OUTCOMES OF COCHLEAR IMPLANTATION AMONG PRELINGUAL CHILDREN WITH DIFFERENT AGE AT IMPLANTATION

BY

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ABSTRACT

Cochlear Implant (CI) is an implanted electronic device designed to produce useful hearing sensations to a person with severe to profound sensorineural hearing loss (SNHL). Early implantation is recommended for prelingual children that receive minimal benefits from the super-power hearing aid. Late of cochlear implantation among prelingual children is often associated with poor outcome and prognosis post-operation due to brain plasticity. Thus, the aims of this study were to compare CI outcome among pre-lingual children with different ages at implantation, and to investigate the correlation of relevant intrinsic and extrinsic factors to the CI outcome. A total of 30 pre-lingual recipients of CI from the University Malaya Medical Centre (UMMC) were divided into two groups, based on their age at implantations; i) early implantation (≤4 years old, n=12) and ii) late implantation (> 4 years old, n=18).The study participants were implanted with MED-EL (n=29) and Oticon Medical (n=1) CIs. The ages at implantation for the early group were between 1.4 and 4 years old (mean 3.2, SD 0.8), and for the late group the ages were between 4.3 and 14.7 years old (mean 6.8, SD 2.9). Five tests were used to evaluate the outcome measures: i) aided thresholds ii) Meaningful Auditory Integration Scale (MAIS), iii) Meaningful Use of Speech Scale (MUSS), iv) Speech Tests, and v) CAEP test. The outcomes of the early and late implanted groups were statistically compared. The results showed improved auditory detection and higher scores from parental questionnaires post-implantation in both groups. The early implanted group was found to have better outcomes in terms of audiological assessments and shorter CAEP P1 latency compared to the late implanted group. However similar improvement gains in parental self-report questionnaires were found in both groups. The intrinsic factors that affecting the CI outcomes were the age of subjects, the duration of implantation and the age at implantation. Whereas the extrinsic factors were the maternal educational and occupational level. In conclusion, both early and late implantations resulted in improved audiological assessments and parental self-report scores post-implantation, with the early implanted group showing rapid brain maturation than the late implanted group. This study recommends early implantation to be prioritised, as it provides better outcome. However, if it is not feasible to implant early, consideration with realistic expectation should be given to implant slightly older children, as they also derive benefits from the CI as observed in this thesis.

خلاصة البحث

إن الزرع القوقعي (CI) جهاز إلكتروني يزرع في الأذن، مصمم لإنتاج أحاسيس سمعية مفيدة لشخص يعاني من فقد السمع العصبي الحسي (SNHL) الشديد إلى العميق. يوصى بالزرع المبكر للأطفال في مرحلة قبل اكتساب اللغة ليحصلوا على الحد الأدنى من الفوائد من المعينات السمعية فائقة القوة. غالبا يرتبط التأخر في زراعة القوقعة لديهم بالنتائج السيئة والتشخيص بعد الجراحة بسبب لدونة الدماغ. وبالتالي، إن أهداف هذه الدراسة هي المقارنة النتائج CI لدى الأطفال في مرحلة قبل اكتساب اللغة من أعمار مختلفة عند الزرع، والتحقيق في ارتباط العوامل الداخلية والخارجية ذات الصلة بنتائج CI. تم تقسيم 30 متلقي CI لدى الأطفال في مرحلة قبل اكتساب اللغة من المركز الطبي بجامعة مالايا (UMMC) إلى المجموعتين، بناء على أعمارهم عند الزرع؛ ا) الزرع المبكر (≤4 سنوات، n = 12)، و ب) الزرع المتأخر (> 4 سنوات، n = 8). وتم زرع المواضع بالماركةMED-EL (n=29، 96.7%) و Oticon Medical (n=1، 3.3%). الأعمار عند الزرع لمجموعة الزرع المبكر تتراوح بين 1.4 و 4 سنوات (متوسط 3.2، 0.8 SD)، و لمجموعة الزرع المتأخر أعمارهم ما بين 4.3 و 14.7 سنة (متوسط 6.8، SD 2.9). استخدمت خمسة اختبارات لتقييم مقاييس النتائج: ا) عتبات المساعدة، ب) مقياس التكامل السمعي الهادف (MAIS)، ج) الاستخدام الهادف لمقياس الكلام (MUSS)، د) اختبارات الكلام، و هـ) اختبار CAEP. تمت مقارنة نتائج بين مجموعتين الزرع المبكر والمتأخر إحصائيا. أظهرت النتائج تحسن الكشف السمعي ودرجات أعلى من استبيانات الوالدين بعد الزرع في كلا المجموعتين. وجدت أن مجموعة الزرع المبكر الحصول على نتائج أفضل من حيث التقييمات السمعية وزمن استجابة CAEP P1 أقصر مقارنة بمجموعة الزرع المتأخر. ومع ذلك، تم العثور على مكاسب تحسن مماثلة في استبيانات التقرير الذاتي للوالدين في كلا المجموعتين. العوامل الداخلية التي تؤثر على النتائج هي أعمار المواضع أثناء الاختبار، و مدة الزرع، والعمر عند الزرع. في حين أن العوامل الخارجية هي المستوى التعليمي والمهني للأم. في الختام، أدت عمليات الزرع المبكر والمتأخر إلى تحسين التقييمات السمعية ودرجات التقرير الذاتي للوالدين بعد الزرع، حيث أظهرت مجموعة الزرع المبكر نضجا سريعا للدماغ مقارنة بمجموعة الزرع المتأخر. يوصى هذه الدراسة بإعطاء الأولوية للزرع المبكر لأنه يوفر نتائج بصورة أفضل. ومع ذلك، إذا لم يكن من الممكن الزرع مبكرا، فيجب مراعاة الواقع لزراعة الأطفال الأكبر سنا قليلا، حيث أنهم يستمدون أيضا فوائد من CI كما هو مذكور في هذه الأطروحة.

