

without the connection. Hearing between each ear was then compared using the t test.

Results: Significant differences between ears with the connection and ears with significant EH of the vestibule and/or cochlea without the connection were seen for air-bone gap at 250 Hz and 3 pure-tone averages (500-, 1000- and 2000-Hz thresholds). Low-frequency air-bone gaps improved after EH medication in some patients.

Conclusions: Ears with significant EH that show a footplate-sacculle connection are associated with not only sensorineural hearing impairment, but also low-frequency air-bone gap. Changes in low-frequency air-bone gaps might reflect this aspect of EH.

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Expression pattern of WOLFRAMIN, the Wolfram syndrome 1 (WFS1) gene product, in the Common Marmoset (*Callithrix jacchus*), a non-human primate, cochlea

Presenting Author: **Noriomi Suzuki**

Noriomi Suzuki¹, Masato Fujioka¹, Makoto Hosoya¹, Naoki Oishi¹, Seiji Shiozawa¹, Takahiro Kondo¹, Reona Kobayashi¹, Takashi Inoue², Hideyuki Okano¹, Kaoru Ogawa¹

¹Keio University, ²Central Institute for Experimental Animals

Learning Objectives:

Introduction: Wolfram syndrome is an autosomal recessive disorder, known as DIDMOAD (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy, and Deafness) syndrome. Its causative gene, WFS1, encodes an 890 amino acid protein, called WOLFRAMIN, which maintains calcium homeostasis and unfolded protein responses in the endoplasmic reticulum (ER). Limited literatures describing temporal bone pathology display loss of hair cells in the basal turn and atrophy of stria vascularis in the apical turn. However, the expression of Wolfram in mice was distributed widely and uniformly in the sensory epithelium and was absent in the stria vascularis. Moreover, WFS1 knockout mice did not suffer deafness.

Learning objectives: In order to elucidate the discrepancy of the phenotype among species, and to explore the pathophysiology of deafness associated with WFS1 mutations, we examined expression of WOLFRAMIN in a non-human primate, common marmoset (*Callithrix jacchus*), cochlea.

Methods: We examined the expression pattern of WOLFRAMIN with double staining of WFS1 with other markers. The primary antibodies used are as follows: anti-WFS1 (rabbit IgG), anti-MYOSIN7a (mouse IgG), anti-CALDESMON (mouse IgG), and anti-CONNEXIN26 (CX26) (mouse IgG).

Results: In marmoset cochlea, WFS1 immunoreactivity was observed in basal cells of stria vascularis, type I fibrocytes, outer hair cells, outer sulcus cells, Claudius cells, Hensen cells, and spiral ganglion. Immunostaining for WFS1 was co-labeled with type I fibrocytes markers, CX26 and CALDESMON. In stria vascularis, immunoreactivity for WFS1 was co-labeled with a basal cell marker, CX26.

Conclusions: The expression pattern of WFS1 in common marmoset cochlea was different from that of mouse. The pattern suggests basal cells may play essential roles in the maintenance of stria vascularis. Clarifying the function of basal cells of primates, including human, may elucidate pathogenesis of hearing loss in Wolfram syndrome patients.

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The canal wall down procedure with soft posterior meatal wall reconstruction in acquired cholesteatoma. Focus on recurrence and postoperative middle ear status

Presenting Author: **Tomoyasu Tachibana**

Tomoyasu Tachibana¹, Michihiro Nakada², Yori-hisa Orita³, Kazunori Nishizaki³

¹Himeji Red Cross Hospital, ²Nakada ENT Clinic, ³Okayama University

Learning Objectives:

Introduction: The aim of procedures performed for acquired cholesteatoma (AC) is the complete removal of lesions, the prevention of disease recidivism, and the restoration of hearing loss. Although two main surgical procedures are canal wall up and canal wall down tympanoplasty (CWDT), it remains controversial which procedure would be appropriate for AC.

Objectives: To review surgical results of CWDT with soft posterior meatal wall reconstruction (SWR) for AC and to identify factors associated with surgical outcomes.

Methods: A retrospective review was made of 119 (flaccida, 99; tensa, 20) ears with AC who underwent CWDT with SWR at Himeji Red Cross Hospital between 2002 and 2015. The mean age was 45 years. The mean postoperative follow-up was 65 months (range, 12 to 156 months). Analyzed factors included sex, age, the type and extent of AC, the type of ossiculoplasty, and so on. We defined postoperative balloon-like retraction (PBR) with web formation, which needed reoperation to clean accumulated earwax, as 'nearly' recurrence. We classified all cholesteatomas according to JOS staging system for middle ear cholesteatoma (2015).

