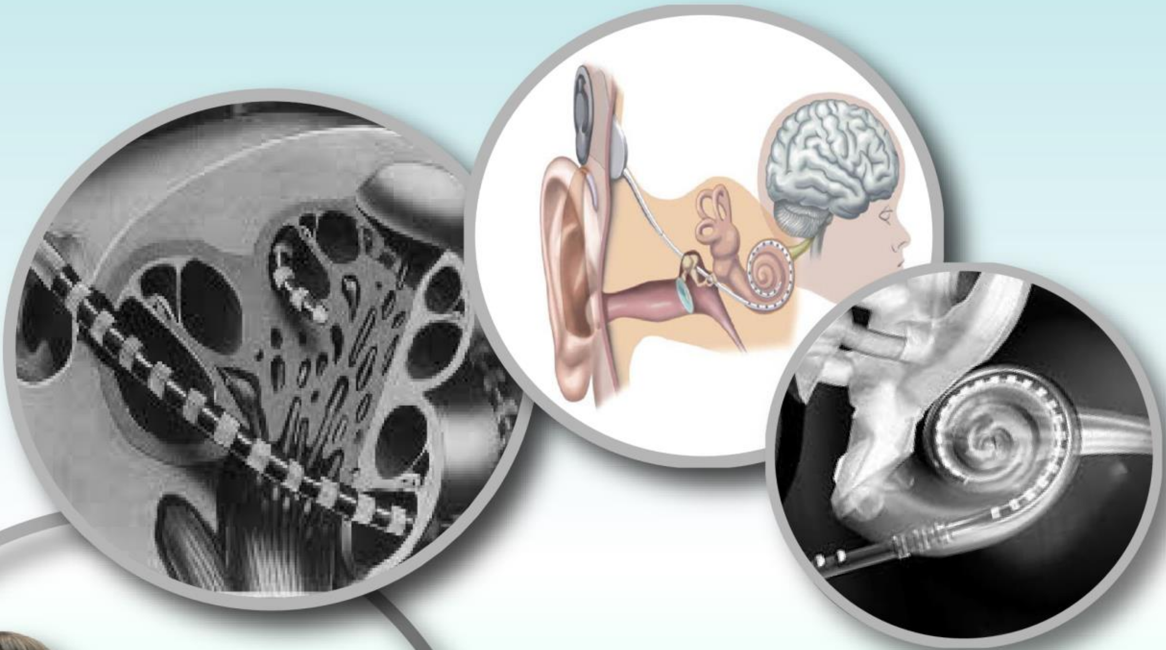
MINISTRY OF HEALTH MALAYSIA
NATIONAL MOH COCHLEAR IMPLANT PROGRAMME

10-YEAR REPORT
2008-2018



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Ministry of Health Malaysia

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March 2020
Medical Development Division, Ministry of Health Malaysia
Jointly published by:
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Data reported was acquired from the National ORL Registry – Hearing and Otology Related Disease/Cochlear Implant & data source providers (MOH facilities). Interpretation and reporting is the responsibility of the editors and do not reflect the official policy of the publisher or the authors affiliated institutions. Caution is advised before drawing conclusions from the data.

The electronic version of this report can be viewed at www.moh.gov.my
ISBN: 978-967-2173-91-5 / 978-967-2173-92-2 (ebook)

ISBN 978-967-2173-91-5
9 789672 173915 >

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ACKNOWLEDGEMENT

The National MOH Cochlear Implant Outcome Report Committee would like to express gratitude and appreciation to the Director General of Health MOH and the Director of Medical Development Division MOH for their support and approval to publish this 10-year report.

We are thankful to chairpersons of the National MOH Cochlear Implant Committee Dato' Dr Siti Sabzah binti Mohd Hashim and Dr Tengku Mohamed Izam bin Tengku Kamalden, and the multidisciplinary team members of the committee for their expert contributions to the programme. Special thanks to the radiology team headed by Dr Noryati binti Mohammad, pediatric team led by Dr Irene Cheah Guat Sim and Dr Chee Seok Chiong, occupational therapy team led by Pn Rokiah binti Alias, medical social officers led by Pn Noraini binti Mohd Zahid and Cik Nur Fatehah binti Mohd Shukri from the Special Education Service Center, Ministry of Education for their dedication and involvement to the programme.

We also thank following satellite hospitals for their commitment, dedication and hard work in managing cochlear implant recipients and timely effort of data collection of the CI recipients;

- Hospital Sultanah Bahiyah, Alor Setar
- Hospital Raja Perempuan Zainab II, Kota Bharu
- Hospital Sultan Ismail, Johor Bahru
- Hospital Sungai Buloh, Selangor
- Hospital Raja Permaisuri Bainun, Ipoh
- Hospital Tuanku Ja'afar, Seremban
- Hospital Queen Elizabeth, Kota Kinabalu
- Hospital Sultanah Nur Zahirah, Kuala Terengganu
- Hospital Kuala Lumpur
- Hospital Umum Sarawak, Kuching

We are grateful to Clinical Research Centre, Hospital Sultanah Bahiyah, Alor Setar and National ORL Registry (Hearing and Otology Related Disease and Cochlear Implant) for the continuous technical supports.

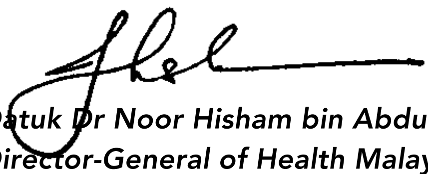
FOREWORD

FOREWORD
Director General Of Health Malaysia

The Ministry of Health Cochlear Implant team was formed in 2008 with the aim to start a cochlear implant programme within the framework of the Ministry of Health. The implementation of the programme over the last ten years, from 2008 to 2018, has been a considerable contribution to the nation, in which a total of 413 ears were implanted through this program. This huge undertaking which incur considerable cost implications has contributed to a tremendous amount of life changing benefit especially in aspects of hearing, speech, academic achievement, psychosocial and career advancements. The success of the programme has been made possible by the dedicated multi-disciplinary national MOH CI team.

Today, the program is firmly established and well into its 12th year. In the first five years report, 137 patients benefitted from the program and 244 more patients in the second five years. The results and outcomes in this report are based on data from the National ORL Registry and CI Outcome Report Committee data collection. This is indeed an important undertaking as data analysis is the most powerful mechanism to express performance. I am well pleased with the outcome, as majority of the recipients have been benefitting from the implants. This is made possible with meticulous candidacy selection, followed by intervention and intensive rehabilitation. Excellent family support and good collaboration with NGOs and related institutions are also instrumental to the success.

On behalf of the Ministry of Health, I would like to convey my sincere thanks and gratitude to all members of the National MOH Cochlear Implant Committee for the hard work and objectivity in running this successful program. I am confident the program will continue to flourish and progress in the coming years as more and more hearing disabled Malaysians will continue to experience this life changing benefit.



Datuk Dr Noor Hisham bin Abdullah
Director-General of Health Malaysia, MOH

FOREWORD
Deputy Director General Of Health Malaysia

Cochlear implantation programme is an expansion of Otorhinolaryngology (ORL) services provided by the Ministry of Health. This essential service first started in 2008 and has rapidly progressed and expanded, servicing from 7 satellite hospitals to 10 satellite hospitals within the Ministry of Health in the past 10 years.

The teamwork of Audiologists, Speech–Language Therapists and ORL surgeons with inter disciplinary collaboration from multiple professional groups has helped strengthen this programme and enabled the management to be more comprehensive.

This 10 year MOH CI Outcome Report shall serve as a benchmark and guiding principle for future improvement and progress. My thanks and gratitude to the various medical fraternities such as pediatric, radiology, occupational therapy, child development specialists, child psychiatrists, social medical workers, educators and others who have contributed in the management and intervention. To meet this specific specialty demands, I ensure the ministry will continue to support the needs and requirement. I would like to congratulate the Medical Development Division and the National MOH CI Outcome Report Committee for initiating and coordinating this effort. I hope that the quality of our medical services will continue to improve in tandem with the Ministry's mission to provide the highest level of care for our nation.

There is no greater disability in society, than the inability to see a person as more. And therefore, our purpose is to help the hearing disabled lead a productive life.

Congratulations and thanks again for a job well done.

Datuk Dr Hj. Rohaizat bin Hj. Yon
Deputy Director-General of Health Malaysia, MOH

FOREWORD
Director of Medical Development Division

According to WHO, 466 million people worldwide have disabling hearing loss (6.1% of the world's population) in 2018 and will increase to over 900 million people by 2050. However, the disability from the hearing impairment can be reduced by early detection and intervention.

Cochlear implant, a surgically implanted electronic device, has been accepted worldwide as one of the treatment for hearing impaired patients who obtain no benefit from hearing aids. Approximately 600,000 people worldwide have received the device with recipients including both adults and children.

Considerable time, effort and costs were involved in commencing and sustaining the Ministry of Health Cochlear Implant Programme since its inception in 2008. The collaboration between the ORL fraternity and Medical Development Division has enabled smooth and rapid implementation of related guidelines and service policies, subsequently a publication of Cochlear Implant Service Operational Policy in July 2009. Development of technology advancement in cochlear implant, along with experiences globally and locally, the second edition of Cochlear Implant Service Operational Policy was published in November 2017.

An audit of this long term programme is no easy task but it is however necessary. This 10 year report of the National MOH CI Outcome provides an in depth overview of the success, strengths and challenges of the programme.

I am pleased with the outcome of this report. The benefits of hearing to the individuals, families and societies are priceless, despite its high costs. The Medical Development Division will continue to facilitate and support the National MOH CI programme. Every hearing impaired individual in our community must never be marginalized and should be given the opportunity to integrate into mainstream society.

Dato' Dr Norhizan bin Ismail
Director Medical Development Division, Ministry of Health

FOREWORD
Head of Otorhinolaryngology Services, MOH

The National Cochlear Implant Committee Ministry of Health in collaboration with The National ORL Registry (Hearing and Otology Related Disease/Cochlear Implant) and Clinical Research Centre is honored to present this 10 year National Cochlear Implant Outcome Report. This report is instrumental for future management and planning of MOH CI related services.

I would like to congratulate chairman of The National MOH Cochlear Implant Outcome Report Committee, Dr Zulkiflee bin Salahuddin and project coordinator, Dr Hafeza binti Ahmad, as well as all CI Outcome Report Committee members. It is indeed a bold step as very few centers worldwide could produce an Outcome Report on cochlear implantation at a national level. I hope that all multidisciplinary team members of the MOH Cochlear Implant at national and satellite level together with the ORL fraternity will continue to strive and maintain high standards of evidence based reports and scientific papers in the future.

A special thank and gratitude to Director of the Medical Development Division and to Head of the CRC Hospital Sultanah Bahiyah and their staff for financial and technical supports.

Dato' Dr Siti Sabzah binti Mohd Hashim
Head of Otorhinolaryngology Services, MOH
Advisor, National MOH Cochlear Implant Programme

FOREWORD
Chairman, National MOH Cochlear Implant Outcome Report Committee

We are honoured and privileged to present this 10 year (2008-2018), Ministry of Health Cochlear Implant Outcome Report, which follows our first 5 year report 2008-2013.

In this report we have expanded our analysis by inclusion of extra functional outcomes in the second cohort of 5 years of our program beside the previous analyzed outcomes.

My sincere gratitude to the outcome report team for their tireless effort and hard work spearheaded by Dr Hafeza Ahmad This was indeed a lengthy and time-consuming process requiring sheer commitment and cohesive teamwork. These include tedious process of data collection, literature review, data cleaning, data analysis and report writing. This huge undertaking could not be accomplished without the constant support and guidance by our Head of Otorhinolaryngology Services, Dato' Dr Siti Sabzah binti Mohd Hashim.

We extend our gratitude to the Director of the Medical Development Division, the Head of the CRC Hospital Sultanah Bahiyah and their staff for their continuous support in preparing this outcome report.

We hope this 10 year report will provide further insight into the MOH National Cochlear Implant Programme which reflects its successful achievements and outcomes. Additionally, we wish the recommendations in this report will strengthen the programme and will assist future planning to improve the health services related to cochlear implant recipients and hearing impaired population at large in our beloved country.

Dr Zulkiflee bin Salahuddin
Deputy Head of Otorhinolaryngology Services, MOH
Chairman, National MOH Cochlear Implant Outcome Report Committee

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- Hospital Queen Elizabeth, Kota Kinabalu, Sabah
- Hospital Kuala Lumpur, Wilayah Persekutuan Kuala Lumpur
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Clinical Research Centre, Hospital Sultanah Bahiyah

- Dato' Dr Muhammad Radzi bin Abu Hassan
- Mrs Ibtisam binti Ismail

ABBREVIATIONS

3FA	Three Frequency Average
3PK	Pusat Perkhidmatan Pendidikan Khas
ABR	Auditory Brainstem Response
APHAB	Abbreviated Profile of Hearing Aid Benefit
ASSR	Auditory Steady State Response
AAC	Augmentative And Alternative Communication
AVT	Auditory Verbal Therapy
CAPII	Categories of Auditory Performance II
CI	Cochlear Implant
CLF	Current Level of Function
COM	Chronic Otitis Media
CRC	Clinical Research Center
dB	Decibel
dBHL	Decibel Hearing Level
EARs	Evaluation of Auditory Response to Speech
ECAP	Electrical Compound Action Potential Test
FPR	Family Participation Rating
HA	Hearing Aid
HL	Hearing Loss
HRNHS	High Risk Newborn Hearing Screening
HINT	Hearing in Noise Test
HRCT	High Resolution Computer Tomography
IP-I	Incomplete Partition Type I
IP-II	Incomplete Partition Type II
LiP	Listening Progress Profile
LVA	Large Vestibular Aqueduct
MAIS	Meaningful Auditory Integration Scale
MEE	Middle Ear Effusion
MOE	Ministry of Education
MOH	Ministry of Health
MRI	Magnetic Resonance Imaging
MUSS	Meaningful Use of Speech Scale
NCIQ	Nijmegen Cochlear Implant Questionnaire
NHEC	National Ear and Hearing Care
NHS	Newborn Hearing Screening
NIHL	Noise Induced Hearing Loss
NORL	National Otorhinolaryngology Registry
ECAP	Electrical Compound Action Potential Test
ORL	Otorhinolaryngology
PEACH	Parent’s Evaluation of Aural/Oral Performance of Children

PTA	Pure Tone Audiometry
RWM	Round Window Membrane
SNHL	Sensorineural Hearing Loss
SIR	Speech Intelligibility Rating
SSI	Surgical Site Infection
UNHS	Universal Newborn Hearing Screening

GLOSSARY

ACQUIRED HEARING LOSS

The loss of hearing that occurs or develops some time during a person's life but was not present at birth.

BEHAVIOURAL ISSUE

Behaviour that is a source of concern or undesirable that may impede the course of rehabilitation process.

COHORT

A cohort study is a particular form of longitudinal study that samples a cohort, performing a cross-section at intervals through time. It is a type of panel study where the individuals in the panel share a common characteristic.

CONGENITAL HEARING LOSS

Congenital hearing loss is a hearing loss present at birth.

DEVICE FAILURE

A complete and irreversible failure of the implanted receiver-stimulator of the cochlear implant

DEVICE REJECTION

Complication due to body reactions towards implanted device.

FACIAL NERVE PARESIS/PALSY

Facial nerve palsy is a neurological condition in which function of the facial nerve is partially or completely lost.

FAMILY PARTICIPATION RATING (FPR)

Scale of rating of families or caregiver participation and commitment in the therapeutic process of prelingual cochlear implant recipients.

ELECTRODE MIGRATION

Electrode migration is displacement of cochlear implant electrode array out of the cochlea.

EXPLANTATION

A surgical procedure removing implanted cochlear implant.

HEARING THRESHOLD

Minimum sound level of a pure tone that an individual can hear with no other sound present.

INTRAOPERATIVE

Occurring or performed during the course of a surgical operation.

MAPPING

"MAPping" is process of getting the CI user a specific listening program (also known as MAP) usually consist of minimum levels of audibility, maximum levels of comfortability, programming strategy and other associated programming options.

MIDDLE EAR EFFUSION

A middle ear effusion occurs when the middle ear cavity is filled up by effusion.

NEWBORN HEARING SCREENING

An objective screening method performed to identify neonates who may have hearing loss and who may need follow up or more in depth testing.

OUTCOME MEASURES

Outcome measures are commonly used to determine the impact of an intervention on an individual's life.

PERILYMPH GUSHER

Perilymph gusher is a condition when perilymph fluid profusely flows out after cochleostomy or round window membrane opening during cochlear implant surgery.

POST-LINGUAL DEAFNESS

Post-lingual deafness is deafness or hearing loss develops after the acquisition of speech and language.

PRELINGUAL DEAFNESS

A prelingual deafness is deafness or hearing loss develops before the acquisition of speech and language.

(RE)HABILITATION

Providing different types of therapies to patients who have deafness, and implementing different amplification devices to aid the client's hearing abilities. Aural rehab includes specific procedures in which each therapy and amplification device has as its goal the habilitation or rehabilitation of persons to overcome the handicap (disability) caused by a hearing impairment or deafness.

SENSORINEURAL HEARING LOSS (SNHL)

Sensorineural hearing loss is a type of hearing loss, or deafness, in which the root cause lies in the inner ear (cochlea and associated structures), vestibulocochlear nerve (cranial nerve VIII), or central auditory processing centers of the brain.

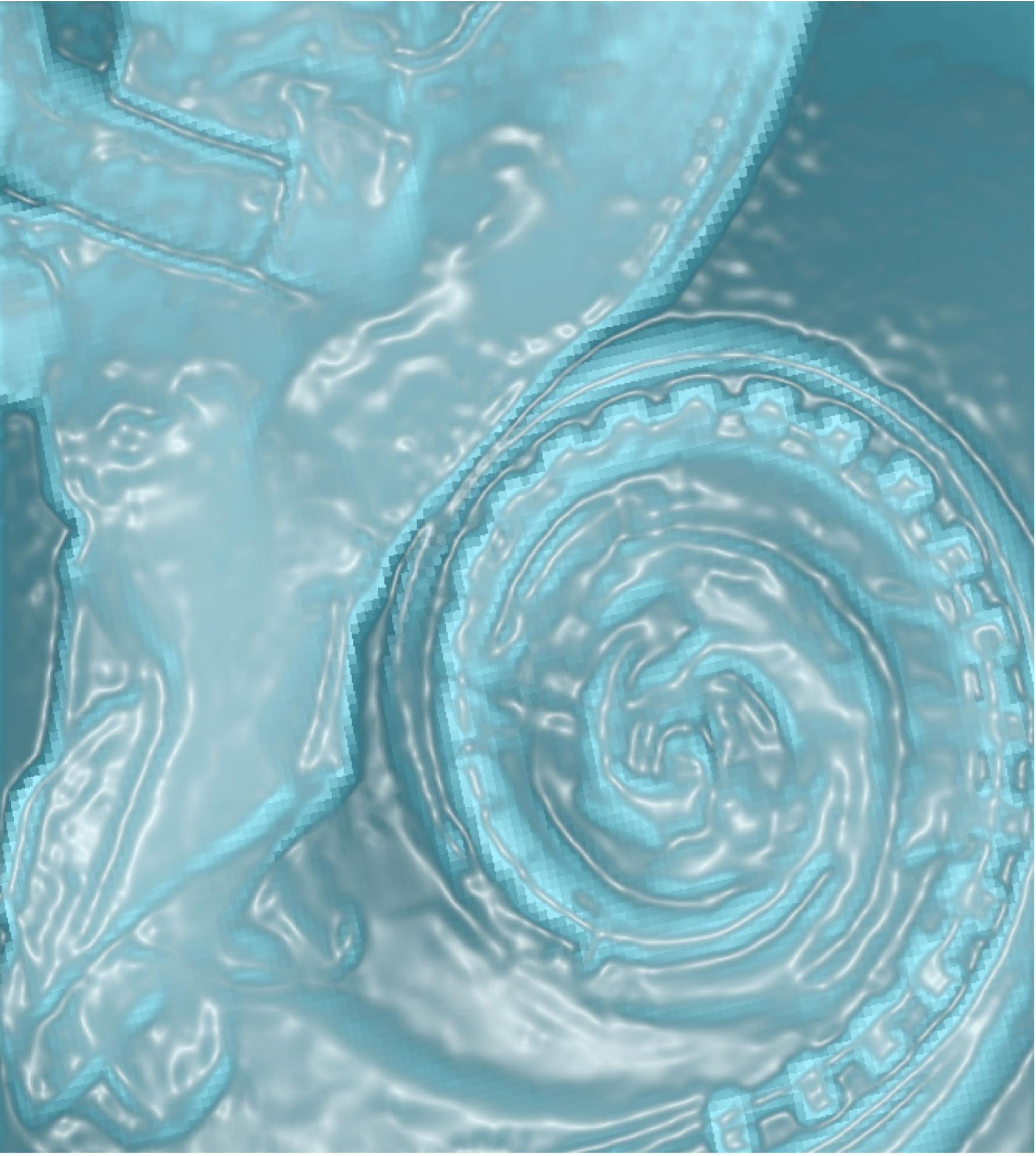
SURGICAL SITE INFECTION (SSI)

A surgical site infection is an infection that occurs after surgery at the site where the surgery took place.

SWITCH ON

"Switch on" is a process whereby the user's cochlear implant is first MAPped (activated) post-operatively. This usually occurs in two to three weeks post-operation.

LIST OF TABLES AND FIGURES



INTRODUCTION
Chapter 1

INTRODUCTION

The first commercial multichannel cochlear implants (CI) were produced in the 1980's. The device was initially used for deaf adults and eventually was successfully used on children. One of the truly amazing aspects of this technology was the outcome in congenitally deaf children; which was the ability and speak and even fit into mainstream education. The challenges to this success were the huge costs in the device, the time required for habilitation and the unpredictability of the outcome. However cumulative experience over the years have enabled professionals to formulate guidelines on candidate selection and expected outcomes.

Cochlear implantation has been accepted as one of the treatment of choice in patients with severe to profound hearing loss with no or minimal benefit from hearing aids (HA). Based on Food and Drug Administration (FDA) information, approximately 600,000 people worldwide have received the device with recipients including both adults and children[1].

The Ministry of Health Malaysia officially started its own cochlear implant programme in the year 2008. A central committee was formed by the Medical Development Division, MOH and comprised of multidisciplinary professionals from MOH, representatives from Ministry of Education and advisors from the local universities. At the inception of the programme, seven hospitals were selected to begin this service. This included Hospital Sungai Buloh, Hospital Sultanah Bahiyah, Hospital Raja Permaisuri Bainun, Hospital Tuanku Ja'afar, Hospital Sultan Ismail, Hospital Raja Perempuan Zainab II, and Hospital Queen Elizabeth. Hospital Kuala Lumpur was later added as one of the satellite hospitals in 2009 followed by Hospital Umum Sarawak in 2016. In addition to the above hospitals, Hospital Sultanah Nur Zahirah, Kuala Terengganu was also designated as a cochlear implant rehabilitation center for the east coast.

