate a grave prognosis. (Stroke 1993;24:132–137)

KEY WORDS • cerebrovascular disorders • hearing • vertebrobasilar circulation

Vertebrobasilar occlusive disease (VBOD) often causes impaired consciousness, bilateral pyramidal tract signs, and variable cerebellar, cranial nerve, and other segmental abnormalities of the brain stem.1,2 Some patients may have premonitory attacks, and some may have a monophasic clinical course with rapid progression to deep coma. Therefore, it is important to recognize the early symptoms of VBOD. Although hearing loss has been reported in patients with brain stem stroke,3–4 only a few patients had sudden bilateral hearing impairment (SBHI).3–7 We report seven patients with SBHI in VBOD seen during the past 5 years, four of whom progressed to a locked-in state. Recognition of this early symptom may help lead to correct diagnosis and proper management.

Subjects and Methods

From January 1986 to June 1991, 503 patients with VBOD were admitted to the neurology service at Chang Gung Memorial Hospital. The diagnosis of VBOD was based on the clinical manifestations indicating involvement of the vertebrobasilar system and exclusion of brain stem or cerebellar hemorrhage by computed tomographic (CT) scan. Six of these patients presented with SBHI without disturbed consciousness, and another developed SBHI on day 5. All seven patients were evaluated by the authors. The onset of hearing impairment was acute and completed within 24 hours of the initial auditory disturbance. Hearing acuity was studied by the authors and otologists. Audiometry was performed in three patients. Brain stem auditory evoked potentials (BAEPs) were obtained in all patients within 72 hours of the onset of SBHI; two patients had follow-up studies several months later. The techniques for recording auditory brain stem responses have been previously described.8 Electrodes were attached at the vertex (Cz) and mastoids, with the ipsilateral mastoid serving as the reference and the contralateral mastoid as the ground. Monaural auditory stimuli consisting of rarefaction clicks of 0.1-msec square pulses were delivered through the electrically shielded earphone. The click rate was 10 per second, and the initial intensity was 70 dB. Higher intensities up to 100 dB were used when the BAEP components could not be adequately identified. The far-field electrical activity was amplified by approximately 10^4, with a bandpass of 10–3,000 Hz, and was averaged over 2,000 click presentations for 10 msec. Cranial CT was obtained within 48 hours. If initial CT study was normal, it was repeated between 1 week to 4 months. Four patients had four-vessel
cerebral angiography when clinically stable. All patients had been followed for 8 months to 4 years.

**Results**

**Clinical Manifestations**

Seven of 503 patients with VBOD had SBHI, giving an incidence of approximately 1.4%. Table 1 summarizes the clinical data of these seven patients. There were four men and three women, with ages ranging from 46 to 71 years (median, 61 years). The occurrence of sudden hearing impairment was almost simultaneous on both sides except for one (patient 1), who first developed sudden hearing impairment on the right side and 4 hours later on the left. In addition to SBHI, four patients presented with vertigo and vomiting. Six patients also had bilateral tinnitus, and two had dizziness; other symptoms included dysarthria, hemifacial palsy, ataxia, hemiparesis, hemianesthesia, and quadripareisis. Acute labyrinthitis or Ménière’s disease was initially diagnosed in four patients (patients 1, 2, 3, and 5). Subsequently, all but one (patient 7) developed other brain stem or cerebellar signs, usually on days 2–7. After hearing acuity began to improve on day 3, patient 3 developed quadripareisis on day 4 and progressed to quadriplegia on day 15. In patient 6, mild left hemiparesis and hemisensory impairment were noted on the first day, and SBHI, tinnitus, diplopia, and dysarthria 5 days later. Hypertension was present in four patients and diabetes in two. Old stroke was found on CT in two (patients 2 and 3). Patient 4 had chronic atrial fibrillation. Two patients (patients 6 and 7) did not have any risk factors for stroke.

**Auditory Function**

Five patients (patients 1–5) had severe hearing loss that caused communication disability. The hearing impairment of patients 6 and 7 was mild and resulted in some difficulty in communication (Table 1). Audiometry performed within 10 days after onset in three patients (patients 3, 5, and 7) was compatible with the clinical auditory status.

