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Paediatric Cholesteatoma (R811)

ID: 811.1

From Retraction Pocket to Cholesteatoma: A Continuum in Pediatric Ears

Presenting Author: **Cuneyt Alper**

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Learning Objectives: Cholesteatoma is not a static condition, and does not develop instantly, changing from the definition of no cholesteatoma to a cholesteatoma all of a sudden. There is gradual transition, from state of cholesteatoma prone conditions to pre-cholesteatoma to cholesteatoma, during which an ear may not fit to the clear definitions. Awareness of this transition may be helpful in identifying the ears that has the risk for cholesteatoma development, and perhaps interventions may moderate or prevent this process.

There is a continuum of conditions from Eustachian tube (ET) dysfunction (ETD) to tympanic membrane (TM) retraction, TM retraction pocket, cholesteatoma prone retraction pocket, pre-cholesteatoma, cholesteatoma, recurrence of ET retraction pocket, recurrence of cholesteatoma. Although this progression of conditions is limited with primary acquired cholesteatoma, it represents majority of the pathogenesis of cholesteatoma cases in both children and adults. While this may be seen as theoretic/logical progression, this transition is observed more clearly in children with recurrent and chronic otitis media with likely underlying ETD followed up by pediatric otolaryngologists through their growth and development.

Current presentation is on the experience with the cholesteatoma prone ears, discussing the definitions, risk factors, management of risk factors, methods of prevention, and the decision making process in assessment and management of ears with retraction pocket, pre-cholesteatoma and early cholesteatoma states that are in transition to cholesteatoma formation.

There is a need for consensus on definitions and classification of these transitional conditions and diagnostic criteria for the underlying ETD, a grading system sensitive in capturing the changes in the state of ears as well as ETD, so that prospective close follow-up generates comparable data for future analysis, making analysis of results from multiple centers and hypothesis driven trials possible.

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Paediatric Cholesteatoma (R811)

ID: 811.2

The Utility of Diffusion Weighted Magnetic Resonance Imaging in Identifying Cholesteatoma in Children

Presenting Author: **Kenneth Lee**

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Learning Objectives: 1) Understand the rationale of DW MRI for identifying presence of cholesteatoma 2) Understand the limits of DW MRI for identifying presence of cholesteatoma 3) Understand the accuracy and utility of DW MRI in determining the presence of cholesteatoma in children.

Cholesteatoma has a significant rate of recidivism. Children in particular are at higher risk of residual disease due to the aggressive nature of congenital cholesteatomas as well as recurrent disease due to ongoing Eustachian tube dysfunction. As a result, historically, "second look" procedures were routinely performed and considered standard of care. Recently, obligate planned revision tympanomastoidectomy procedures have become challenged due to concerns of repeated risks of anesthesia and surgery as well as added health care costs. While the diagnosis of cholesteatoma is primarily made clinically, imaging, particularly computed tomography, has been used as a tool to assist in confirming the diagnosis and determining the extent of the disease. In patients who have previously undergone primary cholesteatoma surgery, diffusion weighted magnetic resonance imaging (DW MRI) has become a useful imaging modality to assist in deciphering the presence of cholesteatoma vs. mere fluid or inflammation in the middle ear and mastoid. Since 2012, we have performed nearly 100 DW MRI studies in children to determine the presence of cholesteatoma. The results of these studies in comparison to subsequent surgical findings will be presented to review the accuracy of DW MRI in identifying cholesteatoma in our pediatric patient population.

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Paediatric Cholesteatoma (R811)

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Predictive factors for recurrent cholesteatoma

Presenting Author: **Adrian James**

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University of Toronto

Learning Objectives: To enhance understanding of the factors which contribute to development of recurrent cholesteatoma in children as part of the Round Table session on Paediatric Cholesteatoma.

Predictive factors for recurrent cholesteatoma

Recurrent cholesteatoma (i.e. development of new disease after previous surgical clearance) may be a consequence of persistent pathogenetic factors and perhaps also of surgical technique. Cholesteatoma is commonly considered to behave more aggressively in children than adults. A clear understanding of factors that predispose to recurrent disease in children may help selection of optimal surgical

technique for disease control for example between canal wall up (CWU) or down (CWD) and mastoid preservation or obliteration.

