



## Cochlear implantation in patients with ossified cochleas

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### ABSTRACT

**Purpose:** The timing of CI for postmeningitic deafness is controversial and differential outcomes have been reported. To review and share our surgical and auditory outcomes.

**Materials and methods:** 17 patients with ossified cochleas who received CI were enrolled. Clinical data including the cause of cochlear ossification, preoperative examination, onset of deafness, age at implantation, surgical findings, and relevant auditory outcomes was analysed.

**Results:** Cochlear ossification was observed in 53% of patients with HRCT, whereas the corresponding value for MRI was 59%. Patients in both stage I and II received complete insertion of the electrode array, however, stage III patients only received partial insertion. 1 patient in stage II received bilateral CI. Hearing tests showed increased average hearing threshold for stage III patients than those in stage I and II ( $P < 0.05$ ). CAP scores were much lower for stage III patients than those in stage I and II ( $P < 0.05$ ). Postlingual deafness patients showed higher SIR scores than prelingual deafness children ( $P < 0.05$ ).

**Conclusions:** HRCT and MRI have comparable value in predicting the occurrence of ossification in cochleas. We recommend fast surgical intervention in the patients with bilateral profound postmeningitic deafness. If possible, bilateral cochlear implantation is recommended.

## 1. Introduction

Cochlear ossification is common in patients who undergo cochlear implantation (CI), especially in postmeningitic deafness, 80% of whom have some degree of cochlear obstruction [1]. Other causes include otosclerosis, Usher syndrome, and Cogan syndrome. CI in patients with cochlear ossification presents many unique challenges [2,3], with the progress in the surgical techniques, however, it's no longer a contra-indication. Various surgical methods were reported in the literature, including partial implantation, double-array implantation, scala vestibuli implantation [2,3], and radical cochleostomy implantation [4]. However, the timing of CI for postmeningitic deafness is still controversial and differential outcomes of the postoperative performance have been reported [5].

The aim of our study was to share our surgical experiences and the rehabilitation outcomes in patients with various degrees of cochlear ossification.

## 2. Materials and methods

### 2.1. Patients

In the retrospective study, 1021 patients were treated with cochlear implantation between 2010 and 2017 at the Department of Otorhinolaryngology of Qilu Hospital of Shandong University. 17 out of these patients (10 males, 7 females) confirmed cochlear ossification during the operation were enrolled in the study.

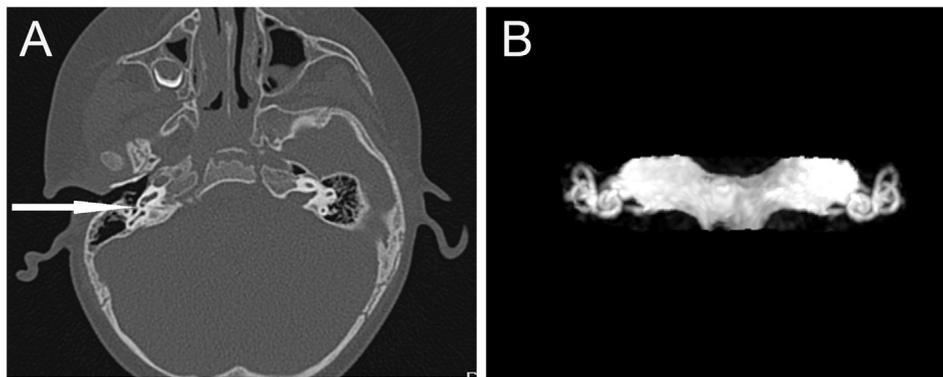
The study was approved by the Ethics Committee of Qilu Hospital of Shandong University, Jinan, China.

### 2.2. Preoperative examination

Electro-otoscopy and acoustic immittance test were carried out to exclude inflammation. Pure-tone audiometry, distortion product otoacoustic emissions (DPOAEs), auditory brainstem response (ABR) and auditory steady-state response (ASSR) tests were performed pre-operatively. Mean hearing threshold was 96.5 dB HL (range 90–110 dB

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**Fig. 1.** Imaging of P3.