Results: Stage I and II were 24 and 95 ears, respectively. Residual was found in 11 ears (9.2%). Of 44 ears with PBR with web formation, 7 ears (5.9%) showed nearly recurrence. Seven residual and 4 nearly recurrent ears underwent

outpatient operation, and the other 7 were operated on under general anesthesia. No significant factors associated with residual or nearly recurrence were evident. In multivariate analysis, the proportion of postoperative mastoid aeration was significantly higher among cases with age

Conclusions: CWDT with SWR showed a low recurrence rate. More than half of residual and nearly recurrent ears could be easily treated with outpatient intervention. This procedure seems to be fully acceptable for surgical treatment of AC.

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Pneumolabyrinth and Perilymphatic Fistula After 27 Years of Head Trauma

Presenting Author: **Emel Tahir**

Emel Tahir¹, Münir Demir Bajin², Shamkhal Jafarov², Levent Sennaroğlu²

¹Ankara Dışkapı Yıldırım Beyazıt Research and Training Hospital, ²Hacettepe University Department of Otolaryngology

Learning Objectives: To discuss possible problematic situations in cochlear implantation following temporal bone fractures such as pneumolabyrinth. To emphasize the importance of careful evaluation of repeated imaging studies to rule out perilymphatic fistula after temporal bone trauma. To discuss timing of cochlear implantation after temporal bone fracture. To interpret temporal bone CT in case of pneumolabyrinth.

Introduction: Pneumolabyrinth is usually associated with a temporal bone trauma or stapes footplate fracture and a part of perilymphatic fistula. In this presentation, a case with footplate fracture results in pneumolabyrinth which is still perpetual 27 years after the trauma and its management is discussed with his radiological data and intraoperative video.

Case: A 56 year old male patient who had a bilateral profound sensorineural hearing loss (SNHL) admitted to our clinic. He experienced a head trauma results in transverse temporal bone fracture 27 years ago. CT demonstrated a fracture line was passing from cochlea and vestibule and pneumolabyrinth on the left side. MRI revealed labyrinthine ossification (LO) on semicircular canals which is characterized by diminished fluid intensity on T2 weighted images. Exploratory tympanotomy was performed and the ossicular chain was mobile. At stapes footplate level there was a fracture line accompanied by perilymph leakage. By the help of a pick a small fenestra was created at the footplate and it was obliterated by packing temporalis fascia. It was so unlikely to encounter a perilymphatic fistula after 27 years from trauma. The patient had no meningitis or encephalitis during that period. Postoperative CT scan verified the resorption of pneumolabyrinth and Weber test was localized to the operation side. He has been discharged the day after the operation without any complication.

Results: On the basis of this case, exploratory tympanotomy should be performed in patients with SNHL in

association with radiologically detectable pneumolabyrinth. If cochlear implantation was performed to this ear without notification of the fistula, the patient would suffer from meningitis because of the electrode and the implantation would not be beneficial. When there is a significant time delay between the temporal bone trauma and the cochlear implantation, LO or other structural abnormalities such as fistula should be ruled out prior to surgery.

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Development of the mastoid air cell system in children with congenital cholesteatoma

Presenting Author: **Masahiro Takahashi**

Masahiro Takahashi¹, Kazuhisa Yamamoto¹, Tsunetaro Morino¹, Manabu Komori¹, Yuitiro Yaguti², Yutaka Yamamoto¹, Nobuyoshi Otori¹, Hiromi Kojima¹

¹Jikei University School of Medicine, ²St. Marianna School of Medicine

Learning Objectives:

Introduction: Development of the mastoid air cell system in children with congenital cholesteatoma is said to be relatively good. However we sometimes encounter cases with poorly developed mastoid air cell system, but there have been few quantitative studies about the matter. The present study was undertaken in order to clarify the relationship between development of the mastoid air cell system in children with congenital cholesteatoma and various clinical factors.

Methods: Development of the mastoid air cell system of 53 children with congenital cholesteatoma was evaluated. The size of mastoid cells were measured from 0.5 mm sliced sectioned computed tomography scan of the temporal bone.

The sum of two areas, one showing the lateral semicircular canal and the other, 3 mm below it, was defined as the cross-section area of the pneumatized mastoid cells. Age, episode of otitis media, extension stage and location of the cholesteatoma, and size of the pneumatized mastoid cells were evaluated. Stage of the cholesteatoma was estimated with the grading system described by Potsic et al.

Results: The cross-section area of the affected side was significantly smaller than that of the unaffected side and this tendency was remarkable especially in the elder cases. The group with episode of otitis media was smaller in area than that without it. Cases with severe invasion of the cholesteatoma had smaller in area than that of others. Whether cholesteatoma present close to auditory tube or not made no difference in the development of the mastoid.

Conclusions: The mastoid cells in the ear of children with congenital cholesteatoma were poorly pneumatized compared with those of the unaffected side, and our data indicate that the suppression factor of pneumatization may be episode of otitis media, high age, and cholesteatoma presented in the mastoid portion.