The presence of the vestibulocochlear nerve (VCN) appears to be the foundation for successful cochlear implantation in patients with sensorineural hearing loss (SNHL). The identification of an intact cochlear nerve is crucial for cochlear implant candidates [1]. High resolution computed tomography (HRCT) of the temporal bone may help in the preoperative planning of cochlear implant surgery by providing information about the anatomy of the cochlear nerve and its relationship with the bony canal. The purpose of this study is to evaluate the significance of the hypoplastic canal for the cochlear nerve in patients with sensorineural hearing loss (SNHL) and the relationship between the hypoplastic bony canal and aplasia or hypoplasia of the cochlear nerve.

Materials and Methods: A retrospective review of high resolution temporal CT (HRCT) and MRI findings was conducted. The narrow bony canal of the cochlear nerve and the relative size of the internal auditory canal were correlated with the cochlear nerve deficiency on MRI. The comparative size of the component nerves (facial, cochlear, superior vestibular, inferior vestibular nerve), and the relative size of the internal auditory canal and the bony canal of the cochlear nerve were measured. The clinical history and the results of the clinical examination were reviewed for each patient.

Results: High resolution MRI showed aplasia of the common vestibulocochlear nerve in one patient and a deficiency of the cochlear nerve in 9 patients. These abnormalities occurred in association with a prominent narrowing of the canal for the cochlear nerve and a stenosis of the internal auditory canal, which was observed on temporal bone CT in 9 patients with congenital SNHL. Three patients had normal IAC, despite the presence of a hypoplastic cochlear nerve on the side on which they had SNHL. In one patient, the narrowing of the canal for the cochlear nerve and internal auditory canal were not found to be associated with acquired SNHL.

Conclusion: The hypoplastic bony canal for the cochlear nerve might be more highly indicative of congenital cochlear nerve deficiency than that of the narrow internal auditory canal, and the position of the crista falciformis should also be carefully.

Index words: Temporal bone
Temporal bone, abnormalities
Temporal bone, MR
Temporal bone, CT
bone remains the method of choice for the evaluation of congenital ear malformation, because it presents information about the middle and external ear and can define the bony labyrinth with the internal auditory canal (IAC) [2]. The association between a narrow IAC and hearing loss has been reported [3]. Aplasia and hypoplasia of the cochlear nerve have been known to occur concomitantly with a very narrow IAC. However, the correlation between the diameter of the IAC and the presence or absence of the vestibulocochlear nerve is disputed. Therefore the visualization of the vestibulocochlear nerve (VCN) with high spatial resolution MR (HRMR) imaging is necessary.

Fatterpekar et al. [4] showed that the length and width of the bony canal for the cochlear nerve were significantly smaller in patients with SNHL than in a control group, and suggested that the hypoplastic bony canal for the cochlear nerve might be an indicator of embryologic malformation of the cochlear nerve. However, they did not compare the CT findings with MR imaging findings to confirm the status of the cochlear nerve. The purpose of this study was to identify the specific imaging findings of HRCT and HRMR of the temporal bone in patients for whom T2-weighted MR imaging showed a deficiency of the VCN as the possible cause of congenital SNHL. The correlations between the aplasia or hypoplasia of the cochlear nerve and the narrow IAC or hypoplastic bony canal for the cochlear nerve in the axial and coronal images were evaluated.

Materials and Methods

We reviewed 10 patients (7 females and 3 males) aged from 5 to 38 years (mean age, 11.7 years) with SNHL, who had undergone temporal bone CT and MR imaging. The clinical history and the results of the clinical examination were reviewed for each patient. CT examinations were composed of axial and coronal, 0.63 mm-thickness, contiguous sections through the temporal bone. The plane of the transverse images was parallel to the infraorbital meatal line, and the coronal images were obtained perpendicular to the transverse plane. The images were reconstructed by using a high spatial-resolution bone algorithm with individual magnification for the right and left temporal bones. The matrix size was 512×512 with a 9.6 cm field of view. We used a window level and width settings of 400/4000 Hounsfield units. An evaluation of the inner ear and related structures, the internal auditory canal, and the bony canal for the cochlear nerve were made by measuring the width, in millimeters, of the bony canal for the cochlear nerve at the fundus of the internal acoustic meatus in the axial plane. The width of the bony canal for the cochlear nerve was obtained at its midportion by drawing a line from the base of the modiolus to the inner margin of the fundus of the internal acoustic meatus [4] (Fig. 1). Consecutive measurements were obtained using calipers and the mean values were calculated for both sides to the nearest 0.1 mm. We measured the vertical diameter of the internal auditory canal at the level of the fundus, followed by of the lower compartment of the IAC [Fig. 2]. The ratio of the lower compartment to that of the IAC was calculated on both the side affected with SNHL and the normal side to see if there was a difference. The widths of the bony canal on both the affected sides and the normal sides were analyzed using the t-test to see if it there was a statistically significant difference. A P value of less than 0.05 indicated statistical significance. The upper and lower compartment of the fundus of the IAC is divided by a transverse crest of bone,

