Cholesteatoma, an Overview

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Grand Rounds Presentation
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Overview

- Introduction
- Classification and Pathogenesis
- Retraction Pockets
- Anatomic Considerations
- Clinical Evaluation
- Management
INTRODUCTION

History
Definition
Microenvironment Behavior
History

1683 – Du Verneey described a steatoma, a mass between the cerebellum and cerubrum.

1838 – Johannes Mueller described a “layered pearly tumor of fat.”

1885 – Luchae described cholesteatoma behind intact TM.

NOTES: Johannes Mueller in 1838 when he described “layered pearly tumor of fat, which was distinguished from other fat tumors by the biliary fat or cholesterin that is interspersed among the sheets of polyhedral cells”
Cholesteatoma Definition

- Cyst-like, expansile lesion of the temporal bone lined by stratified squamous epithelium that contain desquamated keratin.
- Most often middle ear and mastoid.
  - Anywhere in pneumatized temporal bone.
Consequences of Cholesteatoma

- infection
- otorrhea
- bone destruction
- hearing loss
- facial nerve paralysis
- labyrinthine fistula (usually HSC)
- lateral sinus thrombosis
- intracranial complications
  - epidural and subdural abscesses
  - parenchymal brain abscesses
  - meningitis

NOTES: Encephalocele, meningoencephalocele
### Molecular Cascade of Events

#### TABLE II

<table>
<thead>
<tr>
<th>Disease condition</th>
<th>Cascade of Events during Cholesteatoma Development*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tympanic membrane trauma (perforations): epithelial hyperplasia (projections) triggered by inflammation (obits external), formation of mass-like chvol (haphazard growth &amp; dispersed keratin deposits)</td>
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<tr>
<td>Tympanic membrane disease (otitis externa): epithelial hyperplasia (growths) from tympanic membrane or retraction pockets, papillary cone formation &amp; fusion with keratin deposits in lacunae, expansion into sac-like chvol</td>
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<tr>
<td>Diseased mucosa of tympanic cavity (COM with or without effusion): impact on tympanic membrane with retraction pocket formations (or invaginations) &amp; progression towards chvol (with or without secondary tympanic membrane perforations)</td>
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<tr>
<td>Note: chvol may coexist with or without COM</td>
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<tr>
<td><strong>Molecular events involved</strong></td>
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<tr>
<td>Excessive LA, including α-LA &amp; AA (via Δ6d inhibition) &amp; PA (via FAS stimulation) may stimulate hyperplasia (&amp;/or metaplasia), &amp; excessive VLCFAs (such as DHA) (via β-oxidation inhibition) are eventually cytotoxic to cells which may cause chvol deterioration (based on lipid research)</td>
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<tr>
<td>Hypoxia stimulates HIF and induces MMP productions which cause chvol deterioration &amp; bone erosion</td>
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<tr>
<td>Inflammatory infiltrates (neutrophils) produce excessive NO (oxidant) via NOS stimulation &amp; depletion of SOD, GP &amp; CAT (antioxidants), which causes oxidative stress, chvol deterioration &amp; bone erosion</td>
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<tr>
<td>Neutrophil granules also release antimicrobial MPO, &amp; toxic agents (such as HOCl) are released (via MPO-HOCl complex (within phagolysosomes)), which are associated with bone erosion</td>
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<tr>
<td>Lipid peroxidation &amp; production of toxic products (such as hydroperoxide &amp; aldehydes) may also contribute to chvol deterioration</td>
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<tr>
<td>LPSs in chvol &amp; tympanic cavity biofilms cause: enhanced AA release via PLA stimulation &amp; enhanced PGE2 production via COX-2 stimulation. COX-2 is a key inflammatory mediator of inflammation during chvol matrix-perimatrix interations &amp; OM</td>
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<tr>
<td>LPSs also stimulate TNF-α production (in a wide variety of cells). TNF-α, PGE2 &amp; oxygen radicals (NO) activate NF-κβ (central mediator of immune responses), which reactivates production of TNF-α &amp; PGE2</td>
<td></td>
</tr>
<tr>
<td>LPS-induced OM results in mucoid MEE, &amp; mucous secretions stimulate NO production (via enhanced NOS activity) which causes bone resorption</td>
<td></td>
</tr>
<tr>
<td>LPSs, TNF-α, IL-1 &amp; IL-6 play prominent roles in pathogenesis of OM, AOM &amp; COM</td>
<td></td>
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<tr>
<td>LPSs &amp; TNF-α can: stimulate IL-1, IL-6 &amp; HEX activity involved in bone erosion; stimulate IL-1 &amp; RANKL (family member of TNF-α) involved in bone resorption; &amp; inhibit peroxisome β-oxidation, whereby accumulation of VLCFAs may cause bone erosion</td>
<td></td>
</tr>
<tr>
<td>Chol debris can cause bone resorption</td>
<td></td>
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<tr>
<td>Note: degree of bone erosion depends on cumulative effects of events involved</td>
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</tbody>
</table>

