KMJ

Clinical Audit

An audit on pediatric cholesteatoma

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Abstract: Cholesteatoma is a complex condition characterized by the abnormal presence of keratinizing squamous epithelium in the middle ear or mastoid, accompanied by subepithelial connective tissue and accumulating keratin debris. Despite its misleading name, cholesteatoma contains neither cholesterol crystals nor tumor tissue. This condition is particularly aggressive in children and, if left untreated, can lead to bone erosion and serious complications. The pathophysiology of bone resorption in cholesteatoma involves multiple mechanisms, including enzymatic-induced necrosis, cytokine-mediated bone remodeling, prostaglandin-induced changes, and potential bacterial involvement. This study aims to describe our methodology and outcomes in treating pediatric patients with cholesteatoma, addressing the unique challenges posed by the condition in this age group. By examining our approach and results, we hope to contribute to the development of more effective management strategies for pediatric cholesteatoma, ultimately improving patient outcomes and reducing the risk of complications.

Keywords: Ear disease; otitis media; chronic otitis media; cochlear implants; hearing loss

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1. Introduction

The ear is a complex sensory organ composed of three main parts: the external ear, middle ear, and internal ear. The external ear consists of the visible portion, known as the pinna, and the ear canal. The middle ear, a small air-filled cavity, houses three tiny bones called ossicles: the malleus (hammer), incus (anvil), and stapes (stirrup). These ossicles form a chain that transmits sound vibrations from the eardrum to the inner ear. The middle ear is further divided into three regions: the epitympanum (upper portion), mesotympanum (middle portion), and hypotympanum (lower portion). This anatomical arrangement allows for efficient sound conduction and amplification. The internal ear, also known as the inner ear, contains the cochlea and semicircular canals, which are responsible for converting sound waves into electrical signals and maintaining balance, respectively.

Facial nerve leaves the brainstem at pontomedullary junction; it travels through the posterior cranial fossa and enters the IAM. At the fundus of the IAM, it takes a turn posteriorly forming a genu, from the genu the nerve passes backward above the oval window and below the SCC till the level of pyramidal eminence (2nd genu). The nerve continues vertically downward along the posterior wall of tympanic cavity and leaves the temporal bone through the stylomastoid foramen

What is cholesteatoma?

Cholesteatoma

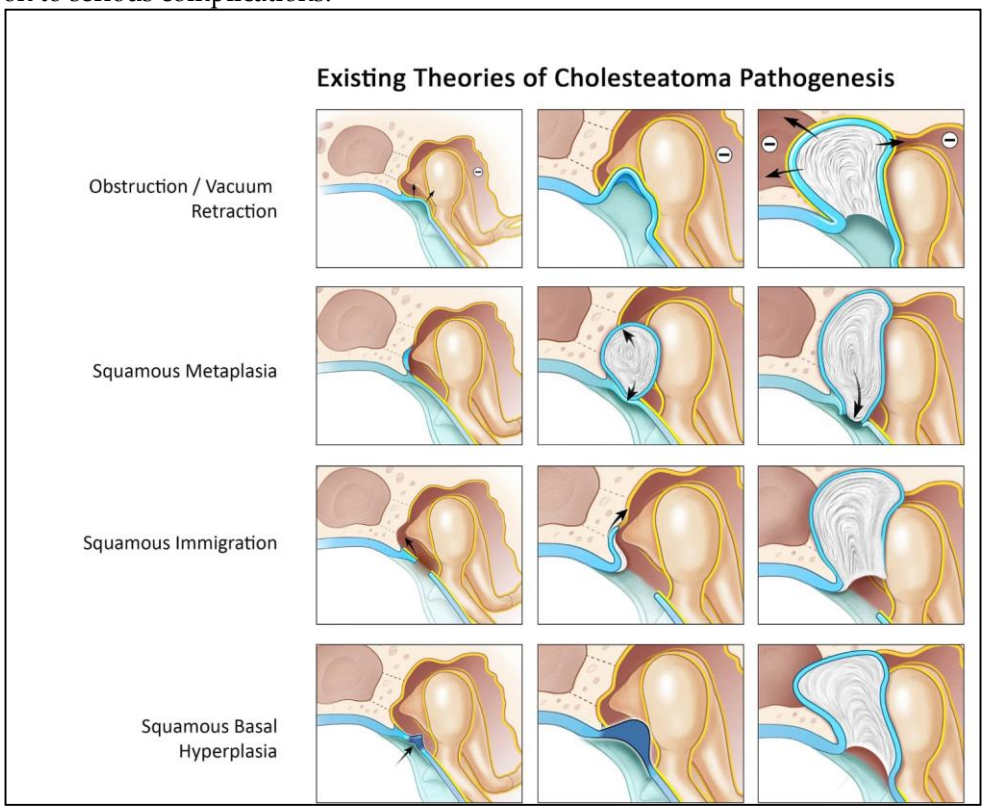
The presence of keratinized squamous epithelium in middle ear or mastoid is characteristic of cholesteatoma – "Skin in the wrong place"

