

New EAONO Cholesteatoma Classification with imaging illustration

Milan Profant, Katarina Sláviková

EAONO/JOS Joint Consensus Statements on the Definitions, Classification and Staging of Middle Ear Cholesteatoma

Matthew Yung, Tetsuya Tono, Ewa Olszewska, Yutaka Yamamoto, Holger Sudhoff, Masafumi Sakagami, Jef Mulder, Hiromi Kojima, Armağan İncesulu, Franco Trabalzini, Nuri Özgirgin

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Classification of cholesteatoma

- Acquired
- Congenital
- Unclassifiable
(cholesteatoma whose origin cannot be accurately determined)

Congenital cholesteatoma

- Congenital cholesteatoma is typically an expanding cystic mass with keratinizing squamous epithelium located medial to the intact tympanic membrane
- It is assumed to be present at birth, but is usually diagnosed during infancy or in early childhood in patients with no prior history of otorrhea, perforation, or previous ear surgery.
- A history of previous bouts of otitis media or an effusion does not exclude congenital cholesteatoma.
- Congenital cholesteatoma is usually located at the anterosuperior quadrant of the middle ear. However, it may be located at the posterosuperior quadrant or other locations.

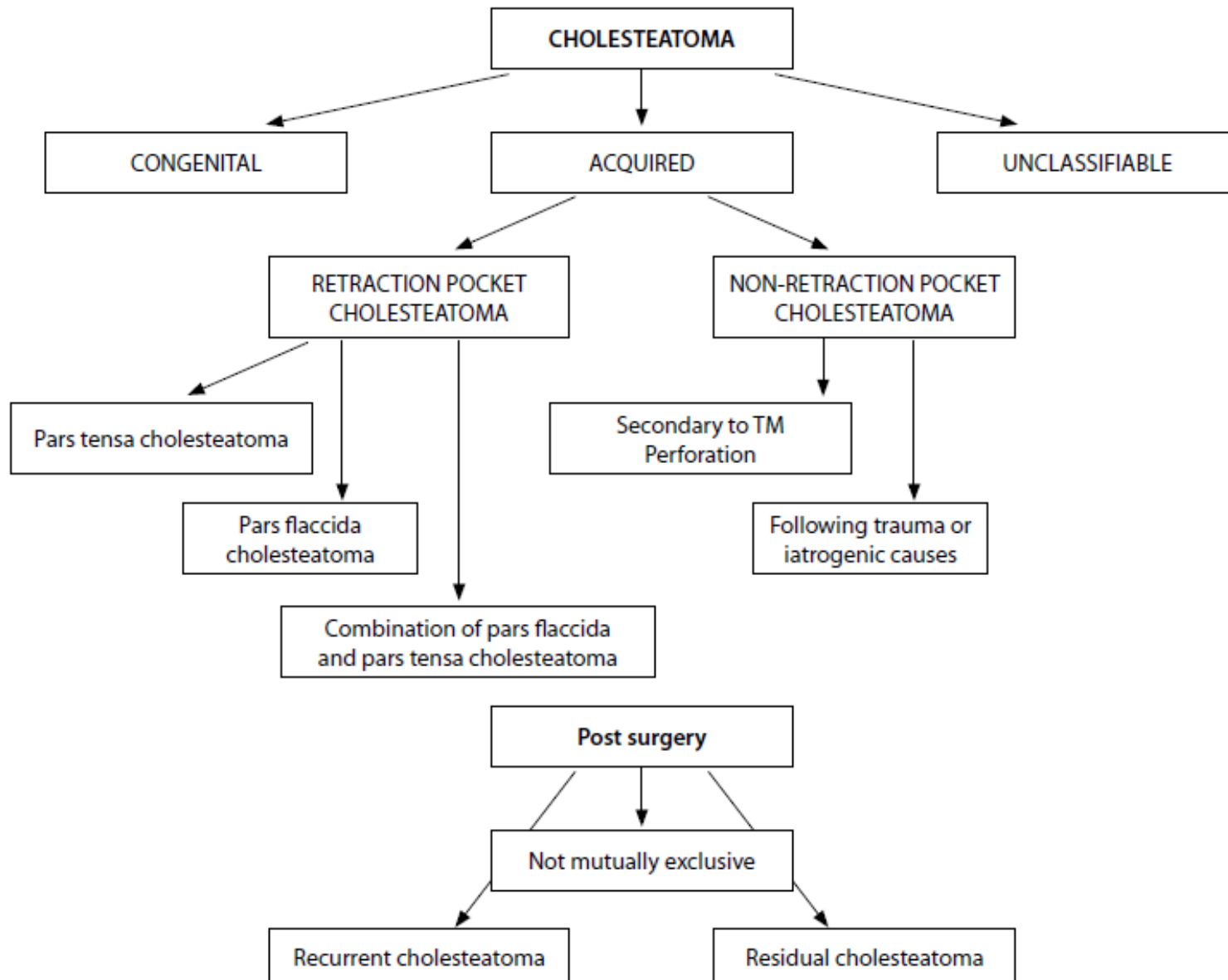
Acquired cholesteatoma

- 1. retraction pocket cholesteatoma
 - a) pars flaccida (attic cholesteatoma)
 - b) pars tensa cholesteatoma
 - c) combination of pars flaccida and pars tensa cholesteatoma
- 2. non-retraction pocket cholesteatoma
 - a) cholesteatoma secondary to tympanic perforation (the so-called secondary acquired cholesteatoma)
 - b) cholesteatoma following trauma and/or otologic procedures

Cholesteatoma recidivism

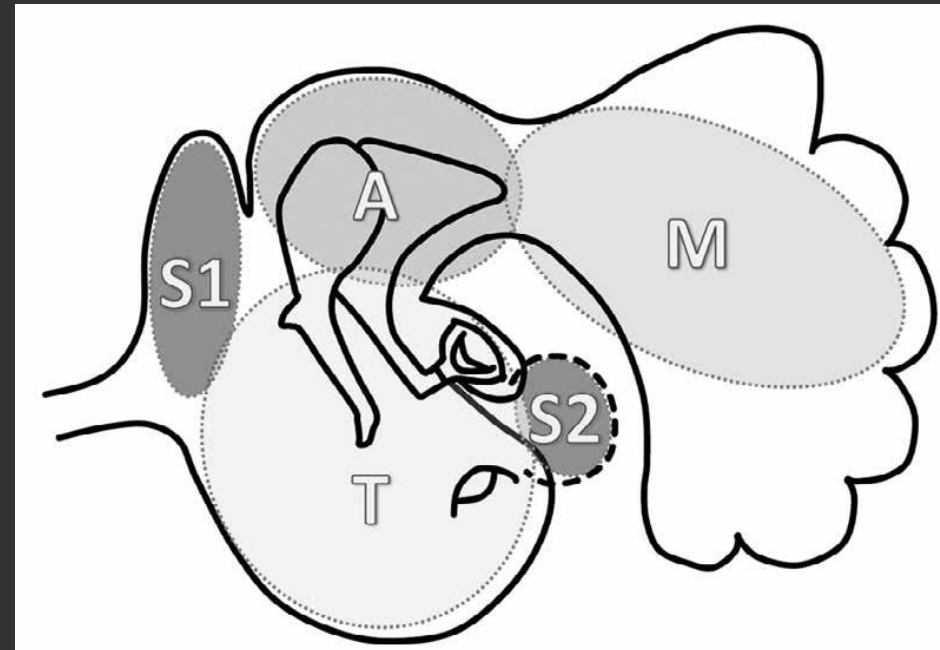
- Residual cholesteatoma
 - Residual cholesteatoma results from the incomplete surgical removal of the cholesteatoma matrix
- Recurrent cholesteatoma
 - results from the reformation of the retraction pocket after a complete previous surgical cholesteatoma removal