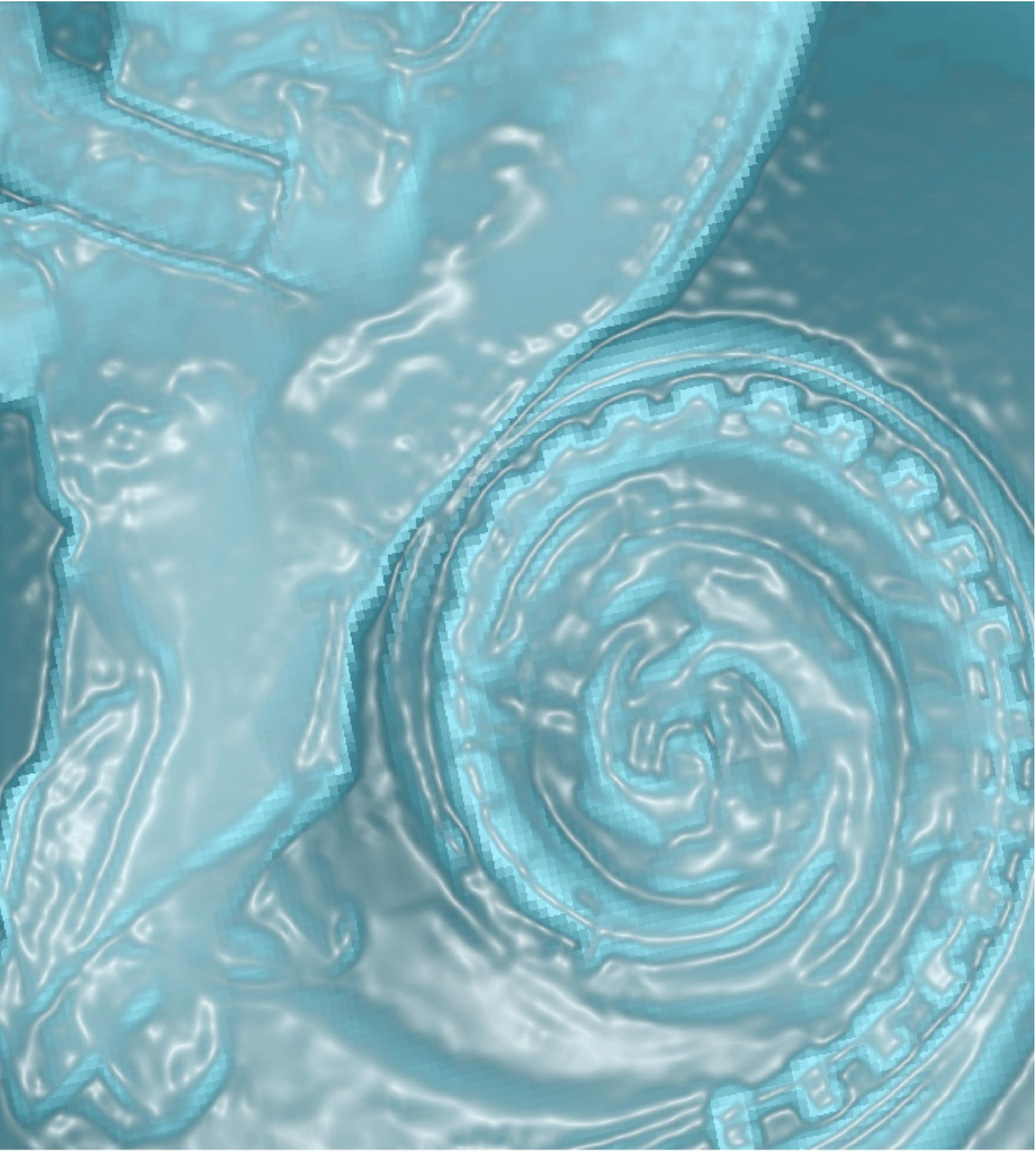
Adequate staffing and necessary medical equipment for cochlear implant surgery and rehabilitation was first addressed to start the service. Staffing requirements included trained ORL surgeons in CI surgery, radiologists, pediatricians, audiologists, speech language therapists, occupational therapists and medical welfare officers. Essential surgical and (re)habilitation equipment were acquired for the satellite hospitals. These include high end operating microscopes, surgical instrument sets, mastoid drills, facial nerve monitors, electrophysiological diagnostic set, hearing aid analyzer and programmer, speech language assessment and stimulation tools.

A Cochlear Implant Service Operational Policy (SOP) was also developed to ensure high professional and ethical standard of practice is achieved in the Ministry of Health hospitals. The first edition of the CI SOP was published in 2009 and the latest edition was published in 2017.

Candidate selection was done through a meticulous multidisciplinary assessment. Primary candidacy evaluations were done at the satellite hospitals with final approval during a centralized MOH National Cochlear Implant Committee Meeting held at regular intervals. Candidacy selection is based on criteria set in the CI SOP. Majority of the candidates were provided with a cochlear implant from an annual grant allocated by the Ministry of Health, Malaysia. The remaining candidates were either funded by other government agencies, zakat, external sources or self-funding[2,3].

Surgeries were performed by trained cochlear implant surgeons from the satellite hospitals in the presence of senior surgeon. Standard surgical techniques of facial recess approach via post auricular skin incision were applied in majority of cases. Mainly, the electrodes were introduced either through cochleostomy or round window membrane (RWM) insertion technique. Once the electrodes were secured in the cochlea, Electrical Compound Action Potential Test (ECAP) and impedance tests were measured by audiologists while the patients were still under general anesthesia. These ECAP provided useful objective information on electrode placement, baseline information for switch on and planning of future mapping session. Switch on was usually scheduled up to two to three weeks post operatively and followed by regular mapping sessions.

Following switch on, cochlear implant recipients were assigned to undergo long term intensive (re)habilitation in audiology, speech language and occupational therapy. Their progress and outcome assessments were monitored and measured at several intervals by cochlear implant team in the satellite hospitals. All data and information of the progress and outcomes were presented to MOH National CI Committee during progress meeting. These data were collected and reported to National CI Outcome Report Committee.



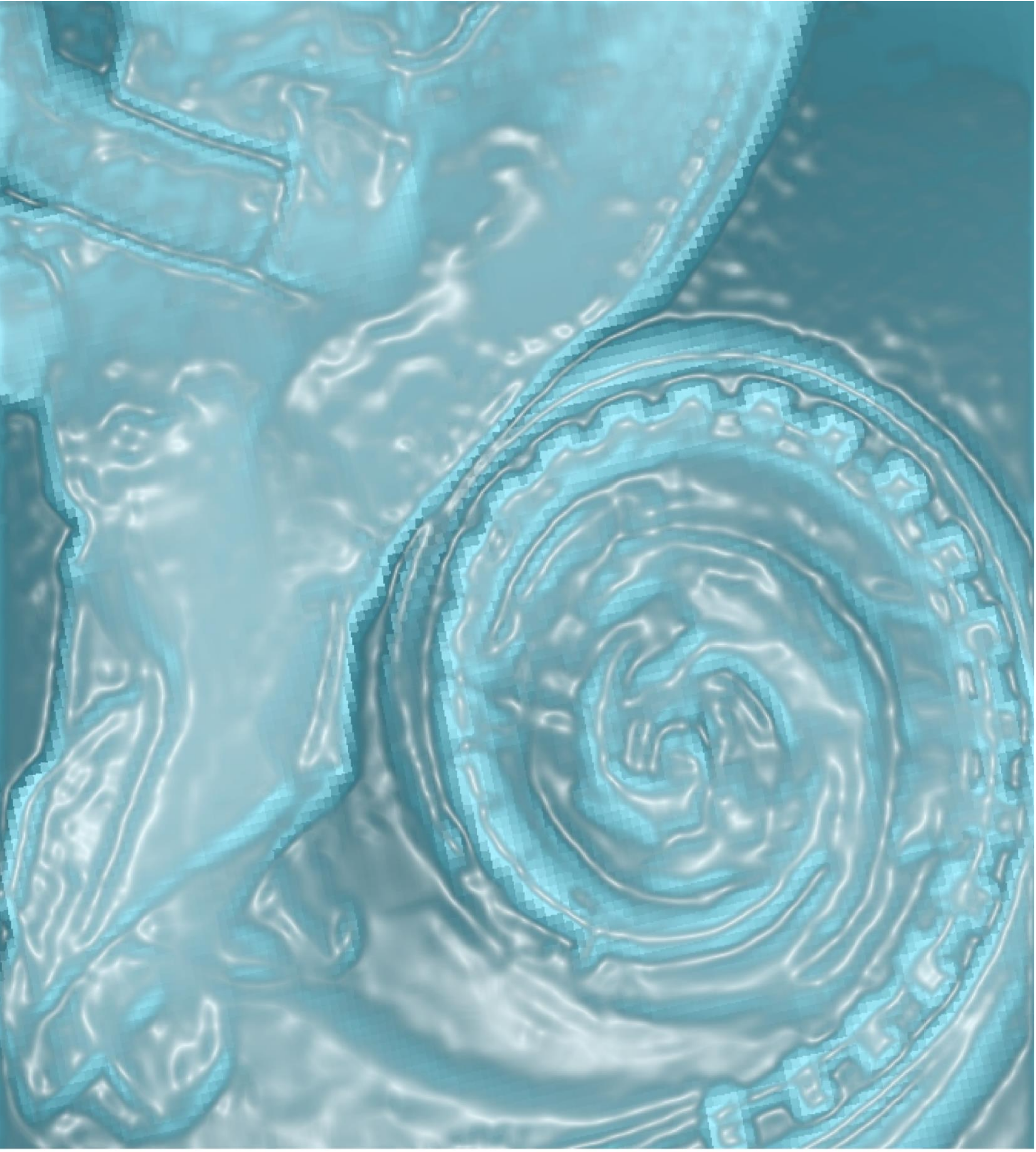
OBJECTIVES
Chapter 2

OBJECTIVES

This MOH National Cochlear Implant 10 year report is aimed as a report to evaluate the success of the program and to identify areas that may require further attention and improvement.

The objectives are:

- To describe the demographic and socioeconomic background of cochlear implant recipients.
- To identify the etiology and associated medical conditions with risks of hearing loss among the cochlear implant recipients.
- To verify the surgical complications outcomes and the safety of cochlear implant surgery.
- To measure the functional outcomes.
- To determine factors that may have effect on surgical and functional outcomes.
- To describe development and changes between the first cohort (2008-2012) and the second cohort (2013-2018).
- To identify the educational status in cochlear implant recipients.
- To evaluate on the quality of life among post-lingual cochlear implant recipients.
- To formulate recommendations in strengthening and improving the National MOH CI Programme.



MATERIAL & METHODOLOGY

Chapter 3

MATERIAL AND METHODOLOGY

A. DATA COLLECTION

Patients enrolled in this retrospective cohort review were all cochlear implant recipients under the MOH Cochlear Implant Programme from 2008 to 2018. Data was collected from the satellite hospitals and the National ORL Registry (Hearing and Otology Related Disease/Cochlear Implant). Post implantation outcomes data was collected until 30 April 2019.

The following data of individual patient were collected, recorded and coded into Excel Microsoft;

- Demographic information
- Clinical characteristics
- Possible etiologies of hearing loss
- Radiology imaging of the inner ear structures, cochlear nerve and brain form high resolution computer tomography (HRCT) and 3D magnetic resonance imaging (MRI)
- Age at diagnosis, age at implantation and age at switch on
- Surgical complications in cochlear implantation
- Audiological data at baseline and at defined follow up intervals after implantation
- Speech language data at baseline and at defined follow up intervals after implantation
- Communication mode post implantation
- Educational status after implantation for cochlear implant recipients
- Quality of life after implantation for post-lingual cochlear implant recipients

In this review, we collected and analysed parameters that were available for 10 years. We also compared parameters that were available for comparison between the first five years (2008-2012)[4] and the second five years (2013-2018). In the second cohort, few additional parameters were added to the database and used for the second cohort analysis only.

B. DEMOGRAPHIC AND CLINICAL CHARACTERISTICS

All cochlear implant recipients under the MOH National Cochlear Implant Programme from 2008-2018 were included in this review. There were categorized into two main groups, prelingual and post-lingual groups. Prelingual group is defined as children who have hearing loss before acquisition of speech and language. Post-lingual group is defined as patients who have hearing loss after acquisition of speech and language. Demographic information and clinical data were analysed descriptively for both categories.

C. OUTCOME MEASURES

Outcomes measured in this review were classified into four main domains namely;

I. Surgical and device related complications
II. Functional outcomes
III. Educational status
IV. Quality of life (QoL)

I. Surgical and Device Related Complications

Surgical complications were defined as an adverse event related to the procedure itself or additional morbidity to a patient. In this review, the surgical complications were classified as major complication or minor complication [5].

Major complications were defined when a patient developed significant medical sequelae such as meningitis and intractable vertigo following surgery, permanent disability of any degree such as complete facial nerve palsy or any other adverse event leading to another surgical procedure either due to patient factors or device related factors such as foreign body reactions and device migration.

Minor complications were defined as any other complications not fulfilling the major complications criteria. These included wound infection or delayed wound healing not requiring surgical intervention, transient facial paresis and transient vertigo[5].

II. Functional Outcomes

a) *Post Implantation Aided Thresholds*

The aided threshold is a valuable tool to assess the improvement in hearing performance of amplification and cochlear implant used. It is widely applicable not only for adult patients but also for pediatric population as it offers behavioral measures [6]. In this review, the aided performance of four frequencies average, 500Hz, 1000Hz, 2000Hz and 4000Hz were measured. The average aided thresholds were categorized into two groups; below and equal to 40dBHL (≤40dBHL), and above 40dBHL (>40dBHL). 40dBHL was taken as a cut off point for the aided threshold as studies had shown that with aided thresholds ≤40dBHL, children were able to hear at least 50% of what is being said[7].

b) Categorical Auditory Performances II (CAPII)

Categorical Auditory Performances II (CAPII) scale is a globally accepted scale to measure auditory perception outcome performance comprising of a nonlinear hierarchical scale of auditory receptive abilities. The lowest level describes no awareness of environmental sounds, and the highest level is represented by the ability to use the telephone with a known speaker[8–10].CAPII (Appendix 1) was used to measure auditory perception outcome in this review and were measured prior to implant as a baseline level and the progress were monitored at 6, 12, 24, 36 and 48 months post implantation.

c) Speech Intelligibility Rating (SIR) Scale

Speech production was measured using Speech Intelligibility Rating (SIR) Scale. We used the SIR (Appendix 2) to measure the outcome of cochlear implantation with respect to speech intelligibility recognizable by the listener. It is a 5-point rating scale ranging from 'pre-recognizable words in spoken language' to 'connected speech is intelligible to all listeners'[11,12]. In this review the SIR were measured prior to implant as a baseline level and the progress were monitored at 6, 12, 24, 36 and 48 months post implantation.

d) Meaningful Auditory Integration Scale (MAIS)

MAIS is parent's report scale of 10 questions to assess the functional ability of a child following the fitting of hearing technologies. The questionnaire includes information about the child's habits while wearing hearing devices, the distance at which the child can hear sound, and the child's ability to obtain more information from these sounds[13]. In this review, we used MAIS to chart progress in functional hearing performance of prelingual implantees in the second cohort at baseline and at 6, 12, 24 and 36 months post implantation.

e) Meaningful Use of Speech Scale (MUSS)

MUSS is parent's report scale of 10 questions to determine the progress in oral communication skills; voice control, spontaneous speech and ability to change communication strategy to improve clarity and intelligibility of speech in hearing-impaired children when using hearing amplifications from parents' perspective[14]. In this review, we used MUSS to chart progress in functional hearing performance of prelingual implantees in the second cohort at baseline and at 6, 12, 24 and 36 months post implantation.

f) Listening Progress Profile (LiP)

LiP (Appendix 3) is an assessment of scoring on auditory detection, discrimination and early identification abilities of environmental sounds and speech at the single word or phoneme level[15,16]. In this review, we used LiP as a functional outcome in prelingual implantees of the second cohort at baseline and at 6, 12, 24 and 36 months post implantation. Parents /caregivers were asked to observe and record patient's listening behaviors in the environment as well as their response to the speech-related sounds within a certain time frame. They have then scored them according to whether the behavior was shown consistently, occasionally, or none at all.

g) Parents' Evaluation of Aural/Oral Performance of Children (PEACH)

The PEACH questionnaire (Appendix 4) is based on a systematic use of parents' observations to measure child's functional performance in everyday life in hearing and communicating in quiet and in noise with others when using hearing amplifications[17]. The overall PEACH scores were measured at baseline and at 6, 12, 24 and 36 months post implantation and converted into percentage and analyzed for the second cohort prelingual implantees in this review.

h) Current Level of Function (CLF)

Recipients' performance in audition, receptive, expressive and speech skills were collected based on Integrated Scales of Development (ISD) that outlines typical stages of development of speech and language developed by the Cochlear Limited. Based on the ISD, current level of function (CLF) can be calculated as a guide to monitor patient's milestone development[18].

The performance of the children in the CLF were assessed prior to implant as a baseline level and the progress were analyzed at baseline and at 6, 12, 24 and 36 months post implantation from the prelingual group in the second cohort.

i) Word count

Word count was measured based on expressive vocabulary (words produced) by implantees. Parents were given a checklist to continuingly charting the words produced in record sheet (diary). This is used as a guide in recording spontaneous word produced by the implantees[19]. Word count data in this review was taken from the prelingual group in the second cohort. The word count was assessed and analyzed at baseline and at 6, 12, 24 and 36 months post implantation in the prelingual group of the second cohort.

j) *Mode of communication*

In our review we categorized the type of communication modes as verbal, total communication, cued speech, gesture, sign language and Augmentative and Alternative Communication (AAC). Data was collected at 24 months and above post implantation from the prelingual group in the second cohort.

III. Educational Status

Educational status in this review was defined based on educational placement under Ministry of Education (MOE) system. Educational placements that related in this review were higher education level, mainstream school, integration class, cued speech school, special education school and pre-school education.

IV. Quality of Life

Data for quality of life (QoL) outcome measure was collected from adult recipients in the post-lingual group of the second cohort. We used two tools Abbreviated Profile of Hearing Aid Benefit (APHAB) (Appendix 5) and Nijmegen Cochlear Implant Questionnaire (NCIQ) (Appendix 6) to measure QoL at before implantation and at 6 and 12 months post implantation[20,21].In this review, a number of parameters were studied to establish their possible association and correlation with the functional outcomes. The parameters were mainly subjective assessments based on verified international and national scales for prelingual and post-lingual groups. In the prelingual group, parameters studied for factors affecting functional outcomes were mean age at surgery, parental education levels, household income, family participation rating (FPR) (Appendix 7)[22], average aided hearing threshold level and CI usage. In the post-lingual group the parameter studied for factors affecting outcome was mean aided threshold.

D. DATA ANALYSIS

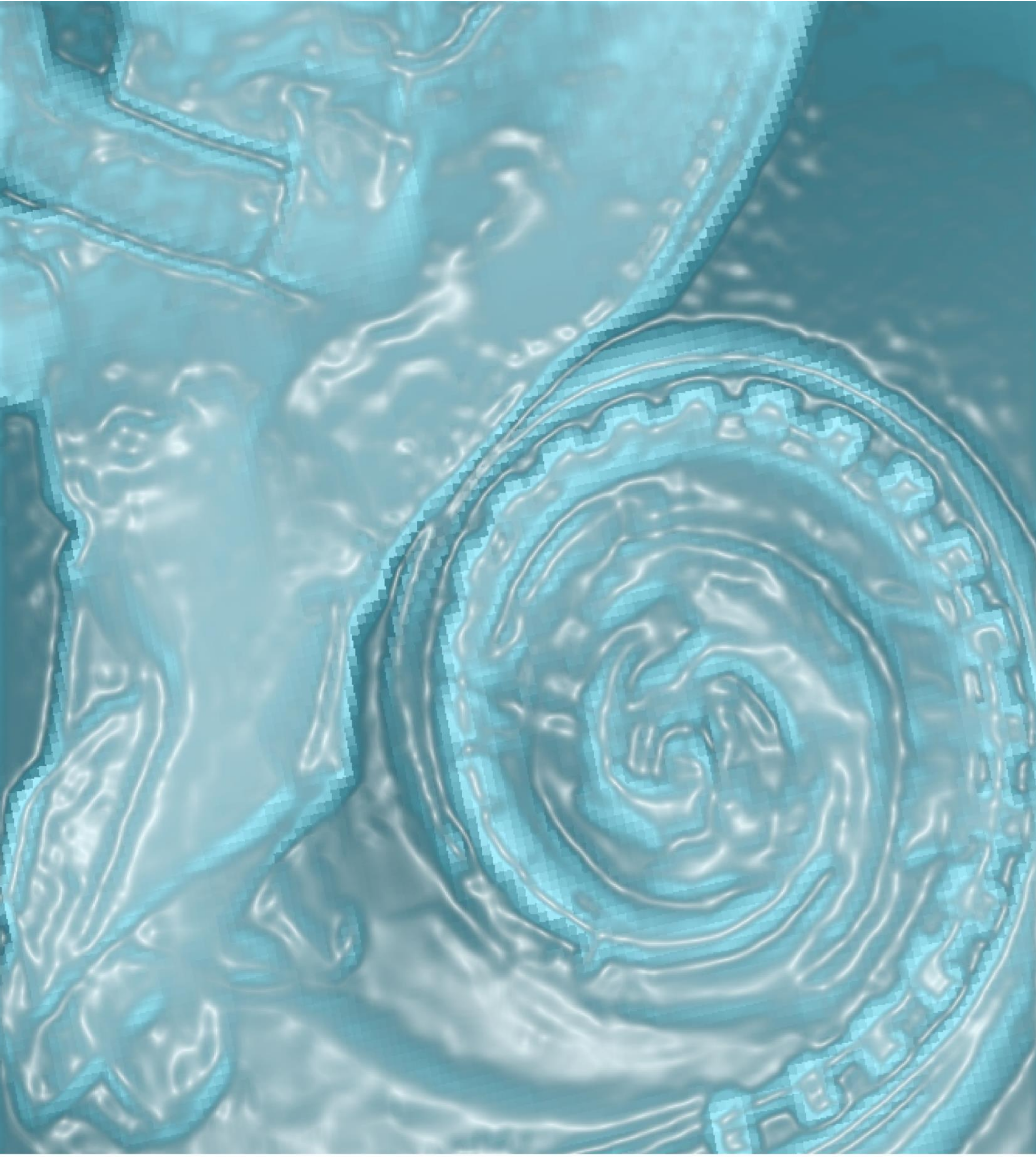
Data were imported from the Excel file into a statistical program for statistical analysis. The analyses were performed with R software version 3.6.2[23]. Descriptive statistics were utilized for selected variables. The results were presented as frequencies and percentage for categorical data while in numerical, which is normally distributed, was presented as mean and standard deviation while the median and interquartile range was presented for numerical data, which is not normally distributed.

Pearson Chi-square test was used to compare differences in categorical data among groups while Fisher's Exact was used as an alternative if the assumptions of Pearson Chi-square not met. Comparing numerical (Mean) data between-subjects were analyzed using the one-way ANOVA test. Repeated measures ANOVA test was used to test changes at 6 months, 12 months, 24 months, 36 months and 48 months of outcomes within-subjects and associated factors.

