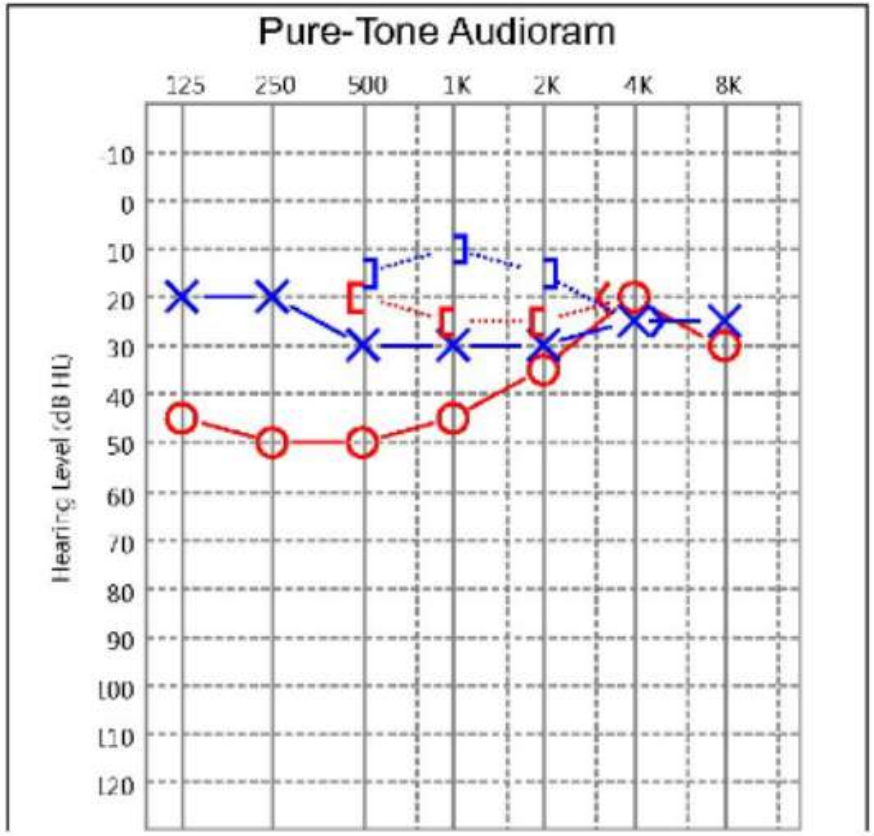
ENT journal reading

Presenter: PGY2 莊政儒
Supervisor: VS 鄭評嘉

Patient profile

- Chart No: A76518
- Name: 高O婷
- Age: 43
- Gender: female
- Hx: (-)
- CC: hearing impairment for long, progress for half a year
  tinnitus(+/-)

