Journal of Current Research in Otolaryngology

Chembio Publishers
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Case Report *Volume 2; Issue 1*

Evaluation of Cochlear Implant Outcome Using Categories of Auditory Performance li in Inner Ear Malformations Patients

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Received Date: March 03, 2019; **Published Date:** March 14, 2019

Abstract

Background: Cochlear implants are used for patient in severe to profound sensor neural hearing loss, where hearing aids no longer could help. Mostly patient with inner ear malformations are suffering from severe to profound sensor neural hearing loss. An Inner ear mal formation occurs in about 20% cases of congenital sensor neural hearing loss. Whether or not the patients with inner ear malformations have cochlear nerve are considered difficult subjects because it will affect the ability of speech perception after cochlear implantation. Hearing ability and speech development of patients with inner ear malformations after cochlear implants are various.

Objective: To report three cases of inner ear malformations which one of the cases was cochlear nerve aplasia that cochlear implants have been used.

Method: Evaluation of patients after cochlear implant was assessed by using Categories of Auditory Performance II. Result: The categories of Auditory Performance II were 8 and 6 consecutively in the first and second case after 2 years of cochlear implant. The third case was assessed a year after cochlear implantation and the result was 5.

Conclusion: It was reported that three cases of inner ear malformations which cochlear implants have been used was compatible and have good result in Categories of Auditory Performance II.

Keywords: Inner ear malformations; Cochlear implant; Categories of auditory performance II

Abbreviations: OAE: Otoacoustic Emissions; ABR: Auditory Brainstem Response; ASSR: Auditory Steady State Response; CT: Computed Tomography; CAP: Categories of Auditory Performance

Introduction

Cochlear implant is a medical instrument that uses electrode array to stimulate spiral ganglion by putting it inside the cochlea [1]. Cochlear implantation is an effective method to help patients with severe to profound sensorineural hearing loss where hearing aids no longer could help [2]. Cochlear implant possibly given a satisfying result especially on children less than one year old, its where the age of normal speech has been occurs or develops. The cause of unsatisfying cochlear implant is multifactor, such as delay in motor development, age

Citation: Dini F and Haris M E. Evaluation of Cochlear Implant Outcome Using Categories of Auditory Performance li in Inner Ear Malformations Patients. J Current Res Oto 2019, 2(1): 180008.

when undergoing implant, and abnormal inner ear anatomy like cochlear nerve deficiency [3].

Minimum requirements in cochlear implantation are a patent cochlear lumen for placement of electrode and the existence of cochlear nerve fibers to carry auditory sensory directly to auditory cortex. Patients with inner ear malformations that have or not have cochlear nerve are considered difficult subject because it will affect the ability of speech perception after cochlear implantation. Kim and Jeong [1] stated that whether in patients with inner ear malformations have or not have cochlear nerve deficiency, it is better to undergo cochlear implantation. Young-aged patients with inner ear malformations that have vestibulocochlear nerve have a higher possibility to succeed in undergoing cochlear implantation.

The purpose of this case report writing was to report the result of categories of auditory performance II (CAP II) on children with inner ear malformations who have undergone cochlear implant.

Case Presentation

The first case was a one year old child; complain about her giving an unclear response when being called. Audiology examination consisted of tympanometry; otoacoustic emissions (OAE), auditory brainstem response (ABR), auditory steady state response (ASSR) and behavioral observational audiometry-aided (BOA-aided) revealed profound sensor neural hearing loss. Magnetic resonance imaging (MRI) showed appearance of normal cochlear nerve while computed tomography (CT) scan showed bilateral Mondini malformations (Figure 1). The cochlear implantation was performed on both ears. Finding during surgery at right ear was perilymph gusher and electrode not curve C-shape while at left ear was five electrodes outside the cochlea. The CAP II one year and two year after cochlear implant was five and eight.

The second case was a four year old child, suspicious of hearing loss and speech disorder. The result of ABR was no response on both ears while ASSR result can be seen at (Figure 2). Audiology examination revealed profound sensor neural hearing loss, MRI and CT scan showed left cochlear nerve aplasia and bilateral Mondini malformations (Figure 3). Cochlear implantation was performed on right ear. The CAP II one year and two year after cochlear implant was four and six.