Kendall's Tau-b correlation was used to study the correlation between variables (SIR, CLF, Word count, MAIS, MUSS, PEACH and LiP score) and CAPII score at each follow-up. The symmetrical distribution of probabilities dividing the alpha level, usually 0.05 into two parts and the probability value of less than 0.05 (p-value < 0.05) was considered as statistically significant[24].

E. LIMITATION OF THE REVIEW

Inherent biases in data recording and collection were unavoidable in this retrospective study. Few subjects were excluded from the functional outcomes analysis because they do not meet the defined interval post implantation assessment and differences in process of data collection in the first and the second cohorts.



RESULTS

Chapter 4

RESULTS

COCHLEAR IMPLANT RECIPIENTS

A total of 413 cochlear implant surgeries were performed involving 380 implantees from 2008 until 2018. Majority of them 347 (84%) were unilateral cochlear implantation and 33 (16%) were bilateral cochlear implantation.

Of the recipients, 283 were children with prelingual deafness and 97 were patients with post-lingual deafness. Distribution of prelingual and post-lingual CI recipients in the satellite hospitals were shown in Table 1 and Table 2.

In the first year of the National MOH Cochlear Implant Programme, four recipients were adult in post-lingual group and only one prelingual recipient. But the scenario changed dramatically in 2009 onward in which more children with prelingual hearing loss were implanted. The youngest recipient was implanted at age of 12 months old due to congenital hearing loss and the oldest recipient was implanted at age of 61 years old due to meningitis in this review.

Ten MOH hospitals are recognized as CI satellite hospitals and are grouped into six zones. The Northern Zone consists of Hospital Sultanah Bahiyah, Alor Setar and Hospital Raja Permaisuri Bainun, Ipoh. The Central Zone consists of Hospital Sungai Buloh and Hospital Kuala Lumpur. The Southern Zone consists of Hospital Tuanku Ja'afar, Seremban and Hospital Sultan Ismail, Johor Bahru. The East Zone consists of Hospital Raja Perempuan Zainab II, Kota Bharu and Hospital Sultanah Nur Zahirah, Kuala Terengganu. The Sabah Zone is represented by Hospital Queen Elizabeth, Kota Kinabalu and the Sarawak Zone is represented by Hospital Umum Sarawak, Kuching.

Table 1. Distribution Of Prelingual CI Recipients MOH Satellite Hospitals 2008-2018

HOSPITALS	YEAR											
	2008	2009	2010	2011	2012	2013	2014	2015	2016	2017	2018	TOTAL
	n	n	n	n	n	n	n	n	n	n	n	n
H Sultanah Bahiyah	0	5	3	5	7	10	5	3	4	6	10	58
H Raja Perempuan Zainab II	0	0	3	2	6	0	2	3	4	2	6	28
H Sultan Ismail	0	4	1	3	6	6	4	1	5	6	4	40
H Sungai Buloh	1	0	3	2	4	3	3	3	1	2	5	27
H Raja Permaisuri Bainun	0	2	0	6	6	2	4	1	5	2	2	30
H Tuanku Ja'afar	0	2	0	2	1	4	1	2	3	3	2	20
H Queen Elizabeth	0	0	1	0	4	0	2	2	1	2	6	18
H Kuala Lumpur	0	2	2	4	3	4	3	5	4	6	4	37
H Umum Sarawak	0	0	0	0	0	0	0	1	3	3	2	9
H Sultanah Nur Zahirah	0	5	1	1	0	2	1	1	2	0	3	16
TOTAL	1	20	14	25	37	31	25	22	32	32	44	283

Table 2. Distribution Of Post Lingual CI Recipients MOH Satellite Hospitals 2008-2018

HOSPITALS	YEAR											
	2008	2009	2010	2011	2012	2013	2014	2015	2016	2017	2018	TOTAL
	n	n	n	n	n	n	n	n	n	n	n	n
H Sultanah Bahiyah	0	2	2	1	3	0	2	2	1	5	3	21
H Raja Perempuan Zainab II	1	0	1	2	1	1	2	1	1	1	0	11
H Sultan Ismail	1	0	1	0	3	1	0	2	1	0	3	12
HSungai Buloh	1	3	1	0	0	4	1	1	1	0	1	13
H Raja Permaisuri Bainun	1	0	3	0	1	0	2	0	0	1	4	12
H Tuanku Jaafar	0	0	0	0	1	1	1	0	1	0	0	4
H Queen Elizabeth	0	1	1	1	3	1	0	0	0	0	1	8
H Kuala Lumpur	0	0	2	1	2	0	1	1	3	1	0	11
H Umum Sarawak	0	0	0	0	0	0	0	0	1	0	1	2
H Sultanah Nur Zahirah	0	0	0	0	2	1	0	0	0	0	0	3
TOTAL	4	6	10	5	15	9	9	7	9	8	13	97

Funding for the procurement of the CI device in the first five years mainly from MOH CI Programme allocation grant. Funding of the CI device in the second five years was summarized in the Table 3 for prelingual group and Table 4 for post-lingual group.

Table 3. Prelingual: Funding for CI devices 2013-2018

FUNDERS	UNILATERAL	BILATERAL	
		First implant	Second implant
	n	n	n
MOH Programme	124	18	1
TBP	20	2	2
JPA	16	0	4
ATM	0	1	1
NGO	0	0	1
Zakat	0	0	2
Insurance	1	0	0
Own funding	1	3	13
Total	**162**	**24**	**24**

Table 4. Post-Lingual: Funding For CI Devices 2013-2018

FUNDERS	UNILATERAL	BILATERAL	
		First implant	Second implant
	n	n	n
MOH Programme	37	0	0
TBP	13	0	0
JPA	3	1	1
ATM	0	0	0
NGO	1	0	0
Zakat	0	0	0
Insurance	1	0	0
Own funding	1	0	0
SOCSO	1	0	0
Total	**57**	**1**	**1**

PRELINGUAL GROUP

Demographic and Clinical Characteristics

From the prelingual group of 283 patients, 253 were unilateral CI recipients and 30 were bilateral CI recipients, resulting in a total of 313 CI surgeries in prelingual children. By gender, 132 were male and 151 were female. Of the recipients 69.6% were Malay, 18.4% were Chinese, 6% were Indian and 6 % were others. In the first cohort (2008-2012), there were 97 CI recipients and in the second cohort (2013-2018) there were 186 CI recipients.

The most common etiology of hearing loss for prelingual group was congenital non syndromic hearing loss 91.5%. There were eight children (2.8%) with congenital syndromic hearing loss. Waardenburg syndrome, Usher syndrome, Charge association and mild dysmorphism without any specific syndrome being diagnosed were recognized in four, one, one and two CI recipients respectively.

The remaining 16 (5.7%) had acquired hearing loss. Of these children seven of them had meningitis, four children had TORCHES, two children had febrile fit, two children had unspecific high grade fever and one child had mastoiditis.

Majority of implanted ears in this review had normal inner ears imaging. 13 ears with congenital cochlear malformation, 3 ears with cochlear ossification and 3 ears with large vestibular aqueduct were implanted. Seven ears with only cochlear nerve hypoplasia were implanted. Five of the thirteen implantees with abnormal cochlea have associated cochlear nerve hypoplasia. All had full electrode insertion except for two patients with incomplete insertion secondary to cochlear ossification.

On parents' education level, more than half of the parents (54%) had a non-tertiary education and majority of the families of the recipients (73.2%) had household incomes of less than RM5000 monthly. These were summarized in Table 5.

Table 5. Prelingual: Demographic And Clinical Characteristics Of CI Recipients From 2008-2018

	n	%
No of patients	283	100
Laterality		
Unilateral	253	(89.4)
Bilateral	30	(10.6)
Gender		
Male	132	(46.6)
Female	151	(53.4)
Ethnic		
Malay	197	(69.6)
Chinese	52	(18.4)
Indian	17	(6.0)
Others	17	(6.0)
Education Level - Father		
Primary	10	(3.5)
Secondary	143	(50.5)
Tertiary	104	(36.7)
No information	26	(9.2)
Education Level - Mother		
Primary	10	(3.5)
Secondary	143	(50.5)
Tertiary	103	(36.4)
No information	27	(9.5)
Household Income		
Less than RM1000	24	(8.5)
RM1000-2499	88	(31.1)
RM2500-4999	95	(33.6)
RM5000 and above	62	(21.9)
No information	14	(4.9)

	n	%
Etiology		
Congenital HL		
Non-syndromic	**259**	(91.5)
Syndromic	**8**	(2.8)
Waardenburg syndrome	4	
Usher syndrome	1	
CHARGE association	1	
Mild dysmorphism	2	
Acquired HL	**16**	**(5.7)**
Meningitis	7	
TORCH infection	4	
Febrile fit	2	
High grade fever	2	
Mastoiditis	1	
Abnormal Imaging		
IP-II	7	
IP-I	3	
Hypoplastic cochlea	1	
Common cavity	1	
LVA	3	
Cochlea ossification	3	
Cochlear nerve hypoplasia	12	

Hearing loss in majority (88%) of these children was detected via parental suspicion (passive detection) i.e., they were prompted by the child's inappropriate or lack of response towards sound. In this review of the 10 years period, 12% of them were detected through newborn hearing screening (NHS). However, there was improvement in detection of hearing loss through NHS in the second cohort 18.8% (n=35) as compared to the first cohort 8.2% (n=8). Of those 35 patients in the second cohort, 13 were screened from universal newborn hearing screening (UNHS) program and 22 were screened from high risk newborn hearing screening (HRNHS) program as Table 6 below.

Table 6. Prelingual: Detection of Hearing Loss

Detection of Hearing Loss	1st Cohort (2008-2012)		2nd Cohort (2013-2018)	
	n	%	n	%
Newborn Hearing Screening	8	(8.2)	35	(18.8)
Passive detection	89	(91.8)	151	(81.2)

Table 7(a). Prelingual: Distribution Of Patients Based On Age Group At Diagnosis, Surgery & Switch On

Variables	2008-2012		2013-2018		2008-2012	
	n=97		n=186		n=283	
	n	(%)	n	(%)	n	(%)
Age of Diagnosis						
<6m	2	(2.1)	18	(9.7)	20	(7.1)
7-12m	8	(8.2)	24	(12.9)	32	(11.3)
13-24m	31	(32.0)	51	(27.4)	82	(29)
25-36m	40	(41.2)	68	(36.6)	108	(38.2)
>37m	11	(11.3)	25	(13.4)	36	(12.7)
No information	5	(5.2)	0	(0.0)	5	(1.8)
Age of Surgery						
<18m	1	(1.0)	5	(2.7)	6	(2.1)
19-24m	3	(3.1)	18	(9.7)	21	(7.4)
25-36m	17	(17.5)	49	(26.3)	66	(23.3)
37-48m	44	(45.4)	85	(45.7)	129	(45.6)
>49m	32	(33.0)	29	(15.6)	61	(21.6)
Age of Switch On						
<18m	1	(1.0)	4	(2.2)	5	(1.8)
19-24m	2	(2.1)	17	(9.1)	19	(6.7)
25-36m	16	(16.5)	48	(25.8)	64	(22.6)
37-48m	40	(41.2)	85	(45.7)	125	(44.2)
>49m	36	(37.1)	32	(17.2)	68	(24)
No information	2	(2.1)	0	(0.0)	2	(0.7)

Table 7(b). Prelingual: Mean Age At Diagnosis, At Surgery And At Switch On

Variables	2008-2018	2008-2012	2013-2018
	n=268	n=97	n=171
AGE OF DIAGNOSIS			
N	262	92	170
Mean	23.4	24.4	22.8
SD	10.58	9.49	11.12
AGE OF SURGERY			
N	268	97	171
Mean	39.2	42.9	37.1
SD	10.22	9.79	9.86
AGE OF SWITCH ON			
N	265	95	170
Mean	40.0	43.9	37.7
SD	10.28	9.81	9.89

Table 7 (c). Prelingual: Interval Between Diagnosis To Surgery And From Surgery To Switch On

MEAN INTERVAL		
Variables	**2008-2012**	**2013-2018**
DIAGNOSIS TO SURGERY IN MONTHS		
N	97	186
Mean	17.9	15.4
SD	(8.86)	(9.25)
SURGERY TO SWITCH ON IN WEEKS		
N	97	186
Mean	3.5	3.0
SD	(1.22)	(1.03)

From 2008 to 2018, of 283 recipients, 268 patients were included in mean age analysis. 15 recipients with prior normal hearing or progressive hearing loss were excluded from this analysis.

The 2nd cohort (22.6%) has shown encouraging results in which the age of diagnosis for patients of less than 12 months as compared to the 1st cohort (10.3%) since we started the CI programme. However, the majority of recipients in both cohorts were diagnosed at the age of 25-36 months old. Age of recipients diagnosed beyond 3 years old almost similar in both cohorts (Table 7(a)).

Age of surgery below 24 months has increased from 4.1% in 1st cohort to 12.4% in 2nd cohort. Similar findings were observed in children implanted at the age group of 36 months and below (21.6% to 38.7%). The percentage of patients undergoing surgery beyond 49 months of age has been markedly reduced by 50% in the second cohort (Table 7(a)).

The overall mean age at diagnosis was 23.4 months, mean age at surgery was 39.2 months and the mean age of switch on was 40 months. The comparison between the two cohorts showed that the mean ages at diagnosis, at surgery and at switch-on of the second cohort were improved as shown in Table 7(b). The mean interval between diagnosis and surgery of the first and second cohort is shortened from 17.9 months to 15.4 months (Table 7(c)).

Surgical and Device Related Complications

Intraoperatively we encountered 9 cases of perilymph gusher, which were successfully managed during the surgeries. Out of 9 cases, 5 had abnormal findings radiologically; 2 patients with IP type II, 2 patients with IP type I and 1 patient with large vestibular aqueduct (LVA) syndrome. The other four patients had normal radiological findings.

Of 313 ears implanted, 16 ears were reported with surgical complication, in which 10 were major and 6 were minor as shown in Table 8. Overall complication rate for prelingual group was 5.11%.

Table 8. Prelingual: Surgical And Device Related Complications

PRELINGUAL SURGICAL COMPLICATIONS		n
Major complication	Device failure	6
	Electrode migration	2
	SSI major	2
Minor complication	SSI minor	5
	Facial nerve paresis/palsy	1
	Total	16

Major Complication

i) *Device failure*

Device failure in 6 patients occurred at 1,3,5,8,16 and 52 months following switch-on. There was no reported trauma to the site of the internal receiver in all cases. All of the patients had undergone uneventful explantation and reimplantation. Three devices had hard failure and the other three had soft failures.

ii) *Electrode migration*

Two patients with electrode array migration that occurred at 6 and 12 months following the implantation. Preoperative radiological imaging and intraoperative findings were normal in both patients. Both of them had uneventful repositioning of electrode array reimplantation using the same device.

iii) *Surgical site infection (SSI) major*

One patient developed post auricular abscess on the surgical site a year following implantation which required surgical drainage with intact implant. Another patient had unresolved granulation tissue with inflammation over the receiver site two months following implantation that needed multiple debridement procedures. She underwent ex-plantation and re-implantation due to associated device failure with a new implant after 34 months of the implantation.

Minor complication

There were six minor complications reported. Five patients had minor SSI wound infection including one patient with delayed wound healing. All of them were managed conservatively with intravenous antibiotics and wound dressing. One patient developed delayed facial paresis, a week following the surgery that recovered completely with conservative measures.

Functional Outcomes

i) Aided Thresholds

The mean aided thresholds with hearing aids measured at four frequencies average (500 Hz, 1000, 2000 Hz and 4000Hz) prior to cochlear implantation for the first cohort was 80dBHL based on previous report and for the second cohort 76.2dBHL (Table 9). Following implantation, 77% patients had improvement of the mean aided thresholds of better than 40dBHL. The remaining 18.7% of patients showed mean aided thresholds of between 40 to 80dBHL.

Table 9. Prelingual: Hearing Aid And Cochlear Implant Aided Thresholds

Variables	2008-2018		2008-2012		2013-2018	
	n=283		n=97		n=186	
	n	(%)	n	(%)	n	(%)
HA AIDED THRESHOLD						
Mean				80		76.2
CI AIDED THRESHOLD						
≤40dB	218	(77.0)	77	(79.4)	141	(75.8)
>41dB	53	(18.7)	20	(20.6)	33	(17.7)
Not Applicable*	12	(4.2)	0	(0.0)	12	(6.5)

* Information not available or CI hearing age less than 12 months

ii) Categorical Auditory Performances II (CAPII) Score

CAPII score was measured at 6, 12, 24, 36 and 48 months interval post implantation. The recipients were grouped according to their age at surgery (Group 1: less than 24 months, Group 2: 25-36 months, Group 3: 37-48, Group 4: more than 49 months). Of 283 recipients, 15 were excluded from functional outcome analysis due to either they have not reached the 6 months progress interval (eight recipients) or due to incomplete data analysis (seven recipients).

The overall CAPII score showed significant improvement at 6 to 48 months follow-up after implantation ($p<0.002$). At 6 months, the mean CAPII score was 2.9 and continued to increase to 5.2 at 48 months.

The CAPII score continued to improve with time in all age groups. Children implanted before 24 months of age showed more rapid improvement (steeper curve) compared to the other groups. Group 1 and 2 showed a higher score in mean CAPII at 24 months follow up as compared to Group 3 and 4 with a mean of

4.8, although they were not statistically significant. Results as summarized in Table 10 as well as in Figure 1 and 2.

iii) Speech Intelligibility Rating Scale (SIR)

Speech Intelligibility Rating Scale (SIR) was measured prior to implantation, and at 6, 12, 24, 36 and 48 months interval post implantation. The children were subdivided into four categories according to their age at surgery. Group 1 were children implanted at age 24 months old or younger, Group 2 were children implanted at age 25 to 36 months old, Group 3 were children implanted at age 37 to 48 months old and Group 4 were children implanted at age 49 months old or older. Of 283 recipients, 15 were excluded from functional outcome analysis due to either they have not reached the 6 months progress interval (eight recipients) or due to incomplete data analysis (seven recipients).

Overall, at 6 months the mean SIR score was 1.5 and it continued to increase to 2.9 at 48 months. There was significant mean SIR different between each group at 6 to 48 months post implantation as shown in Table 11.

At 12 months post implantation, more than half (55%) of the children were able to produce intelligible speech in single words when context and lip reading cues are available (SIR score 2 and above). At 48 months post implantation, 28% of our patients; mainly from Group 1 and 2 are able to produce intelligible speech to a listener who concentrates and lip reads (SIR score 3 and above).

The mean SIR score continued to improve with time. Children in Group 1 and Group 2 showed more rapid improvement compared to children in Group 3 and Group 4. They also had a higher mean score at 24 months follow up. Group 3 and Group 4 were noted to have poorer performance than the overall score as shown in Figure 3 and Figure 4.

Table 11a demonstrates that auditory perception showed improvement across all age groups. The mean SIR improvement is better in those implanted at younger age groups. Those implanted below the age of 24 months showed a mean SIR improvement of 2.0 as compared to 0.9 in those implanted above 48 months.

Table 10. Prelingual:CAPII Score Post Implantation 2008-2018

Variables	CAPII SCORE 2008-2018															
	6 months			12 months			24 months			36 months			48 months			*Within-subjects*
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	p-value
OVERALL	268	2.9	(1.40)	238	3.7	(1.39)	193	4.5	(1.36)	158	4.9	(1.44)	136	5.2	(1.56)	0.002
AGE OF SURGERY																1.000
≤24m	26	2.8	(1.33)	20	3.6	(1.47)	13	4.8	(0.90)	11	5.3	(0.79)	7	5.6	(0.53)	
25-36m	64	3.2	(1.42)	56	3.9	(1.50)	45	4.8	(1.62)	37	5.4	(1.72)	31	5.5	(1.98)	
37-48m	120	2.6	(1.26)	106	3.4	(1.30)	90	4.3	(1.23)	69	4.7	(1.36)	58	5.0	(1.47)	
≥49m	58	3.3	(1.54)	56	3.9	(1.33)	45	4.6	(1.37)	41	4.9	(1.37)	40	5.1	(1.43)	
Between-groups																
p-value		0.004			0.029			0.090			0.089			0.411		

Table 11. Prelingual: SIR Score Post Implantation 2008-2018

Variables	SIR SCORE 2008-2018															
	6 months			12 months			24 months			36 months			48 months			Within-subjects
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	p-value
OVERALL	268	1.5	(0.82)	238	1.9	(0.94)	193	2.4	(1.03)	158	2.6	(1.10)	136	2.9	(1.25)	0.245
AGE OF SURGERY																0.974
<24m	26	1.3	(0.56)	20	2.0	(0.83)	13	2.5	(1.05)	11	2.6	(1.03)	7	3.3	(0.76)	
25-36m	64	1.7	(0.92)	56	2.1	(1.07)	45	2.8	(1.15)	37	3.1	(1.23)	31	3.4	(1.39)	
37-48m	120	1.4	(0.68)	106	1.7	(0.85)	90	2.1	(0.93)	69	2.4	(1.02)	58	2.8	(1.25)	
>49m	58	1.8	(0.99)	56	2.0	(0.94)	45	2.3	(1.00)	41	2.5	(1.03)	40	2.7	(1.11)	
Between-groups																
p-value		0.009			0.018			0.008			0.031			0.049		

Table 11a. Prelingual: Post Implantation Mean CAP & SIR Score Differences Based On Age Group

Age of Surgery	<24m	25-36m	37-48m	>49m
Mean CAP Difference	2.5	2.1	1.8	2.4
Mean SIR Difference	2.0	1.7	1.4	0.9

Figure 1. Prelingual: Overall Mean CAPII Score Post Implantation 2008-2018

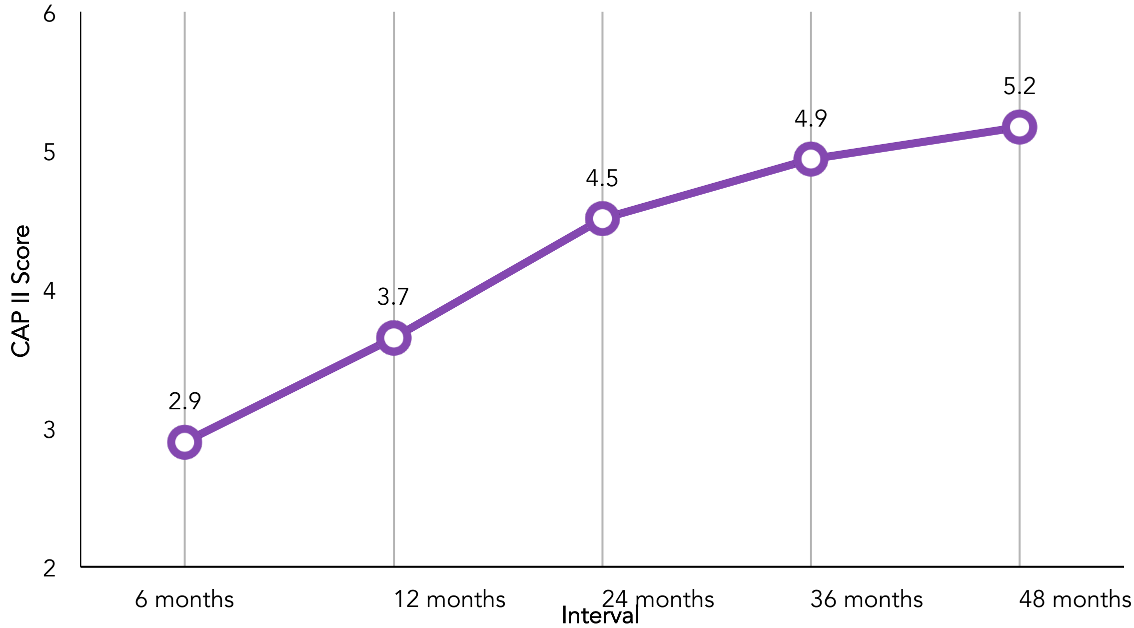


Figure 2. Prelingual: Mean CAPII Score Post Implantation And Age Of Surgery 2008-2018

