A new experience of auditory brainstem implantation in Malaysia

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SUMMARY

Auditory brainstem implantation (ABI) is the only solution to restore hearing when cochlear nerves are disrupted together with the pathologies where bilateral cochleae do not provide a suitable location for cochlear implantation. We reported first two successful auditory brainstem implantation cases in patients with neurofibromatosis Type II (NF2) with bilateral acoustic neuroma causing bilateral profound sensorineural hearing loss in Malaysia. A good candidate selection, dedicated surgeons and rehabilitation team as well as strong family support are the crucial factors in achieving the best possible surgical, audiological and speech outcomes.

INTRODUCTION

Neurofibromatosis Type II (NF2) is a rare autosomal dominant condition resulting from mutations in the NF2 tumour suppressor gene located on chromosome 22q. The incidence and prevalence of NF2 in Malaysia have not been previously studied but they are estimated to be lower than western countries. Bilateral acoustic neuromas are common manifestations in NF2 patients causing deafness. The growth of the tumour affecting the cochlear nerves and the surgical resection of these lesions, usually result in these patients being not suitable candidates for cochlear implantation as a mode of hearing rehabilitation. Auditory brainstem implant was designed to bypass the cochlear nerve and to directly stimulate the cochlear nucleus complex. Auditory brainstem implantation (ABI) was first performed in 1979 by House and Hiltselberger using a single electrode.1 Here, we report the first 2 cases of auditory brainstem implantation (ABI) which were performed in Malaysia with its promising outcome.

CASE REPORT

Case report 1

A 29-year-old gentleman presented with bilateral profound sensorineural hearing loss in 2008. Further radiological investigations revealed bilateral acoustic neuroma. He was also diagnosed to have NF2. He subsequently underwent multiples cranial and spinal surgeries to excise the neuromas, followed by radiosurgery. As the remaining tumours at cerebellopontine angle were stable after five years of surveillance and with good family support, he was offered ABI to restore his hearing. He underwent an auditory brainstem implantation (Med-EL Company, Innsbruck, Austria, Fig. 1) via a right translabyrinthine approach in June 2016. Two months post-implantation switch-on revealed 12 electrodes being stimulated with good response. His verbal communication skill was enhanced with lip reading.

Case report 2

A 27-year-old gentleman presented with bilateral profound sensorineural hearing loss in 2006. Further radiological investigations revealed bilateral acoustic neuroma. He was subsequently diagnosed with NF2. He underwent multiples cranial surgeries to excise the neuromas, followed by radiosurgery. As the remaining tumours at cerebellopontine angle were stable and with good family support, he was offered ABI to restore his hearing. Two months post-implantation switch-on revealed 11 electrodes being stimulated with good response.

After one year of follow-up, both of them achieved mean pure tone hearing level at 500Hz to 4000Hz of 45dB HL with the range of 40 to 50 dB HL. In term of speech recognition at one year of follow-up, both of them achieved average closed set sound recognition at 80% and average closed set word recognition at 81%.

DISCUSSION

Bilateral acoustic neuromas in NF2 patients can cause hearing loss either from the tumour progression or from the complication of the surgical resection of the tumour. The aim of ABI in NF2 patients is to provide auditory sensations to enhance the patients' lip-reading skills by an average of 30% so that oral communications can still be possible and also to help the patients in detecting environmental sounds which will further improve their communication skills.[1]

The indications of ABI can be divided into two: postlingual and prelingual profound sensorineural hearing loss patients. Postlingual hearing loss patients can be subdivided into two groups, with or without tumours at cerebellopontine angle. NF2 patients are the group with tumours. The non-tumour group consists of patients with abnormal cochlea like cochlear ossification that prevents the patients from having normal cochlear implantation. Meanwhile, cochlear aplasia and cochlear nerve aplasia or other similar conditions that exclude normal cochlear implantation with intact cochlear nuclei at brainstem are the main prelingual hearing loss patients who will benefit from ABI.[2]

