6th SYDNEY

ENDOSCOPIC EAR SURGERY

DISSECTION COURSE

Royal North Shore Hospital
September 21st & 22nd 2019

SYDNEY
ENDOSCOPIC
EAR SURGERY
RESEARCH GROUP

RACS
CPD Approved

Course Directors
A/Prof Nirmal Patel
A/Prof Nicholas Jufas

Local Faculty
A/Prof Alexander Saxby
A/Prof Jonathan Kong

Guest Local Faculty
Dr Andrew Ma

International Faculty
Prof Adrian James
A/Prof Arun Iyer
Prof Daniele Marchioni

Introduction

Welcome to the 6th Sydney Endoscopic Ear Surgery Dissection Course, this year held at Royal North Shore Hospital. We hope you enjoy and benefit from two full days of dissection, lectures and live surgery, focused entirely on the new technique of endoscopic access to the ear.

Background

Although ear surgery has been performed successfully with a microscope for many years, recent key improvements in endoscope and camera technology have allowed keyhole (transcanal) approaches with high fidelity. Minimally invasive techniques have a number of advantages and have been used to great effect in laparoscopic, thoracic and orthopedic surgery. In otolaryngology specifically, Functional Endoscopic Sinus Surgery (FESS) has utilized endoscopes for decades with excellent results. The ear canal provides the perfect corridor for access to the middle ear, enabling the possibility of removal of diseases such as cholesteatoma without having to drill the canal or mastoid.

With transcanal surgery patients can enjoy the benefit of less invasive surgery with no post-auricular scar or disfiguring meatoplasty. Day surgery possibilities and decrease in disposables such as drill pieces also have important health dollar cost saving implications. Clinically it is expected that the increased visualisation enabled by the endoscopes may reduce recidivism rates and that the decreased disturbance and/or removal of healthy tissue will result in faster and less painful recovery.

Fundamental Principles of EES

- Improved visualization of the middle ear
- Minimally invasive surgical techniques
- Potentially improved functional outcomes

Endoscopic ear surgery around the world

Endoscopic ear surgery was introduced to Australia in 2012 and worldwide the method is becoming increasingly popular. The third EES Congress was be convened in Boston, USA in 2019. The 4th EES World Congress will be held in Kyoto, Japan in 2021.

An international society has been set up with invited delegates from all around the world, with the purpose of developing the technique, called the “International Working Group for Endoscopic Ear Surgery” http://iwgees.org/.

Sydney Endoscopic Ear Surgery Research Group

The Sydney Endoscopic Ear Surgery (SEES) Research Group is an Australian not for profit organisation dedicated to research, teaching and advancing the technique of Endoscopic Ear Surgery to Otolaryngologists and trainees (http://sydneyearendoscopy.com/). The founding members are Nirmal Patel, Jonathan Kong, Alexander Saxby and Nicholas Jufas. All of the SEES group are members of the International Working Group on Endoscopic Ear Surgery (IWGEES). The SEES Group offers an International Clinical Fellowship in Endoscopic Ear Surgery & Otology (applications can be made through the website).

COURSE TIMETABLE

Participants will be split into Groups A and B. Each will contain dissectors & observers.

During the split sessions, one group will dissect / observe (Level 6 Surgical Skills Lab, Kolling Building) whilst the other group listens to lectures (Level 6 Lecture theatre). All dissection and lectures are on Level 6, Kolling Building Royal North Shore Hospital Main campus, St Leonards.

All food and drink will be consumed on Level 5 of the Kolling Building.

DAY 1 - SATURDAY 21ST SEPTEMBER

7:30	8:00	Coffee / Breakfast / Registration	
8:00	8:30	**WELCOME** Welcome & Introduction - N Patel Introduction to EES: What are the advantages and disadvantages? - A James	
8:30	9:00	**DEMONSTRATION** Holding The Scope, Raising a TM Flap & Inspection of middle ear anatomy - A Iyer	
9:00	11:00	**DISSECTION GROUP A** STEP 1 Ear prep, TM flap & Prussak's space STEP 2 Protympanum, ET, Tensor Fold STEP 3 Epitympanum and Lateral Atticotomy	**LECTURES GROUP B** Endoscopic Anatomy & Ventilation N Jufas Paediatric EES & Myringoplasty A James Control of Bleeding in EES J Kong
11:30	13:00	**LECTURES GROUP A** As Above	**DISSECTION GROUP B** As Above
13:00	13:30	**LUNCH (with Conference Photograph)**	
13:30	15:00	**DISSECTION GROUP A** STEP 4 Retrotympanum & Hypotympanum STEP 5 Ossicle Removal & Extended Atticotomy STEP 6 Tympanic Plexus & Facial Nerve	**LECTURES GROUP B** Cholesteatoma in EES N Patel What is the evidence that EES works? A Iyer Non Cholesteatoma EES A Saxby
15:00	16:30	**LECTURES GROUP A** As Above	**DISSECTION GROUP B** As Above
16:30	18:00	**LIVE SURGERY (from Verona, Italy) - D Marchioni** Q & A and roving microphone during the live presentation	
19:00	23:00	**CONFERENCE DINNER** Sails on Lavender Bay - **sailslavenderbay.com**	

DAY 2 - SUNDAY 22ND SEPTEMBER

7:30	8:00	Coffee / Breakfast	
8:00	9:00	**RECAP & PANEL DISCUSSION- N Patel** Revision of yesterday's dissections with Q&A (Whole Faculty) Tips & Pearls, Difficult Cases	
9:00	9:30	**DEMONSTRATION** Inner Ear Anatomy - Lateral Skull Base Corridors - N Patel	
9:30	11:00	**DISSECTION GROUP A** STEP 7 Major vessels & Infracochlear Approach STEP 8 Transpromontorial Approach to IAC (Dissectors can start second ear specimen early here if ahead)	**LECTURES GROUP B** Instrumentation & Equipment N Jufas Managing difficult scenarios - stapes, dehiscent facial nerve, difficult access A Iyer Role of combined approach - when to convert A Saxby
11:00	12:30	**LECTURES GROUP A** As Above	**DISSECTION GROUP B** As Above
12:30	13:00	**LUNCH**	
13:00	13:30	**DEMONSTRATION** Applied EES: Stapedotomy, Myringoplasty, Cholesteatoma techniques (A James)	
13:30	15:00	**DISSECTION GROUP A** Second ear specimen: Dissectors can either run through the antomical dissection again or simulate clinical applications , eg. Stapedotomy, OCR, myringoplasty, cholesteatoma resection	**LECTURES GROUP B** Use of cartilage in EES A Ma Safety & Improving outcomes in EES: Tips and Pearls A James Setting up an EES Practice and getting started J Kong
15:00	16:30	**LECTURES GROUP A** As Above	**DISSECTION GROUP B** As Above
16:30		**END OF COURSE (with Course Feedback & Concluding Remarks)**	

SYDNEY
ENDOSCOPIC
EAR SURGERY
RESEARCH GROUP

Course Faculty

Invited International Faculty

Professor Adrian James

One of North America's first endoscopic ear surgeons from the University of Toronto, Canada. Adrian is an International Working Group of Endoscopic Ear Surgery (IWGEES) Board Member and the scientific advisor to the Board. He is a paediatric otolaryngologist and a very deliberate and critical thinker. Adrian has made enormous contributions to the endoscopic field particularly in outcomes research and the use of laser in endoscopic ear surgery. He is a dynamic speaker and teacher.

Associate Professor Arun Iyer

Arun is a Board Member of the International Working Group on Endoscopic Ear Surgery (IWGEES). He is one of the UK's first endoscopic ear surgeons from the University of Glasgow. He ran the first course in Scotland and regularly teaches at the British Endoscopic Ear course. Arun is a critical thinker and has made significant contributions to the field in particular with our understanding of imaging in EES and outcomes research.

Professor Daniele Marchioni (Live Surgery)

Professor and Head of Department at the University of Verona. Daniele is world re-knowned for progressing the method of EES and has been performing the technique for over 10 years. He is the author of the first comprehensive textbook on the subject "Endoscopic Ear Surgery" Thieme Publishing. Professor Marchioni is a founding and Board member of IWGEES.

Dr Andrew Ma (SEES Otology/Neurotology Fellow)

Andrew is the current SEES Otology/Neurotology fellow. He completed his Otolaryngology residency training at the University of Toronto. Andrew has presented at multiple international endoscopic ear conferences and is actively involved in EES outcomes research. His long term plans are to build an EES practice in Canada, with a focus on surgical education.

Local Faculty - Sydney Endoscopic Ear Surgery Research Group

A/Prof Nirmal Patel

A/Prof Nicholas Jufas

A/Prof Jonathan Kong

A/Prof Alexander Saxby

DISSECTORS

GROUP A

Celeste Ann CHUA
Anton HINTON-BAYRE
Jiaying LIU
Niell BOUSTRED
Helmi BALFAS
Rohit UDAYA PRASAD
Robert EISENBERG
Daniel YAFIT
Fatia Permata SARI
Dian Ayu RUSPITA

GROUP B

Susana FATCHUR RACHMAN
Diana ROSALINA
Rini FEBRIANTI
Patrick DE WAAL
Shannon WITHERS
Alice STRINGER
David HOUGHTON
Alessandra LOCATELLI SMITH
Ena SARIKENCANA
David LOW

NON-DISSECTORS

GROUP A

Fikri Mirza PUTRANTO
Ahmad Dian WAHYUDIONO
Bintang Berthaliana Mangantar NAPITUPULU
Orit SAMUEL
Raul ROSAS

GROUP B

Diamanti DIAMANTARAS
Anna Mailasari Kusuma DEWI
Alexander TREBLE
Joanne MALEK

Course Location and Venue

The course is held at the Sydney Skills Simulation Centre (SSSC) in the Kolling Building at Royal North Shore Hospital, St Leonards, Sydney. The SSSC is at the corner of Westborne St and Reserve road, St Leonards. Lectures and catering will be on Level 5 and the Dissection/ Dry stations will be on Level 6 of the Kolling Building.

The Royal North Shore Campus, St Leonards is 6 km north of the Sydney CBD and can be easily accessed by train on the T1 North Shore Line.

Course Sponsorship

A course of this size, with complex equipment and prestigious international speakers would not be possible without the extremely generous support of our sponsors. In particular, our Platinum sponsor, Karl Storz, has flown from Germany 10 HD stacks, scopes and dedicated endoscopic ear surgery equipment. They have been an integral part of all our courses so far. Many other sponsors including Coremed, Endotherapeutics and Medtronic have graciously supported the course with specialized equipment. Other sponsors have also been very kind in their support and provision of specialized protheses and grafts.

We appreciate their time and effort in providing for the course. We are grateful for their efforts and urge you to visit their tables to talk to the reps and make the most of their expertise.

Course Dinner

The course dinner is included in the registration fee and will be at Sails at Lavender Bay at 7pm on Saturday, September 21st. Sails on Lavender Bay is perched on the shore of Sydney's magnificent harbour with postcard views of the Sydney Opera House and Harbour Bridge. The menu offers what can be described as a contemporary interpretation of classic French cooking techniques, with an emphasis on fresh seafood. The award winning wine list comprises of over 130 Australian and International wine varietals.

www.sailslavenderbay.com

Conclusion

Endoscopic ear surgery represents one of the greatest steps forward in otology since the introduction of the microscope some 75 years ago.
This is an exciting time in otology. Parallels can be drawn with the introduction of the endoscope to sinus surgery and the tremendous success and uptake of that technique over the past decade.

The Royal North Shore Hospital Otolaryngology Department and Sydney Endoscopic Ear Surgery Research Group (SEES) welcomes all of the participants and we hope you thoroughly enjoy your time in Sydney.

A/Prof Nirmal Patel
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A/Prof Nicholas Jufas
drnicholasjufas@gmail.com

With thanks to our sponsors who made this course possible:

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Chapter 1

Endoscopic Ear Surgery

Nicholas Jufas, Nirmal Patel

INTRODUCTION

In recent years, a number of key technological advances have allowed the field of endoscopic ear surgery (EES) to progress significantly. These include the development of cooler light-emitting diode (LED) light sources, high-definition three charge-coupled device (3-CCD) cameras and improved optics in compact narrow scopes. The unique advantage of the endoscope is in the placement of the objective lens into the middle ear space, giving a broad width and depth of field to the visualization of middle ear structures. This view allows a better understanding of traditionally hidden anatomical recesses of the middle ear such as the sinus tympani, anterior epitympanum and protympanum. This anatomical knowledge, in turn, has allowed a more in depth analysis of the pathophysiology of middle-ear disease. The surgeon can then redefines and reimagine the surgical anatomy with a greater appreciation for the disease/anatomy interface during the procedure.

With more experience, EES allows a more comprehensive, directed surgical clearance in the middle ear in a less-invasive manner. The technique has also demonstrated itself as ideal for teaching, with simultaneous view of the operative field by the surgeon and trainee. Conversely, when the trainee is performing the surgery, the surgical mentor can closely supervise and follow their progress with the same view. In fact, all operating room staffs have the same image, encouraging interest and engagement throughout the operation.

In this chapter, endoscopic anatomy of the middle ear will be discussed, along with indications, advantages, disadvantages and instrumentation of the EES approach. Furthermore, suggestions for introduction of the technique into practice will be offered, as well as pitfalls and pearls.

ENDOSCOPIC ANATOMY OF THE MIDDLE EAR

The classic subdivisions of the middle ear are based on the medial projection of the tympanic membrane onto the medial wall of the middle ear, which is known as the mesotympanum. All surrounding areas are confluent with the mesotympanum: the protympanum is anterior, the epitympanum superior, retrotympanum posterior and hypotympanum inferior (Fig. 1.1).

In the last 10 years, these areas have been studied in depth with the visualization afforded by the endoscope. Key regions and structures contained within have been classified, and knowledge of these areas is essential to confident progression with the technique.

Mesotympanum

The defining feature of the mesotympanum is the promontory, covering the basal turn of the cochlea. The tympanic branch of the glossopharyngeal (Jacobson's) nerve leaves the inferior ganglion above the jugular foramen, traverses the inferior tympanic canaliculus to enter the middle ear through or just anterior to the finiculus and travels on the promontory. The nerve is the main contributing nerve to the tympanic plexus, which lies on the promontory. The plexus is usually

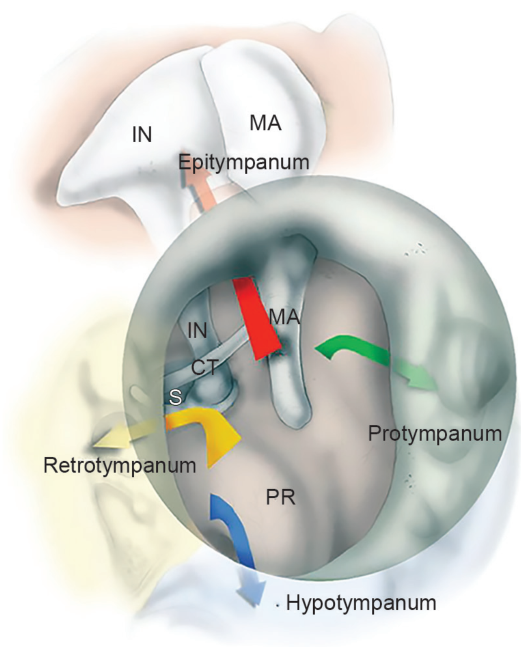


Fig. 1.1: Schematic diagram of mesotympanum, showing the confluent regions in a right ear. (CT: Chorda tympani; S: Stapes; IN: Incus; MA: Malleus; PR: Promontory)
Source: Prof Daniele Marchioni, University of Verona, Italy. Endoscopic Ear Surgery: Principles, Indications & Techniques (Presutti & Marchioni, Thieme, 2016).

submucosal, however, the nerves may lie deeper and groove the bone and rarely are embedded in the bone of the promontory. Jacobson's nerve provides sensation to the protympanum and Eustachian tube (ET) as well conveying parasympathetic fibers that arise in the inferior salivary nucleus to the parotid gland via the otic ganglion.[1]

Epitympanum

The epitympanum contains the head of the malleus as well as the body and short process of the incus. The malleus and incus form the epitympanic diaphragm, a concept introduced by Chatellier and Lemoine in 1946,[2] building on the 19th century work of Prussak.[3] The diaphragm also contains the three malleal ligamental folds (anterior, lateral and posterior) the posterior incudal ligamental fold, the tensor fold and the lateral incudomalleal fold. The latter two are duplicate folds and have variable position and orientation dependent on outgrowth of the sacci from the ET in utero, while the other composite folds envelop fixed ligaments limiting their variability[4] (Fig. 1.2).

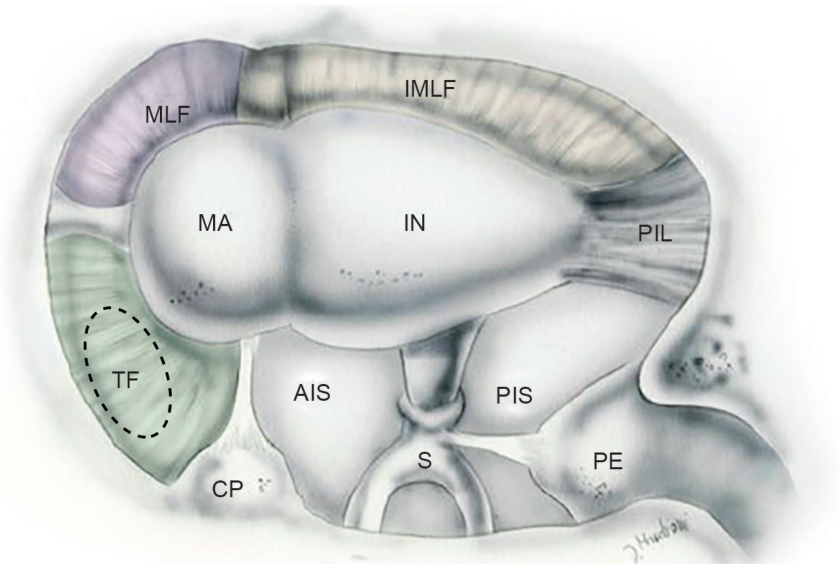


Fig. 1.2: The epitympanic diaphragm—with the main ventilation pathway through the anterior isthmus (AIS) and accessory pathways through the posterior isthmus (PIS). An alternate pathway exists in 25% of cases through an incomplete tensor fold (TF, dashed ellipse).
(S: Stapes; CP: Cochleariform process; MA: Malleus; IN: Incus; PIL: Lateral and medial posterior incudal ligaments; MLF: Lateral malleal fold; IMLF: Lateral incudomalleal fold; PE: Pyramidal eminence)
Source: Prof Marchioni—Endoscopic Ear Surgery: Principles, Indications & Techniques (Presutti & Marchioni, Thieme, 2016).

The ventilation route primarily travels through the epitympanic diaphragm via the isthmus. The main anterior isthmus is situated between the tensor tympani tendon anteriorly and the stapes posteroinferiorly, with a variable diameter between 1 mm and 3 mm. The accessory posterior isthmus is between the short process of the incus and the stapedial muscle (Fig. 1.3).