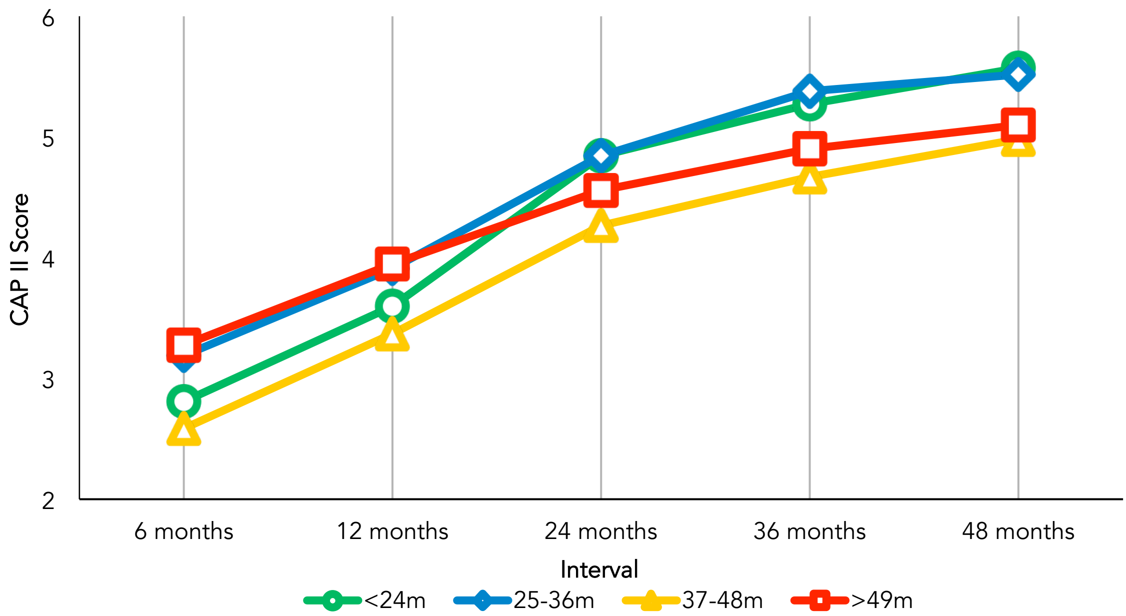


Figure 3. Prelingual: Overall Mean SIR Post Implantation 2008-2018

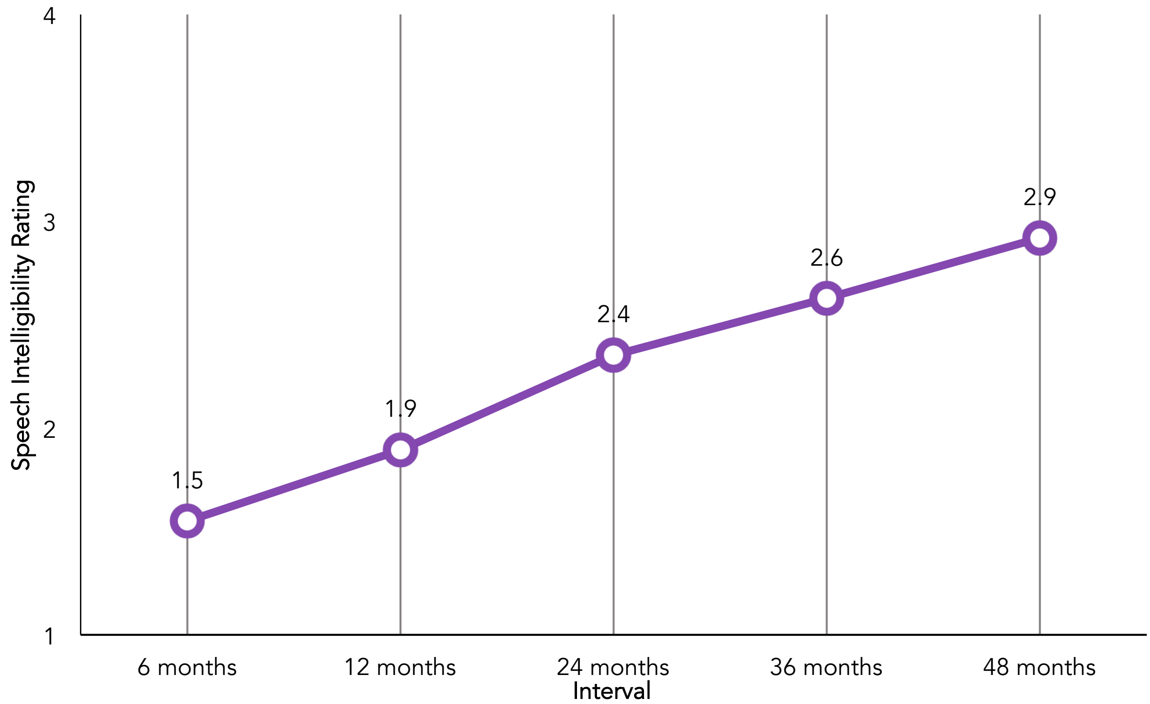
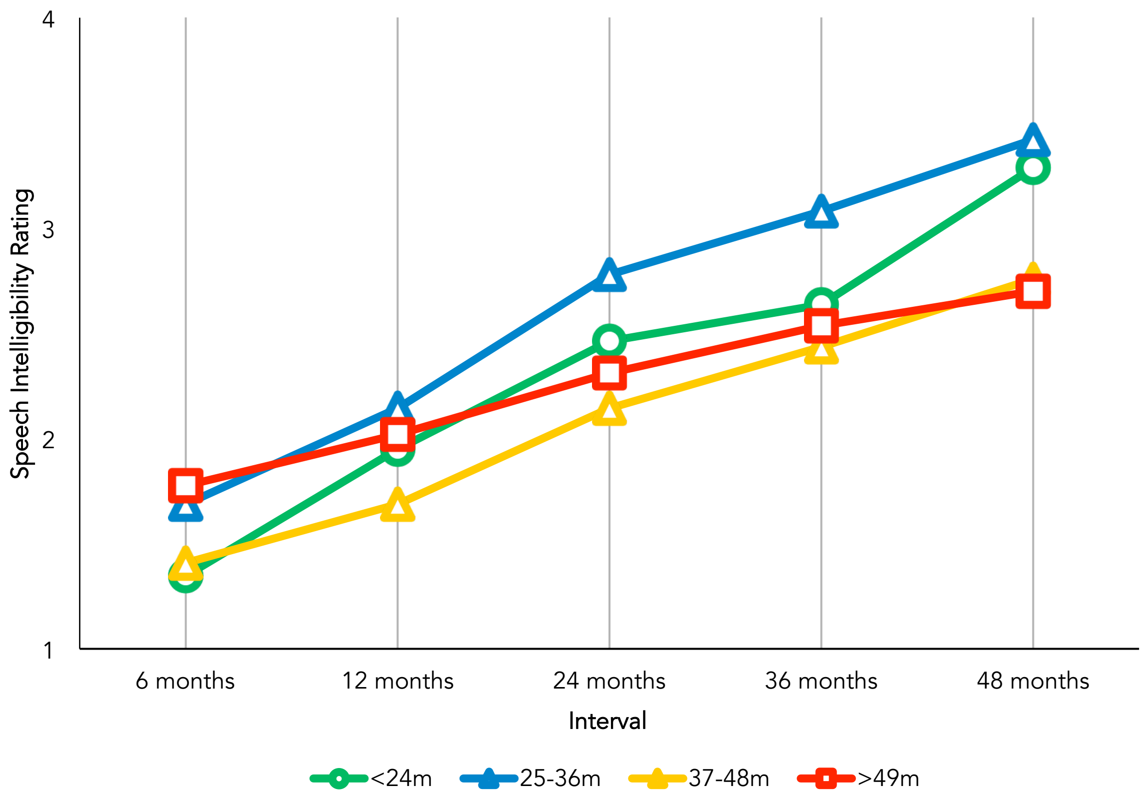


Figure 4. Prelingual: Overall SIR Post Implantation And Age Of Surgery 2008-2018



b) Meaningful Auditory Integration Scale and Meaningful Use of Speech Scale

All patients showed improvement over time. As shown in Table 12, in each interval, the score recorded was markedly higher than the baseline, indicating the improvement of speech understanding and the use of speech in a different natural context. Moreover, MAIS and MUSS score gradually increased as the time of rehabilitation extended, as demonstrated in Figure 5. The maximum score for each questionnaire is 40.

Overall, MAIS improved from a mean of 11.6 at the baseline to 36.6 at 36 months after implantation. For MUSS, our results showed clinically remarkable increment from a mean of 6.6 at the baseline to 30.3 at 36 months after implantation (Table 12 and Figure 5).

c) Parents' Evaluation of Aural/Oral Performance of Children (PEACH)

PEACH rating scale was used to evaluate the effectiveness of hearing technology for patients under our series. We analyzed the overall functional ability of these children in both quiet and noisy environment descriptively. Our PEACH analysis shows that the children under this series continuously receiving benefits from their implants with the remarkable growth of performance as shown in Figure 6 throughout the follow-ups.

At the initial observations before implantation, parents scored 20.1% on the aural/oral performance, and at 36 months, they rated 75.6%, more than three-fold increment than before as in Table 12 and Figure 6.

d) Listening Progress Profile (LiP)

The Listening Progress Profile performance scores of prelingual patients in this series, show marked improvement at the first six months of implantation compared to baseline and continued to improve with time. Prior to implantation, they scored 13.8 and by the 36 months interval they were able to reach the score of 39.1 over 42 (Table 12 & Figure 7).

Table 12. Prelingual:Overall Scores of MAIS, MUSS, PEACH and LiP of Prelingual CI Recipients 2013-2018

Variables	Baseline			6 months			12 months			24 months			36 months			Within-subjects
MAIS, MUSS, PEACH AND LiP OVERALL SCORES																
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	*p*-value
MAIS	149	11.6	(7.38)	119	21.9	(9.07)	82	27.0	(8.50)	52	32.6	(6.65)	25	36.6	(6.62)	0.077
MUSS	106	6.6	(5.26)	99	12.0	(7.42)	69	16.4	(8.54)	51	22.0	(10.06)	23	30.3	(10.78)	0.336
PEACH	26	20.1	(20.86)	41	46.4	(25.38)	32	53.8	(25.99)	20	60.8	(20.53)	12	75.6	(25.70)	0.668
LiP	103	13.8	(10.39)	92	26.2	(10.87)	64	31.6	(9.25)	41	35.8	(7.08)	23	39.1	(3.87)	0.873

Figure 5. Prelingual:Overall Scores of MAIS and MUSS 2013-2018

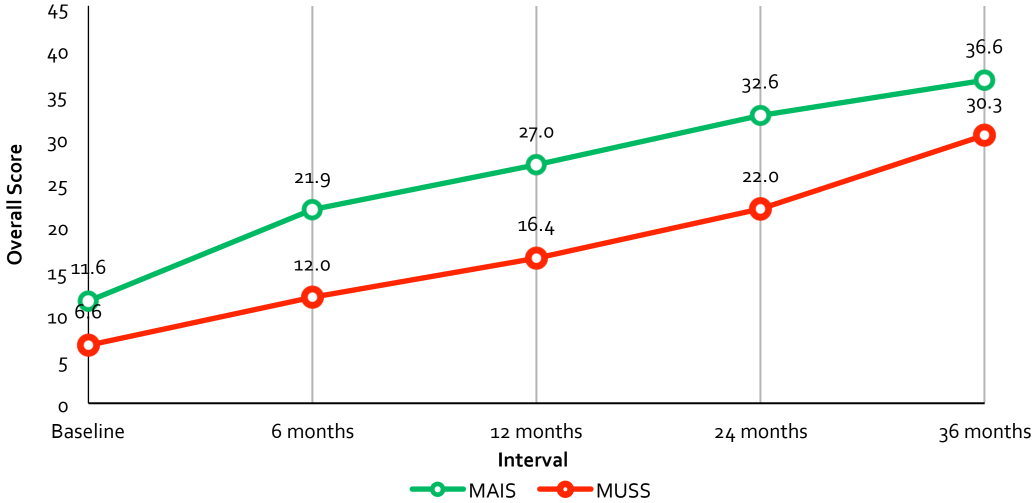


Figure 6. Prelingual:Overall Scores of PEACH 2013-2018

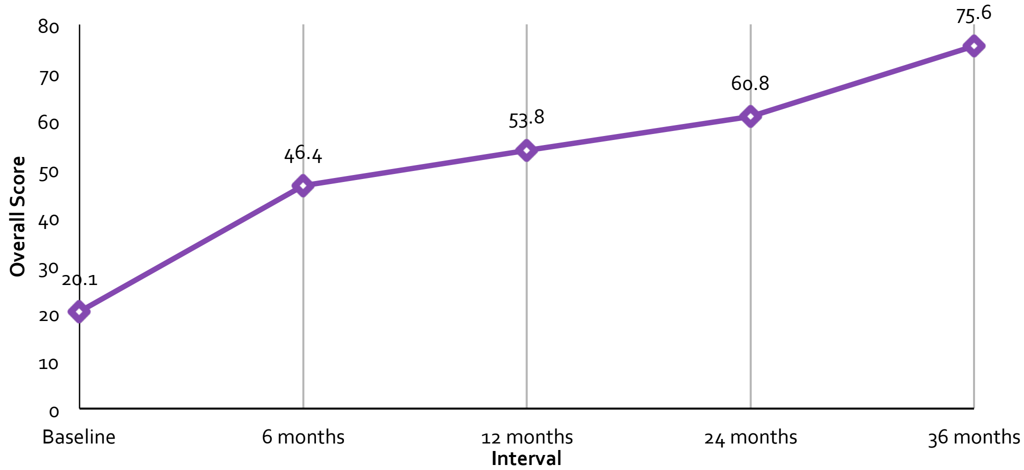
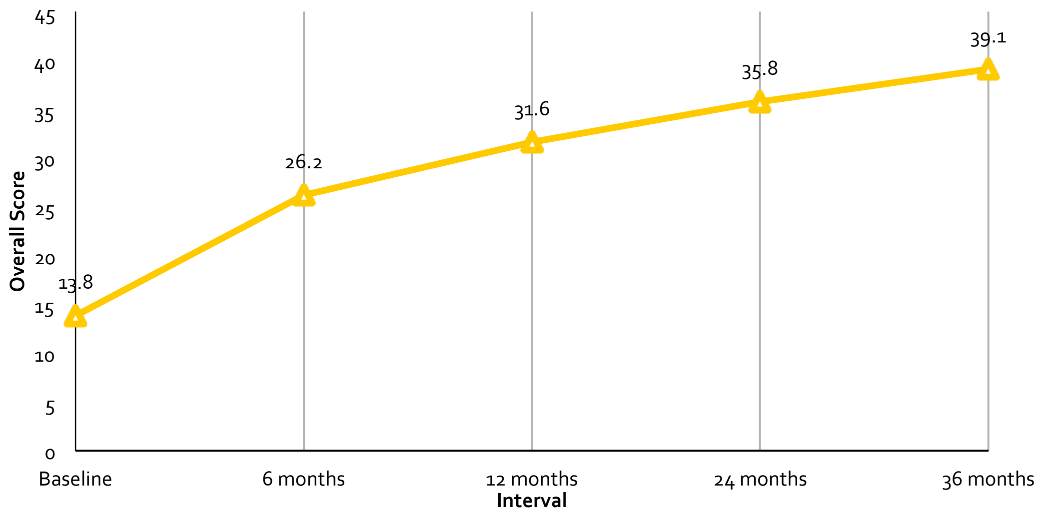


Figure 7. Prelingual:Overall Scores of LiP 2013-2018



e) *Current Level of Function (CLF)*

CLF score was measured prior to implantation and at 6, 12, 24 and 36 months intervals post implantation. The implantees were grouped according to their age at surgery (Group 1: less than 24 months, Group 2: 25-36 months, Group 3 37-48, Group 4 >49 months).

Overall mean CLF score showed improvement from 4.0 at baseline up to 21.9 at 36 months post implantation as in Table 13 and Figure 8. All groups showed improvements in the mean CLF score throughout follow up as shown in Figure 9. However, it is not statistically significant. After 36 months post implantation, the Group 1 achieved mean CLF score of 20 months. The Group 2 achieved mean CLF score of 27.4 months, the Group 3 achieved mean CLF score of 17.4 months and the Group 4 achieved mean CLF 23 months.

From the result, children implanted at age 49 months old and older showed the highest mean CLF score prior to implantation indicated that the recipients had already gained some language function with hearing aids and the score continued to increase throughout follow up.

f) *Word count*

Word count score was measured prior to implantation and at 6, 12, 24 and 36 months post implantation in patients from the second cohort.

Overall, mean word count at the baseline was 0.3 words and it continued to increase to 197.9 words at 36 months shown in Figure 10. Although it was not statistically significant, clinically the word count score for all age groups showed improvement throughout the follow-up as shown in Table 14 and Figure 11.

Table 13. Prelingual: Mean CLF At Baseline And Post Implantation 2013-2018

Variables	CLF score (Prelingual)														
	Baseline			6 months			12 months			24 months			36 months		
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)
Overall	**169**	**4.0**	**(2.76)**	**156**	**7.7**	**(4.25)**	**131**	**11.4**	**(6.10)**	**89**	**17.1**	**(9.63)**	**63**	**21.9**	**(13.30)**
Age of Surgery															
<24m	23	2.9	(1.94)	22	7.3	(2.49)	16	11.4	(4.91)	8	17.4	(7.89)	7	20.0	(6.35)
25-36m	45	3.6	(2.08)	43	8.3	(4.74)	38	12.4	(7.46)	27	20.0	(12.06)	22	27.4	(17.25)
37-48m	82	4.1	(2.86)	74	7.1	(4.23)	60	10.3	(5.15)	44	15.1	(7.87)	26	17.4	(10.20)
>49m	19	5.9	(3.61)	17	9.4	(4.54)	17	13.5	(6.47)	10	18.2	(9.89)	8	23.0	(9.53)
Between-groups															
p-value	0.002			0.174			0.183			0.224			0.069		

Figure 8. Prelingual: Overall Mean CLF 2013-2018

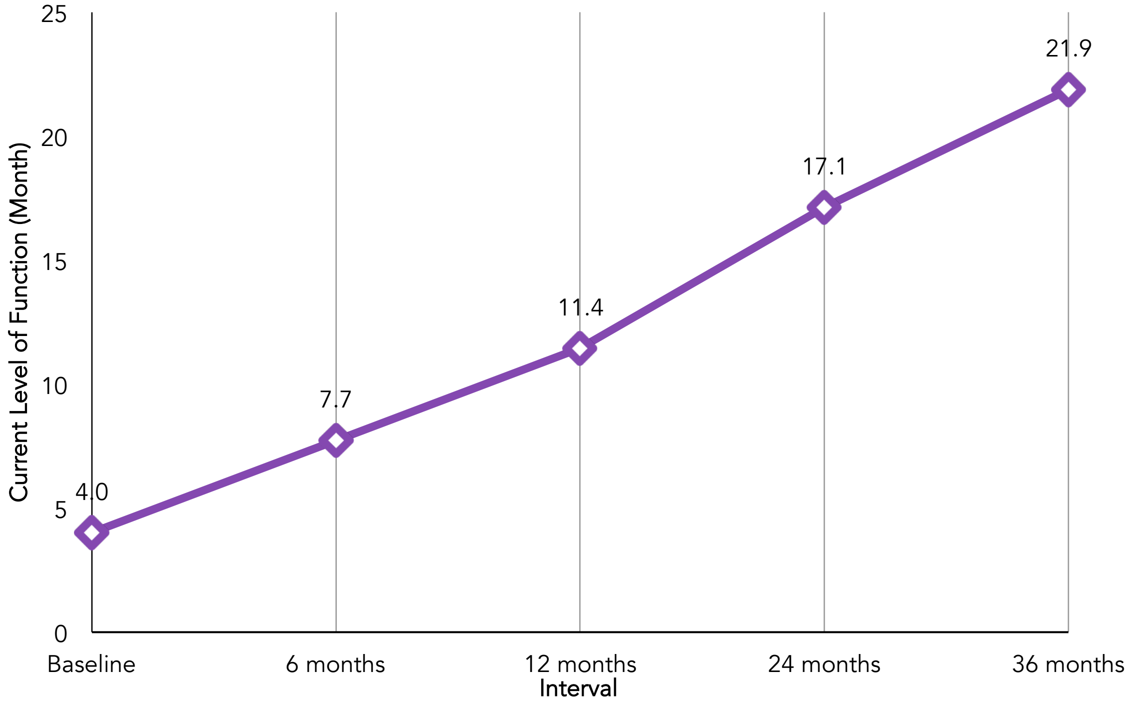


Figure 9. Prelingual: Mean CLF Post Implantation Based On Age Groups 2013-2018

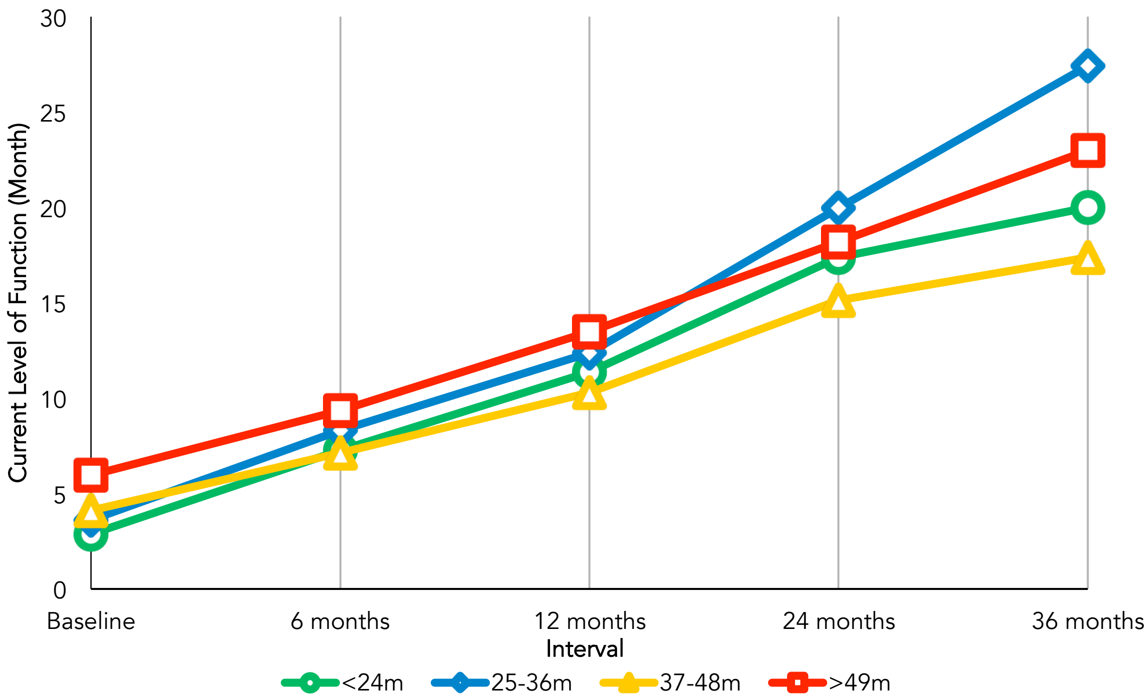


Table 14. Prelingual: Mean Word Count At Baseline And Post Implantation 2013-2018

