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-saccule 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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ID: IP206

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

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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 Wolframin 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-labled 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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ID: IP207

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

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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 cholesteatetomas 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