The tensor fold occupies a strategic position, separating the anterior epitympanic space of the epitympanum from the supratubal recess of the protympanum. This provides an alternate ventilation pathway from the middle ear to the mastoid, if present, however, studies have shown that it is only incomplete and therefore able to do so in around 25% of cases.[5]

There are two described configurations of the anterior epitympanic space: (1) type I (around 80%) consists of an oblique (and occasionally vertical) tensor tympani fold that creates a smaller anterior epitympanic space and the presence of a supratubal recess; (2) type II (around 20%) consists of a more horizontally placed tensor fold, which does not allow for the presence of a supratubal recess.[6]

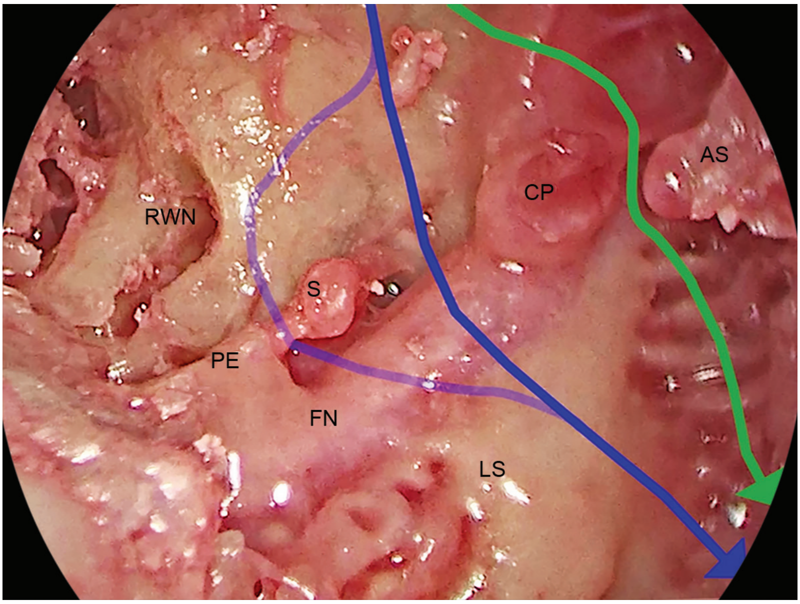


Fig. 1.3: Endoscopic view of left ear, with incus and malleus removed. Blue arrow indicates the main ventilation pathway from the Eustachian tube into the mastoid through the isthmus—with the dominant anterior pathway (solid blue) and accessory posterior pathway (faded blue) indicated. The alternate ventilation pathway through the tensor fold if incomplete is shown in green.
(S: Stapes; CP: Cochleariform process; PE: Pyramidal eminence; LS: Lateral semicircular canal; RWN: Round window niche; FN: Facial nerve; AS: Anterior spine)

Retrotympanum

The retrotympanum is the region posterior to the mesotympanum, confluent inferiorly with the hypotympanum and superiorly with the epitympanum. The retrotympanum is divided into superior and inferior divisions by the subiculum (Fig. 1.4). The region is best viewed with angled endoscopes either in the conventional-operating position or standing on the opposite side of the patient and rolling the patient toward the surgeon.

Except where indicated, the bony structures of the retrotympanum occur as a complete ridge of bone (type A), a bridge (type B) or are absent (type C), while the spaces have variable pneumatization (Figs. 1.5 and 1.6A to C).

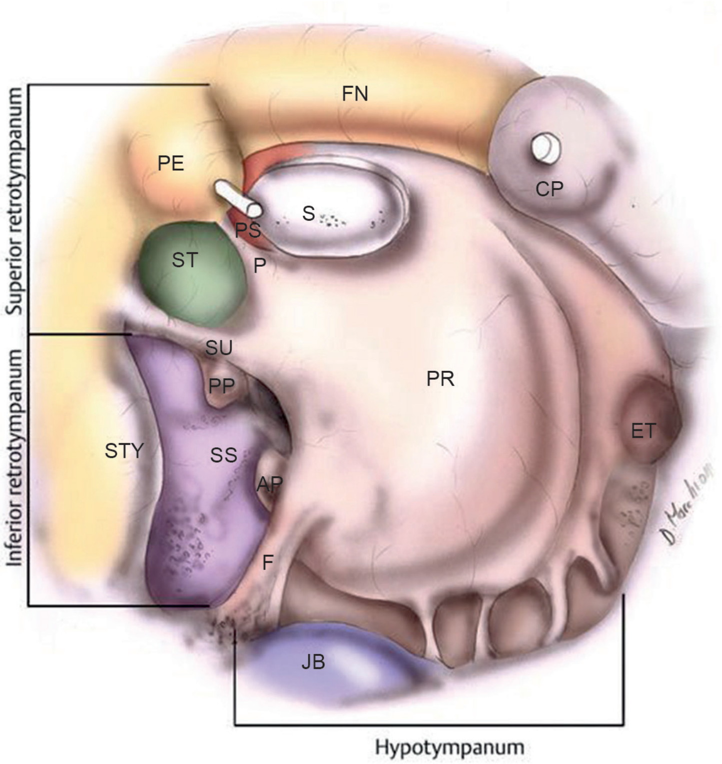


Fig. 1.4: Schematic diagram of the divisions of the retrotympanum with surrounding spaces also shown.
(PR: Promontory; S: Stapes; ET: Eustachian tube; F: Finiculus; AP: Anterior pillar; PP: Posterior pillar; JB: Jugular bulb; SS: Sinus subtympanicus; STY: Styloid complex; SU: Subiculum; P: Ponticulus; ST: Sinus tympani; PS: Posterior sinus; PE: Pyramidal eminence; FN: Facial nerve; CP: Cochleariform process)
Source: Prof Marchioni—Endoscopic Ear Surgery: Principles, Indications & Techniques (Presutti & Marchioni, Thieme, 2016).

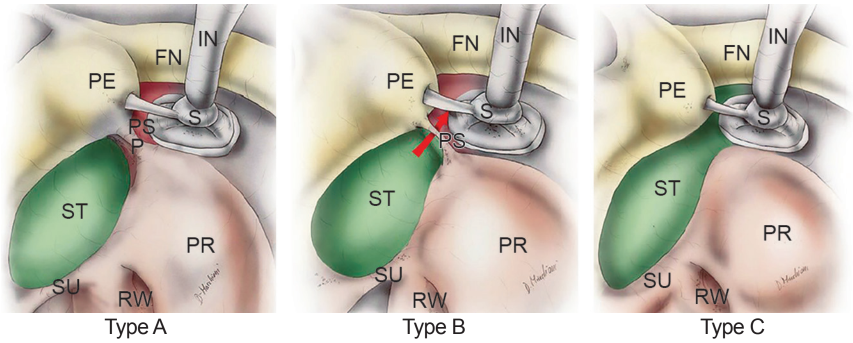


Fig. 1.5: Conformational variations on the ponticulus—type A (ridge), type B (bridge), type C (absent). Other bony ridges, such as the subiculum, finiculus and protiniculum all share the same classification.
(S: Stapes; IN: Incus; FN: Facial nerve; PE: Pyramidal eminence; ST: Sinus tympani; P: Ponticulus; PS: Posterior sinus; PR: Promontory; SU: Subiculum; RW: Round window)
Source: Prof Marchioni—Endoscopic Ear Surgery: Principles, Indications & Techniques (Presutti & Marchioni, Thieme, 2016).

Superior Division

Ponticulus: A bony structure from the pyramidal eminence to promontory.

Subiculum: A bony structure extending from the styloid eminence to the posterior pillar region of the round window (RW), separating both the superior and inferior retrotympanum and the sinus tympani from sinus subtympanicum.

Posterior sinus: A space medial to the stapedial tendon, posterior to the posterior crus of stapes, and separated from the sinus tympani by the ponticulus.

Subpyramidal space: A space medial to the pyramidal eminence and contiguous with the sinus tympani and/or posterior sinus.

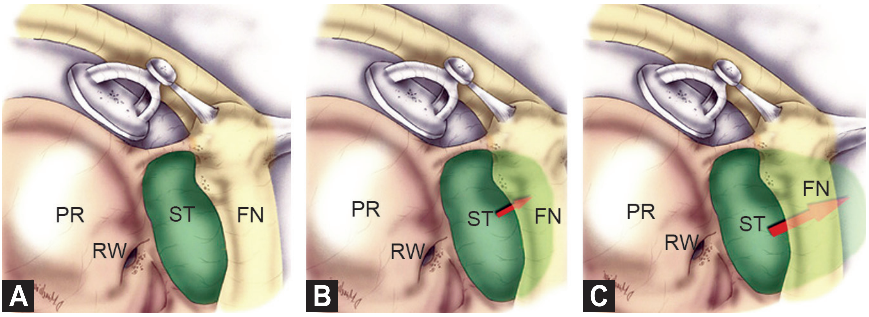
Sinus tympani: A space between the ponticulus and subiculum, classified relative to pneumatization medial to the facial nerve (Figs. 1.6A to C):

- *Type A*: Anterior to the facial nerve
- *Type B*: Medial to the facial nerve
- *Type C*: Posterior to the facial nerve (inaccessible via totally endoscopic methods).[7]

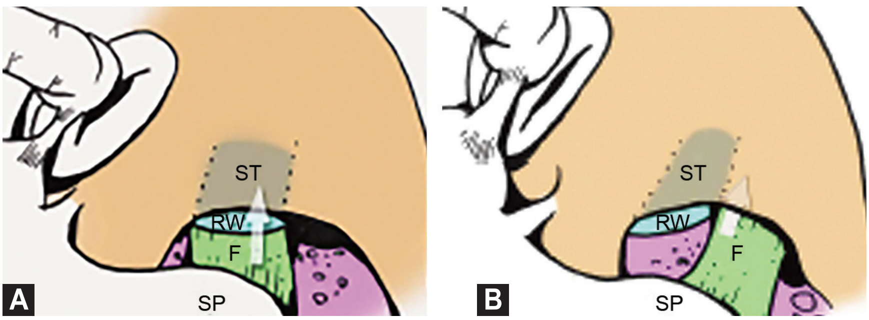
Inferior Division

Fustis: A thick club-like bony prominence, extending from the styloid eminence to the RW niche, forming the basal turn of the cochlea, with two conformations (Figs. 1.7A and B):

- *Type A*: Points directly to RW membrane, leads directly to scala tympani
- *Type B*: Points inferior to RW membrane, becomes floor of scala tympani.[8]



Figs. 1.6A to C: Conformational variations of the sinus tympani depth—(A) type A, shallow; (B) type B, deep; and (C) type C, very deep.
(PR: Promontory; RW: Round window; ST: Sinus tympani; FN: Facial nerve)
Source: Prof Marchioni—Endoscopic Ear Surgery: Principles, Indications & Techniques (Presutti & Marchioni, Thieme, 2016).



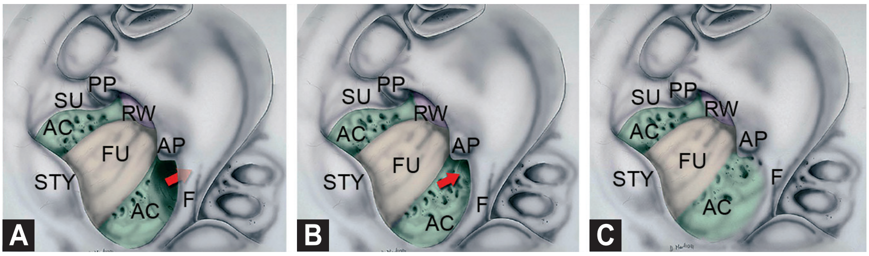
Figs. 1.7A and B: Schematic diagram depicting the two conformations of fustis (A) type A, and (B) type B.
(F: Finiculus; RW: Round window; ST: Scala tympani; SP: Styloid prominence).
Source: Marchioni D, Soloperto D, Colleselli E, Tatti MF, Patel N, Jufas N. Round window chamber and fustis: endoscopic anatomy and surgical implications. Surg Radiol Anat. 2016;38:1013-9.

Round window niche: A space covered by a bony tegmen that connects the posterior and anterior pillars, creating a niche in front of the RW membrane, which separates the middle ear from the scala tympani.

Finiculus: A bony structure variably containing Jacobson's nerve and/or the inferior tympanic artery, extending from the anterior pillar region to the floor of the hypotympanum and the region of the jugular bulb. This structure defines the inferior limit of the retrotympanum and separates it from the hypotympanum.[9]

Sinus subtympanicus: A space medial to the facial nerve and styloid prominence, contained by the subiculum superiorly and finiculus inferiorly.

Subcochlear canaliculus: A space of variable pneumatization extending between the fustis and anterior pillar/finiculus region toward the petrous apex (Figs. 1.8A to C).



Figs. 1.8A to C: Conformational variations of the subcochlear canaliculus—(A) type A, deep; (B) type B, shallow; and (C) type C, absent.
(AC: Aria concamerata; F: Finiculus; FU: Fustis; AP: Anterior pillar; PP: Posterior pillar; RW: Round window; STY: Styloid complex; SU: Subiculum).
Source: Prof Marchioni, from Marchioni D, Alicandri-Ciufelli M, Pothier DD, Rubini A, Presutti L. The round window region and contiguous areas. Eur Arch Otorhinolaryngol. 2015;272:1103-12.

- *Type A*: Deep tunnel extending inferiorly to the petrous apex
- *Type B*: Shallow tunnel easily accessible endoscopically
- *Type C*: No tunnel.[10]

Protympanum

The protympanum, previously referred to as the bony portion of the ET, is anterior to the mesotympanum, and confluent with the epitympanum superiorly, hypotympanum inferiorly and the cartilaginous ET anteriorly. It is bound by the tegmen tympani, tensor canal and supratubal recess superiorly, and the protiniculum inferiorly. The medial boundary is the lateral wall of the carotid canal, and the lateral boundary, the protympanic recess, is a bony wall separating it from the mandibular fossa (Fig. 1.9). Carotid canal dehiscence on the medial wall of the protympanum has been identified in up to 7% of temporal bones.[11]

The protympanum can have a quadrangular or triangular conformation, when viewed in a plane perpendicular to the long axis of the ET. Demarcation between the two types is based on whether the inferior wall is more (quadrangular) or less (triangular) than half the length of the superior wall in equivalent transverse dimension.

The tensor tympani canal either sits flat or rises on the superior boundary. When an area of pneumatization is inferomedial to the tensor tympani canal is called the subtensor recess (SbTR):

- *Type A*: Flat tensor canal and absent SbTR
- *Type B*: Raised tensor canal, shallow SbTR and easily visible fundus
- *Type C*: Raised tensor canal, deep SbTR, difficult to see limits of fundus.

Demarcation between a Type B and Type C SbTR is when the fundus of the SbTR extends superior to the midpoint of the tensor tympani canal.

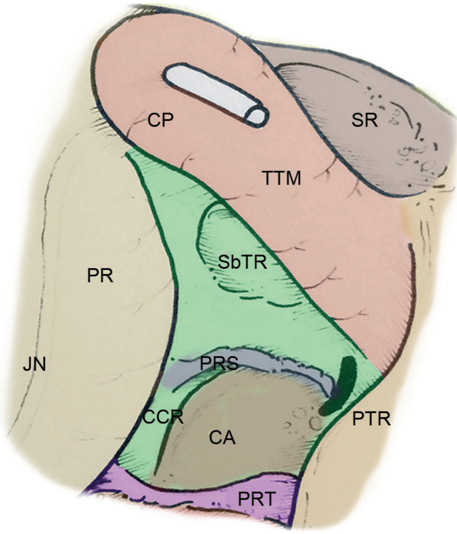


Fig. 1.9: Schematic overview of the protympanum in a right ear showing the relevant anatomical features.
(JN: Jacobson's nerve; PR: Promontory; CCR: Caroticocochlear recess; CA: Carotid artery canal; PRT: Protiniculum; PRS: Protympanic spine; PTR: Pretympanic recess; TTM: Tensor tympani canal; CP: Cochleariform process; SbTR: Subtensor recess; SR: Supratubal recess).
Source: Jufas N, Marchioni D, Tarabichi M, Patel N. Endoscopic anatomy of the protympanum. Otolaryngol Clin North Am. 2016;49:1107-19.

The protiniculum is a bony ridge commonly found extending from the promontory on the medial wall, across the inferior wall and merging with the lateral wall. It separates the hypotympanum from the protympanum and has three conformations (A: ridge, B: bridge and C: absent), similar to the bony ridges of the retrotympanum.[12]

The caroticotympanic nerves and arteries exit through channels in the bone overlying the carotid canal on the medial aspect of the protympanum. The caroticotympanic nerves carry sympathetic fibers posteriorly and cross the caroticocochlear recess, between the anterior aspect of the promontory and the carotid canal, to reach the tympanic plexus.

Occasionally, a rough spine of bony ridges or spicules is seen over the carotid artery prominence, called the protympanic spine. It is likely related to a fusion of the two laminae of the carotid canal in embryological development. If so, it may be indicative of a decreased likelihood of carotid canal dehiscence in the protympanum.[12]

Hypotympanum

The hypotympanum lies between the finiculus posteriorly and protiniculum anteriorly. It has variable pneumatization with air cells, which can track through into the petrous apex. The hypotympanic space can be restricted or even obliterated by a high-riding jugular bulb, which occurs in around 25% of cases.[13]

INDICATIONS AND APPLICATIONS

The endoscope should be considered an essential visualization tool for otologic surgery, complimentary to the microscope. Endoscopic and microscopic visualization should be utilized on a spectrum, which is dependent on patient, surgeon and disease factors. A proposed classification for EES, to facilitate comparison and discussion is:

- *Class 0*: Microscope only and no endoscope
- *Class 1*: Endoscopic used for inspection only
- *Class 2*: Mixed microscopic or endoscopic case
  - *Class 2a*: Endoscope used for less than 50% of dissection
  - *Class 2b*: Endoscope used for more than or equal to 50% of dissection
- *Class 3*: Endoscope only and no microscope [transcanal EES (TEES)].[14]

Cholesteatoma

Cholesteatoma of the middle-ear spaces represents the ideal situation to demonstrate the strengths of the endoscopic method. Cholesteatoma is usually subdivided into congenital and acquired forms (Boxes 1.1 and 1.2).

Box 1.1: Preoperative assessment for endoscopic ear cholesteatoma surgery.

- The lateral meatus and bony canal relative to disease location should be assessed to determine the largest endoscope size that can be used and whether a canal widening procedure (meatoplasty or canaloplasty) is required.
- The status of middle ear—whether inflamed or not will help to assist the surgeon in determining the need for preoperative topical therapy and microscopy during some of the middle ear work.
- Fine cut computed tomography of the temporal bones is essential in preoperative evaluation of cholesteatoma to determine the extent of disease. With the limitations of current instrumentation, a mastoidectomy will be required, if disease extends into the mastoid well beyond the posterior aspect of the lateral semicircular canal or into a deep type C sinus tympani.
- A non-echo-planar diffusion-weighted magnetic resonance imaging (non-EPI DWI MRI) is useful to determine mastoid, intralabyrinthine and intracranial spread. Caution is required regarding negative prediction rates in discharging ears and with disease less than 4 mm.