**Brain Stem Auditory Evoked Potential Findings**

Figure 1 shows BAEP findings. In patient 1, initial BAEPs disclosed a total absence of all waveforms (Figure 1, tracing 1a). Fourteen months later, waves III and V on the left appeared to be present, although BAEPs on the right were still absent (Figure 1, tracing 1b). In patient 2, the BAEPs failed to show any waveforms in the acute stage (Figure 1, tracing 2) and 6 months later. Patient 3 demonstrated an absence of waves I and III and a prolonged wave V on both sides (Figure 1, tracing 3). Patient 4 showed an absence of BAEP waves, except for possible marked attenuation of waves III and V on the left (Figure 1, tracing 4). The BAEPs of patient 5 exhibited abolilition of all recognizable waves, except for a normal wave I on the left side (Figure 1, tracing 5). In patient 6, no recognizable waves were noted from either ear stimulation on day 6 (Figure 1, tracing 6). In patient 7, there was a normal BAEP on the right and possible attenuation of wave IV-V complex, with a decreased V/I voltage ratio on the left (Figure 1, tracing 7). Thus, six patients had abnormal BAEPs, with a possibly abnormal BAEP in the remaining one.

**Computed Tomographic Findings**

All seven patients underwent serial CT studies. Figure 2 illustrates multiple low density lesions in the cerebellar hemispheres and upper pons on day 8 and 14 months later in patient 1. The CT scans of patient 2 showed multiple low density lesions in the pons and the left cerebellar hemisphere (Figure 3). Three other patients (patients 3–5) had multiple hypodense lesions in the cerebellar hemispheres or cerebellar peduncles. Patient 6 showed a right pontine low density on day 2. Patients 2 and 3 also had old infarcts in the frontoparietal areas and left putamen, respectively. The remaining patient (patient 7) failed to reveal any abnormality on CT.

**Cerebral Angiographic Findings**

Cerebral angiograms were obtained in four patients (patients 1, 2, 5, and 7). Patient 1 had an occlusion of the left vertebral artery and a small and irregular caliber of the right vertebral artery. The internal auditory
arteries (IAAs) were not visualized on either sides. Marked arteriosclerosis with multiple segmental narrowing over intracranial vessels was also noted. There was a fetal origin of the posterior communicating arteries, with partially retrograde perfusion to the basilar artery (Figure 4). Patient 2 also showed a complete occlusion of bilateral vertebral arteries. However, a collateral circulation from a muscular branch of the left extracranial vessels to the basilar artery was found by the left subclavian artery injection. There was no opacification of bilateral IAAs (Figure 5). In patient 5, there were severe stenosis of bilateral vertebral arteries and complete obliteration of the basilar artery. Bilateral IAAs were not visualized, although anterior inferior cerebellar arteries were opacified. Posterior cerebral arteries and superior cerebellar arteries were perfused by the carotid circulation. Patient 7 showed a mild arteriosclerosis of the vertebral arteries. There was a faint opacification of IAAs on both sides.

**Hearing Recovery and Neurological Outcome**

Table 2 summarizes the clinical outcome and its correlations with BAEP, CT, and cerebral angiography. All seven patients had slight to complete hearing recovery within 1 month. Hearing acuity was almost completely recovered in two patients (patients 6 and 7) and incompletely in the remaining five. The recovery of hearing occurred within 10 days in five patients and between 15 days and 1 month in two.