Variables	Word Count (Prelingual)														
	Baseline			6 months			12 months			24 months			36 months		
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)
Overall	170	0.3	(1.01)	157	4.8	(10.76)	130	23.5	(49.76)	89	78.9	(160.37)	63	197.9	(411.92)
Age of Surgery															
<24m	23	0.0	(0.00)	22	2.3	(3.65)	16	13.1	(16.53)	8	81.3	(131.45)	7	86.7	(104.26)
25-36m	46	0.3	(1.06)	44	6.3	(12.01)	37	34.8	(54.98)	27	138.5	(234.46)	22	386.1	(618.23)
37-48m	82	0.2	(0.74)	74	4.5	(10.99)	60	17.8	(44.32)	44	35.1	(49.22)	26	76.0	(138.01)
>49m	19	1.1	(1.87)	17	6.0	(12.41)	17	28.9	(71.44)	10	108.4	(216.04)	8	174.3	(315.27)
Between-groups															
p-value		0.003			0.497			0.310			0.059			0.056	

Figure 10. Prelingual: Overall Mean Word Count 2013-2018

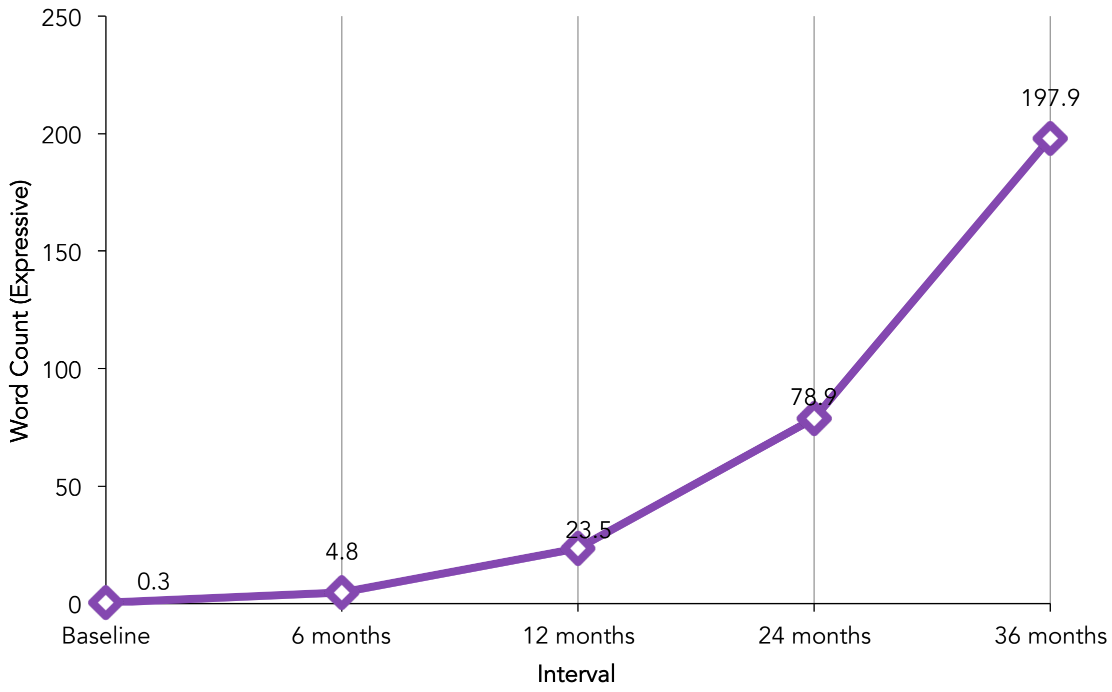
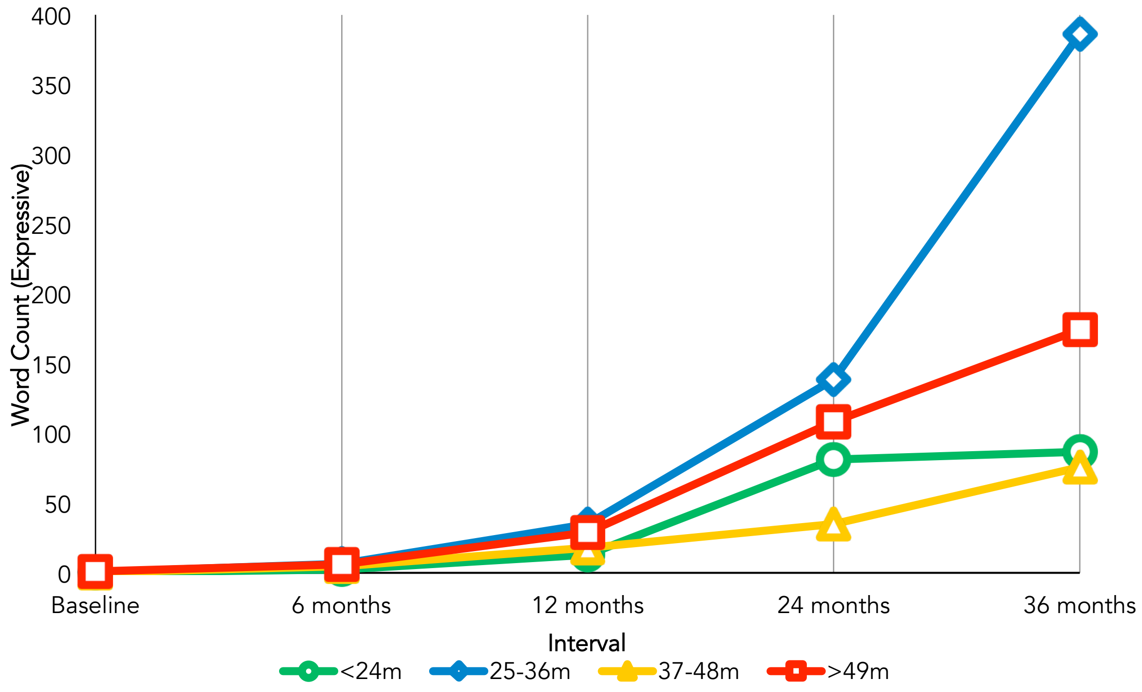


Figure 11. Prelingual: Word Count Based On Age Groups 2013-2018



g) Mode of communication

The mode of communication for the second cohort 2013-2018 was reviewed at 24 months and above post implantation. Out of 186 recipients, 100 had reached at least 24 months post implantation rehabilitation and were included for this analysis. 41 children (41%) were using verbal, 15 children (15%) were using total communication, 4 children (4%) were using cued speech, 30 children (30%) were using gesture, 9 children (9%) sign language, and 1 child (1%) was using AAC. Results are summarized as in Table 14(a).

Table 14 (a). Prelingual: Mode Of Communication Among CI Recipients at 24 Months Post Implantation

Mode of Communication	Number of Patient
Verbal	41
Total communication	15
Cued speech	4
Sign Language	9
Gestures	30
AAC	1

Factors Affecting Outcomes

To look at the trend of outcome changes across follow-ups (within subjects), a series of repeated measures ANOVA were conducted to explore the relationship between changes of the CAPII scores and SIR with factors affecting it. Overall, for the CAPII scores, there was a significant change from baseline to 48 months, p=0.002. Nonetheless, only household income has shown a statistically significant result as the only factor affecting the changes in the CAPII score. SIR steadily increased in trend from baseline to 48 months. Although SIR improved clinically, it was not statistically significant.

Similarly, a series of repeated measures one way ANOVA were conducted to explore the relationships between CAPII scores and SIR with factors that may have influenced the functional outcomes. Of all the factors analyzed in this review, the age of surgery, household income, aided threshold and family participation rating (FPR) had statistically significant associations with CAPII score and SIR. These were summarized in Table 15(a), Table 15(b), Table 16(a) and Table 16(b).

In the second cohort, some new functional outcomes parameters were added (MAIS, MUSS, PEACH, LiP, CLF and Word count) and statistical analysis was used to analyse the factors for association as shown in Table 12, Table 17,Table 18 and Table 19.

i) Age of Surgery

For CAPII score, the differences among groups were statistically significant at 6 and 12 months after the device activation. Figure 2 showed at 24 months onwards, the younger age groups had better CAPII scores than the older age groups, however it was not statistically significant.

Age of surgery was shown to have significant correlation with SIR. The younger age groups showed better SIR across the test intervals compared to the other two groups as in Figure 4.

MAIS, MUSS, LiP, and PEACH continued to improve with time after implantation. However, these improvements were not statistically significant. MAIS, MUSS, LiP, and PEACH score at 36 months post-implantation for those recipients who had surgeries at the age of 24 months and below were higher compared to those implanted at a later age as shown in Table 12.

In all age groups, both word count and CLF at 36 months was not statistically significant with a p-value of p=0.056 and p=0.069, respectively Table 12.

ii) Parents' Education Level

In our review, there was no significant association between education level of parents with CAPII, SIR, CLP, word count, PEACH, MAIS, MUSS and LiP (Table 17).

iii) Household Income

For household income, all groups derived substantial benefit from their implant. Better CAPII score were observed among recipients with household income RM1000 and above and this was statistically significant (p=0.016).

Recipients with household income RM1000 and above showed better SIR. However, it was not statistically significant (p=0.057). Those recipients with household income of less than RM1000 showed a stagnant score from 36-48 months.

There were no associated correlation between MAIS, MUSS, LiP, PEACH, word count and CLF scores at 36 months post implantation among recipients with the household income (Table 18).

iv) Family Participating Rating (FPR)

Patients with family participation rating of scale 4 and 5 showed positive impact to the performance of CAPII score and SIR across test intervals (p<0.001).

For the second cohort, family with FPR 4 and 5 scored higher in all measurements, especially in LiP where the differences were statistically significant. Significant correlation also showed in CLF and word count (p<0.001) (Table 18).

v) CI Usage

The recipients with higher data logs, CI usage 12 hours and more per day have higher score in MAIS, MUSS, LiP, CLF, word count and PEACH as compared to the recipients whom CI usage were less than 12 hours per day. However the correlations between CI usage and the parameters measured were not statistically significant (Table 19).

vi) CI Aided

Patients with better-aided thresholds (≤40dBHL) showed marked improvement in CAPII score and SIR than the group with poorer aided thresholds (>40 dBHL). Results were all statistically significant across test intervals except for at 36 months (p=0.051). Across post implantation interval, the recipients with better aided thresholds had better SIR than the poorer aided thresholds. This was statistically significant at 24 months and 36 months follow up with p=0.027 and p=0.035 respectively (Table 19).

Improvement in the MAIS and the MUSS scores were observed after implantation. The range of score in MAIS was from 33 to 37 and in MUSS was from 30 to 31 for MUSS at 36 months post implantation. However, the MAIS and the MUSS improvement with better aided threshold and poorer aided threshold were not statistically significant in this review.

The recipients with better aided thresholds exhibited better behavioural response towards sounds compared to the recipients with poorer aided threshold in the LiP performance. There was strong correlation and significant distinction statistically between the LiP performance and level of aided threshold in this review.

Patients with better aided thresholds (<40dBHL) showed improvement in CLF scores than the group with poorer aided thresholds. Results are all statistically significant across test intervals. However, this result was not statistically significant in word count.

Table 15(a). Prelingual: Correlations Of CAPII Scores With Factors Affecting Outcomes (CI Aided,Education Level-Father & Education Level-Mother) From 2008 To 2018

Variables	CAPII Score from 2008 to 2018															
	6 months			12 months			24 months			36 months			48 months			Within-subjects
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	p-value
Overall	268	2.9	(1.40)	238	3.7	(1.39)	193	4.5	(1.36)	158	4.9	(1.44)	136	5.2	(1.56)	0.002
CI Aided																0.787
<40	212	3.0	(1.35)	195	3.8	(1.31)	159	4.6	(1.31)	131	5.0	(1.37)	113	5.3	(1.45)	
>41	50	2.4	(1.50)	40	3.1	(1.49)	32	4.0	(1.47)	27	4.4	(1.72)	23	4.6	(1.95)	
Between-groups																
p-value		0.003			0.002			0.011			0.051			0.041		
Education level - Father																0.834
Primary	9	2.6	(1.51)	9	3.4	(1.51)	6	4.5	(1.38)	6	4.8	(1.47)	5	4.8	(1.64)	
Secondary	136	3.0	(1.31)	120	3.7	(1.34)	99	4.5	(1.30)	83	4.9	(1.39)	71	5.2	(1.49)	
Tertiary	101	3.0	(1.52)	91	3.6	(1.44)	73	4.5	(1.44)	58	5.0	(1.52)	50	5.2	(1.64)	
Between- groups																
p-value		0.693			0.709			0.998			0.915			0.825		
Education level - Mother																0.932
Primary	9	2.2	(1.56)	7	3.1	(1.46)	7	3.9	(1.46)	4	4.5	(1.73)	4	4.5	(1.73)	
Secondary	136	2.9	(1.34)	120	3.9	(1.28)	99	4.6	(1.29)	90	5.0	(1.38)	76	5.3	(1.47)	
Tertiary	99	3.0	(1.49)	90	3.5	(1.50)	70	4.6	(1.44)	52	4.9	(1.54)	45	5.1	(1.68)	
Between- groups																
p-value		0.228			0.103			0.386			0.777			0.419		

Table 15(b). Prelingual: CAPII Score Correlations With Factors Affecting Outcomes (Household Income & FPR At 12 Months) From 2008 To 2018

Variables	CAPII SCORE FROM 2008 TO 2018															
	6 months			12 months			24 months			36 months			48 months			Within-subjects
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	p-value
Household Income																0.016
Less than RM1000	22	2.7	(1.13)	19	3.5	(1.17)	16	4.0	(1.10)	13	4.3	(1.84)	10	4.1	(2.13)	
RM1000 and above	232	3.0	(1.37)	205	3.7	(1.33)	163	4.7	(1.29)	133	5.1	(1.32)	115	5.4	(1.39)	
Between- groups																
p-value		0.312			0.399			0.053			0.047			0.009		
FPR At 12 Months																0.456
1	12	1.9	(1.51)	11	2.9	(1.14)	10	3.6	(1.07)	6	3.8	(1.33)	6	3.8	(1.33)	
2	45	2.5	(1.16)	45	2.9	(1.22)	33	3.5	(1.23)	26	3.7	(1.51)	21	3.8	(1.41)	
3	81	2.7	(1.20)	81	3.5	(1.28)	66	4.4	(1.18)	53	4.8	(1.05)	44	5.0	(1.09)	
4	69	3.2	(1.52)	69	4.1	(1.46)	58	4.9	(1.34)	51	5.5	(1.45)	44	5.7	(1.62)	
5	30	3.9	(1.28)	29	4.6	(1.02)	23	5.5	(1.12)	20	6.0	(0.89)	19	6.3	(1.06)	
Between groups																
p-value		<0.001			<0.001			<0.001			<0.001			<0.001		

Table 16(a). Prelingual: SIR Correlations With Factors Affecting Outcomes (Education Level-Father, Education Level-Mother & Household Income) From 2008 To 2018

Variables	SIR SCORE FROM 2008 TO 2018															
	6 months			12 months			24 months			36 months			48 months			Within-subjects
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	p-value
Overall	268	1.5	(0.82)	238	1.9	(0.94)	193	2.4	(1.03)	158	2.6	(1.10)	136	2.9	(1.25)	0.245
Education level - Father																0.836
Non-Tertiary	145	1.6	(0.80)	129	1.9	(0.92)	105	2.3	(1.03)	89	2.6	(1.04)	76	2.9	(1.20)	
Tertiary	101	1.6	(0.91)	91	1.9	(1.01)	73	2.4	(1.08)	58	2.8	(1.17)	50	3.0	(1.29)	
Between-groups																
p-value		0.679			0.653			0.568			0.191			0.749		
Education level - Mother																0.952
Non-Tertiary	145	1.5	(0.81)	127	2.0	(0.91)	106	2.4	(1.02)	94	2.6	(1.08)	80	3.0	(1.22)	
Tertiary	99	1.7	(0.89)	90	1.9	(1.02)	70	2.4	(1.08)	52	2.7	(1.15)	45	2.9	(1.29)	
Between-groups																
p-value		0.166			0.440			0.995			0.900			0.603		
Household income																0.547
Less than RM1000	22	1.5	(0.51)	19	1.7	(0.73)	16	1.9	(0.85)	13	2.2	(0.99)	10	2.2	(1.23)	
RM1000 and above	232	1.6	(0.83)	205	2.0	(0.94)	163	2.4	(1.04)	133	2.7	(1.08)	115	3.0	(1.20)	
Between-groups																
p-value		0.567			0.315			0.059			0.071			0.039		

Table 16(b). Prelingual: SIR Correlations With Factors Affecting Outcomes (CI Aided & FPR At 12 Months) From 2008 To 2018

SIR SCORE FROM 2008 TO 2018																
Variables	**6 months**			**12 months**			**24 months**			**36 months**			**48 months**			Within-subjects
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	p-value	n	Mean	(SD)	n	Mean	(SD)	p-value
CI Aided																0.973
<40dB	212	1.6	(0.82)	195	2.0	(0.96)	159	2.4	(1.05)	131	2.7	(1.09)	113	3.0	(1.25)	
>41dB	50	1.4	(0.84)	40	1.7	(0.80)	32	2.0	(0.80)	27	2.2	(1.05)	23	2.7	(1.23)	
Between-groups																
p-value		0.266			0.088			0.027			0.035			0.262		
FPR at 12 months																0.527
1 – Limited	12	1.1	(0.67)	11	1.4	(0.81)	10	1.7	(0.95)	6	2.2	(1.17)	6	2.2	(1.17)	
2 – Below average	45	1.3	(0.66)	45	1.4	(0.61)	33	1.6	(0.61)	26	1.8	(0.75)	21	1.8	(0.81)	
3 – Average	81	1.4	(0.61)	81	1.7	(0.66)	66	2.2	(0.85)	53	2.3	(0.81)	44	2.7	(1.00)	
4 – Good	69	1.7	(0.90)	69	2.3	(1.05)	58	2.8	(1.02)	51	3.1	(1.16)	44	3.4	(1.23)	
5 – Ideal	30	2.2	(1.12)	29	2.6	(1.05)	23	3.1	(1.00)	20	3.4	(0.94)	19	3.8	(1.13)	
Between-groups																
p-value		<0.001			<0.001			<0.001			<0.001			<0.001		

Table 17. Prelingual: Outcome Measurement Scores At 36 Months In Association With Parents Education Level

Variables	OUTCOME MEASUREMENT SCORES AT 36 MONTHS FOR SECOND COHORT																	
	MAIS			MUSS			LiP			PEACH			CLF			WORD COUNT		
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)
Overall	25	36.6	(6.62)	23	30.3	(10.78)	23	39.1	(3.87)	12	75.6	(25.70)	63	21.9	(13.30)	41	147.8	(222.15)
Education level - Father																		
Non-Tertiary	11	33.5	(9.07)	10	26.4	(15.18)	11	37.9	(5.13)	4	82.4	(11.79)	32	19.7	(12.20)	19	107.5	(214.59)
Tertiary	12	38.8	(1.85)	11	33.5	(4.23)	10	40.2	(1.81)	6	68.6	(35.31)	24	26.5	(15.24)	17	210.4	(251.17)
Between-groups p-value	0.057			0.150			0.197			0.479			0.069			0.194		
Education level - Mother																		
Non-Tertiary	11	37.4	(2.80)	11	32.0	(9.64)	11	39.5	(3.45)	6	82.2	(13.59)	29	20.4	(11.85)	20	121.1	(206.04)
Tertiary	11	35.3	(9.67)	9	28.1	(13.89)	9	38.7	(5.05)	4	64.2	(42.98)	27	24.6	(15.20)	17	175.7	(247.92)
Between-groups p-value	0.499			0.470			0.650			0.356			0.263			0.469		

Table 18. Prelingual:Outcome Measurement Scores At 36 Months In Association With Household Income And CI Aided Thresholds

Variables	OUTCOME MEASUREMENT SCORES AT 36 MONTHS FOR SECOND COHORT																	
	MAIS			MUSS			LiP			PEACH			CLF			WORD COUNT		
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)
Household income																		
Less than RM1000	0	-	-	0	-	-	0	-	-	0	-	-	5	15.8	(5.22)	4	15.0	(2.45)
RM1000-2499	6	36.5	(2.88)	6	34.7	(4.50)	7	39.4	(4.28)	4	89.2	(8.17)	18	20.8	(10.22)	13	208.1	(259.57)
RM2500-4999	10	34.3	(9.92)	10	25.3	(14.39)	8	39.4	(2.56)	3	71.2	(12.52)	20	21.2	(13.81)	11	44.6	(49.11)
RM5000 and above	8	39.0	(1.41)	6	33.5	(5.13)	6	39.8	(1.72)	4	62.5	(41.43)	16	28.3	(16.57)	11	236.5	(282.16)
Between-groups p-value		0.496			0.278			0.393			0.551			0.134			0.154	
CI Aided																		
<40dB	24	36.7	(6.72)	22	30.3	(11.03)	21	40.1	(2.00)	11	74.2	(26.47)	53	23.4	(13.62)	36	142.1	(195.13)
>41dB	1	33.0	-	1	31.0	-	2	28.5	(2.12)	1	90.9	-	10	13.9	(7.87)	5	189.4	(397.31)
Between-groups p-value		-			-			<0.001			-			0.037			0.661	

Table 19. Prelingual:Outcome Measurement Scores At 36 Months In Association With FPR & CI Usage

Variables	OUTCOME MEASUREMENT SCORES AT 36 MONTHS FOR SECOND COHORT																	
	MAIS			MUSS			LiP			PEACH			CLF			WORD COUNT		
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	n
Family Participation Rating																		
1 – Limited	0	-	-	0	-	-	0	-	-	0	-	-	0	-	-	0	-	-
2 – Below average	0	-	-	0	-	-	1	27.0	-	0	-	-	13	10.7	(4.52)	6	21.3	(9.42)
3 – Average	14	34.7	(8.38)	13	26.8	(12.92)	10	37.9	(3.38)	5	70.5	(38.60)	27	18.0	(8.25)	22	61.0	(90.44)
4 – Good	7	38.9	(2.04)	6	34.5	(4.89)	8	40.9	(1.46)	5	80.5	(8.74)	20	30.6	(12.87)	11	326.5	(306.65)
5 – Ideal	3	38.7	(1.53)	3	37.0	(4.36)	3	41.3	(1.15)	1	95.5	-	3	47.7	(11.59)	2	500.0	(0.00)
Between-groups																		
p-value	0.195			0.080			0.001			0.378			<0.001			<0.001		
CI Usage At 6 Month																		
Less than 8 hours	9	36.8	(8.94)	8	30.1	(10.15)	7	38.3	(5.38)	6	65.9	(33.49)	26	16.1	(6.14)	20	76.1	(96.81)
8 to 11.9 hours	10	34.6	(5.95)	9	26.4	(13.37)	11	38.5	(3.39)	4	81.8	(11.13)	19	22.4	(14.50)	11	258.3	(335.16)
12 hours and More	4	39.3	(0.96)	4	35.8	(2.87)	3	41.0	(1.73)	2	92.0	(4.82)	8	27.9	(12.74)	7	218.6	(248.77)
Between-groups																		
p-value	0.516			0.385			0.602			0.425			0.019			0.073		

POST-LINGUAL GROUP

Demographic and Clinical Characteristics

There were 97 post-lingual patients who underwent cochlear implantation in the period between 2008-2018. 40 patients were implanted in the first five years (2008-2012) of the programme and 57 patients were implanted in the second five years (2013-2018). Majority 94 (96.9%) patients had unilateral implantation and 3 (3.1%) patients had bilateral implantation.