The term 'cholesteatoma' is a misnomer; it contains neither cholesterol crystals nor a tumor.

Definition of Cholesteatoma

Cholesteatoma is a mass formed by keratinizing squamous epithelium in the middle ear and/or mastoid, subepithelial connective tissue and by progressive accumulation of keratin debris with/without surrounding inflammatory reaction.

It is aggressive in children, if left untreated, can cause bone erosion which in turn can lead on to serious complications.



Bone resorption in Cholesteatoma

1) Enzymatic-induced necrosis and resorption - Collagenase, Acid phosphatase, proteolytic enzymes
2) Cytokines induced bone remodelling – IL-1a and 1b, TNF-alpha, TNF-beta
3) Prostaglandin induced bone remodelling - PG E2
4) Pyogenic osteitis theory – Pseudomonas aeruginosa, Streptococcus sp, proteus, E.coli
5) Pressure necrosis theory – unlikely

Objectives

To describe our methodology and outcome in treating pediatric patients with Cholesteatoma.

2. Materials and Methods

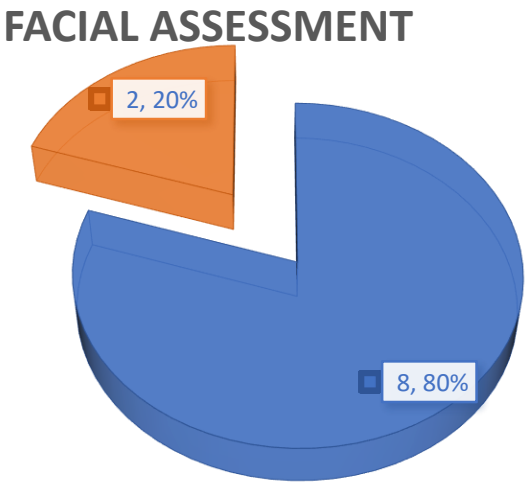
1) A retrospective study was carried out from July 2023–June 2024.
2) The study includes 10 patients, of whom
   4 were boys
   6 were girls
3) Between the age of 7 and 16 years.

The evaluated parameters

1) Clinical symptoms
2) Site of infection
3) Extension of the disease

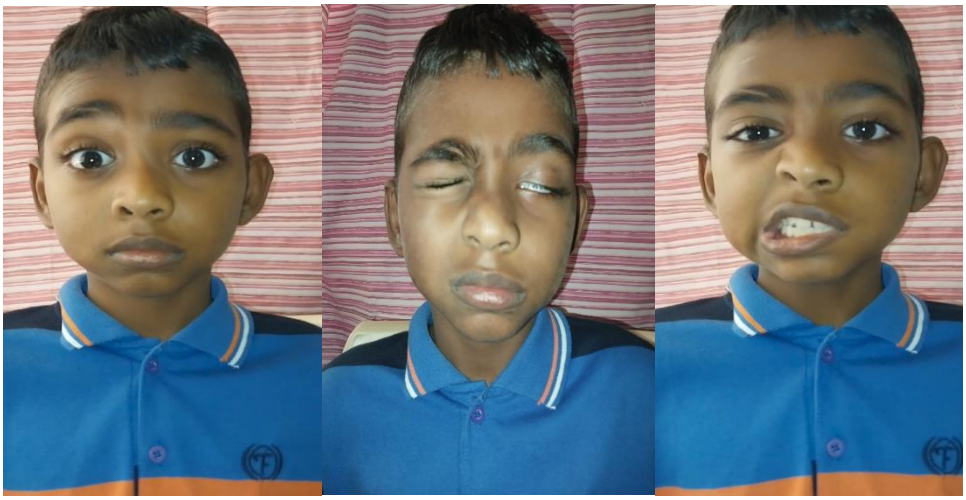
4) Ossicular chain involvement
5) Facial nerve integrity
6) Imaging
7) Treatment modality
8) Outcome
9) Complications

Symptoms and Signs



1) All patients invariably presented with Foul smelling ear discharge and Hard of hearing.
2) 50% of patients had blood stained ear discharge.
3) 20% of patients had facial asymmetry.