Box 1.2: Optimal disease and patient characteristics for endoscopic ear surgery.

- Congenital cholesteatoma.
- Acquired cholesteatoma in mesotympanum and confluent areas (especially retrotympanum, protympanum and anterior epitympanum), with minimal mastoid extension.
- Sclerotic mastoid.
- Low tegmen.

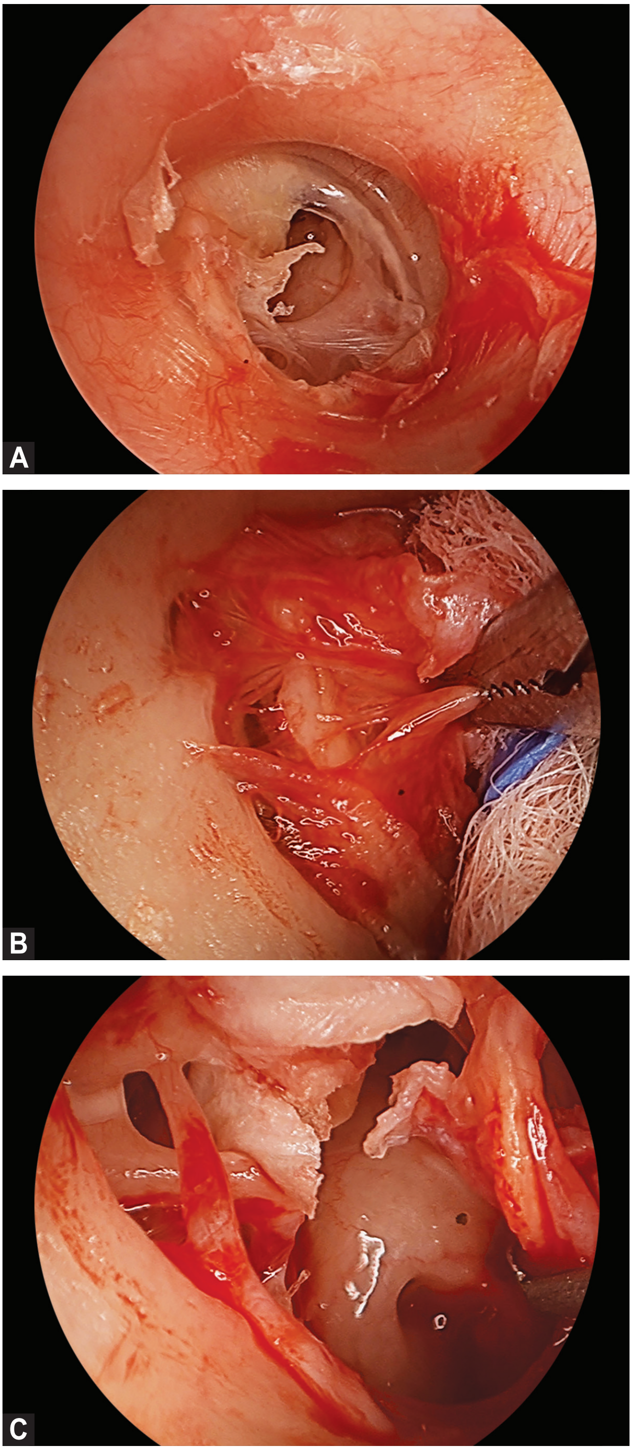
The congenital form, in general, is more common in children and usually presents in a subspace of the mesotympanum, the most common site being anterosuperior. Using an angled endoscope the surgeon can directly visualize and elevate an inferior based tympanomeatal flap in the restricted confines of a pediatric external ear canal. Furthermore, the surgeon can directly see the disease in front of the handle of the malleus and anterior tensor tympani tendon, where this is typically blind dissection with the microscope. In most cases, little to no bone removal is required.

Acquired cholesteatoma most commonly arises in Prussak's space, the attic or mesotympanum. In all these instances, the disease, as it spreads from its origin tends to migrate into the subspaces of the middle ear including difficult to visualize regions. The endoscope allows the surgeon to trace the disease, with direct and complete visualization, from its origin to endpoint (Figs. 1.10A to F). This provides a different mindset to the microscopic approach, which in these scenarios requires drilling a significant amount of normal external canal and mastoid bone to access the disease from posterior to anterior.[15] Often with the combination of carbon dioxide (CO_2) or potassium titanyl phosphate (KTP) laser, ossicular-preserving surgery is possible with minimal to no bone removal.[16]

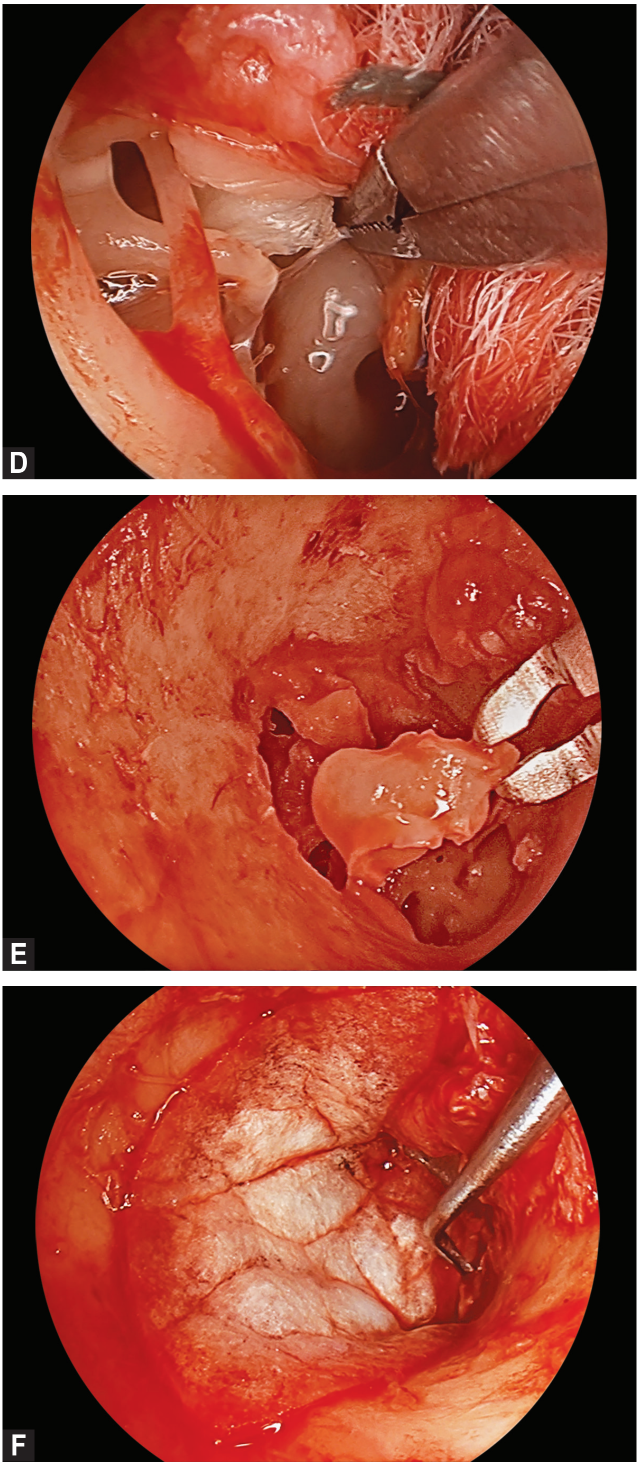
Visual access to certain regions of the middle ear, such as the retrotympanum, protympanum and lateral anterior epitympanum, is difficult using the microscope.[17] Using the endoscope nearly all the spaces and subspaces of the middle ear can be completely visualized with the exception of a very deep type C sinus tympani and type A subcochlear recess, allowing the surgeon a greater confidence with disease removal (Figs. 1.11 and 1.12).

Tympanoplasty

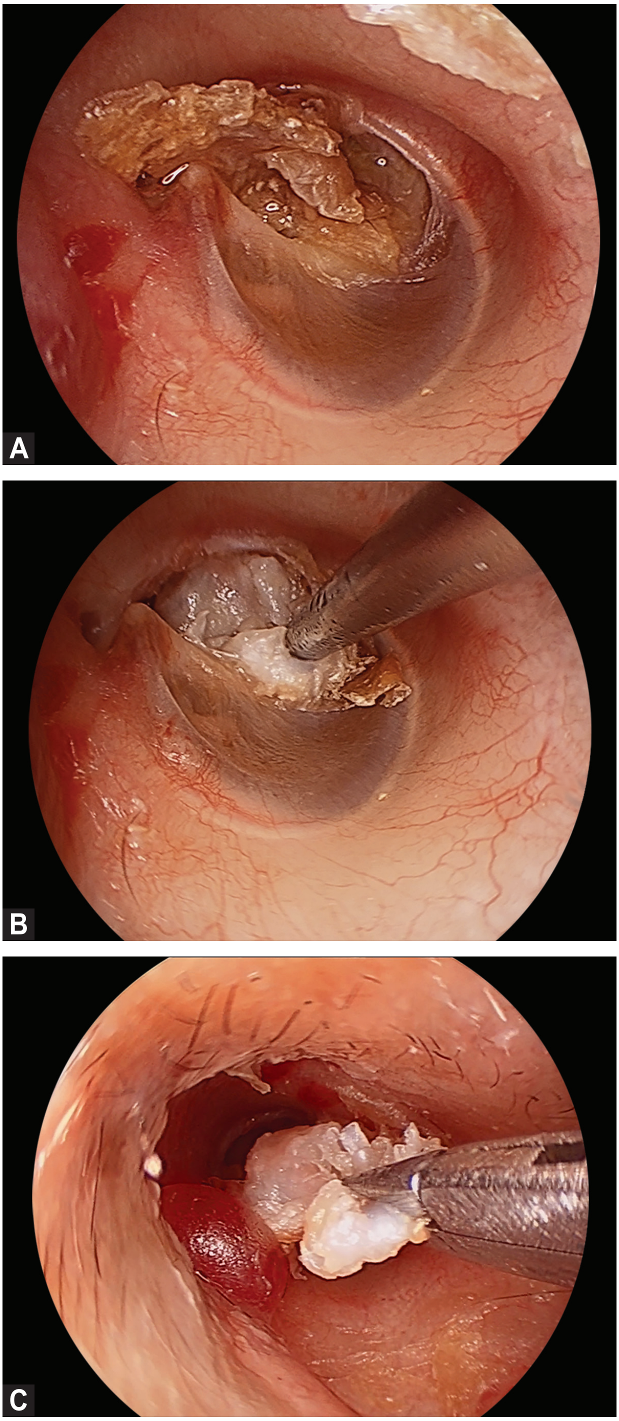
The visualization that the endoscope offers can be used to significant advantage in myringoplasty and ossicular reconstruction. Total or subtotal perforations often have an anterior margin that is difficult to see



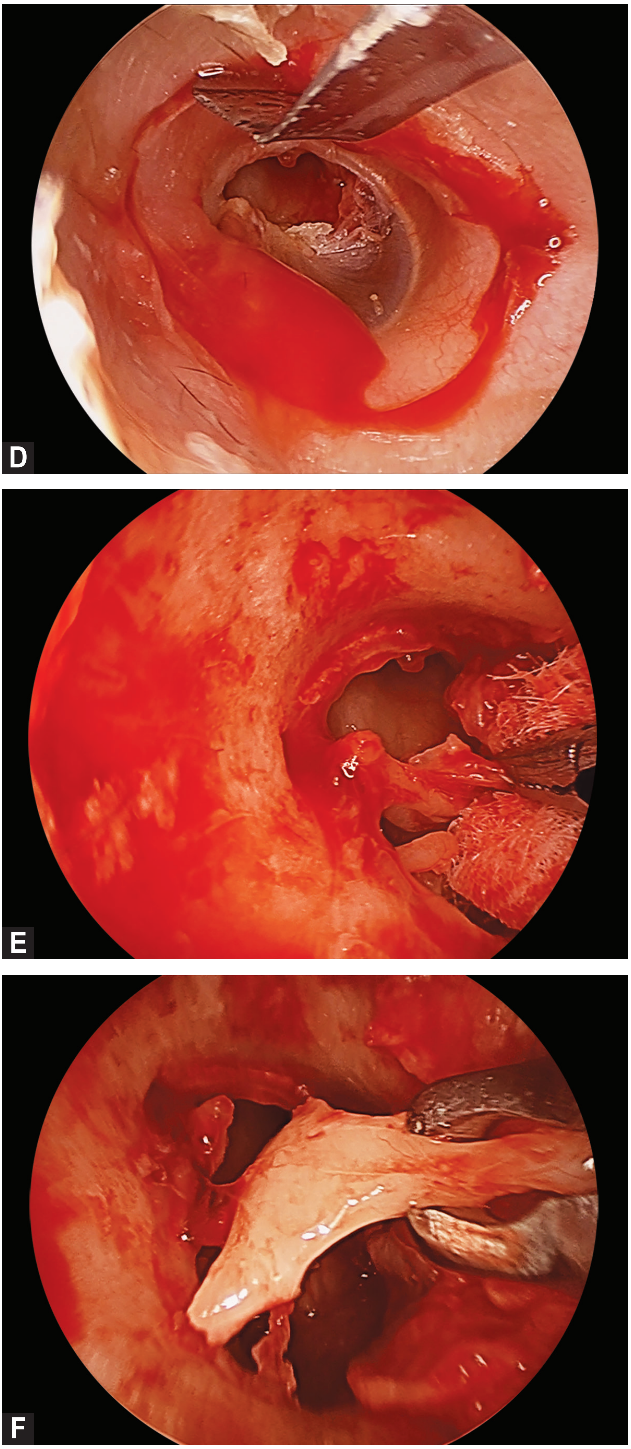
Figs. 1.10A to C: Endoscopic dissection of en plaque cholesteatoma in a right ear. (A) Initial endoscopic transcanal view. (B) Lifting and separating the tympanomeatal flap. (C) Cholesteatoma visible on the incus.



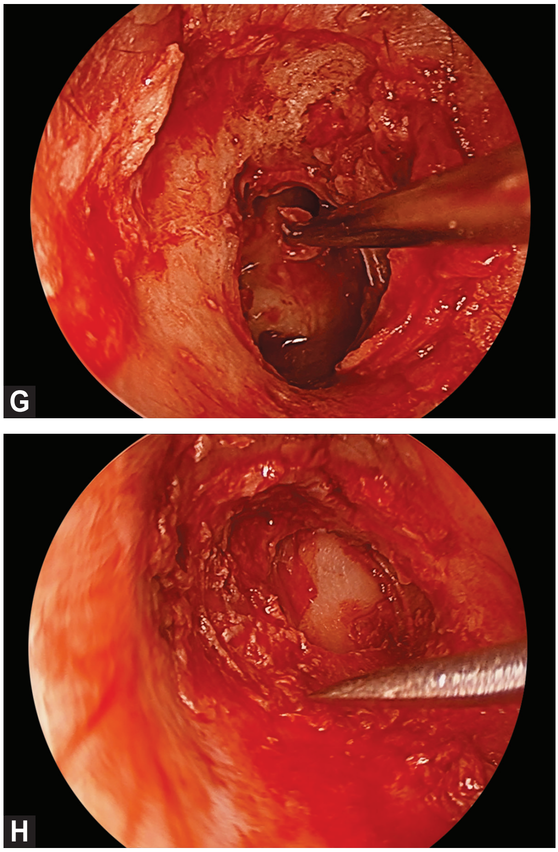
Figs. 1.10D to F: (D) Dissecting cholesteatoma off the incus. (E) Removal of the malleus head. (F) Laying in the biodesign graft.



Figs. 1.11A to C: Endoscopic protympanic cholesteatoma removal in a right ear. (A) Initial endoscopic transcanal view. (B) Decompression of cholesteatoma with suction. (C) Removal of further cholesteatoma sac contents with forceps.



Figs. 1.11D to F: (D) Transcanal incision to raise tympanomeatal flap. (E) View of mesotympanic and protympanic spaces. (F) Removal of incus.



Figs. 1.11G and H: (G) Removal of protympanic cholesteatoma. (H) Replacement of tympanomeatal flap after graft.

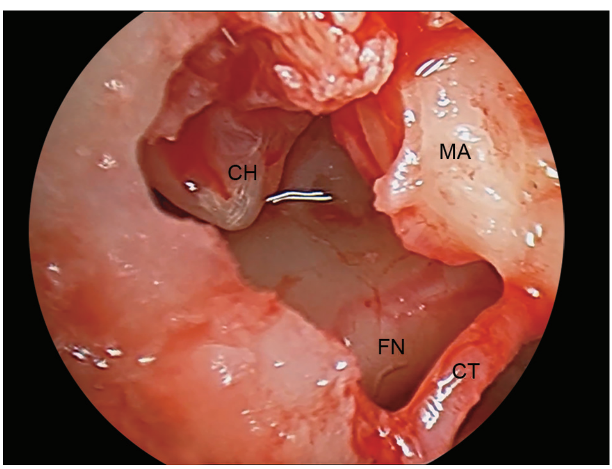
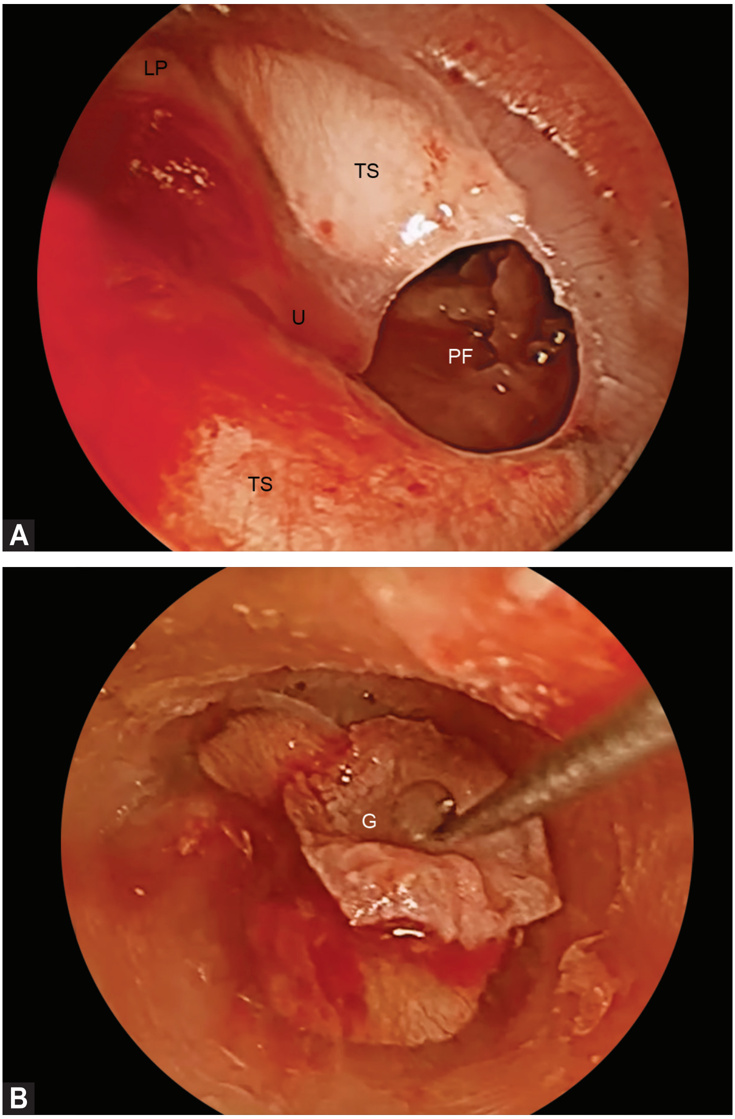


Fig. 1.12: Endoscopic view of cholesteatoma sac in right ear medial to ossicles in epitympanum after atticotomy and removal of incus.
(CH: Cholesteatoma; MA: Malleus head; FN: Facial nerve; CT: Chorda tympani)

using a microscope with a bony canaloplasty. Small and limited anterior perforations can also be easily visualized and repaired (Figs. 1.13A to C).