2025/4/12 PTA

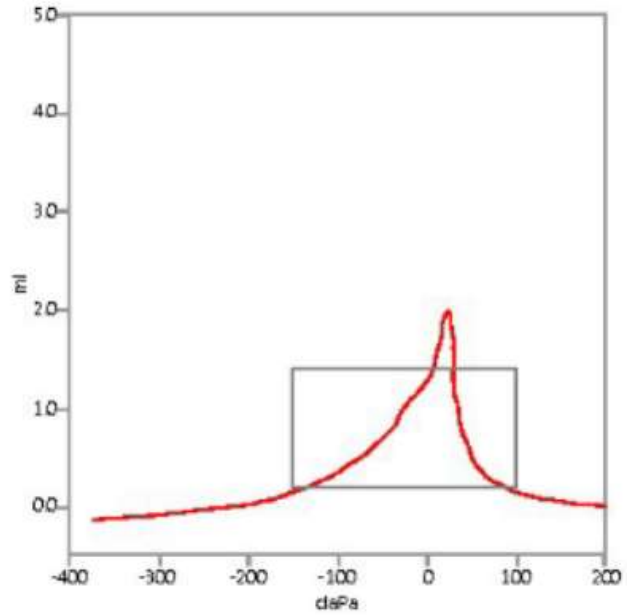


AC PTA 38 dB BC PTA 22 dB SII 46 %

AC PTA 29 dB BC PTA 16 dB SII 76 %

Tympanometry Graph Right

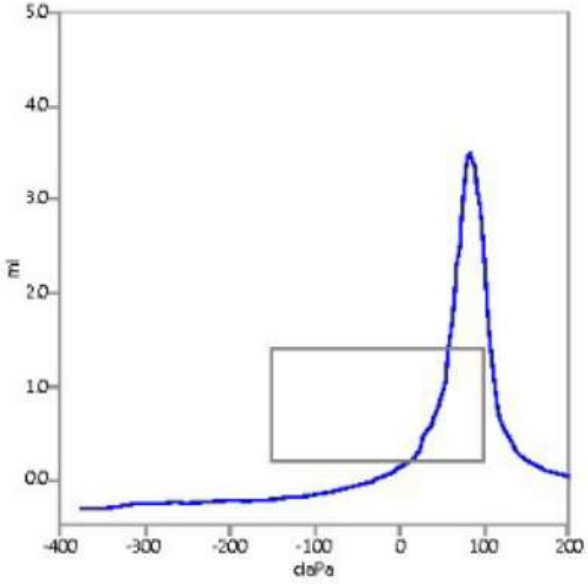
Right Ytm 226 Hz



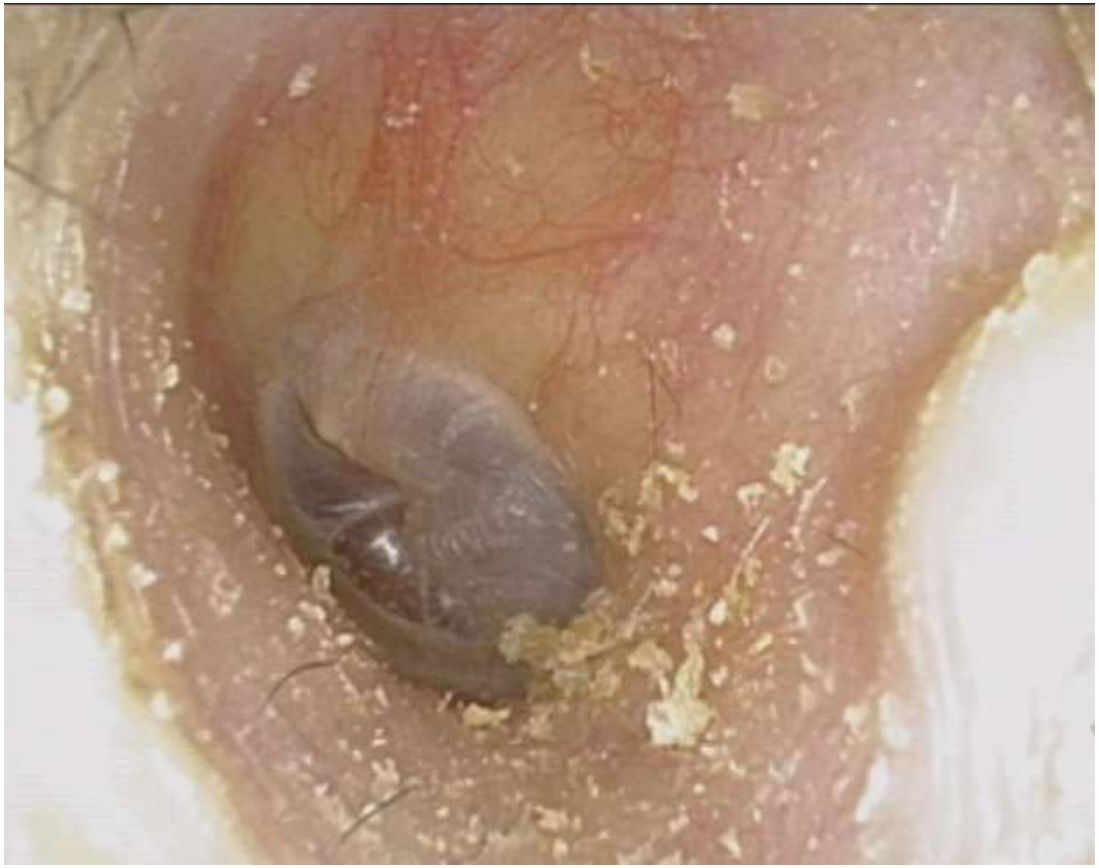
	Tymp			Sweep daPa/sec		ECV	Peak daPa	Peak ml	Width daPa	Type
1	226 Hz	Y	←	600/200		1.4	25	2.00	N/A	