Out of these 97 patients, 47 were male and 50 were female. By ethnicity, 59 (60.8%) were Malay, 27 (27.8%) were Chinese, 5 (5.2%) were Indian and 6 (6.2%) were other races. Age at implantation for the post-lingual group ranged from 5.6 to 63.2 years old. 67.0% had undergone implantation at age of 19 years old and above, 16.5% between 13 to 18 years old and 16.5% between 5 to 12 years old.

Of 97 patients, 83 patients had acquired hearing loss and 14 patients had congenital progressive hearing loss. The most common etiology of acquired hearing loss was idiopathic with 59 cases (60.8%). Other etiologies were meningitis with 7 cases (7.2%), post motor vehicle accident 6 cases (6.2%), chronic otitis media with or without cholesteatoma 4 cases (4.2%) and following non-specific high grade fever 4 cases (4.2%). Other less common reported etiologies were labyrinthitis, sudden sensorineural hearing loss (SSHL) and noise induced hearing loss (NIHL).

There were six patients with first degree family history of hearing loss with two of them were siblings with Usher syndrome and one with Klippel-Fiel syndrome. All ears implanted had normal inner ear anatomy except in three patients with IP-II, cochlear ossification and cochlear nerve hypoplasia. All patients had full insertion of the electrodes including three patients with inner ear anomalies. These were summarized in Table 20.

Table 20. Postlingual:Demographic And Clinical Characteristics Of Post-Lingual CI Recipients 2008-2018

Variables	Overall 2008-2018		(Cohort 1) 2008-2012		(Cohort 2) 2013-2018	
	n	(%)	n	(%)	n	(%)
	97		40		57	
Laterality						
Unilateral	94	(96.9)	38	(95.0)	56	(98.2)
Bilateral	3	(3.1)	2	(5.0)	1	(1.8)
Gender						
Male	47	(48.5)	23	(57.5)	23	(41.8)
Female	50	(51.5)	17	(42.5)	32	(58.2)
Ethnic						
Malay	59	(60.8)	33	(82.5)	25	(45.5)
Chinese	27	(27.8)	5	(12.5)	22	(40.0)
Indian	6	(6.2)	2	(5.0)	3	(5.5)
Others	5	(5.2)		-	5	(9.1)
Aetiology						
Idiopathic	59	(60.8)				
Meningitis	7	(7.2)				
Post MVA	6	(6.2)				
COM ±cholesteatoma	4	(4.2)				
Non specific high grade fever	4	(4.2)				
Labyrinthitis	1	(1.0)				
Sudden SNHL	1	(1.0)				
NIHL	1	(1.0)				
Progressive HL	14	(14.4)				
Mean Age Of Diagnosis (Year)						
	27.9		23		31.5	
(SD)	(15.9)		(14.4)		(16.1)	

Surgical And Device Related Complications

There were 100 ears operated in 97 patients in the post-lingual group. All 100 ears were analyzed for surgical outcome measures. Intra-operatively we encountered perilymph gusher in one patient with normal radiological findings, which was successfully managed during surgery.

Of 100 ears, 7 ears had surgical complications, in which 5 were major and 2 were minor, as shown in the Table 21. Overall complication rate for post-lingual group was 7%.

Table 21. Post-Lingual: Surgical And Device Related Complications

Surgical Complications		n
Major complication	Electrode migration	3
	Device failure	1
	Device rejection	1
Minor complication	Facial nerve paresis/palsy	1
	SSI minor	1
	Total	7

Major Complications

i) Electrode migration

Three patients had electrode migration that occurred at 1, 3 and 14 months following surgery. Preoperative radiological imaging and intraoperative findings were normal in all of them except for one patient who had middle ear effusion intraoperatively. None had any history of trauma or head injury. All had uneventful explantation and reimplantation.

ii) Device failure

There was hard failure in one patient, which occurred 14 months after the surgery. There was no history of trauma or head injury prior to the event. The patient had undergone an uneventful explantation-reimplantation with a new device.

iii) Device rejection

One case of device rejection was encountered in our series. Eight months following implantation the patient developed recurrent post auricular swelling, which was controlled with antibiotics. However nine months later, serous discharge was noted from the recurrent swelling and subsequently was complicated with skin breakdown, exposing the implant receiver. Despite vigorous dressing and antibiotics the skin breakdown was never healed. Four different procedures of skin flaps were attempted within 2 years period but to no avail due to unhealthy friable skin tissues surrounding the implant. Explantation of the device was performed upon patient requests. The wound healed well following the explantation. Histopathology report of the skin tissue showed acute on chronic inflammation with no growth noted.

Minor complications

There were two minor complications reported. One patient developed delayed facial paresis a week following the surgery, which recovered completely with conservative measures. One patient had minor surgical site infection (SSI) wound infection, which was managed with intravenous antibiotic and wound dressing.

Functional Outcomes

i) Aided hearing threshold

Of 97 patients in the post-lingual group, two patients were excluded from aided threshold outcome measures due to missing data.

Mean aided threshold was measured in four frequencies average (500Hz, 1000Hz, 2000Hz and 4000Hz). The mean aided hearing threshold with hearing aid prior to implantation was 80 dBHL. The mean aided threshold at 6 month post implantation of less than or equal to 40 dBHL was observed in 86 patients (90.5%). The remaining 9 patients (9.5%) showed mean aided threshold above 40 dBHL ranging from 41.25 dBHL to 58.7 dBHL as in Table 22.

Table 22. Post-Lingual: Mean Aided Thresholds 6 Months Post Implantation

	Mean Aided Threshold (dBHL)	n	%
Pre implantation	80 dBHL	95	100
Post implantation	≤ 40 dBHL	86	90.5
	> 41 dBHL	9	9.5

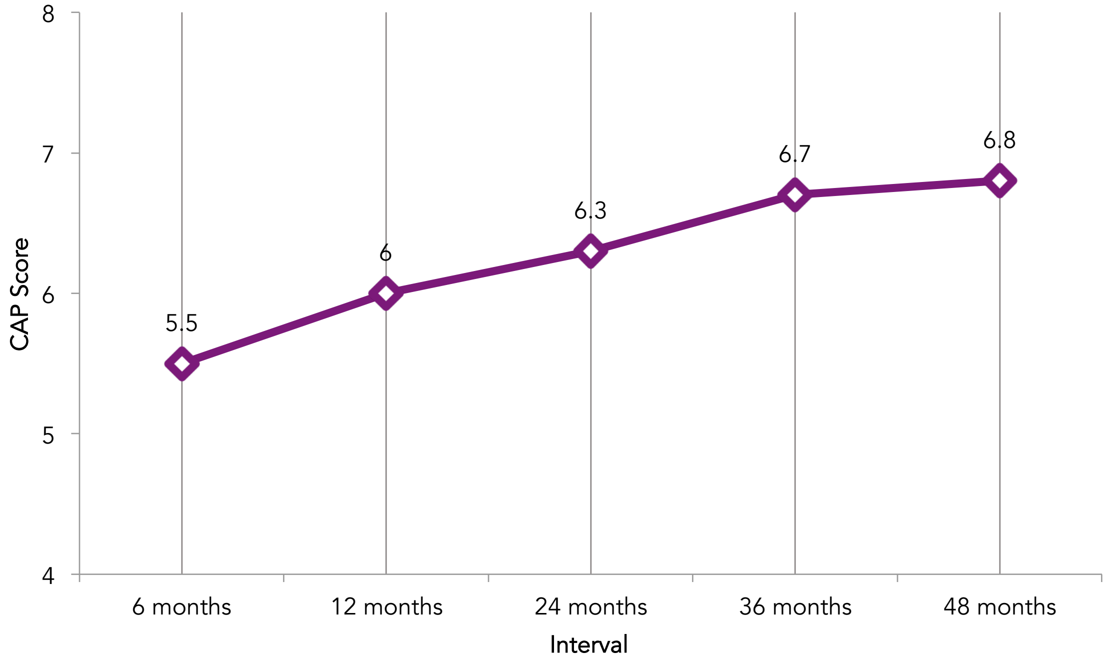
ii) Categorical Auditory Performance II (CAPII)

CAPII scores were measured at 6, 12, 24, 36 and 48 months post implantation. Out of 97 patients, 85 were included in CAPII score analysis. Six patients were excluded due to incomplete data. Four patients were also excluded because their CI age had not met the minimum 6 months post implantation assessment. Of 85 patients, 58 of them had reached at least 4 years of implantation age. Overall mean CAPII score showed improvement from 5.5 to 6.8 in post-lingual recipients as in Table 23 and Figure 12.

Table 23. Post-Lingual: Mean CAPII Score Post Implantation Up To 48 Months

	Post Implantation Interval				
	6 months	12 months	24 months	36 months	48 months
n	85	78	68	63	58
Mean CAPII score	5.5	6.0	6.3	6.7	6.8
SD	1.32	1.34	1.31	1.61	1.63

Figure 12. Post-Lingual: Mean CAPII Score Post Implantation Up To 48 Months



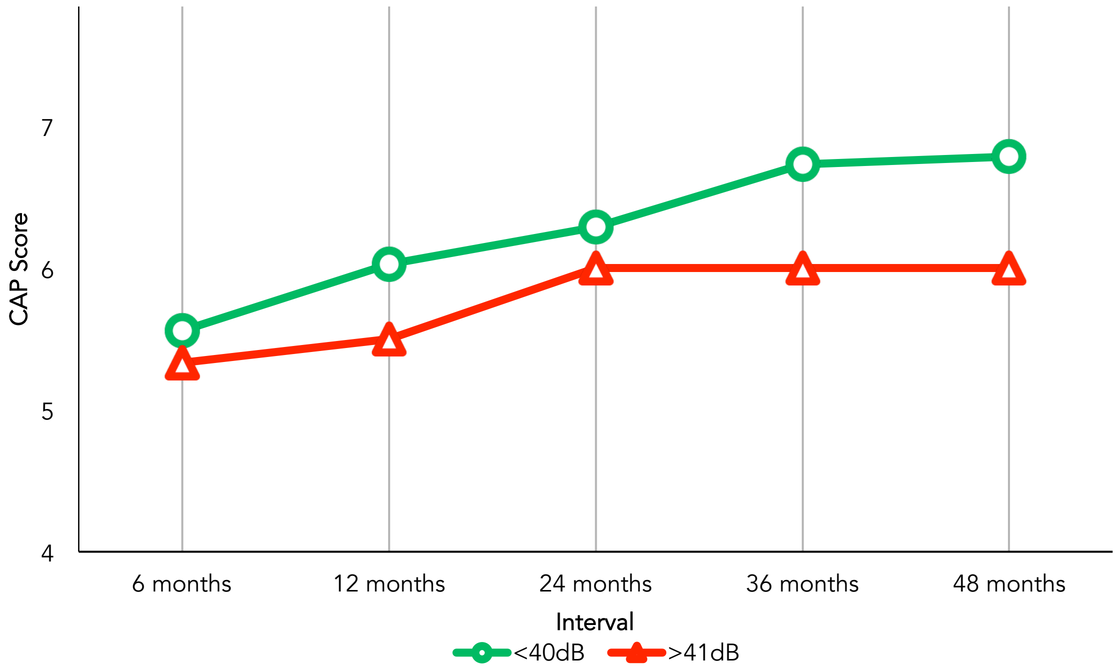
Factors Affecting Outcomes

Patients with aided threshold of 40dBHL and lower had higher mean CAPII score compared to patients with aided threshold of 41dBHL and above at all follow up intervals as shown in Table 24 and Figure 13.

Table 24. Post-Lingual: CAPII Score Post Implantation In Correlation With Aided Threshold

	MEAN CAPII SCORE FROM 2008 TO 2018															
Interval	6 months			12 months			24 months			36 months			48 months			Within-subjects
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)	p-value
OVERALL	85	5.5	(1.32)	78	6.0	(1.34)	68	6.3	(1.31)	63	6.7	(1.61)	58	6.8	(1.63)	0.031
CI AIDED																0.999
≤40dB	79	5.6	(1.33)	74	6.0	(1.34)	65	6.3	(1.33)	60	6.7	(1.64)	56	6.8	(1.64)	
>41dB	6	5.3	(1.37)	4	5.5	(1.29)	3	6.0	(1.00)	3	6.0	(1.00)	2	6.0	(1.41)	
Between-groups																
p-value	0.692			0.447			0.709			0.447			0.507			

Figure 13. Post-Lingual: CAPII Score Post Implantation In Correlation With Aided Threshold



EDUCATIONAL STATUS

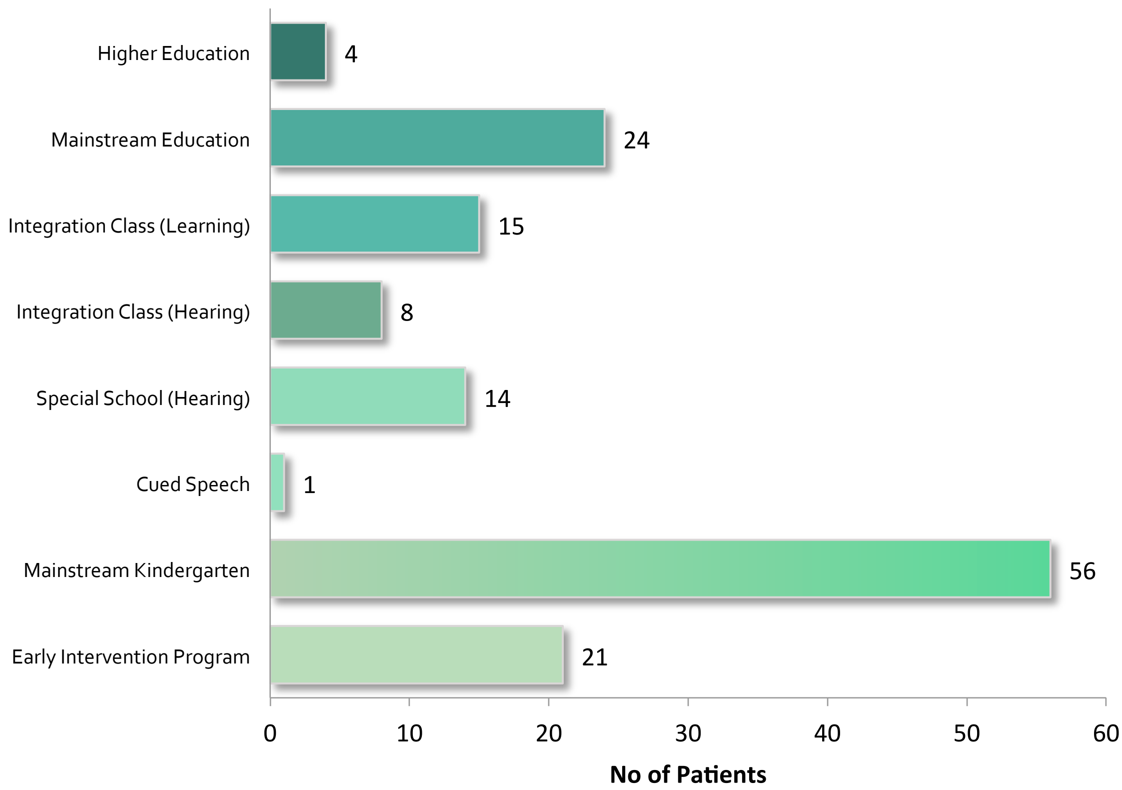
The educational status of 143 CI recipients in the year 2018 from the second five years cohort is reviewed. Their educational status of the recipients is as follows (Figure 14):

i) Preschool: Total 77 of who 56 are in mainstream kindergarten and 21 are in Early Intervention Program Centre (EIP).

ii) Formal Schooling: Total 62 school age recipients of whom 24 are in mainstream education, 8 are in integration classes (hearing), 15 are in integration classes (learning), 14 are in special school placement and 1 is in cued speech school.

iii) Higher Education: Total 4 recipients.

Figure 14. Educational Status Of CI Recipients 2013-2018



QUALITY OF LIFE

a) Abbreviated Profile of Hearing Aid Benefit (APHAB)

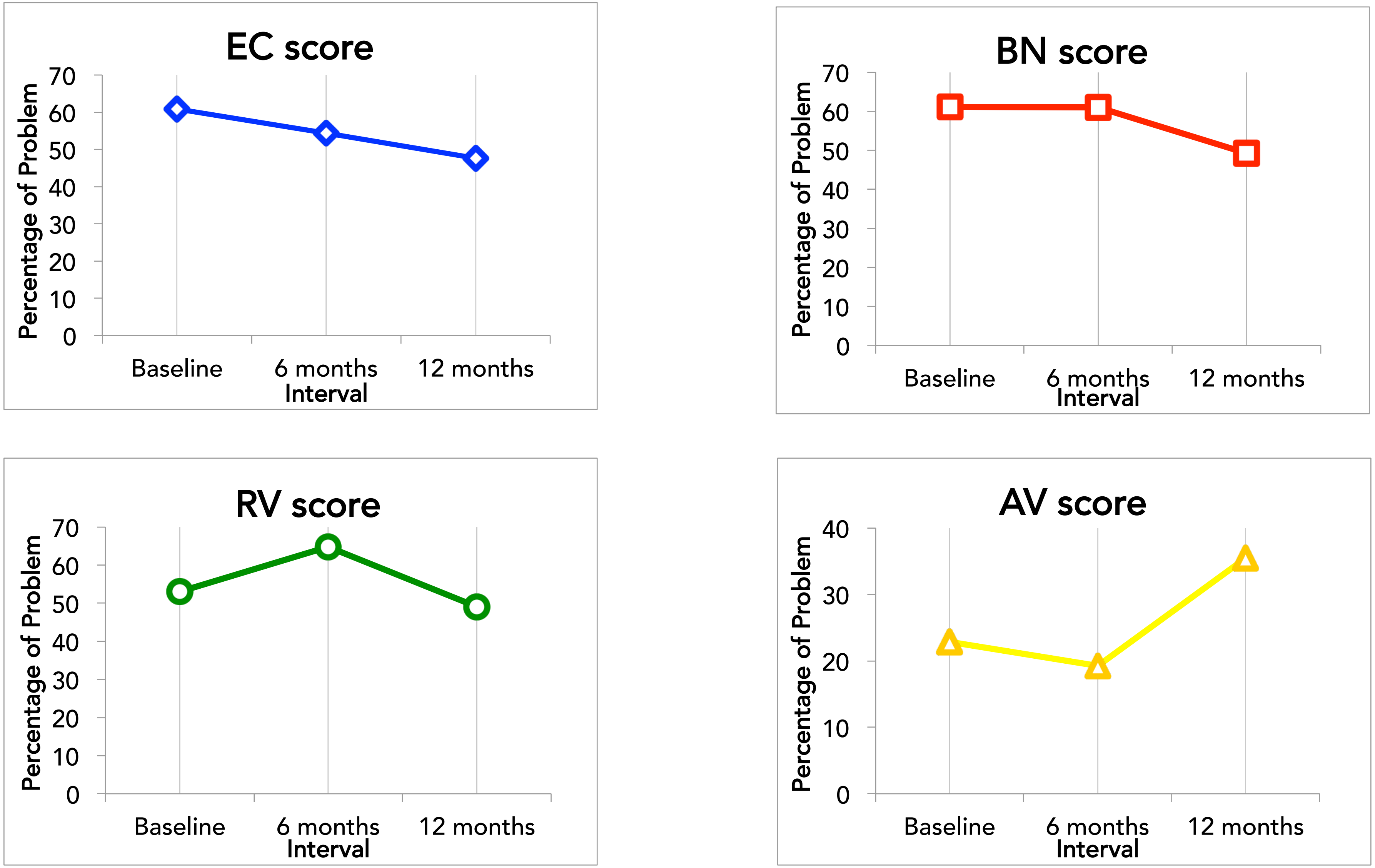
APHAB is used to measure the amount of difficulty faced by patients with hearing impairment in communication or noises in various everyday situations before and after amplification. We used APHAB to analyze the difficulty faced by patients at baseline and at 12 months post implantation in 28 post-lingual adult patients from the second cohort.

The mean percentage of problems for three subscales, Ease Of Communication (EC), Background Noise (BN) and Reverberation (RV) were reduced after implantation in comparison to baseline. These scores reflect of better benefit perceived by patients following cochlear implantation. However, for Aversiveness (AV) subscale, the mean percentage reduced at 6 months post implantation and increased again at 12 months post implantation. These were shown in Table 25 and Figure 15.

