Case Report

Intraoperative Electrically Evoked Auditory Brainstem Response (eABR) examination in cochlear implant candidacy

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ABSTRACT

Background: Cochlear implantation is one of the most promising hearing habilitation methods for patients with profound sensorineural hearing loss (SNHL). One of the difficulties associated with the method is predicting the outcome of the surgery, especially in patients with cochlear nerve abnormalities. **Purpose:** To evaluate the intraoperative cochlear nerve response using Electrically Evoked Auditory Brainstem Response (eABR), and determine if cochlear implantation was required. **Clinical question:** How significant was intraoperative eABR in determining the outcome of cochlear implantation? **Case report:** This study considered two cases with profound bilateral SNHL that underwent intraoperative eABR. The first case involved 10-year-old girl who had used conventional hearing aids for eight years before surgery. She practiced lip reading for communication. The second case involved 4-year-old boy with delayed speech and a history of febrile seizure when he was two years old. He had used a conventional hearing aid for six months. **Method:** Evidence based literature was conducted through PubMed, Embase, and Wiley. **Result:** On the first case, eABR examination evoked no response in both ears and the parents decided not to carry on with the surgery. In the second case, eABR examination aroused a significant response in both ears, so the implantation was performed. Three months postoperative, the child was babbling and able to detect sound. **Conclusion:** eABR could give valuable input in identifying profound bilateral SNHL patients with cochlear nerve abnormality. The high cost of implant devices makes this examination beneficial for the patient's family in deciding if implantation surgery is required.

Keywords: cochlear implant, cochlear nerve hypoplasia, eABR, hearing habilitation, profound sensorineural hearing loss

ABSTRAK

Latar belakang: *Implantasi koklea merupakan salah satu metode habilitasi pendengaran yang paling menjanjikan bagi pasien dengan gangguan pendengaran sensorineural berat. Salah satu kesulitan yang terkait dengan metode ini adalah memprediksi hasil pembedahan, terutama pada pasien dengan kelainan saraf koklearis.* ***Tujuan:*** *Untuk mengevaluasi respons saraf koklearis intraoperatif memakai* Electrically Evoked Auditory Brainstem Response (eABR)*, guna menentukan apakah implantasi koklea dapat dilanjutkan.* ***Pertanyaan klinis:*** *Seberapa signifikan* eABR *intraoperatif dapat menentukan hasil akhir implantasi koklea?* ***Laporan kasus:*** *Mendiskusikan dua kasus dengan tuli sensorineural sangat berat bilateral yang dilakukan eABR intraoperatif. Kasus pertama adalah seorang anak perempuan berusia 10 tahun dan telah menggunakan alat bantu dengar konvensional selama delapan tahun sebelum operasi. Ia berkomunikasi menggunakan baca bibir. Kasus kedua adalah seorang anak laki-laki berusia empat tahun dengan keterlambatan bicara, dan mempunyai riwayat kejang demam pada usia dua tahun. Ia telah menggunakan alat bantu dengar selama enam bulan.* ***Metode:*** *Penelusuran literatur berbasis bukti dilakukan melalui* PubMed, Embase, *dan* Wiley. ***Hasil:*** *Pada kasus pertama pemeriksaan eABR tidak memberikan respons pada kedua telinga dan orang tuanya memutuskan untuk tidak melanjutkan operasi. Pada kasus kedua, pemeriksaan eABR memberikan respons yang signifikan pada kedua telinga sehingga implantasi tetap dilakukan. Tiga bulan pasca operasi, anak ini sudah bisa mengoceh dan mampu mendeteksi suara.* ***Kesimpulan:*** *eABR dapat memberikan masukan berharga dalam mengidentifikasi pasien tuli sensorineural bilateral dengan kelainan saraf koklearis. Mahalnya biaya untuk perangkat implan koklea membuat pemeriksaan ini bermanfaat bagi keluarga pasien dalam memutuskan apakah akan melanjutkan operasi implantasi.*

Kata kunci: *implan koklea, hipoplasi saraf koklearis,* eABR, *habilitasi pendengaran, tuli sensorineural sangat berat*