Tympanometry Graph Left

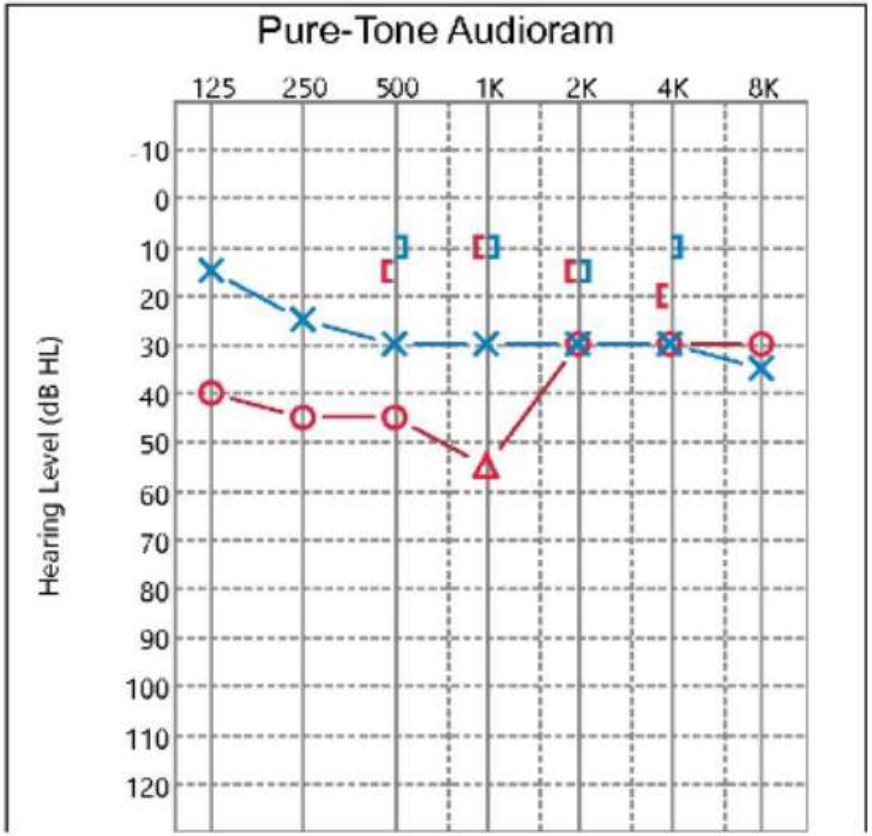
Left Ytm 226 Hz



	Tymp			Sweep daPa/sec		ECV	Peak daPa	Peak ml	Width daPa	Type
1	226 Hz	Y	←	600/200		1.5	85	3.50	N/A	



2025/4/19 PTA



AC PTA 40 dB BC PTA 15 dB SII 42 %

AC PTA 30 dB BC PTA 11 dB SII 72 %

Finding, possible diagnosis and plan

- 43 y/o female, hearing impairment for long, progress for half a year

- Test finding
  - right SNHL, bil ABG
  - tymp: bil type Ad
  - PTA: R 38dB, L 29dB

    => suspect otosclerosis

- Plan
  - suggest Hearing advice use first
  - regular f/u 1 yr or any discomfort

SAGE journals

Review

Diagnosis and Management of Unexplained Conductive Hearing Loss With Intact Tympanic Membrane: A Systematic Review

Ear, Nose & Throat Journal
1–12

DOI: 10.1177/01455613241262129
journals.sagepub.com/home/ear
Sage

Yuzhe Hao, MM[1], Xuan Yu, MS[1], Yi Wang, MD[1], Davood Hosseini, MD[2], Shimin Zong, MD[1], Haiying Sun, MD[1], and Hongjun Xiao, MD[1]

Ear Nose Throat J. 2024 Jun 19:1455613241262129.

Conductive Hearing Loss (CHL) overview

- CHL results from abnormal sound transmission through the external, middle, or inner ear.

- Common causes:
  - Genetic abnormalities
  - Embryonic developmental issues
  - Inflammation (most common)
  - Trauma
  - Tumors

Unexplained Conductive Hearing Loss (UCHL)

- Definition:
  - CHL without clear inflammatory, traumatic, or tympanic membrane abnormalities.

- Primarily manifests as CHL, additional symptoms may include:
  - Tinnitus
  - Ear fullness
  - Dizziness
  - Ear pain

Unexplained Conductive Hearing Loss (UCHL)

- Types of UCHL
  - Congenital UCHL:
    - Often detected in childhood or found incidentally
  - Acquired UCHL (e.g., otosclerosis)
    - Develops gradually over time

Unexplained Conductive Hearing Loss (UCHL)

- Diagnostic challenges
  - Normal tympanic membrane
  - Absence of otitis media or trauma history
  - Lack of significant abnormal clinical, audiological, or imaging findings

- Diagnosis and Treatment:
  - Exploratory tympanotomy is the primary diagnostic & therapeutic approach for UCHL