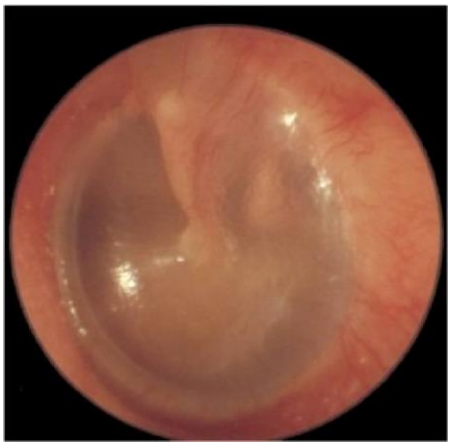
Preoperative Assessment of Facial Nerve



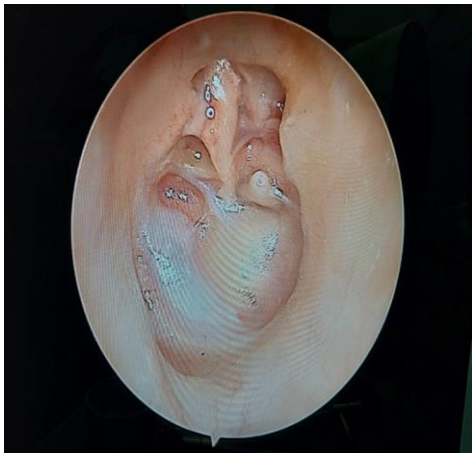
a. House-Brackmann grade 4
b. He also had Giddiness, Vomiting, Headache

(1) - Complete left eye closure only with maximum effort
(2) - Asymmetry of mouth with maximum effort
(3) - Inability to blow the cheek
House-Brackmann grade 3