(A) CT scan image with a white arrow pointing to ossification restricted in the round window niche, without any signs of ossification in the lumen. (B) Inner ear fluid MRI and 3D reconstruction image showing complete cochlear cavity.

HL). All DPOAEs were negative.

High-resolution computed tomography (HRCT), magnetic resonance imaging (MRI), inner ear fluid MRI and 3D reconstruction were performed preoperatively in all patients.

As described previously [6], we divided the ossified cochleas into 3 stages: stage I, ossification is limited to the round window area (Fig. 1); stage II, ossification involves the entire inferior segment of the basal turn up to 180° (Fig. 2); stage III, complete cochlear ossification involves more than two turns (Fig. 3).

### 2.3. Surgical methods

The transmastoid facial recess approach was applied to all the patients. The round window is located 2 mm below the inferior edge of the stapes footplate. For stage I, since the round window is the sole site of involvement, after grounding off the round window niche and picking away new bone around, an electrode can be fully inserted through round window.

For stage II, using the round window as a mark, drilling is continued for up to 8 mm at the basal turn or until the lumen of the basal turn is opened. A metal probe with a C-shaped head would be useful in opening the non-ossification zone upward and forward.

For stage III, open the basal turn of the cochlear using the method in stage II, followed by removal of the incus. Furthermore, drill out the lateral wall of the middle turn of the cochlear, clear the newly formed bone or soft tissue to make a 540° man-made bone tunnel around the modiolus. Continuous irrigation is essential to avoid thermal damage to residual spiral ganglia cells. Electrodes surrounding the modiolus were secured in the tunnel with small pieces of temporalis muscle fascia.

Types of cochlear implant were Nucleus 24 (Cochlear™, Sydney,

Australia), C40+ (MED-EL GmbH, Wien, Austria), PULSAR (MED-EL GmbH, Wien, Austria), SONATA (MED-EL GmbH, Wien, Austria), and HiRes 80KTM (Advanced Bionics, Los Angeles, USA).

### 2.4. Postoperative treatment and follow up

The patients were followed up for 6–48 months. The outcome was assessed using sound field audiometric test by the same audiologist with the background noise less than 30 dB SPL. The auditory receptive abilities and speech intelligibility of the patients were rated using Categorical Auditory Performance (CAP) and the Speech Intelligibility Rating (SIR) scales as previously reported [7]. The tests were carried out by professional staff every 3 months in the first year, every 6 months in the second year, and once annually until 4 years after implantation.

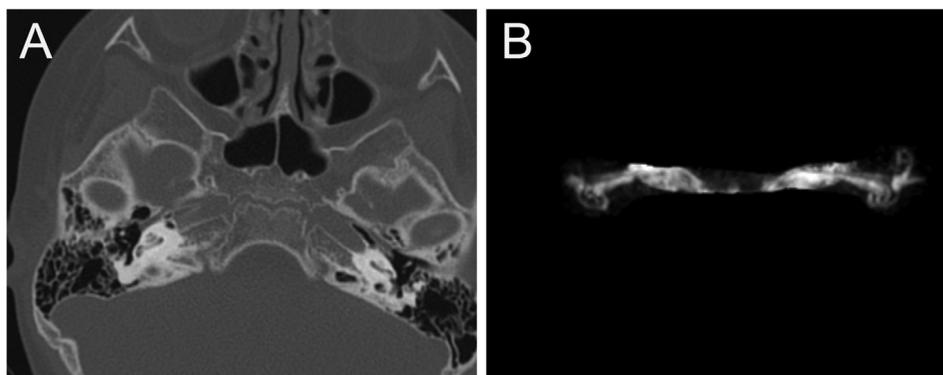
### 2.5. Statistical analysis

Statistical analysis of the data was conducted with SPSS18.0 (SPSS, Inc., Chicago, IL, USA). The CAP and SIR scores were compared using Mann-Whitney *U* test. A *P* value lower than 0.05 was regarded as statistically significant.