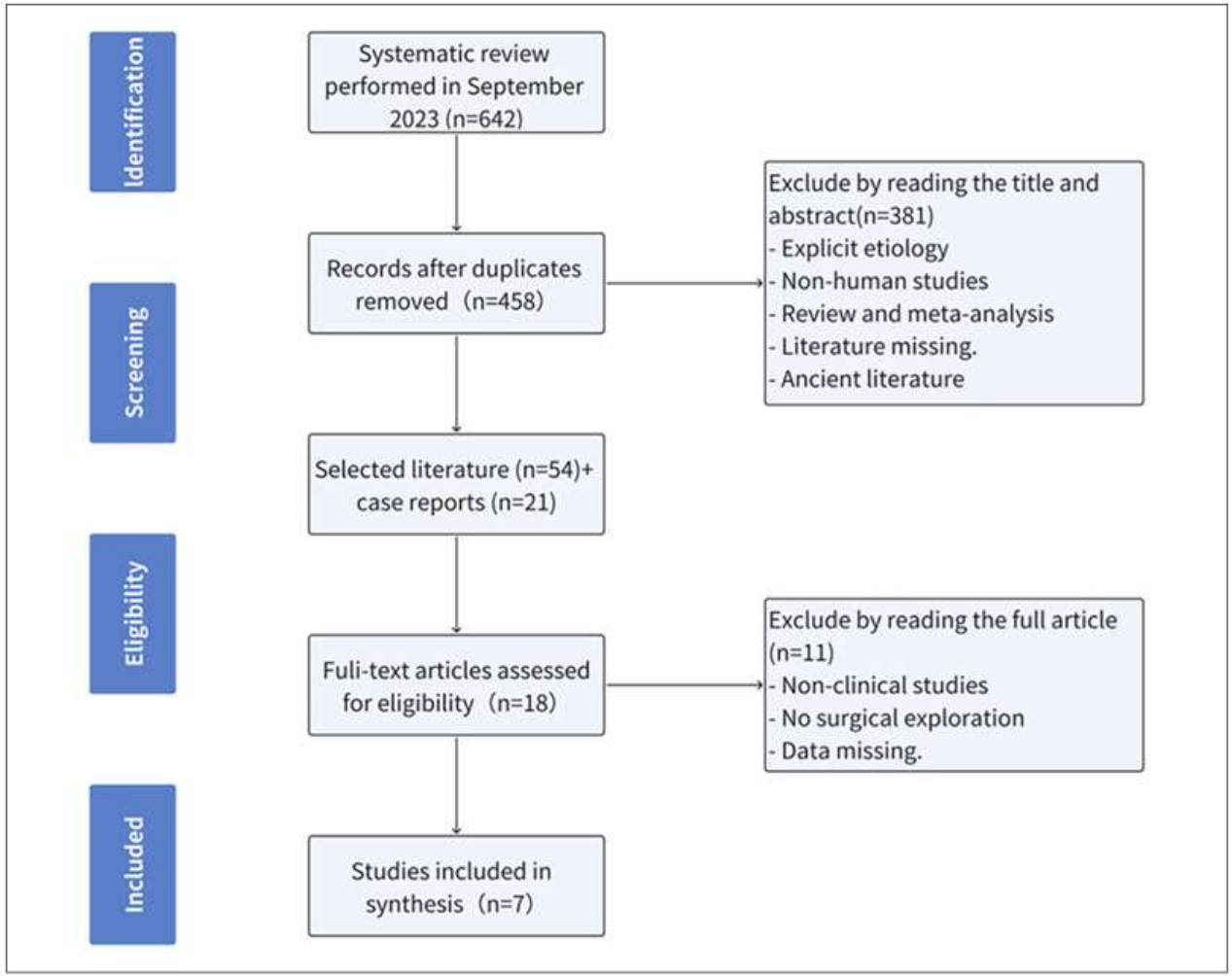
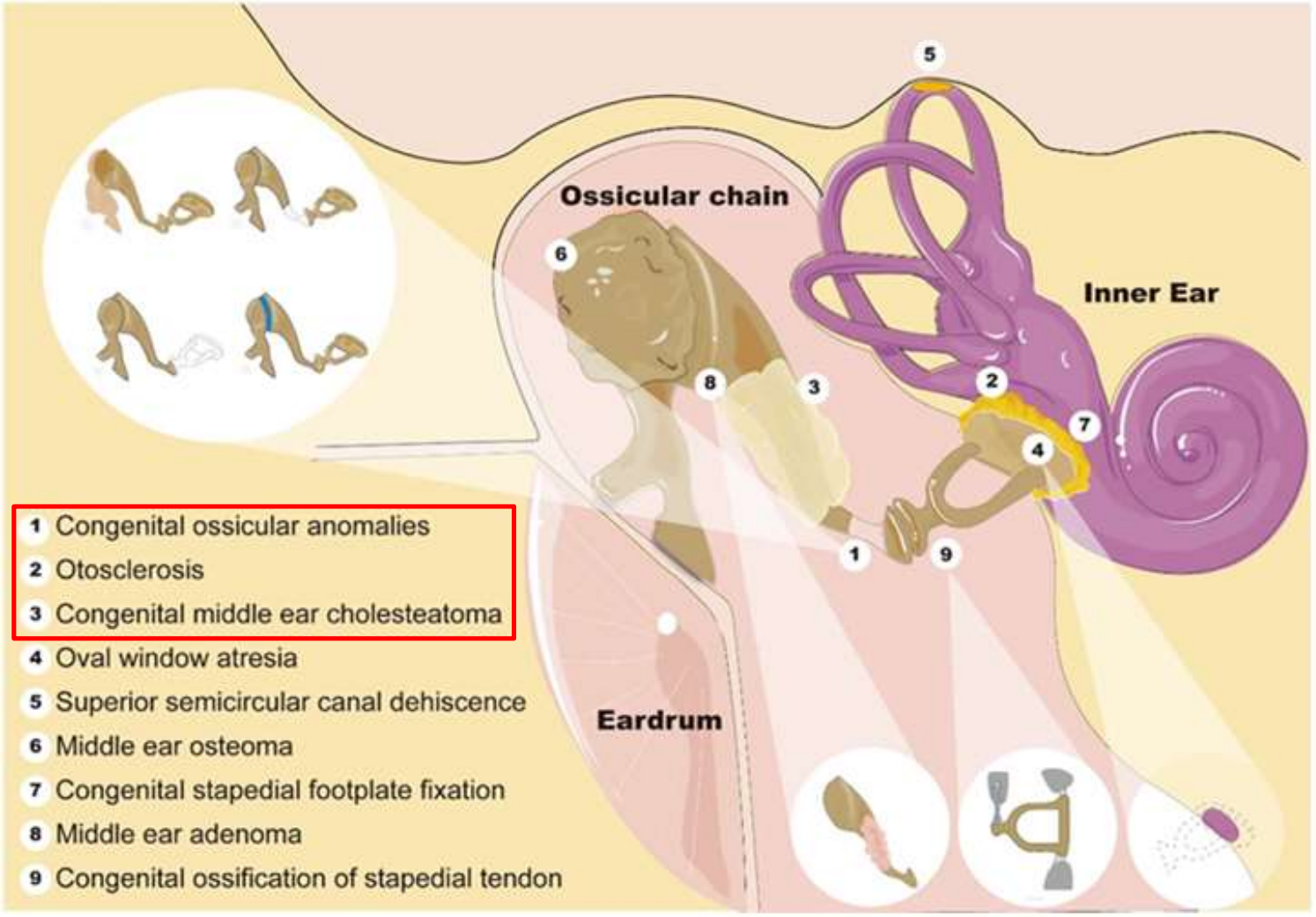