The third case was a 4.5 year old child, suspicious of hearing loss and speech disorder. The result of ABR was no response on both ears while ASSR result can be seen at (Figure 4). Audiology examination revealed profound sensor neural hearing loss, MRI and CT scan showed bilateral cochlear nerve aplasia and bilateral Mondini malformations (Figure 5). Cochlear implantation was performed on left ear. The CAP II one year after cochlear implant was five.

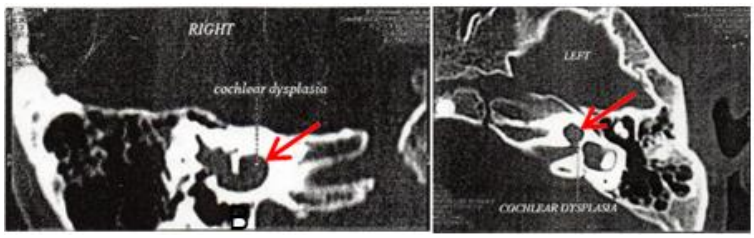

Figure 1: The result of CT scan of patient in case 1.

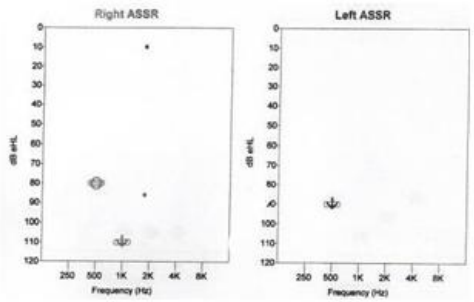

Figure 2: ASSR result in patient case 2.

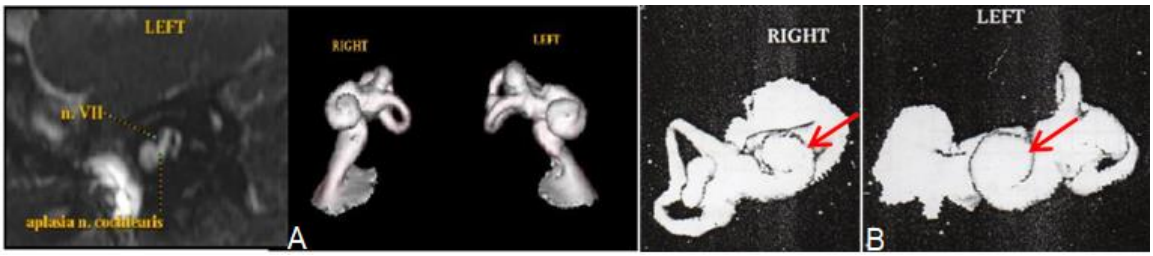

Figure 3: (A) MRI and (B) CT scan result of patient in case 2.

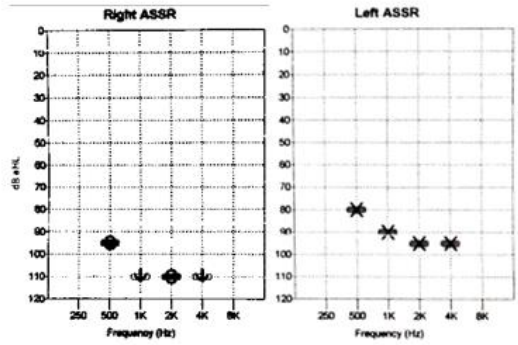

Figure 4: ASSR result of patient in case 3: Right 95 dB/ >110 dB/ 110 dB/ >110 dB; Left 80 dB/ 90 dB/ 95 dB/ 95 dB.

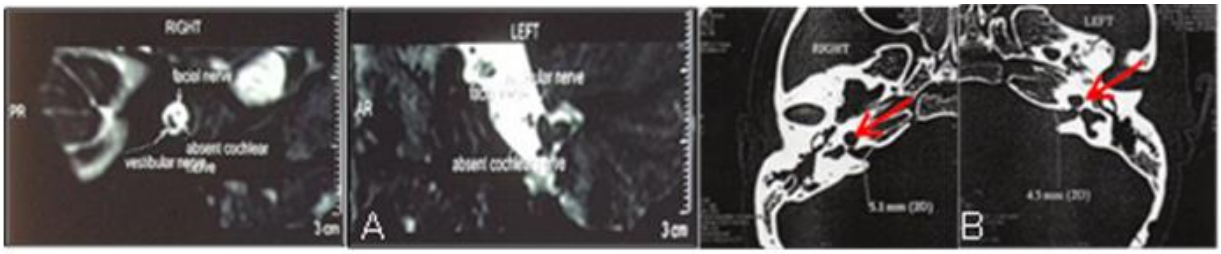

Figure 5: (A) MRI and (B) CT scan result of patient in case 3.

