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# Post Meningitis Hearing Loss, Labyrinthitis Ossificans and Cochlear Implantation: A Case Report and Review of Literature

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**ABSTRACT**

Cochlear implant is a known neural prosthesis for children with severe to profound deafness. Meningitis causes fibrosis and ossification within the cochlea lumen (Labyrinthitis Ossificans). We present a 15-months boy with bilateral profound hearing loss, post Streptococcus meningitis. We highlighted challenges in managing this case and recommended ideal management.



## 1. INTRODUCTION

Hearing is an essential element of communication skill, particularly in young children who have not fully developed speech and language. Cochlear implant (CI) is a known neural prosthesis for children with severe to profound deafness. About 90% of bilateral profound hearing loss (HL) occurs due to post bacterial meningitis [1]. Meningitis causes fibrosis and ossification within the cochlea lumen. These may prevent complete CI electrode insertion and thus lead to poorer hearing rehabilitation outcomes. So, what are the red flags in bacterial meningitis that increase the chance of deafness? How early should we intervene after the infection? This paper evaluates the recent literatures and recommends the ideal guidelines for early diagnosis and management of post meningitis deafness, by understanding the pathology of Labyrinthine Ossificans (LO).

## 2. Case report

This is a 15-month, Malay, boy, who was born healthy at term. At 6 months, he was admitted for Streptococcus Pneumonia meningitis complicated with subdural effusion and status epilepticus. One month after discharge, parents noticed the child was not responding to sound. Also, his previous speech of babbling had regressed to cooing. Audiology assessment concluded bilateral profound HL. High Resolution Computer Tomography (HRCT) (at 10 months) showed complete obliteration of right cochlear by new bone and left cochlear in normal configuration (Figure 1). Bilateral lateral semicircular canal (SCC) showed partial obliteration (Figure 2). T2 weighted Magnetic Radiological Imaging (MRI) showed absent of right cochlea

configuration due to loss of fluid in ossified lumen and the left cochlear showed high intensity due to presence of perilymph fluid in lumen (Figure 3). Patient underwent left cochlear implantation at 11months post meningitis. Unexpectedly, intra-operatively, noted the left cochlear fully ossified- resistance on electrodes insertion. Then proceeded with the drill out procedure and inserted short length implant. Post op was uneventful. Upon switching on CI, auditory nerve response telemetry detected 4 active electrodes.

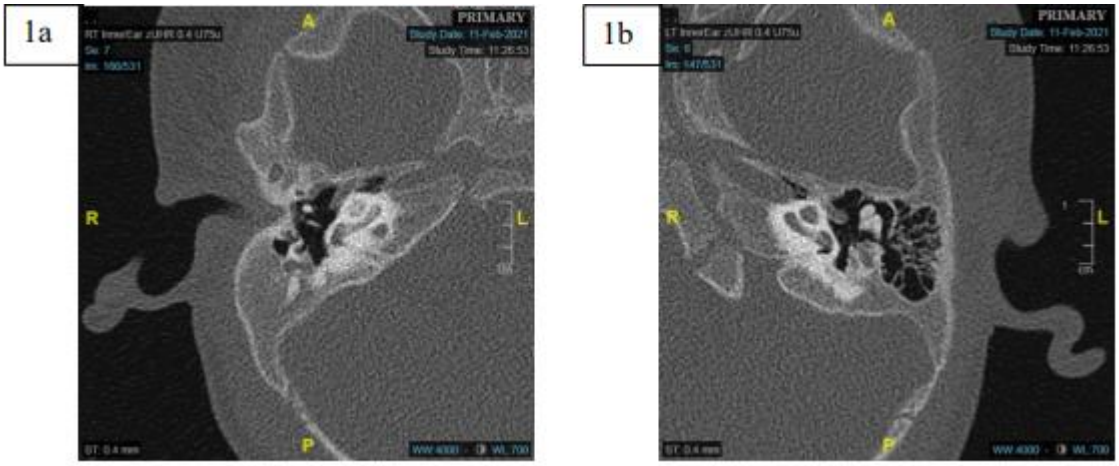


**Figure 1** shows HRCT Temporal (Axial, Bone Window)
(1a) HYPERDENSE Right cochlear likely calcification and final "Ossification''
(1b) Left cochlear normal configuration -appear to have patent lumen -no fluid loss