Figure 1. Project flow chart. Literature evaluation and selection, according to PRISMA criteria (http://www.prisma-statement.org/). PRISMA, Preferred Reporting Items for Systematic Reviews and Meta-Analyses.


Ossicular chain
Inner Ear
Eardrum
1 Congenital ossicular anomalies
2 Otosclerosis
3 Congenital middle ear cholesteatoma
4 Oval window atresia
5 Superior semicircular canal dehiscence
6 Middle ear osteoma
7 Congenital stapedial footplate fixation
8 Middle ear adenoma
9 Congenital ossification of stapedial tendon

UCHL- Congenital Ossicular Anomalies (COA)

- Most common cause of unexplained conductive hearing loss (UCHL).
- Originates from abnormal embryonic development of first & second pharyngeal arches.
  - Stapes superstructure deformity: 50.9%
  - Incus deformity: 48.0%
  - Malleus deformity: 18.3%
- Clinical
  - Typical: Moderate to moderate-severe unilateral hearing loss
  - Tinnitus
  - Ear fullness

UCHL- Congenital Ossicular Anomalies (COA)

- Diagnosis
  - High-resolution computed tomography (HRCT) of the temporal bone is essential.
  - Effective in detecting:
    - Long crus deformity of the incus
    - Stapes superstructure deformity
    - Oval window atresia (OWA)
  - Ossicular fixation is often missed in imaging

- Treatment
  - Primary treatment is endoscopic ossicular reconstruction.

UCHL- Otosclerosis (OTS)

- Spongy degeneration in the bony labyrinth of the inner ear, cause remains unknown.
  - Potential Contributing Factors:
    - Genetic predisposition
    - Environmental influences
    - Viral infections
    - Autoimmune mechanisms
  - Most commonly affects middle-aged and women, often bilateral

- Symptom
  - Progressive conductive hearing loss (CHL) of moderate to moderate-severe degree
  - When the cochlea is involved, mixed symptoms of
    - Tinnitus
    - Ear fullness

UCHL- Otosclerosis (OTS)

- Radiologic Findings (High-Resolution CT - HRCT):
  - Abnormal low-density areas around the vestibular window and cochlea
  - Sensitivity: 61.9%~76.3%
    - Variability may be due to disease severity and equipment differences
- Diagnosis:
  - Based on clinical symptoms and audiologic examination
  - Definitive confirmation requires intraoperative evaluation of stapes footplate mobility
- Treatment:
  - Fenestration of the stapes footplate
  - Piston prosthesis implantation
  - Outcome:~90% of patients experience significant hearing improvement

UCHL- Congenital Middle Ear Cholesteatoma (CMEC)

- Invasive lesion believed to arise from residual ectodermal squamous epithelium.
  - Mostly affecting children, 2% to 5% of all middle ear cholesteatoma
- Symptom
  - Typically causes unilateral conductive hearing loss (CHL) in childhood.
  - As progresses, symptoms include: Ear fullness, Otorrhea, Dizziness, Facial paralysis

- Endoscopic findings:
  - Pearly white, mass-like structures on the inner surface of the eardrum
- HRCT findings:
  - Intra-tympanic, circular like soft tissue density shadows

- Treatment: endoscopic endoaural incision complete removal of the cholesteatoma

UCHL- Oval and Round Window Atresia (ORWA)

- ORWA is a congenital malformation of the labyrinthine wall.
- Presents as moderate to severe conductive hearing loss (CHL) starting in childhood.
  - 69.1% lacked the long crus of the incus
  - 61.9% lacked the stapes
  - 45.1% had stapes superstructure deformities
  - 69.1% exhibited facial nerve abnormalities

- HRCT: Irregularities or absence of oval and/or round window ± middle ear anomalies

- Treatment:
  - Vestibule fenestration is the main surgical treatment
  - Effectiveness is variable and often suboptimal, long-term success rates range from 12.5% to 75%

UCHL- Superior semicircular canal dehiscence (SSCD)

- SSCD is a bony defect in superior semicircular canal, by genetic, infectious, or traumatic.
- Most commonly affects middle-aged and elderly individuals.