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INTRODUCTION

Currently, hearing habilitation using a cochlear implant is a very promising treatment for people living with profound sensorineural deafness who experienced little to no improvement using conventional hearing aids (HA). Cases involving inner ear malformation present significant challenges considering intraoperative risk and expectations related to postoperative outcome. The outcome of cochlear implantation also depends on myriad other factors, including brain capability in perceiving and interpreting sounds. Technological advancement and increasing demands placed on such devices have together contributed to rapid development in the devices' design, including the number of channels and electrodes. This development has indirectly caused the price of these devices to escalate.[1]

Since 2012, Cipto Mangunkusumo Hospital, Jakarta has provided cochlear implantation surgery, however, it remains exceptional due to exorbitant appliance cost and a lack of coverage by health insurance. In 2015, cochlear implant surgery became more common because the medical cost became covered by government health insurance, though the cost of the implant device still should be borne by the patient's family.

Cochlear implant candidates must undergo several routine examinations including an aural physical examination and audiology examinations including a behaviour test, tympanometry, an otoacoustic emission (OAE) test, Brainstem Evoked Response Audiometry (BERA), Audiometry Steady-State Response (ASSR), and a hearing examination using hearing aids. Radiologic examinations such as Computed Tomography Scanning and Magnetic Resonance Imaging are performed to evaluate cochlear and cochlear nerve anatomy, which is necessary to predict factors of surgical relevance and its likely outcome. In the presence of cochlear malformation or cochlear nerve deficiency, cochlear implantation is often controversial. The results of all these examinations merit consideration in deciding whether a patient is eligible for surgery.[1] Even though cochlear implantation has been found to have positive outcomes for both hearing and speech improvement in several studies, Niparko et al.[2] reported that 15-30% of the cohort performed poorly during two years of postoperative observation.

Electrically Evoked Auditory Brainstem Response (eABR) is an Auditory Brainstem Response (ABR) test variant using direct electrical stimuli to the cochlea. Two types of eABR are currently available, corresponding to the method of delivering the stimuli. Transtympanic eABR (TT-eABR) or Round Window eABR are performed before inserting electrodes into the cochlea, while Implant eABR (Imp-eABR) is an electrical stimulation performed after electrode insertion. Response-related latency periods vary between ABR and eABR due to the presence of shortcut from the outer to the inner ear in eABR; resulting in the absence of cochlear-derived waves I. As a result, latency periods in eABR are 1-1.5 ms faster than in ABR.[3,4] According to Gibson,[5] the amplitude observed in eABR examinations reflects

the stimulability of remaining cochlear nerves, while the wave latency period reflects auditory pathway synchronization quality extending to the brainstem.

This paper reported the results of eABR examinations on two cochlear implant candidates before electrode insertion, conducted as a part of tests to determine which ear would receive the implant. The results had significance not only to the patient and the patient's family, but also to the hospital and cochlear implant vendor. Conducting eABR examinations in cases with an uncertain outcome could help medical providers avoid inappropriate implantation of cochlear implants. Kileny et al.[6] demonstrated the efficacy of TT-eABR in predicting the benefit of cochlear implantation surgery in cases involving cochlear malformation, or cochlear nerve hypoplasia. In addition, Kim[4] reported significant differences in the rate of language comprehension development, between subjects with cochlear malformation who responded well to the TT-eABR examination and those with subpar responses. Accordingly, eABR examinations could help prevent patients and their families from being disappointed by unfavourable surgical outcomes, and also help prevent the incursion of high medical costs related to insalubrious cochlear implantations, which can range from USD 14,000-37,000.