Using angled endoscopes, the anterior margin of the perforation can be easily seen and when placing underlay grafts the medial aspect of the perforation is also accessible to ensure that no epithelium has migrated medially and graft placement is appropriate. Additionally



Figs. 1.13A and B: Endoscopic composite cartilage button tympanoplasty in a right ear. (A) Preoperative image showing tympanosclerotic plaques within the fibrous layer of the tympanic membrane abutting the perforation. (B) Intraoperative image showing graft being placed and locked into position with an instrument.

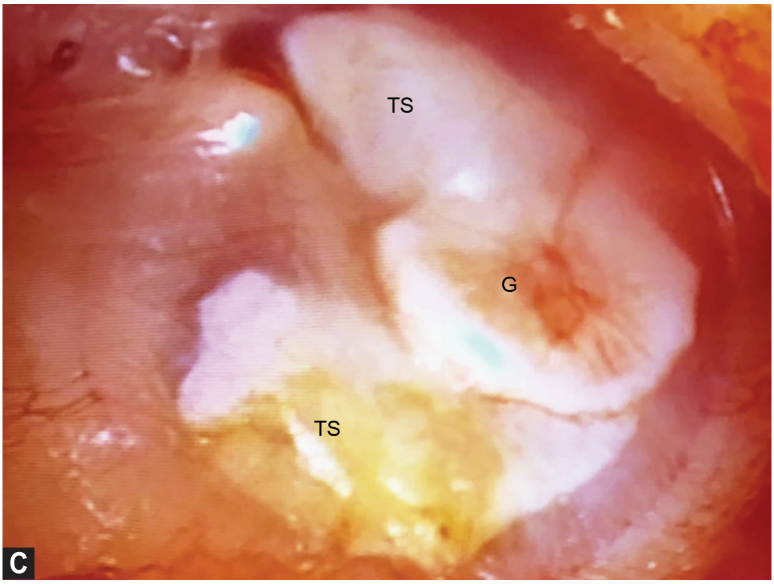


Fig. 1.13C: (C) Postoperative image showing incorporation of the cartilage graft and full epithelialization.
(LP: Lateral process of malleus; TS: Tympanosclerotic plaque; U: Umbo; PF: Perforation; G: Graft).

in ossiculoplasty, prosthesis placement may be aided by the improved view particularly with the respect to the anterior crus of the stapes and using the RW reflex to check coupling (Fig. 1.14).

Stapes Surgery

Stapedotomy and stapedectomy have been performed using the endoscope, demonstrating similar audiometric and postoperative outcomes to the microscopic method.[18] The loss of one hand in operating is certainly a challenge and this technique is generally reserved for the experienced endoscopic ear surgeon. The theoretical advantages here include a reduction in chorda tympani movement and therefore potentially less dysgeusia. Secondly, the anterior crus can be more directly visualized and often directly observed and manipulated with a hook or laser to reduce trauma in stapes superstructure removal (Figs. 1.15 and 1.16).

Cochlear Surgery

Cochlear implantation has been performed via postauricular and permeatal approaches with no mastoidectomy in several centers.[19]

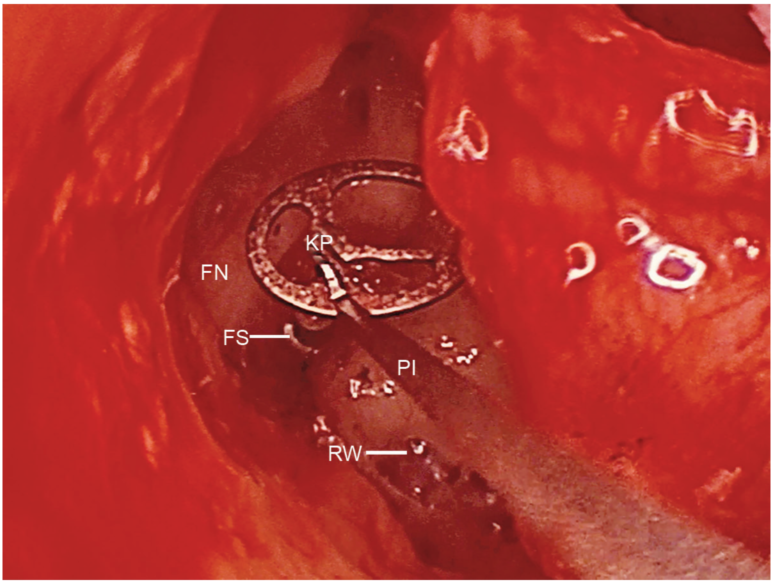


Fig. 1.14: Endoscopic view of right ear, checking coupling of a total ossicular prosthesis with footplate shoe by palpating and checking round window reflex.
(FN: Facial nerve; PI: Pick; KP: Kurz-Variac total ossicular prosthesis; FS: Kurz omega connector; RW: Round window).

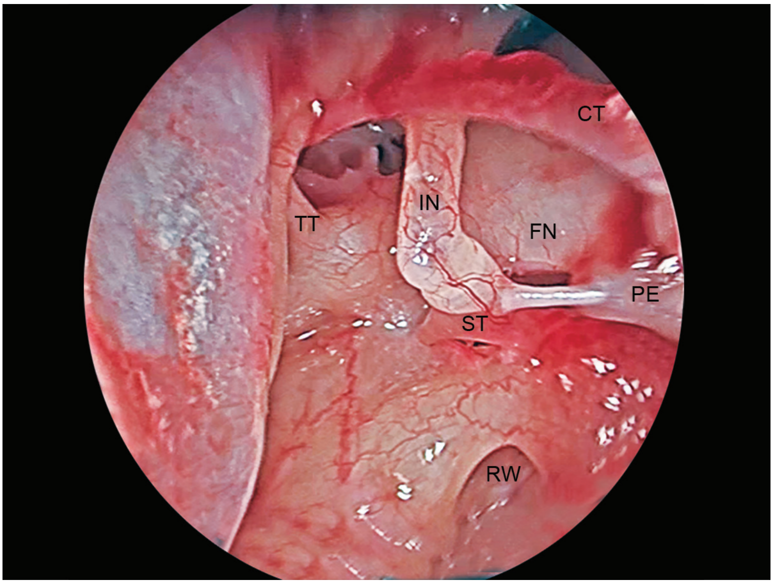
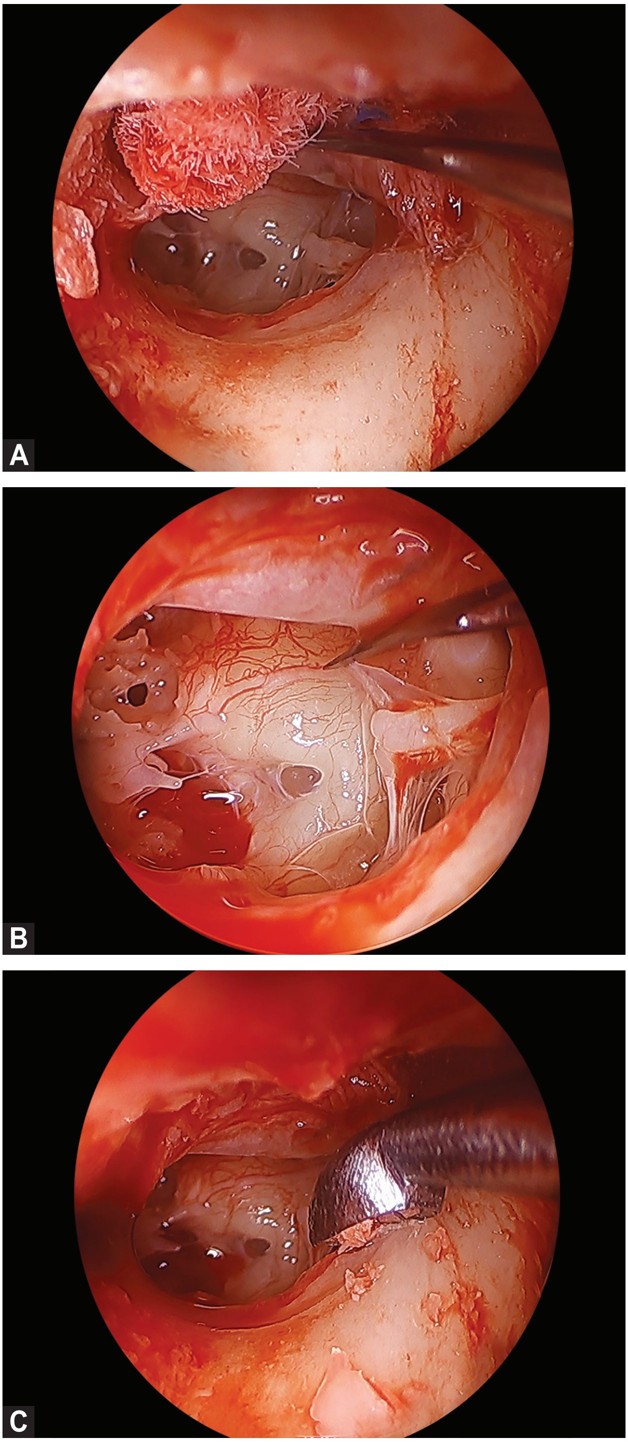
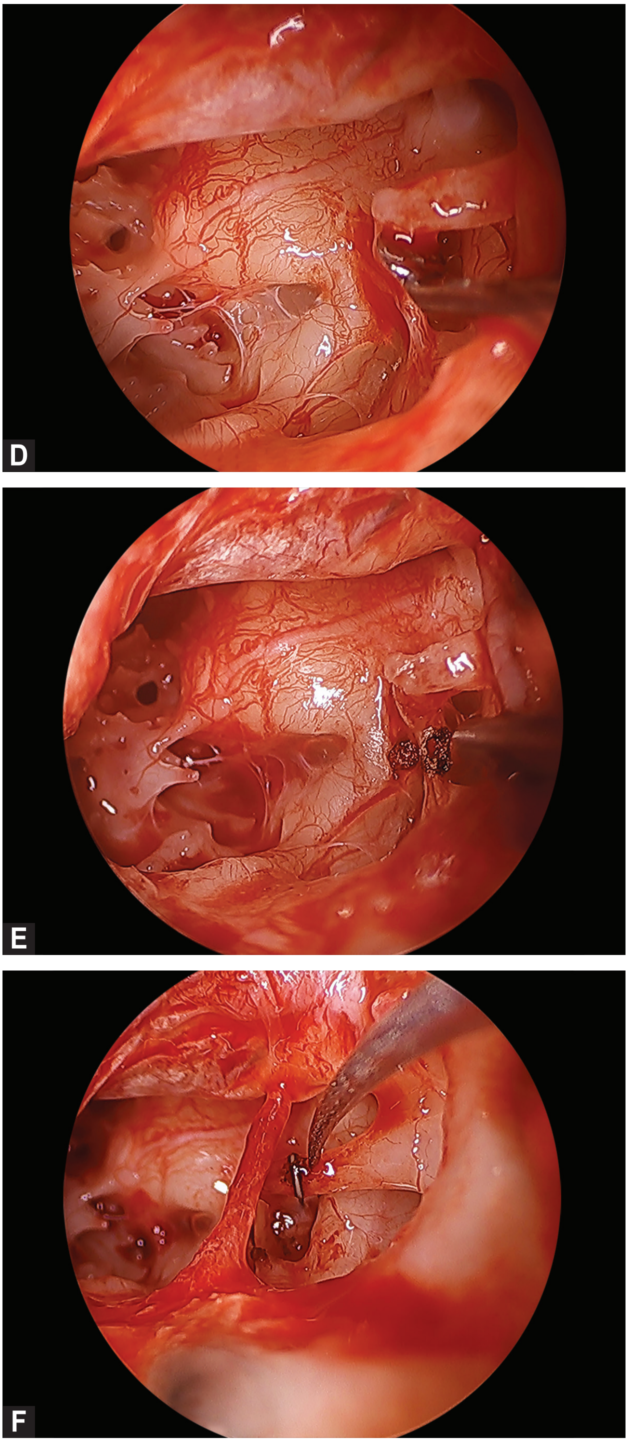


Fig. 1.15: Endoscopic view of left ear during stapedotomy. Note the fenestral and cochlear otosclerosis easily visualized with the angled endoscope.
(IN: Incus; ST: Stapes; FN: Facial nerve; PE: Pyramidal eminence; RW: Round window niche; CT: Chorda tympani; TT: Tensor tympani)



Figs. 1.16A to C: Endoscopic stapedotomy performed transcanal on left ear: (A) Lifting the tympanomeatal flap. (B) Division of adhesions around incudostapedial complex. (C) Minimal curettage of bone for access.



Figs. 1.16D to F: (D) Division of incudostapedial joint. (E) CO_2 laser used to obliterate posterior crus. (F) Checking coupling and placement of prosthesis prior to lowering tympanomeatal flap.

The purpose of this technique is to avoid the mastoidectomy, however, the method requires elevating a tympanomeatal flap and may leave the electrode exposed under the skin of the external ear canal. A more conventional use of the endoscope is to aid in viewing the RW and basal turn of the cochlea anatomy through a posterior tympanotomy, when inserting the electrode, particularly when challenging anatomy is present, such as malformation or far-advanced otosclerosis. Endoscopes have also been utilized in a transcanal and transpromontorial corridor to remove intracochlear disease, such as schwannoma.[20]

Eustachian Tube Surgery

Disease Clearance

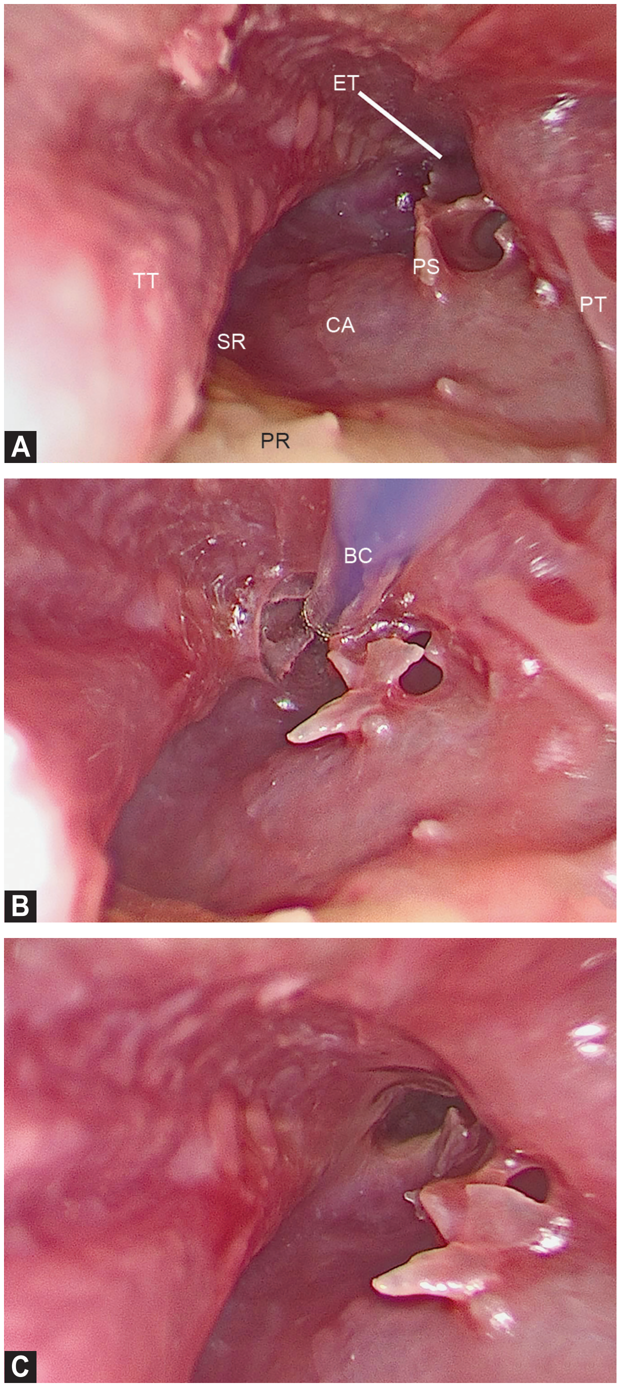
Endoscopes allow a complete view of the all the boundaries of the protympanum. Cholesteatoma in this region is uncommon, but may occur with the congenital form of the disease, extensive mesotympanic spread or anterior epitympanic disease and an incomplete tensor fold. With the utilization of angled endoscopes and instruments, granulation and cholesteatoma can be removed in a deliberate and methodical fashion.

Eustachian Tube Dilatation

Transnasal endoscopic balloon dilatation has been performed for many years for intractable ET dysfunction with moderate success. In larger studies, there is a failure rate of at least 30%.[21,22] The narrowest portion of the ET is typically just lateral to its junction with the protympanum and this region is often not reached with the transnasal approach.[23] To overcome the limitations of transnasal approaches a longer balloon or a more lateral insertion would be required. Given the variable anatomy in the region and the potential risk to the carotid artery, this has been avoided. Direct transtympanic dilation may be an approach to overcome the deficiencies of the transnasal approach in a potentially safe manner,[24] particularly in cases of repeated failure with the transnasal approach (Figs. 1.17A to C). Furthermore, the ear is often already being operated upon for associated chronic ear disease.

Patulous Eustachian Tube Management

Transtympanic stenting of the ET via the protympanum has been performed for the patulous ET. The rationale is that the flange of the stent can remain in the protympanum, avoiding prolapse and extrusion of the stent.[25] The technique has the disadvantage of requiring a tympanotomy and potential dissection around the ossicles.