- Typical symptoms (reflect vestibulocochlear involvement)
  - Episodic vertigo
  - Conductive hearing loss (CHL)
  - Tinnitus

- HRCT is useful for detecting the bony defect, small or subtle defects may be missed

- Treatment
  - Mild: conservative treatment
  - Severe: Superior semicircular canal plugging, canal wall reinforcement

UCHL- Middle ear osteoma

- A rare, slow-growing osseous tumor in tympanum, most in younger individuals
- Most commonly arises from the tympanic promontory (39.5%).

- Symptoms:
  - Unilateral conductive hearing loss (CHL)
  - Tinnitus
  - Sensation of ear tightness
  - Small osteomas may remain asymptomatic.

- Otoscopy: May reveal a creamy-white mass on the inner surface of the eardrum.

UCHL- Middle ear osteoma

- HRCT
  - localized, high-density bony lesion in the tympanum.
  - May demonstrate:
    - Fusion with the ossicular chain
    - Attachment to the tympanic wall

- Intraoperative Findings
  - During exploratory tympanotomy, the tumor appears as a bone-like neoplasm.
  - It is often fused to adjacent bone structures, making complete removal difficult.

- Treatment and Outcomes
  - Ossicular reconstruction can be performed following tumor removal.
  - Majority of patients experience improvement in hearing after surgery.

UCHL- Congenital Stapedial Footplate Fixation (CSFF)

- A non-progressive form of conductive hearing loss (CHL) that begins in childhood.
- Caused by abnormal development of annular ligament of oval window during embryonic.

- Clinical Features
  - Poor hearing, often associated with an air-bone gap (ABG) > 30 dB

- Differentiation from Juvenile Otosclerosis (JO)
  - CSFF:
    - Non-progressive
    - Typically no family history
  - JO:
    - Progressive
    - May have a familial link

UCHL- Congenital Stapedial Footplate Fixation (CSFF)

- Radiological Findings (HRCT)
  - Subtle abnormalities may be more telling than direct footplate fixation:
    - Pyramidal eminence abnormalities
    - Narrowed aperture of the stapedial tendon

- Treatment and Outcomes
  - Surgical approach is similar to that used for otosclerosis (OTS).
  - Surgical outcomes for CSFF are generally less favorable compared to OTS.

UCHL- Neuroendocrine Adenomas of the Middle Ear (NAME)

- Rare, benign, and slow-growing primary tumors of the middle ear

- Symptom
  - Conductive hearing loss (CHL), most common
  - Ear fullness
  - Tinnitus
  - Ear pain

- Otoscopy Findings:
  - Non-pulsatile white or pink mass on the inner surface of the tympanic membrane

UCHL- Neuroendocrine Adenomas of the Middle Ear (NAME)

- HRCT and MRI
  - Nonvascular mass surrounding the ossicles
  - No bone destruction
  - Possible extension into the posterior tympanum or Eustachian tube

- Treatment Strategy
  - Complete surgical resection is the primary treatment
  - Postoperative pathological and immunohistochemical analysis is essential to:
    - Confirm diagnosis
    - Evaluate tumor characteristics
  - Adjuvant radiation or chemotherapy may be considered in selected cases

- Long-term monitoring: Recurrence, Progression, Metastasis (rare but possible)

UCHL- Salivary gland choristoma in middle ear

- A rare congenital tumor of the middle ear
  - Believed to result from abnormal development of the second branchial arch
  - Involves heterotopic (misplaced) salivary gland tissue within the tympanic cavity
  - May accompanied by other middle ear malformations

- Symptom
  - Unilateral conductive hearing loss (CHL)
  - Tinnitus
  - Ear fullness
  - Facial paralysis

UCHL- Salivary gland choristoma in middle ear

- HRCT
  - Reveals a well-defined soft tissue mass surrounding the ossicles
  - No evidence of bone destruction
- Treatment
  - Complete surgical excision of the tumor is recommended
  - Followed by ossicular chain reconstruction to restore hearing
- Diagnosis requires pathological examination to confirm
- Prognosis
  - Generally favorable with appropriate surgical treatment

UCHL- Congenital Ossification of the Stapedial Tendon (COST)

- A rare, hereditary middle ear condition
  - ossification (bone formation) of the stapedial tendon

- Symptom
  - Unilateral or bilateral conductive hearing loss (CHL)
  - Can be clinically indistinguishable from:
    - Otosclerosis (OTS)
    - Congenital stapedial footplate fixation (CSFF)

    => Requires exploratory tympanotomy, findings include:
      - Ossified stapedial tendon
      - Limited mobility of the stapes

UCHL- Congenital Ossification of the Stapedial Tendon (COST)

- HRCT:
  - linear, bony density extending from the pyramidal eminence to the stapes superstructure

- Treatment and Outcomes
  - Surgical release of the ossified stapedial tendon can restore stapes mobility
  - Leads to a favorable auditory prognosis

Table 2. Demographic Information.