Methods of eABR examination had been explained by Kileny et al.[6] in previous studies reported that examinations were performed intraoperatively under general anaesthesia, using neuromuscular blockades as needed to prevent artefacts produced by facial nerve stimulations. The response to electric-auditory stimulus of the brainstem was obtained using a simulator producing biphasic waves with a beta output up to 999 µA and a wave duration configured at 200 µs. Responses were recorded through a subdermal needle using electrodes placed on the forehead (non-inverting) and the non-stimulated ear (inverting), with a grounding electrode placed on the patient's neck. The stimulus was given at low speed, approximately 7.7 waves per second, to avoid the formation of artefacts. The brainstem electric-auditory threshold response was evaluated based on the presence of waves V, the peak-to-trough amplitude of waves V and the peak latency measured at the threshold.[6]

CASE REPORT

The first case concerned a 10-year-old girl who was non-responsive towards calls and loud noises, such as the sound of slammed doors and vehicle horns. The non-responsiveness was first recognized by the parents when the child was 15 months old. The mother denied any illness or medication consumption during the gestational period. The patient had a normal birth history and healthy motoric development. Parents suspected their child had a hearing problem, and brought her to Krakatau Medika Cilegon Hospital, where the patient was advised to undergo hearing examination in Otolaryngology-Head and Neck Department, Cipto Mangunkusumo Hospital.

The examination results revealed profound hearing impairment in both ears, and the patient was further advised to use super-power hearing aids (HA) in both ears. The patient began using HA at age two and routinely underwent biannual medical check-ups. During the habilitation process, the patient attended speech therapy and attempted several Audio Verbal Therapy (AVT) procedures, however she ceased to follow the regime due to inconvenience related to time and distance. During the process, the patient gained the ability to pronounce muddled words and speak short sentences such as "want to eat" or "I want to bathe". Currently, the patient was a fifth-grade student in a public elementary school and was able to socialize with peers, self-reliant and active. During history taking, the patient cooperated in communicating, and

tried to understand questions by lip-reading.

Examination of the patient's ears, nose and throat showed no abnormality, and intact tympanic membranes. Tympanometry assessment resulted in tympanogram type A on both ears, interpreted as no dysfunction in the middle ear. Otoacoustic Examination (OAE) resulted in REFER for both ears, suggesting disturbances in outer hair cell emission on the cochlea of both sides. Brain Evoked Response Audiometry (BERA) was unable to detect waves V on both ears using 80 dB sound stimuli. The examination was followed by Auditory Steady-State Response (ASSR), through which a hearing threshold was acquired in the range of 100-110 dB. Obtained results suggested moderate to severe bilateral sensorineural hearing loss, and HA response evaluation on 70 dB.

Examinations in other departments such as paediatrics, neurology, child psychiatry, growth and development, and medical rehabilitation showed no apparent contraindications for cochlear implantation. Radiological examination showed the good anatomical structure of both cochleae, with bilateral posterior and horizontal semi-circular canal hypoplasia, alongside bilateral cochlear nerve hypoplasia, especially on the right side (Figure 1).