Figs. 1.17A to C: Endoscopic view of protympanum on right ear, (A) before (B) during and (C) after balloon dilatation of the Eustachian tube.
(ET: Eustachian tube lumen; TT: Tensor tympani canal; SR: Subtensor recess; PR: Promontory; CA: Carotid artery; PS: Protympanic spine; PT: Protiniculum; BC: Balloon catheter)

Facial Nerve

Angled endoscopes have been used to study and treat pathology of the facial nerve. Using a totally transcanal approach the entire tympanic segment of the facial nerve can be exposed without disturbing the otic capsule. Expanding this approach in a transpromontory manner allows for the facial nerve to be exposed from the root entry zone to the mastoid segment. Such approaches have been used to manage perigeniculate cholesteatoma and facial hemangioma; the method could be extended for other pathologies such as management of facial nerve injury.[26]

Lateral Skull Base Surgery

Suprageniculate Approach

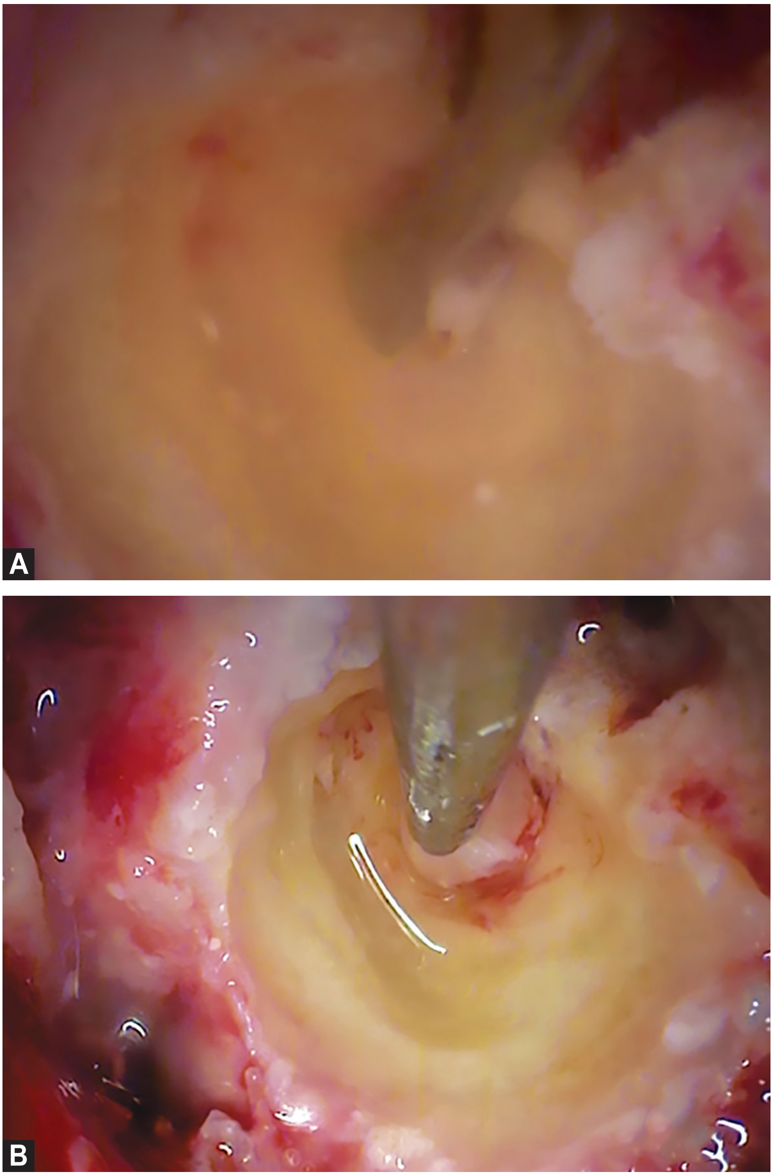
This approach is possible when disease is contained by the facial nerve, labyrinthine bloc and middle cranial fossa. It allows access to the geniculate ganglion and perigeniculate area. Bone can be removed from the fallopian canal, tegmen tympani, as well as in front of the labyrinthine bloc. This approach is most commonly used in managing cholesteatoma extension in the region, where following removal of the disease, the space can be obliterated. It also preserves sensorineural hearing by working above the cochlear and labyrinth.[27]

Transcanal Transpromontorial Approach

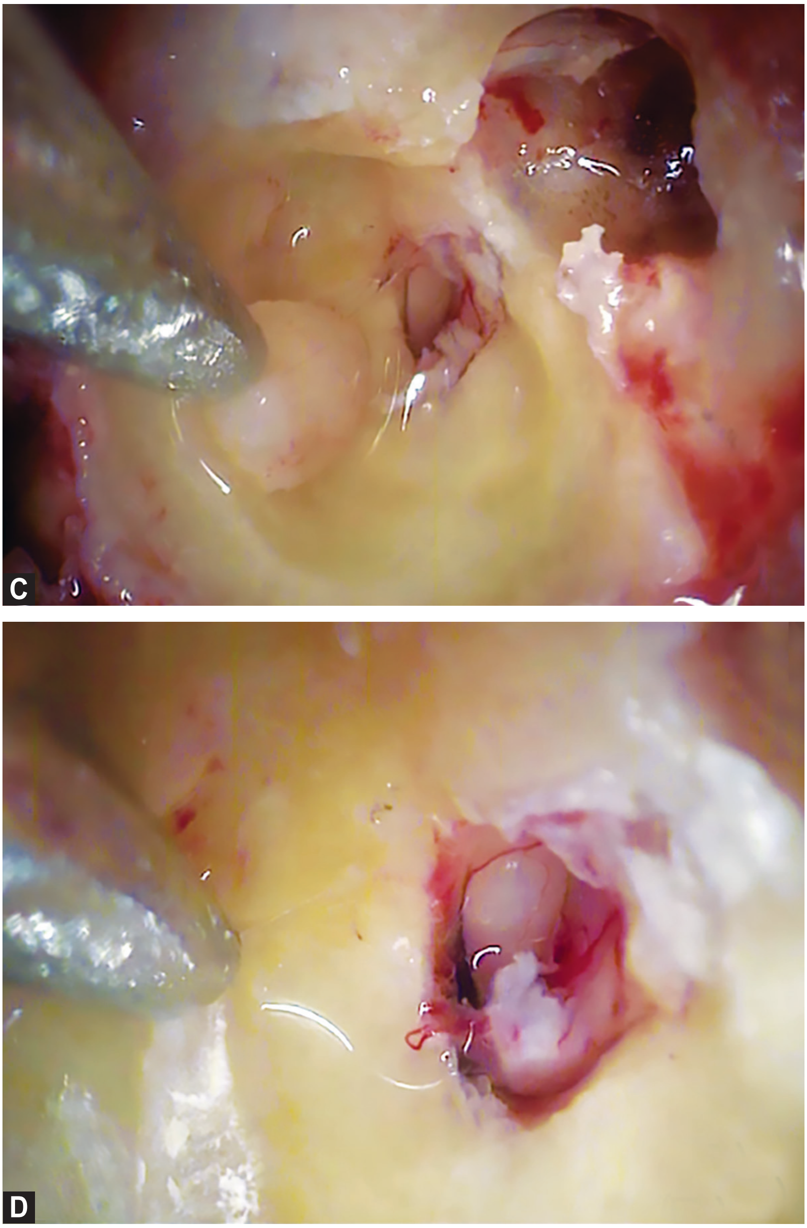
This approach, first described by Marchioni and Presutti, is a totally endoscopic technique that involves the removal of the cochlea to gain direct access to pathology in the cochleovestibular region and internal acoustic canal (IAC). The approach involves direct lateral to medial access to the IAC, without manipulation of the dura or meninges. The bony defect is plugged with fat and the tympanic membrane and skin replaced. The approach has not only been used to remove smaller cochlear schwannomas, but also larger vestibular schwannomas of the IAC[28] (Figs. 1.18A to D). Owing to the minimal invasiveness of the technique, it often provides a postoperative course that is shorter and not requiring intensive care. An extended approach that involves an endaural incision and combination endoscopic/microscopic technique has also been described, allowing for lesions up to 1.5 cm to be safely removed by exposing the whole length of the IAC.[29]

Infracochlear Approach

In this approach, bone is extensively removed after raising a tympano-meatal flap, starting along the inferior and posterior annulus, as well as over the carotid artery and jugular bulb, inferior to the cochlear. This allows for the infracochlear regions to be followed to the petrous apex. The technique is important to know in managing cholesteatoma in this region as well as a method for draining cholesterol



Figs. 1.18A and B: Endoscopic images of a transcanal, transcochlear approach to the internal auditory canal (IAC) to remove a small intracanalicular vestibular schwannoma in this left ear. (A) Piezoelectric device to remove cochlear bone. (B) Suction on the tumor within the IAC.



Figs. 1.18C and D: (C) Continued suction to remove tumor from IAC. (D) Transcochlear view into the IAC after tumor removal, showing the cochlear and facial nerves, and residual vestibular nerve fibers.
Source: Prof Marchioni—Endoscopic Ear Surgery: Principles, Indications & Techniques (Presutti & Marchioni, Thieme, 2016).

granuloma. Although, this approach had previously been described microscopically,[30] the endoscopic facilitates access often with less bone removal.[27]

Endoscope-assisted Vestibular Schwannoma Surgery

Angled endoscopes have been used in conjunction with traditional suboccipital approaches to remove IAC extensions of either vestibular schwannoma or meningioma.[31] The concept is to reduce drilling of the porus acusticus, which may in turn reduce operative time and cerebrospinal fluid leak rates. Fully endoscopic resection of vestibular schwannomas via standard, but smaller craniotomies has also been reported.[32]

Endoscope-assisted Superior Canal Dehiscence Management

Angled endoscopes have been used in conjunction with middle fossa craniotomy to limit temporal lobe retraction and reach medial or anterior aspects of superior canal dehiscence.[33] The endoscopic method using mini-craniotomy to access the superior semicircular canal has also recently been published.[34]

ADVANTAGES OF ENDOSCOPIC EAR SURGERY

Lack of External Scar

Traditional microscopic ear surgery requires incisions within the ear canal incorporating the tympanic membrane, creating a tympanomeatal flap. This incision is generally made in conjunction with a large postauricular incision, or less commonly endaural. With transcanal endoscopic surgery, the tympanomeatal flap is placed back into position, healing remarkably well, often with no evidence of scar, even on otoscopy. A less than 1-cm incision is often made behind the tragus to take a cartilage or perichondrial graft—this incision is small and well hidden.

Faster Operating Time and Reduced Cost

Once the learning curve for EES is achieved, then there is generally a reduction in operating time due to the lack of a postauricular incision, canaloplasty and mastoidectomy. This reduction in time along with the reduction in burr use reduces the cost of ear surgery compared to traditional methods (Fig. 1.19).

Less-deforming Surgery

Many major ear surgeries involve the creation of a canal wall down or modified radical mastoid cavity. This is disfiguring due to the removal of cortical bone and the posterior canal wall creating a collapse of the postauricular skin, displacement of the pinna, and a visible and palpable divot in the bone. This bone removal is not required with the EES approach, so the method is less disfiguring.

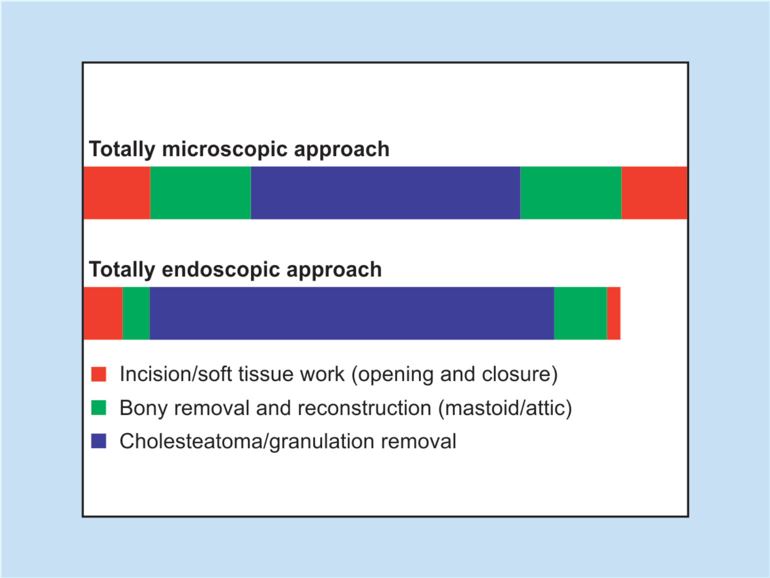


Fig. 1.19: Comparison of times taken for the various stages of surgery for an extensive cholesteatoma that can be approached by either a totally microscopic or totally endoscopic approach. Timings shown are indicative only for a surgeon that has achieved proficiency in both approaches.

Better Visualization

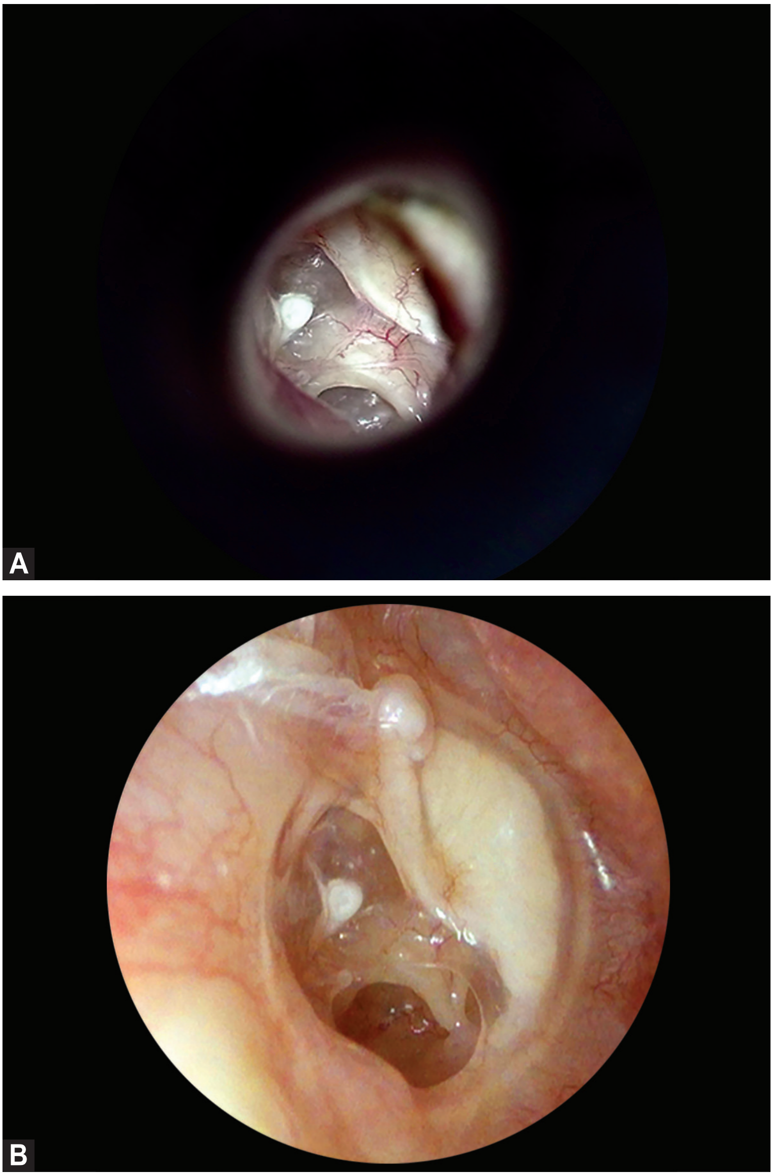
Endoscopy provides a field of view that significantly larger. With the light source delivered to the tip of the endoscope, illumination even on low levels is excellent, and angled scopes allow vision around corners. In contrast, the microscope relies on direct line-of-sight, which means that three key areas in the middle ear—retrotympanum, anterior epitympanum and protympanum—are not seen well with a microscope, even with surrounding bone removed. They can easily be seen by endoscopy, often without any bone removal, and sometimes even without using an angled endoscope (Figs. 1.20A and B).

Less Pain and Faster Healing

A significantly smaller incision, plus decreased normal tissue and bone removal, result in less pain—leading to decreased/nil postoperative analgesia. Healing time is faster and hospital stay, in the author's experience, reduced to day-only.

Potentially Better Function

Extensive mastoid surgery may result in patients having their lateral semicircular canal exposed. Cold water (or air) enters the ear canal



Figs. 1.20A and B: Comparison of (A) microscopic view through a speculum, and (B) endoscopic view of the same right ear. Erosion of the long process of the incus, a myringostapediopexy, a posteroinferior perforation and anterior myringosclerosis are all clearer seen through the endoscopic view.

causing profound, disabling vertigo. Canal wall down mastoid surgery is often not self-cleaning, and patients require lifelong ENT surgeon surveillance/toilet. EES avoids both of these drawbacks, likely better preserving the ear's innate function.

Improved Teaching

It is difficult for others to obtain the same view that the primary surgeon does in microscopic ear surgery. Sidearms and television screen outputs often deliver an inferior view. In EES, all present in the room share the same view as the primary surgeon. Teaching, therefore, transpires as the view and detail from EES simultaneously engages all in the operating theater. It also allows the supervisor an equal view to the trainee to closely follow their progress.

DISADVANTAGES AND LIMITATIONS OF ENDOSCOPIC EAR SURGERY

Proof of Concept

Conceptually, the benefits of EES are easy to understand, the evidence for the short-term results on quality-of-life, including healing time and postoperative pain, as well as long-term results regarding disease recurrence and stable hearing are still required.

Loss of One Hand

This is one of the biggest downsides of endoscopy. Microscopic work allows two hands to work simultaneously—needed during periods of excess bleeding or for delicate two-handed manipulation. Specifically designed suction dissection instruments do help, but using the microscope for this task remains the better option at present.

Loss of Three-dimensional Perception

The binocular microscope can see in three dimensions. The endoscope cannot being essentially monocular with current technology. The endoscope does have two features giving a simulated three-dimensional experience—haptic feedback between scope-holding and operating hand as well a number of optical effects, which allow the brain to perceive three dimensions in a two-dimensional image, including: linear perspective, occlusion, shadowing, texture and prior knowledge.

Optimum Instrumentation

Using the presently developed instruments can at times be frustrating, as the right tool for the job has not yet been invented. More specific and a greater range of instrumentation will come with time. Instruments combining suction and dissection exist and are being refined. Analogous to EES, but ahead by more than 20 years, improved instruments in functional endoscopic sinus surgery are still being developed.

Retraining

While the endoscope offers improvements, the amount that can be done with the endoscope versus the microscope is minimal. As a result, established surgeons are more likely to use the microscope, rather than adopt new technology for minimal gain. Eventually, if patient experience and long-term results prove the expected advantages, further uptake will ensue.

INSTRUMENTATION IN ENDOSCOPIC EAR SURGERY

Dedicated Instruments

There are two primary manufacturers of dedicated and specific endoscopic ear instruments: Karl Storz GmbH and Spiggle & Theis Medizintechnik GmbH. The instruments they make are varied, unique and complement each other well (Figs. 1.21A and B). Karl Storz has a number of curved suction instruments of varying lengths and diameters, and specific dissection instruments. Spiggle & Theis, on the other hand have suction capability built into all of their instruments, which have varying dissection tips. As previously stated, newer and improved instruments will no doubt be created in coming years.