*For both sacs or mass-like growths, Chol = cholesteatoma; COM = chronic otitis media; α-LA = alpha linolenic acid; AA = arachidonic acid; Δ6d = Δ6 desaturase; PA = palmitic acid; FAS = fatty acid synthase; VLCFA = very long chain fatty acid; DHA = docosahexaenoic acid; HIF = hypoxia inducible factor; MMP = metalloproteinase; NO = nitric oxide; NOS = nitric oxide synthase; SOD = superoxide dismutase; GP = glutathione peroxidase; CAT = catalase; MPO = myeloperoxidase; HOCL = hypochlous acid; LPS = lipopolysaccharide; PLA = phospholipase A; PGE2 = prostaglandin E2; COX-2 = cyclo-oxygenase-2; OM = otitis media; TFN-α = tumour necrosis factor α; NF-κβ = nuclear factor κβ; MEE = middle-ear effusion; IL = interleukin; AOM = acute otitis media; HEX = hexosaminidase; RANKL = receptor activator of nuclear factor κβ ligand
Molecular Cascade of Events

- Accumulation of fatty acid chains -> hyperplasia/metaplasia
- Hypoxia stimulates HIF and induces MMP -> bone erosion
- NO -> bone erosion
- Lipid peroxidation -> cholesterol deteriorization
- Cox-2 inflammatory mediator
- TNF-alpha, PGE2, oxygen radicals
- IL-1, IL-6, among cytokines -> bone erosion

- Molecular cascade paper
Destructive Properties

- Proteolytic activity
- Bone remodeling/resorption
  - Multinucleated OC within subepithelial matrix release acid phosphotase, collagenase, other proteolytic enzymes.
- Fibroblast activation in perimatrix
- Matrix metalloproteinases (MMPs)
  - Zinc and Calcium dependent endopeptides made from fibroblasts, keratinocytes, macrophages, endothelial cells
  - MMP2, MMP3, MMP9 first identified in 1996
  - MMP8, MMP13 also mentioned in literature.
Destructive Properties

- MMP paper
  - Accumulation of fatty acid chains -> hyperplasia/metaplasia
  - Hypoxia stimulates HIF and induces MMP -> bone erosion
  - NO -> bone erosion
  - Lipid peroxidation -> cholesterol deteriorization
  - Cox-2 inflammatory mediator
  - TNF-alpha, PGE2, oxygen radicals
  - IL-1, IL-6, among cytokines -> bone erosion

- Molecular cascade paper
### Microbiology

<table>
<thead>
<tr>
<th>Isolated pathogens</th>
<th>No. cases (%)</th>
<th>Pure/mixed culture (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Aerobic</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pseudomonas aeruginosa</td>
<td>65 (31.1)</td>
<td>50 (76.9)/15 (23.1)</td>
</tr>
<tr>
<td>Staphylococcus aureus</td>
<td>40 (19.1)</td>
<td>10 (25)/30 (75)</td>
</tr>
<tr>
<td>Proteus mirabilis</td>
<td>16 (7.7)</td>
<td>0 (0)/16 (100)</td>
</tr>
<tr>
<td>Escherichia coli</td>
<td>3 (1.4)</td>
<td>0 (0)/3 (100)</td>
</tr>
<tr>
<td>Klebsiella pneumoniae</td>
<td>2 (1)</td>
<td>0 (0)/2 (100)</td>
</tr>
<tr>
<td><strong>Anaerobic</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peptococcus</td>
<td>26 (12.4)</td>
<td>10 (38.5)/16 (61.5)</td>
</tr>
<tr>
<td>Peptostreptococcus</td>
<td>10 (4.8)</td>
<td>4 (40)/6 (60)</td>
</tr>
<tr>
<td>Bacteroides</td>
<td>26 (12.4)</td>
<td>8 (30.8)/18 (69.2)</td>
</tr>
<tr>
<td>Clostridium</td>
<td>8 (3.8)</td>
<td>2 (25)/6 (75)</td>
</tr>
<tr>
<td>Fusobacterium</td>
<td>6 (2.9)</td>
<td>2 (33.3)/4 (66.6)</td>
</tr>
<tr>
<td>Propionobacterium</td>
<td>4 (1.9)</td>
<td>0 (0)/4 (100)</td>
</tr>
<tr>
<td><strong>Mycetes</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aspergillus niger</td>
<td>3 (1.4)</td>
<td>0 (0)/3 (100)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>209 (100)</td>
<td></td>
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</tbody>
</table>