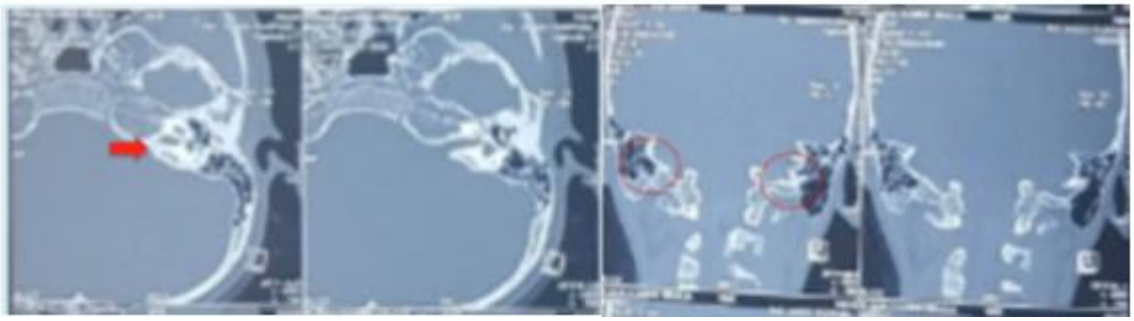

Figure 1. High resolution mastoid CT scan examination

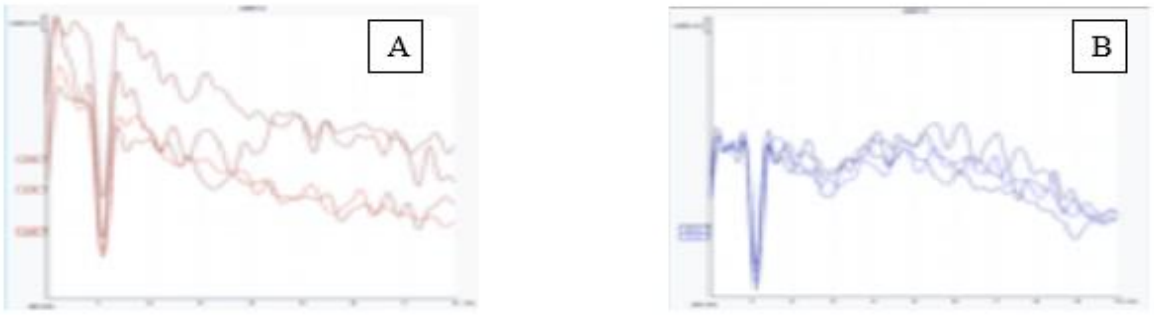

Figure 2. A. TT-eABR result of the right ear, B. TT-eABR result of the left ear

Based on these results, the patient was strongly recommended to undergo a TT-eABR examination to evaluate nerve response towards electrical stimuli, to determine which ear would undergo cochlear implantation. The examination showed no response; waves V could not be identified in either ear (Figure 2). Because no adequate response was obtained from either ear, cochlear implantation was cancelled after another informed consent was taken from the parents.

The second case being reported was a 4-year-old boy with the chief complaint of being non-responsive towards calls or loud sounds since the age of three. According to his parents, the patient was able to respond

to loud sounds or when he was called, previously. He was already able to sing and to count numbers when he was two. It happened that he had a high fever when he was three, and afterward showed no improvement in his vocabulary, and did not respond when his name was called. The patient was brought to the Otolaryngology Department of Hermina Kemayoran Hospital, and was diagnosed with severe hearing impairment; the use of HA was suggested, and he had used HA for six months. The patient could pronounce muddled words. The patient had participated in speech therapy for four months, followed by AVT therapy once a week for a month. During the physical examination, the tympanic membranes on both ears were found to be intact. An ear and throat examination also revealed no apparent abnormality.

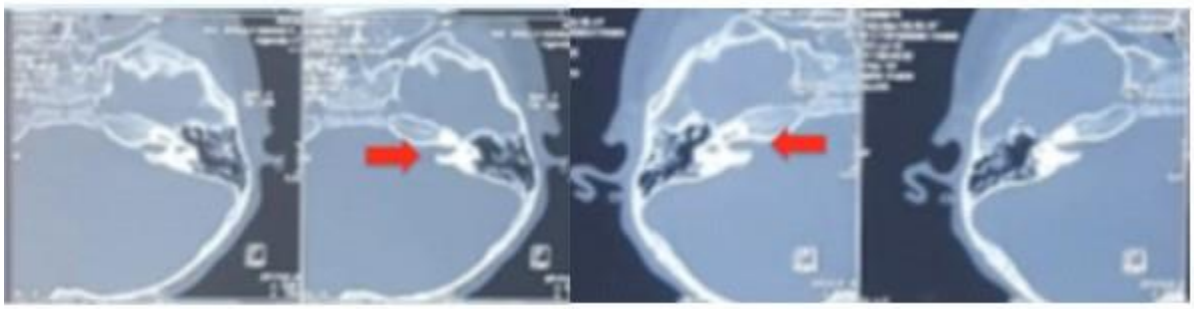

Figure 3. High resolution mastoid CT scan examination

Tympanometry resulted in type A (proper middle ear function), while OAE showed outer hair cell emission dysfunction on the cochlea of both ears. In the BERA examination, waves V were not detected on either ear until 90 dB, followed by ASSR procedure. The examination results suggested very severe bilateral sensorineural hearing loss. Examination by paediatrics, neurology, child psychiatry, growth and development and medical rehabilitation showed no contraindications for cochlear implantation. Radiological findings showed normal cochlea on both ears (Figure 3).

