Methods: Mouse induced pluripotent stem cells (iPS) were used for generation of Cx26-expressing cells with proper gap junction plaque between the cells. Adeno associate virus (AAV) were used for the GJB2 gene transfer and restoration of GJP.

Results: By diffentiation of iPS cells, we generated the Cx26-expressiong cells with large gap junction plaque as cochlear cells. Cochlear delivery of Gjb2 using AAV significantly improved the auditory responses and development of the cochlear structure of Cx26f/fP0Cre mice (Iizuka, *Hum Mol Genet*, 2015, 24(13):3651–61).

Conclusions: Using cell therapy or gene therapy to restore hearing in the mouse models of Gjb2-related deafness may lead to the development of effective therapies for human hereditary deafness.

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Genetics in Otology (R831)

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Is Cholesteatoma heritable and how can we find the genes involved?

Presenting Author: **Mahmood Bhutta**

Mahmood Bhutta

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Learning Objectives: To review evidence that cholesteatoma is heritable, and to discuss methods that can be used to ascertain genetic pathways involved.

Abstract is for the round table on "Genetics in otology"

The aetiology of cholesteatoma remains elusive. Those with a history of chronic mucosal disease are susceptible, but only a few such individuals will develop cholesteatoma. What evidence is there that cholesteatoma is a heritable disorder, and what methods can we use to elucidate genetic susceptibility?

I will discuss evidence from a recent systematic review of the heritability and genetics of cholesteatoma. This evidence includes reports of familial clustering of disease, and family history in the Kibutz population of Israel. Presence of disease in certain syndromes, in particular congenital malformation syndromes of the head or ear, also suggests genetic pathways are perturbed in cholesteatoma, and that a relatively small number of loci may be relevant.

I will introduce the other speakers for this session, and outline epidemiological and laboratory methods that can be exploited to further research molecular and genetic pathways involved in cholesteatoma. I will discuss how the discovery of such pathways could lead to potential clinical benefit.

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Free Papers (F832)

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Anatomical understanding in canal wall down mastoidectomy using a medical image processing system – simulation and education of ear surgery

Presenting Author: **Kazunori Nishizaki**

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Learning Objectives: To understand surgical anatomy of the temporal bone using a medical imaging processing system.

Introduction: Although canal wall down mastoidectomy still plays an important role in treatment of cholesteatoma, the chance of performing this procedure appears to be decreasing by appropriate intervention for ear diseases that develop cholestatoma. The decreasing chance to master this technique should be compensated by other methods. As one of the alternatives we introduce a simulation and education method of ear surgery using a medical image processing system.

Methods: Sagittal 2 and 3 dimensional reconstructive images (DRI) of the temporal bone CT scan are made for this purpose using a three-dimensional image analysis system volume analyzer (SYNAPSE VINCENT, Fuji Film Co, Tokyo, Japan).

Results: Sagittal 3DRIs introduced here show, in the order from lateral to medial, the antrum cavity, the prominence of the lateral semicircular canal, the incus body, the malleus head, the bridge being formed, the second genu of the facial nerve canal, the bridge resected at the level of the malleus neck, the mastoid segment of the facial nerve canal, the completely resected bridge, the lateral semicircular canal, and the completely opened facial recess. These images also show that the lateral wall of the attic has anterior-posterior and superior-inferior slants. 2DRIs parallel to the lateral wall of the attic show that the resection of the bridge parallel to the lateral wall is safe without risk of injury to the ossicles, the facial nerve, and the inner ear. However, sagittal 2 and 3DRIs should be evaluated for each patient due to individual differences in the temporal bone anatomy and bone structural changes affected by the disease.

Discussion and Conclusions: Since ear surgery usually progresses from lateral to medial, sagittal 2 and 3DRIs from lateral to medial simulate ear surgery including canal wall down mastoidectomy. Medical imaging processing systems are a useful and inexpensive tool to