Discussion

Inner ear malformations in patients with congenital sensor neural hearing loss were occurs about 20 to 40%. Cochlear implant was standard therapy for children with severe to profound hearing loss. Anatomy of patients with inner ear malformations could increase the risks of surgical complication, such as cerebrospinal fluid gusher, facial nerve lesions, electrode insertion into internal auditory canal, or difficulty to spot cochlea [1]. Patients with cochlear nerve deficiency are not subjects for cochlear implantation, but auditory brainstem implant (ABI), which is the main therapy for cochlear nerve deficiency [2]. Auditory brainstem implant was major neurosurgical procedure where the long- term result is still unknown until now so cochlear implant is still an option before undergoing ABI [4]. In three cases above, risks of surgical complication and prognosis surgical outcome have been explained to the parents yet they were insisted on undergoing cochlear implantation. An examination that is currently used to assess results of patient hearing progress after cochlear implant in US and Europe cochlear implant centers is CAP II. Categories of auditory performance II is an index consisting of 10 auditory performances that in every level-up arranged to increasing difficulty. From category 0 (no awareness of environmental sound) to 9 (use of phone with unknown speaker in unpredictable context). This assessment is

easy for parents and medical staff to conduct daily. The good outcome of CAP II score, the good speech ability.5 Hearing evaluation after cochlear implantation in three cases above was using CAP II.

The ability to hear and discriminate speech sounds is important aspect in speech and language development. Human brain has high adaptability and sensitivity to the sounds. Even small noises still could be processed by brain that transferred the noise to be recognized by the hearer [6]. Sharma and Campbell [7] have proven that hearing system has the capability to non- degenerate and/or to exhibited plasticity until the age of 3.5 years old. Central hearing pathway still develops normally although it was not stimulated and still will not degenerate until age of 3 to 4. Central hearing pathway degeneration starts after the age of four through synapse elimination. However, some children less than 7 years still exhibit plasticity. The ages of patients in three cases above were respectively 2, 4, and 4.5 years old. These ages were still categorized as sensitive periods to develop ability so the patients still able to hear normally like other children in general.

The rate of successful cochlear implant depends on patients selection, surgery of cochlear implant, and post implant habilitation. These three factors were equally important and none of them should be eliminated. Key, Porter and Bradham6 stated that reorganizing of auditory cortex occurs in the first 6 to 8 months after cochlear implantation. Patient in case 1 had CAP II score by 5 and 8 after a year and 2 years of cochlear implant, while at the same time, patient in case 2 had the score of 4 and 6. The result was in accordance with study by Bakhshaee et al. [8]. Who stated that the outcome of cochlear implant could not directly known right after the surgery was performed. It needs an auditory habilitation in order to improve auditory performances.

Study by Chen et al. [9]. has shown that hearing developments of children with Mondidi deformity and normal ear were the same that hearing will be develops rapidly in three years after cochlear implant. Number of spiral ganglion cells in people with Mondini deformity was enough to stimulate the hearing nerve. This research has stated that inner ear malformations were not an absolute contraindication for cochlear implantation. Instead, cochlear implantation was effective intervention for patients with Mondini deformity. The three cases above were Mondini deformity cases and CAP II evaluation showed that patients could discriminate speech sound and understand common phrases without lip-reading precisely a year after cochlear implantation. Different research has been conducted by Nair et al. [10] who have stated that hearing ability of children with inner ear malformations will gradually increases after cochlear implant, but it cannot reach the same level as children with normal cochlea. Final result of cochlear implantation cannot be fully predicted because there were various factors affecting the result. Many reports regarding cochlear implant outcome in patients with inner ear malformations have been published, but the number of patients is still limited. An outcome of hearing evaluation after cochlear implant will be better in patients with mild inner-ear malformations than severe. Dettman et al. [11] have stated that speech-language ability is more related to the age. Jeong and Kim [12] have also stated that speech perception in children with inner ear malformations was determined by age and size of cochlear nerve.

