were observed in MEC matrix and perimatrix. There was no meaningful difference between congenital and acquired MEC with respect to p21 contrary to p53. A statistical significance was obtained for APO2.7-positive cells in MEC epithelium (43.23 ± 4.8%) as compared to CS (29.89 ± 6.2%).

More extensive positive immunohistochemical reaction with anti-TGF-alpha, Ki67 and PCNA was observed in MEC matrix and perimatrix compared with CS.

RAGE expression levels was present in all cholesteatoma tissues (strong in 86 %) vs skin 25% (weak) respectively ($p < 0.0001$).

*Conclusion*: Selected markers of apoptosis, proliferation, angiogenesis and inflammatory response are associated with cholesteatoma development. The co-expression of HMGB1 and RAGE in MEC may result in activation of the intracellular signaling pathways. This process may be responsible for faster accumulation of keratin debris, more invasive process, and affect the clinical course and the treatment outcome.

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## Congenital Cholesteatoma (R634)

### ID: 634.1

### Congenital cholesteatoma of the middle ear: a report of 62 cases

Presenting Author: **Katsumi Doi**

Katsumi Doi

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*Introduction*: Congenital cholesteatoma (CC) of the middle ear is a rare clinical entity that classically presents as a white mass situated in the anterior-superior quadrant of the middle ear behind an intact tympanic membrane (TM). Derlacki and Clemis established the diagnostic criteria for CC: 1) A pearly white mass medial to an intact TM, 2) Normal Pars Tensa and Pars Flaccida, 3) No history of otorrhea, perforation or previous otologic procedures. CC is seen far more frequently in children, but House and Sheehy remarked adult patients with cholesteatoma behind an intact TM.

*Materials and Methods*: A retrospective analysis was conducted of the clinical charts of all patients with CC in both children (n = 56) and adults (n = 6) from 1992 to 2015. CCs of the petrous apex (n = 15) were excluded. 1445 cases of acquired and congenital cholesteatomas were treated, therefore, the prevalence of CC should be 4.3% (62/1445).

*Results*: Based on the staging system by Potsic 54 patients were classified into stage1–4 according to the surgical findings: 11 cases in stage1, 7 in stage 2, 24 in stage3, and 20 in stage4. It was suggested that most CCs could be derived from the epidermoid formation (EF) in 53 cases. A planned two-staged surgery was conducted in 54 cases (87%), while one-stage surgery was adopted in 8 cases. The residual cholesteatoma at the time of second stage surgery was detected in 19 out of 48 cases (40%). The most common residual sites were at oval window (n = 7). Hearing assessment was done in 55 cases: success in 46 cases (84%), moderate in 8 cases, and failure in one.

*Discussion*: As the stage of CC advanced, the area of its invasion could be enlarged, which should result in a higher risk of CC residual. Considering that CC is usually discovered in its advanced stages (stage 3–4), the establishment of a screening program including otoscopic and CT examinations and hearing tests for early CC diagnosis should be required.

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## Back to the future: the evolution of cholesteatoma diagnosis and management (N635)

### ID: 635.1

### Back to the Future: The Evolution of Cholesteatoma Diagnosis and Management

Presenting Author: **John McElveen**

John McElveen

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Confucius once said, “Study the past if you would define the future.” As an introduction to the 10th International Conference on Cholesteatoma and Ear Surgery, the American Neurotology Society has assembled panelists (Mr. David Moffat (Addenbrooks Hospital), Dr. Jack Lane (Mayo Clinic), Dr. Clough Shelton (U. of Utah), Dr. Moises Arriaga (LSU) and Dr. Dennis Poe (Harvard) to discuss the evolution of cholesteatoma diagnosis and management. Dr. John McElveen (Carolina Ear Research Institute) will moderate the panel.

Mr. Moffat will trace the history of the diagnosis of cholesteatoma from ancient times to the present. Based on the research in 1967 by McKenzie and Brothwell, the existence of chronic suppurative otitis media in prehistoric times has been clearly documented. It was the French anatomist Joseph-Guichard Du Verney who in 1683 first described a temporal bone tumour which was probably a cholesteatoma. However, the term, “cholesteatoma”, was first used by Johannes Peter Muller in 1838. Although a misnomer, it has continued to be used to describe “keratomas” involving the temporal bone and skull base. Abramson *et al* in 1977 provided a more detailed definition of cholesteatomas at the First International Conference on Cholesteatoma.

The classification of cholesteatoma into congenital and acquired and the latter’s subdivision into primary and secondary acquired was the natural sequel of refinements in diagnostic capability which accompanied the use of the microscope both in histopathology and in the clinical examination of the ear (Nylen, 1921).

Since the dawn of medical imaging, radiographic examination of the temporal bone has been used in the evaluation and management of cholesteatoma. X-ray modalities have evolved from plain radiographs (1900–1940s) to polytomography (1950–60s) to single slice Computed Tomography (CT) acquired separately in the axial and coronal planes (1970–1980s) to multislice CT with multiplanar

reconstruction (1990–2010s). MR with diffusion-weighted imaging (DWI) (1990–2000s) has been employed most effectively in the detection of recurrent cholesteatoma, particularly in the setting of canal wall up procedures. In some centers, DWI MR has obviated the need for many second look procedures. Dr. Lane will explore the evolution of these radiographic techniques and discuss recent advances in temporal bone imaging, including Photon Counting CT and high field MR (3 T and 7 T). These imaging modalities are anticipated to achieve greater degrees of resolution and sensitivity in the detection of acquired and congenital cholesteatomas involving the temporal bone.