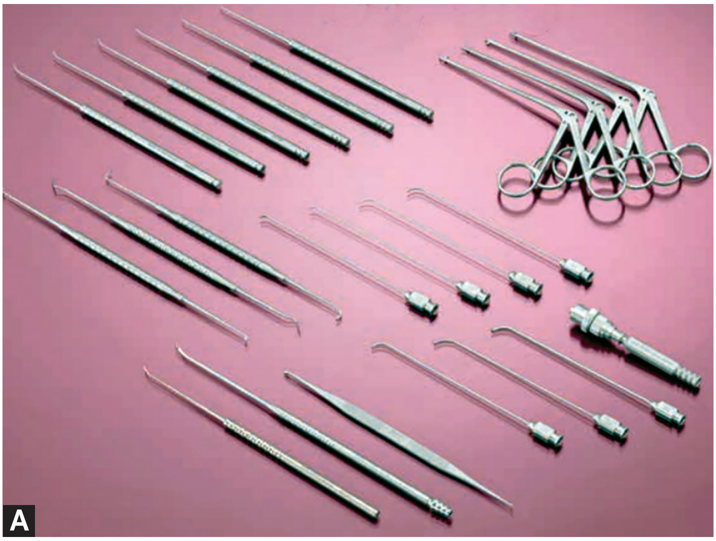
Endoscopes

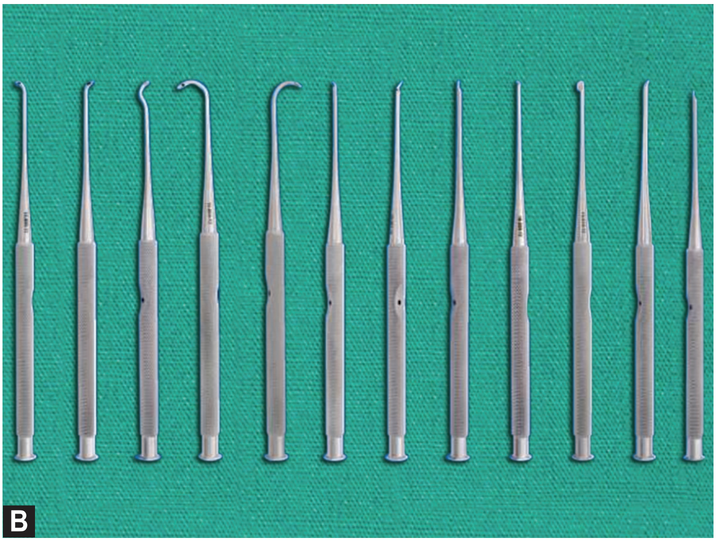
There are a number of manufacturers of rigid Hopkins rod-lens endoscopes. They come in a variety of diameters, but for EES, the most useful of these are 2.7, 3 and 4 mm. Better picture quality and size is obtained with a larger diameter scope, so the best one to use is the largest one that can fit into an ear canal. The 3-mm diameter appears to give the best compromise on image and ability to fit in most ear canals.

They also come in a variety of lengths with around 14 cm being the standard for most EES scopes. This size was chosen as it gives a good compromise between a longer scope, which is too unwieldy and a shorter scope, which may clash with the operating instrument in the dominant hand. For more lateral work, such as simple tympanoplasty and ossiculoplasty, a shorter scope may prove feasible.

Cameras

There are two main types of camera sensors: charged-coupled device (CCD) and complementary metal-oxide semiconductor (CMOS). Traditionally, CCD was the better of these technologies in terms of picture quality, color reproduction and performance in low light. The technology behind CMOS is now dramatically and quickly improving, with many professional digital still and movie cameras now taking up this technology.





Figs. 1.21A and B: A selection of specific instruments available for endoscopic ear surgery from Karl Storz GmbH (A) and Spiggle and Theis Medizintechnik GmbH (B).

Nevertheless, medical technology still predominantly employs CCD, which can either be 1-chip or 3-chip. 1-chip uses a Bayer filter, which means that a dominant color in the red-green-blue spectrum can washout the others—leading to a phenomenon called "red-out" if bleeding is present. 3-chip has prisms, which split the red-green-blue inputs into three separate chips, preventing this from occurring—and so is preferred in EES where small amounts of bleeding in a confined space can make a big difference.

Weight is also a factor to consider with some camera manufacturers targeting the endoscopic markets of other surgical disciplines where weight of the camera is of less importance than in EES. The smaller the camera, the less fatigue the surgeon is likely to experience.

Zoom

Some cameras have a manual focusing ring, while others rely on digital zoom. As with digital photography, manual zoom provides a crisper or clearer image.

Postprocessing

Some manufacturers are now starting to offer postprocessing options, which make darker areas stand out more, or blood vessels more vibrantly displayed. These are certainly helpful options to have available but not essential.

High- and Ultra-high Definition

Standard definition in medical equipment is being phased out and replaced with high definition. High definition means at least 720 pixels in height per frame. Newer ultra-high-definition televisions, which begin at 2,160 pixels in height per frame will likely be phased in next, if the example of the television industry is followed.

LEARNING ENDOSCOPIC EAR SURGERY AND INTRODUCTION TO PRACTICE

Modern Learning Theory and Its Application to Endoscopic Ear Surgery

Certain concepts from modern learning theory need to be understood by any surgeon looking to introduce a technique into their practice. At first, learning is directly affected by the learning surgeon's emotional state and their cognitive bias, at the time of receiving new information. Cognitive bias in the instance of EES refers to the how the traditional microscopic methods have been used in surgical scenarios in the past. Secondly, certain scenarios, referred to as "desirable difficulties" enhance the long-term uptake of a new operative technique:[35]

- *Vary conditions of learning*—in this instance moving from standing to sitting and interspersing different first cases enhances long-term hand eye coordination

- *Distribute sessions*—an inter-training interval of approximately 7 days is ideal, not block learning (such as repeated 2 days intensive courses)
- *Test yourself*—regular review of the relevant anatomy is important to establishing the long-term memory.

Learning Endoscopic Ear Surgery in Residency

Residents generally bring low-cognitive bias and more erratic emotional state control to the learning situation of EES. Residents should ideally have been through a period of autonomous scope holding prior to progressing onto live surgery. Most often this can be achieved through cadaver or three-dimensional printed temporal bone courses. Frequently trainees are already adept with scope usage through functional endoscopic sinus surgery.

Knowledge of anatomy is critical and review of anatomy in standard texts, as well as through purpose designed endoscopic cadaveric dissections online (http://www.sydneyearendoscopy.com) is of paramount importance.

A curriculum-based stepwise training schedule (Box 1.3) using operant conditioning in the form of relatively neutral click prompts has been demonstrated to show uniform improvement in cohorts of orthopedic surgical residents and medical students.[36] This method could be applied to EES-resident training.

Learning as an Established Surgeon

Established surgeons generally bring a high-cognitive bias (regarding the benefits of traditional microscopic methods) and a better ability to control their emotional state, than residents. Learning in this situation commences with prereading and watching anatomy and surgical videos online. The established surgeon should then at least attend one course on the method and begin soon after with a step wise progression of cases (*see* below). Prior to progression into more advanced techniques the established surgeon should consider either visiting another surgeon familiar with EES methods or asking such a surgeon to attend their operating sessions as a mentor.

Starting out in Endoscopic Ear Surgery

Initially, simple procedures will serve as ideal training for the hand-eye coordination required to progress to more advanced endoscopic procedures. Suggestions for initial EES experience include (Box 1.4):

- Middle-ear ventilation tube insertion
- Raising a tympanomeatal flap
- Myringoplasty for small central or posterior perforations

Box 1.3: Sydney endoscopic ear surgery task specific checklist (modified from Lin, et al.[37]).

Assessment criteria:

- Basic inventory and setup of equipment:
  - Knowledge of endoscopes, equipment and instruments
  - Appropriate draping
  - Instrument/tissue handling
- Assessment of candidacy, pathology and canal size:
  - Endoscopic assessment of canal
  - Endoscopic assessment of disease location
  - Assessment of disease radiologically
  - Determination of canal widening and or mastoidectomy
- Ear preparation, injection and hair trimming:
  - Injection
  - Application of otowick or neuropathy with adrenaline
  - Hair removal
- Flap elevation:
  - Incision location
  - Management of bleeding
  - Demonstration of Prussak's space
  - Elevation off handle of malleus; observe the anterior malleus ligament
- Middle-ear exploration with 3-mm scopes 0, 30 and 45:
  - Demonstrate safe insertion of angled scopes
  - Naming structures of retrotympanum
  - Observing ventilation routes
  - Naming structures of the hypotympanum and protympanum
  - Curetting to show limits of Prussak's space and lateral epitympanum; name the regions of the epitympanum
- Ossicle manipulation:
  - Division of incudostapedial joint
  - Removal of incus and identification of the facial nerve and relationships
  - Division of the neck of the malleus and head removal
  - Name the regions of the epitympanum
- Bone removal methods, drill, curette, sonopet and piezoelectric
- Extended middle ear—dissection with angled scopes and instruments; extended protympanum, antrum, hypotympanum and retrotympanum
- Beyond ME—infracochlear, perigeniculate and transpromontorial

(ME: Middle ear).

Box 1.4: Pitfalls and pearls when starting.

- Spend time trimming hair
- Slow down and add time to the operating list when starting EES
- Start in a deliberate manner with wide ear canals and simple uninflamed pathology
- Have the microscope in the OR ready for use
- Avoid 45- and 70-degree scopes when starting
- Manage bleeding by:
  - Anesthetic control of mean arterial pressure and pulse
  - Neuropatties or cottonoids with 1:1,000 adrenaline, ensuring to allow it time to work before suctioning
  - Hemostatic agents like surgical or floseal
  - Warm saline irrigation
  - Microbipolar forceps
  - Low power protected time monopolar for the vascular strip incision

(EES: Endoscopic ear surgery; OR: Operation room).

- Using the scope for inspection during conventional microscopic surgery.

Once the surgeon has gained some familiarity with the principles of EES, specialized EES instruments are useful for more advanced indications such as extensive tympanoplasty, cholesteatoma surgery and surgery beyond the middle ear. These instruments include:

- High-definition cameras and high-definition screen
- 3-mm rigid 0, 30, 45 degree endoscopes
- Short- and long-angled dissectors
- Angled EES curettes
- Angled suckers
- Angled microscissors
- Angled cup forceps
- Bone removal tools such as guarded drill burrs, osteotomes, piezoelectric device or ultrasonic aspirator.

Once the surgeon gains confidence and basic as well as more advanced equipment is available, then progression can occur:

- Subtotal or total perforations using angled scopes
- Cholesteatoma using angled scopes
- Ossicular reconstruction
- Stapes surgery
- Beyond the middle ear: ET and lateral skull base surgery.

CONCLUSION

Endoscopic ear surgery is already established as an alternative approach to perforations, cholesteatoma and chronic ear disease. Future development of improved technology and instrumentation will allow the approach to advance into areas that area traditionally has not accessed with minimally invasive approaches such as lateral skull base and ET surgery. Ongoing research and quality assurance will help to safely advance the indications while improving morbidity and quality-of-life.

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Review

Endoscopic Management of Pediatric Cholesteatoma

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ABSTRACT

Pediatric cholesteatoma occurs in one of two forms: congenital cholesteatoma, developing from embryonic epidermal cell rests or acquired cholesteatoma, associated with a focal defect in the tympanic membrane. This disease has been traditionally managed with the operating microscope, often requiring mastoidectomy for adequate visualization of and access to the middle ear and mastoid cavities. Recently, advances in endoscopic equipment have enabled otologists to manage most cases of pediatric cholesteatoma via a minimally-invasive, transcanal endoscopic approach. This review discusses the current literature relating to the etiopathogenesis, assessment and endoscopic management of pediatric cholesteatoma. Early outcomes of endoscopic treatment, emerging trends and technologies are also reviewed.

Contents

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1. Introduction

Congenital and acquired pediatric cholesteatoma has been successfully managed with the microscope for many decades. Minimally invasive approaches that seek to minimize trauma to normal tissue yet achieve similar surgical goals would be logical to apply to the pediatric population. In the last 5 years, with the introduction of high quality narrow diameter endoscopes, cold light sources, ultra-high definition cameras and screens, endoscopic ear surgery (EES) has evolved into a method to successfully treat pediatric cholesteatoma in a minimally invasive manner. This review seeks to outline the current methods and results of EES in the pediatric cholesteatoma population.

2. Epidemiology

The true incidence of pediatric cholesteatoma is not known, and epidemiological studies are sparse. A retrospective review of 122 children operated on for cholesteatoma in Denmark between 1965 and 1978 inferred an annual incidence of pediatric cholesteatoma of 2.9 per 100 000 children (Tos, 1983). This reported incidence remains a good estimate despite its limitation to a single centre in a well-developed country with excellent access to health care. In children, there appears to be a slight male predilection (relative risk 1.4) (Olszewska et al., 2004). The mean ± SD age of children at diagnosis is 5.6 ± 2.8 years in congenital cholesteatoma and 9.7 ± 3.3 years in acquired cholesteatoma (Nelson et al., 2002). A large historical cohort study (Djurhuus et al., 2015) demonstrated a 20-fold increase in the incidence of surgically-treated middle ear cholesteatoma in individuals with cleft palate and a 14-fold increase in the incidence among individuals with cleft lip and palate. However, no difference in incidence was observed in individuals with cleft lip alone. Girls with Turner syndrome are at a higher risk of developing middle ear disease, including cholesteatoma (Hall et al., 2009) with a reported lifetime incidence in this population of approximately 4% (Lim et al., 2014).

3. Classification

Broadly, pediatric cholesteatoma is classified as congenital or acquired. Congenital cholesteatoma occurs as a "closed type" congenital cholesteatoma (CTCC), the more common encapsulated cyst; or an "open type" congenital cholesteatoma (OTCC) where the stapes is replaced by a medial and lateral plaque-like keratinizing epithelium. Acquired pediatric cholesteatoma is further subdivided into more common middle ear/mastoid or rarer external ear canal.

3.1. Congenital cholesteatoma

Congenital cholesteatoma occurs behind an intact tympanic membrane in a patient with no history of perforation or otorrhea and no prior otologic surgery (Levenson et al., 1989). Congenital cholesteatoma accounts for 10–28% of pediatric cholesteatomas (Potsic et al., 2002a) although this may be underestimated as congenital cholesteatomas resulting in perforation of the tympanic membrane may be incorrectly classified as acquired disease (Darrouzet et al., 2002). Fig. 1 shows the atypical otoscopic findings of an OTCC that may be mistaken for acquired cholesteatoma.

Consistent with an increasing awareness of the disease and improved otoscopic equipment, the most common presentation of congenital cholesteatoma is an asymptomatic middle ear mass (82% of cases) (Potsic et al., 2002a) as seen in Figs. 2 and 3. In this retrospective cohort study, the authors found that 13% of congenital cholesteatomas were found at myringotomy for serous otitis media.

The anterosuperior quadrant is most commonly involved (82% of cases), followed by the posterosuperior quadrant (49% of cases). Ossicular chain involvement and mastoid extension is present in 42% and 23% of cases respectively. Single-quadrant disease is confined to the anterosuperior quadrant in 77% of cases and the posterosuperior quadrant in 22% of cases (Potsic et al., 2002a).

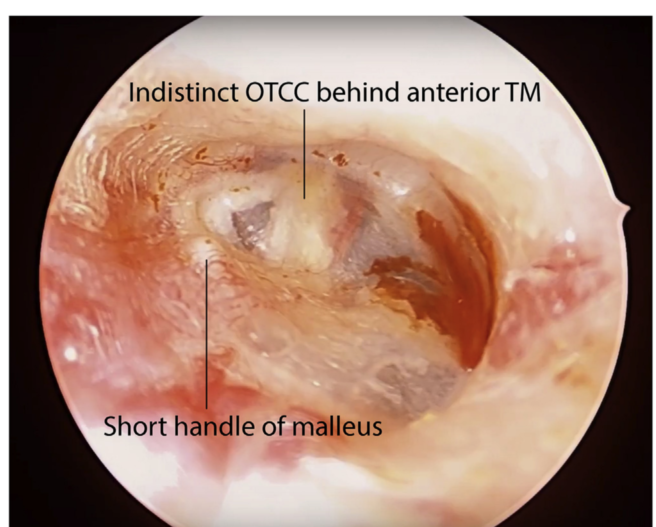

Fig. 1. Atypical otoscopic findings in open type congenital cholesteatoma may be mistaken for acquired cholesteatoma in a 10-year-old, right ear.

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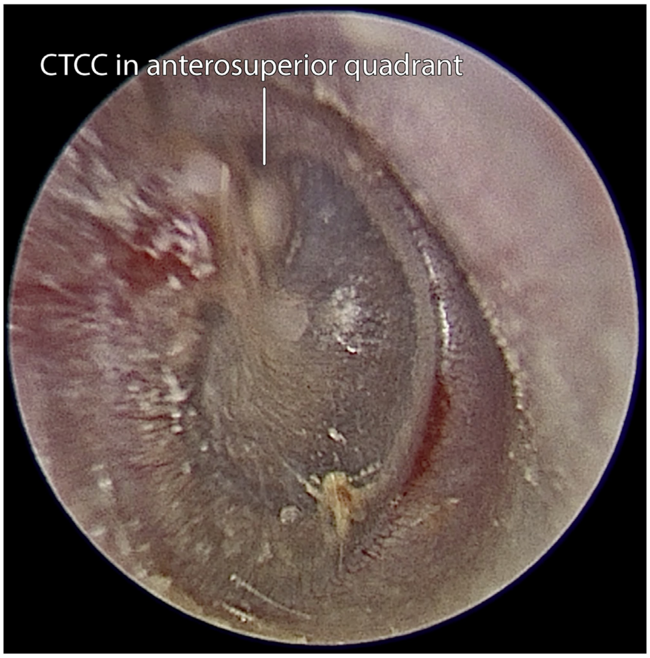

Fig. 2. Otoscopic view of a CTCC in a 2-year-old, right ear.

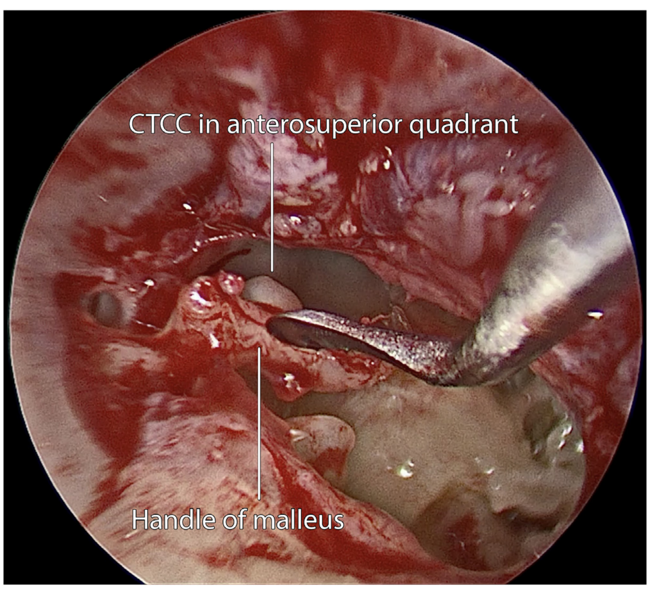

Fig. 3. Operative findings of the CTCC in the same 2-year-old shown in Fig. 2.

