



## Labyrinthitis Ossificans and Cochlear Implant

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### Abbreviations

LO: *Labyrinthitis Ossificans*; CA: Cochlear Aqueduct; HRCT: High-Resolution Computed Tomography; MRI: Magnetic Resonance Imaging.

### Introduction

*Labyrinthitis Ossificans* (LO) is the 2nd most common pathology of the inner ear (most common is inner ear malformations). It is defined as pathological ossification of the membranous labyrinth in response to inflammatory or destructive process. LO usually develops as a sequela of previous infection which gets disseminated via various routes: hematogenic (via cochlear vasculature secondary to viral infections), meningogenic (via cochlear aqueduct from the subarachnoid space), tympanogenic (as a sequela of otitis media, i.e. the spread of infection via the round window), or posttraumatic. LO affects the enchondral layer of the osseous labyrinth but does not cross the endosteal layer. In pathological conditions like meningitis, otosclerosis, Paget's disease, trauma, ototoxicity, malignant infiltration, vascular obliteration of labyrinthine artery, or other infections, the new disorganized woven bone replaces the normal healthy bone and obliterates the spaces of the otic capsule [1].

Bacterial meningitis is the most common cause of LO. The most common organisms are streptococcus pneumoniae and Neisseria meningitidis. Streptococcus pneumoniae is the most common organism causing deafness post meningitis which occurs due to robust inflammatory response to the teichoic acid content in the bacterial cell wall. This highlights the importance of proper immunization at least 2 weeks prior to cochlear implant surgery. According to previous studies the pathophysiology of deafness after meningitis is due to the spread of infection from the meninges of the

posterior cranial fossa via the cochlear aqueduct (CA) to the scala tympani of the basal turn of the cochlea. The most common region to be involved is the scala tympani of the basal turn of the cochlea close to the round window due to 2 reasons: (i) increased concentration of the inflammatory mediators in this region where the CA drains into the scala tympani (ii) relatively decreased blood supply in this area.

Paparella and Sugiura have classified LO into three stages:

**Acute stage:** bacteria and leucocytes get accumulated in the perilymphatic spaces but spare the endolymphatic space.

**Fibrous stage:** increased angiogenesis with fibroblastic proliferation and osteoid deposition (undifferentiated mesenchymal cells) in the perilymphatic spaces.

**Ossification stage:** both perilymphatic and endolymphatic spaces are obliterated with unorganized woven bone.

LO causes sensorineural hearing loss due to progressive ossification of the cochlea. Early radiological evaluation, audiological follow-up and early surgical intervention is required in such cases. The best option for hearing rehabilitation is CI surgery at an early stage. In cases of bilateral LO, bilateral CI should be considered keeping in mind the progressive nature of the disease. Radiological diagnosis of LO can be made by high-resolution computed tomography (HRCT) or magnetic resonance imaging (MRI) of the inner ear. HRCT has high sensitivity in identifying the ossification stage of LO but fails to do the same in the early stages (acute and fibrous). This can be overcome by MRI which can identify the perilymphatic space fibrosis prior to ossification. Post-meningitis acute phase can be detected early in contrast enhanced T1-weighted images. T2W images will show loss of fluid signal in fibrosis or calcified tissue [2].