NOTES: 150 pts with CSOM found to have cholesteatoma
CLASSIFICATION AND PATHOGENESIS

Congenital

Acquired

• Primary (retraction pocket)
• Secondary
Congenital Cholesteatoma

- “epidermal inclusion cyst” behind intact TM
- In anterior superior region
- Theories
  - 1. invasion of misdirected ectodermal cells within
    EAC migrate through tympanic isthmus into ME.
  - 2. Embryonic rest remnants form epithelial tissue
- Criteria:
  - WITHOUT TM perforation
  - WITHOUT history of ear infections
  - WITHOUT history of otologic surgeries
  - NORMAL pars flaccida, pars tensa

NOTES: Majority of embryonic rests occur within anterior limb of tympanic ring (AS region)
Congenital Cholesteatoma

- 2/3 present as white mass in anterior-superior quadrant.
- Also found in tympanic membrane and in petrous apex
- Mean age of presentation is 4.5 yo
- M:F 3:1
- Incidence is 0.12 per 100,000 people
Congenital Cholesteatoma
Pathogenesis

- Teed in 1936 – fetal human temporal bones
  - Ectodermal epithelial thickening involutes to form mature middle ear lining.
  - Failure of involution is source of cholesteatoma.
- Michaels in 1980s – fetal human temporal bones
  - Identified squamous cell tuft present from 10-33 wk of gestation.
  - This “epidermoid formation” was noted in AS wall of ME cleft.
  - Failure of involution could be basis of cholesteatoma in AS mesotympanum
  - Documented existence of congenital cholesteatoma with epithelial rests.
Staging of Congenital Cholesteatoma

- Derlacki and Clemis (1965) first to stage congenital cholesteatoma:
  - 1. Petrous pyramid cholesteatoma
  - 2. Cholesteatoma involving the mastoid cavity
  - 3. Cholesteatoma involving the middle ear cavity.

- Potsic’s staging:
  - Stage I: Single quadrant involvement with no ossicular / mastoid involvement.
  - Stage II: Multiple quadrant involvement with no ossicular / mastoid involvement
  - Stage III: Ossicular involvement without mastoid involvement
  - Stage IV: Mastoid extension

- Nelson's staging:
  - Type I: Involvement of mesotympanum without involvement of incus / stapes
  - Type II: Involvement of mesotympanum / attic along with erosion of ossicles without extension into the mastoid cavity
  - Type III: Involvement of mesotympanum with mastoid extension
Theories for Acquired Cholesteatoma

1. Metaplasia
2. Implantation
3. Proliferation
4. Retraction
Metaplasia

- Transformation of chronically inflamed ME mucosa into keratinizing epithelium
  - i.e. Barrett’s Esophagus
Implantation

- After perforation whether traumatic or 2/2 OM
- Keratinizing epithelium introduced directly into ME or migration from edges of perforation
- Properties of TM epithelium shared with cholesteatoma epithelium
Proliferation

- Keratinocytes of the basal layer of the TM form conelike extensions that grow into the ME rather than externally.

NOTES:
- Molecular cascade paper also supports this
- Sudhoff and Tos provide IHC support of this
Retraction

- Chronic ETD leads to formation of a retraction pocket in the weakest portion of the TM, the pars flaccida and PS pars tensa.
- Continued negative pressure deepens retraction pocket and keratin debris accumulates.
- Pars flaccida lacks a fibrous layer, making it weaker.
Combination of Retraction and Proliferation Theory

- IHC of attic cholesteatomas showed proliferation of keratinocytes within epithelial cones growing toward underlying stroma.
- Cones with focal discontinuities of basement membrane.
- 4-step Theory
  - 1. retraction pocket stage
  - 2. proliferation stage of into cone formation and fusion
  - 3. expansion stage
  - 4. bone resorption
Combination of Retraction and Proliferation Theory

Sudhoff and Tos

FIG. 9. Schematic illustration of the proliferation stage. A: Normal intraepithelial keratinocyte differentiation and maturation in an epithelial cone orientated vertically toward the surface. Because of the initial increase of keratinocyte proliferation, the cone has started to grow (thick arrow). B: The keratinocyte differentiation is orientated toward the center of a long cone (arrows), forming small lakes of keratin—the microcholesteatoma. C: The microcholesteatoma lakes are expanding, opening to the surface of the retraction and to the neighboring cones, making the cholesteatoma expand for the length of a cone.

