INTRODUCTION: Large vestibular aqueduct syndrome, or enlarged endolymphatic duct (EED) and sac (EES) syndrome, is an inner ear malformation which manifests itself as progressive sensorineural hearing loss starting in infancy or childhood. The deformity of the endolymphatic duct and sac is congenital; however, the hearing loss that develops in this syndrome is acquired. A characteristic feature is that hearing loss related to the large endolymphatic duct and sac is triggered by minor head trauma. Although, the true etiology of the hearing loss observed in this syndrome remains unknown, a variety of pathophysiological theories have been proposed. To investigate the pathophysiology of the progressive sensorineural hearing loss in EED and EES syndrome, the volumes of the EED and EES, the diameter of the EED, the area of the cochlear modiolus, and the signal intensities of the EED and EES were measured by SD-FastASE images obtained using the following parameters: TR 4000/TE 240, echo train length 79, field of view (FOV) 16 cm, slice thickness 0.8 mm, 512 x 512 x 40 matrix axial slab, voxel size 0.3 mm x 0.3 mm x 0.8 mm, 1 excitation, and scan time 11 min 48 s.

Two radiologists traced the areas of the EED and EES manually on the console for each slice, and the volumes were calculated (Fig). The area of the cochlear modiolus, the diameter of the EED, and the signal intensities of the EED and EES were also measured by drawing regions of interest manually. The ratios of the signal intensities of the EED and EES relative to the signal intensity of the adjacent cerebrospinal fluid were calculated. These measured values were compared against audiogram data. The degree of hearing loss was calculated as the average air conduction level of the audiogram data at three frequencies (500, 1000, and 2000 Hz). All audiograms were obtained within 1 month of MR examination. Statistical analysis was performed using stepwise regression analysis.

RESULTS: The volumes of the EED and EFS, the area of the modiolus, the diameter of the EED, and the signal intensities of the EED and EES did not show significant correlation with the degree of hearing loss.

DISCUSSION: In a previous CT study, it was reported that a deficient modiolus allows the transmission of CSF pressure waves into the labyrinth, resulting in damage to the hair cells in the organ of Corti (1). A recent MR study confirmed that a large endolymphatic duct and sac is frequently associated with modiolar deficiency, but some cases have a normal modiolar area (2). The results of that study do not necessarily support the previously proposed hypothesis regarding the cause of progressive hearing loss in the presence of a large vestibular aqueduct. The results of the present study also show that the degree of cochlear deficiency does not correlate with the hearing level.

Another theory suggests that hyperosmotic proteins in the enlarged endolymphatic sac reflux into the ductus cochlearis (scala media) through a widely patent endolymphatic duct, causing osmotic damage to the neuroepithelium (3). The signal intensities of the EED and EES did not correlate with hearing level in the present study, and the findings of this study do not directly support the latter theory. However, the signal intensities observed on T2-weighted images of the EED and EES may differ from those at the time of the insult to the neuroepithelium, and the findings of this study therefore cannot rule out the latter theory.

CONCLUSION: Although several pathophysiological theories have been proposed for hearing loss in EED and EES syndrome, the findings of the present study failed to directly support any of these theories. Therefore, the pathogenesis of EED and EES syndrome may not be so simple. Further studies, perhaps employing serial high-resolution MRI before and after the development of hearing loss, may be necessary to clarify the pathophysiology of this syndrome.

REFERENCES: