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Segmental Enhancement of the Cochlea on Contrast-Enhanced MR: Correlation with the Frequency of Hearing Loss and Possible Sign of Perilymphatic Fistula and Autoimmune Labyrinthitis

Alexander S. Mark¹ and Dennis Fitzgerald²

PURPOSE: To relate the finding of selective enhancement of different turns of the cochlea to the frequency range of the hearing loss measured by audiogram. **METHODS:** Six patients aged 23 to 53 years, four men and two women, who presented with sudden hearing loss and had segmental enhancement of different turns of the cochlea on contrast-enhanced MR imaging were included in this retrospective study. The patients were imaged on a 1.5 T MR imaging system using pre- and postcontrast axial T1-weighted images and postcontrast coronal T1-weighted images through the temporal bone. **RESULTS:** The basal turn of the cochlea enhanced selectively in five of the six patients and the apical turn enhanced in the sixth patient. All patients had sensorineural hearing loss. Three of the patients with basal turn enhancement had predominantly high-frequency hearing loss, whereas the patient with apical turn enhancement had predominantly low-frequency hearing loss. The two other patients with basal turn enhancement had complete hearing loss. Three patients had presumed autoimmune labyrinthitis. The other three patients had strong clinical and surgical evidence of a perilymphatic fistula. **CONCLUSION:** Contrast-enhanced MR imaging may demonstrate selective enhancement of different turns of the cochlea. In certain patients, the areas of enhancement may correlate with specific hearing loss in the frequencies mapped by the enhancing segment. Enhancement of the cochlea may represent radiologic evidence of cochlear inflammation secondary to a perilymphatic fistula.

Index terms: Ear, magnetic resonance; Hearing, loss; Fistula

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Several recent reports (1-3) have described enhancement of the membranous labyrinth on gadolinium-enhanced magnetic resonance (MR) imaging. This enhancement has been demonstrated to be a specific radiographic sign of a severe inner ear abnormality often associated with loss of cochlear and/or labyrinthine function both subjectively and on audiologic and vestibular testing (1, 2). The cause of enhancement is usually an infectious process (viral, bacterial, luetic) (1, 2) or posttraumatic inflammation (3). Intense enhancement may also be seen in patients with vestibular schwannomas (4, 5). On MR different

turns of the cochlea can be identified individually. We describe five patients with selective enhancement of different turns of the cochlea and relate this finding to the frequency range of the hearing loss measured by the audiogram. We also discuss possible etiologies of this phenomenon.

Materials and Methods

Six patients aged 23 to 53 years, four men and two women, who presented with sudden hearing loss and had segmental enhancement of the different turns of the cochlea on contrast-enhanced MR imaging were included in this retrospective study. Four of the patients were included in a previous report (2). All patients were imaged on a 1.5 T MR scanner. The imaging protocol included sagittal T1-weighted images (300/15 (TR/TE) 5 mm thick, number of signals average (NSA) was 1, 256 × 192 matrix) for location, precontrast axial T1-weighted images (400/11, 3 mm thick with a 0.2-mm intersection gap and a 20-cm field of view with 2 NSA through the posterior fossa), postcontrast axial and coronal T1-weighted images (600/13, 14-cm field of view, 256 × 192 matrix, 4 NSA), and

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postcontrast axial proton density and T2-weighted images (2000/30 and 80, field of view 20, 0.75 NSA, 5 mm thick with a 2.5-mm intersection gap). The purpose of the precontrast T1-weighted axial images is to detect any high-intensity lesion such as an intralabyrinthine hemorrhage or a lipoma. T2-weighted images through the whole brain are obtained to exclude an intraaxial lesion such as multiple sclerosis or an infarct, which may present occasionally with sudden sensorineural hearing loss. Because the patient's head was often tilted and the right, and left labyrinth did not appear on the same image, the illustrations are presented as composite right and left images to allow immediate comparison between the right and left labyrinth. A similar windowing was used for each half of the image.

The patients' clinical histories and audiologic examinations were reviewed with particular attention to the range of frequency of the hearing loss and to any clinical history of trauma or recent viral infection.

Results

The patients' clinical and radiographic findings are summarized in Tables 1 and 2.