FIG. 10. A: Expansion of attic cholesteatoma; the lakes of keratin are opened to the surface of the retraction wall at the depth the cones are proliferating and growing. B: The cholesteatoma has expanded by the length of the cone in the depth new microcholesteatoma are formed within the new cones. C: The keratin lakes are fused, moving the border of the matrix further toward the attic. D: Further expansion of attic cholesteatoma. Establishment of a vicious circle: proliferation at the bottom of the cone, keratin formation within the cones, fusions of microcholesteatomas, and further accumulation of keratin, leading to further deterioration of the self-cleansing mechanism.
Retraction pockets of pars flaccida
Retraction pockets of pars tensa
Grading

RETRACTION POCKETS

Fig. 1
Grade III retraction pocket (seen intra-operatively with 0° scope).
Retraction pockets

- **Causes**
  - 2/2 ETD
    - AR
    - LPRD
  - 2/2 repeated OM
    - Histological degeneration of lamina propria of TM
  - Weak pars flaccida
    - No fibrous middle layer

- **Treatment**
  - Observation
  - Treat underlying cause of ETD
  - Tympanostomy tubes
    - Walsh showed excision with tube corrected grades 2-4 (Sade)
  - Tympanoplasty

**NOTES:**
- Ramakrishnan review of retraction pockets
- Walsh results
Pars Flaccida Retraction Pocket Grading (Tos)

Tos’s grading system (1982):

- **Grade I:** retracted pars flaccida is not in contact with neck of the malleus.
- **Grade II:** retracted pars flaccida is in contact with the neck of the malleus “clothing” the neck.
- **Grade III:** retracted pars flaccida is in contact with the neck of the malleus AND limited erosion of the outer attic wall or scutum.
- **Grade IV:** retracted pars flaccida is in contact with the neck of the malleus AND severe erosion of the outer attic wall or scutum.

Sudhoff and Tos paper 2000
Sade’s atelectasis staging system (1976):

- Stage 1 - Mild retraction
- Stage 2 - Retraction onto incudostapedial joint
- Stage 3 - Retraction onto promontary
- Stage 4 - Adhesion of pars tensa to medial wall

In stage 3 the tympanic membrane can be lifted off the middle ear medial wall whereas in stage 4 it is not possible.

Sade and Berco in 1975 from Israel
Posterior Superior Retraction Staging (Sade)

- 1. slight, self-cleansing retraction
- 2. deeper retraction needing cleansing by clinician
- 3. deeply partly hidden retraction requiring excision
- 4. deep retraction pocket with exposing scutum

Sade – 1993 – tx of chol and retraction pockets
Pars Tensa Retraction Pocket Staging (Charachon)

Charachon proposed a different classification for pars tensa retractions (1992):

- **Stage 1** - Mobile retraction pocket
- **Stage 2** - Fixed and controllable retraction pocket (totally visible under otomicroscopy)
- **Stage 3** - Fixed and uncontrollable retraction pocket (deepest part is invisible)

- Charachon paper, France
- Used this classification to see which pts can be medically managed vs surgery for retraction pocket
Retraction Pocket Staging (Black)

Black and Gutteridge staging (2011):

- **Stage 1:** TM collapse w/o HL
- **Stage 2:** collapsed w/ CHL
- **Stage 3:** collapsed and OC fixation/necrosis
- **Stage 4:** cholesteatoma
Retraction Pocket Staging (Black)

- Stage 1 – no intervention needed or tube
- Stage 2 – drum stiffening with composite material
- Stage 3 – OCR with myringoplasty
- Stage 4 – TC/CWU/CWD/atticotomy

Black and Gutteridge staging (2011)
ANATOMIC
CONSIDERATIONS

Anatomy of ME
Definitions
Common routes of spread
Anatomy Considerations

ME divisions
- Mesotympanum
- Hypotympanum
- Epitympanum

Pathways
- Follows ligaments and folds
ME Divisions

- Attic (Epitympanum)
- Incus
- Tympanic Membrane
- Oval Window
- Round Window
- Stapes
Most frequent origins

- Posterior epitympanum (most common)
- Posterior mesotympanum
- Anterior epitympanum
Epitympanum Cholesteatoma

- Originates in Prussak’s Pouch
  - Between pars flaccida and neck of malleus
  - Floor is lateral process of malleus and associated folds

Posterior epitympanic cholesteatoma passing through the superior incudal space and the aditus ad antrum
Posterior Epitympanum Spread

- Through superior incudal space lateral to body of incus
- Then through aditus ad antrum to mastoid
Posterior Mesotympanum Spread

