

Original Article

Preoperative Evaluation and Surgical Outcome of Cochlear Implantation in NIENT

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Abstract:

Background: Sensory neural hearing loss (SNHL) is the most common congenital sensory deficit, with an incidence of one to three per 1000 live births. Acoustic deprivation during the first 3 years of life can hinder speech and language acquisition with significant negative consequences on a child's educational and psychosocial development. The gold standard intervention for permanent severe to profound hearing loss is cochlear implantation. Cochlear implant (CI); is a semi implantable electronic device that bypass the cochlea.

Objectives: An observational study was carried out on 40 cases of pre-lingual deaf to find out the causes of pre-lingual deaf, to evaluate the preoperative procedures to set ideal criteria for pre -lingual cochlear implantation and to evaluate surgical procedure and outcome of cochlear implantations.

Methods: Evaluation of the candidates included patient medical history, general health check-up, ENT examination, audiometric evaluation, CT and MRI scans, psychological profile of the candidate. A limited cortical mastoidectomy was performed. The facial recess was opened using the fossa of incudis as an initial landmark. The round window niche was visualized through the facial recess about 2 mm inferior to the stapes. A cochleostomy created by drilling over the basal turn of the cochlea anterior and inferior to the annulus of the round window membrane. The electrode array was then carefully inserted through the fenestra into the scala tympani of the cochlea. Electrophysiological testing (Neural Response Telemetry: NRT) was performed to verify the correct placement of active electrodes.

Result: Among them 22 (55%) were male and 18 (45%) were female. Male female ratio was 1.2:1. Age distribution at implantation was 3.3 ± 1.054 (SD). Average hearing loss was 96.4 ± 5.3 (SD) dB and in aided audiogram was 63.7 ± 4.6 (SD) dB. Overall complications

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occurred in 10 (25%) cases. Transient facial nerve paresis in 2 (5%), injury of tympanic membrane in 1(2.5%), seroma 4(10%) and delayed otitis media in 1(2.5%) were observed. Major postoperative complications occurred in 2 cases including facial nerve palsy in 1(2.5%) case and spontaneous device failure in 1(2.5%) case.

Conclusion: The result of this survey was find out the risk factor of congenital hearing loss. With a thorough preoperative evaluation, we can select proper candidates for CI which is a reliable and safe procedure with a low percentage of severe complication.

Key words: Cochlear implant, Preoperative evaluation, Surgical procedures, Postoperative complications.

Introduction:

Cochlear implant (CI) is a semi implantable electronic device that bypasses the cochlea by means of an electrode array stimulating directly the cochlear nerve, thereby transmitting an electrical signal to the auditory cortex. A CI has an external component, worn behind the ear just as a hearing aid and an internal component, surgically embedded in the mastoid. The external part consists of a microphone for obtaining sound, a speech processor that analyzes and encodes sound into a digital code and a magnetic headpiece transmitting the coded signal to the internal part via a transcutaneous radiofrequency link to the internal part. The internal part contains a receiver-stimulator that receives and decodes the data, and in turn sends the decoded signal to the electrode array. The latter is the core of the system and consists of a flexible silicone carrier containing a variable number of electrodes. The electrode array is surgically inserted into the scala tympani of the cochlea and stimulates directly the residual cochlear nerve fibers. Since their introduction more than 30 years ago, CIs have improved their performance to the extent that are now considered to be standard of care in the treatment of children with severe to profound SNHL¹. Today the use of cochlear implant for hearing rehabilitation of deaf individuals is widespread all over the world with more than 100000 users in the world^{2,3}.

Acoustic deprivation during the first 3 years of life can hinder speech and language acquisition with significant negative consequences on a child's educational and psychosocial development.

Hearing loss affects about 5.3% (approximately 360 million) of the world's population⁴. SNHL is the most common congenital sensory deficit, with an incidence of one to three per 1000 live births; this incidence mounts up to 4-5% in neonates with risk factors for SNHL⁵. This incidence is estimated to be 4.8% in children aged 0–1 years and 6.4% in children aged 1-4 years⁶. Genetic causes account for approximately 50% to 60% of pediatric SNHL, while 15% to 40% is due to an acquired cause, such as infections, ototoxic drugs, anoxia, low birth weight, hyperbilirubinemia, traumas, metabolic and autoimmune diseases^{7,8,9}. Prevalence and severity of hearing loss vary with some factors including socioeconomic status, exposure to infections and consanguinity¹⁰. Severe to profound hearing loss is defined as hearing loss of 61 dBHL or more in the better ear¹¹.

The gold standard intervention for permanent severe to profound hearing loss is cochlear implantation. Candidates include children born with the permanent bilateral sensorineural hearing loss¹².