Five patients demonstrated enhancement of the basal turn of the cochlea (Figs. 1 and 2) and one patient had enhancement of the apical turn of the cochlea (Fig. 3). Three of the patients with enhancement of the basal turn of the cochlea had predominantly high-frequency hearing loss. The patient with enhancement of the apical turn had predominantly low-frequency hearing loss. The two other patients with enhancement of the basal turn had complete hearing loss. Three of the patients had clinical and surgical evidence of a perilymphatic fistula. The other three patients had presumed autoimmune inner ear disease.

Discussion

Enhancement of the membranous inner ear has been described recently and correlated with both subjective and objective loss of cochlear and vestibular function (1–3). This finding proved to be a highly characteristic finding of inner ear pathology. However, its sensitivity remains to be determined. There is clearly a threshold effect and only the more severe cases of hearing loss and/or vertigo result in enhancement. Milder cases of inner ear disorders may not reach the threshold and may not show enhancement with this technique.

The cause of enhancement is usually an infectious process, most often viral, even though specific viruses are rarely detected. Syphilitic and bacterial inner ear infections have also been reported as a cause of inner ear enhancement.

Traumatic lesions may also result in inner ear enhancement or hemorrhage (3), even in the absence of a demonstrable temporal bone fracture. Intralabyrinthine schwannomas produce intense enhancement that does not change over time, contrary to the inflammatory enhancement which usually resolves over several months with or without improvement in the patient's hearing (3–5).

Enhancement of the cochlea and vestibule needs to be separated from pericochlear and perivestibular enhancement seen in patients with active otospongiotic plaque (3). The differentiation is easy on high resolution studies demonstrating no enhancement in the membranous labyrinth but linear foci of perilabyrinthine enhancement in patients with otospongiosis. A precontrast study should always precede the contrast examination in order to exclude labyrinthine hemorrhage (6).

Each segment of the cochlea maps specific hearing frequencies, with progressively higher frequencies from the apical to the basal turn. In four of our patients, there was a relationship between the location of the enhancement and the range of frequency of hearing loss. However, in two of the patients with basal turn enhancement, there was complete hearing loss. This may be due to the above-mentioned threshold effect in which milder abnormalities were present in the rest of the cochlea sufficient to cause hearing loss in that frequency range but not enough to cause enhancement. Compromise of the vascular supply to the apical turn by the abnormality resulting in enhancement of the basal turn may also explain the lack of enhancement in the apex of the cochlea. It is also possible that the process affected the cochlear nerve itself, suppressing the signals from the entire cochlea, thus explaining the complete hearing loss.

The etiology of the enhancement in these six patients seems somewhat different from the reported patients who had enhancement of the entire cochlea. Three of our patients had clinical and surgical findings suggesting a perilymphatic fistula. Perilymphatic fistulas result from a tear in the round window or membrane of the ligamentous attachment of the stapedial footplate to the oval window resulting in leakage of perilymph into the middle ear (7–9). The etiology of this condition is not fully understood, but it is most often associated with head or direct ear trauma or sudden change in barometric pressure such as during takeoff and landing in airplanes or diving

TABLE 1: Clinical findings

Patient	Age	Sex	Clinical History ^a	Diagnosis
1	50	F	Sudden left SNHL. 3 years before had mild left SNHL after airplane ride. Positive fistula test.	Perilymphatic fistula. Surgery: perilymph leak at round window. Severe inflammatory reaction round window niche.
2	52	M	Profound right SNHL. Severe left SNHL 7 years before. No history of viral illness. No trauma. Elevated sedimentation rate. Borderline positive lymphocyte transformation test against inner ear membrane protein. Negative VDRL.	Presumed autoimmune labyrinthitis. Cochlear implantation planned.
3	53	M	Right SNHL after barotrauma from descent in an airplane. Negative fistula test.	Perilymphatic fistula. Surgery: perilymph leak at the round window. Slight improvement of hearing after surgery.
4	30	F	Left SNHL. No history of viral illness. VDRL negative. Positive lymphocyte transformation test against inner ear membrane protein.	Presumed autoimmune labyrinthitis.
5	30	M	Right-sided sudden SNHL after weight lifting.	Perilymphatic fistula. Improved postsurgery. Spontaneous recurrence 4 months later. Improved after second surgery and remained stable for the past year.
6	23	M	Bilateral hearing loss. Bilateral joint effusions. Elevated sedimentation rate.	Presumed autoimmune labyrinthitis.

