Classification of Middle Ear Cholesteatoma

I. Acquired cholesteatoma
1) Retraction pocket cholesteatoma *(so-called primary acquired cholesteatoma)*
   - a) Pars flaccida cholesteatoma (Attic cholesteatoma)
   - b) Pars tensa cholesteatoma
   - c) Combination of pars flaccida and pars tensa cholesteatoma
2) Non retraction pocket cholesteatoma
   - a) Cholesteatoma secondary to a chronic tensa perforation *(so-called secondary acquired cholesteatoma)*
   - b) Transplanted cholesteatoma following trauma or otologic procedures

II. Congenital cholesteatoma
III. Unclassifiable cholesteatoma

Staging of Middle Ear Cholesteatoma

1. Divisions of the middle ear space (PTAM system)

   Schematic drawing of divisions of the tympanomastoid space. The tympanomastoid space is divided into four sections: the protympanum (P), the tympanic cavity (T), the attic (A) and the mastoid (M) in order to represent the extent of cholesteatoma.

2. The JOS staging system applies to 4 types of middle ear cholesteatoma *(Pars flaccida cholesteatoma, pars tensa cholesteatoma, congenital cholesteatoma and cholesteatoma secondary to a tensa perforation)*

   - **Stage I**
     - Cholesteatoma localized in the primary site *
     - *The site of cholesteatoma origin, i.e. the attic (A) for a pars flaccida cholesteatoma; the tympanic cavity (T) for pars tensa cholesteatoma, congenital cholesteatoma and cholesteatoma secondary to a tensa perforation.*
     - **Stage Ia**
       - Cholesteatoma involving two or more sites
     - **Stage Ib**
       - Cholesteatoma involving two or more sites
   - **Stage III**
     - Cholesteatoma with extracranial complications and/or intratemporal pathologic conditions
     - Facial palsy (FP), labyrinthine fistula (LF): with conditions at risk for membranous labyrinth, labyrinthine disturbance (LD): scale out BC values for more than two speech frequencies (0.5, 1, and 2kHz), canal wall destruction (CW): more than half the length of the bony ear canal, adhesive otitis (AO): total adhesion of the pars tensa, petrous bone or skull base destruction (PB), neck abscess (NA).
   - **Stage IV**
     - Cholesteatoma with intracranial complications including
     - Purulent meningitis, epidural abscess, subdural abscess, brain abscess, sinus thrombosis, etc.

3. Sub-classification of stage I for respective cholesteatoma types

   - **Pars flaccida cholesteatoma**
     - **Stage Ia:** Cholesteatoma localized in the attic
     - **Stage Ib:** A retraction pocket with epithelial self-cleaning function
     - **Stage Ic:** A retraction pocket with persistent accumulation of keratin-debris
   - **Pars tensa cholesteatoma**
     - **Stage Ia:** Cholesteatoma localized in the tympanic cavity
     - **Stage Ib:** A retraction pocket with epithelial self-cleaning function
     - **Stage Ic:** A retraction pocket with persistent accumulation of keratin-debris
   - **Cholesteatoma secondary to a tensa perforation**
     - **Stage Ia:** Cholesteatoma localized in the tympanic cavity
     - **Stage Ib:** Epithelial invasion confined to the underside of the pars tensa
     - **Stage Ic:** Epithelial invasion extending to the tensor tympani tendon and the promontorial wall
   - **Congenital cholesteatoma**
     - **Stage Ia:** Cholesteatoma localized in the tympanic cavity
     - **Stage Ib:** Cholesteatoma confined to the anterior half of the tympanic cavity
     - **Stage Ic:** Cholesteatoma confined to the posterior half of the tympanic cavity
     - **Stage Id:** Cholesteatoma involving both of sides of the tympanic cavity