Post-surgical cholesteatoma may be residual or recurrent, although these are not mutually exclusive.



Divisions of the middle ear space (STAM system)

- Difficult access sites (S)
 - S1, the supratubal recess (also called the anterior epitympanum or protympanum)
 - S2, the sinus tympani
- Tympanic cavity (T)
- Attic (A)
- Mastoid (M)



Cholesteatoma staging

- **Stage I:** Cholesteatoma localized in the primary site
- **Stage II:** Cholesteatoma involving two or more sites
- **Stage III:** Cholesteatoma with extracranial complications or pathologic conditions including
- **Stage IV:** Cholesteatoma with intracranial complications including

Stage I: Cholesteatoma localized in the primary site

- Pars flaccida cholesteatoma (attic cholest
- Pars tensa cholesteatoma
- Cholesteatoma secondary to a tensa perforation
- Congenital cholesteatoma

Stage II: Cholesteatoma involving two or more sites

- Pars flaccida cholesteatoma (attic cholest
- Pars tensa cholesteatoma
- Cholesteatoma secondary to a tensa perforation
- Congenital cholesteatoma

Stage III: Cholesteatoma with extracranial complications

- Facial palsy
- Labyrinthine fistula
- Labyrinthitis
- Postauricular abscess or fistula
- Zygomatic abscess
- Neck abscess
- Canal wall destruction: more than half the length of the bony ear canal
- Destruction of the tegmen

Stage IV: Cholesteatoma with intracranial complications

- Purulent meningitis
- Epidural abscess
- Subdural abscess
- Brain abscess
- Sinus thrombosis
- Brain herniation into the mastoid cavity

CT

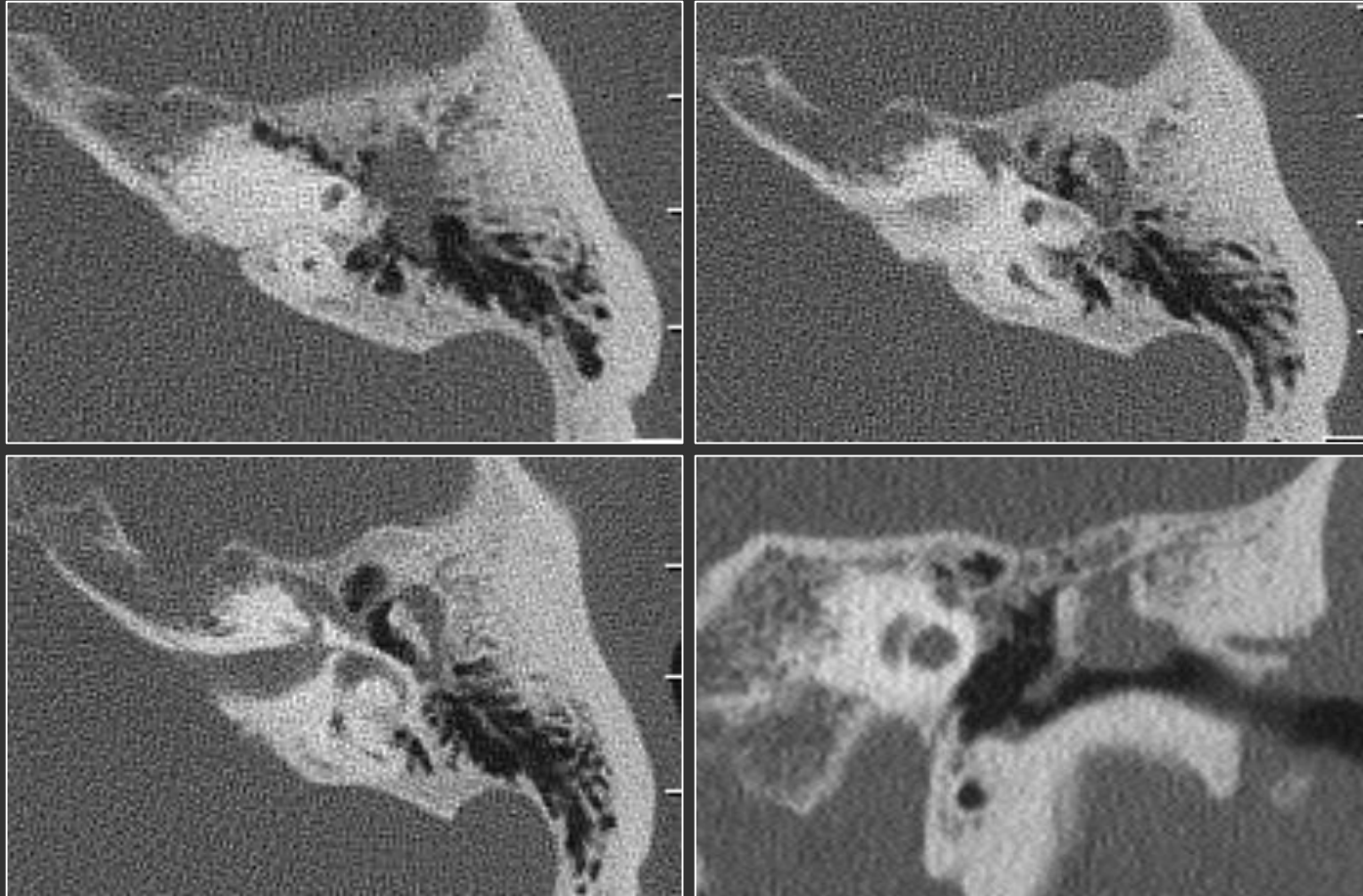
- Gold standard to image middle ear pathology
- High space resolution (Temporal bone anatomy before surgery)
- High sensitivity imaging in normal middle ear cleft
- Low specificity in case of full middle ear cleft