Author	Years	Nation	Study design	Study time	Number of patients	Average age (y)	Sex
Tomasoni et al[7]	2022	Italy	Retrospective	2011-2019	48	51.08 ± 14.63	F 23, M 17
Zhang and Tong[3]	2021	China	Retrospective	February 2016-February 2019	77	26.40 ± 16.00	F 40, M 37
Zhang et al[4]	2020	China	Retrospective	January 2013-December 2019	83	MD = 7	F 30, M 53
Tan et al[8]	2020	China	Retrospective	January 2018-December 2019	16	20.3 ± 12.8	F 8, M 8
Tang et al[9]	2016	China	Retrospective	April 2011-September 2013	82	26.5 ± 13.7	F 41, M 41
Min and Woo[10]	2015	Korea	Retrospective	January 2009-June 2011	37	41	
Xu et al[5]	2023	China	Retrospective	January 2019-November 2022	179	8.5 ± 3.1	F 54, M 125

Abbreviations: F, female; M, male.

Table 3. Number of Ears for Different Diseases Reported in Each Article.

Author	Number of ears	COA	OTS	CMEC	COA and CMEC	COWA	TOI	AOM	TS
Tomasoni et al[7]	48	17	23						8
Zhang and Tong[3]	82	32	28	6		6	10		
Zhang et al[4]	83	7	2	52			9	13	
Tan et al[8]	16	6	2	5	2		1		
Tang et al[9]	82	40	22	8	3	3	6		
Min and Woo[10]	37	18	10	2			7		
Xu et al[5].	174	31	6	132					5
Total	522	151	93	205	5	9	33	13	13

Abbreviations: AOM, adhesive otitis media; CMEC, congenital middle ear cholesteatoma; COA, congenital ossicular anomalies; COWA, congenital oval window atresia; OTS, otosclerosis; TOI, traumatic ossicular injuries; TS, tympanosclerosis.

Analysis of Articles About UCHL

- Common Symptoms
  - Hearing loss (CHL)
  - Tinnitus
  - Dizziness
  - Ear fullness
  - Ear pain
  - Facial paralysis

- Disease Duration
  - Ranged from 7 days to 50 years

Analysis of Articles About UCHL

- Symptom Patterns by Disease
  - COA
    - CHL in 87.5%–93.0%
    - Tinnitus in 15.6%–30.2%
    - Onset typically in childhood
  - OTS:
    - Progressive CHL in 86.4%–96.4%
    - Tinnitus in 60.1%–90.9%
  - CMEC:
    - Sudden CHL with tinnitus in 50.0%–80.0%

Analysis of Articles About UCHL

- Radiological Diagnosis (Temporal Bone CT)
  - Positive diagnosis rates ranged from 33.8% to 87.1%
    - COA: 28.6%–64%
    - CMEC: 83.3%–100%
      - CMEC more easily identified due to soft tissue density in tympanic cavity

Table 4. Postoperative Hearing Improvement and Complications.

Author	Surgery	PTA (dB)			ABG (dB)			Follow-up time	Postoperative complication
		Pre	Post	△	Pre	Post	△		
Tomasoni et al[7]	Tympanotomy	61.6	42.6		38.39 ± 12.25	14.77 ± 12.38	24.03 ± 16.17		Chorda tympani nerve injury in 7 cases; dizziness in 3 cases
Zhang and Tong[3]	Endoscopic tympanotomy	60.09 ± 8.83	29.77 ± 8.70	30.32 ± 10.01	40.82 ± 8.83	11.83 ± 6.83	28.99 ± 10.33	>3 mo	Taste abnormalities in 5 cases; transient vertigo in 2 cases; short-term tinnitus in 6 cases
Zhang et al[4]	Tympanotomy	42.6 ± 14.9	35.6 ± 12.6					>6 mo	Failure to report
Tan et al[8]	Endoscopic tympanotomy	61.7 ± 6.5	29.8 ± 10.7		36.8 ± 3.2	10.7 ± 6.9		3-12 mo	Taste abnormalities in 1 case; transient vertigo in 1 case
Tang et al[9]	Microscopic tympanotomy	60.0 ± 11.4	32.2 ± 12.1		43.2 ± 12.0	16.3 ± 9.4		>2 y	None
Min and Woo[10]	Endoscopic tympanotomy in 28 cases and microscopic tympanotomy in 9 cases	64.3 ± 16.3	42.0 ± 20.8		39.0 ± 10.8	20.7 ± 12.7			Failure to report
Xu et al[5]	Tympanotomy	50.8 ± 12.9	36.1 ± 14.5		30.8 ± 9.4	20.0 ± 8.6			Failure to report

Abbreviation: ABG, air-bone gap

Table 5. Preoperative and Postoperative Audiological Data of Different Diseases (Excluding Traumatic Inflammation).