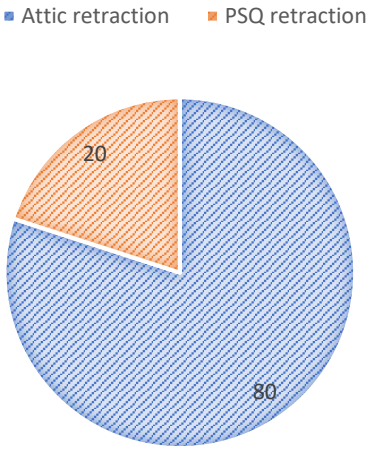
Otoendoscopy

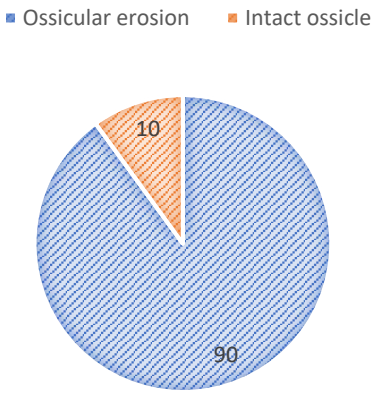


Normal TM

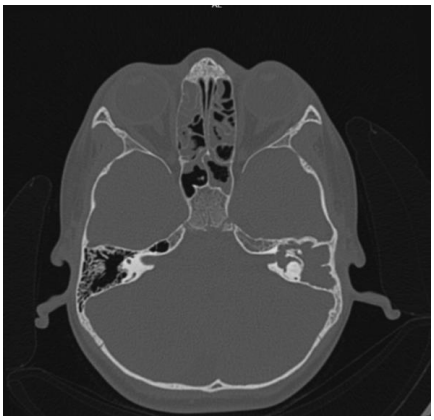


Retracted TM with eroded incus



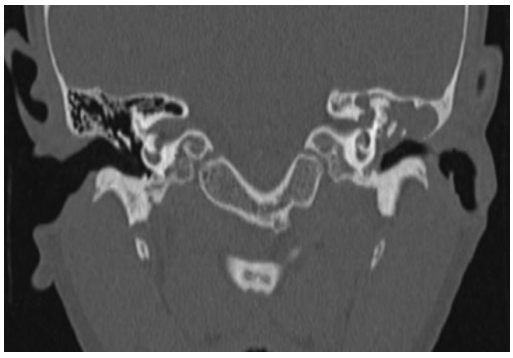


CT Temporal Bone – Axial Plane

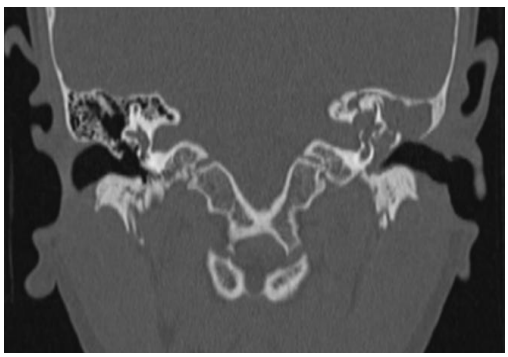


Soft tissue mass extending deep to involve the semicircular canals, encompassing the labyrinthine segment of fallopian canal and causing erosion of internal auditory meatus

CT Temporal Bone – Coronal Plane



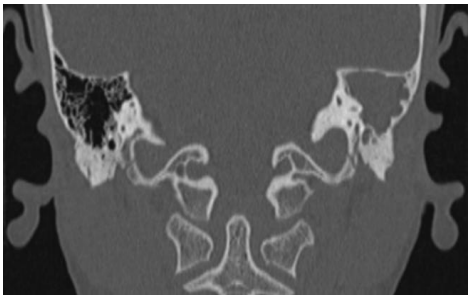
Erosion of Lateral epitympanic wall (the scutum). Erosion of ossicles



Sagging of Posterior wall of external auditory canal Soft tissue mass in middle ear

CT Temporal Bone – Coronal Plane

Extensive bone destruction of the mastoid mimicking the appearance of mastoidectomy–Automastoidectomy



Diagnosis

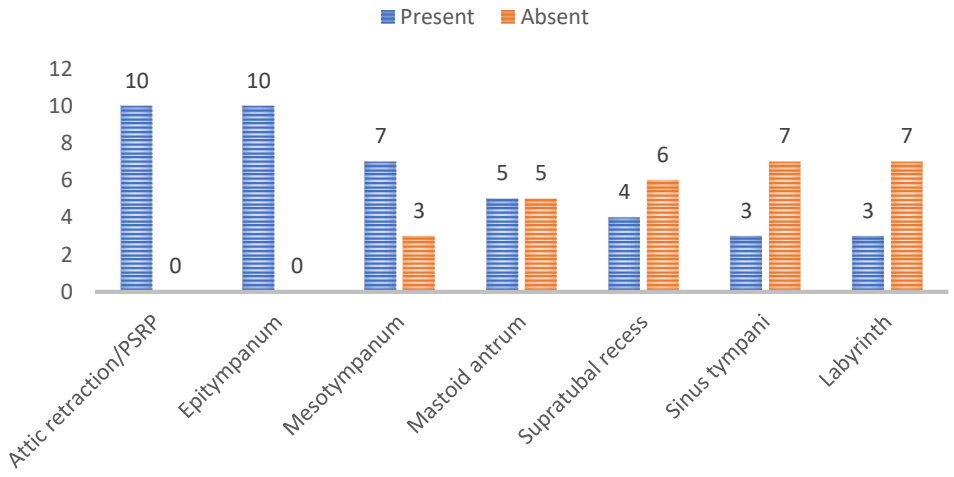
Diagnosis	No. of. Cases
Chronic otitis media with cholesteatoma, active, squamosal disease with facial nerve compression	2
Chronic otitis media with cholesteatoma, active, squamosal disease with intact facial nerve	8