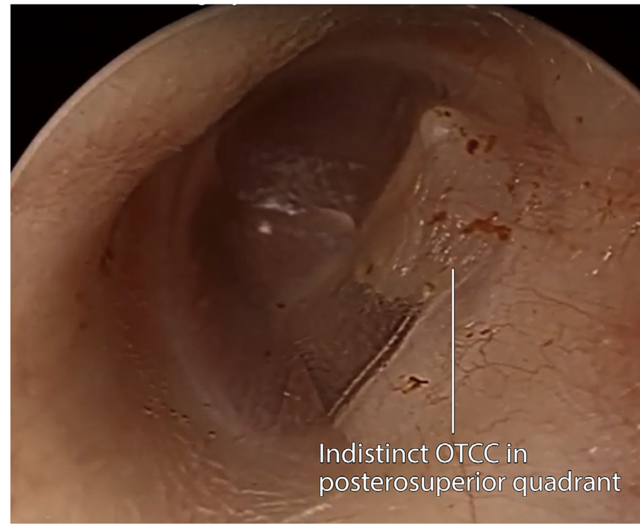

Fig. 4. Otoscopic view of an OTCC in a 12-year-old, left ear.

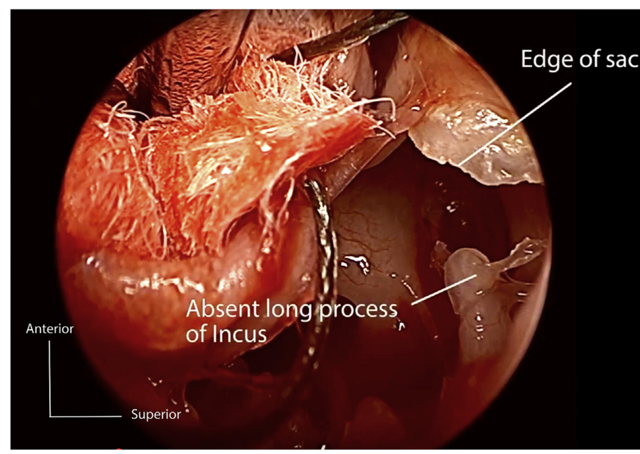

Fig. 5. Operative findings of the OTCC in the same 12-year-old shown in Fig. 4.

3.1.1. Classification

Histopathologically, congenital cholesteatoma takes one of two forms. CTCCs occur as an epithelial cyst without exposure of keratin (Figs. 2 and 3), whereas OTCCs occur as plaque-like lesions of keratinizing epithelium usually replacing a portion or all of the stapes (Figs. 4 and 5). CTCC has a higher prevalence than OTCC (approximately two-thirds of cases), confirmed in most published series (Bacciu et al., 2014). A 15-year retrospective review of 96 congenital cholesteatomas compared the clinical features of the two forms: CTCC was associated with a younger age at diagnosis (6.5 years compared to 11 years in OTCC). 100% of CTCC were diagnosed with otoscopy compared to 40% of OTCC. All cases of OTCC were of a more advanced stage at diagnosis (64% Stage III; 36% Stage IV). The authors demonstrated a significantly increased rate of residual cholesteatoma (based on postoperative otoscopy) in OTCC (OR 7.39, 95% CI 1.1–49.8, $p = 0.03$) (Bacciu et al., 2014).

3.1.2. Etiology

The etiology of congenital cholesteatoma is not known with certainty. Multiple etiopathogenic theories have been proposed and discussed over more than a century, however many remain without convincing supportive evidence (Persaud et al., 2007). The most widely accepted etiopathogenic theory was proposed by Michaels (1986). He studied stained sections of 76 fetal temporal bones and demonstrated the presence of an epidermoid (distinct squamous cell nest with unknown function) in the anterosuperior mesotympanum (Persaud et al., 2007). In his series, this epidermoid formation was not found beyond a gestational age of 33 weeks and Michaels proposed that its persistence could lead to the development of congenital cholesteatoma. In a more recent study, Levine, et al. (1998) demonstrated persistence of epidermoid formations in fetal and postnatal temporal bones beyond 33 weeks gestation to an age of 2 years and 7 months. This work alone did not explain congenital cholesteatomas originating in other areas of the middle ear. More recently, however, epidermoid formations have been demonstrated in other regions of the middle ear, strongly

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supporting epidermoid formation as the anlage for congenital cholesteatoma (Persaud et al., 2007).

3.1.3. Staging

Potsic et al. (2002b) proposed the widely used staging system for congenital cholesteatoma, shown in Table 1. The authors demonstrated that the incidence of residual disease increased with disease stage, from 13% in stage I disease to 67% in stage IV disease. A similar classification was employed by Nelson et al. (2002), that likewise demonstrated higher rates of residual cholesteatoma with more advanced disease.

3.2. Acquired cholesteatoma

The development of acquired cholesteatoma (Fig. 6) is associated with a defect in the tympanic membrane, most commonly a focal retraction pocket (Persaud et al., 2007).

3.2.1. Classification

Historically, acquired cholesteatoma has been classified “primary acquired” cholesteatoma where it originates in a limited retraction of the pars flaccida and “secondary acquired” cholesteatoma where it occurs as a result of a posterosuperior tympanic membrane perforation. Lau and Tos (1989) proposed an alternative classification that is more clinically useful and based on otoscopic findings: (1) attic cholesteatomas originate in a retraction or perforation of Shrapnell's membrane; (2) sinus cholesteatomas originate in a retraction or perforation of the posterosuperior tympanic membrane, spreading to the stapedial niche and tympanic sinus; (3) tensa retraction cholesteatomas arise from a retraction or perforation of the whole pars tensa, including anterior and inferior segments.

Acquired cholesteatoma can occur outside of the middle ear cavity in other areas of the skull, intracranially and in the external auditory canal (EAC). Pediatric EAC cholesteatoma is a rare entity that may be confused with keratosis obturans (Olszewska et al., 2004) and has been associated with significant morbidity including formation of a labyrinthine fistula and invasion into the bony wall of the jugular bulb (Jang et al., 2016b). EAC cholesteatoma is, therefore, an important consideration in a child presenting with otalgia, otorrhea and no apparent middle ear disease.

3.2.2. Etiology

Like congenital cholesteatoma, the etiology of acquired cholesteatoma is not known with certainty. Local infection is commonly found in the setting of cholesteatoma, however its role in cholesteatoma etiology is not confirmed.

The most widely accepted etiopathogenic theory was proposed by Tos (1988), in which a cholesteatoma forms within a deep, advancing retraction pocket most commonly of the pars flaccida. The tympanic membrane is composed of three embryologic layers: an inner endodermal mucosal layer, a middle fibrous mesodermal lamina, and an outer ectodermal epithelial layer, the latter possessing a unique radial migratory capability (Louw, 2010) that confers to the tympanic membrane an ability to self-cleanse (Preciado, 2012). In the setting of chronic eustachian tube dysfunction, as a retraction pocket advances and its neck narrows, the ability of the retracted epithelium to self-cleanse becomes impaired, debris and actively proliferating epithelium becoming trapped to form a cholesteatoma sac.

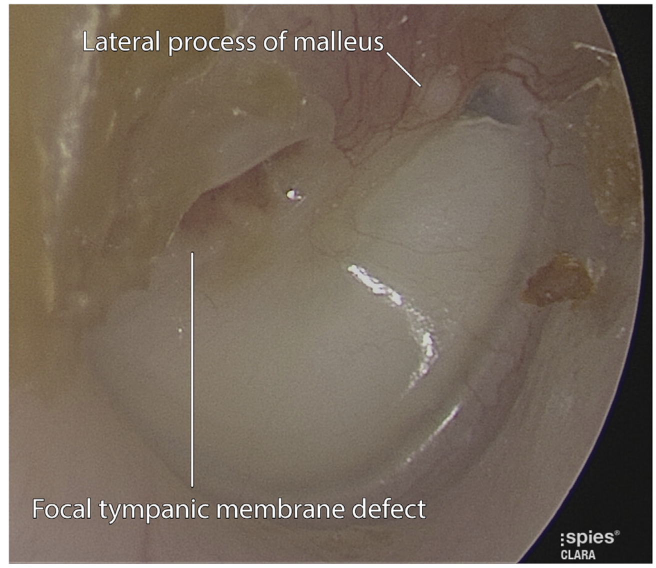

Fig. 6. Acquired pediatric cholesteatoma resulting from a focal defect in the tympanic membrane in a 5-year-old, right ear.

Several other theories explain the origin of cholesteatoma in other areas of the middle ear cavity. Epithelial migration around the margin of a perforation can occur anywhere on the tympanic membrane. Intervening infection can arrest migration, invoke hyperplasia and induce cholesteatoma formation (Louw, 2010). Blast injuries, middle ear surgery and fractures of the petrous temporal bone can result in implantation of epithelium in the middle ear cavity, potentially resulting in cholesteatoma formation.

4. Preoperative assessment and operative planning

Complete transtympanic visualization of the middle ear cavity with the operating microscope is difficult in all but the largest canals, with the view limited by the narrowest segment of the ear canal (Tarabichi and Kapadia, 2017). For this reason, the preferred traditional approach to pediatric cholesteatoma has involved a postauricular canal wall up (CWU) mastoidectomy, canalplasty and a posterior tympanotomy to provide access to the tympanic cavity for visualization and instrumentation. Due to slightly lower rates of recidivism, CWD procedures may be appropriate in patients for whom follow-up presents difficulties or resources available for relook procedures are scarce (McGuire et al., 2016; Osborn et al., 2012). However, in the pediatric population, the requirement for long-term follow-up and mastoid cavity care, poorer hearing outcomes, water exposure restrictions and future considerations such as fitting of hearing aids mean that canal wall down (CWD) procedures are largely out of favor except for markedly aggressive

Table 1
Staging of congenital cholesteatoma (Potsic et al., 2002b).

Stage	Description
I	Single quadrant disease without ossicular involvement or mastoid extension
II	Disease involving multiple quadrants without ossicular involvement or mastoid extension
III	Ossicular involvement, defined as ossicular erosion or necessity of surgical removal for disease eradication
IV	Disease with any mastoid extension

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recurrent disease (Shirazi et al., 2006).

The wide viewing angle of modern endoscopes (Fig. 7) overcomes many of the limitations of the operating microscope, making minimally-invasive, transcanal endoscopic ear surgery (TEES) feasible for the management of pediatric cholesteatoma of the tympanic cavity (Marchioni et al., 2015). When cholesteatoma extends through the aditus into the mastoid, microscopic removal remains the gold standard.

Careful preoperative assessment is essential to determine the extent of disease and therefore, parent/patient preoperative counseling. Clinical and radiological assessment proceeds from lateral to medial, determining whether resection will be totally endoscopic, require a canal widening procedure and/or mastoidectomy.

4.1. Clinical assessment for TEES

Three particular domains should be assessed preoperatively to identify limitations or obstacles to TEES. This assessment is possible in the otologist's rooms or clinic prior to surgery.

4.1.1. Lateral meatus

Rigid endoscopy performed preoperatively with a 3 mm scope is useful to identify the meatal diameter and determine its capacity to accommodate both scope and instruments during TEES. Pediatric cartilage as well as skin is thinner and relatively more elastic than the adult. Therefore, a “collapsing canal” is more likely with suction, which in turn may cause barotrauma of the ear canal skin and tympanic membrane, impairing the surgeon's view.

4.1.2. Bony canal

The pediatric canal doubles in length from 11 to 20 mm between the ages of 5 and 18 (Isaacson, 2014). During that time the external bony canal diameter increases from 4 to 6 mm. Concerns regarding the narrow pediatric ear canal limiting the utility of endoscopic approaches to the tympanic cavity are valid but appear not to be well-founded with several series reporting successful removal of pediatric cholesteatoma confined to the tympanic cavity in all included cases (Ito et al., 2015; Marchioni et al., 2015). A Japanese series demonstrated successful cholesteatoma removal by TEES using rigid endoscopes with a 2.7 mm outer diameter through canals as narrow as 3.2 mm anteroposteriorly and 3.4 mm superoinferiorly (Ito et al., 2015). Nevertheless, a narrow canal can significantly limit the view and passage of instruments during EES as demonstrated in Figs. 8 and 9.

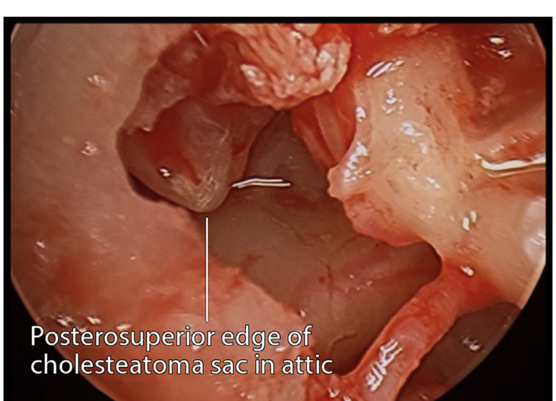

Fig. 7. Angled scope view of attic cholesteatoma sac in a right ear, exemplifying the wide viewing angle of modern endoscopes in a 12-year-old.

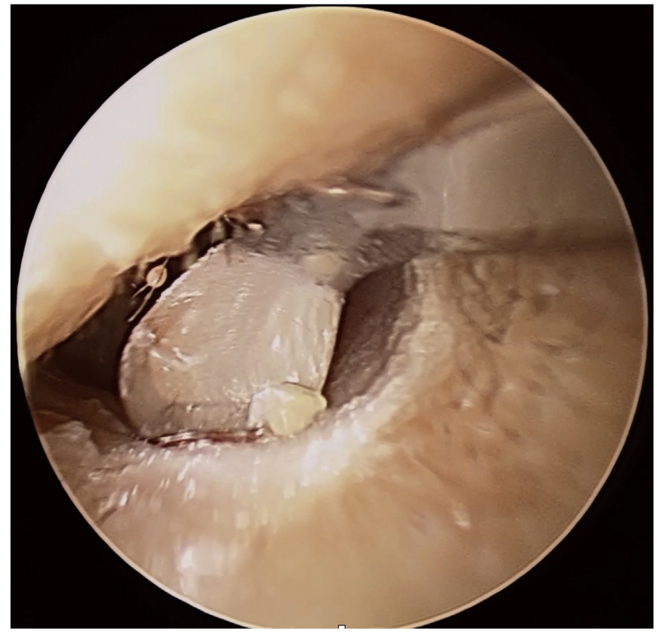

Fig. 8. Narrow left ear canal with angled beaver tympanoplasty blade demonstrating the limitation a narrow canal can place on the view and passage of instruments in a 3-year-old, left ear.

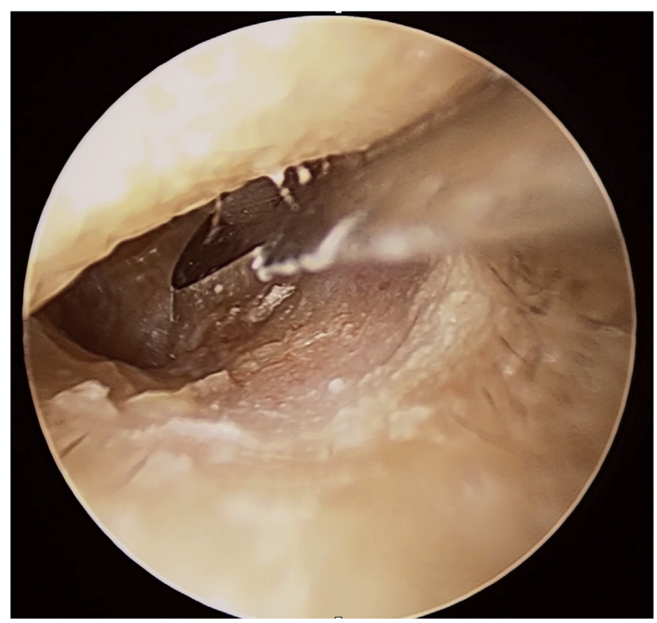

Fig. 9. Straight beaver tympanoplasty blade permitted more easily in the same narrow canal as Fig. 8.

4.1.3. Meatus and canal relative to disease burden

Once the meatus and bony canal have been assessed, the approach should be assessed relative to the disease burden. If the disease can be visualized with a 0-degree scope at the bony-cartilaginous junction then complete endoscopic removal is likely. If there is a narrow canal or large anterior overhang and the view of the disease is limited, then the parents/patient may need to be counseled regarding canal widening.

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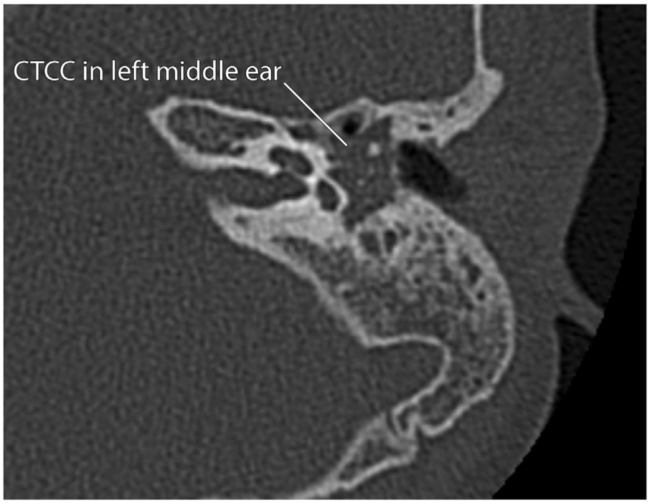

Fig. 10. Axial CT image of a left CTCC in a 5-year-old.

4.2. Radiological assessment for TEES

Radiological assessment is used to identify the presence and extent of cholesteatoma spread into the mastoid and temporal bone.

4.2.1. Computed tomography

CT (Fig. 10) is the ideal modality for assessment of disease and bony erosion however, certain considerations need to be made in the pediatric population. Firstly, children below 4 may find lying still difficult even for the short acquisition times of modern machines. Secondly, radiation concerns of the parents need to be balanced with the requirement for accurate preoperative planning. The effective radiation dose of computed tomography in children is relatively high compared with other imaging modalities and is increased due, in part, to their small body size (Kim et al., 2017). The effective radiation dose of a low-dose temporal bone CT protocol in a 5-year-old child is 0.25 mSv compared with up to 1.7 mSv in literature-derived protocols (Nauer et al., 2011). The annual background radiation in the United States is 2.28 mSv (United States Environmental Protection Agency, 2018). Low dose temporal bone CT appears sufficient to define the bony anatomy of the middle ear however the detail provided is insufficient for the identification of smaller structures such as the stapes (Nauer et al., 2011). Pediatric temporal bone CT potentially exposes radiosensitive organs such as the eyes and thyroid. The effective radiation dose of a single, high-dose temporal bone CT scan in a child is not sufficient to confer carcinogenic or other health-related risks (Box 1) (Kim et al., 2017). However, subjecting patients to the lowest possible dose that achieves the image quality and detail required for preoperative planning is appropriate.

Disease that extends posterior to the lateral semicircular canal, into a deep type c sinus tympani or complications of cholesteatoma such as fistulae or tegmen erosions will generally require preoperative counseling for mastoidectomy.