- Reaches ME descending through floor of Prussak’s space into posterior space of von Troelsch
  - Von Troelsch’s space
    - between TM and posterior mallear fold
    - Inferior edge is chorda tympani nerve
  - Open to mesotympanum
  - May involve stapes, round window, sinus tympani, facial recess
Posterioal Mesotympanum Origin

- Pars tensa retraction
- Passes medial to malleus and incus
- Invade:
  - Sinus tympani (between facial nerve and medial wall of mesotympanum)
  - Facial recess (bounded by fossa incudis and facial nerve medially and chorda tympani nerve laterally)
Anterior Epitympanum Origin

- Retraction anterior to head of malleus
- Bordered by:
  - Anteriorly is middle cranial fossa, petrous tip, zygoma root
  - Posteriorly is cog extending to cochleariform process
  - Superiorly by middle cranial fossa
  - Laterally by tympanic bone and chorda tympani nerve
  - Floor associated with facial nerve -> may affect CN7
- Extend to supratubal recess via anterior pouch of von Troeltsch’s space.
CLINICAL EVALUATION

H&P
Imaging
Pre-operative considerations
Clinical Work-up

- Complete History
- Otologic History
  - Hearing loss, usually unilateral
  - Tinnitus
  - Otalgia
  - Otorrhea
  - Vertigo
  - h/o ear infections
  - h/o otologic surgeries
  - h/o TM perforation
  - h/o allergic rhinitis
Clinical Work-up

- Complete H&N Exam
- Otologic Exam
  - Auricle and mastoid tip
  - EAC
    - Looks for any polyps, granulation tissue, erosion of bony canal
    - Take any cultures if indicated
  - TM
  - Otomicroscopy
    - Remove any debris in canal
  - Pneumatic Otoscopy
    - Fistula test
      - Positive suggests erosion of inner ear, most commonly HSC.
  - Tuning Fork Exam
    - Rinne and Weber
- Neurological Exam
  - Cranial Nerves, especially CN7
  - Dizzy tests
Complete Audiological Exam (CAE)

- Audiometry
  - PTA with air and bone conduction
  - SRT
  - WR

- Tympanometry
Audiometric Findings

- Degree of conductive loss will vary considerably depending on the extent of disease depending disease
  - Moderate conductive deficit in excess of 40 dB indicates ossicular discontinuity
    - Usually from erosion of the long process of the incus or capitulum of the stapes
  - Mild conductive deafness may be present with extensive disease if cholesteatoma transmits sound directly to stapes or footplate (natural myringostapediopexy).
Radiological Studies

- CT temporal bones w/o contrast
  - Axial and coronal planes
  - 1 mm cuts
- Visualize:
  - Scutum erosion
  - Expansion of antrum
  - Ossicular destruction
  - Facial canal erosion
  - Tegmen dehiscence
  - Otic capsule erosion, especially HSC
  - Petrous ridge involvement
Radiological Studies

- Plain films are useless
- MRI only specific situations
  - Dural involvement
  - Sub or epidural abscess
  - Herniated brain
  - Inflammation of labyrinth or facial nerve
  - Sigmoid sinus thrombosis
CT Indications

- CT is not essential for preoperative evaluation
- CT evaluation should be obtained for:
  - CSOM
  - Preop risk assessment
    - Ossicle involvement/post op CHL risk
    - Parents counseled on congenital cholesteatoma on postoperative HL
  - Revision cases
    - Underlying anatomy indeterminate
  - Congenital anomalies expected
    - Craniofacial abnormalities
  - Cases of cholesteatoma in which SNHL, vestibular symptoms, or other hearing complication evidence exists
  - Presence of complicating factors
    - Involvement of labyrinth, dura, brain, sigmoid sinus
  - Identify associated conditions
    - CRS, septal deviation, NP masses
CT Controversy

- Some believe CT in all cases
- Some believe CT only in handful of cases:
  - Dx in doubt with small attic retraction
    - Bony erosion can be seen
    - White mass behind TM may be tympanosclerosis, cartilage, or cholesteatoma
  - Pt wishes to avoid surgery
    - Poor medical health
    - Assess watchful waiting risks
    - If only hear, avoiding surgery reduces postop HL

Per Roland in Pensak
MANAGEMENT

“GOOD JUDGMENT COMES FROM EXPERIENCE, BUT EXPERIENCE COMES FROM BAD EXPERIENCE.”
Preventative Management

- Retraction 2/2 ETD precedes Cholesteatoma
- Tympanostomy tube may be indicated
- TM may adhere to ossicles or lose elasticity
- Tympanoplasty may be indicated
Concurrent Infection

- Otomicroscopy with debridement
- Antibiotic otic drops
  - Ciprodex
- Acetic Acid otic drops
Medical Management