Management

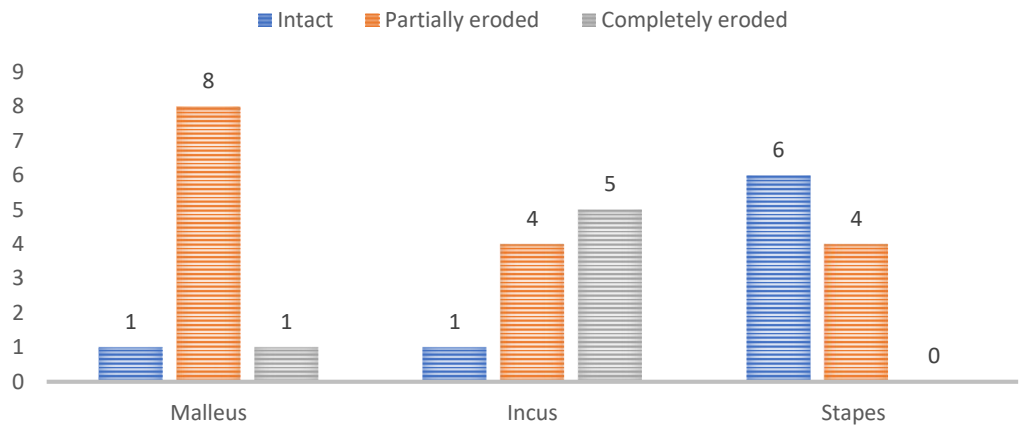
Surgery	No. of. Cases
Canal wall down mastoidectomy, tympanoplasty with facial nerve decompression	2
Canal wall down mastoidectomy with tympanoplasty	8

Intraoperative Findings

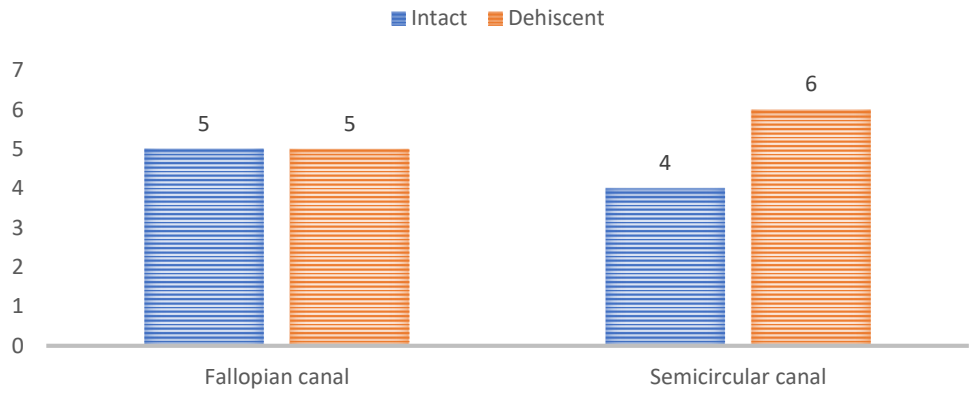
Cholesteatoma Site of Involvement



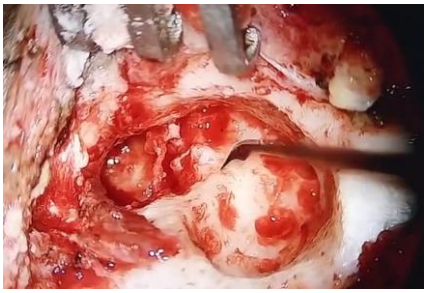
Ossicular status



Fallopian Canal and Semicircular Canal Status

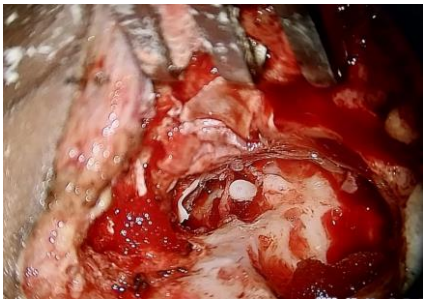


Left ear CWD Mastoidectomy cavity of a 7-year-old girl



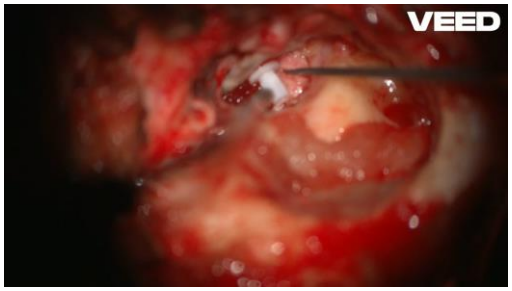
Handle of malleus with head being nipped off
Dome of horizontal semicircular canal
Chorda tympani nerve
Head of stapes