Box 1
Comparative radiation doses. Compiled from (NSW ACI, 2012) and (Nauer et al., 2011)

Exposure	Effective dose (mSv)
Chest X-Ray	~0.02
7-h flight	0.05
Mammogram	0.1 to 1
Low-dose CT petrous temporal bones	0.25
Return flight (Sydney to London)	~0.25
CT petrous temporal bones (literature-derived protocols)	0.9 to 2.6
Annual background radiation (USA)	2.28
CT pulmonary angiogram	>10

4.2.2. Diffusion-weighted magnetic resonance imaging

Diffusion weighted MRI (Non-EPI DWI) is the imaging of choice for identifying cholesteatoma in the mastoid. Children have a lower sensitivity and specificity (65–70%) (Clarke et al., 2017) likely due to higher prevalence of inflammatory disease in the developing ear compared to the adult. MRI-identification of disease is also poor when its size is < 4 mm or in an OTCC where the keratin is flat.

5. Intraoperative considerations

5.1. Anesthetic

Endoscopic work is possible under assisted local anesthesia; however, the vast majority of pediatric cases require a general anesthetic with intubation using a standard endotracheal tube or laryngeal mask. South RAE tubes, commonly used in otolaryngological procedures, should be avoided as long procedure times may cause compressive injury to the lower lip.

Volatile anesthetics, propofol and older opioids such as morphine cause vasodilation (Saad and Aladawy, 2013), thereby increasing bleeding.

A total intravenous anesthetic (TIVA) using a remifentanil and propofol infusion has been reliably demonstrated in endoscopic sinus and craniofacial surgery to reduce intraoperative bleeding, post-operative edema and ecchymoses (Eberhart et al., 2003; Wormald et al., 2005). Controlled hypotension is more easily achieved in TIVA; remifentanil, a very short-acting μ-opioid agonist and propofol enable more precise control of intraoperative blood pressure compared with volatile inhalational anesthetics such as sevoflurane/isoflurane plus fentanyl/alfentanil (Eberhart et al., 2003; Wormald et al., 2005). The pulse and mean arterial blood pressure should be kept as low as possible, within the safety profile of the patient.

5.2. Positioning and wound preparation

The patient's head is placed on a small head ring and if the child is < 12 years of age then often a small gel bag is required under the shoulders to slightly extend the neck and offset the occipital protuberance. A facial nerve monitor is used in all cases and the surgeon should avoid over-rotating the neck which may lead to neurapraxia and jugular compression. Slight reverse Trendelenburg is preferred by some surgeons to theoretically reduce bleeding.

If the need for a postauricular incision is anticipated with preoperative planning, then hair is shaved and the skin marked up now.

Injection is performed, either preoperatively or after sterile preparation of the skin, with 0.75% Ropivacaine and 1:50000 adrenaline. Caution is required in children under 3 years as the mastoid tip is not formed yet. When injecting near the mastoid tip a finger is placed in the tympanomastoid groove to disperse local anesthesia away from the facial nerve.

A single canal injection (25G – 30G) very slowly in the vascular strip is often all that is required, as well as tragal and conchal

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injections. Overinjection should be avoided as this may greatly reduce vision in the canal. Neuro Patties or an Otowick (Medtronic, 2018) with 1:1000 adrenaline is placed in the bony medial canal whilst hair is then cut from the lateral meatus.

Equipment that is required for most endoscopic pediatric cholesteatoma surgery include, the smaller diameter 3 mm endoscopes (11–14 cm, 0, 30 and 45°) as well as, a dedicated endoscopic ear surgery tray with angled instruments (Karl Storz GmbH, 2018; Spiggle & Theis Medizintechnik GmbH, 2018).

6. Removal of cholesteatoma in hidden areas and special considerations

6.1. Attic and antrum

Bone removal for access to the attic and antrum includes the standard variety of methods: curette, protected tip burr, piezoelectric devices and osteotome. Minimal bone removal is all that is usually required to see all aspects of the anterior and posterior attic with angled scopes. Care is required in this region with insertion and manipulation of angled scopes and instruments. Furthermore, excessive removal of the scutum may create a problem with reconstruction as pediatric tragal cartilage is smaller than the adult.

6.2. Retrotympanum

An area that is difficult to access with traditional canal wall up methods, the retrotympanum usually requires 30- or 45-degree scopes to visualize the lateral aspects of region (Fig. 11). Moving to the contralateral side of the patient's head may help the surgeon with visualization and more natural hand eye movements. An intimate knowledge of the anatomy including sub-classification of recesses allows the surgeon to be confident of removal, when for example bridge configurations need to be removed to fully access disease.

6.3. Protympanum

The protympanum is the region in front of the cochleocarotid groove, bound superiorly by the tensor canal and supratubal recess, inferiorly by the protiniculum and laterally by the lateral lamina which separates the region from the mandibular fossa. Pediatric cholesteatoma often occurs in pneumatized bones so this region is well developed and subclassifications, which have been recently described (Jufas et al., 2016) should be understood by the surgeon.

The protympanum usually requires removal of the tympanic membrane off the handle of the malleus and angled scopes to be completely visualized (Figs. 12 and 13). Manipulation of angled instruments in the region should be undertaken with care to avoid injury to the stapes bone as well as the facial nerve. When beginning to dissect this region, the novice endoscopic surgeon should start with either division of the incudostapedial joint or removal of the ossicles to minimize the risk of sensorineural hearing loss.

6.4. Limited canal wall up mastoidectomy (LCWU) with endoscope assistance

With extensive mastoid disease a mastoidectomy is required. In combination with the endoscopic approach, no canalplasty or disturbance of the lateral vascular strip occurs, so a limited canal wall up mastoidectomy (LCWU) is preferred. The limited form still requires identification of the tegmen, posterior canal wall and sigmoid sinus, however, bony removal does not need to extend posteriorly to view the attic. Furthermore, angled endoscopes may be used to assist in the lateral attic and antral dissection. A wet sponge can be placed at the posterior margin of the mastoidectomy incision and the endoscope rested on the sponge to minimize slipping of the scope on a wet mastoidectomy site.

Since no external canal surgery occurs in the LCWU, the neotympanic membrane heals quicker and there is a reduced risk of blunting and external ear canal healing problems.

6.5. Special considerations for acquired cholesteatoma

The acquired form of pediatric cholesteatoma has two macroscopic forms, an encapsulated sac or infiltrative. Furthermore, in children the disease is often concurrent with acute or chronic inflammation making dissection particularly difficult. Using methods outlined previously to reduce bleeding including TIVA anesthetic, reverse Trendelenburg position, proper infiltration, frequent use of 1:1000 adrenaline-soaked Neuro patties, irrigation and patience will help.

Laser ablation of dissected regions using CO_2, Argon or KTP (Potassium titanyl phosphate) laser, has been shown to reduce recurrence rates in small series, particularly in the pediatric population (Hamilton, 2005; James, 2013; le Nobel and James, 2016).

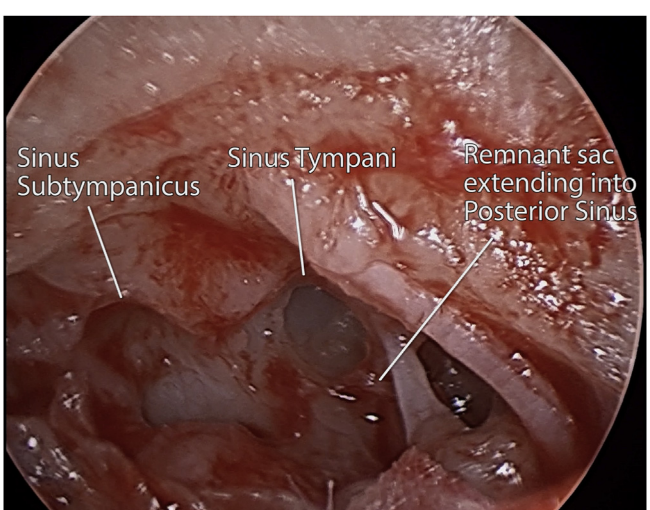

Fig. 11. Contralateral view of retrotympanum after resection in an 8-year-old, right ear.

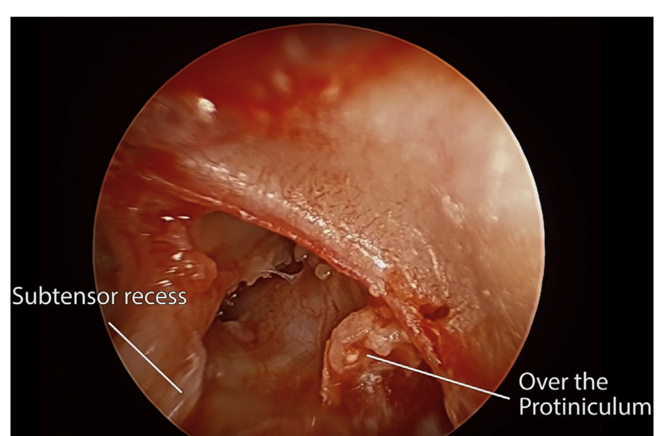

Fig. 12. Protympanum: cholesteatoma in subtensor space and over protiniculum in a 10-year-old, right ear.

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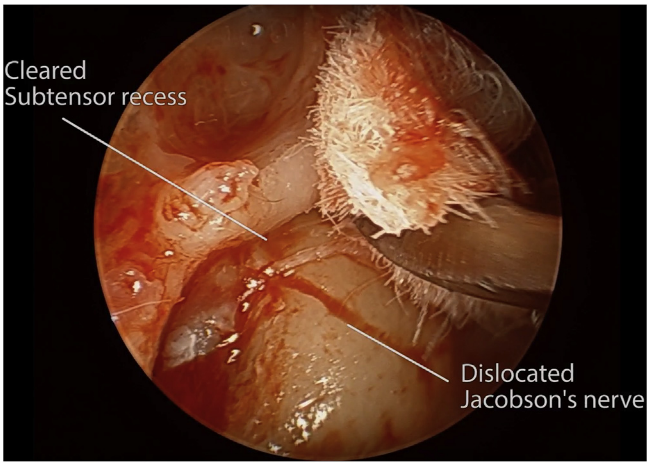

Fig. 13. Protympanum: clearing the subtensor recess in a 10-year-old, right ear.

Care of course should be taken in region of the facial nerve, oval and round windows.

6.6. *Special considerations for congenital cholesteatoma*

CTCC represents an ideal indication for EES. Angled scopes may provide a view of the anterior aspect of the handle of the malleus and tensor tympani to facilitate direct-view dissection of disease in this region (Fig. 14). A recent Korean publication highlights the possibility of successfully removing cysts applied to the tympanic membrane with a transtympanic incision and gelfoam patch (Jang et al., 2016a). Also, a larger series of congenital cholesteatoma demonstrated that lesions that are involving the tensor tympani tendon tend to have a higher recurrence rate (Lee et al., 2017).

The OTCC has a variable relationship to the oval window. Care may often be required here as the epithelial plaque may replace the footplate and dissection reveals membranous labyrinth which requires simultaneous perichondrial or fascial patching, to reduce the risk of sensorineural hearing loss.

6.7. *Considerations in endoscopic pediatric ear reconstruction*

Reconstruction of the attic and tympanic membrane following cholesteatoma resection in EES most commonly uses composite cartilage grafts (Figs. 15 and 16). Cartilage has been reliably demonstrated to retract less and provides similar hearing results (Dornhoffer, 1997). Two unique challenges are encountered compared to the adult reconstruction (see Fig. 17).

- Pediatric tragal cartilage is relatively smaller and thinner than the adult – care must be taken to minimize the defect size to only that which is required. Other possible sites of composite cartilage graft harvest include the conchal bowl and triangular fossa.
- Ongoing Eustachian tube dysfunction and exposure to environmental risks means that in the younger child, post reconstruction infections and chronic otitis media may occur. Some authors advocate the use of ventilation tubes in such recurrent cases in front or through the cartilage graft (Russell et al., 2015).

7. Outcomes with EES compared to CWU

Several recent studies have compared outcomes in TEES for removal of cholesteatoma with outcomes in exclusively microscopic and combined transcanal, transmastoid approaches. Currently reported series are limited by relatively small sample sizes inhibiting the authors' ability to achieve statistical significance in important outcomes and the bias introduced by early endoscopic approaches being reserved for limited, early stage disease.

Two studies (Hunter et al., 2016; Marchioni et al., 2015) demonstrated lower rates of residual disease in endoscopic approaches to cholesteatoma removal (20%) compared with 40% in exclusively microscopic cases and 34.4% with CWD approaches ($p > 0.05$). Hunter et al. (2016) compared pure tone average, (PTA), air-bone gap (ABG) and word recognition (WR) scores pre- and post-operatively in patients undergoing exclusively microscopic, combined transcanal, transmastoid and TEES approaches. PTA, ABG and WR scores were tested in accordance with the guidelines of the Hearing Committee of the American Academy of Otolaryngology – Head and Neck Surgery (Gurgel et al., 2012). The authors demonstrated that pure tone averages improved postoperatively from 33.2 to 26.6 dB HL in exclusively microscopic approaches ($p = 0.054$), from 33.9 to 24.3 dB HL in transcanal, transmastoid approaches ($p = 0.047$) but most significantly from 27.4 to 16.7 dB HL ($p = 0.011$) in TEES approaches. Only patients undergoing TEES

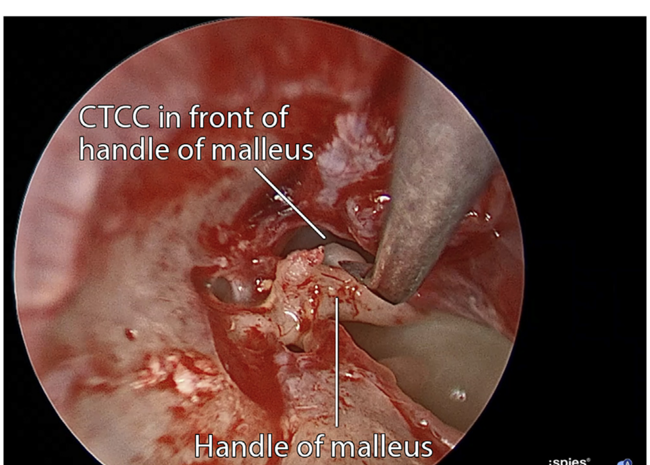

Fig. 14. Dissection of CTCC off the handle of malleus in a 2-year-old, right ear.

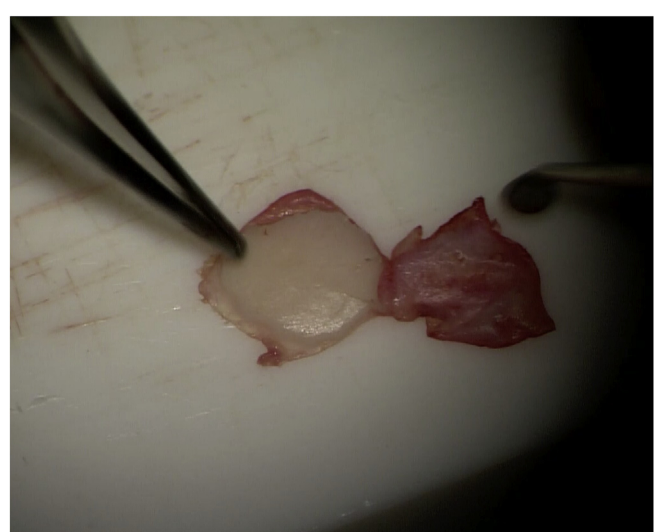

Fig. 15. Thin tragal cartilage graft used in reconstruction in a 3-year-old.

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Fig. 16. Otoscopic view of a right post-operative attic and tympanum reconstruction (at 8 weeks), 12-year-old.

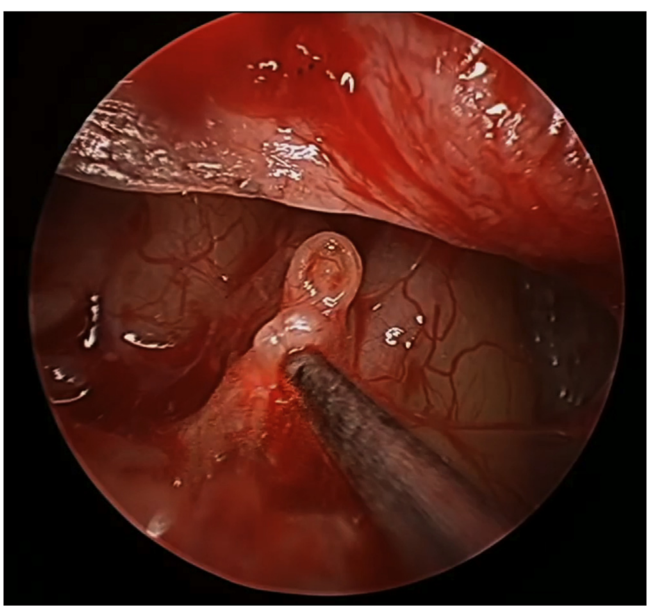

Fig. 17. KTP laser in use over the stapedius tendon during EES for pediatric cholesteatoma in a 6-year-old, left ear.

demonstrated a statistically significant improvement in the air-bone gap, although it is worth noting this group had the lowest preoperative PTA thresholds. At the time of reporting, mean length of follow-up for these two studies was 36 months (Marchioni et al., 2015) and 18.8 months (Hunter et al., 2016). There is evidence that TEES is associated with improved health-related quality of life (HRQoL) postoperatively (Lailach et al., 2015). Patients undergoing TEES demonstrated the lowest restriction in the hearing function, ear symptoms and mental health domains compared with trans-canal, transmastoid and CWD approaches. The difference in ear symptoms (otalgia, otorrhea, aural fullness, headache, hearing loss) was statistically significant (Lailach et al., 2015).

As the number of TEES cases continues to rise, newer equipment and techniques enable the treatment of more advanced disease via this approach. These early studies should be repeated to test the persistence and significance of promising but early outcomes.

8. Future considerations

Instrument and equipment improvements will likely expand the repertoire of EES.

Malleable and directable instruments such as the Malleable sinus suction catheters (MedTronic, 2018) available for endoscopic sinus surgery will likely soon be produced for the ear.

Scope technology continues to improve with chip on tip CMOS technology allowing autofocus and improved dynamic range of light. 3D endoscopes as small as 4 mm are now commercially available (Karl Storz GmbH, 2018) and some manufacturers have 3D bendable scopes to look around structures (Olympus Corp.). In time, with miniaturization, these technologies will translate into the ear.

In-camera live post processing with narrow band imaging (NBI) is available in several specialties to better identify tissue and blood vessels. Staining of keratin and identification with such technology is also a promising idea that may further reduce residual cholesteatoma rates.

Lastly, argon retinal lasers similar in outer diameter to current argon, CO2 and KTP tips have been shown to be applicable to the ear in microscopic surgery (Yau et al., 2015). These lasers have an illuminated tip and are steerable. With miniaturization these may become a very useful tool in managing difficult to reach areas with TEES.

9. Conclusion

Most cases of pediatric cholesteatoma can now successfully be managed with a minimally invasive transcanal endoscopic approach. The otologist should consider this method, along with canal wall up and canal wall down techniques when managing cholesteatoma in the child.

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