Purpose: To evaluate the CT and MRI findings of the large endolymphatic duct or sac syndrome (LEDS) and its associated anomalies, with clinical features.

Materials and Methods: We retrospectively reviewed the MR and CT images of 52 ears obtained from 26 patients with LEDS. We reviewed the clinical findings, audiology testing, and treatment results. The degree of hearing loss was classified from normal to profound, based on pure tone audiometry. The largest areas were measured at each endolymphatic duct and analyzed to determine whether a correlation exists with the degree of hearing loss. We also analyzed the differences in measurements between CT and MRI findings.

Results: All 26 patients had some degree of sensorineural hearing loss, which resulted in 18 ears to undergo a cochlear implantation. One patient was diagnosed with Cornelia de Lange syndrome. Five patients had a sudden hearing loss onset. Ten ears had incomplete cochlear partitions, whereas 28 ears had enlarged vestibules. All patients had severe to profound hearing loss. We found no statistical correlation between the size of the largest area of the endolymphatic duct and the degree of hearing loss. The mean area of the endolymphatic ducts, as per an MRI examination, revealed slightly greater areas than the CT findings, although the differences were not significant.

Conclusion: Enlarged vestibules and incomplete partitions of the cochlea were common anomalies associated with LEDS. We found no statistical correlation between the largest area of the endolymphatic duct or sac with the degree of hearing loss.

Index words: Computed tomography (CT)
Magnetic resonance (MR)
Ear, inner
Endolymphatic duct
Hearing loss
Large endolymphatic duct and sac (LEDS) syndrome is an inner ear malformation which manifests itself as progressive sensorineural hearing loss (SNHL), which begins during infancy or childhood (1). LEDS is recognized as one of the most common morphologic findings associated with congenital SNHL as well as the most commonly identified radiologic anomaly on cross-sectional imaging studies of the inner ear (2, 3). Thin-section T2-weighted MR imaging takes advantage of the intrinsic fluid contrast of the membranous labyrinth to yield clear inner ear images that can be used to assess the membranous labyrinth in patients with LEDS (4). The enlargement of the endolymphatic duct and sac corresponds to an enlargement of the vestibular aqueduct. LEDS was diagnosed when the diameter of the endolymphatic sac exceeded that of the posterior semicircular canal (Fig. 1A) or when the width of the vestibular aqueduct, at the midpoint between the common crus and its external aperture, was larger than 1.5 mm (5-8).

The purpose of this study was to evaluate the CT and MR imaging findings of LEDS and its associated anomalies with clinical findings.

**Materials and Methods**

We retrospectively reviewed the MRI or CT images of 52 ears, belonging to 26 patients (12 men and 14 women; mean age, 13.3 years) with LEDS. Fifteen patients had both MRI and CT scans performed. Three patients only had an MRI, whereas eight patients only underwent a CT scan. Of the 15 patients that underwent both MRI and CT scans, three CT scans were excluded due to poor image quality. The CT scans were performed with a spiral CT (GE, Hispeed CT/I pro / Milwaukee, U.S.A.) and a 16-row MDCT (Siemens, Somatom Sensation 16 / Erlangen, Germany) at a 1 mm slice thickness. All the CT scans were viewed at a bone window setting. Three-dimensional T2-FSE images were obtained by 1.5 T (GE SIGNA / Milwaukee, U.S.A.) at a 1 mm thickness and FIESTA images by 3.0 T (GE, SIGNA HDx / Milwaukee, U.S.A.) at a 0.77 mm thickness.

The largest areas of endolymphatic ducts and sacs were obtained by the ROI measurement using PiViewSTAR (Ver. 5.0, INFINITT / Seoul, Korea) (Fig. 1B). Previous studies have reported on the volume measurements of the endolymphatic duct and sac (9-11). The volume measurement is a more accurate parameter for comparing the endolymphatic ducts and sacs of different sizes. However, the measurement of the largest area is more practical, because it takes less time to determine. Experimentally, we measured the whole area of the endolymphatic ducts and sacs of eight patients. An example of one of these results is illustrated in Fig 2. as a bar graph. All of the graphs showed a “bell-shaped” pattern. This pattern indicates that the largest area is easily obtained by taking three to five measurements. If we intuitively measured the true largest area on the first mea-

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**Fig. 1.** Normal endolymphatic ducts and LEDS.  
A. Three-year-old female with normal inner ear structures.  
An axial FIESTA image shows a normal endolymphatic duct (arrow) which was smaller than the posterior semicircular canal (arrowhead)  
B. Two-year-old male with LEDS  
An axial FIESTA image shows an enlarged endolymphatic duct and sac with the ROI measurement (dotted line) and display of the measured area
measurement, the additional two measurements are needed on the previous image section and subsequent section to confirm that the first measurement was the largest or not. Using this method, we measured the largest areas of the LEDS in all patients in a short time. The degree of hearing loss was classified as normal (10–25 dB), mild (26–40 dB), moderate (41–55 dB), moderate to severe (56–70 dB), severe (71–90 dB), and profound (more than 91 dB) based on pure tone audiometry.