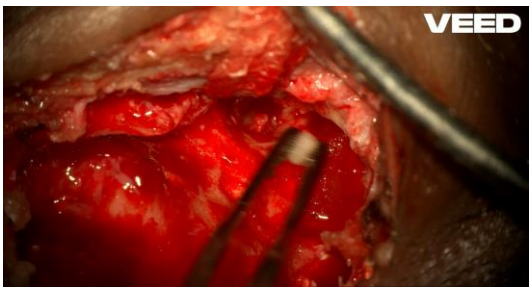
Ossicular Chain Reconstruction



Sculptured head of malleus placed over head of stapes

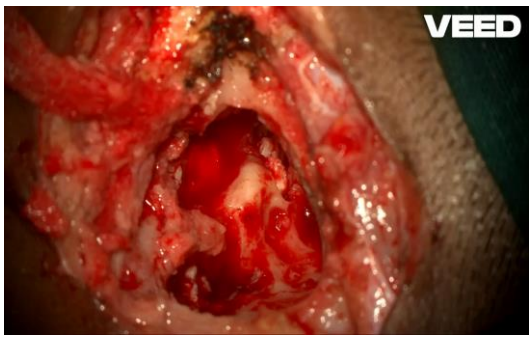
Graft materials	No.of. Cases	Placed over	Tympanoplasty type
Harvested conchal cartilage	2	Head of stapes	III
Septal cartilage	1	Head of stapes	III
Sculptured Malleus	1	Head of stapes	III
Sculptured Incus	1	Head of stapes	III
Sculptured Incus	2	Footplate of stapes	IV
PORP	1	Head of stapes	III
TORP	1	Footplate of stapes	IV



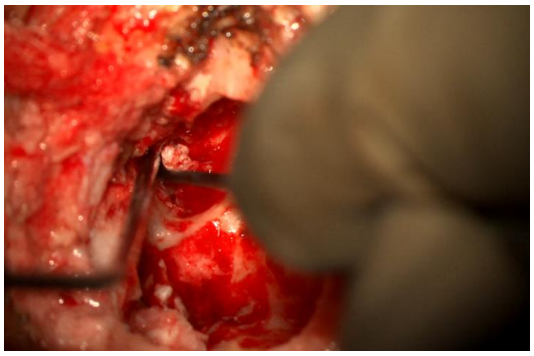


Left ear CWD Mastoidectomy of a 9-year-old boy

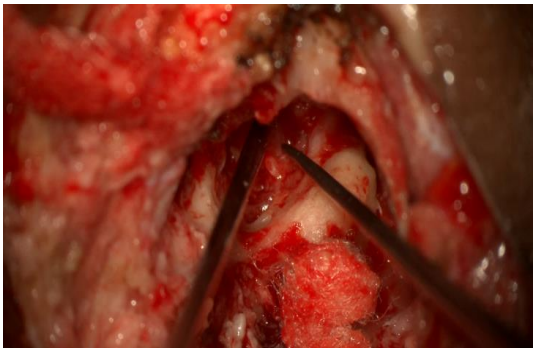
Cholesteatoma flakes in supratubal recess



