Learning Objectives: Hearing loss is the most widespread sensory disorder, with an incidence of congenital genetic deafness of 1 in 1,600 children. For many ethnic populations, the most prevalent form of genetic deafness is caused by recessive mutations in the gene gap junction protein, beta 2, 26 kDa (*GJB2*), which is also known as connexin 26 (*Cx26*). For more than 15 years, we have developed and evaluated a mouse model of *Gjb2*-related deafness as follows, i) a dominant-negative *Gjb2* R75W transgenic mouse model shows incomplete development of the cochlear supporting cells, resulting in profound deafness from birth (Kudo et al., Hum Mol Genet 2003; Inoshita et al., Neuroscience 2008), ii) the outer hair cells (OHCs from the dominant-negative mutation of *Gjb2* are deformed, but reveal normal development and maturation (Minekawa et al. , Neuroscience 2009), iii) Cx26 dysfunction is associated with delayed apoptosis and retention of the greater epithelial ridge cells (Inoshita et al., BMC Genet 2014), iv) the disruption of the cochlear gap junction plaques is associated with the *Gjb2*-related deafness and the the assembly of cochlear gap junction plaques is dependent on Cx26 (Kamiya et al., J Clin Invest 2014), vi) the deformation of OHCs and the accumulation of caveolin-2 in the organ of Corti plays a crucial role in the progression of, or secondary OHC loss in, *Gjb2*-associated deafness (Anzai et al., Plos One 2015). In the next, we focused on the development of fundamental therapies for *Gjb2*-related deafness. Successful transgene expression was obtained through the round window membrane in the supporting cells of the neonatal mouse cochlea using adeno-associated viral (AAV) vectors without causing additional damage to the cochlear function (Iizuka et al., Huma Gen Ther 2008). Perinatal cochlear delivery of *Gjb2* using an AAV significantly improved the auditory responses and development of the cochlear structure (Iizuka et al., Hum Mol Genet 2015).

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Ossicular Reconstruction (R776)

ID: 776.1

The use of the Dresden partial clip prosthesis in ossicular reconstruction

Presenting Author: **Christopher Aldren**

Christopher Aldren
Wexham Park Hospital

Learning Objectives: To demonstrate the use of the Dresden partial clip prosthesis and show results.

The Dresden partial clip prosthesis is a titanium prosthesis used for ossicular reconstructin in the presence of a mobile stapes. Video will be shown to demonstrate its ease of application. Results will be presented and compared to the authors experience with other prostheses. Cases requiring revision will be discussed with video illustration.

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Endoscopic Ear Surgery: Concept and Technique (1) (V777)

ID: 777.1

Direct Cost Comparison of Totally Endoscopic versus Open Ear Surgery

Presenting Author: **Nirmal Patel**

Nirmal Patel[1], Aydin Mohammadi[2], Nicholas Jufas[2]
[1]Macquarie University, Sydney, [2]Kolling Deafness Research Centre| Macquarie and Sydney University

Learning Objectives: Objective: The introduction of new surgical techniques requires not only an evaluation of safety and clinical efficacy but also cost justification. Totally Endoscopic Ear Surgery (TEES) is a relatively new technique for managing chronic ear disease. The cost of specialised equipment required may be a barrier to implementation of the technique. This study aims to test the null hypothesis that open and endoscopic approaches have similar direct costs for the management of attic cholesteatoma in an Australian private hospital setting. Study Design: A retrospective direct cost comparison from a hospital perspective, of TEES and traditional canal wall up mastoidectomy for the management of attic cholesteatoma in the private tertiary setting was undertaken. Indirect and future costs were excluded. Methods: A cost comparison of anaesthetic set up and resources, operative set up and resources, average cost of running an operating theatre and cost of overnight admission was performed between the two techniques. Results: TEES has a mean reduction of AUD$2998.63 per operation from the hospital perspective when compared to an open procedure for attic cholesteatoma. Conclusion: Once the learning curve is achieved, TEES is more cost effective from a hospital perspective, than canal wall up mastoidectomy for attic cholesteatoma. When indirect and future costs are considered as well, the economic gain of managing attic cholesteatoma endoscopically could possibly be even greater.