Indications:

- Advanced age
- Poor health
- Refusal of surgery
Surgical Goals

- Safe ear
  - Treating or decreasing risk of complications
- Removal of disease
  - Bone, mucosa, polyps, cholesteatoma
- Reduce recidivism
- Dry ear
- Preserve normal anatomy
  - i.e. posterior canal wall
- Improve/Preserve hearing
Surgical Management

- Radical Mastoidectomy
- Modified Radical Mastoidectomy
- Canal Wall Down Mastoidectomy (CWD)
- Canal Wall Up Mastoidectomy (CWU)
- Others
  - Canal Wall Reconstruction Mastoidectomy
  - Transcanal anterior atticotomy
  - Bondy Modified Radical Mastoidectomy
History of Mastoidectomy

- Before mid 1950s, there was only RM and MRM.
- Wullstein and Zollner introduced tympanoplasty.
- 1958, House started T-mastoids (CWU)
- In 1963, 50% of cholesteatoma managed with CWU.
- In 1960s, plastic sheeting through facial recess decreased retraction pockets therefore reducing recurrent cholesteatoma.
Radical Mastoidectomy

- Mastoid antrum, tympanum, EAC converted into common cavity exteriorized through external meatus
- Removal of tympanic membrane and ossicular remnants
- Spares stapes
- No OCR/grafts
- Eustachian tube plugged
- Meatoplasty to allow bowl debridement
- Infrequently performed
Radical Mastoidectomy

OPEN CAVITY

RADICAL MASTOIDECTOMY

CHOLEST. MATRIX
Modified Radical Mastoidectomy

- Epitympanum, mastoid antrum, EAC converted into common cavity.
- TM and ossicles spared.
- Infrequently performed.
Tympanoplasty with Mastoidectomy

3 variations

1. permanent exteriorization of epitympanum and mastoid (CWD)
2. CWU
3. CWD + obliteration of mastoid cavity and/or reconstruct EAC
CWD Creation

- Removal of all air cells including retrofacial, retrolab, subarcuate
- Remove lateral and posterior walls of epitympanum
- Amputate mastoid tip
- Saucerize lateral margins
CWD Creation (cont)

- Lower posterior bony EAC wall to facial nerve level
- Exteriorize anterior epitympanic recess by removing cog
- Enlarge meatus, remove conchal cartilage
- Lower medial end of EAC toward hypotympanum
CWD Mastoidectomy

- **Indications**
  - Cholesteatoma in only hearing ear
  - Bilateral Dz
  - Multiple previous procedures
  - Erosion of posterior bony EAC
  - Labyrinthine fistula (pre-op, +fistula test)
  - Poor ET function
  - Previous failure of CWU
    - Recurrence 2/2 retraction pocket
  - Intra-operative decision
    - Inadequate surgical access
      - Sclerotic mastoid limiting access to epitympanum
    - Extensive cholesteatoma
    - Labyrinthine fistula
    - Facial Nerve Involvement
CWD Mastoidectomy

- **Advantages**
  - Residual disease easily detected
  - Recurrent disease is less frequent*
  - Facial recess exteriorized

- **Disadvantages**
  - Open cavity takes months to heal
  - Mastoid bowl needs to be maintained for life
  - Dry ear precautions
  - Shallow ME space makes OCR difficult
  - HA does not fit well

*recurrent disease is less... is controversial, depending on specific otologist
CWU Mastoidectomy

• Preserve posterior bony EAC
• Staged 2\textsuperscript{nd} look procedure 6-18 mo
  ◦ Removal of residual cholesteatoma
  ◦ OCR
  ◦ May not be needed depending on disease extent and skill of otologist*
CWU Mastoidectomy

- Relatively contraindicated in:
  - Cholesteatoma in only hearing ear
  - Labyrinthine fistula
  - Poor ET function
CWU Mastoidectomy

- **Advantages**
  - Takes only weeks to heals
  - No mastoid bowl
  - HA fit better

- **Disadvantages**
  - Technically more difficult
  - 2\textsuperscript{nd} stage operation needed
  - Residual disease harder to detect
  - Recurrent disease at higher rate*
**Summary**