## 3. Results

The patients' mean age at implantation was 14.1 years, ranging from 0.8 to 50.5 years. All patients had suffered from bilateral profound hearing loss for more than 4 months, including 6 prelingually deafened children (Table 1). 15 out of the 17 deaf patients were due to bacterial meningitis, however, 2 patients were of unknown cause.

HRCT scan was normal in 8 patients and abnormal in 9 patients.



**Fig. 2.** Imaging of P12.

(A) CT scan, axial section of the petrous temporal bone, partial ossification of the basal turn of the cochlea. (B) Inner ear fluid MRI and 3D reconstruction, partial extinction of the T2 hyperintensity of the endolymph of both cochlea suggesting ossification and fibrosis.

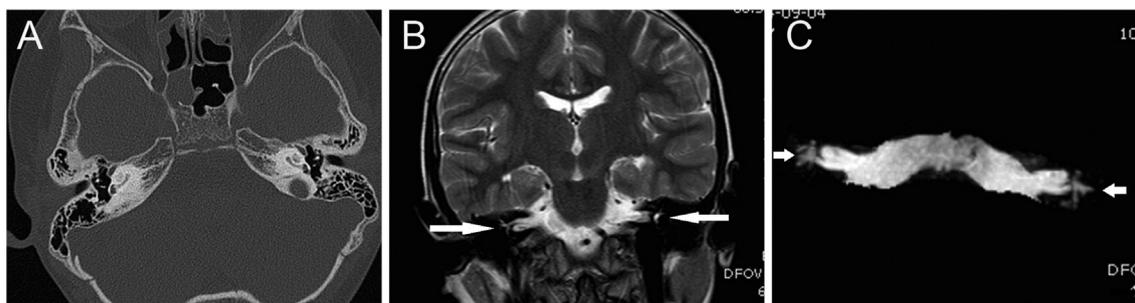


Fig. 3. Imaging of P16.

(A) CT scan, axial section of the petrous temporal bone, complete ossification of the entire cochlea. (B–C) White arrow show completely absent cochlea bilaterally on MRI.

Table 1

Characteristics of subjects with ossified cochleas and post-operative test results. P, Patient; M, male; F, female.

Subject	Sex	Age of onset (yrs)	Duration of deafness (yrs)	Causes of ossification	Degree of ossification	CAP	SIR
P1	M	0.3	0.5	Meningitis	Stage I	6	4
P2	M	0.4	0.6	Meningitis	Stage I	5	3
P3	F	1.4	0.6	Meningitis	Stage I	6	4
P4	M	30	5	Unknown	Stage I	8	5
P5	F	35.5	15	Meningitis	Stage I	8	5
P6	F	0.8	0.6	Meningitis	Stage I	7	4
P7	F	14.2	0.8	Unknown	Stage I	9	5
P8	M	0.7	0.6	Meningitis	Stage II	5	3
P9	M	4.6	0.9	Meningitis	Stage II	6	4
P10	F	11	1	Meningitis	Stage II	7	5
P11	M	13.3	0.7	Meningitis	Stage II	8	5
P12	F	11.7	0.3	Meningitis	Stage II	7	5
P13	F	25	21	Meningitis	Stage II	8	5
P14	M	15	1	Meningitis	Stage II	7	5
P15	M	10	1	Meningitis	Stage II	6	5
P16	M	8.5	0.5	Meningitis	Stage III	3	5
P17	M	7.3	0.7	Meningitis	Stage III	4	5

Among the 9 patients, whose CT evaluation corresponded to the intraoperative findings, partial ossification was found in 7 cases, and complete ossification was found in the 2 others. 10 showed attenuation of hyperintense signal on T2-weighted MRI for whom fibrosis or ossification were confirmed at surgery. In 3 cases with a normal CT, signs of ossification were seen on MRI, whereas ossification was seen on CT in 2 cases with a normal MRI.