Objective: The introduction of new surgical techniques requires not only an evaluation of safety and clinical efficacy but also cost justification. Totally Endoscopic Ear Surgery (TEES) is a relatively new technique for managing chronic ear disease. The cost of specialised equipment required may be a barrier to implementation of the technique. This study aims to test the null hypothesis that open and endoscopic approaches have similar direct costs for the management of attic cholesteatoma in an Australian private hospital setting.

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Endoscopic Ear Surgery: Concept and Technique (1) (V777)

ID: 777.2

Protympanum: Endoscopic Anatomy, Variations and Applications

Presenting Author: **Nicholas Jufas**

Nicholas Jufas, Nirmal Patel, Alexander Saxby, Jonathan Kong

Sydney Endoscopic Ear Surgery Research Group

Learning Objectives:

The protympanum is a middle ear space lying anterior to the mesotympanum. Historically, the region has often been overlooked due to its difficulty to visualize microscopically, despite optimum tissue and bone removal. Angled endoscopes with attached high definition cameras have meant that this region is now able to be fully appreciated.

This presentation will revisit the anatomical boundaries of this region, which will be demonstrated using endoscopic visualization. It will also detail and explain the newly defined classification systems for the region, including: (1) the protympanum conformation; (2) the protiniculum, a bony ridge from the promontory to the medial wall, separating the hypotympanum from protympanum; and (3) the subtensor recess, an area of pneumatisation inferomedial to the tensor tympani canal.

The presentation will utilise high definition endoscopic video and photographs taken during human cadaveric and live surgery. Normal anatomy will be demonstrated, as well as management of pathology in the area. Particular emphasis will be given to relevance of the region in both the development and surgical management of cholesteatoma.

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Endoscopic Ear Surgery: Concept and Technique (1) (V777)

ID: 777.3

Transcanal Endoscopic Ear Surgery for Pediatric Population with a Narrow External Auditory Canal

Presenting Author: **Tsukasa Ito**

Tsukasa Ito, Toshinori Kubota, Tomoo Watanabe, Kazunori Futai, Takatoshii Furukawa, Seiji Kakehata

Yamagata University Faculty of Medicine

Learning Objectives: To confirm that TEES with optional transcanal atticoantrotomy can be used for pediatric patients with a narrow EAC.

Introduction: Transcanal endoscopic ear surgery (TEES) is less invasive and functional procedure that offers the wide angle view of surgical field, higher magnification of fine anatomical structures and better visualization of hidden anatomical areas. We demonstrated the feasibility of TEES for the pediatric population with a narrow external auditory canal (EAC).

Methods: Thirty-nine patients ranging in age from 2 to 14 years old (median: 7.9 yrs) underwent TEES between November 2011 and March 2015. Twenty-one of these patients had surgery for cholesteatomas; fifteen for chronic otitis media; and five for malformation of the middle ear. A preoperative CT scan was performed to evaluate the middle ear disease. We evaluated the bony EAC retrospectively using ImageJ as the image processing program. The EAC bony portion was defined as the bone which surrounds the canal in sagittal CT images. The minimum and maximum Feret diameters were used as the EAC minor and major axis respectively. Transcanal endoscopic tympanoplasty was performed with a rigid endoscope with an outer diameter of 2.7 mm coupled to the full HD system. Transcanal atticoantrotomy was also performed, as necessary, on some patients to remove cholesteatomas in the antrum. We will show a video of the surgical procedure for cholesteatomas.

Results: The minor axis ranged from 3.2 to 5.9 mm (mean: 4.7 mm), while the major axis ranged from 4.8 to 10.2 mm (mean: 7.8 mm). TEES was successfully performed on each patient without having to make a retroauricular incision. Postoperative hearing levels and air-bone gap fell into an acceptable range and are comparable to those results obtained by microscopic ear surgery.

Conclusions: TEES is feasible using rigid endoscopes with an outer diameter of 2.7 mm and is effective and less invasive for pediatric patients with a narrow EAC.