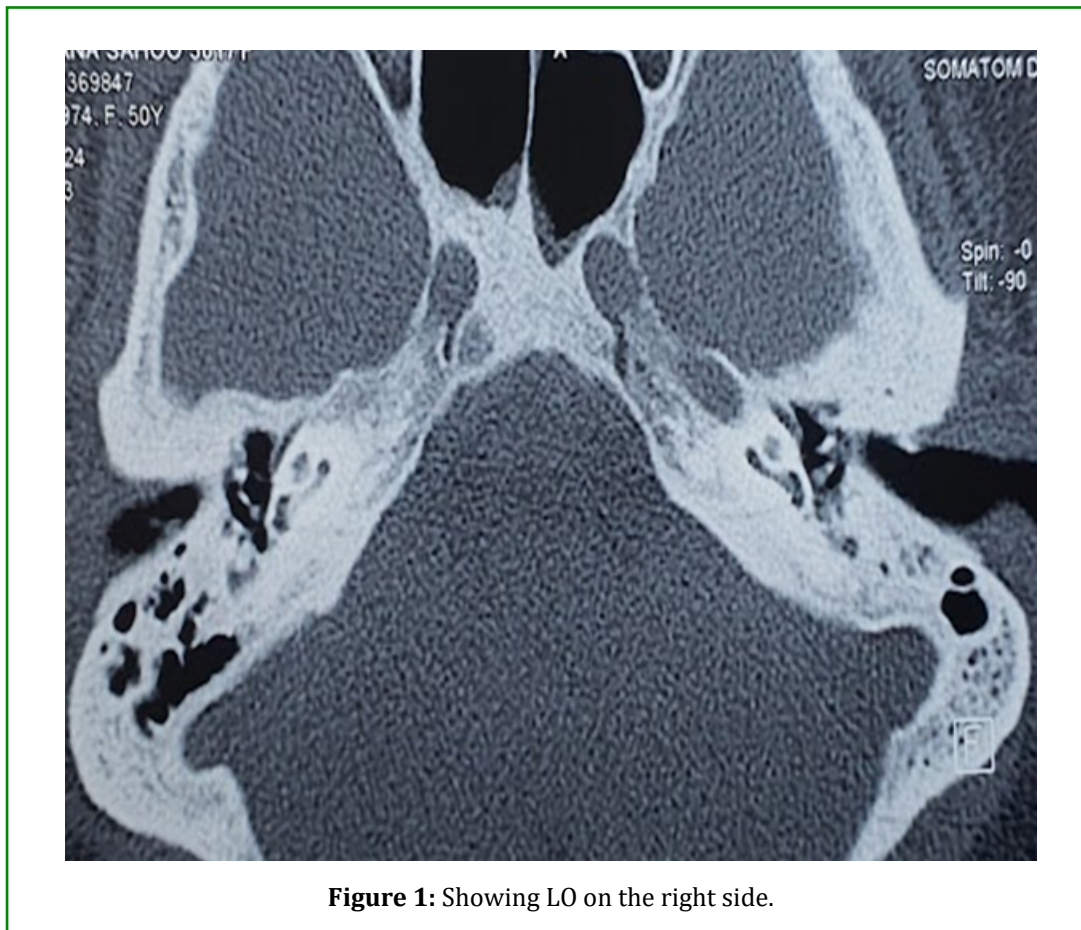
Based on the opacification on HRCT scans, Balkany and Dreisbach proposed a classification system as follows:

**C0:** Normal cochlea.  
**C1:** indistinctness of endosteum of the basal turn.  
**C2:** Definite narrowing of the basal turn.  
**C3:** Bony obliteration of at least a portion of the basal/middle

turn or the entire cochlea.  
 Based on the degree of ossification and surgical approach, Balkany, et al. classified LO into 4 stages:

Stage		Extent of ossification	Surgical approach
Stage I		Round window niche	Cochleostomy
Stage II		Inferior turn of basal turn (less than 180 degrees)	Partial drill out through round window niche
Stage III	IIIa	More than 180 degrees but less than 360 degrees of basal turn	Scala vestibuli insertion Complete basal turn drill out middle turn cochleostomy with compressed electrode array insertion
	IIIb	More than 360 degrees of basal turn	Total drill out of basal and middle turn with canal wall up or canal wall down/ auditory brainstem implant (ABI)

**Table 1:** Stages of *Labyrinthine ossification*.



**Figure 1:** Showing LO on the right side.

Specifically designed CI device with compressed or double or split arrays may be used in LO. The outcomes of CI in LO depend on the residual spiral ganglion cells, extent of insertion electrode arrays and duration of deafness before implantation.

### References

1. Paparella MM, Sugiura S (1967) The pathology of suppurative labyrinthitis. *Ann Otol Rhinol Laryngol* 76(3): 554-586.
2. Balkany T, Gantz BJ, Steenerson RL, Cohen NL (1996) Systematic approach to electrode insertion in the ossified cochlea. *Otolaryngology Head Neck Surg* 114(1): 4-11.