Table 25. Post-Lingual: APHAB Score Post Implantation 2013-2018

	APHAB								
Interval	**Baseline**			**6 months**			**12 months**		
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)
SUBSCALES									
EC score	28	60.8	(28.70)	22	54.3	(21.97)	14	47.6	(17.29)
BN score	28	61.1	(29.45)	19	60.9	(19.23)	11	49.2	(14.35)
RV score	28	53.1	(29.76)	19	64.8	(17.17)	11	49.1	(25.49)
AV score	28	22.8	(35.03)	19	19.2	(28.97)	11	35.5	(30.02)

Figure 15. Postlingual:APHAB Score Of Each Subscale Post Implantation 2013-2018



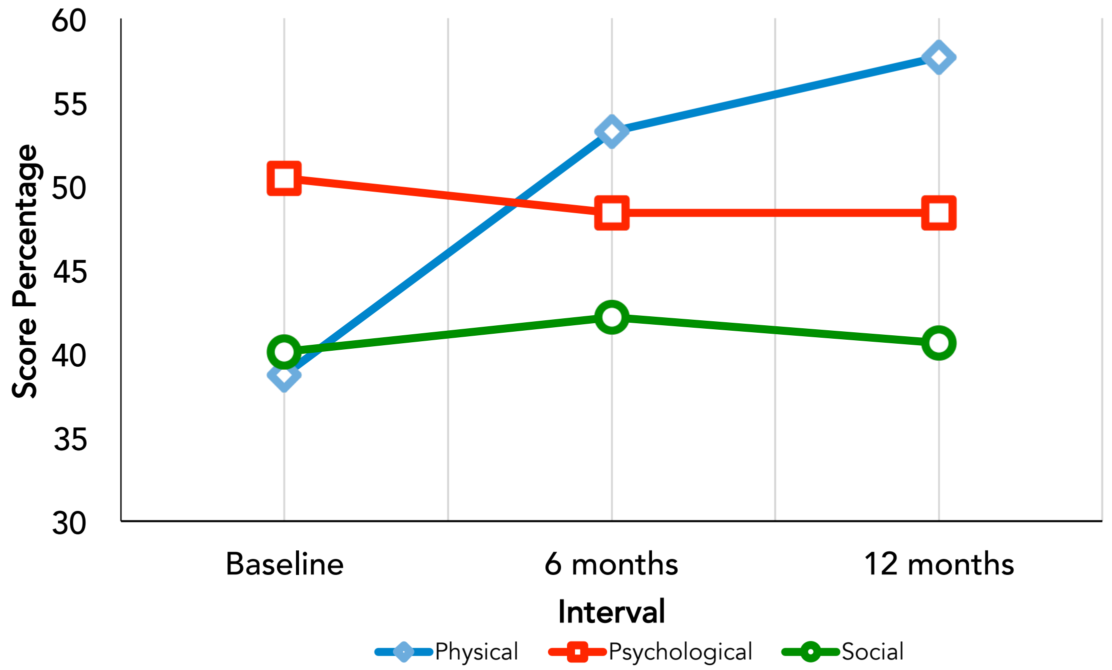
b) Nijmegen Cochlear Implant Questionnaire (NCIQ)

We evaluated health related quality of life (QoL) in 21 post-lingual adult patients from the second cohort using NCIQ. There was marked improvement in mean percentage of physical subdomain (basic sound perception, advanced sound perception, speech production) at 6 months and 12 months post implantation. However, there were no changes in psychological (self-esteem) and social (activity limitations and social interactions) subdomains as shown in Table 26 and Figure 16.

Table 26. Post-Lingual: NCIQ Score Post Implantation 2013-2108

	NCIQ Score								
Interval	Baseline			6 months			12 months		
	n	Mean	(SD)	n	Mean	(SD)	n	Mean	(SD)
SUBDOMAIN									
Physical	21	38.7	(17.71)	17	53.3	(17.09)	17	57.7	(26.79)
Psychological	21	50.5	(25.12)	17	48.4	(11.46)	17	48.4	(21.50)
Social	21	40.1	(26.04)	17	42.2	(12.65)	17	40.6	(25.05)

Figure 16. Post-lingual: NCIQ Score Post Implantation 2013-2018



DISCUSSION
Chapter 5

DISCUSSION

Since the inception of the cochlear implant programme in MOH in 2008, a total of 380 individuals have been implanted (2008-2018). In the second five years (2013-2018) the numbers of recipients has doubled compared to the first five years (2008-2012).

The MOH National CI Programme is one of its kind whereby it is a structured and centralised programme serving the whole country under the MOH facility. All potential candidates for cochlear implant are meticulously screened and prepared at the satellite hospitals prior to final selection by the central committee at regular intervals. Upon approval, surgery and intensive rehabilitations will take place in the respective satellite hospitals. Their progress and success are monitored periodically.

In the past ten years, we have increased the number of professionals; audiologists, speech-language therapists and trained CI surgeons to expand the service. As a result of this, the number of satellite hospitals has increased from eight to ten. We have published the First 5 year CI Outcome Report in 2016[4] and several papers based on the programme.

In this review, we compared the mean age of implantation from the first five year and second five year cohort. The mean age of surgery in the second five years was reduced to 38.6 months of age compared to the first five years, which was 42.9 months. Despite that, the mean age at surgery was higher than that of developed countries. The mean age of surgery in Netherlands was reduced to 14 months old from 28 months following the introduction of newborn hearing screening (NHS) program[25]. Similar findings were obtained from the CI program of Chicago which shortened the mean age of implantation to 20 months old after strengthening their NHS program[26].

Availability of NHS is a significant contributory factor resulting in children implanted at an earlier age[27,28]. Research worldwide has shown that the age at which children are implanted is an important factor in the development of speech perception and intelligibility. In 2008, when we first embarked on the CI programme, NHS was only available in ten MOH hospitals. To date there are 38 MOH hospitals with NHS program. Of these, 17 have UNHS program and 21 have HRNHS program. The percentage hearing loss detection through NHS has increased in the second five years. However, overall percentage of detection through NHS program in this 10 years report was still low and this is of considerable concern. Still, majority of the prelingual recipients' detection of hearing loss were through passive detection. Low level of awareness on NHS program and impact of hearing loss in children among parents were possible reasons and might lead to delay and unsuccessful interventions[29–31]. Therefore it is important to further strengthen and expand the NHS program in the MOH facilities and to improve the level of awareness on impact of hearing loss throughout the country.

Indications for cochlear implantation continue to expand with the advancement of CI technology and evidence of positive impact of CI worldwide. These criteria include those with inner ear anomalies, implantation at early age (12 months and below), those with additional disabilities, bilateral implantation and implantation in single sided deafness[32–35].

In current practice, presence of inner ear malformations is no more an absolute contraindication for cochlear implantation. Studies have shown that up to 35% of prelingual CI candidates have inner ear malformation[36]. Children with bilateral inner ear malformations accounted for about 65% of whom 93% had mirrored anomalies of right and left inner ears structures[37]. In the beginning, our recipients were mostly patients with normal inner ear. In keeping with these expanding criteria for CI, in the second cohort we have implanted 16 patients with inner ear malformations, incomplete partition type I (IPI) and incomplete partition type II (IPII), common cavity, enlarged vestibular aqueduct and cochlear nerve hypoplasia. There were also four recipients with acquired hearing loss following meningitis and labyrinthitis ossificans, a condition in which the cochlea becomes ossified and may result in incomplete or failed electrode insertion. In these groups of patients, preoperative imaging evaluation is important to assist in preoperative planning regarding choice of electrode array, technique for surgery and to anticipate complications[38].

Majority of recipients in the program received unilateral implantation primarily due to resources limitation. This is also practiced in other developing countries[38–40]. Szyfter et al. 2019 suggested priority for unilateral implantation to benefit as many candidates possible before bilateral implantation is considered when the funding is limited[32].

Basis for bilateral implantation is for binaural hearing allowing recipients to hear more effectively in noisy environments, minimise problems with head shadow effect and better sound localisation[40,41]. In this first 10 years review, we have 33 patients who had undergone bilateral cochlear implantation. Time interval between the first and the second surgery in sequential implantation was between 4 to 67 months. Simultaneous and short interval implantation compared to long interval implantation have a noticeable advantage in hearing and speech outcomes[42,43].However even long interval sequential implantation of greater than 2 years and up to 10 years have been found to be beneficial compared to unilateral implantation[44–47].These benefits on bilateral implantation continued to increase with time after bilateral implantation regardless of the duration[44,45,48–50].

The number of CI recipients was doubled in the second five years of this 10 years review. This reflects the good support we have had from the MOH right from the beginning of the program. Studies by Khan et al. (2007) and Chundu et al. (2013) reported that the biggest limiting factor for cochlear implantation in developing countries was the costs of getting and maintenance of the device and the rehabilitation costs after the implantation[39,51]. The level of financial support from their governments in these developing countries will impact the recipients and the CI programme[39,51–54].

Since the establishment of the program to date, the main funder for device procurement is by the MOH CI Programme allocation. Other sources of funding were obtained from JPA, TBP, NGO, zakat, insurance and self-funding. There is concern about the waiting period between funding and surgery in this review. The median interval between the central committee approval and the surgery were 3.3, 4.4 and 4.6 months from the MOH allocation, JPA and TBP respectively in the prelingual group for the second five-year of the program (2013-2018). The waiting period can be minimised if the procurement process and funding approval can be expedited. Funding for second implant is limited and mainly from self-funding. The CI also incurs substantial costs associated with maintenance of the external device and long term rehabilitation process, which are borne by the recipients and caregivers throughout their lifetime.

Surgical Outcomes

Cochlear implantation is a widely accepted and safe procedure for severe to profound hearing loss, and is associated with relatively low rate of complications[5,55–57]. In this report we use the consensus on CI complications proposed by Hansen et al. (2010). Overall, our CI surgeries have shown to be safe with low complication rate of 5.57%. This complication rate is comparable with other large series of 5-20%[5,55–57].

We reported perilymph gusher as an intraoperative finding, rather than a complication. All 9 cases were successfully managed during the surgery. There was no encounter of severe CSF gushers that needed further measures such as lumbar drainage. More than half of these patients had abnormal radiological findings; 2 patients with IP type II, 2 patients with IP type I and one patient with enlarged vestibular aqueduct syndrome. The other four patients had normal radiological findings, which indicate that the surgeon should be ready to encounter the condition should it arise during surgery.

Cochlear implantation is associated with a low incidence of major complications, including flap necrosis, device failure, electrode migration, bleeding, permanent facial weakness, otitis media and persistent perilymph leakage complications of cochlear implant surgeries in both children and adults[5,55–58]. In our review, we encountered device failure, electrode array migration, device rejection, surgical site infection (SSI) and transient facial nerve paresis.

1) Device failure

Device failure is divided into hard and soft failures. Hard failure has been previously defined as a CI that is out of compliance with manufacturer's specifications and which provides poor performance. Soft failure is a diagnosis of exclusion identified after extensive medical workup, reprogramming, external hardware exchange and integrity testing by the manufacturer fails to identify an etiology[5,59,60].

Device failure was the most common causes of explanation and reimplantation with the rate of 1.7% in our program. Seven patients had this complication, which occurred at 1, 3, 5, 8, 14, 16 and 52 months following switch-on. Of these, four had hard failures and three had soft failures.

2) Electrode migration

Electrode migration may occur more commonly than was once thought. Rivas et al. in (2007) described electrode migration as an important leading cause of cochlear re-implantation, second to device failure. The clinical symptoms associated with electrode migration include change of sound, poor auditory performance, aversive stimuli such as pain, shocks and facial nerve stimulation. It may be associated with ossified cochleae and cochlear malformations, but most subjects had not associated with any reported etiologic factor[42,61].

In our series electrode migration is the second common major complication. We encountered 5 cases of electrode migration that occurred at 1, 3, 6, 12 and 14 months post operatively. All the patients had shown a decreased in auditory performance at time when the migration of electrode array were confirmed. There were no documented history of trauma and HRCT scans did not show any evidence of temporal bone trauma or cochlear ossifications that may have led to the migration of the electrodes. None of our cases have associated factors that may lead to electrode migration as mentioned above, which is comparable to other studies. All our patients with electrode migration have undergone uneventful repositioning of electrode array or reimplantation.

3) Device rejection

We encountered one case of device rejection in our series, which started to manifest eight months following implantation in post-lingual group. Literature review showed few cases of similar complication[62,63].The explanation of this rare incident could be attributed to foreign body reaction, contact dermatitis or allergy to the cochlear implant materials. Foreign body reaction is diagnosed with typical histological findings and clinical history. Explantation of the device was performed upon patient request. The wound healed well following explantation.

4) Surgical Site Infection (SSI)

Infection is always a major concern in CI surgery. In our series overall SSI rate was 1.9 %, involving 8 patients. This rate is comparable and within 1.7-20% with other reported series of infection in CI surgery[5,56,64,65].

We had two children with major SSI. One child required explantation and reimplantation with a new implant device as the SSI had affected the integrity of the implant. Minor SSI occurred more commonly in children than adult among our recipients and none had issued with theirs auditory and speech performance during the episode of SSI.

5) Facial Nerve Paresis

Facial nerve injury is one of the dreaded complications in otological procedures including cochlear implant. None of our patients had intraoperative facial nerve injury because we practice intraoperative facial nerve identification in all our implanted cases to ensure the integrity of the nerve and facilitated by the usage of facial nerve monitor in all the cases.

However, we had two patients with postoperative delayed facial nerve paresis. These two cases had no inner ear anomalies, which could be associated with abnormal course of facial nerve and presented without any clinical features of ear or systemic infection. The paresis recovered completely within few weeks after treatment with corticosteroids in both of the patients. We did not encounter permanent facial nerve palsy or facial nerve stimulation post operatively.

There was no documentation of other complications in our series as reported in other studies such as excessive intraoperative bleeding, vertigo, labyrinthitis, meningitis, cholesteatoma or magnet migration[5,55,56,59].

Functional Outcomes for The Prelingual Group

1) CI Aided Thresholds

We used cut off point of mean CI aided threshold at 40dBHL in our CI programme because at this threshold allows audibility of average input level sounds which are appropriate for people with severe hearing loss[6,66,67]. This is consistent with the study by Madell et al. (2004) which showed patients with hearing threshold of 40dBHL or better were able to hear at least fifty percent of what is being said[68]. Other studies suggest that aided thresholds of 35dBHL and below provide the best opportunity for the patients to hear the acoustic cues of soft speech as in the speech spectrum[66,67,69]. However, this mean low aided threshold may not be achieved in patients with profound hearing loss or inner ear anomalies due to desensitisation and dead regions as suggested by few studies[66,67].

In this review, mean CI aided threshold for prelingual group was measured at 12 months post implantation meanwhile for post-lingual group it was measured at 6 months post implantation. Generally post lingual adult patients require shorter duration to get stable mapping as compared to children [6,70,71].

Mean CI aided threshold of 40dBHL or lower was associated with better CAPII score in both prelingual and post-lingual patients in this review. These results were in agreement with other studies which report the significant association of high CAPII scores with aided thresholds of 40dBHL and lower in CI recipients[6,66,67,70,71].

Following implantation, the CAPII score and SIR improved with CI age. These findings showed that there was an improvement in auditory performance and speech intelligibility in the majority of our patients after the switch on in both prelingual and post-lingual groups. The significant benefit of CI also can be seen in Current Level of Function (CLF) score and expressive word count produced among the prelingual recipients. These findings are consistent with other studies that found CI as a viable and effective strategy to help children with bilateral sensorineural hearing loss to hear and develop language[43,46,49,61].

2) Age Of Surgery

In the prelingual group, patients implanted at age less than 36 months old performed fairly well in their CAPII score and SIR at the baseline compared to those implanted at age more than 36 months old. They continued to improve further in performance surpassing the recipients whom were implanted at a later age. They were able to achieve a CAPII score of more than 5 at 36 months post implantation. By 48 months post implantation they began to show evidence of understanding conversation without lip reading with a familiar person. At 48 months post implantation these groups were able to express intelligible connected speech to a listener who concentrates and lip reads.

The children implanted at age of 36 months and above have shown improvement in CAPII score and SIR throughout the follow up. At 48 months post implantation, these later age recipients were able to understand simple commands without lip reads and developing to produce intelligible single words when context and lip reading cues were available. However their scores were lower than those children implanted at younger age, i.e. below 36 months old.

The above findings emphasises the positive impact of early implantation that allows the child to get access to sound and spoken language from a younger age, which in turn will facilitate better speech and language development. Our results were compatible with speech and language outcomes as reported by other studies[70–73]. For this review, Integrated Scales of Development (ISD) by Cochlear Limited[18] was used as a checklist to identify Current Level of Function (CLF) of speech and language of prelingual recipients. Our findings showed that cochlear implants enable the recipients to gain better speech and language skills over follow up. The mean CLF was 21.9 months at 36 months post implantation indicates that after three years intensive CI rehabilitation our recipients were able to gain speech and language skill equivalence to 21 months old level. This implies that smaller language gap can be achieved if a child is implanted at a younger age.

Most of studies indicate that the earlier the implantation, the better the language acquisition[73–75]. Manrique et al. (2004)[76] found that children who implanted between ages 2 and 6 years exhibited a slower growth rate and a lag of between 2 and 3 years below achievement levels for hearing age-mates. In this review, we did not find a statistical correlation between the age of implantation and CLF performance. This is probably due to discrepancy of sample size among the age groups. It may not have been sufficient to show differences in the result of children implanted at age below 24 months.

Overall mean word count performance at 36 months post implantation is 197.9 words which indicated that children with cochlear implants demonstrate lower expressive vocabulary count than children with normal hearing. This finding was similar to other studies that indicate children with CI did not develop expressive vocabulary knowledge comparable to their peers[77,78]. However, in this review, we found that children implanted before the age of 36 months old able to attain higher mean expressive vocabulary count which was 386.1 words, compared to older subgroups.

3) Family Participation Rating

Family Participation Rating (FPR) Scale by Moeller (2000)[22] was utilised to categorise the quality of family participation in the intervention program. In this scale, families with level 4 and level 5 were considered above average participation. From our review, higher levels of family participation correlate strongly with better functional outcomes. The parents who achieved these levels were highly involved in learning and advocacy at home. They were also supportive of aspects regarding managing the children. The commitment of parents and caregivers are important in long-term rehabilitation program.

Young children develop their skills through daily experiences and interactions with their parents or caregivers. Hence, parents' or caregivers' involvement in carrying over facilitative techniques as per guided by clinicians are very important.

In Malaysia, centres that promote early intervention program for the hearing impaired are still limited. Therefore, it is crucial to empower the parents, particularly in their role to provide adequate auditory and speech-language input at their home settings.

Bhamjee et al. (2019)[79] reported that continuous parental guidance, assistance and support by CI teams, especially in delivering adequate informational and emotional counseling throughout the CI process may assist parents in better managing and balancing their existing and new-found responsibilities in a more constructive manner. Our review showed that up to 48 months post implantation, about 46% of the family continued to show good (level 4) to ideal (level 5) family participation.

In this review, FPR was the factor that had strong correlation with better CLF score throughout the follow up intervals ($p<0.001$). Higher levels of family involvement from the beginning will lead to better management of a child, especially in facilitating auditory and speech language skills at home. These will definitely enhances the child's overall performance. Spencer,2004[80] mentioned that positive parent involvement during pre implantation period was an indication that was closely associated with the level of parent's post implant involvement with the child's development.

In our review, recipients with higher level of FPR gain better performance in number of word produced. This inferred that family involvement in the intervention program is crucial as parents and family are the main language facilitators.

4) Household Income

In our review at 48 months post implantation interval, the higher household income group had achieved better CAPII score and SIR compared to lower household income group. This raises a concern whether the lower household income group might have difficulty in sustaining long term rehabilitation needs such as attending the rehabilitation sessions, external device maintenance and long term family participation. Other studies reported that financial difficulties were the most difficult challenge faced by recipients from low household income families for device cost and maintenance costs in long term period[39,51,59,79]. These issues have to be addressed in detail by the CI team to facilitate recipients in the low household income group to be evaluated and assisted for social and financial support, as long-term commitment is mandatory in the rehabilitation process of the CI program.

5) Parents' Education Level

Numerous researchers have demonstrated the importance of parental factors on child development. In our series, we studied the outcomes of hearing, speech and language development in association with the parents' educational level and on the rating of family participation in the CI implantee's aural habilitation journey.

Scholars reported a discrepancy in conclusions on the relationship between parental education levels and language performance of children with hearing loss. Some studies showed good association; especially on the level of maternal education level with a positive speech and language performance, however, some other studies have contradicted these[81].

In our series, we found that there is no significant relationship between parents' education levels with all of the outcome measures throughout the test intervals.

6) CI Usage At 6 Month

Consistency in the use of hearing technology is vital for auditory, speech, and language development in children. Therefore, it is crucial to monitor the daily device use among these implantees regularly.

Daily CI usage is defined as "time-on-air" estimate for a patient. This is objectively measured using datalog technology. The datalog system allows the recording of "time-on-air" which takes place while the CI system is on and coil in place.

We examined the correlation between the outcome measures and the recipients datalogs. Although statistically not significant, our review showed that recipients with higher datalogs showed better results compared to other recipients with lesser CI usage (<12 hours). Gagnon et al., 2020 and Easwar et al., 2018, showed that higher CI usage yielded higher receptive language scores. This receptive language outcomes has a significant impact on their future expressive language outcomes[82,83]. We also learned that while device use time is a critical aspect for positive language development, it remains just one of the variables contributing to successful outcomes following cochlear implantation.

7) Mode Of Communication

Communication through the use of spoken language is one of the objectives of cochlear implantation. Majority of our prelingual recipients from the second cohort (2013-2018) after minimum interval of 24 months post implant did use verbal as communication mode 41%, another 49% are using total communication, cued speech and gesture. Only 9% of them ended up with sign language. However this maybe still early for us to make conclusion as study by Watson et al. showed many subjects changed their mode of communication from sign language to oral communication, 5 years after implantation[84].

One recipient is using AAC despite intensive rehabilitation had been diagnosed with additional disability, Autism Spectrum Disorder after the implantation. Studied by Datta et al. showed that profound hearing impaired children with multiple learning difficulties and associated complex need including autism, had negative effect of the functional outcomes. Most of implantees with additional needs were using AAC for their communication mode[85] .

8) Educational Status

Historically, majority of the hearing impaired children especially with severe to profound hearing loss were placed in special schools instead of mainstream. It has been our main objective since the inception of this program to provide them with an opportunity to attend mainstream education in order to optimise their potential and thus, quality of life.

In 2018, 73% of our preschool-age recipients attending mainstream kindergarten. Meanwhile, 76% of our school-age recipients attend normal school, of which 51% are in mainstream while the rest are in integrated program. The condition in normal school is not without challenges because majority of our implantees are unilateral recipients.

Krijger S et al. (2020)[86] in their study found that although children with CI seemingly fit well in mainstream-school they still experience significantly more listening difficulties than normal peers due to multiple factors in the classroom. Among factors that may contribute to these conditions are background noise, distortion of the speech signal, lack of visual support and speaker to implantee's distance in the classroom. They emphasise that awareness among teachers is important to minimise these difficulties[86,87].

Children with CI may benefit from audio assistive listening devices to improve the listening situation in background noise and also for distant hearing in the classroom. Silva et al. (2017)[88], reported that the use of FM system in the classroom has improved the listening situation and understanding instruction among 113 patients with cochlear implant. Thus, to enhance the learning process among hearing-impaired children, the use of this technology could replicate the same results in our classrooms.

Functional Outcomes for The Post-Lingual Group

Similar to the prelingual group, the mean CAPII score achievement in the post-lingual group was increased over time up to 48 months post implantation. Mean aided threshold of 40dBHL or lower was associated with better CAPII score for the post-lingual group in this review. These findings were similar and consistent with other post-lingual CI recipients studies[66,70,71] which report that post-lingual recipients with severe to profound hearing loss achieve good auditory benefit and speech perception post implantation.

Hearing loss in adulthood results in major difficulty in communication and may cause social isolation, depression and negative feelings that can seriously affect personality[89,90]. Difficulties in communication is worse for those with severe and profound hearing loss, possibly affecting personal relationships and even lifestyle[91,92].