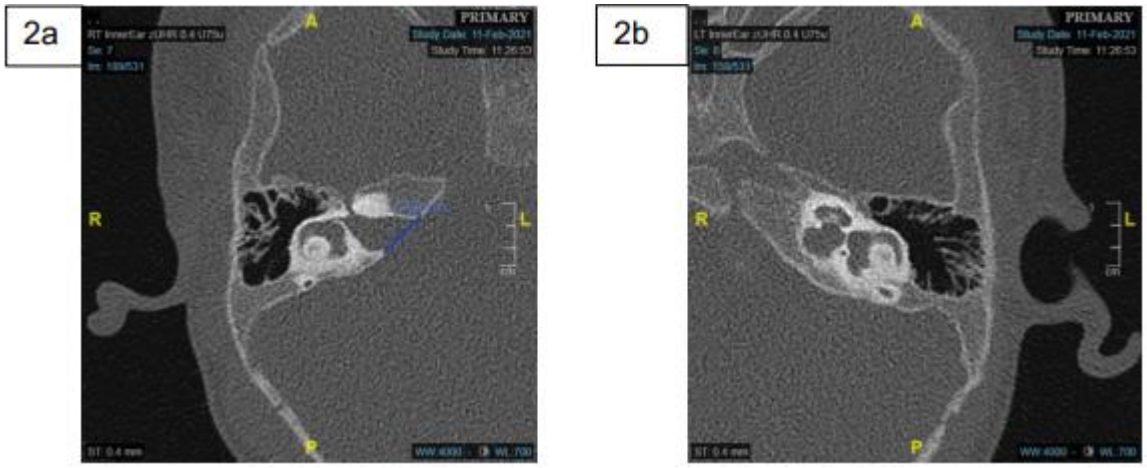


**Figure 2** shows HRCT Temporal (Axial, Bone Window)
(2a), (2b) Part of lateral SCC bilaterally appears partially hyperdense and lumen not visualised especially posterior part likely due to sclerosis

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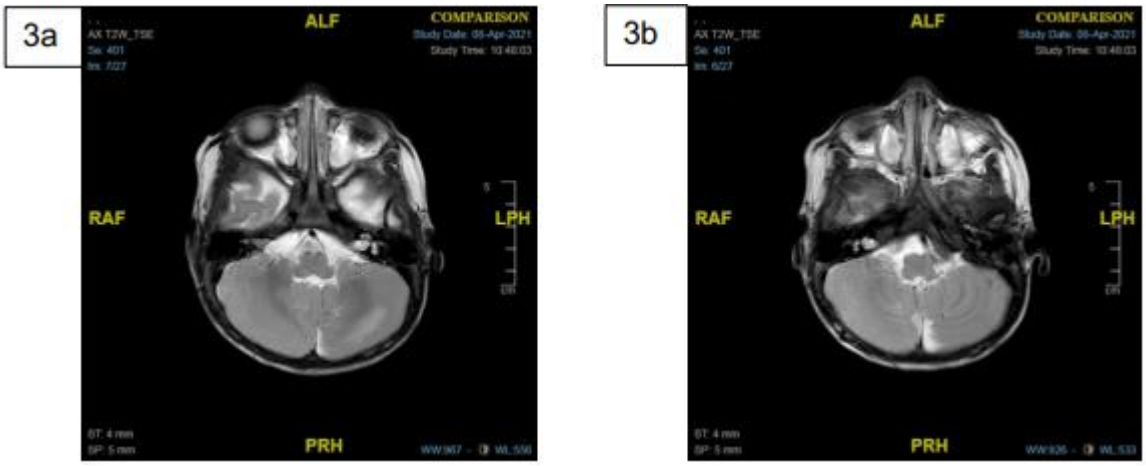