This study was conducted in specialized Ear, Nose and Throat hospital in Bangladesh and

concentrated on preoperative evaluation, surgical procedure and outcome of cochlear implantation indicated for pre (<2 years of age) and perilingual (2 -5 years of age) deafness.

Objectives:

- To find out the cause of pre -lingual and perilingual deafness.
- To evaluate the preoperative procedures.
- To set ideal criteria for pre -lingual cochlear implantation.
- To evaluate surgical procedure and outcome of cochlear implantations.

Methods:

Study design

- Type of study: Cross sectional observational study.
- Study population: 40 cases of prelingual deaf were selected for this study.
- Study period: 1st June 2018 to 31st May 2019 (12 months).
- Place of study: National Institute of ENT, Tejgaon, Dhaka.
- Inclusion criteria:

Prelingual deaf child under 5 years of old who were physically and psychologically fit for cochlear implantation.

- Exclusion criteria:

Post lingual deaf. Deaf more than 5 years old.

Materials:

Evaluation of the candidates was performed according to a protocol created in our department that included patient medical history, general health check-up, ENT examination, audiometric evaluation, CT and MRI scans, psychological profile of the

candidate. Every candidate was evaluated by taking proper medical history including marriage consanguinity of parents, prenatal TORCHES infections, birth history, mode of delivery, birth weight, birth asphyxia, use of neonatal ICU. Pediatric audiological evaluation was based on several subjective and objective hearing tests like Impedance audiometry, Behavioral Observation Audiometry (BOA), Otoacoustic Emissions (OAEs), Auditory Brainstem Response (ABR), Auditory Steady-State Response (ASSR) and Aided Audiogram. Preoperative imaging assessment is mandatory to verify the presence of minimal requirements for cochlear implantation, i.e, a patent cochlea and an intact cochlear nerve. High resolution computed tomography (HRCT) was done to provide information regarding the structure of the bony labyrinth, the number and patency of the cochlear turns, the size of the internal acoustic meatus, the position of the facial nerve and the vestibular structures, the anatomy of the middle ear and the mastoid. Magnetic resonance imaging (MRI) was done to confirm the presence of cochlear nerve as well as in searching for central auditory pathway abnormalities and fibrous obliteration of the membranous labyrinth.

Results:

In this study 40 pre-lingual deaf cases were included. Among them 22 (55%) were male and 18 (45%) were female. Male female ratio was 1.2:1.

Age distribution at implantation was 3.3 ± 1.054 (SD) years (Mean age was 39.6 months with a standard deviation of 12.64 months). Most of the cases were age group 4-5 years (32.5%).

Analysis of the socio-economic profile of the cases revealed both fathers and mothers are educated up to secondary school level were 55%. Among the all cases 40% mothers were illiterate. Most of the fathers were

employed in mid-level non-professional jobs (85%) and almost all mothers were housewives (90%). Around 30% and 65% patients belonged to the middle-income (2100-7000 USD per annum) and low-income (<2100 USD per annum) family respectively. The etiology of deafness was unknown in 9 cases (22.5%), however in 13 cases (32.5%) and 18 cases (45%) had multiple and single risk factor respectively.

Average hearing loss was 96.4±5.3(SD) dB and in aided audiogram was 63.74.6(SD) dB. Hypoplastic cochlear nerve in 1 (2.5%) case and hypocellularity of mastoid process in 4 (10%) cases were observed in radiological evaluation. Intraoperative abnormally placed facial nerve was found in one (2.5%) case. Per-operative NRT was absent in 3 (7.5%) cases, whereas post-operative NRT was absent in only one case.

Overall complications occurred in 10 (25%) cases, including minor complications in 8 (20%) and major complications in 2 (5%) cases. Transient facial nerve paresis in 2 (5%), injury of tympanic membrane in

1(2.5%), seroma 4(10%) and delayed otitis media in 1(2.5%) were observed. Major postoperative complications occurred in 2 cases including facial nerve palsy in 1(2.5%) case and spontaneous device failure in 1(2.5%) case. Postoperative nausea and vomiting (PONV) was present in 4 (10%) cases. No drop out was found in this study.