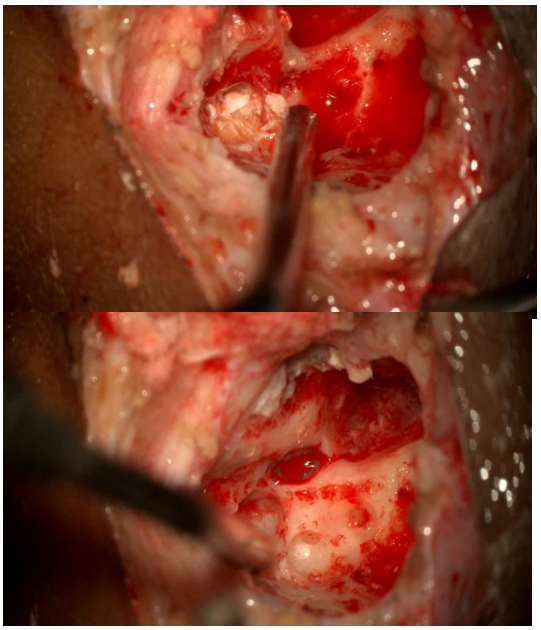
Disease clearance from sinus tympani



Granulation tissue being dissected from the head of stapes



Cholesteatoma flakes filling the mastoid tip cells



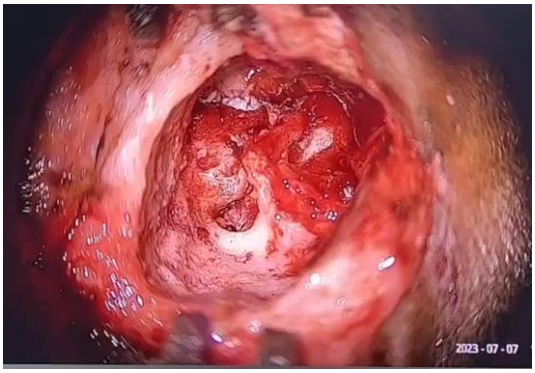
Mastoid cavity after disease clearance

Right ear CWD Mastoidectomy of a 16-year-old girl

Dome of horizontal semicircular canal eroded in the superior aspect

Facial nerve being exposed from 1st genu till chorda tympani branching

Cholesteatoma matrix in superior semicircular canal



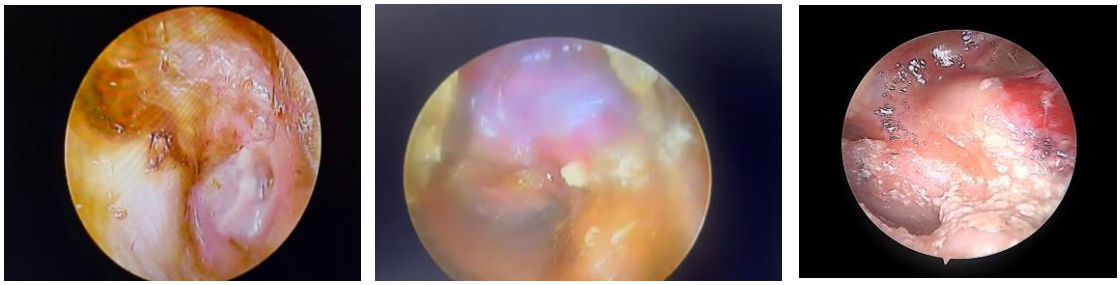
Pre OP – HB Grade 4

Post OP – Normal

Pre OP – HB Grade 3

Post OP – Normal

Otoendoscopy of Postoperative Mastoid Cavity



Comparison

Variables	Our study	Acta Ital
Number of patients	10 in 1 year	36 in 6 years
Age group	7–16 years	6–14 years
Surgery		
Canal wall down mastoidectomy	100%	39.39%
Canal wall intact mastoidectomy	0	60.60%
Complications		
Residual disease	Nil	18.18%
Recurrence	Nil (On Avg 5 months follow-up)	6.06%
Hearing		
Subjective test	Hearing improvement present	Hearing improvement present
Objective test (Audiometry)	Not done	ABG between 0 and 20 db in 54%

Surgery

Primary role of surgery	Secondary role of surgery
Remove all original cholesteatoma squamous epithelial matrix Prevent further erosion and complications Give a dry, watertight ear Give a ear that will be self-cleaning Prevent the occurrence of recurrent cholesteatoma.	Improve hearing (aim is to restore the best possible hearing in an ear that will never be normal).

What to be cautious in pediatric Cholesteatoma?	Why/how?
More extensive and destructive in children than adults	Hence early intervention is important to prevent further damage
Increases risk of complications	More prone to facial paralysis, Meningitis, Brain abscess
Impact on development	if left untreated, it affects speech, language and cognitive development
Difficulty in diagnosis	Due to nonspecific symptoms
Surgical challenges	Surgery can be more complex in children due to extensive spread in developing temporal bone
High recurrence rates	More likely to recur due to poor Eustachian tube function. Hence long term follow-up will be necessary
Impact on hearing and balance	As it cause erosion of ossicles and labyrinth, patient develops hard of hearing and giddiness respectively
Potential for congenital associations	Down's syndrome, Craniofacial syndrome, di-George syndrome

Emotional and psychological impact	Children and families may experience emotional distress and concerns about hearing

3. Conclusion

Management of pediatric cholesteatoma requires a highly individualized approach that takes into account anatomic, clinical and social factors to determine the most successful treatment paradigm.

Prompt evaluation, management, and early intervention are very important to avoid life- threatening events.