HRCT

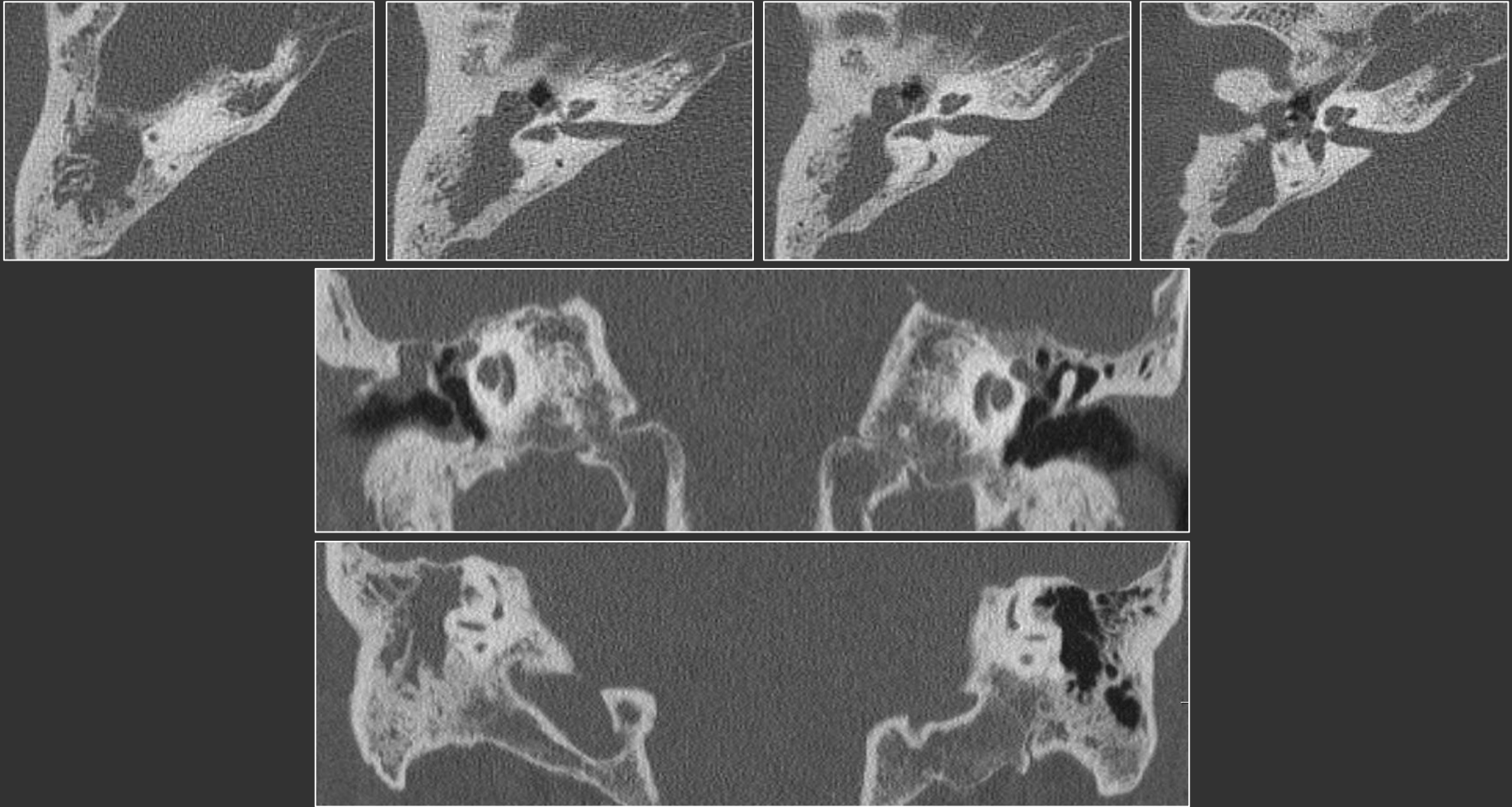
- Axial and coronal plane with additional reconstruction
- Colimation (0,5 – 0,6mm), slice thickness \leq 1mm
- Bony algorithm
- Native imaging (+C in case of vascular middle ear lesion)

Stage I



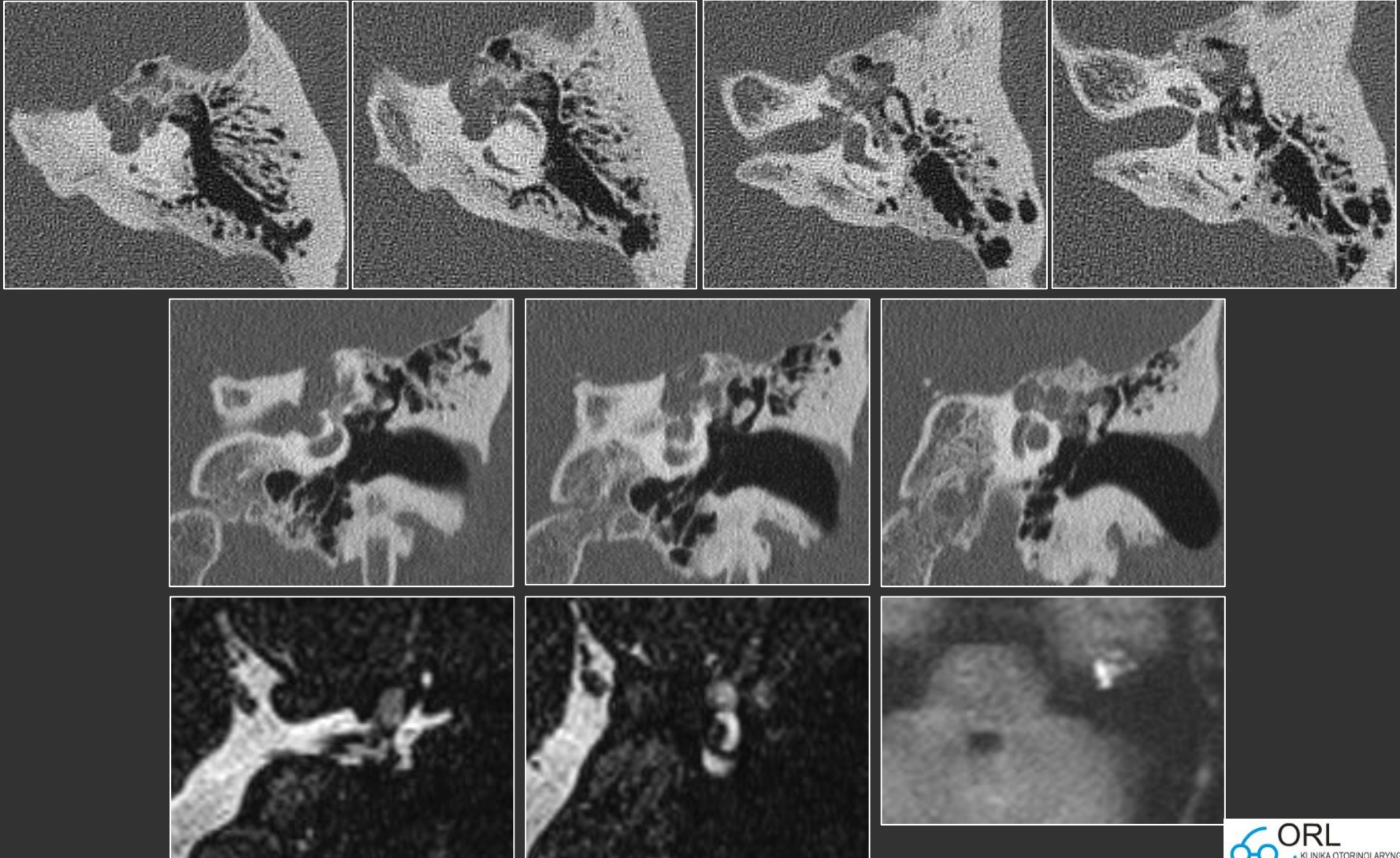
Cholesteatoma localised to the single primary site