Fig. 1. The width of the bony canal for the cochlear nerve was obtained at its midportion by drawing a line from the base of the modiolus to the inner margin of the fundus of the internal acoustic meatus.
called the crista falciformis and, normally, the upper compartment occupies about 40% and the lower about 60% of the vertical dimension of the canal. This ratio can be used as a significant indicator of a hypoplastic or aplastic cochlear nerve, if the ratio is less than 0.5 (50%). We designated the bony canal for the cochlear nerve as hypoplastic, if its width was asymmetrically smaller than that on the contralateral side. If the position of the crista falciformis lies below the mid-point, it should be considered abnormal (5). We also evaluated the size and shape of the IAC. The IAC was designated as abnormal if it was <3 mm in either the vertical or transverse diameter, if it was irregularly shaped, or if it was appreciably smaller than that on the contralateral side (5, 6).

The MR images were evaluated for the presence and relative size of the nerves in the IAC, with correlations being made of the size of the IAC, as shown on temporal bone CT for the same patients. We evaluated the nerves from the mid to lateral aspect of the IAC after the separation of the three divisions of the VCN. In the parasagittal plane, the caliber of the cochlear nerve was compared with that of the facial nerve, the superior and inferior vestibular nerves, and the contralateral cochlear nerve (Fig. 3). The cochlear nerve was defined as hypoplastic or deficient when it appeared smaller in size compared to the other nerves of the IAC and as aplastic when it could not be seen on the axial and parasagittal images, irrespective of whether the nerve was imperceptibly small or truly absent. The inner ear structures (cochlea, vestibule, modiolus, semicircular canals and endolymphatic duct and sac) were evaluated for abnormalities in contour, size and signal intensity. The clinical history and audiometry findings were reviewed. The parameters used for the MR images were as follows: 3D T2-weighted turbo spin-echo MR imaging for the axial images 4000/250/1 (TR/TE/NEX); TSE fact, 57; matrix, 256×256; field of view, 14 cm; slice thickness, 0.7 mm. The acquisition time was 6 minutes 4 seconds. The parasagittal images were obtained through the IAC perpendicular to its axial plane, by using the following parameters: 4500/250/1; TSE fact, 57; matrix, 256×256; field of view, 14 cm; slice thickness, 0.5 mm. The acquisition time was 6 minutes 4 seconds. The parasagittal images were obtained as a direct parasagittal sequence using parameters similar to those of the axial 3D sequence.

Results

The CT images of 6 patients with SNHL showed nar-
rowing of the diameter of the IAC, while 4 patients had an IAC with a normal diameter. The canal size for the cochlear nerve was conspicuously reduced on the same side as the SNHL in 9 patients, while in the remaining patient, who was suspected of having acquired SNHL after suffering from a febrile disease of unknown origin during childhood, the canal size was normal. Details of the clinical presentation of the patients and the imaging data are listed in Table 1. The mean size of the IAC for all 10 patients on the side of the SNHL was 4.18 mm, while that on the contralateral normal side was 5.01 mm, which implies that the reduction in the mean size

during childhood, the canal size was normal. Details of the clinical presentation of the patients and the imaging data are listed in Table 1. The mean size of the IAC for all 10 patients on the side of the SNHL was 4.18 mm, while that on the contralateral normal side was 5.01 mm, which implies that the reduction in the mean size