^a Abbreviations used in table: SNHL, sensorineural hearing loss; VDRL, Venereal Disease Research Laboratory Test.

TABLE 2: Radiographic findings

Patient	Frequency of Hearing Loss	Cochlear Enhancement	Vestibular Enhancement
1	High	Basal turn	Yes
2	Total (bilateral)	Basal turn	No
3	Low	Apical turn	No
4	High	Basal turn	No
5	Total	Basal turn	No
6	High (bilateral)	Basal turn (bilateral)	Yes

(9). Sudden increases in intracranial pressures may also cause perilymphatic fistulas.

Experimental evidence in guinea pigs (10) exposed to barotrauma in a pressure chamber indicates that barotrauma can induce injuries to the cochlea and membranous labyrinth. There was a tendency for the damage to be more pronounced in the basal turn rather than in the apical turn (10). Inner ear hemorrhage occurred more frequently in the cochlea than in the semicircular

canal. Blood was commonly found in the scala tympani of the basal turn close to the round window. Rupture of the round window membrane was also seen in some animals. There was a tendency for the inner ear hemorrhage to be more severe in ears with rupture of the round window membrane.

Another author reported damage to the inner ear hair cells due to barotrauma in guinea pigs and also stated that the basal turn sustained more

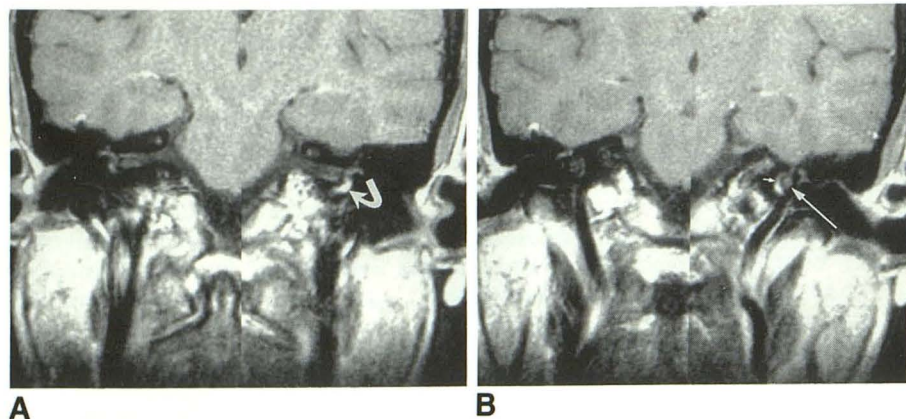


Fig. 1. A 50-year-old woman (patient 1) with left sensorineural hearing loss.

A, Composite coronal T1-weighted image (600/13) postcontrast through the basal turn. Notice marked enhancement of the basal turn of the left cochlea (arrow).

B, Composite coronal T1-weighted image (600/13) through the apical turn of the cochlea (3 mm anterior to Fig. 1A). No enhancement is noted in the apical turn (large arrow). The cutoff between the enhancing segment and the nonenhancing segment is clearly demonstrated (small arrow).