Stage II



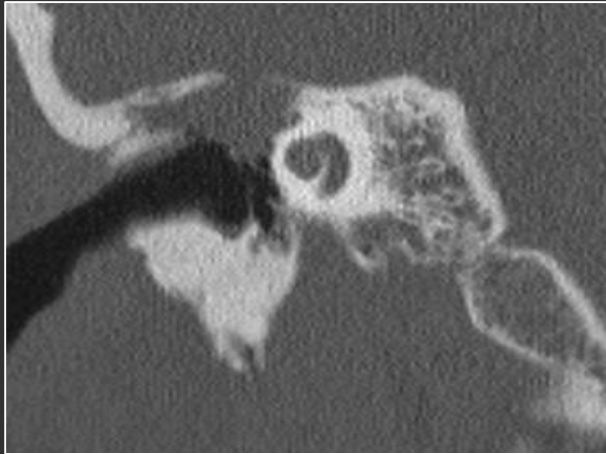
Cholesteatoma occupying 2 or more sites

Stage III



Cholesteatoma with extracranial complications

Stage IV



Cholesteatoma with intracranial complications

MR protocol

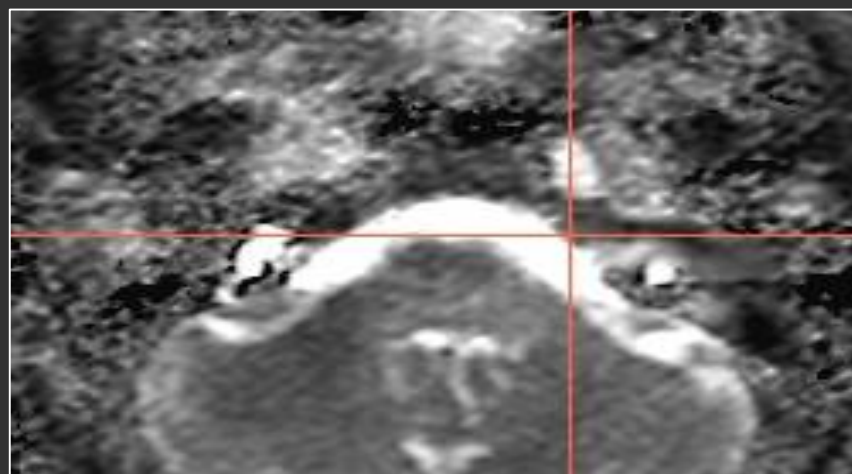
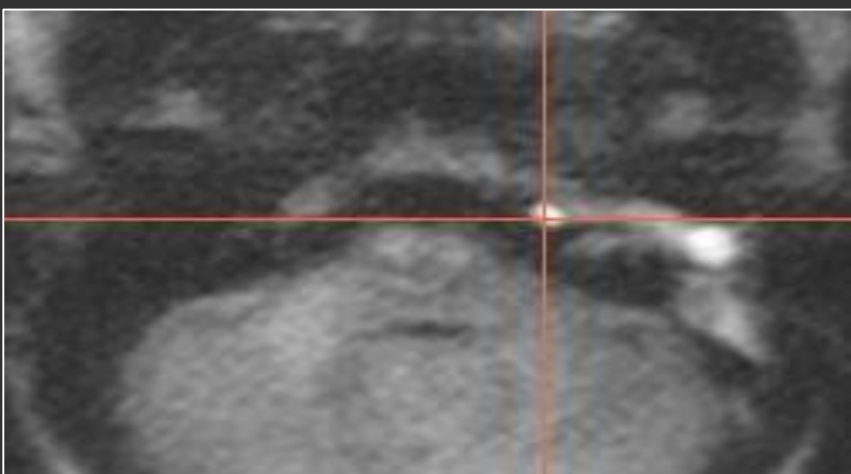
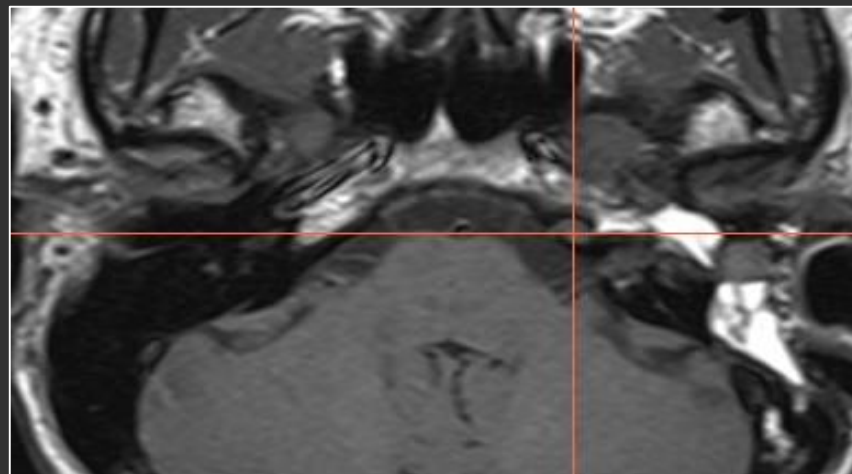
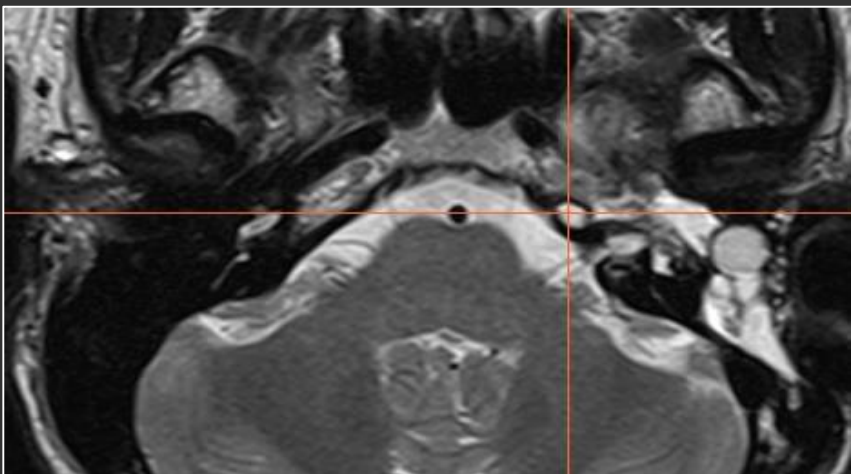
1,5 T Magnetom Avanto