Table 1. Clinical and Imaging Data of Patients with Sensorineural Hearing Loss

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex/Age</th>
<th>Significant medical history</th>
<th>History</th>
<th>CT canal size IAC (R/L)</th>
<th>BCCN (R/L)</th>
<th>MR findings of inner ear and nerves</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>7/M</td>
<td>None</td>
<td>Right side HL of unknown duration</td>
<td>N/N</td>
<td>Narrow/N</td>
<td>Cochlear nerve hypoplasia</td>
</tr>
<tr>
<td>2</td>
<td>5/F</td>
<td>Prematurity IUP 35weeks</td>
<td>Left side HL since neonatal</td>
<td>N/Narrow</td>
<td>N/Narrow</td>
<td>Cochlear nerve aplasia</td>
</tr>
<tr>
<td>3</td>
<td>9/F</td>
<td>None</td>
<td>Right side HL since several years</td>
<td>Narrow/N</td>
<td>Narrow/N</td>
<td>Cochlear nerve aplasia</td>
</tr>
<tr>
<td>4</td>
<td>7/M</td>
<td>None</td>
<td>Left side HL of unknown duration</td>
<td>N/Narrow</td>
<td>N/Narrow</td>
<td>Cochlear nerve aplasia</td>
</tr>
<tr>
<td>5</td>
<td>8/M</td>
<td>None</td>
<td>Right side HL for 1 month</td>
<td>N/N</td>
<td>Narrow/N</td>
<td>Cochlear nerve hypoplasia</td>
</tr>
<tr>
<td>6</td>
<td>13/F</td>
<td>None</td>
<td>Left side HL of unknown duration</td>
<td>N/Narrow</td>
<td>N/Narrow</td>
<td>Cochlear nerve aplasia</td>
</tr>
<tr>
<td>7</td>
<td>7/F</td>
<td>Down syndrome</td>
<td>Right side HL since Birth</td>
<td>Narrow/N</td>
<td>Narrow/N</td>
<td>Cochlear nerve hypoplasia</td>
</tr>
<tr>
<td>8</td>
<td>6/F</td>
<td>None</td>
<td>Right HL of unknown duration</td>
<td>N/N</td>
<td>Narrow/N</td>
<td>Cochlear nerve hypoplasia</td>
</tr>
<tr>
<td>9</td>
<td>17/F</td>
<td>None</td>
<td>Right HL of unknown duration</td>
<td>Narrow/N</td>
<td>Narrow/N</td>
<td>Cochlear nerve aplasia</td>
</tr>
<tr>
<td>10</td>
<td>38/F</td>
<td>Febrile disease of unknown cause during childhood</td>
<td>Both since childhood</td>
<td>N/N</td>
<td>N/N</td>
<td>Cochlear nerve deficiency</td>
</tr>
</tbody>
</table>

Note.-  R: right, L: left, N: normal, HL: hearing loss, IAC: internal auditory canal, BCCN: bony canal for cochlear nerve

Fig. 4. Cochlear nerve aplasia in a 17-year-old patient with right sensorineural hearing loss.

A. Axial HRCT scan of the temporal bone through the midportion of the modiolus shows a conspicuous hypoplastic bony canal for the right cochlear nerve (arrows). The width of the canal was obtained by measuring the distance between the two arrows.

B. Contralateral normal side shows a comparatively large bony canal for the left cochlear nerve (arrows).

C. Aplasia of the VCN on the right side on axial T2-weighted fast spin echo MR image is noted. Only one nerve, the right facial nerve, can be visualized. Contralateral normal side shows two nerves (facial and vestibulocochlear nerves) of normal size.
of the IAC on the side of the SNHL compared with that on the contralateral normal side was not statistically significant. The mean width of the canal for the cochlear nerve on the normal side was 1.82 mm, whereas that on the contralateral side with SNHL was 0.54 mm. The difference in the average width between the side affected with SNHL and the contralateral normal side was statistically significant ($p < 0.05$). The coronal image of the temporal bone CT also showed a reduced width in the lower compartment, which corresponds to the anterior cochlear area. The mean ratio of the lower compartment to the fundus of the IAC was about 34% ipsilateral to the congenital SNHL. All patients had normal-appearing ossicles and a normally formed middle ear cavity.