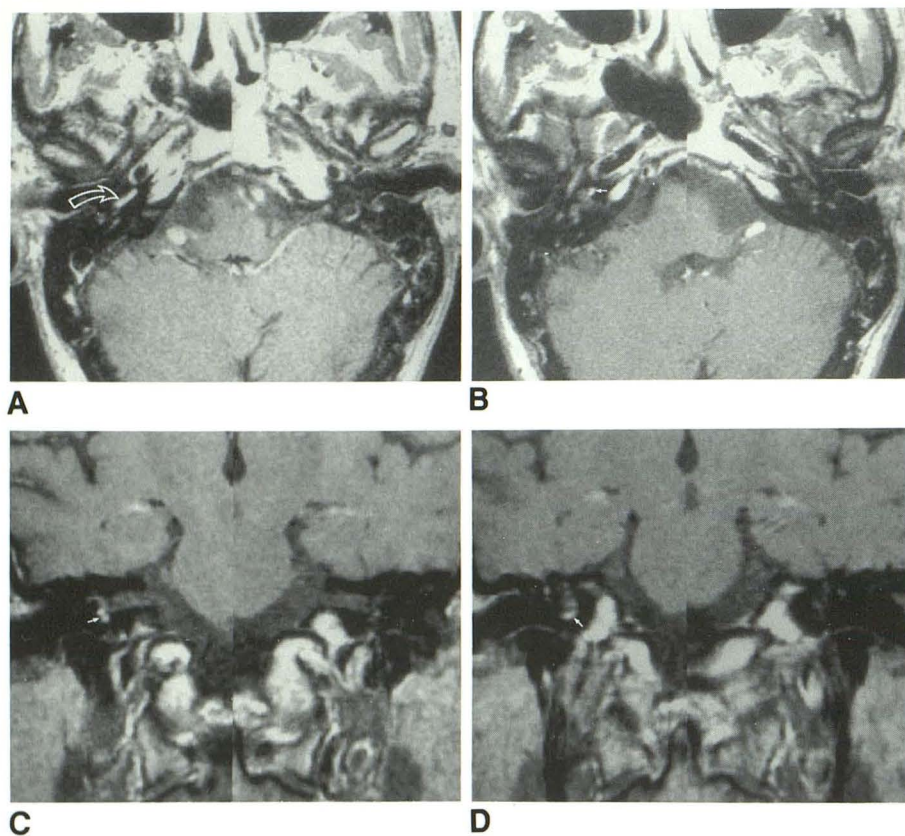


Fig. 2. A 52-year-old man (patient 2) with profound acute right sensorineural hearing loss. He had had acute left sensorineural hearing loss 7 years earlier without any improvement.

A and B, Composite axial postcontrast T1-weighted images (600/13) through the basal turn of the cochlea demonstrate marked enhancement of the basal turn (arrows). The apical turn does not enhance.

C and D, Consecutive composite coronal postcontrast T1-weighted (600/13) images through the cochlea confirm the basal turn enhancement (arrows).

damage than the apical turn in most cases (11, 12). However, occasionally the apical turn was the one most severely damaged. The proposed origin of the hemorrhage is the rupture of the blood vessels of the round window membrane. The variability of the auditory and vestibular dysfunction in round window membrane ruptures may be caused by variations in the lesions inside the inner ear rather than differences in the extent of the rupture (10).

This experimental data may explain our observations. The lack of visible hemorrhage on the

precontrast study may be explained by the small amount of blood present, which may be below that able to be detected by this technique or, more likely, by the delay between the occurrence of the trauma and the imaging. The predilection of barotrauma to damage of the basal turn of the cochlea described in experimental studies may explain the pattern of enhancement we describe. Two of our three patients with labyrinthine fistula had enhancement in the basal turn. The enhancement most likely represents the posttraumatic disruption of the tight junctions of the labyrinthine

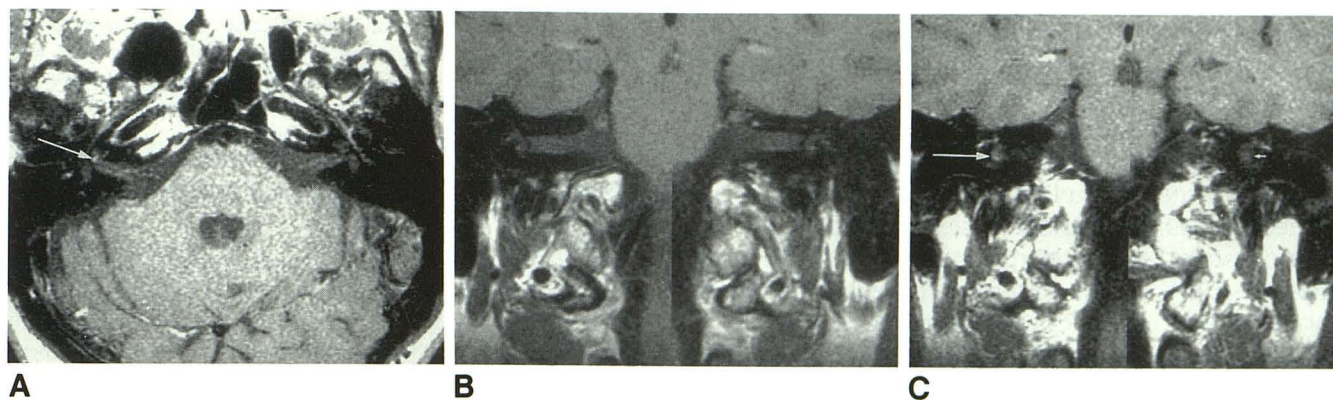


Fig. 3. A 53-year-old man (patient 3) with low-frequency hearing loss.

