Objective: To share our experience of difficulties encountered while implanting different types of cochlear implants and ascertain the affordability of cochlear implant surgery among the Pakistani population to treat sensorineural hearing loss in a novel cochlear implant programme.

Study design: Retrospective longitudinal study.

Place and Duration of Study: Department of ENT, Combined Military Hospital (CMH), Rawalpindi Pakistan, from Apr 2017 to May 2021.

Methodology: The study included patients who underwent cochlear implant surgeries. Patient demographics and causes of deafness were collected. Intraoperative difficulties were encountered, and round window accessibility was also recorded.

Results: Out of 200 patients, 112(56%) were males, while 88(44%) were females. 193(96.5%) patients were pre-lingual under 12 years of age, while post-lingual patients were 7(3.5%) and were adults. Cochlear implants from Med-El 186(93%), Cochlear 2(1%) and Advance Bionic 10.5% were implanted through a transmastoid, facial recess approach, while Neubio 11(5.5%) was implanted through a postauricular transcanal approach. Congenital deafness with consanguinity was the leading cause of deafness, while round window variation was the most frequently encountered surgical challenge.

Conclusion: In developing countries, financial restraints cause delays in surgery, leading to limited access to cochlear implant programmes. Surgical challenges are frequently encountered during surgery, and better understanding is required for easy implantation.

Keywords: Bilateral Deafness, Cochlear implant, Cochlear Round Window


INTRODUCTION

Cochlear Implant (CI) is a surgically implanted device that converts acoustic signals into electric pulses by directly stimulating auditory nerve neurons bypassing cochlear hair cells. It is a gold standard technique for treating profound sensorineural hearing loss (SNHL) that does not benefit from hearing aids.

An effective CI programme demands a well-trained team of Otolaryngologists, Audiologists, Speech and Language pathologists, Child Psychologists, Child Specialists, and Implant surgeons. Unfortunately, like other developing countries, the CI programme in Pakistan is still limited to the private healthcare sector due to financial limitations, unawareness of the general population, lack of financial support from the Government, and scarcity of trained surgeons and equipment in the Public sector. However, with the development of infrastructure, training of local surgeons, and availability of post-operative rehabilitation, the CI programme has been started in a few major hospitals of this country regularly. Our hospital, being novel in the field, is looking after a Pakistani population through the Armed Forces Welfare Project (AFWP) and the Federal Government of Pakistan (FGOP) along with self-financed (SF) affording patients. We implanted CI by Med-EL in AFWP and FGOP-funded patients. In contrast, based on affordability and availability, SF patients were implanted with Med-EL, Neubio, Advance Bionic, and Cochlear CI.

Though in Pakistan, the private sector did involve charity-based CI, elevating the affordability threshold for patients, limited surgeries are performed in contrast to the high incidence of childhood SNHL in Pakistan. Similarly, data and local studies about challenges related to CI surgery are meagre. Few studies have discussed surgical complications, but surgical challenges encountered during surgery related to the round windows, facial nerve, CSF gusher, etc, are less reported.
Experience of Patients in a Novel Cochlear Implant Programme

This study aims to share our experience of successfully treating 200 CI cases in a novel CI programme run in our tertiary care hospital. We intend to contribute to the literature about the demographic distribution of patients who benefitted from different types of implants, causes of SNHL, difficulties encountered while operating these cases and the affordability of CI surgery. Our objective is to identify causes of deafness among CI patients, determine challenges experienced during CI surgery and ascertain the affordability of CI among the Pakistani population. This research will aid CI surgeons in effective planning before surgery and facilitate researchers for future studies in the region.

METHODOLOGY

After approval of the Hospital Ethical Committee (No.243/2/22), the retrospective longitudinal study was commenced from April 2017 to May 2021 at the Department of ENT, Combined Military Hospital (CMH), Rawalpindi Pakistan.

Inclusion Criteria: Patients of either gender or age with bilateral profound SNHL of >90 db did not benefit from a trial of six months or more of hearing aid and speech therapy (for pre-lingual congenitally deaf children, the maximum age for inclusion was five years, while for post-lingual patients, it was 50 years. However, pre-lingual children between 5-12 years of age, who started using hearing aids and speech therapy within the first five years of life but remained unbenefited), were included once their families consented to postop regular speech therapy and followed up with a realistic outcome. Only those patients who were medically fit for surgery and whose families consented to post-operative speech rehabilitation were included.

Exclusion Criteria: Children below 01 years, having post meningitis cochlear ossification, diagnosed cases of Cochlear aplasia/deformity/Cochlear nerve agenesis and pre-lingual children >5 years of age not using any hearing aid or speech therapy were excluded.