Just as imaging techniques have continued to evolve, so have the surgical techniques used to manage cholesteatomas. However, surgical management of cholesteatomas involving the temporal bone continues to be a source of some controversy. Some advocate a one stage procedure with revision of the failures. Others promote modern techniques for post-operative surveillance by employing imaging or endoscopic inspection. Dr. Shelton will discuss the surgical options currently used and review the rationale, controversy and history of the two stage strategy as advocated by the late Jim Sheehy.

Skull base cholesteatomas can be particularly destructive and potentially life threatening. Dr. Arriaga will discuss the traditional and modified versions of skull base neurotologic approaches such as Middle Cranial Fossa, Middle Fossa Transpetrous (extended middle fossa), Retrolabyrinthine, Retrosigmoid and Translabyrinthine approaches. In addition, he will explore the use of four-hand skull base surgery techniques, fallopian bridge strategies and simultaneous application of the endoscope to microscopic visualization to deal with challenging skull base cholesteatomas.

Dr. Poe will bring us "Back to the Future" with an update on the current understanding and treatment of Eustachian tube dysfunction as it relates to chronic otitis media. His focus will be on tubal dilatory dysfunction which typically involves the cartilaginous portion of the Eustachian tube. He will explore the use of Eustachian tuboplasty to treat tubal dysfunction and discuss its potential role in standard tympanoplastic and tympanomastoid procedures.

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## Back to the future: the evolution of cholesteatoma diagnosis and management (N635)

### ID: 635.2

### Evaluation of Eustachian tube Function and Practical Physiology for Surgeons

Presenting Author: **Dennis Poe**

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The Eustachian tube (ET) bridges the realm of the nasal cavity and upper aerodigestive tract with the ear and serves to optimize its special sensory role of hearing. The middle ear and mastoid system behaves as an auxiliary sinus and the ET can be thought of as a long, dynamic ostium with a functional valve located within the cartilaginous portion. Failure of the "valve" to function properly can occur if it dilates insufficiently to adequately aerate the middle ear or if it fails to close following dilation. It is affected by all of the same pathophysiologic processes as the nose and other sinuses.

The net effect of the middle ear gas exchange is to cause a constant absorption of gases from the middle ear air space into the venous blood system, creating an ongoing tendency toward developing negative pressure compared to ambient atmospheric pressure. When appropriate, the ET should dilate, typically with a swallow or yawn for about 400 msec, to restore the middle ear pressure toward ambient, optimizing the function of the tympanic membrane. If the dilatory effort is consistently insufficient to adequately aerate the middle ear, ET dilatory dysfunction results with the possible consequences of negative middle ear pressure, retraction of the tympanic membrane, otitis media with effusion, tympanic membrane perforation, conductive hearing loss, fixation of retraction pockets and ultimately cholesteatoma.

Most of the pathology that is responsible for dilatory dysfunction has been observed within the cartilaginous portion and is most commonly due to inflammatory disease, which can be readily diagnosed with transnasal endoscopy. A careful assessment of the dynamics of the ET by endoscopy can be very effective in determining the etiology, location and severity of dilatory and patulous dysfunction within the functional valve in the cartilaginous portion. Vocalizing "K-K-K" demonstrates isolated excursions of the Levator Veli Palatini (LVP) muscle. Swallows start with elevation of the LVP that acts as a scaffold upon which the additional contraction of the Tensor Veli Palatini muscle should be seen to dilate the valve open under normal circumstances. Yawns or vocalizing "Ahhh" can demonstrate a maximal dilatory effort. Disorders of dilation may be observed and classified. Inflammatory disease can be graded on a recently validated mucosal inflammation score instrument. The etiology of the inflammation can be investigated and treated, with the most common causes being infectious or reflux in younger children and over age 6, allergic disease, reflux, rhinosinusitis, adenoid hypertrophy and other commonly known causes of nasopharyngeal inflammation.

Treatment of the underlying medical conditions can result in improvement of ET function and resolution of middle ear disease. When the medical causes have been optimally treated, but ET dilatory dysfunction persists, possibly due to irreversibly injured mucosa, biofilms or other pathology, tympanostomy tubes are usually recommended. When tubes fail to resolve the problem, treatment of the underlying pathology with surgery can be offered. Surgery may involve turbinate reduction, sinus surgery, adenoidectomy, or balloon dilation of the ET. All of these procedures are designed to remove irreversibly injured tissue and provide a fresh start, assuming the underlying medical conditions are adequately controlled. Failure to control the medical problems can lead to recurrence of inflammatory disease.

Failure of the functional valve to close results in patulous dysfunction. Once thought to be rare, it is now clear that the diagnosis is frequently missed. It can be related to weight loss, chronic illnesses (especially rheumatologic), but it often occurs after long-standing inflammatory dilatory dysfunction with atrophy and decreased mucus production. This may occur particularly with chronic allergic rhinitis. Patients develop frequent sniffing strategies to minimize their symptoms, despite negative middle ear pressure or effusions, raising suspicion that dilatory dysfunction has transitioned to patulous dysfunction. Examination of the tympanic membrane by otoscopy or tympanometry for excursions with ipsilateral nasal breathing can be diagnostic and endoscopy of the ET will reveal a defect in the functional valve, usually within