A, Postcontrast axial T1-weighted image (600/13). Notice a small focus of enhancement in the apical turn of the cochlea (*arrow*).

B, Composite coronal T1-weighted image (600/13) through the basal turn of the cochlea. No enhancement is seen.

C, Composite coronal T1-weighted image (600/13) through the apical turn of the cochlea (3 mm anterior to Fig. 1C). Notice enhancement of the right apical turn of the cochlea (*large arrow*). The apical turn of the left cochlea (*small arrow*) does not enhance.

vessels secondary to trauma with secondary leakage of the contrast agent.

Thus, while the rupture of the round window or oval window cannot be directly imaged, contrast-enhanced MR may be able to demonstrate the secondary changes of the trauma leading to the perilymphatic fistula. We believe these secondary inflammatory changes, which may occur in the cochlea secondary to the fistula, may take several days to develop; thus, one would anticipate seeing enhancement within the first week following the symptoms. However, it is also likely that only the most severe inflammatory changes will result in enhancement and patients with milder degrees of inflammation may show no abnormalities on contrast-enhanced MR.

Since the oval window is involved twice as often as the round window in patients with perilymphatic fistula, one may wonder whether enhancement of the vestibule can be seen in this condition. We have encountered a patient who developed sensorineural hearing loss immediately following head trauma; the precontrast MR performed shortly after the accident excluded labyrinthine hemorrhage. The postcontrast study demonstrated vestibular enhancement. Patient 1 in our series also exhibited vestibular enhancement on the axial images (not illustrated here). Thus, based on these preliminary observations, one should add the diagnosis of perilymphatic fistula to the list of differential diagnoses for vestibular enhancement. We have seen enhancement of the semicircular canals in addition to vestibular enhancement in two patients with labyrinthitis, but not in patients with perilymphatic fistulae.

The diagnosis of perilymphatic fistula is difficult. The fistula test consisting of mechanically increasing the middle ear pressure and looking for nystagmus or vertigo is not always positive. Since often there is no active leaking of perilymph at the time of surgery due to the intermittent nature of this disorder and the very small amounts of perilymph (microliters) involved, the diagnosis may be difficult even at the time of surgery. In the past, imaging of patients with perilymphatic fistulae both by computed tomography and MR has been disappointing. Experimental studies with MR following subarachnoid injection of gadolinium-DTPA with surgically produced perilymphatic fistulae have shown leakage of the gadolinium in the middle ear (M. Morris, personal communication, 1992). However, currently gadolinium-DTPA is not approved by the Food and Drug Administration for subarachnoid injection. Since the entire notion of perilymphatic fistulae is somewhat controversial, one can understand that management of this condition is also variable from institution to institution. In our hospital, if surgical intervention is considered, it is usually not performed until at least 7 days after the acute event.

Sudden hearing loss has recently been attributed to autoimmune inner ear disease (13). Evidence of autoimmunity in these patients includes a positive lymphocyte inhibition test, a gross test of autoimmunity in which prepared inner ear antigens are used to challenge the patient's leukocytes. Other evidence of autoimmunity includes substantial hearing improvement achieved from steroid therapy and histopathologic evidence of vasculitis in one case. The lack of other

explanations for the hearing loss in patients 2, 4, and 6, the high sedimentation rate in patient 6, as well as a borderline positive lymphocyte transformation test against pooled inner ear membrane protein in patients 2 and 4, suggests autoimmune inner ear disease as a likely cause for the findings in these cases. With these cases in mind, perilymphatic fistula and autoimmune inner ear disease must be part of the differential diagnosis when enhancement of the cochlea is found.

Selective enhancement of different turns of the cochlea may be demonstrated by high resolution contrast-enhanced MR. All these patients had sensorineural hearing loss on the enhancing side. In certain patients, the frequency range of the hearing loss corresponded to the range of frequency mapped by the enhancing segment. Enhancement of the cochlea may be a radiographic finding suggesting a perilymphatic fistula.

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