



# Cochlear Implantation in a Patient with Paget's Disease

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## INTRODUCTION

Paget's disease is the second most common metabolic bone disease [1]. The disease is characterized clinically by painful bony lesions, skeletal deformities, fractures and bone tumors. Other manifestations include cranial and spinal nerves involvement, cardiac abnormalities, electrolytic imbalance & hyperparathyroidism [2,3]. Development of a significant hearing loss is seen in as much as 60% of the cases [4,5].

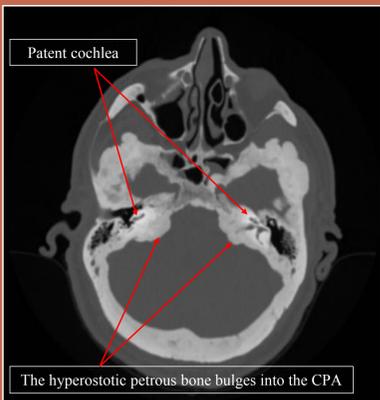
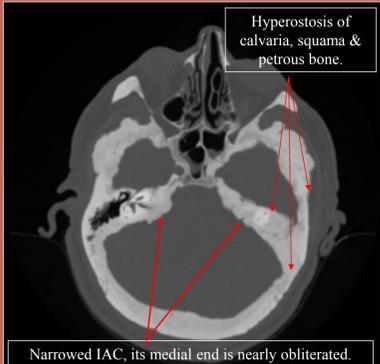


Fig 2: HRCT images demonstrating homogeneously diffused hyperostotic skull, patent cochlea and obliterated lateral part of IAC bilaterally.

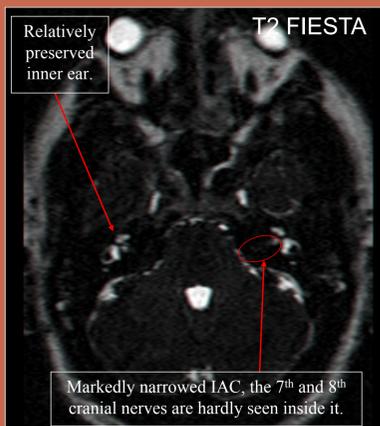
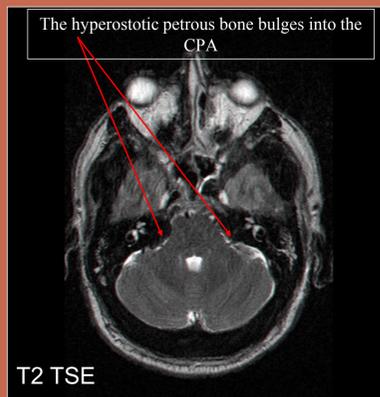


Fig 3: MRI images, demonstrating homogeneously diffused hyperostosis of the skull, nearly obliterated IAC bilaterally. Markedly narrowed CPA, with 7<sup>th</sup> and 8<sup>th</sup> cranial nerves hardly seen.

## REFERENCES

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## CASE REPORT

We report of a 59-year-old male with Paget's disease related severe-to-profound SNHL (Fig. 1) who was referred to our center for cochlear implantation. No responses could be recorded on ABR. His communication skills were lower than expected according to the audiogram and raised concerns regarding post implantation outcome.

Imaging showed diffuse hyperostosis of the calvaria and the temporal bone. The cochlea was patent but the internal auditory canal (IAC) was nearly obliterated bilaterally on its medial end [Fig. 2] with cranial nerves 7th & 8th hardly seen even in it. The hyperostotic bone bulged into the CPA to obstruct the possible view of the nerves [Fig. 3]. In order to maximize chance for reasonable outcome it was decided to implant the better hearing ear (right ear).

## COCHLEAR IMPLANTATION

At implantation, markedly rough skull bone was encountered. The bone was firmer than usual and tended to bleed more easily. A Nucleus® Freedom™ CI24RE with contour advance™ electrode was implanted. All electrodes were inserted into the scala tympani.

## POST IMPLANTATION SWITCH-ON AND OUTCOME

At switch-on all electrodes were activated. Neural Response Telemetry (NRT), a non-invasive direct physiological measurement of the evoked compound action potentials of the auditory nerve via the implant, revealed no response. Nevertheless, the patient did detect pitch difference across the electrode array during the NRT measurement. The ability to differentiate pitch difference across the electrode array could be considered as an indication for future good speech understanding.

Further assessments failed to record NRT responses nor electrically derived auditory brainstem responses (EABR) via the implant, since technically it was not possible to reach sufficient intensity level for recording EABR. At stimulation with relatively low current intensity levels (150 currents unities) for which normally EABR can be easily elicited, an “Out of Compliance” message was displayed.

Nevertheless, during the following months his communication skills continued to improve. At nine months post implantation he could easily follow a conversation with familiar talkers, his monosyllabic words (AB) recognition in quiet score was 65% and the CID sentence recognition in noise S/N + 15 score was 44%. The patient was using 900Hz ACE map, and the behavioral T and C levels were within normal range [Fig 4].