In an attempt to clarify this relationship between disease, patient and surgery, a single-surgeon prospective database of consecutive cholesteatoma surgeries was analysed. Analysis was restricted to cases with no prior history of cholesteatoma surgery. Out of 368 paediatric cases, 328 (89%) were completed with CWU of which 69 were totally endoscopic (TEES), and 40 were completed with CWD of which 10 had primary obliteration. Overall 34 (9%) were found to develop recurrent cholesteatoma. Kaplan Meier survival (KM) analysis was used to control for the cumulative increase in recurrence with time, giving an overall recurrence rate of 12% at 5 years and 18% at 10 years. Perhaps surprisingly, KM analysis suggests that gender and younger age had no impact on likelihood of recurrence. Similarly the same rate of recurrence was found for congenital and acquired cholesteatoma and whether disease was acquired from pars tensa or pars flaccida retraction. The most significant pre-operative determinant of outcome was extent of cholesteatoma, with cholesteatoma involving 4 sub-sites (meso- and epitympanum, antrum and mastoid) having significantly greater risk of recurrence than smaller disease (30% at 5 years; KM log rank statistic $p = 0.002$).

While canal wall down surgery (CWD) is commonly considered to have a lower risk of recurrence than intact canal wall surgery (CWU), in this series, KM analysis showed no difference in rate of recurrent cholesteatoma between these different techniques. Subgroup analysis, in which the sample sizes are small, suggests (a) the same recurrence rate with TEES and (b) no difference in revision surgery for uncontrolled disease for CWD cases with or without obliteration of the mastoid. In contrast to the perhaps unreachable gold-standard of a randomized prospective trial to control for differences between patients, it must be noted that selection of surgical technique was allocated according to patient and disease factors, for example CWD surgery was used more often for larger cholesteatoma.

Careful recording and analysis of surgical intervention and outcome provides valuable insight into the effectiveness of otologic intervention for cholesteatoma.

Survival analysis is required to control for the increasing incidence with longer follow up. It is also important to control for other risk factors such as the extent of cholesteatoma. Understanding of the mechanisms of recurrent cholesteatoma is confounded by selection of surgical technique according to the extent of disease.

Nevertheless, as it seems that "*Bad ears do badly and good ears do well*" it is arguably most important maximise strategies to prevent recurrent disease in those thought to be most at risk.

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Paediatric Cholesteatoma (R811)

ID: 811.4

Managment of Pediatric Cholesteatoma: The Gruppo Otologico Experience in the Management of 618 Cases

Presenting Author: **Enrico Piccirillo**

Enrico Piccirillo, Enrico Piccirillo, Flavia D'Orazio, Melissa Laus, Sampath Chandra Prasad Rao, Mario Sanna

Gruppo Otologico

Learning Objectives: Management of pediatric cholesteatoma and the long term outcomes of canal wall up mastoidectomy.

The objectiveof this study is to report the Gruppo Otologico experience in the surgical treatment of paediatric cholesteatoma. This is a retrospective study wherein 572 charts of young patients who underwent surgery between 1983 and 2015 were analysed. 46 patients had bilateral disease and the average age was 10.6 years old. The extension of cholesteatoma was defined using Sanna and Zini's cholesteatoma classification. The most commonly surgical procedure used in children were Canal Wall Up and a Canal Wall Down tympanoplasty. Canal Wall Up Tympanoplasty (CWU) was performed in 263 patients, while, Canal Wall Down Tympanoplasty (CWD) that includes Modified Bondy Technique (BT) and Radical Mastoidectomy (RM), was used in 258 patients. There were more numbers of revision surgeries in CWU (34%) than CWD (10%) tympanoplasty. In all surgeries put together, we had an improvement of hearing of a mean of 8.5 dB HL. The mean follow-up was 10 years. In conclusion, the technique of choice for pediatric cholesteatoma is CWU tympanoplasty. However, the use of a CWD approach to surgically treat extensive cholesteatoma in children results in a low recurrence rate with a high rate of trouble-free ear in the long term.

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Paediatric Cholesteatoma (R811)

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Long term functional and hearing outcomes of surgery in pediatric cholesteatoma

Presenting Author: **Enrico Piccirillo**

Enrico Piccirillo, Flavia D'Orazio, Sampath Chandra Prasad, Gianluca Piras, Mario Sanna

Gruppo Otologico

Learning Objectives: The aim of this study is to retrospectively analyze the functional and hearing outcomes of surgery for cholesteatoma in pediatric population.

Study Design: Retrospective study

Setting: Gruppo Otologico, a quarternary referral center for Otology and Skull Base Surgery in Italy.

Materials & methods: A retrospective analysis is presented of 664 cases of cholesteatoma in pediatric population who were treated by surgery. The surgical approach was chosen according to the hearing loss, symptoms, status of tympanic membrane and radiological finding. Surgical procedures included Canal Wall Down mastoidectomies, Modified Bondy mastoidectomies, Canal Wall Up mastoidectomies, Radical mastoidectomies, revision surgeries and Subtotal Petrosectomies. In some of these patients