**FIGURE 1.** Tracings show brain stem auditory evoked potentials (BAEPs) of patients 1–7. Tracing 1: Initial BAEPs showed absence of all waveforms (a); follow-up BAEPs showed possibly attenuated waves III and V on the left (L) but absence of all waves on the right (R) (b). Tracing 2: BAEPs failed to show any waveforms. Tracing 3: An absence of waves I and III but a prolonged wave V on both sides. Tracing 4: An absence of waves except for possible attenuated waves III and V on the left. Tracing 5: BAEPs revealed abolition of all recognizable waves except a normal wave I on the left. Tracing 6: Recognizable waves were absent from either ear stimulation. Tracing 7: BAEPs were normal on the right; wave IV-V complex was attenuated with a decreased VII voltage ratio on the left.
FIGURE 2. Computed tomographic scans of patient 1 show multiple low density lesions in the pons and cerebellar hemispheres on day 8 (panel A) and brain stem and cerebellar atrophy 14 months later (panel B). r, Right.

Four patients received anticoagulation therapy after development of subsequent brain stem dysfunction. Three remained in a locked-in state, and one had severe truncal ataxia. Three patients (patients 1, 6, and 7) received antiplatelet therapy with aspirin (150–300 mg daily). Patient 6 had mild left hemiparesis, and patient 7 recovered completely and returned to work in the following year. The neurological status of these seven patients remained unchanged during the follow-up periods ranging from 8 months to 4 years, with a mean of 1 year and 5 months.

Discussion

Hearing impairment may occur, though it is uncommon, in patients with VBOD. The pathogenesis of this clinical setting may be an ischemic nature of the auditory pathways secondary to thromboembolic occlusion of the vertebrobasilar artery or its branches. A few reports identified pathological changes consistent with occlusion of the IAA,9,10 which supplies the cochlear nerve and the cochlea. In addition to the cochlea, involvement of the cochlear nucleus7 or lesions that interrupted ascending and descending auditory pathway in the pons and midbrain11,12 was also reported to result in hearing loss. In our series, the bilateral auditory pathways may be damaged first, and then the brain stem was involved. These diffuse lesions may account for the high frequency of poor outcome.

Although animal studies have revealed that the cochlea has a low tolerance to ischemia,13,14 hearing impairment is not a frequent symptom of VBOD, occurring in only one of 653 and 10 of 1124 patients. However, the IAA may have variable origins and anastomoses.15,16 When the proximal IAA is occluded, the cochlea is less involved because adequate collateral circulation may develop.17 According to the findings of Sohmer et al,18 when the cerebral perfusion pressure is decreased, blood flow to the cochlea is still sufficient

FIGURE 3. Computed tomographic scans of patient 2 show bilateral low density lesions in the upper pons (panel A) and low density lesions in the mid-pons and left cerebellar hemisphere (panel B). r, Right.
because intracochlear pressure is usually lower than intracranial pressure. This allows preferential blood flow in the larger intracranial arteries to supply the cochlea.

In certain clinical conditions, hearing impairment may be underestimated. Elderly patients may attribute their mild sudden hearing impairment to presbycusis. Hearing impairment is difficult to evaluate in patients with altered consciousness. If major neurological deficits progress rapidly, particularly to death, hearing impairment tends to be neglected. Nevertheless, Stefan et al. reported that one patient with basilar artery occlusion presented with bilateral hearing loss, dysarthria, and hyperventilation, then died 10 hours later.

Because four of seven patients in this series were in the locked-in state, SBHI in vertebrobasilar occlusive stroke seems to foretell a grave prognosis. As it is unusual that auditory pathways are bilaterally involved with sparing other structures of the brain stem and cerebellum, SBHI is frequently presumed to be accom-

Figure 4. Cerebral angiography of patient 1 shows complete occlusion of the left vertebral artery (panels A and B, arrow), small caliber of the right vertebral artery after right innominate artery injection (panel C, arrowheads), and marked arteriosclerosis of the intracranial vessels and an infantile type of the posterior communicating artery with partial retrograde perfusion to the basilar artery (panel D, arrowheads) after right common carotid artery injection.