Speech perception in children with cochlear nerve deficiency that undergone cochlear implant were various. Vincenti et al. [13] have reported cochlear implant outcome of some children with cochlear nerve deficiency were only able to detect sound, the others were able to understand common speech. Wu et al. [14] have proven that children with nerve hypoplasia have a good outcome after cochlear implant, while children with nerve aplasia have worst result. However, evaluation outcome of patient who had cochlear nerve aplasia in the third case shown a different result. The CAP II score of the patient after a year of cochlear implant was five, which means that patient was not only able to detect sound but also able to understand common phrases. This score was in accordance with the scores of patients in the first and second case in which patients have normal cochlear nerve. Three cases of inner ear malformations in cochlear implants have been reported. These cases obtained a good result of CAP II score. Even in the third case, which patient had cochlear nerve deficiency has a good result as well. Patients with inner ear malformations still have probability for cochlear implantation, although this therapy is still considered controversial. Expensive cost of cochlear implant instrument, high complication risk, and uncertain outcome are allegation reason for parents to choose this habilitation therapy. This case report was expected to become an brand new information for parents to decide strategic therapy for their children with inner ear malformations.

References

1. Kim LS, Jeong SW (2017) Outcome of cochlear implantation in children with cochlear nerve deficiency and/or inner ear malformations. In: Kaga K (Ed.), Cochlear implantation in children with inner ear malformation and cochlear nerve deficiency. (1st edn), Springer, Singapore. pp. 169-913.

2. Surmelioglu O, Tarkan O, Ozdemir S, Tuncer U, Kiroglu M, Atik FA et al. (2016) Cochlear implantation in patient with cochlear nerve deficiency: a case report. J Int Adv Otol 12(2): 213-215.

3. Birman CS, Powell HR, Gibson WP, Elliot EJ (2016) Cochlear implant outcomes in cochlea nerve aplasia and hypoplasia. Otol Neurotol 37(5): 438-445.

4. Sennaroglu L, Bajin MD (2017) Classification of inner ear malformations. In: Kaga K (Ed.), Cochlear implantation in children with inner ear malformation and cochlear nerve deficiency. (1st edn), Springer, Singapore. p. 61-84.

5. Gilmour L (2010) The inter-rater reliability of category of auditory performance-II (CAP)-II. Thesis. Southampton: University of Southampton.135: 14-20.

6. Key APF, Porter HL, Bradham D (2010) Hearing processing following sequential bilateral cochlear implantation: a pediatric case study using event-related potentials. J Am Acad Audiol 21(4): 235-238.

7. Sharma A, Campbell J (2013) A sensitive period for cochlear implantation in deaf children. J Matern Fetal Neonatal Med 241(1): 151-153.

8. Bakhshaee M, Sharifian MR, Ghasemi MM, Naimi M, Moghiman T (2007) Speech development and auditory performance in children after cochlear implantation. MJIRI 20(4): 184-191.

9. Chen X, Yan F, Liu B, Liu S, Kong Y (2014) The development of auditory skills in young children with mondini dysplasia after cochlear impantation. PONE 9(9):1-6.

10. Nair G, Vadivu S, Sampathkumar R, Pradhananga RB, Kameswaran M (2017) A study of anomalies of cochlea and cochlear nerve in children with congenital profound hearing loss - an indian perspective. JSRR 13(1): 1-11.

11. Dettman S, Barzalighi AS, Ambett R, Dowell R, Trotter M (2011) Cochlear implants in forty-eight children with cochlear and/or vestibular abnormality. Audiol Neurotol 16(4): 222-232.

12. Jeong SW, Kim LS (2015) A new classification of cochleovestibular malformations and implications for predicting speech perception ability after cochlear implantation. Audiol Neurotol 20(2): 90-101.

13. Vincenti V, Ormitti F, Ventura E, Guida M, Piccinini A (2014) Cochlear implantation in children with cochlear nerve deficiency. Int J pediatric otorhinolaryngol 78(6): 912-917.

14. Wu CM, Lee LA, Chen CK, Chan KC, Tsou YT et al (2014) Impact of cochlear nerve deficiency determined using 3-dimensional magnetic resonance imaging on hearing outcome in children with cochlear implants. Otol Neurotol 36(1): 14-21.