Sequence	Thickness (mm)	TR (ms)	TE (ms)	FOV (mm)	Matrix	b factor
TRA tse T2	3	3850	108	230	384x216	
TRA se T1	3	490	14	230	320x168	
TRA 3D tse T2 (spc T2)	0,6	1200	258	200	324x320	
TRA HASTE DWI	3	2000	105	220	190x144	0,1000 s/mm ²
COR tse T1 FS	3	500	12	220	320x180	
TRA se T1 (Gd)	3	490	14	230	320x168	
COR tse T1 FS (Gd)	3	500	12	220	320x180	
3D T1 mp-range (Gd)	1	1900	3,37	256	256x192	

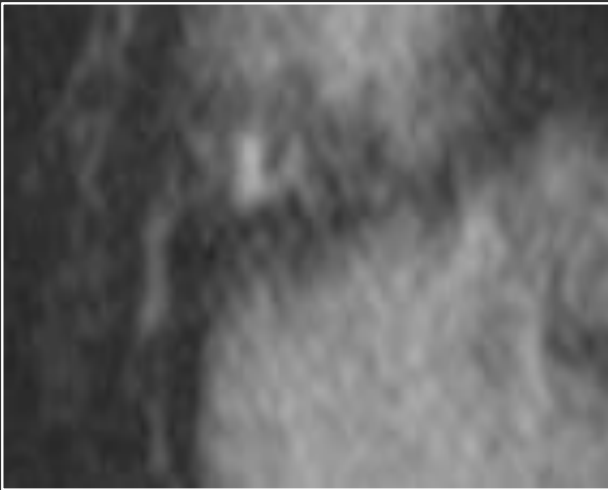
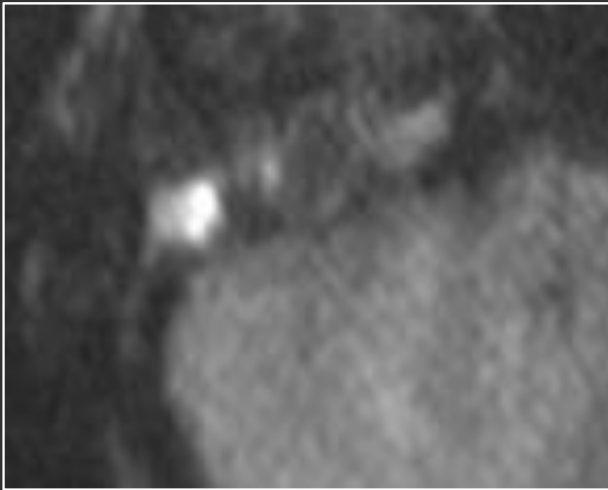
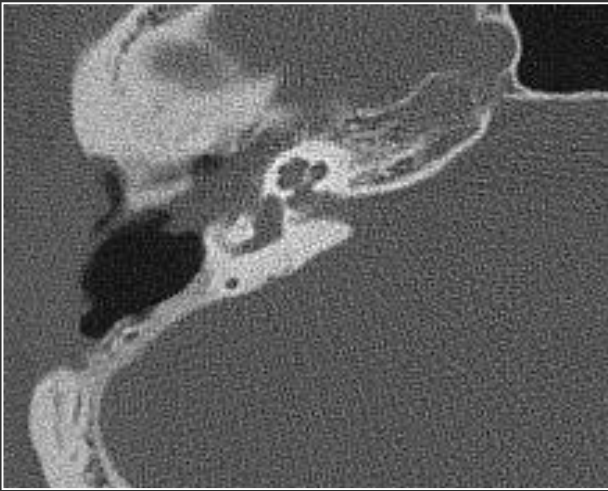
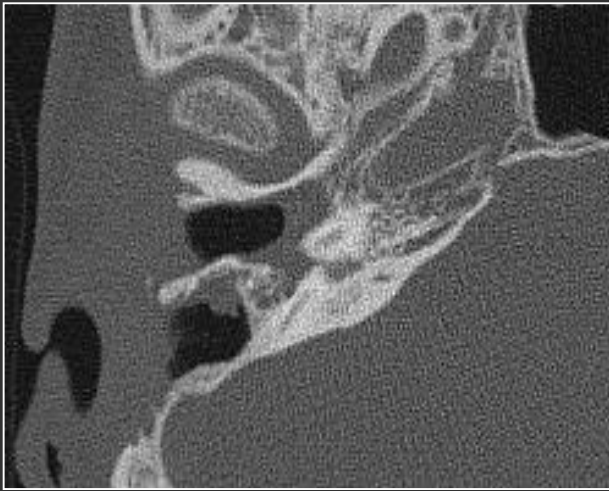
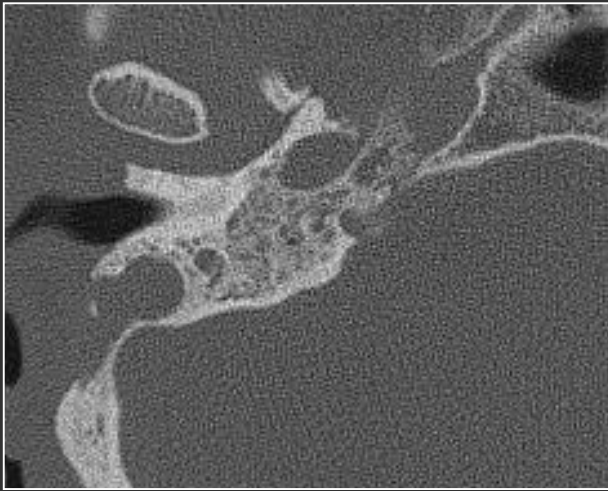
MR

- $\downarrow T1 Vo, \uparrow T2 Vo$
- CH after +C ring enhancement in periphery (perimatrix) / no enhancement
- CH equal to granulation tissue in T1 Vo and T2 Vo
- granulation tissue after +C significant enhancement