APPROVAL PAGE

I certify that I have supervised and read this study and that in my opinion, it conforms to acceptable standards of scholarly presentation and is fully adequate, in scope and quality, as a thesis for the degree of Master of Allied Health Sciences (Audiology).

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Assoc. Prof Dr Ahmad Aidil Arafat Bin Dzulkarnain
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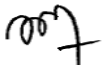

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DECLARATION

I hereby declare that this thesis is the result of my own investigations, except where otherwise stated. I also declare that it has not been previously or concurrently submitted as a whole for any other degrees at IIUM or other institutions.

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This thesis is dedicated to all my children, Ainul Mardhyyah, Eusoff, Hurun Ain, Hanis Adneen and Husna Naim and my beloved husband, Jeffry for being patience with me for the past 4.5 years part time study's journey.

For my nephew, Luqman and all hearing-impaired children,

you are such an inspiration to me.

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LIST OF SYMBOLS

<	Smaller than
>	Larger than
%	Percent
=	Equal to
:	Ratio to
R	Correlation coefficient
≥	Larger or equal than
≤	Smaller or equal than
ms	Milliseconds
dB	Decibels
n	Sample size
p	Significance value
d	Cohen’s d
g	Normalised gain
SD	Standard deviation

LIST OF ABBREVIATIONS

ABR	Auditory Brainstem Response
AEP	Auditory Evoked Potential
AC	Auditory Cortex
dBHL	Decibel Hearing Level
dBnHL	Decibel in normal Hearing Level
dBSPL	Decibel in Sound Pressure Level
CAEP	Cortical Auditory Evoked Potential
CAP	Categorical Auditory Performance
CDI	Child Development Inventory
CI	Cochlear Implant
CNC	Cochlear Nuclear Complex
CANS	Central Auditory Nerve System
EARS	Evaluation of Auditory Responses
EEG	Electroencephalography
EHDI	Early Hearing Detection and Intervention
EOG	Electrooculography
FDA	Food and Drug Administration
FSP	Fine Structure Processing
HA	Hearing Aid
HDCIS	High Definition Continuous Interleaved Sampling
HRHS	High Risk Hearing Screening
HRQOL	Health- Related Quality of Life
IC	Inferior Colliculus
IHS	Intelligent Hearing System
IIUM	International Islamic University Malaysia
IREC	IIUM Research Ethics Committee
IT-MAIS	Infant-Toddler Meaningful Auditory Integration Scale
LiP	Listening Profile
LLR	Late Latency Response
MAIS	Meaningful Auditory Integration Scale
MED-EL	Medical Electronics Corporation
MGB	Medial Genus Body
MLR	Middle Latency Response
MOH	Ministry of Health
MTP	Monosyllabic Trochee Polysyllabic
MPIS	Main Peak Interleaved Sampling
MUSS	Meaningful Use of Speech Scale
NLL	Nuclei of the Lateral Lemniscus
PET	Positron Emission Tomography
PTA	Pure Tone Audiometry
SDT	Self Determination Theory
SES	Size Economy Status
SLM	Sound Level Meter
SNHL	Sensorineural Hearing Loss
SNR	Signal To Noise Ratio

SPSS	Statistical Package for the Social Sciences
SOC	Superior Olivary Complex
UMMC	University Malaya Medical Centre
UMREC	University Malaya Research Ethics Committee
UNHS	Universal Newborn Hearing Screening
WHO	World Health Organization

