

# Cochlear implantation in adults with prelingual deafness - a case report

## Abstract

Cochlear implantation in adult with prelingual deafness is not a common practice in Malaysia. There are limited data and publication indicating the outcomes in adult with prelingual deafness. These cases are usually not accepted in government hospitals and hence they will seek for help in the private hospitals. A gentleman with prelingual deafness was first seen at our hospital in the year of 2011 for cochlear implantation consultation. Finally, he had his cochlear implantation done in the year of 2014. His last aided evaluation showed the responses at 15-20dB across frequency 500-4000Hz with the usage of cochlear implant.

**Keywords:** cochlear implant, prelingual, deafness, adult

Volume 7 Issue 4 - 2017

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**Received:** October 07, 2016 | **Published:** June 07, 2017

**Abbreviations:** CI, cochlear implant; SNHL, sensorineural hearing loss; HRCT, high-resolution computed tomography; NRT, neural response telemetry; N6 SP, nucleus 6 speech processor

## Introduction

Cochlear implantation has been proven to be an effective treatment for patients with severe to profound sensorineural hearing loss (SNHL) especially in children with prelingual deafness or individuals with post-lingual deafness. Earlier implantation usually leads to a better outcome for children with prelingual deafness and they have higher possibility to develop their speech to be on par with the typical developing children with normal hearing. However, the outcomes for adults with prelingual deafness are not commonly seen in the papers and published data often showed enormous variability among individual. This case presents a 33-year-old gentleman with prelingual deafness that had undergone cochlear implantation in the year of 2014 and the outcomes after two years of re-habilitation.

## Case presentation

This 33-year-old gentleman was first seen at our clinic on 4<sup>th</sup> March 2011 following the referral by Consultant ENT-Head + Neck Surgeon for the consideration of CI candidacy. He had been diagnosed with hearing loss since three years old. He discontinued wearing the hearing aids because he rejected the hearing aids. He was not using any device until the age of 16. He had his hearing aid fitted on the right ear and had been using it consistently until now. During the initial session, he was only using the hearing aid on the right ear but not on his left ear, following the advice given by a “doctor” at his hometown. According to his sister, he responded to sounds at home but relied solely on sign language for communication. Sometimes, he will write in English language to communicate.

An otoscopic examination demonstrated clear ear canals and intact tympanic membranes in both ears. Pure Tone Audiometry indicated profound SNHL in both ears. The aided evaluation revealed the under amplification of the hearing aid on the right ear. He started using the hearing aid on his left ear since March 2011, followed our advice. Several sessions of hearing aids fine tuning and CI counseling were done to provide a clear and realistic picture on his expectation.

He decided to proceed with the cochlear implantation on August 2014. The High-resolution computed tomography (HRCT) revealed

that the internal auditory meatus was normal and symmetrical. The cochlear loops were normal as well. The baseline hearing aid evaluation showed the under amplification at the high frequency. He was able to detect /a/, /u/ and /m/, responded inconsistently on /i/ and unable to detect /s/ and /ʃ/. He was not able to discriminate the six Ling sounds. Realistic expectation, motivation and commitment were once again emphasized. We also found out that he showed psychosocial disadvantages resulting from his hearing loss. He was not close with the other family members except his sister. He had not been sharing his thoughts and ideas to his parents and brother for some times.

These also indicated that there was a need to continue with the counseling and psychological adjustment even after the cochlear implantation. He underwent cochlear implant surgery on 26th August 2014. He was implanted in his left ear using Cochlear Nucleus CI 422 with slim straight electrode. The surgery went well. Intra-operation measurement showed good impedance for all the electrodes. We were able to obtain the responses for all the electrodes except electrodes one to three and 19 using Neural Response Telemetry (NRT). The Nucleus 6 CP910 speech processor was switched on on 11th September 2014. We were able to get good impedance for all the electrodes. The NRT was obtained at the ranges of 150 to 200 cu for all the electrodes except at electrodes one to three. The gap between the C-level and T-level were noted too narrow. Hence we decided to set C-level behaviorally and set the T-level 40cu below the C-level. He commented that the sound was louder than his hearing aids when we turned the N6 SP to live at the clinic. He accepted the initial settings well and wanted to maintain the initial setting despite the louder sensation level.

He came back for follow up two weeks later after the switch on. The family members reported that he was able to identify cutlery and footsteps sounds. He was able to detect all 6 Ling sound - /a/, /m/, /u/, /i/, /s/ and /ʃ/ but still struggled with identification on 3weeks after the switch on. His listening performance continued improving, he was able to identify more environmental sounds such as water from tap, door bell and phone ringing on the 7thweek after the switch on. He was also able to identify the number of the syllables presented. Aided responses were obtained at the range of 20-30dB across the tested frequency.

Up to date, his aided responses were obtained at 15-20dBHL across the tested frequency. Currently, he communicates mostly by sign language and writing but he is now more confident in using

speech to express himself. He is able to produce at least 50 single words spontaneously and he also has a number of two to three words phrases, such as 'thank you', 'good morning', 'last time', 'how are you' and 'I am sorry'. He is motivated in learning to speak when he comes for rehabilitation. Also, during speech therapy, he relies less on lip-reading to understand what is being spoken as his listening skills starts to improve. He practices at least 3-5 times a week at home using phone application to enhance his identification skills on environmental sounds as well as 6 Ling sounds. His sister will do the practice with him when he goes over to her house. However, there is still a lack of practice at home with his parents and brother.

## Discussion

Unlike children with prelingual deafness and patients with post lingual deafness, the listening performance of adults with prelingual deafness was usually observed with enormous variability among individual and lower speech perception abilities.<sup>1</sup> due to the cortical colonization and reorganization by other sensory modalities.<sup>2,3</sup> The slow progress of listening performance was shown in this 33 years old gentleman. He still struggles in words identification especially in noisy environment even though he has received aural rehabilitation with the Audiologist and Speech-Language Therapist for two years now.

Another possible reason that leads to the slow progress with this patient was due to the limited support from the family members and patient's characteristic. According to Teoh et al.,<sup>2</sup> they realized that patients' characteristics played a main role in determining the observed outcome measures instead of the implants brands Cochlear Limited, MED-EL or Advanced Bionics.<sup>1</sup> Our patient is willing to have the training done by his sister but not the other family members. Unfortunately, he can only meet his sister once in a month due to distance issue. Hence, he will practice his listening skills using phone application. Adults with prelingual deafness and/or limited listening experience should not be the reasons for not continuing with the cochlear implantation. They do have the possibilities to develop oral

language even in late implantation.<sup>4</sup> Our patient was implanted late which was at the age of 31 and had no listening experience until he was fitted with the hearing aid at the age of 16. There was also limited auditory training received prior his cochlear implantation. He is now able to produce at least 50 single words spontaneously and started to use two to three words phrases. However, it is very important to explain to the patient and the family members and set a realistic expectation on speech and language level because all the implantees have different outcomes.<sup>4</sup>

## Acknowledgements

The author thank Mr. Saravanan Selanduray, Senior Audiologist, and Ms. Christine Yu, Speech-Language Therapist for their assistance in this case report.

## Conflicts of interest

Author declares there are no conflicts of interest.

## Funding

None.

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