New EAONO Cholesteatoma Classification with imaging illustration

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

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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.
CHOLESTEATOMA

CONGENITAL

ACQUIRED

UNCLASSIFIABLE

RETRACTION POCKET CHOLESTEATOMA

Pars tensa cholesteatoma

Pars flaccida cholesteatoma

Combination of pars flaccida and pars tensa cholesteatoma

NON-RETRACTION POCKET CHOLESTEATOMA

Secondary to TM Perforation

Following trauma or iatrogenic causes

Post surgery

Not mutually exclusive

Recurrent cholesteatoma

Residual cholesteatoma
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 cholesteatoma)
- Pars tensa cholesteatoma
- Cholesteatoma secondary to a tensa perforation
- Congenital cholesteatoma
Stage II: Cholesteatoma involving two or more sites

- Pars flaccida cholesteatoma (attic cholesteatoma)
- 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
- 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 ≤ 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

<table>
<thead>
<tr>
<th>Sequence</th>
<th>Thickness (mm)</th>
<th>TR (ms)</th>
<th>TE (ms)</th>
<th>FOV (mm)</th>
<th>Matrix</th>
<th>b factor</th>
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<tr>
<td>TRA tse T2</td>
<td>3</td>
<td>3850</td>
<td>108</td>
<td>230</td>
<td>384x216</td>
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<td>TRA se T1</td>
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<td>230</td>
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<td>TRA 3D tse T2 (spc T2)</td>
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<td>TRA HASTE DWI</td>
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<td>2000</td>
<td>105</td>
<td>220</td>
<td>190x144</td>
<td>0,1000 s/mm²</td>
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<tr>
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<td>500</td>
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<td>220</td>
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<tr>
<td>TRA se T1 (Gd)</td>
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<td>490</td>
<td>14</td>
<td>230</td>
<td>320x168</td>
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<tr>
<td>COR tse T1 FS (Gd)</td>
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<td>500</td>
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<td>220</td>
<td>320x180</td>
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<tr>
<td>3D T1 mp-rage (Gd)</td>
<td>1</td>
<td>1900</td>
<td>3,37</td>
<td>256</td>
<td>256x192</td>
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MR

- ↓T1 Vo, ↑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

- ↑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)

### Differential diagnosis in MR

<table>
<thead>
<tr>
<th>Pathology</th>
<th>T1 Vo</th>
<th>T2 Vo</th>
<th>+C T1 Vo</th>
<th>DWI</th>
<th>ADC</th>
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<tbody>
<tr>
<td>Cholesteatoma</td>
<td>↓</td>
<td>↑</td>
<td>-</td>
<td>↑</td>
<td>↓</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(Ring)</td>
<td></td>
<td></td>
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<tr>
<td>Cholesterol granuloma</td>
<td>↑</td>
<td>↑</td>
<td>-</td>
<td>-</td>
<td>-</td>
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<tr>
<td>Glomus TU</td>
<td>Slightly hypoSi</td>
<td>Slightly hyperSi</td>
<td>↑↑↑</td>
<td>-</td>
<td>-</td>
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<tr>
<td>(„salt and spice“)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Schwannoma</td>
<td>Slightly hypoSi</td>
<td>Slightly hyperSi</td>
<td>↑↑</td>
<td>-</td>
<td>-</td>
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<tr>
<td>Meningeoma</td>
<td>Slightly hypoSi</td>
<td>Slightly hyperSi</td>
<td>↑↑</td>
<td>-</td>
<td>-</td>
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<tr>
<td>Fresh granulation tissue</td>
<td>↓</td>
<td>↑</td>
<td>↑↑</td>
<td>↓</td>
<td>↑</td>
</tr>
<tr>
<td>Old scars</td>
<td>↓</td>
<td>↑</td>
<td></td>
<td>Late</td>
<td>↓</td>
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<tr>
<td>Absces</td>
<td>↓</td>
<td>↑</td>
<td></td>
<td>Ring</td>
<td>↑</td>
</tr>
<tr>
<td>Liquid</td>
<td>↓</td>
<td>↑</td>
<td></td>
<td>-</td>
<td>↓</td>
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Abscess
<table>
<thead>
<tr>
<th>Author</th>
<th>DWI Method</th>
<th>P</th>
<th>Size (mm)</th>
<th>Sensit. (%)</th>
<th>Specific. (%)</th>
<th>PPH (%)</th>
<th>NPH (%)</th>
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<tbody>
<tr>
<td>De Foer et al. (2008)</td>
<td>nEPI-DWI</td>
<td>19</td>
<td>2</td>
<td>90</td>
<td>100</td>
<td>100</td>
<td>96</td>
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<tr>
<td>Dhepnorrarat et al. (2009)</td>
<td>nEPI-DWI</td>
<td>22</td>
<td>3</td>
<td>100</td>
<td>100</td>
<td>100</td>
<td>100</td>
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<tr>
<td>Khemani et al. (2011)</td>
<td>nEPI-DWI</td>
<td>38</td>
<td>3</td>
<td>82</td>
<td>90</td>
<td>96</td>
<td>64</td>
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<tr>
<td>Profant, Sláviková (2012)</td>
<td>nEPI-DWI</td>
<td>42</td>
<td>3</td>
<td>97</td>
<td>62</td>
<td>91</td>
<td>83</td>
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</table>
Indications for imaging in cholesteatoma (our experience)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>CT</th>
<th>MR (HASTE DWI)</th>
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</thead>
<tbody>
<tr>
<td>Chronic otitis with CH</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>(primary dg, preoperative evaluation)</td>
<td></td>
<td>(complementary in complicated cases)</td>
</tr>
<tr>
<td>Chronic otitis with CH</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>(revision surgery to improve hearing)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chronic otitis with CH</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>(revision surgery „second look“)</td>
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<td>(negative MR, no revision)</td>
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<tr>
<td>Chronic otitis with CH</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>(follow up)</td>
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</table>
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
• Recurrence 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*