Cases were selected after a detailed history and clinical examination, hearing assessment by Brainstem Evoked Response Audiometry (BERA), Auditory Steady State Response (ASSR), Pure Tone Audiometry (PTA) and Tympanometry were done. Laboratory and radiological Investigations, including blood complete picture, coagulation profile, hepatitis screening, renal, hepatic, and thyroid function tests, computerized tomographic (CT) scanning and Magnetic Resonance Imaging (MRI) as per CI protocol to find out any associated syndromes, an anatomical or neurological disorder in brain, cochlea or in cochlear nerve were performed. Patients were also evaluated in detail by specialists in ENT, Speech and language pathologists, child specialists, child psychologists and audiologists individually to rule out any associated cause of speech delay, disorder or syndrome and cause of deafness before final recommendation of fitness for surgery. Patients were also evaluated for anaesthesia fitness. Children were also vaccinated against pneumococcal infections. All patients were thoroughly informed about surgical procedures and possible complications, and detailed informed consent was taken from adult patients and parents of paediatric groups. Affordability by patients was asked through the questionnaire. Funds from AFWP or FGOP were demanded for unaffordable patients.

Three surgeons with nearly the same surgical experience and skills operated on the patients. For patients undergoing Med-El, Advance Bionics and Cochlear devices, the gold standard technique of transmastoid posterior tympanotomy approach with the drilling of receiver bed was performed. At the same time, Neubio was installed through a transanal approach without bed creation. Postoperatively, patients were kept on injectable antibiotics and analgesics for 24 hours, along with pressure dressing. A transorbital/Stenvers X-ray was conducted to confirm that the electrodes were placed correctly. Patients were discharged on oral Co-Amoxiclav and Paracetamol after 48 hours in the hospital, depending upon their general condition. The stay was prolonged in patients who had nausea and vertigo. Third-generation cephalosporin was added only if infection was suspected. As per culture and sensitivity, an appropriate antibiotic was advised for the patients with post-operative infective discharge. Dressing lightened after 24 hours was entirely removed on day five, while stitches were removed on day-seven.

Demographic data and causes of deafness, were collected. In addition, intraoperative difficulties, and round window (RW) accessibility was recorded per St Thomas Hospital (STH) classification. Perioperative visibility of RW after doing adequate posterior tympanotomy was measured in frequency. Accordingly, RW visibility was classified into Type I (100%), Type IIA (> 50%), Type IIB (<50%) and Type III (0%).

Statistical Package for Social Sciences (SPSS) version 23.0 was used for the data analysis. Quantitative variables were expressed as Mean±SD and
qualitative variables were expressed as frequency and percentages.

RESULTS

Out of 200 patients who operated between 2017 and May 2021 (Figure), 112(56%) were males, while 88(44%) were females. There were 116(58%) patients who were below 5 years of age, 77(38.5%) were between 5 and 12 years and 7(3.5%) were above 12 years of age. The average age of patients below five years was 3.48±1.19 years, between 5 and 12 years was 6.28±1.70 years, and above 12 years patients was 27.71±10.01 years. 193 (94.27%) were post-lingual, the majority under 12 years of age, while 07(3.5%) were above 12 years of age and post-lingual. CI was used from Med-El, Neubio, Cochlear and Advance Bionic 186(93%), 11(5.5%), 2(1%) and 1(0.5%) respectively. Except for all Neubio cases, which recommend a postauricular transcanal approach, the rest of the implantations were performed through transmastoid, facial recess approach 189(94.27%). Table-I elaborates on the apparent causes of deafness in our patients, with congenital deafness of unknown origin being the leading cause of hearing loss.

<table>
<thead>
<tr>
<th>Causes</th>
<th>n(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital Unknown</td>
<td>180(90%)</td>
</tr>
<tr>
<td>Noise Induced Hearing loss</td>
<td>4(2%)</td>
</tr>
<tr>
<td>Progressive Sensorineural Hearing loss</td>
<td>8(4%)</td>
</tr>
<tr>
<td>Post Meningitis</td>
<td>4(2%)</td>
</tr>
<tr>
<td>Post CMV Infection</td>
<td>2(1%)</td>
</tr>
<tr>
<td>Syndrome Associated</td>
<td>2(1%)</td>
</tr>
<tr>
<td>Jervell and Lange-Nielsen Syndrome</td>
<td>1(0.5%)</td>
</tr>
<tr>
<td>Teitz albinisim Deafness Syndrome</td>
<td>1(0.5%)</td>
</tr>
</tbody>
</table>

RW variation was the most frequently encountered challenge 67(33.5%) while other problems faced during surgery included middle ear effusion 16(8%), CSF gushers 8(4%), high riding jugular bulb 5(2.5%), basal turn ossification 3(1.5%), early bifurcation of facial nerve 2(1%), and dehiscent jugular bulb 1(0.5%) (Table-II).