We tested the relationship between the largest areas of the LEDS measured by MR and CT scanning, with the degree of hearing loss using Pearson’s correlation (SPSS Ver. 14.0 for Windows, \( p < 0.05 \)). In addition, we analyzed the difference in the measurements between the CT and MRI scans using the Wilcoxon signed rank test and a paired t-test. Furthermore, we reviewed the clinical findings, audiology tests, and treatments.

**Results**

All patients had unilateral or bilateral SNHL. Twelve patients underwent a speech-language evaluation and showed decreased language ability due to the decreased hearing. Eighteen ears underwent a cochlear implantation. Four ears with LEDS also had mastoiditis. One patient was diagnosed with the Cornelia de Lange syndrome and had mental retardation as well as the characteristic facial appearance (Fig. 3). Five patients had a definite history of trauma before developing or aggravating their hearing loss. Theses clinical data and associated anomalies are listed on Table 1. All patients had severe (11 ears) or profound (41 ears) hearing loss (Table 2). Enlarged vestibules were observed in 28/52 (53.8%) ears (Fig. 3C, D, 4A). An incomplete partition was present in

| Table 1. Clinical Data and Associated Anomalies in 52 Ears from 26 Individuals |
|---------------------------------|-----------------|---------|
| SNHL                                           | 52 [5*]       |
| Mastoiditis                                     | 4              |
| Cochlear implantation                           | 18             |
| Enlarged vestibule                              | 28             |
| Incomplete partition                            | 10 [2**]      |
| SNHL: Sensorineural hearing loss                 |
| *Sudden onset hearing loss                       |
| **Cornelia De Lange syndrome                    |

| Table 2. Degree of Hearing Loss |
|-------------------------------|-------|
| Degree of hearing loss   | [dB] | N   |
| Normal                  | 10–25 | 0   |
| Mild                     | 26–40 | 0   |
| Moderate                 | 41–55 | 0   |
| Moderate to severe       | 56–70 | 0   |
| Severe                   | 71–90 | 11  |
| Profound                 | >91   | 41  |

![Image](image_url)

**Fig. 2.** The bar graph presents the measured areas of the endolymphatic duct and sac according to the image section (number) in one patient. This graph shows a “bell shaped” pattern, allowing the largest area to be easily obtained.
Fig. 3. A five-year-old male with LEDS and incomplete partition type I. He presented with the typical facial appearance and mental retardation, compatible with Cornelia de Lange syndrome.

A, B. Axial MR images of right (A) and left (B) inner ears show a common cavity formation of the cochlea (arrows).

C, D. Axial MR images of right (C) and left (D) inner ears show enlarged vestibules bilaterally (arrowheads) and right LEDS (curved arrow).

E. A plain AP radiograph of the right hand shows decreased bone age (36 M, 29–45 M). This finding is common in Cornelia de Lange syndrome.
10/52 (19.2%) ears with LEDS (Fig. 3A, B, 4B). No statistically significant correlations were found between the largest areas of the LEDS measured by MRI and CT scans and the degree of hearing loss (Fig. 5). The mean area obtained by MR imaging was slightly larger than the mean area obtained by the CT scan in 22 ears that had both forms of imaging. However, no significant differ-

<table>
<thead>
<tr>
<th>Degree of Hearing Loss</th>
<th>Area in CT (mm²)</th>
<th>Area in MR (mm²)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe (71-90)</td>
<td>17.42</td>
<td>20.57</td>
</tr>
<tr>
<td>Profound (&gt;91)</td>
<td>20.92</td>
<td>23.29</td>
</tr>
<tr>
<td>Mean</td>
<td>19.81</td>
<td>22.42</td>
</tr>
</tbody>
</table>

Table 3. The Mean Areas Measured in the CT and MRI Results of 22 Ears with Severe to Profound Hearing Loss

Fig. 4. Ten-year-old female with LEDS diagnosed as an enlarged vestibule and incomplete partition type II
A. Axial MR image shows an enlarged vestibule (arrowhead) and endolymphatic duct and sac (curved arrow)
B. An axial MR image showing a cystic appearance of the apical and middle turns of the cochlea (arrow), which are compatible with incomplete partitioning.