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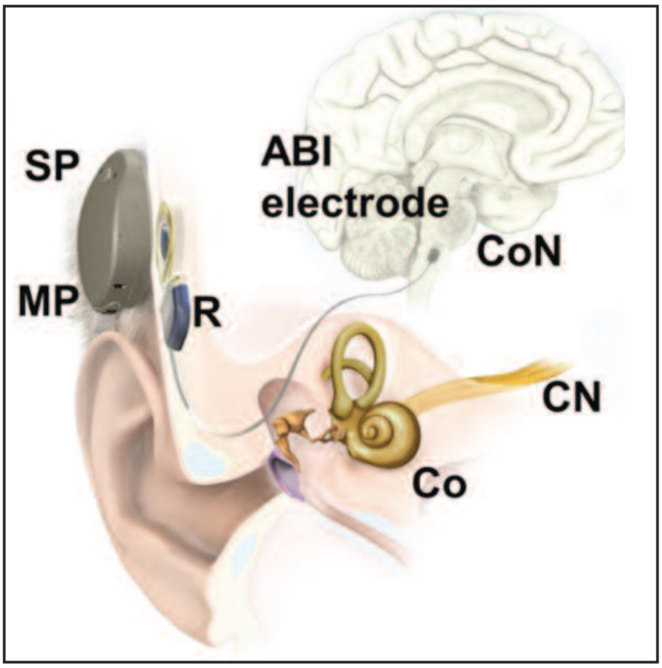

Fig. 1: [2]Scheme of the Auditory Brainstem Implant (ABI) supplied from MED-EL (Innsbruck, Austria). CN: Cochlear Nerve, Co: Cochlea, CoN: cochlear nucleus, MP: microphone part, R: receiver, SP: Speech processor part.
Illustration Courtesy of MEDEL

A thorough preoperative clinical, audiological and speech assessment is needed before embarking on surgery. It is important to counsel potential ABI recipients to understand and manage their expectations. [1] In both of our patients, clinical and radiological examinations were performed. In view of the previous history of surgery and radiosurgery done on both patients, thorough counseling regarding the realistic expectations in terms of speech and hearing outcomes was given to the patients and family members. They understood clearly the procedures and the need for long term speech and audiology rehabilitation.

The surgical approach for ABI consists of translabyrinthine and retrosigmoid approaches. Both of our patients were operated via translabryinthine approach as this approach enabled the surgeons to remove the remaining tumour with direct vision at internal acoustic meatus that blocked the pathway to cochlear nucleus during the insertion of ABI electrode array. The cochlear nucleus located within the lateral recess of the fourth ventricle was identified using the facial nerve, lower cranial nerves (glossopharyngeal and vagal nerves), and choroid plexus as landmarks. Evoked auditory brainstem response (EABR) was used to confirm proper positioning of the implant electrode array during surgery.

The device usually activated at about eight weeks post-operatively. Threshold and maximum comfortable levels are set during the initial session. Electrodes are activated sequentially and the intensity gradually increased to obtain thresholds while avoiding adverse effects.

Implant recipients have to be patient as the outcome of the surgery may not be so evident in the short term. Learning and acclimatization to the implant may take months to years in achieving the maximum benefit of the device. Behr et al3 showed that patients who had been deaf for a year or less had a more favorable audiological outcome compared to patients who have been deaf longer. Patients' age, the size of the tumour and the tumour staging did not play a role in affecting the outcome.[3] The most important surgical factors in preserving hearing were good microsurgical skills with minimal manipulation of the brainstem and proper placement of the electrodes. Both of our patients achieved reasonable progress in regaining hearing and speech recognition after a year of surgery.

CONCLUSION

ABI is the only form of speech rehabilitation for patients with NF2. These first two cases that were performed in Malaysia would pave a way and served as a guide for future ABI surgeries in this region. An experienced and dedicated team of otologist, neurosurgeon, audiologist and speech therapist is essential in ensuring the best possible audiological and speech outcomes for the ABI recipients.

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