<table>
<thead>
<tr>
<th>Operative Difficulties</th>
<th>n(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type II B Round Window (RW) Variation</td>
<td>56(28%)</td>
</tr>
<tr>
<td>Type III RW Variation</td>
<td>11(5.5%)</td>
</tr>
<tr>
<td>Middle ear effusion</td>
<td>16(8%)</td>
</tr>
<tr>
<td>CSF Gusher</td>
<td>08(4%)</td>
</tr>
<tr>
<td>High riding Jugular Bulb</td>
<td>05(2.5%)</td>
</tr>
<tr>
<td>Basal Turn Ossification</td>
<td>03(1.5%)</td>
</tr>
<tr>
<td>Early Bifurcation of Facial Nerve</td>
<td>02(1%)</td>
</tr>
<tr>
<td>Dehiscent Jugular bulb</td>
<td>01(0.5%)</td>
</tr>
</tbody>
</table>

DISCUSSION

According to a WHO report of 2018, Pakistan had approximately 14.5 million people suffering from hearing loss, yet there are very few hearing rehabilitation centres in the private and public sectors. Initially, CI surgery was performed only on an SF basis in the private sector, but recently, CI programmes have also been started in the public sector. With a population of over 220 million (the world’s 5th largest), Pakistan had a GDP per capita of only $1,186 in 2020-21, which is 154th in the world. This limits resources to rehabilitate deaf individuals, involving expensive CI devices for the general public.

In our programme, out of 200 patients, 112 were males. At the same time, 88 were females, similar to the study by Ahmed et al. (154 males versus 97 females) and Garrada et al., which depicts male predominance over females, which was not established by other international research.

Selection criteria below 5 years of age were exercised to prevent neural plasticity. Patients above five years of age were implanted only when the patient had residual hearing, were hearing aid users, or had partial speech. It is supported by Freni et al. who claim no difference in the pragmatic development of pre-lingual hearing impaired children if operated before the age of 7 years. Forli et al. also claims that cochlear implants are beneficial even in pre-lingual adults.

Among the 200 cases we operated on, 90% of patients had congenital deafness of unknown origin but mostly associated with consanguinity, which is a proven cause across the globe. Shafique et al. and Korver et al. reported that congenital causes are
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associated with hearing loss in 70% of cases,\textsuperscript{16,17} while Khan et al. published a 94% incidence of congenital deafness in children for cochlear implants.\textsuperscript{6} Syndrome-associated causes of deafness included Jervell and Lange-Nielsen syndrome and Teitz albinism deafness syndrome. Jervell and Lange-Nielsen Syndrome were tricky in the pre-anaesthesia period; the patient remained asymptomatic but developed arrhythmias per-operatively, which were managed appropriately. Teitz albinism deafness syndrome was easily diagnosed due to obvious fair skin, light colour hairs and abnormal retinal pigmentation. Maternal and neonatal infections, including meningitis and CMV, were found in a total of 6 (2 and 1\% respectively) patients, which is quite less than the international literature in which hearing loss is associated with meningitis, and CMV is 6\% and 14\% in the paediatric group.\textsuperscript{18} In different regions, noise-associated SNHL is 7-21\%, but in our study, it was only 2\%, which was in only the adult population.

In this study, various operative challenges faced during CI surgery were identified. According to STH classification, we faced type IIB RW in 28\% of patients and type III RW in 5.5\%, comparable to Lim et al.\textsuperscript{1} and Stuermer et al. Where they claim 22\% and 16\% incidence of difficult RW accessibility,\textsuperscript{18,19} The study is a shared understanding of the CI programmes funded through AFWP, FGOP and SF. Though it is the only local study identifying surgical challenges and the experience of a public sector CI programme, it is novel and limited and requires a larger sample size. We suggest further multi-centre research to determine epidemiological causes, challenges encountered by surgeons, and the financial impact of the cost-effectiveness of CI programmes on society. Simultaneously, public awareness and neonatal hearing screening are imperative for early diagnosis and intervention for desirable results.

CONCLUSION

Cochlear implant entails handling an effective but expensive device that requires proper training, infrastructure, and a well-trained team for its successful outcome. Congenital deafness of unknown origin is the commonest cause of deafness in pre-lingual deaf children. Moreover, surgical challenges are frequently encountered by CI surgeons, who require a better understanding of variations in anatomy for easy implantation.

Conflict of Interest: None.

Authors’ Contribution

Following authors have made substantial contributions to the manuscript as under:

MA & SNR: Conception, study design, drafting the manuscript, approval of the final version to be published.

AHK & OKN: Data acquisition, data analysis, data interpretation, critical review, approval of the final version to be published.

SA & RAK: Data acquisition, drafting the manuscript, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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