Fig. 5. Scatter plots with Pearson’s correlation coefficients of the largest areas, along with MRI and CT scanning results and the degree of hearing loss in dBHL
A. The degree of hearing loss by pure tone audiometry is increased as the area measured on MRI is increased ($r = 0.213$) without a statistically significant difference ($p = 0.226$).
B. The degree of hearing loss by pure tone audiometry is decreased as the area measured on the CT scan is increased ($r = 0.042$), although the difference is not statistically significant difference ($p = 0.800$)
ference was found between the means obtained by MRI and CT scans (Wilcoxon signed rank test \( p = 0.200 \); paired t-test \( p = 0.080 \)) (Table 3).

**Discussion**

LEDS has been identified as the most common morphologic finding associated with SNHL as well as the most commonly identified radiological anomaly on cross-sectional imaging studies of the inner ear [2, 3]. The normal endolymphatic duct originates in the anteromedial wall of the vestibule, coursing posterolaterally in the bony vestibular aqueduct, and merging with the endolymphatic sac while remaining within the bony canal. The endolymphatic sac then emerges from the bony vestibular aqueduct in a shallow impression on the posterior face of the petrous temporal bone. The function of the endolymphatic duct and sac is not well understood. Endolymph produced in the cochlea travels from the inner ear to the endolymphatic sac. The endolymphatic sac functions as a metabolically active filter to regulate the volume and composition of the endolymphatic fluid. The loss of function of the endolymphatic sac or abnormalities in this region has been implicated in Meniere’s disease [4]. The mean diameters of the endolymphatic duct, 0.16 to 0.41 mm at the internal aperture of the vestibular aqueduct and 0.09 to 0.20 mm at the isthmus, are below the resolution of current resolution limits of MR imaging. The corresponding measurements of the vestibular aqueduct, 0.32 to 0.72 and 0.18 to 0.31 mm, also challenge the resolution limits of current CT scanning [9]. Thin-section T2-weighted MR imaging takes advantage of the intrinsic fluid contrast of the membranous labyrinth to yield clear inner ear images that can be used to assess the membranous labyrinth in patients with LEDS [3].

LEDS is commonly associated with other cochlear and vestibular abnormalities [3]. The recognition of the associated anomalies in LEDS is critical to the imaging assessment of these patients, since the presence of other abnormalities, especially of the cochlea, will affect the patient prognosis and therapeutic planning [3]. In this study, various anomalies such as enlarged vestibules and incomplete partitions were diagnosed according to the new classification [12]. One patient was diagnosed with Cornelia de Lange syndrome, which is a rare genetic syndrome with an incidence of 1/10,000- to 1/60,000 newborns. Cornelia de Lange syndrome is characterized by delays in growth and development, hirsutism, structural anomalies of the limbs, and distinctive facial characteristics [13, 14]. Limited information exists on Cornelia de Lange syndrome associated with temporal bone anomalies. However, the pathophysiology of LEDS or other inner ear anomalies are not well recognized as part of this syndrome [14, 15].

Deformity of the LEDS is congenital; however, the hearing loss accompanying this syndrome is acquired [1]. A distinct feature of Cornelia de Lange syndrome is that the hearing loss associated with LEDS syndrome is triggered by minor head trauma [1]. In this study, five patients had a definite history of trauma before developing or aggravating their hearing loss.

In previous studies, various techniques were used to measure the LEDS. None of the prior measurements showed a significant correlation with the degree of hearing loss [1, 10, 11]. It is unlikely that the hearing loss associated with the LEDS is caused by transmission of subarachnoid pressure forces into the labyrinth through a deficient modiolus [10]. We measured the largest area of the LEDS by a simple method. Because the pattern of the areas, based on the imaging, were “bell shaped”, we could measure the largest area of the LEDS with three or four measurements in each patient. However, we observed no statistically significant relationship between the LEDS with hearing loss in this study. The measurements by CT scan and MRI can differ due to the differences in imaging techniques. For the CT scans, the margins of the LEDS were delineated by bone. In contrast, the MRI showed strong fluid signal intensity with LEDS, therefore the posterior extension of the endolymphatic sac as well as the interface between the sac and the adjacent cerebellum were more clearly visualized. Our measurements showed that the largest mean area in the MRI was slightly larger than in the CT scan; however, these differences were not statistically significant.

In this study, 18/52 (34.6%) of the ears tested had cochlear implantations without significant complications. The appropriate diagnosis of the LEDS by CT scanning or MRI allow for improved patient management.

High resolution MRI and CT scanning were effective for the evaluation of LEDS with associated anomalies. Enlarged vestibules and incomplete partitioning of the cochlea were commonly associated the anomalies observed with cases of LEDS. No significant statistical relationship was found between the largest area of endolymphatic duct and the degree of hearing loss. The correct imaging diagnosis of LEDS improves patient
management leading to cochlear implantation where indicated.

References
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