Electrodes were fully inserted via scala tympani in 15 of the 17 patients. 7 patients (P1–P7) belonged to stage I, who had round window ossification. After removing the new bone around the round window niche, the patent cochlear lumen exist without further drilling procedures. 8 patients (P8–P15) had varying degrees of ossification in the basal turn and upper basal turn of the cochlear. They were drilled until the lumen of the basal turn was opened (Fig. 4A). 2 patients (P16–P17) had complete cochlear ossification belonging to stage III. Total drill-out technique was applied and a cochlear tunnel surrounding the modiolus was created (Fig. 4B). Both of the patients received Medel C40+ cochlear implantation, however, with the following up on the patients, only 4 electrodes worked in P16 and 7 electrodes worked in P17. Patient 13 received bilateral cochlear implantation.

All surgeries were successful and no surgical complications, including facial nerve paralysis, wound infection, or cerebrospinal fluid leak occurred in any case.

Sound field audiometric test were performed 6 months after surgery by the same audiologist. The average hearing threshold was 30 dB HL for patients both in stage I and stage II, however, it increased in stage III group, with an average of 40 dB HL for P16 and P17 ( $P < 0.05$ ).

CAP scores were comparable for patients in stage I and stage II

( $P > 0.05$ ), on the contrary, they were much lower for patients in stage III ( $P < 0.05$ ). SIR scores were comparable for prelingual deafness children in stage I and stage II ( $P > 0.05$ ), however, in postlingual deafness patients they were higher ( $P < 0.05$ ).

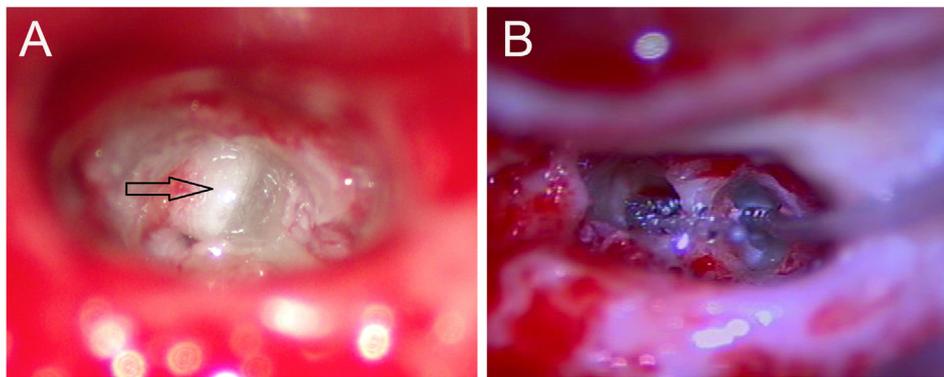
#### 4. Discussion

Bacterial meningitis is the most common cause of ossified cochleas reported in the literature [8]. Infection in the subarachnoid space reaches the cochlea mainly via cochlear aqueduct, which is the natural link between CSF to the inner ear [9]. Ossification centered at the basal turn of the cochlea could be explained by the anatomic route. Indeed, in our study 15 out of the 17 patients were caused by bacterial meningitis, of whom round window niche and basal turn were the most frequently areas involved.

Preoperative radiographic assessment is essential for determination of implantation candidacy, as well as preoperative planning. In accordance with the literature [10], our data showed no difference in predicting the occurrence of ossification between CT and MRI. Since the accuracy of both modalities appears inadequate, we recommend both of them in preoperative evaluation. Indeed, established ossification may be more evident using CT, fibrosis as an incipient sign of ossification is more likely to be discernible using MRI [10,11].