Figure 5. Cerebral angiography of patient 2 shows collateral circulation from a muscular branch of left extracranial vessels (arrows) to basilar artery system after left subclavian artery injection (panels A and B), faint right vertebral artery (arrows) after aortic arch injection (panel C), and marked arteriosclerosis of the intracranial vessels and posterior cerebral artery perfused by the carotid system (panel D).
TABLE 2. Correlation Among Clinical Outcome, Brain Stem Auditory Evoked Potential, Computed Tomographic, and Cerebral Angiographic Findings

<table>
<thead>
<tr>
<th>Patient</th>
<th>Hearing recovery</th>
<th>Neurological outcome</th>
<th>BAEP (R/L)</th>
<th>Hypodense in CT</th>
<th>Cerebral angiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Incomplete</td>
<td>Locked-in</td>
<td>I</td>
<td>-/-</td>
<td>Occlusion of L vertebral artery, small caliber of R vertebral artery, occlusion of bil IAA</td>
</tr>
<tr>
<td>2</td>
<td>Incomplete</td>
<td>Locked-in</td>
<td>II</td>
<td>-/-</td>
<td>Bil midbrain, pons, frontoparietal areas</td>
</tr>
<tr>
<td>3</td>
<td>Incomplete</td>
<td>Locked-in</td>
<td>III</td>
<td>+/+</td>
<td>Bil cerebellar peduncles, L putamen</td>
</tr>
<tr>
<td>4</td>
<td>Incomplete</td>
<td>Locked-in</td>
<td>IV</td>
<td>-/A</td>
<td>Bil cerebellar hemispheres</td>
</tr>
<tr>
<td>5</td>
<td>Incomplete</td>
<td>Truncal ataxia</td>
<td>V</td>
<td>-/-</td>
<td>R cerebellar hemisphere, L cerebellar peduncle</td>
</tr>
<tr>
<td>6</td>
<td>Complete</td>
<td>L hemiparesis</td>
<td>I</td>
<td>-/-</td>
<td>R pons</td>
</tr>
<tr>
<td>7</td>
<td>Complete</td>
<td>Recovery</td>
<td>II</td>
<td>N/N</td>
<td>Normal</td>
</tr>
</tbody>
</table>

BAEP, brain stem auditory evoked potential; R, right; L, left; CT, computed tomography; -, absence of peak; bil, bilateral; IAA, internal auditory artery; +, increased latency; A, attenuated amplitude; N, normal latency; AICA, anterior inferior cerebellar artery.

panned by other severe neurological deficits. Thus, bilateral hearing involvement due to ischemia of the cochlea and possibly other auditory pathways was presumed to reflect a severe degree of hypoperfusion of the vertebrobasilar artery territories.

In basilar artery thrombosis, an absence of BAEP or a delayed small wave V with absence of earlier waves indicates peripheral sensorineural hearing loss that was likely due to the involvement of IAA, 19 but a preexisting bilateral hearing loss must be excluded. Six of our patients (11 ears) had this abnormality on BAEP. However, other BAEP abnormalities in patients 5 and 7 indicated involvement of the brain stem.

Hearing impairment alone or associated with vestibular symptoms may be the presenting symptom of acute labyrinthitis, Ménière’s disease, and VBOD. In our study, SBHI occurred before (three patients), during (three patients), or after (one patient) other symptoms and signs of VBOD. Therefore, the possibility of VBOD should be considered when clinicians are confronted with a patient, especially the elderly, presenting with sudden hearing impairment and no prior history of hearing or vestibular dysfunction. We suggest that these patients should be admitted and observed for possible progression of vertebrobasilar insufficiency. Furthermore, a high proportion of patients with SBHI and vertigo developed devastating vertebrobasilar ischemia that evolved on a delayed basis (2–7 days). Early anticoagulation may prevent the delayed progression, 20 particularly in patients with severe SBHI.

References

Sudden bilateral hearing impairment in vertebrobasilar occlusive disease.

M H Huang, C C Huang, S J Ryu and N S Chu

*Stroke*. 1993;24:132-137
doi: 10.1161/01.STR.24.1.132

*Stroke* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1993 American Heart Association, Inc. All rights reserved.
Print ISSN: 0039-2499. Online ISSN: 1524-4628

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://stroke.ahajournals.org/content/24/1/132

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in *Stroke* can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to *Stroke* is online at:
http://stroke.ahajournals.org//subscriptions/