**“Open” versus “Closed” Cavity Techniques**

<table>
<thead>
<tr>
<th>Closed Cavity</th>
<th>Open Cavity</th>
</tr>
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<tbody>
<tr>
<td>1. Normal appearance</td>
<td>1. Enlarged meatus</td>
</tr>
<tr>
<td>2. Easy-to-fit hearing aid</td>
<td>2. Difficulty to fit hearing aid</td>
</tr>
<tr>
<td>3. No routine cleaning required</td>
<td>3. Annual or semiannual canal cleaning needed</td>
</tr>
<tr>
<td>4. Relatively high rate of recurrent or persistent cholesteatoma</td>
<td>4. Low rate of persistent or recurrent cholesteatoma</td>
</tr>
<tr>
<td>5. High tolerance for water exposure</td>
<td>5. Occasional problem with water exposure</td>
</tr>
<tr>
<td>6. Usually staged</td>
<td>6. Usually single procedure</td>
</tr>
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CWU vs. CWD Controversy

- Hearing: CWU=CWD (experienced otologists)
- Healing: CWU (6-8 wk) < CWD (6-8 mo)
- Residual Dz: CWU=CWD (experienced otologists)
- Recurrent Dz: only CWU can have PS retraction. CWU (~5%) > CWD (0%)

- Syms and Sheehy (House)
CWD Mastoidectomy

- Retrospective review of one surgeon (Chang) of 104 CWD for extensive/advanced cholesteatoma with high grade atelectasis and severely destructed ossicles.
- Recurrence rate was 3.8%
- Recurrent otorrhea in 9.6%
- 35.6% had closure of ABG within 20 dB
- Availability of stapes superstructure influenced postop hearing level significantly.
  - +stapes -> 67.8% with <20 dB ABG
  - -stapes -> 23.7% with <20 dB ABG
- Conclusion – low recurrence rate, high dry ear rate, preserves adequate hearing.
CWD Mastoidectomy

- Ossiculoplasty performed by 1-stage surgery if indicated.

- Even in advanced cholesteatoma, CWD mastoidectomy provides a low recurrence rate, establishes a high dry ear rate, preserves adequate hearing when stapes superstructure is available for reconstruction.

- Chang and Chen 2000
CWU vs. CWD Controversy

- Roland
  - No significant difference in hearing results in either
  - Avoid recurrence -> CWD
  - Poor follow-up -> CWD
  - Refuse meatoplasty -> CWU
  - Poor ETD -> CWD

- Intra-op decision
  - CWD if: sinus tympani, medial canal wall, hypotympanum, canal wall, lab fistula

Per Roland in Pensak
Clinical and Surgical Pitfalls

1. inadequate surgical objectives
   ◦ Plan for cholesteatoma removal as well as surrounding reaction
2. remember the hypotympanum
3. adequate intraop exposure
   ◦ Size of mastoid, height of facial ridge, adequate meatoplasty
4. Utility of mastoid obliteration
5. Remember limitations of ETD
6. Skin grafting
7. Limitations by histological changes from long standing CSOM
8. OCR: staged vs. unstaged
   ◦ Nadol says to OCR at primary surgery
   ◦ No need for max CHL for 6-18 mo
Decision Making for Surgery

- Is Surgery Necessary?
  - Sooner if impending complication
  - Elective if only intermittent otorrhea
  - Earlier to prevent complication & simpler surgery
- CWD is to “avoid disturbing” the hearing and avoid 2\textsuperscript{nd} look
- Sheehy and House guys stage 70\% of T-plasties because of:
  - Mucosal disease
  - Possible residual cholesteatoma
  - CWU and CWD residual cholesteatoma incidence is same for them*
- CT not routine
- CWU more likely in children, avoid bowl
- CWD more likely in elderly, “simple and safe”
- FN Monitoring only in impending complication or revision performed “elsewhere”

* Per Sheehy from Pensak
Functional Results of CWD

- Retrospective review of 259 cases by Kos et al.
- f/up avg
  - 10 x for 1st 6 mo.
  - 2 x per yr for 6 yr after
  - <2 x per yr for 6 yr after that
- Surgical revisions for:
  - 6.1% for residual or recurrent cholesteatoma
  - 7.3% for TM perforations
  - 12.2% to improve hearing
- 1-24 yr after surgery
  - 95% were dry and self cleaning
  - 5% with persistent otorrhea

Kos et al in 2004
Functional Results of CWD

• Hearing outcomes
  ◦ 41.3% unchanged
  ◦ Increase
    • 15.4% had 10-19 dB increase
    • 11.5% had 20-29 dB increase
    • 3.8% had >29 dB increase
  ◦ Decrease
    • 11.9 had 10-19 dB loss
    • 6.5% had 20-29 dB loss
    • 9.2% had >29 dB loss
  ◦ 0.7% (n=2) had >60 dB SNHL

Kos et al in 2004
Functional Results of CWD

- 0.3% (n=1) case of FN paralysis
- 1.5% of persistent vertigo

CWD adequately treats cholesteatoma with acceptably low rates of complications.