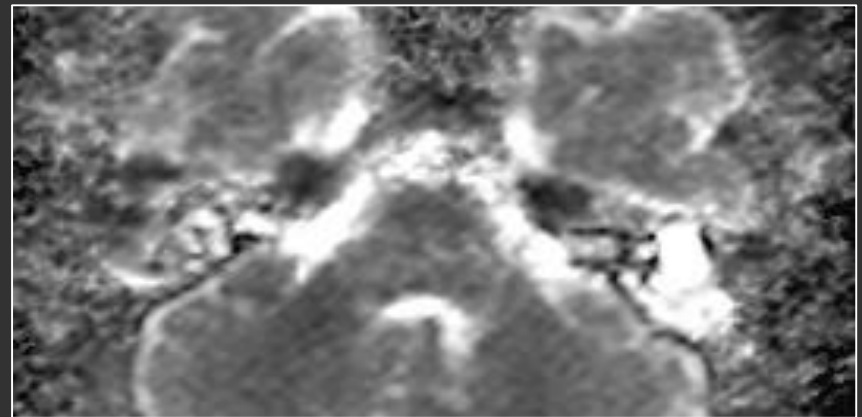
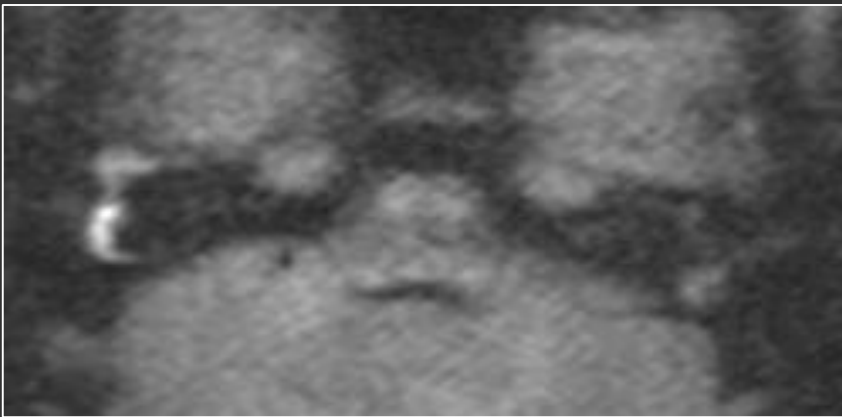
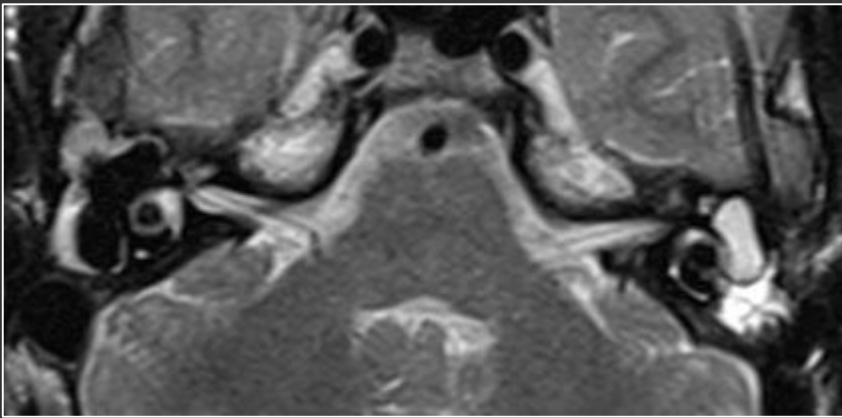
- $\uparrow DWI$ – to differentiate postoperative changes from residual/recurrent cholesteatoma



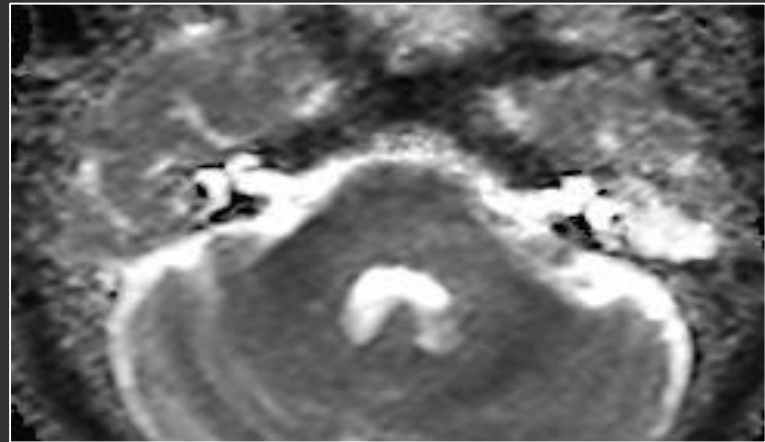
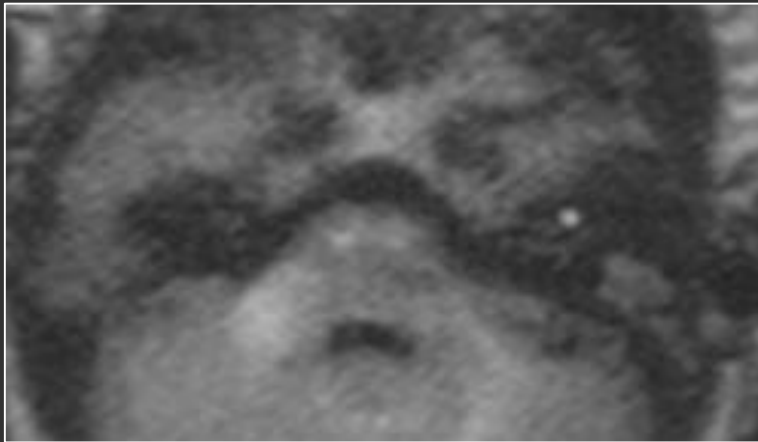
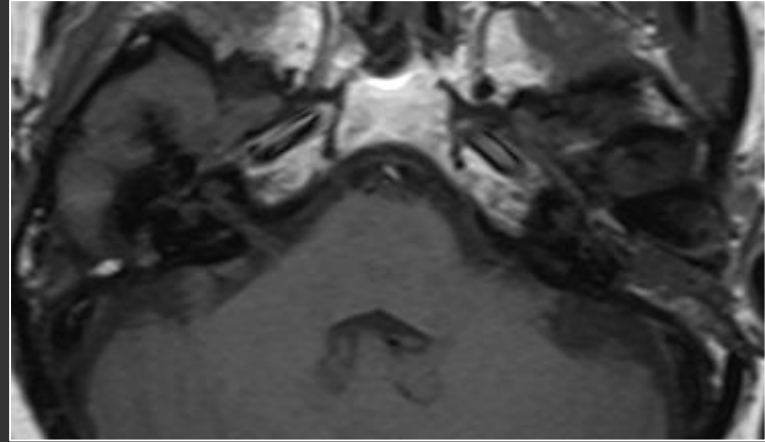
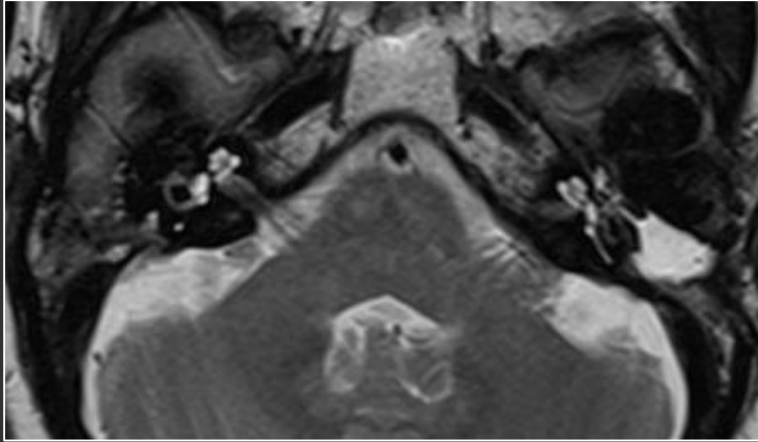
Subtotal petrosectomy