Submillimetric 3D T2-weighted turbo spin-echo images showed aplasia of the cochlear nerve in 5 patients and hypoplasia of the cochlear nerve on the side with SNHL in 4 patients. In two patients, we had difficulty differentiating between aplasia and hypoplasia of the cochlear nerve, because it was very small and was lying against the anterior or inferior wall near the fundus of the IAC. No associated inner ear abnormalities were observed with MR and CT imaging in our patients, except for a large semicircular canal and vestibular dysplasia in one patient. In one patient with congenital right side hypoplasia of the cochlear nerve, whose bony canal for the cochlear nerve appeared markedly narrow on HRCT, the cochlear nerve was hardly visible on parasagittal MRI (Fig. 5C). Another patient, who showed a conspicuous hypoplastic bony canal for the cochlear nerve, showed a hypoplastic cochlear nerve on MRI (Fig. 4A). One patient, who had acquired bilateral SNHL, did not have a narrow IAC or hypoplasia of the bony canal for the cochlear nerve, but showed bilateral cochlear nerve deficiency on the MRI study. Only the facial nerve, inferior vestibular branch and superior branch of the VCN were well depicted on the axial and parasagittal turbo-spin echo T2WI for this patient (Fig. 6).

**Discussion**

SNHL results from a dysfunction of the cochlea, the VCN or the central auditory pathways. Short scan times, good bony details and easy availability have made CT the method of choice for the evaluation of congenital deafness. However, HRCT is not able to show in detail...
the membranous labyrinth and branches of the VCN. The diameter of the IAC is not always correlated with the presence or absence of the VCN. For this reason, Casselman et al. (7) suggested obtaining submillimetric T2-weighted gradient-echo images in all cochlear implant patients. Thanks to the latest developments in MR imaging techniques, cochlear nerve deficiency can be detected in congenital SNHL cases using high resolution MRI. It is essential to use images with a thickness of 1 mm or less, in order to individually depict the facial nerve and the three branches of the VCN inside the IAC. The value of the parasagittal plane is superior to that of the axial plane in depicting the size and relationship of the facial nerve and VCN, because in general these nerves run medially laterally within the IAC. The cochlear and vestibular nerves usually separate 3-4 mm from the lateral end of the IAC (8). In a study by Casselman et al., the cochlear nerves were found to be larger than either the superior or inferior vestibular nerve in 90% of cases, and were of similar size or larger than the facial nerve in 64% of cases. The relative size of the four nerves were symmetrical with the contralateral IAC in 70% of the cases (9) (Fig. 3). Aplasia of the complete VCN and aplasia or hypoplasia/deficiency of its cochlear branch were demonstrated on MR images in all 10 patients of this study. These abnormalities oc-

Fig. 5-2. same patient
A. A normal sized right inferior vestibular branch (Vin) can be seen, but the right cochlear nerve is hypoplastic (white arrow). On the left side, both the cochlear and Vin branches are normal in size (white arrow head).
B. Parasagittal 3D spin echo image shows a hardly visible cochlear nerve (Cn) (white arrow), but the facial nerve (Fn) and the vestibular branches (Vsn) are well visualized in the middle third of the internal auditory canal.
C. On the left side, a normal facial nerve and a normal sized cochlear nerve and dividing vestibular branches can be well visualized. The cochlear nerve appears larger than the facial nerve.
D. Parasagittal 3D spin echo image obtained at the level of the lateral third of the IAC. The cochlear nerve lying against the anterior or inferior wall of the narrow IAC is hardly visible (white arrow).
E. On the left side at the level of the fundus, the IAC is wider. High signal intensity cerebrospinal fluid delineates the four nerves. In the anterior aspect of the canal, the facial nerve (FN) lies superiorly to the cochlear nerve (CN). The superior and inferior vestibular nerve lie posteriorly.
curred in association with a stenosis of the IAC in 9 patients. The MR findings in all 10 patients with aplasia or hypoplasia/deficiency of the cochlear branch of the VCN were in accordance with the clinical findings.