Kos et al in 2004
Status of Canal Wall

• Retrospective chart review of 486 ears in 6 yrs at House (Syms&Luxford).
  ◦ CWU in 68.5%
  ◦ CWD in 31.3%
  ◦ Planned Procedures in CWU and CWD
    • 341 (70.2%) had planned 2nd procedure
    • 21 (14.3%) had planned 3rd procedure
    • Residual Dz in 26.9% of planned 2nd procedures
    • Residual Dz in 2.7% of planned 3rd procedures
    • Residual Dz in 66/203 (32.5%) of planned 2nd procedures for residual cholesteatoma
    • Residual Dz 3/8 of planned 3rd procedures for residual cholesteatoma
  ◦ Unplanned Procedures in CWU and CWD
    • 6 ears had unplanned 2nd procedure
    • 21 ears had unplanned 3rd procedure
    • 3 ears had unplanned 4th procedure
    • Cholesteatoma found in 12 (40%) of the 30 unplanned procedures
  ◦ Age
    • <10 yo, 11.8% had CWD
    • >10 yo, 29.2% had CWD

Syms and Luxford paper 2003
Status of Canal Wall

- CWD – 14.6% had residual cholesteatoma
  - 6-13% has been reported in literature
- CWU – 3.2% had residual cholesteatoma AFTER 2nd planned procedure
- CWU results in 10% of pts undergoing addition procedures in addition to planned 2nd procedure.

- Syms and Luxford paper 2003
- Most common location of cholesteatoma was the epitympanum (84.8%) and middle ear (78%)
Primary vs. Secondary OCR

- Reality – either depends on situation
- Primary
  - Avoid period with CHL
- Secondary
  - Permits ME and TM to heal
  - Motivation for pt to return for 2nd procedure
  - Concern for post-op aeration with ET at initial surgery
  - Previous failure of OCR
  - Damaged mucosa -> scaring, fibrosis, retraction
- Black et al. in retrospective study undergoing cholesteatoma removal performed OCR at first procedure, unless severe disease was present (4%, 19 cases).
Intraoperative FN Monitoring

- 1990 survey showed most experienced otologists do NOT believe FN monitoring is obligatory.
  - Extra expense and time
  - “Silent” transections occur
  - May start use intra-op if difficult

- Others argue FN course if not predictable.
  - “safety net”
  - Routine use for experience
  - Medical/legal reasons
  - Feel bad if FN cut and did not use

- Intermediate course used by others
  - Use for revision cases
  - Previous postop FN weaknesses
  - Preop FN weakness
  - Preop CT shows abnormalities such as dehiscence in fallopian canal
Transcanal Anterior Atticotomy

- Indicated for cholesteatoma limited to only middle ear, ossicular chain, epitympanum
- Procedure involves:
  - Elevation of tympanomeatal flap with removal of scutum
  - Aditus obliterated
  - Reconstruction of lateral attic wall with cartilage to prevent retraction
Transcanal Anterior Atticotomy
Bondy Modified Radical Mastoidectomy

- In classic modified radical mastoidectomy
- Epitympanum and canal wall down mastoidectomy and EAC are converted into common cavity
- TM and ME are left undisturbed
Mastoid Obliteration

- Soft tissue, bone pate, biocompatible materials used to fill space of mastoid cavity.
  - Muscle, ceramics, hydroxyapatite granules, demineralized bone matrix, bioactive glass
- Obliteration prevents re-retraction of TM into attic or mastoid.
- Decrease air absorption from ME and mastoid. -> prevent retraction pocket.
Canal Wall Reconstruction
Tympanomastoidectomy with Mastoid Obliteration

- Gantz et al. performed in 130 ears at Iowa.
- Original procedure by Mercke.
- Posterior canal wall removed with miter cuts by saw.
- Bone pate used for obliteration.
Canal Wall Reconstruction Tympanomastoidectomy with Mastoid Obliteration

- 2\textsuperscript{nd} look ossiculoplasty in 78%
- No recurrence in 98.5%
  - 1.5% had revision CWD
- Infection rate from 14.3% to 4.5% after 48 hr post op IV abx started
Complications of Surgery

- Facial paralysis/paresis
- Dysguesia
- Vertigo
- Hearing loss
- Tinnitus
- Recurrent/residual cholesteatoma
- CSF leak
- Meningitis
Conclusion

- Cholesteatoma pathogenesis is debated but a combination of proliferation and retraction theories likely.
- Goal is to first have a safe ear and then to have a well functioning ear.
- The best procedure for cholesteatoma has been debated. Ultimately, management is individualized to the patient with surgeon experience taken into account.
Bibliography