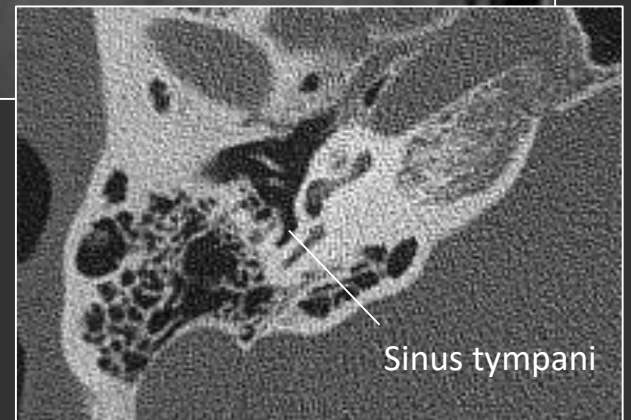
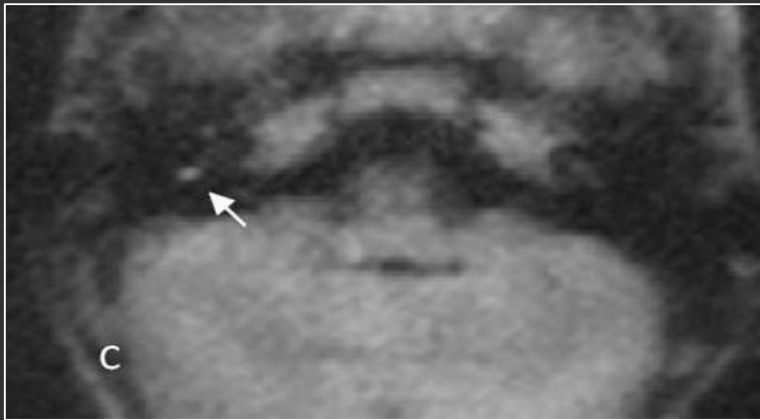
CWU tympanomastoidectomy