In one patient with congenital right side hypoplasia of the cochlear nerve, who had a markedly narrow bony canal for the cochlear nerve, the cochlear nerve was scarcely visible on parasagittal MRI (Fig. 5C white arrow). However, one 38 year old patient, who had an acquired SNHL, and whose bony canal for the cochlear nerve exhibited normal calibers, had a bilaterally hypoplastic cochlear nerve (Fig. 6C, D white arrows).

Before interpreting the imaging findings associated with congenital SNHL, it is crucial to understand the embryology of the inner ear and IAC. Labyrinthine development starts at about 3 weeks' gestation with the formation of the otic placode. This otic placode subsequently invaginates and forms an otic vesicle. During

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**Fig. 6.** Cochlear nerve deficiency in a 38-year-old patient with hearing loss after otitis media during childhood.
A. Axial T2-weighted fast spin echo MR image of both sides shows a small calibered cochlear nerve (arrow heads).
B. Axial T2-weighted fast spin echo MR image of both sides at upper level shows a normal calibered facial and superior vestibular nerve in a normal sized IAC.
C, D. Corresponding direct parasagittal view shows the facial nerve, superior vestibular nerve and inferior vestibular nerve at the level of the lateral third of the IAC, but the small calibered cochlear nerve is faintly visualized (arrow).
FN indicates facial nerve, Vsn, superior vestibular nerve, Vin, inferior vestibular nerve, CN, cochlear nerve, Cvn, common vestibular nerve, VCN, vestibulocochlear nerve.
the first 7 weeks, the spiral organ of Corti develops from the cochlear duct. The fibers from the spiral ganglion form the cochlear nerve of the VCN. The simultaneous development of the ampulla and the vestibular ganglia leads to the formation of the vestibular branches of the VCN. At approximately 9 weeks’ gestation, the mesenchyme surrounding the otic vesicle begins to chondrify and forms the otic capsule that subsequently ossifies (10). The IAC is formed by the inhibition of cartilage formation at the medial aspect of the otic vesicle. This inhibition requires the presence of the VCN nerve. In the absence of this nerve, no canal will be formed, because the formation of the IAC is dependent on the inhibition of capsule chondrogenesis by the VCN root (11). Not only does the VCN allow the formation of the IAC, but the survival and promotion of this nerve seems to require the presence of a nerve growth factor from the otic vesicle (12). In all of the patients with congenital SNHL there was a narrowing of the diameter of the IAC and conspicuous reduction of the canal for the cochlear nerve on the same side as the SNHL. In this study, in the submillimetric 3D T2-weighted turbo spin-echo images, the aplasia or hypoplasia of the cochlear nerve was clearly demonstrated in 9 patients ipsilateral to the SNHL, but no associated inner ear abnormality was unequivocally observed with MR and CT imaging. The only inner ear abnormality observed was a small modiolus in four cases. Only one patient showed a lateral semicircular canal-vestibular dysplasia (LSCV) of the left inner ear. Fatterpekar et al. (4) found that the hypoplasia of the bony canal for the cochlear nerve was an isolated finding with a normal appearing cochlear. In the studies presented by Casselman et al. (7) and Glastonbury et al. (6), the abnormality of the inner ear structures was normal or subtle in MR imaging, despite the abnormality of the nerve caliber. They also postulated that the cochlear abnormality might be microscopic or that there might have been a direct insult to the nerve before the IAC completely formed. The normal range of vertical diameters for the IAC vary from 2 to 8 mm, with the average being 4 mm. Canals smaller than 2 or 3 mm are generally considered stenotic (1, 5). Shelton et al. (3) stated that a very narrow IAC (1 to 2 mm in diameter), as seen on HRCT of the temporal bone in patients with congenital, profound SNHL, is a contraindication to a cochlear implant. However, according to several reports, stenotic IAC is not always correlated with the presence or absence of the VCN (2). There was a case report of a child who showed a narrow IAC, but who nevertheless was given a cochlear implant and for whom the postoperative results were encouraging (13). By obtaining submillimetric T2-weighted gradient-echo images, Casselman et al. identified 10 patients with aplasia or hypoplasia of the VCN. The cochlear nerve was absent in 1 ear for 3 patients with stenosis of the IAC, while the cochlear nerve was absent but the vestibular nerves were present in those patients with a normal IAC. Therefore, they concluded that the relationship of IAC stenosis/ataresia to congenital SNHL was unclear. Nelson et al. (14) also suggested that cochlear nerve aplasia can occur in both a narrow or a normal-sized IAC. However, Glastonbury et al. (6) stated that hypoplasia of the IAC is an indicator of congenital cochlear nerve deficiency because, in their study, a marked reduction in caliber of the IAC ipsilateral to the deficient cochlear nerve was evident in 11 out of 12 patients with congenital SNHL. Our 9 patients with congenital SNHL showed considerable narrowing in width. Hypoplasia of the canal for the cochlear nerve of the IAC was not seen in those patients whose cochlear nerve was of normal caliber. We did not find any reports which described a relationship between a normal caliber cochlear nerve shown by MRI and a hypoplastic canal for the cochlear nerve or a markedly narrow IAC. Therefore, we consider that the presence of a hypoplastic canal for the cochlear nerve is correlated with aplasia or hypoplasia of the cochlear nerve.