**Figure 3** shows images of MRI T2 weighted Brain and Internal Auditory Meatus
(3a) On left cochlear, there is uptake in scala tympani and vestibule indicating perilymph fluid still presents in cochlear
(3b) On right cochlear, there is a loss of fluid in the cochlea -indicated calcification

**3. Discussion**

CI is a well-established neural prosthesis for children with severe to profound deafness. LO is a final sequela of meningitis that produces reaction in the endosteum of the cochlea resulting in formation of new bone that fills the lumen of the cochlea and SCC. The LO has been seen in 80% of post meningitis cases [2]. Degree of ossification can range from minimal immature bone in the proximal end of the basal turn to complete ossification and obliteration of the cochlea lumen.

Factors that may predict the future deafness in post meningitis patients are male gender, streptococcus pneumoniae causative organism, nuchal rigidity, imaging evidence of raised intracranial pressure and low cerebral spinal fluid glucose levels [2]. There are several routes for the infection in the subarachnoid space to spread to the labyrinth. One is via the cochlear aqueduct to the scala tympani at basal turn of cochlea. Others are through the lateral end of internal auditory meatus via canaliculi of cochlear nerve or a direct dehiscence in congenital anomalies [3].

The understanding of the chronology of LO is crucial in surgical planning of CI. It determines a timeline at which intervention may alter the process of ossification. An animal study to determine the chronology of LO revealed that osteoneogenesis of cochlear lumen begins as early as 3 weeks and may precede over 12 months duration [3]. Histopathology studies of the temporal bone concluded a negative correlation of number of spiral ganglion cells to the degree of cochlear ossification, degree of bony occlusion and total years of deafness [4]. Some authors recommend allowing grace period of 2–4 weeks upon discharge to resolve any middle ear pathology of deafness [4]. We suggest hearing assessment to be done within 2 weeks upon discharge and follow up for 12 months post meningitis.

Bacterial meningitis commonly occurs in children, less than 2 years, who have immature immunity [5]. Also, good hearing is compulsory for good language development in children less than 3 years old. The Auditory Brainstem Response (ABR) is a mandatory objective hearing assessment. With any failed Otoacoustic Emission at discharge, an early audiology appointment must be made for ABR. Those children with ABR result of moderate to profound HL (≥30dBhl), must be referred immediately to CI center for complete hearing evaluation and an urgent MRI to detect any early changes of LO. Any evidence of fibrosis in MRI alongside progressive HL, decision for CI is mandatory.

The MRI is superior to the CT in diagnosing early stages of LO. In acute stage, the gadolinium enhanced T1 weighted MRI shows enhancement within cochlea due to inflammation. In fibrosis stage, the T2 weighted MRI shows hypointense cochlear lumen due to loss of fluid. In advanced stage, the whole cochlea configuration could be absent due to calcification of cochlea lumen, evidently a contraindication for CI. CT is also important in knowing surgical landmarks pre operatively like the jugular bulb position and the presence of infra-cochlear air cells. Some authors proposed the CT findings of a partial obliteration of the lateral SCC which is a valuable indicator for early ossification of the cochlea, although cochlea may seemingly appear patent in the scan. The ossification may start in the SCC which predicts the development of cochlear ossification [6].

The medical management like the corticosteroid has been recommended for post meningitis LO. It helps in reducing the inflammatory response within cochlear lumen [7]. The dexamethasone is an ideal choice because it has high penetration of blood brain barrier and has longer half-life. A systemic study advocated four-days course of dexamethasone in children (0.4 or 0.6 mg/kg/day) divided into four daily doses [7]. It should be administrated before or along the first dose of antibiotic therapy [7].

## 4. Conclusion

As we know it is time-critical to optimize success rate of CI. An early diagnosis, assessment and intervention is vital especially in children a less than 3 years old. Hence, we should treat HL as a neurodevelopmental emergency. A consistent meaningful hearing is required for developing a neural connection with auditory and speech centre in the brain. The joint effort of paediatrician, otologist, audiologist and speech therapist is important for the prevention of speech and language disability or handicap in a child.

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