Modified radical surgery

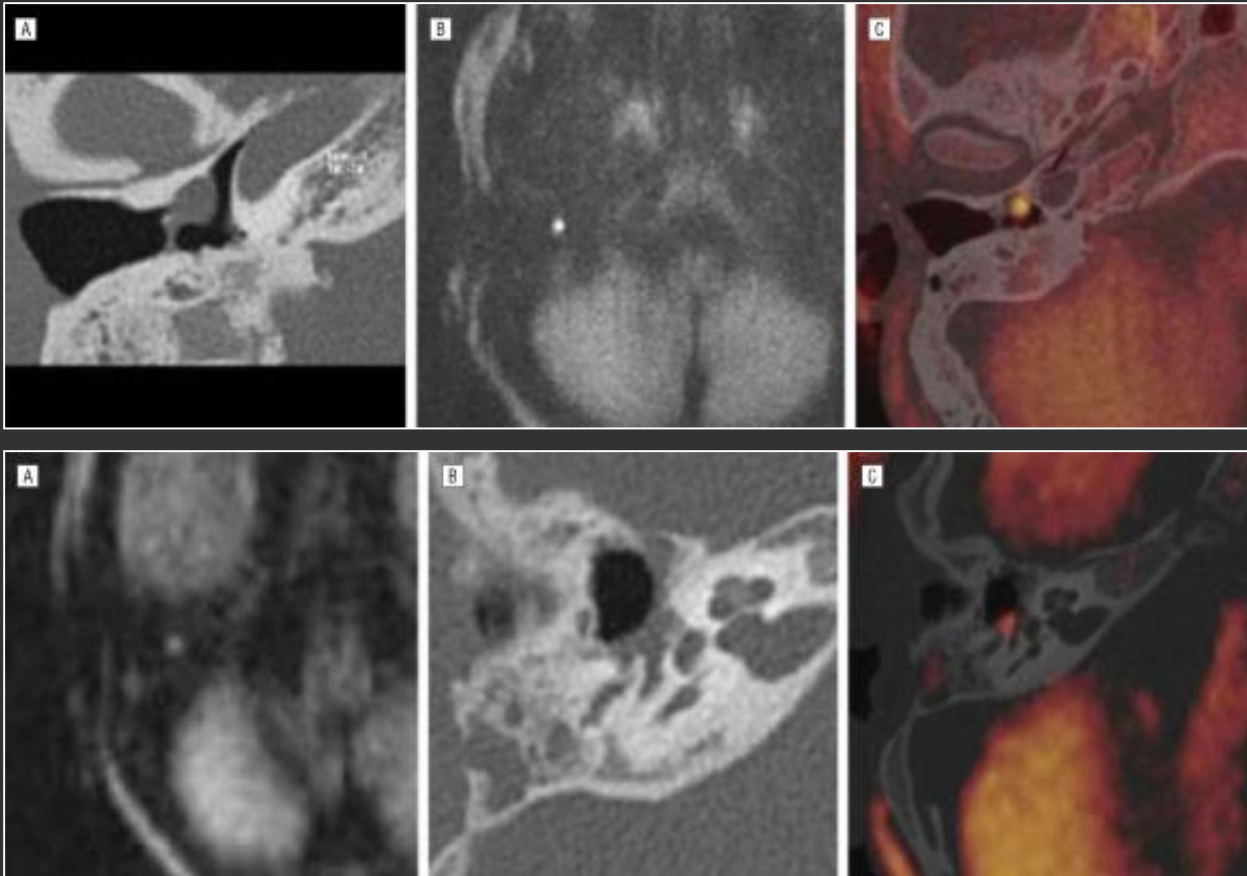


Cholesteatoma pearl



Cholesteatoma pearl in sinus tympani

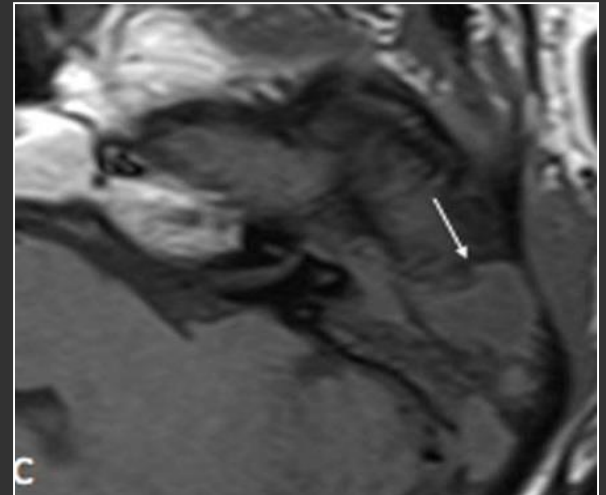
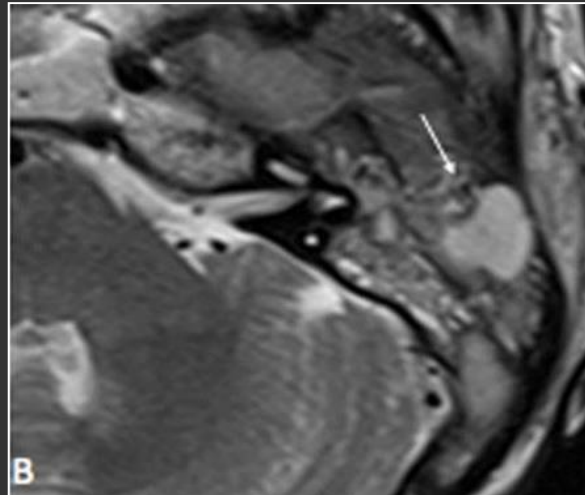
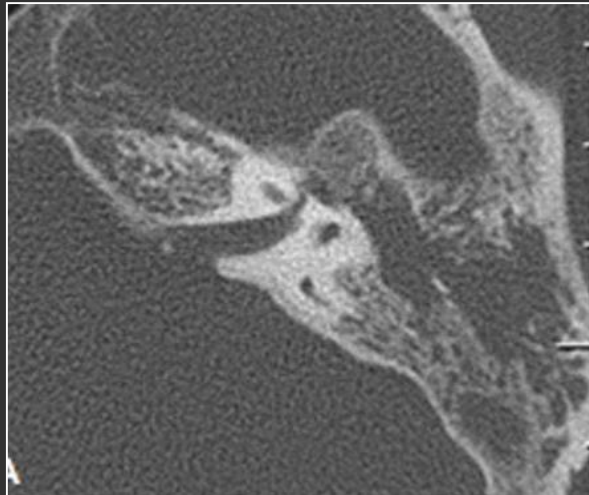
Localization (fusion of CT and MR)



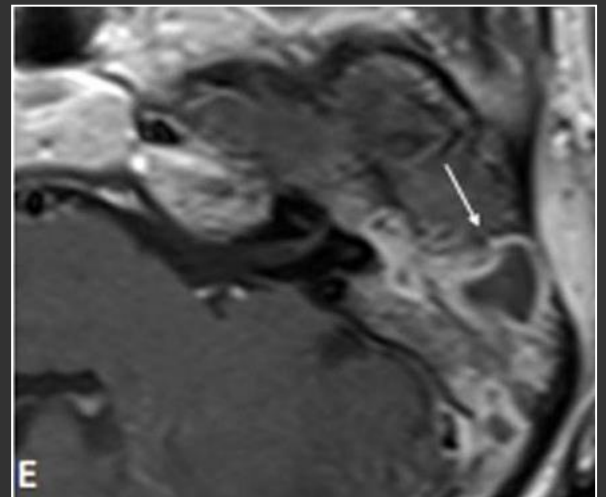
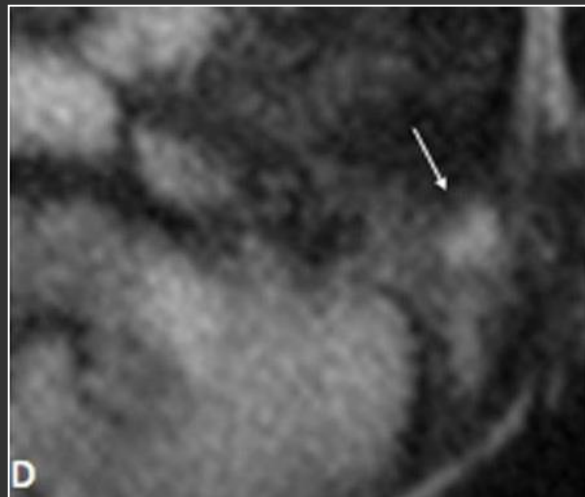
Plouin-Gaudon I. et al. „Fusion of MRIs and CT scans for surgical treatment of cholesteatoma of the middle ear in children, Arch Otolaryngol Head Neck Surg., 2010 Sep;136(9):878-83. doi: 10.1001/archoto.2010.151.

Differential diagnosis in MR

Pathology	T1 Vo	T2 Vo	+C T1 Vo	DWI	ADC
Cholesteatoma	↓	↑	- (Ring)	↑	↓
Cholesterol granuloma	↑	↑	-	-	-
Glomus TU („salt and spice“)	Slightly hypoSi	Slightly hyperSi	↑↑↑	-	-
Schwannoma	Slightly hypoSi	Slightly hyperSi	↑↑	-	-
Meningeoma	Slightly hypoSi	Slightly hyperSi	↑↑	-	-
Fresh granulation tissue	↓	↑	↑↑	↓	↑
Old scars	↓	↑	Late	↓	↑
Absces	↓	↑	Ring	↑	↓
Liquid	↓	↑	-	↓	↑



Abscess



Author	DWI	P	Size (mm)	Sensit. (%)	Specific. (%)	PPH (%)	NPH (%)
De Foer et al. (2008)	nEPI-DWI	19	2	90	100	100	96
Dhepnorrarat et al. (2009)	nEPI-DWI	22	3	100	100	100	100
Khemani et al. (2011)	nEPI-DWI	38	3	82	90	96	64
Profant, Sláviková (2012)	nEPI-DWI	42	3	97	62	91	83

Indications for imaging in cholesteatoma (our experience)

Diagnosis	CT	MR (HASTE DWI)
Chronic otitis with CH (primary dg, preoperative evaluation)	+	- (complementary in complicated cases)
Chronic otitis with CH (revision surgery to improve hearing)	-	-
Chronic otitis with CH (revision surgery „second look“)	-	+ (negative MR, no revision)
Chronic otitis with CH (follow up)	-	+

Conclusions

- New classification will not change biological behaviour of cholesteatoma
- Nowadays there is no method of choice to manage cholesteatoma to be respected by all otosurgeons
- Rerurrence rate is varying from 0% to 30%
- Obliteration of mastoid cavity with bone dust and separation from the tympanic cleft is a hit of recent period
- ***Cholesteatom itself at the end is the master with the final word***