According to the literature, studies reported differential outcomes after cochlear implantation for patients with ossified cochleas. As presumed, several studies showed patients with partial insertion of the electrode array performed more poorly than patients with complete insertion [4,12]. The number of working electrodes in an ossified cochlea is an important factor influencing postoperative hearing and audiometric performance [13]. Parallel to the findings above, in our study patients in stage I and stage II had comparable audiometric performances with patients with nonossified cochleas (data not shown). This is primarily attributed to complete electrode insertion in the two groups. However, audiometric performances of cases with complete cochlear ossification in stage III were poor. The average hearing threshold was 40 dB HL, as a result, average CAP scores for P16 and P17 were much lower than those in other two groups. In fact, only 4 and 7 electrodes worked in P16 and P17 respectively, which could be the explanation of the differential results. However, Tokat et al. [7] and Nichani et al. [5] reported that patients with ossified cochleas might express satisfactory CAP scores as patients with nonossified cochleas, regardless of full or partial electrode insertion. The conflicting results might be attributed to different patients' conditions in different studies.

The timing of cochlear implantation for postmeningitic deafness is controversial. The length of time required for a trial with conventional amplification has been a subject of debate. Brookhouser et al. [14] followed up 64 children with postmeningitic deafness and found that auditory thresholds changed in 9 children over a variable interval ranging from 1 month to 12 years. Later on he conducted another follow-up study on 25 children with profound deafness after meningitis



**Fig. 4.** Intraoperative views of P12 and P16.

(A) Black arrow showing newly formed bone tissue in the lumen of the basal turn, with a much lower density than the lateral wall. (B) The electrodes wind around the modiolus through the man-made cochlear tunnel.

and found 3 children exhibited delayed improvement of hearing thresholds after 16 to 25 months [15]. Two other case reports also showed late hearing recovery of meningitic deafness and proposed that cochlear implantation should be delayed from 6 to 12 months to detect cases of spontaneous hearing recovery [16,17]. However, several other studies showed intracochlear osteogenesis started within 4 to 8 weeks after the acute meningitis [6,18]. Since CT and MRI might fail to reveal any sign of ossification in the early stage of labyrinthitis, they suggest fast cochlear implantation in patients with postmeningitic deafness [6,19,20]. In agreement with the later data, our results imply the time of cochlear implantation is an important factor in post-operative outcomes. P12 and P16, suffering from profound bilateral sensorineural hearing loss due to traumatic bacterial meningitis, opted for a conservative approach due to financial difficulties. However CT and MRI revealed stage III ossification in P16 6 months postmeningitis and stage II ossification in P12 4 months postmeningitis. Eventually P12 received complete insertion, whose audiometric performances were comparable to normal cochlear implantation; P16 ended up with partial insertion, only 4 electrodes confirmed working in the cochlear. No hearing recovery occurred on both of the patients based on 4-year follow-up. So, according to our experiences we advocate fast cochlear implantation to avoid dealing with labyrinthitis ossificans. As pointed out, the spiral ganglion cell count is inversely proportional to the degree of cochlear ossification, the survived auditory neurons in patients with complete cochlear ossification might be too small to provide useful speech recognition [9,21]. Moreover, early electric stimulation delivered by cochlear implants promotes increased survival of the auditory neurons and prevents the retrograde neural degeneration [12].

In our study, one patient received bilateral cochlear implantation, as stated previously [22], she conferred listening advantages for both speech recognition in noise and sound localization. Since potential ossification could occur in any ear in the long term, we recommend bilateral cochlear implantation in cases with postmeningitic deafness. Indeed, relatively small size of the patient population, especially the very small number of individuals in stage III might be considered as the limitation of our study. Larger numbers of subjects with longitudinal data sets should be considered in future studies.

## 5. Conclusions

In the light of our experiences, CT and MRI have comparable value in predicting the occurrence of ossification in cochlea. We recommend fast surgical intervention in the patients with bilateral profound postmeningitic deafness. If possible, bilateral cochlear implantation is recommended for each patient due to the unique development of deafness secondary to meningitis.

## Disclosure statement

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this article.

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