The results of these examination determined that this case was a candidate for cochlear implantation, and on the same day, TT-eABR was performed. The results from this examination revealed waves V in response to electrical stimulation (Figure 4).

The patient underwent cochleaer implantation on his right ear, and six months after surgery, the patient was able to pronounce new words.

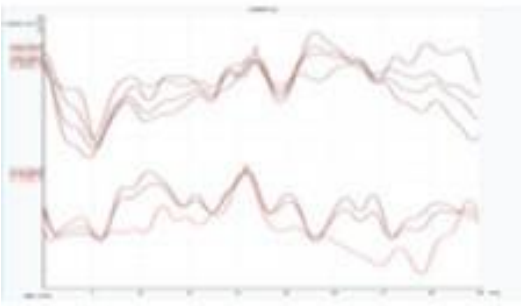

Figure 4. TT-eABR result with recognizable wave V

CLINICAL QUESTION

How significant intraoperative eABR in determining the outcome of cochlear implantation?

REVIEW METHOD

Literature search was conducted on December 2023 with the keywords "evoked auditory brain response" AND "cochlear implantation". We obtained 320 articles in Pubmed, 73 articles in Embase, and 49 articles in Wiley. There were 3 articles included in the study which were relevant to the topic.

RESULT

There were 3 studies included which were relevant to the topic. Based on a study by Lundin et al.[7], it was revealed that eABR latencies increased towards base stimulation of Cochlea with Wave V. The study was conducted in Uppsala University Hospital, Sweden with 74 adults and 4 children. Children were further investigated with high-resolution MRI before the cochelar implantation together with eABR. Similar results was also reported by Wang et al.[8] It was reported that average C value and V-wave I/O curve slope had positive correlation with postoperative auditory perception. A study by Kim et al.[9] also reported that eABR was useful in determining the outcome of cochlear implant procedure. It revealed that larger Wave V amplitude and shorter latency had much better outcome in speech performance.[4,9]

DISCUSSION

Decision whether to conduct a cochlear implantation in patient with hearing loss could be difficult. One of the objective techniques to evaluate is through eABR examination. The latency period and threshold of waves V identified through eABR examination could provide valuable information about the likely clinical outcome of cochlear implantation

In the first case discussed in this paper, eABR revealed that the patient had no response to electrical stimuli in either ear. Factors that could impede response recordings such as a false round window membrane, and the presence of liquid or blood were negative. In a study, Kileny[6] stated waves V could still be identified, for example in the case of narrowing Internal Auditory Canal (IAC) or cochlear nerve hyperplasia, even though with very low amplitude due to reduced nerves components, or responses are not well-synchronized. Kileny[6] also stated that TT-eABR examinations were important for predicting abnormal adaptation to electrical stimuli and helped the patient, family, and practitioner to educate and inform on cochlear implantation. As such, the first case was deemed to be inoperable, since no electrical stimuli-driven response was recorded in either ear.

Conversely, Buchman et al.[1] documented several cases involving inner ear malformations in which cochlear implantation led to excellent results, as long the abnormality was well-addressed radiologically, audiologically, and intraoperatively. In the second case presented in this paper, the patient was prepared as a cochlear implantation candidate, and eABR examination before electrode insertion showed a good response. As a result, the operation was conducted and led to an excellent outcome, as indicated by the patient's ability to respond to sounds and pronounce new words six months postoperatively.

In conclusion, cases involving congenital temporal bone anomaly, the eABR examination has two purposes: to predict patients' postsurgical performance, and to support in developing a management plan, including the potential insertion of an Auditory Brainstem Implant (ABI). The examination is also useful for assessing nerve pathway integrity, especially when it is hard to evaluate the child's behaviour.

A narrow IAC anatomical structure, as revealed in radiological findings, and the impression of cochlear nerve hypoplasia are not necessarily contraindications against cochlear implantation, especially if a response can be obtained from electrical stimulation during a perioperative eABR examination delineating auditory nerve pathway integrity.

CONFLICT OF INTEREST

The authors declare no conflict interests.

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