Author	Disease	Number of cases	PTA (dB)			ABG (dB)		
			Pre	Post	△	Post	Pre	△
Tomasoni et al[7]	COA	17				39.56 ± 11.14	18.01 ± 14.21	21.58 ± 13.43
	OTS	23				35.33 ± 12.99	9.73 ± 9.99	25.82 ± 18.00
Zhang et al[4]	COA	7	58.2 ± 9.9	49.3 ± 12.1				
	OTS	2	30.2 ± 9.9	28.5 ± 7.7				
	CMEC	52	40.8 ± 13.1	36.8 ± 12.6				
Tang et al[9]	COA	22				47.6 ± 8.9	20.6 ± 10.8	27.0 ± 12.4
	OTS	15				34.1 ± 9.1	13.3 ± 4.4	20.8 ± 9.0
	CMEC	6				54.2 ± 7.7	15.3 ± 3.4	38.9 ± 9.3
	COA and CMEC	3				45.6 ± 21.1	8.3 ± 11.1	37.2 ± 14.4
Xu et al[5]	COA	31	57.1 ± 11.7			35.4 ± 9.8		
	OTS	6	63.5 ± 7.81)			35.3 ± 7.5		
	CMEC	132	47.8 ± 14.5			28.8 ± 10.5		

Abbreviations: ABG, air-bone gap; CMEC, congenital middle ear cholesteatoma; COA, congenital ossicular anomalies; COWA, congenital oval window atresia; OTS, otosclerosis.

Impact on Pediatric Population

- UCHL may negatively affect speech and language development
  - Delayed diagnosis is common due to:
    - Limited cognitive awareness in children
    - Parental unawareness
    - Children's tendency to adapt to hearing loss
  - Average delay from symptom recognition to diagnosis: 2.2 ± 2.9 years
    - CMEC diagnosed fastest (~1.3 years)
    - COA & OTS took longest (~5 years)

    => Delay leads to worse hearing outcomes and higher recurrence (e.g., CMEC)

- Surgical treatment of UCHL in children aged 3–6 years:
  - No increased risk of complications
  - Good prognosis and hearing recovery

Impact on Pediatric Population

- Early Detection
  - Monitor speech delays or abnormal behavior
  - Implement routine hearing screenings for school-age children

- Exploratory tympanotomy should be considered at an appropriate age
  - Surgical decision-making may include:
    - Ossicular reconstruction
    - Auditory implants
  - For suspected COA or OTS:
    - Start with hearing aids
    - Surgery for bilateral cases: 5–6 years old; Unilateral cases: consider after 10 years old
  - Opposing view: early unilateral intervention is beneficial due to:
    - Temporal bone ossification completing by age 6

Surgical Considerations

- Inner ear window procedures:
  - Some recommend waiting until 15 years old
  - Stapes surgery:
    - Safe in children
    - Higher success rate than adults (due to less severe pathology)
  - CMEC surgery:
    - Safe after 3 years old
    - If minimal lesion and <1 year old: defer to 1–2 years
    - If >3 years and suspected CMEC: prompt surgery recommended

Importance & Limitations of Temporal Bone HRCT

- HRCT is essential for preoperative diagnosis of UCHL.
- However, routine HRCT often fails to accurately visualize delicate middle ear structures.
- Risk of misdiagnosis or missed diagnosis due to:
  - Complex middle ear anatomy
  - Disease overlap
- Reported diagnostic accuracy:
  - *40% ~ 62.1% for CMEM
  - **HRCT identified abnormalities in 9/10 UCHL cases, but surgery revealed all had COA or CMEC

*Chin J Otorhinolaryngol Head Neck Surg. 2016;51(5):348-354. ; Ann Otol Rhinol Laryngol. 2020;129(3):216-223.
**ORL J Otorhinolaryngol Relat Spec. 2020;82(3):139-149.

UCHL Etiology and Diagnostic Complexity

- Most congenital UCHL cases stem from genetic/embryonic maldevelopment
- Malformation of 1st or 2nd branchial arches may lead to multiple coexisting pathologies
- Nonspecific symptoms may mask concurrent conditions:
  - Example: CMEC often coexists with COA
  - Example: SFF and SSCD may occur together, and misdiagnosis can lead to poor hearing outcomes
- Recommendation:
  - Carefully evaluate all soft tissue shadows and abnormal densities in HRCT scans

Key Recommendations for Otologists

- Improve imaging interpretation, and look for:
  - Abnormal soft tissue shadows
  - Density differences in key anatomical areas (e.g., promontory, footplate)

- Thorough Intraoperative Exploration:
  - Completely remove pathological tissue
  - Perform ossicular reconstruction where suitable

- Monitor for:
  - Residual tumors
  - Recurrence (especially for cholesteatoma, adenoma)
  - Follow-up should last at least 5 years for high-recurrence conditions

Limitation

- Heterogeneity of included studies
- Small sample size
- Variations in research themes and methods limit data synthesis

Conclusion

- UCHL encompasses a spectrum of diseases with similar clinical presentations
  - Conditions often present with unilateral conductive hearing loss, typically without inflammatory or traumatic history
- Exploratory tympanotomy is the main method for both diagnosis and treatment
- Surgical success depends on:
  - Detailed intraoperative exploration of the middle ear structures
  - Complete lesion removal
  - Effective ossicular chain reconstruction

Back to our patient

- 43 y/o female, hearing impairment for long, progress for half a year

- Test finding
  - right SNHL, bil ABG
  - tymp: bil type Ad
  - PTA: R 38dB, L 29dB

    => suspect otosclerosis

- Plan
  - suggest Hearing advice use first
  - regular f/u 1 yr or any discomfort