A major advance in CT technology was achieved with the introduction of multislice spiral CT. Multislice spiral CT with submillimeter spatial resolution enables the delineation of subtle anatomical structures and pathological changes of the inner ear and middle ear. The bony canal of the labyrinthine segment of the facial nerve and the cochlear nerve can be delineated from the branches of the vestibular nerve on images acquired in the axial and coronal planes. Fatterpekar et al. (4) showed that the length and width of the bony canal for the cochlear nerve were significantly smaller in patients with SNHL than in a control group, and suggested that the presence of a hypoplastic bony canal for the cochlear nerve may be indicative of an embryologic malformation of the cochlear nerve. In our 9 patients with congenital SNHL, the mean width of the bony canal for the cochlear nerve of the normal ear was 1.82 mm, whereas the mean width for those ears with SNHL was 0.54 mm. The lower compartment of the fundus of the IAC occupies ap-
proximately 34% ipsilateral to the congenital SNHL. In our cases, the narrowing of the bony canal for the cochlear nerve appeared more obvious than the narrowing of the IAC. The lower compartment, which corresponds to the anterior cochlear area showed a marked reduction [Fig. 5C white arrow]. This finding suggests that the narrowing of the IAC is more related to a reduction in the size of the anterior cochlear area than to that of the [internal auditory canal/posterior cochlear area?]. Therefore, the measurement of the bony canal for the cochlear nerve is more sensitive in detecting cochlear nerve deficiency than that of the internal auditory canal. For patients who need to undergo further evaluation with MR imaging, high resolution T2-weighted, fast spin-echo MR imaging may help evaluate the status of the cochlear nerve and would also be helpful in documenting the size of the bony canal for the cochlear nerve.

The limitations of this study include the small number of patients and the difficulty in differentiating between aplasia and hypoplasia of the cochlear nerve in those cases in which the nerve was lying against the anterior or inferior wall of the IAC. To the best of our knowledge, although there have been previous articles which described the presence of a narrowed IAC in relation to aplasia or hypoplasia of the cochlear nerve canal, hypoplasia of the bony canal for the cochlear nerve and narrowing of the anterior cochlear area have not previously been described or emphasized.

In conclusion, a hypoplastic bony canal for the cochlear nerve may be indicative of a congenital cochlear nerve deficiency and needs to be carefully observed in patients with congenital sensorineural hearing loss. The measurement of the bony canal for the cochlear nerve is more sensitive to the detection of cochlear nerve deficiency than that of the internal auditory canal. The position of the crista falciformis should also be carefully observed in the coronal image of the HRCT.

References

Yoon Jung Choi, et al. The Significance of a Hypoplastic Bony Canal for the Cochlear Nerve in Patients with Sensorineural Hearing Loss

CT와 MRI

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이상의 결과를 바탕으로, 이 연구는 보다 정확한 진단과 치료를 위한 중요한 기여를 하였습니다.

결과:

100명의 환자 중 90명은 CT 및 MRI를 통해 진단을 받았습니다. 이 중 90%의 환자가 전방호와 후방호를 동시에 비후하였고, 90%의 환자가 전방호와 후방호를 동시에 비후하였습니다.

이상의 결과를 바탕으로, 이 연구는 보다 정확한 진단과 치료를 위한 중요한 기여를 하였습니다.