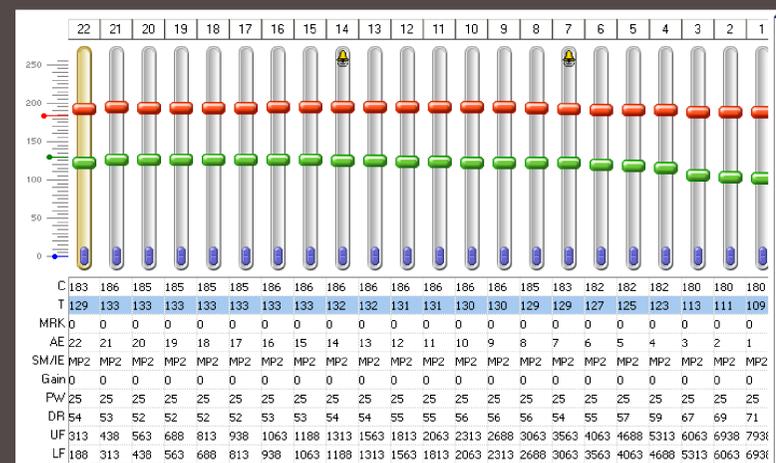


Fig. 4: Mapping: ACE 900Hz, T and C levels within normal ranges. T level: Threshold level – the minimum stimulation level that the subject can detect on a channel. C level: Comfort level – the loudest comfortable stimulation level on a channel.

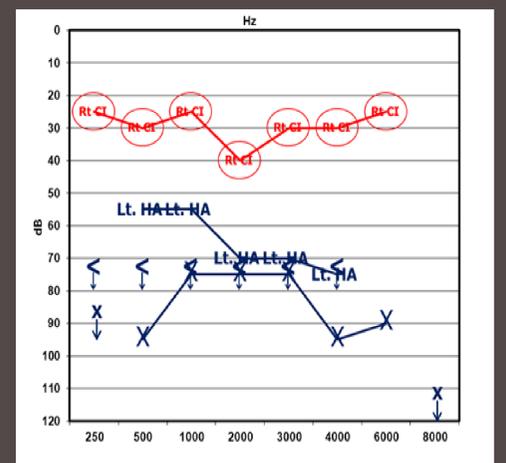


Fig. 5: Hearing thresholds 5 years post right cochlear implantation.

One year post cochlear implantation, the patient suffered significant fluctuations in his speech perception abilities. Integrity test results returned normal. Close follow-up and re-mapping enabled the patient to regain his previous post-implantation function. 4 years later, another drop in hearing function occurred, leading to a presumed diagnosis of soft failure, and replacement of the device was considered. However, few more weeks later hearing function stabilized and returned to previous levels [fig 5]. As speech understanding was still not satisfactory, and the fact that there was no benefit from a hearing aid on the left side, decision was made to proceed with a sequential contralateral cochlear implantation, thus maximizing the bilateral binaural effect, and minimizing the risk of losing exciting benefit from the cochlear implant on the right side. The patient is now scheduled for a contralateral cochlear implantation in the near future.

## COMMENTS

Reviewing the literature, we found 3 case reports about patients with Paget's disease who underwent cochlear implantation in whom similarly to our patient open set speech understanding was achieved [6–8]. Fluctuations in hearing performance post CI were not documented in those case reports. Implantation in our patient was straight forward, since the cochlea was patent. As commonly accepted today, a narrow IAC is not a contra-indication for implantation, since it is possible that the amount of surviving auditory neural elements is still high enough to allow benefit from CI [9]. And yet, level of auditory nerve degeneration might be significant in these cases so that it might not be possible to rely on NRT/EABR, especially when the disease responsible for the bony IAC narrowing consists of a neurologic deficit as well [10]. When fluctuations in cochlear implant function occur, device failure must be ruled out. When formal integrity test is normal, a “soft” device failure can be proved only by gaining better function after re-implantation.

In the present case, the risk of hampering speech perception function by re-implantation is high, and a sequential bilateral cochlear implantation is indicated, with anticipation for a similar outcome to that of the first CI, that would compensate for any future events of unilateral fluctuations in CI function.

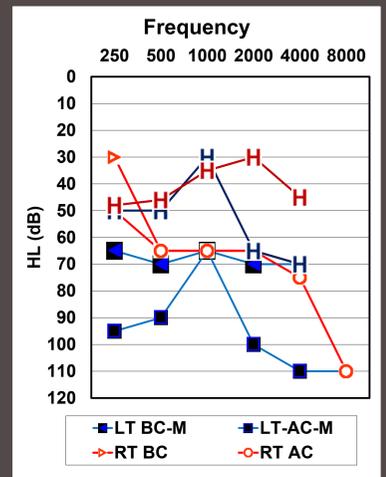


Fig. 1: Audiometry