to be solved in the discussion. Authors will present also changing opinion in surgery in the only hearing ear with possibility to manage unexpected deafness by cochlear implantation. Special case reports will be the subject of discussion after short communication presented by the panel members.

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CI in chronic ears (R644)

ID: 644.2

Subtotal Petrosectomy for cochlear implantation in cases of Chronic Otitis Media

Presenting Author: **Miguel Arístegui**

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Learning Objectives: We will show the safety of subtotal petrosectomy applied to cochlear implantation in cases chronic otitis media, to prevent future infections that might compromise the implant

Expanding indications for cochlear implantation require adaptation of surgical techniques in special cases.

The presence of chronic otitis media (relapsing acute otitis media, chronic supurative otitis media or cholesteatoma) require special protection in cases of cochlear implantation.

Subtotal petrosectomy offers the best protection option against future infection in these cases.

Cul di sac closure of the esternal auditory canal, sealing of the Eustachian tube orifice and elimination of middle ear mucosa provides a secure scenario to avoid infections and risk cochlear implant explantation in the future.

Out of 41 cases in which we have used this technique we have 17 cases that were applied to chronic otitis media of the above mentiones¡d cathegories. We will report on rationale, technique and complications.

Follow up is made with MRI techniques adapted to the type of implant.

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New diagnostic method in otology (N645)

ID: 645.1

Trends in genetic diagnostics of hereditary hearing loss

Presenting Author: **Ronald Pennings**

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Introduction: Over the past decades, many deafness genes have been identified to cause hereditary hearing impairment (HI). It therefore, has become possible to screen for these genes in the out-patient clinic. The importance of genetic screening of HI is that patients can be counseled about the cause and prognosis of their hearing loss and effects of rehabilitation.

Hearing impairment is genetically heterogeneous and testing of several single HI-related genes is laborious and expensive. This study evaluates the diagnostic utility of whole exome sequencing (WES) targeting a panel of HI-related genes.

Methods: Two hundred index patients, mostly of Dutch origin, with presumed hereditary HI underwent WES followed by targeted analysis of an HI gene panel of approximately 100 genes. 206 additional patients underwent single gene testing guided by phenotype analyses.

Results: We found causative variants underlying the HI in 67 of 200 patients (33.5%). Eight of these patients have a large homozygous deletion involving a known HI gene, which could only be identified by copy number variation detection. Variants of uncertain significance were found in 11 patients (5.5%). In the remaining 122 cases no potentially causative variants were detected (61%). The diagnostic yield of single gene testing in the 206 additional patients was 7.6%.

Conclusion: The diagnostic yield for HI using WES targeting a HI gene panel is higher (33.5%) than targeted sequencing of single genes (7.6%). In our patient cohort, causative variants in *GJB2*, *USH2A, MYO15A*, *STRC*, and in *MYO6* were the leading causes for autosomal recessive and dominant HI, respectively. Segregation analysis of variants of uncertain significance will further increase the diagnostic yield of WES. A practical workflow for genetic testing of hereditary HI for screening in the out-patient clinic will be presented.

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New diagnostic method in otology (N645)

ID: 645.2

N645 session : A review of automated audiometry devices and portable smartphone or tablet-based hearing testing systems in otology

Presenting Author: **Allan Ho**

Allan Ho

University of Alberta

Learning Objectives: Recent advances in portable and automated hearing testing systems has enabled testing to occur outside the traditional sound treated booths. This has far reaching implications for otologists and the patients they treat. It expands the utility of these devices in the community and in the developing world where diagnostic audiology services are scarce. We aim to review automated hearing testing systems which do not require testing in traditional sound treated booths. We will discuss the evidence supporting portable automated hearing testing systems which are available on the web and those that are independent applications for smartphones or tablet computers.

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