CHAPTER ONE

INTRODUCTION

1.1 INTRODUCTION

According to the WHO (2018), an estimated 466 million people worldwide experience hearing loss. Seven percent from this population are children ranging from the ages of 0 to 14 years old and represent 34 million patients. A recent systematic review shows that approximately 2 per 1000 neonates worldwide were identified with neonatal permanent hearing loss (Bussé et al., 2020). This permanent hearing loss among children would lead to three major issues as follows; i) functional effect on speech development and inability to communicate with others ii) socio-emotional impact that makes the child susceptible to loneliness, insecurity, and dissatisfaction iii) economic impact related to health sector costs, educational support, and societal costs (Yoshinaga et al., 1998; Hyde, Punch, & Grimbeek, 2011). The severity of the hearing loss makes the impact of the hearing loss worse. This adverse effect may influence not only the children but also their family (Zaidman-Zait et al., 2016).

One of the best technologies to help patients with severe to profound hearing loss children is through the Cochlear Implant (FDA, 2018). However, intervention through the Cochlear Implant (CI) should start as soon as possible during the sensitive period so that the benefits of the device will be maximised (Sharma et al.,2005; Yoshinaga et al., 2018). To achieve this target, the Universal Newborn Hearing Screening (UNHS) programme has been widely adopted throughout North America, Europe and in most other developed regions since 1994 (Patel et al., 2011). As a result, age at detection of hearing loss has been reported to reduce and children with

severe to profound hearing loss can be implanted before they reach the age of 9 months to achieve the goal of normal speech development (Karltorp et al., 2020). Many studies have demonstrated that speech development and language acquisition in children implanted under the age of 12 months is very similar to normal hearing children (Dettman et al., 2007; Miyamoto et al.,2008; Roland et al., 2009). However, for some countries that are unable to implement the UNHS program, implantation is typically conducted at a later stage in life and this may affect the outcome of the implantation. Thus, this study aims to assess the outcome among late implanted pre-lingual children with a broader focus rather than assessing only on the aspects of oral communication (receptive and language skills). This study measures the outcome of children that received early and late implantation, using different aspects of outcome measures apart from standard measures used clinically in Malaysia, including the cortical auditory evoked potential (CAEP) and from the parents' perspective such as Meaningful Auditory Integration Scale (MAIS) and the Meaningful Use of Speech Scale MUSS (Robbins, 1991). The terms such as CAEP, MAIS, MUSS and MTP will be discussed in Chapter 2 subtopic 2.4).

1.2 PROBLEM STATEMENT

1.2.1 Potential Age at Implantation Issue

In Malaysia, not all children with severe to profound hearing loss are being detected at an early age by the UNHS program as in other developed countries. This is due to the unavailability of the UNHS program at many government hospitals. As reported by Ministry of Health Malaysia (2018), only 16 government hospitals have the UNHS program while 22 hospitals are practising high risk hearing screening (HRHS). There

are many factors affecting the program such as manpower, facilities and other factors (Low et al., 2005; Majid, Zakaria, & Hamzah, 2017, Mazlan & Min (2018). The Malaysian Ministry of Health's (MOH) Cochlear Implant Programme reported that only 9.9 % or a total of 121 pre-lingual implantees children underwent the UNHS program (Md Yusoff, Umat, & Mukari, 2017). Based on these program findings, a majority of the cochlear implant patients did not undergo the UNHS (Mohd Hasim et al, 2016) and were only referred to audiology clinics when symptoms of delayed speech and language development appeared between the ages of 1 and 4 years old (pre-school age).

Late detection of pre-lingual hearing loss is also possible due the delay in confirming the diagnosis. As reported by Elizabeth et al. (2017), late confirmation and intervention may be experienced by a significant number of children referred for early assessment. A few explanations were suggested for this longer duration of confirmation, such as developmental and medical conditions, including middle ear disorders. Some children required more time to confirm the diagnosis due to inconclusive audiological results, missed or cancelled appointments, and family issues**.** In contrast, some patients were detected early but faced issues in gaining funding or had rehabilitation issues such as inconsistency of hearing aid (HA) usage and attending the speech therapy session (Karandikar & Valame, 2020). These were considered as barriers to achieving timely cochlear implantation for these children. By the time the child gets the funding for the CI operation, the child has already exceeded the sensitive period to get good prognosis from the CI operation (Kothari et al., 2015). As a result, the age at implantation will be later than 4 years old and this exceed the age recommendation for pre-lingual CI candidate by National Cochlear Implant Program (Medical Development Division, 2016). Because of the factors above and

despite the strong recommendation for early hearing detection and intervention (EHDI), there are still numerous pre-lingual children with severe to profound hearing loss that have been late diagnosed, which leads to late intervention including late implantation.