In this review the eldest recipient was implanted at the aged of 61 years old whom had bilateral profound hearing loss following meningitis. She became dependent on family members and depressed following the disease. It took her two months after the switch on to adapt to the CI, and became fully independent in activities of daily living for almost 4 years, as she was before the meningitis.

For post-lingual recipients, many studies focused on improvement in quality of life (QoL) after implantation. QoL is a complex task and assessing its improvement in hearing impaired patients is challenging due to its subjectivity. In this review, we analyzed QoL using two questionnaires; APHAB and NCIQ among post lingual adult recipients in the second cohort.

ABHAB scores for post lingual recipients in this review showed a reduction of the mean percentage in three subscales which addressed speech understanding in various everyday environments namely ease of communication (EC), background noise (BN) and reverberation (RV). These findings indicated that the amount of difficulty faced by patients with hearing impairment after the implantation have reduced. However, the result for aversiveness (AV) subscale, which quantifies negative reactions to environmental sounds, was the opposite.

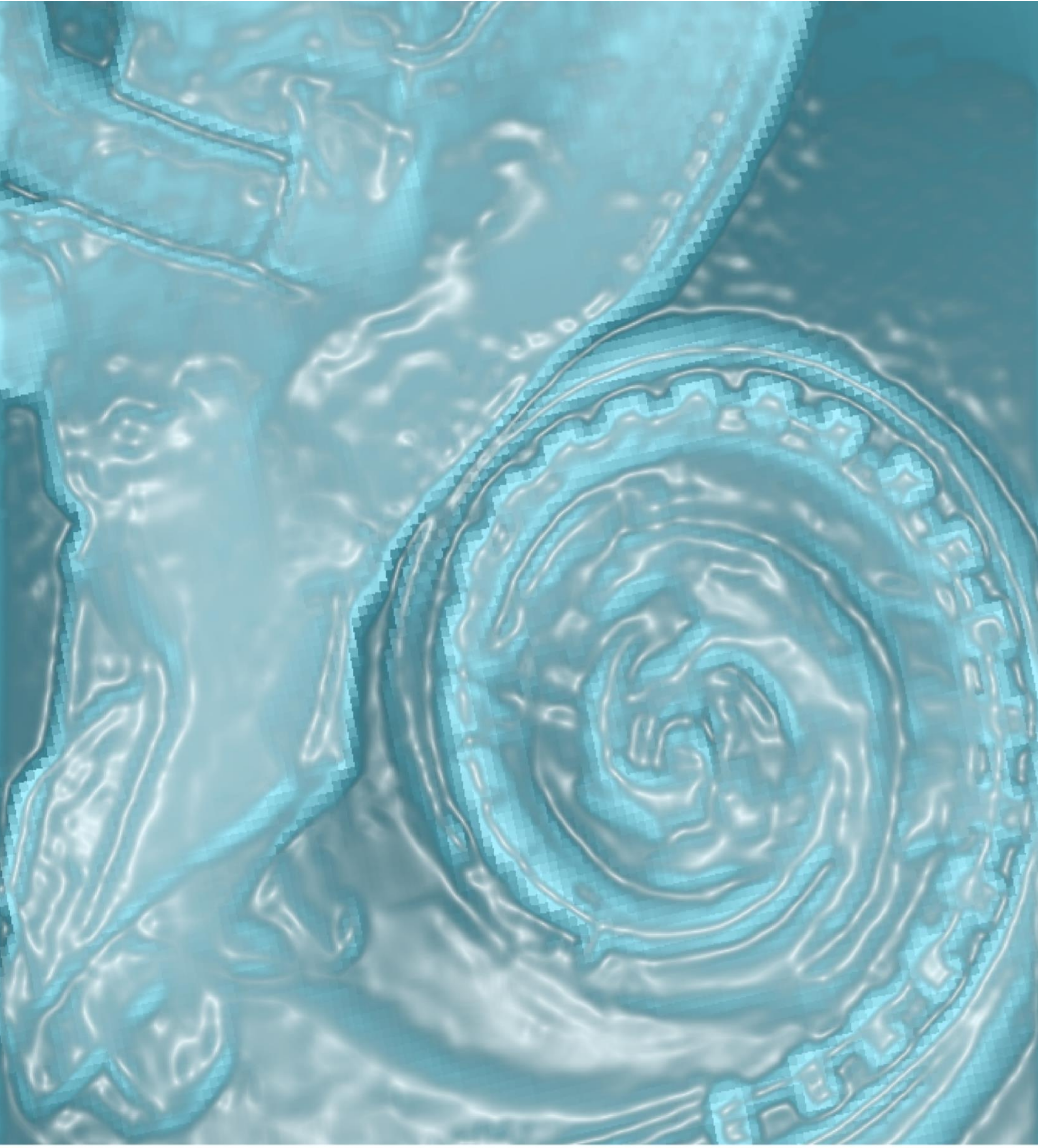
In other research done by Skarzynski et al. (2006) [94] using APHAB questionnaire to study QoL at 6 and 12 months after receiving cochlear implant revealed Global APHAB scores show a significant decrease over time, which means an improvement in benefit. Significant improvements were found in ease of communication (EC), background noise (BN) and reverberation (RV) subscales. However aversiveness subscale showed no significant difference. Their findings on these four subscales were similar to our findings in this review. These findings may suggest that CI provides significant improvements benefit in listening conditions for individuals with hearing experiences and still these individuals need some time to become accustomed to the sound from the implant. It should be considered as adverse effect in one listening situation and period for accustomization to the sound from implant should be minimized.

NCIQ is used as a quality of life questionnaire in many different countries all over the globe to investigate the application of the equipment, hearing ability, and emotional exchange ability. A study in 2008 by Hirschfelder et al.[95] to assess the impact of cochlear implantation on health-related quality of life for 56 adult CI users concluded that NCIQ was able to detect significant effects of cochlear implantation on health related quality of life and its correlation with audiologic measures, thus supporting its use as a measurement tool in cochlear implantation[95]. Their findings showed significant improvements in the total score and in all subdomains; physical, psychological and social after implantation among CI recipients.

In Beijing, China Liu B et al.[97] (2008) studied the QoL using NCIQ in 32 postlingually adult deaf recipients after cochlear implantation. Their findings showed that improvement was seen in sound recognition, speech recognition, confidence, daily life, social exchange and emotional behavior ability at different degrees. The improvement of confidence was very obvious. Their study also revealed that the speech communication ability of those with a hearing deprivation time longer than 5 years was lower than those with a shorter hearing deprivation time. They concluded that NCIQ could be used to evaluate the QoL among postlingually deaf adult cochlear implant recipients in China.

A continuation research by Dong R et al. in 2010[98] was done to investigate the reliability and validity of the Chinese version of NCIQ. A total of 94 CI recipients over 18 years old were included in this study. Test-retest analysis was administered randomly to 30 users without significant changes in health and social status during a two weeks' interval between test and retest. In this study, test-retest reliability of the NCIQ was proved to be satisfactory. All domains had significant coefficients ($p<0.01$). For validity, the correlation coefficients between overall NCIQ scores and the subdomains were also significant ($p<0.01$). They concluded the NCIQ meets many psychometric criteria of a robust instrument, which possesses appropriate validity with good reliability, and can be used to measure the outcome of cochlear implant adults in China.

Santos et al. in 2017[96] translated NCIQ into Brazilian Portuguese version, in their study on QoL of 24 adults CI users. Similarly to our review, the high overall score obtained in this study were for the subdomains basic and advanced sound perception, which belong to the physical domain. These subdomains reflect more on the direct benefits of CI use, providing better access to speech sounds, thus reducing the impact of hearing impairment on communication situations.



SUMMARY, RECOMMENDATIONS & PLAN OF ACTIONS

Chapter 6

SUMMARY

1. The MOH Cochlear Implant Programme is a successful programme. The Medical Development Division, MOH has taken the right course and direction to implement the Cochlear Implant Programme in Ministry Of Health hospitals.
2. The surgical procedures have been shown to be safe with successful cochlear implantation and low overall complication rate of 5.56%.
3. The majority of implantees have significantly gained improvement in auditory and speech-language performance following implantation.
4. In the pre-lingual group, children implanted at younger age showed better auditory and speech-language performance, and hence the need of early detection and intervention of hearing loss in neonates.
5. Post-lingual implantees require a shorter duration of rehabilitation period to achieve targeted auditory performance.
6. Participation of family members in providing appropriate audiological and speech-language rehabilitation is vital for successful outcome. It is important that they are fully involved and committed in providing adequate auditory and speech-language input to the implanted child.
7. Although there is improvement, compared to the first cohort (2008-2013), late age of detection of deafness in pre-lingual children and waiting time for implantation remain issues which need to be addressed.

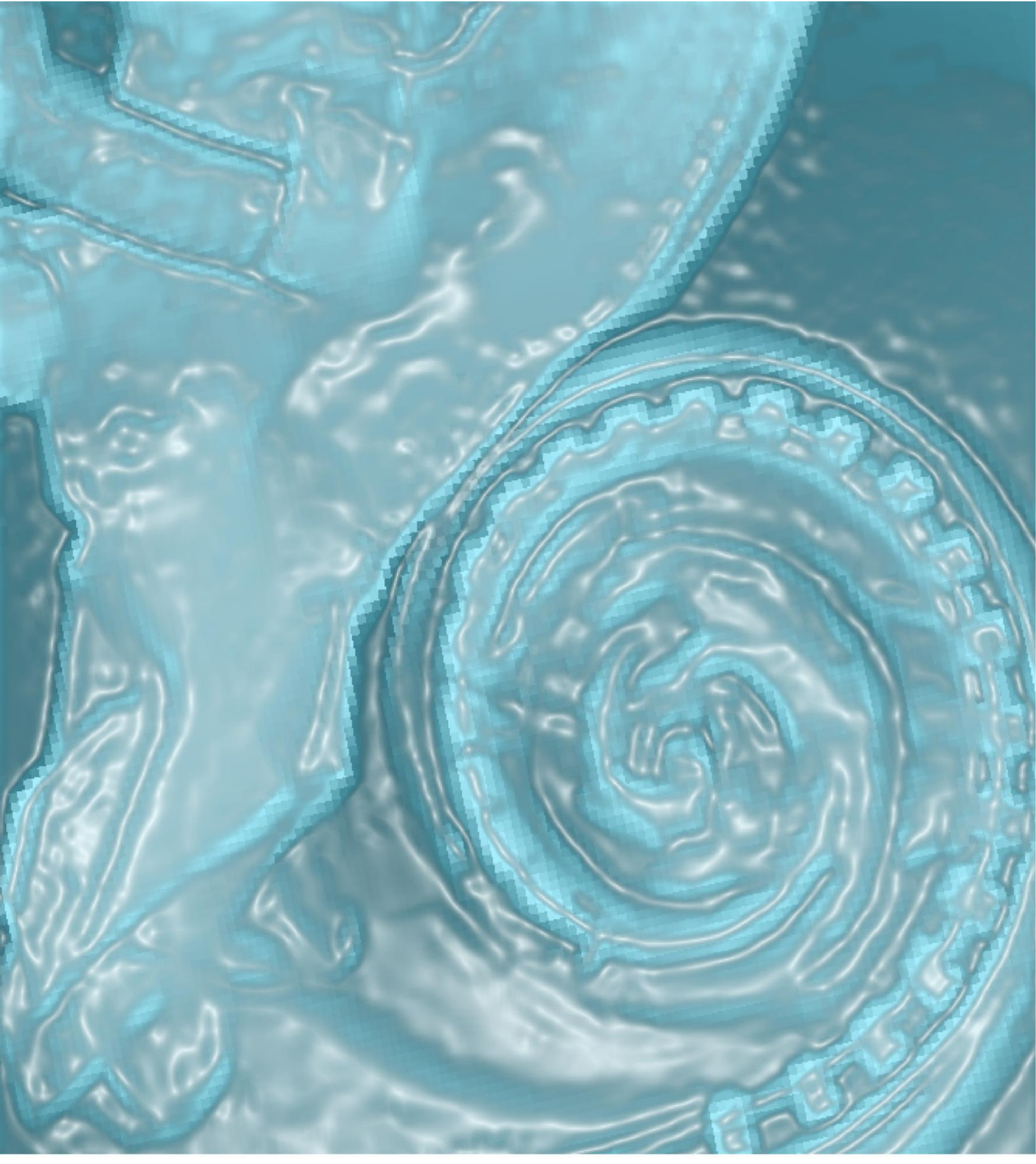
RECOMMENDATIONS

Based on this 10 years outcome review, the following are our recommendations to improve and maintain successful outcomes:

1. To strengthen and expand the Universal Neonatal Hearing Screening programme throughout the MOH facilities including primary care facilities to allow early detection and hearing intervention in the prelingual hearing impaired children.
2. To increase awareness of hearing loss and importance of early intervention among all levels of health care workers and public at large in concordance to World Health Organization (WHO).
3. To address hearing loss through a multidisciplinary approach and promoting, involving inter disciplinary and inter agencies collaboration; one of which being through National Ear and Hearing Care programme.
4. To ensure adequate numbers of personnel in the satellite hospitals which include Surgeons, Audiologists, Speech-Language Therapists and trained paramedics for optimum service delivery.
5. To provide continuous training for members in the CI team to ensure that the programme is up to date and conform to international standards of practice.
6. To create awareness and to emphasis on the importance of parental commitment in home-based program to maximize rehabilitation outcome.
7. To support the initiation of early intervention centers for hearing impaired children in Malaysia in order to promote oral and aural method of communication.
8. To expedite hearing aid acquisition in order to reduce the waiting time for hearing aid provision and cochlear implantation, especially in the prelingual age group.
9. To ensure a consistent and adequate cochlear implant grant from MOH in order to cater for the increasing number of candidates for implantation.
10. To shorten the waiting time from approval to surgery.
11. To develop a suitable national assessment tool in audiological and speech outcomes according to the needs of the local population.
12. To collaborate with other agencies in relation to cochlear implantation research.

PLAN OF ACTIONS:

1. To expand and strengthen the Universal Neonatal Hearing Screening Program (UNHS) in MOH hospitals and facilities.
2. To strengthen the workflow process between satellite and non satellite hospitals within the designated zone to expedite candidacy selection.
3. To expedite hearing aids and cochlear implants acquisition for prelingual (less than 4 years old) and postlingual deaf children through networking with funding agencies e.g. Tabung Bantuan Perubatan (TBP) and Jabatan Perkhidmatan Awam (JPA) or other sources to fast-track approval for application of hearing aids.
4. To conduct awareness program including regular workshops and continuous medical education (CME) to improve awareness and knowledge among primary care physicians, pediatricians, allied health care professionals, other health care professionals, NGOs and public for purpose of early referrals of suspected hearing loss cases.
5. Provision of training to all categories of personnel involved in Cochlear Implant Program on yearly basis.
6. To encourage the establishment of Parent Support Groups in all satellite hospitals and to strengthen their roles in order to improve parents understanding and commitment in the rehabilitation process.
7. To establish multidisciplinary early intervention centres in MOH hospitals for children with hearing and speech-language problems via inter-agency collaboration.



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Appendix 1:Categories of auditory performance II (CAPII) (Adapted from Nikolopoulos et al., 2005).

Category	Categories of Auditory II (CAPII) Criteria
0	No awareness of environmental sounds.
1	Awareness of environmental sounds.
2	Response to speech sounds
3	Identification of environmental sounds.
4	Discrimination of some speech sounds without lip-reading.
5	Understanding of common phrases without lip-reading.
6	Understanding of conversation without lip-reading.
7	Use of telephone with known listener.
8	Follows group conversation in a reverberant room or where there is some interfering noise, such as a classroom or restaurant.
9	Use of phone with unknown speaker in unpredictable context.

Appendix 2: Speech Intelligibility Rating (SIR) Scale (Cox & Mcdaniel, 1989)

Category	Speech Intelligibility Rating (SIR) Scale
Category 1	Connected speech is unintelligible. Prerecognizable words in spoken language, primary mode of communication may be manual.
Category 2	Connected speech is unintelligible. Intelligible speech is developing in single words when context and lip-reading cues are available.
Category 3	Connected speech is intelligible to a listener who concentrates and lip-reads.
Category 4	Connected speech is intelligible to a listener who has little experience of a deaf person's speech.
Category 5	Connected speech is intelligible to all listeners. Child is understood easily in everyday contexts.

Appendix 3: Parents' Evaluation of Aural/Oral Performance of Children (P.E.A.C.H.)

No.	Question	Never 0%	Seldom 1 - 25%	Sometimes 26 - 50%	Often 51 - 75%	Always 75-100%
1.	How often has your child worn his/her hearing aids and/or cochlear implant?					
2.	How often has your child complained or been upset by loud sounds?					
3.	When you call, does your child respond to his/her name in a quiet situation?					
4.	When asked, does your child follow simple instructions or do a simple task in a quiet situation?					
5.	When you call does your child respond to his/her name in a noisy situation when he/she can't see your face? (examples of responses include looks up, turns, answers verbally)					
6.	When asked, does your child follow simple instructions or do a simple task in a noisy situation?					
7.	When you are in a quiet place reading with your child, how often does he/she pay close attention to what you are saying? OR if your child is listening to stories/songs on the TV or CD when there is no other background noise how often can he/she follow what is being said?					
8.	How often does your child initiate/ participate in conversation in a quiet situation?					
9.	How often does your child initiate/ participate in conversation in a noisy situation?					
10.	How often does your child understand what you say in the car/bus/train?					
11.	How often does your child recognise peoples' voices without seeing who was talking?					
12.	How often does your child successfully use a phone?					
13.	How often does your child respond to sounds other than voices?					

Appendix 4: Listening Progress Profile (LiP) (Sue Archbold,1994a)

Listening Progress Profile (LiP)		
No	**Behaviour / Skill**	**Accepted Response**
1.	Response to environmental sounds	Any awareness of environmental sounds shown by spontaneous behavioural response
2.	Response to drum (elicited)	A behavioural response to loud drum elicited in play format, e.g. emerging from playhouse on sound of drum.
3.	Response to musical instruments (elicited)	A behavioural response to at least 2 different musical instruments (elicited) instruments: maracas, bells, tambourine, etc, e.g. child rolls ball at skittles in response to sound.
4.	Response to voice (elicited)	The child performs a task on the spoken signal, for example 'go' or 'boo' with voice at conversational sound level.
5.	Response to voice (spontaneous)	Reactions to voice in stories, singing, games formats have (spontaneous) been observed.
6.	Discrimination between 2 different instruments	The ability to discriminate (without being able to watch 2 different instruments when the sound is made) between 2 differing musical instruments.
7.	Discrimination between loud/quiet drum	The ability to discriminate between loud and quiet drums, loud/quiet drum for example, by pointing to appropriate picture, or imitating sound.
8.	Discrimination between single/repeated drum	Again, pictures may be used to denote a drum being played single/repeated drum once, or several times, and the child indicates by pointing at the picture. Alternatively, the child may be given a drum of his own and imitate the sound being made by the teacher out of sight.
9.	Identification of environmental sounds	Score 'S' if the child is reported as identifying some environmental sounds environmental sounds at home/school and the observer has observed the child identifying by sound alone at least one environmental sound. Score 'A' if the observer has observed the child identifying a range of environmental sounds at home/school and the child is monitoring his environment auditorily.
10.	Response to Ling's Five Sounds ('oo', 'ah', 'ee', 'sh', 'ss'):	The observer, in a variety of game Sounds (oo, ah, ee, sh, ss) formats, observes the child's detection of the five sounds. For example, does the child move the sleeping baby on the sound 'sh'? Score S if the skill is becoming established; score A if the child has been observed to carry out the tasks consistently.

11.	Discrimination between: • Long/short sounds • Single and repeated speech sounds • Loud and quiet speech sounds	The child can discriminate, for example, between the large long/short sounds **toy dog** giving **a loud** 'woof woof' and the **toy puppy** giving **a quiet** 'woof woof'.
12.		
13.		
14.	Discrimination between two of Ling's Five Sounds.	Score 'S' if the child can discriminate between at least one two of Ling's Five Sounds combination of 2 of Ling's Five Sounds. Score A if the child (for example oo versus ee, can discriminate between any combination of 2 of Ling's sh versus ss) Five Sounds.
15.	Recognition of all of Ling's Five Sounds	Score 'S' if the child is becoming able to identify any one sound out of the five. Score 'A' if the child is consistently able to identify all five sounds.
16.	Discrimination between 2 family names of differing syllabic length	Using family pictures, written names or the people 2 family names of differing themselves, can the child discriminate between 2 names of syllabic length differing syllabic length, e.g. John v. Mummy.
17.	Identification of own name	Score 'S' if the child has been observed to identify his own name at home and school on at least one occasion. Score 'A' if the child can identify his own name (in reasonable listening conditions) consistently. The child may, of course, choose to ignore his name!

- **Score N (Never/not known) if the skills never been observed. 'N' carries 0 mark.**
- **Score S (Sometimes) if the skill is becoming established and has been observed more than chance level. 'S' carries 1 mark**
- **Score A (Always) if the skill is reported to be well established and the observer has observed it consistently in more than 2 situations. 'A' carries 2 marks.**
- **Total marks/score= 42**

Appendix 5: Family Participating Rating Scale (Moeller,2000)

Rating of 1 (Limited Participation)

Family faces significant life stresses that may take precedence over the child's needs (e.g. domestic abuse, homelessness). Family has limited understanding of deafness and its consequences for the child. Participation may be sporadic or less than effective. Parent/child communication is limited to very basic needs.

Rating of 2 (Below Average)

Family struggles in acceptance of the child's diagnosis. The family may be inconsistent in attendance. They may be inconsistent in maintaining the hearing aids and keeping them on the child outside of school. They may have some significant life stressors that interfere with consistent carryover at home. Management of the child presents daily challenges to the family. Communicative interactions with the child are basic. Family lacks fluency in the child's mode of communication.

Rating of 3 (Average Participation)

Family is making efforts to understand and cope with the child's diagnosis. Family members participate in most sessions/meetings. Busy schedules or family stresses may limit opportunities for carryover of what is learned. Family may find management of the child challenging. Family attends Individual Family Service Plan and Individual Education Plan meetings but may rely primarily on professional guidance. Family attempts to advocate but may be misdirected in some of their efforts. Selected family members (e.g. mother) may carry more than their share of responsibility for the child's communicative needs. Family members develop at least basic facility in child's communication mode. Family members are willing to use language expansion techniques but need ongoing support and direction.

Rating of 4 (Good Participation)

Family members make a better than average adjustment to the child's deafness. Family members regularly attend parent meetings and sessions. Parents take an active role (perhaps not the lead) in Individual Family Service Plans and Individual Education Plans. Family members serve as good language models for the child and make an effort to carry over techniques at home. Some family members have fairly good facility in the child's communication mode and/or in techniques for language stimulation. Efforts are made to involve extended family members.

Rating of 5 (Ideal Participation)

Family seems to have made a good adjustment to the child's deafness. The family is able to put the child's disability in perspective within the family. Family members actively engage in sessions. They attend sessions and meetings regularly and pursue information on their own. They serve as effective advocates for their child with professionals/school districts, etc. Family members become highly effective conversational partners with the child and serve as strong and constant language models. Family members become fluent/effective users of the child's mode of communication. They are capable of applying techniques of language expansion. Extended family members are involved and supportive.