1.2.2 Potential Limitation of not using objective Outcome Measures among Late Implantation

The majority of the outcome measures among pre-lingual CI focused on the goal of EHDI which is to maximise the linguistic competency. Whilst synchronising the outcome measure with the EHDI goal is important, it may not be straightforward for those who had been implanted at a slightly later stage of their life. The language assessment tool is highly dependent on the language ability of the child before the period of implantation, which could have been severely impaired in those who received delayed amplification. Late implantation usually results in poor functional performance and poor predicted progress (Hanvey et al., 2017). Assessment that either uses language assessment tools or focuses only on oral communication may limit the overall conclusion on the potential benefits of CI that can be offered in the long term. This will result in a poor performance among the assessed late implantees. Therefore, an objective test such as cortical response evoked potential (CAEP) is suggested as one of the tools in measuring the outcomes of implantees. As the late implantation decision becomes a dilemma as the children's age has passed the sensitive period to acquire language (Kos et al., 2009; Karandikar & Valame, 2020), more study on the late implantees is needed to give some meaningful information to the decision for the operation and to meet parents' expectations.

1.3 OPERATIONAL DEFINITION

1.3.1 Cochlear Implant

According to the Food Drug Association (FDA, 2018), Cochlear Implant (CI) is an implanted electronic hearing device, designed to produce useful hearing sensations in a person with severe to profound nerve deafness by electrically stimulating nerves inside the inner ear. Patients who are unable to get maximum benefits from conventional superpower HA are recommended to have the cochlear implant. The cochlear implant comprises two parts namely internal and external parts. The internal part consists of a coil inserted in the cochlea through a particular approach by an otology surgeon. The external part is the processor of the device, which is programmed by an audiologist in the switch on and mapping session. The CI processor must be appropriately mapped based on the needs of the recipient (Vaerenberg et al., 2014).

1.3.2 Outcomes measures

The outcomes of amplification must involve their social skills change, language, speech production, and functional performance in everyday life, via a combination of direct assessments directed at the child and reporting tools based on the observations of parents and teachers (Ching et al., 2013). Further explanation of the outcome measures will be explained in subtopic 2.2.2 in Chapter 2 based on different aspects of outcome measures.

1.3.3 Factors Affecting the Outcomes

The outcomes of cochlear implants vary based on intrinsic and extrinsic factors. Intrinsic factors are embedded within the subject in terms of current age of patient, age at implantation, duration of implantation, and aetiology of the impairment. Extrinsic factors refer to external factors such as quality time spent by the parents with the implanted children, family economic status, maternal education, and occupational level. Details of the factors will be discussed in subtopic 2.3 in Chapter 2.

1.4 SIGNIFICANCE OF THE STUDY

The findings of this study are expected to be a source of reference for evaluating the outcomes between early and late implanted patients. The study will provide data on the outcomes of pre-lingual children locally that can assist the professionals in CI teams and families in the implantation decision once they exceed the sensitive period. The rationale of this study is to know whether the existing patient with CI that being late implanted shows benefit in auditory detection, parental observation, speech test and CAEP test. This provides future clinician whether they should implant or not patient that exceeds critical period and factors associated with the outcomes in each group.

1.5 RESEARCH QUESTION

This study aims to investigate the outcomes of CI among patients who received cochlear implantation at different ages. In particular, this study seeks to answer the following questions:

1. Is there any difference in audiological outcome measure between pre and post implantation in early and late implanted group?
2. Is there any difference in audiological outcome measure between in early and late implanted group?
3. What are the differences in the CAEP P1 latency between early and late implanted group and what are the intrinsic factors affecting it?
4. What is the correlation of the intrinsic factors with the outcomes (average aided difference, normalised MAIS, normalised MUSS and speech score MTP3, MTP6 and MTP12) in early and late implanted group?
5. What is the correlation of the extrinsic factors with the outcomes (average aided difference, normalised MAIS, normalised MUSS and speech score MTP3, MTP6 and MTP12) in early and late implanted group?

1.6 SPECIFIC OBJECTIVES

1. To evaluate within group outcomes of CI* in pre and post implantation for early implanted group and late implanted group.
2. To evaluate between group outcomes of CI** in early and late implanted groups.
3. To determine the correlation between the CAEP P1 latency with the intrinsic factors among prelingual implantees.
4. To determine the correlation of the outcomes*** with the intrinsic factors among prelingual implantees.