Table II :
Risk Factors

Risk Factor	Frequency	Percentage
Consanguinity of marriage	4	10%
TORCHES infection	2	5%
Low Birth weight	4	10%
Preterm delivery	2	5%
Birth asphyxia	5	12.5%
USE of NICU	1	2.5%
Multiple Risk factors	13	32.5%
No risk factors	9	22.5%
	n=40	Total= 100%

Table III
Postoperative Complications

Complications:	Name of complications	Frequency	Percentage	P value
Minor Complications	Injury of the Tympanic membrane	1	2.5%	0.025
	Transient facial nerve paresis	2	5%	0.05
	Seroma	4	10%	0.1
	Delayed otitis media	1	2.5%	0.025
Major Complications	Facial nerve palsy	1	2.5%	0.025
	Spontaneous device failure	1	2.5%	0.025
		n=10	25%	

Discussion:

Cochlear implantation is a surgical procedure performed in numerous centers around the world. Expanding the criteria for CI leads to a significant increase in the number of patients using such devices. 1990 FDA approved the use of CIs in congenitally deafened children; since then, the candidacy criteria in pediatric age have changed and expanded over the years. The lack of auditory information from the environment during early childhood impedes the normal development of the auditory system, and interferes with the acquisition of language skills. In fact, there is a window of time in the first 3 years of life (the "sensitive period"), during which the child's brain is very plastic and has the ability to develop new neural pathways in response to auditory stimuli¹³. Behind this period, the auditory cortex can no longer be recruited by auditory input because the intact senses take over the auditory neural network through a process of cross-modal reorganization¹⁴. The rationale behind early cochlear implantation is to minimize the consequences related to sensory deprivation during the sensitive period. Several studies have shown that children implanted before 2 years of age perform significantly better than children implanted at older ages¹⁵.

It is evident that earlier implantation yields superior cochlear implant performance in children, because we utilize the mental plasticity to create acoustic memory^{16,17}. That's why congenitally or prelingually deafened children implanted prior to age 3 years may yield improved results¹⁸.

In this study, age distribution at implantation was 3.3 ± 1.054 (SD) years (Mean age was 39.6 months with a standard deviation of 12.64 months). Most of the cases were age group 4-5 years (32.5%). Collettiet al reported on 12 children implanted at or

before the age of 6 months; four years after implantation, these children had receptive and expressive language skills similar to normal-hearing peers. However, other studies did not confirm clear evidence of improved outcomes in children implanted in the first year of life compared with those implanted a year later^{19,20}.

Among the all cases 40% mothers were illiterate. Both fathers and mothers are educated up to secondary school level were 55%. Almost all mothers were housewives (90%). Around 65% patients belonged to low-income family (<2100 USD per annum). S. Singh et al. found that education of fathers and mothers was mostly at secondary school level. While most mothers were housewives only, fathers were in all kinds of low to medium level skilled jobs with almost none in high paying professional roles like Lawyers and Doctors. The annual income of the vast majority of these patients was less than 8000 USD. The etiology of hearing loss could not be established in 9 (22.5%) cases. Within our study setup, the investigation of etiology of disease did not compulsorily include genetic testing due to extra costs involved there. Low birth weight, birth asphyxia, premature delivery, viral infections and consanguinity of marriage were the most common etiologies observed in this study. S. Singh et al²¹ reported that viral infections prematurity and history revealing an inherited pattern were the most common etiologies.

Average hearing loss was 96.4 ± 5.3 (SD) dB and in aided audiogram was 63.7 ± 4.6 (SD) dB. Hypoplastic cochlear nerve in 1 (2.5%) case and hypocellularity of mastoid process in 4 (10%) cases were observed in radiological evaluation. Intraoperative abnormally placed facial nerve was found in one (2.5%) case. Per-operative NRT was absent in 3 (7.5%) cases, whereas post-operative NRT was absent in only one

case. In our series, overall complications occurred in 10 (25%) cases during 4 months follow up. The number of major complications was 2 (5%) and the number of minor complications was 8 (20%). Sefin IK²² reported in his series that the overall rate of complication was 18.75% during a maximum follow-up period of 20 months. The number of major complications was 12 (10.7%), the number of minor complications was nine (8.03%). Venail *et al.*²³ reported in their series that the overall rate of complication was 16% during a maximum follow-up period of 18 years and the rate of major complications (5.6%) was lower than that found in previous studies (18.3%)²⁴ and 11.8% excluding device failures²⁵. Thus, our study showed comparatively less complications reported in other studies. However, we should take into consideration that the number of patients in our study is lower, with a shorter period of follow-up. Facial palsy remains a rare and transient complication of CI. Transient facial nerve paresis in 2 (5%), injury of tympanic membrane in 1 (2.5%), seroma 4 (10%) and delayed otitis media in 1 (2.5%) were observed in our study. Complete facial nerve palsy was occurred in one case (2.5%).

Transient facial nerve paresis occurred in three cases (2.678%) in our series immediately postoperatively due to overheating during drilling compared with one of 500 cases (0.002%) in the study by Venail *et al.* [34] and 0.33 and 2.22% [37] in other studies. As demonstrated elsewhere, facial palsy is usually of late onset and moderate, implying that the underlying cause is an inflammatory and edematous mechanism rather than direct trauma during drilling²⁶. Spontaneous device failure in 1 (2.5%) case compare to 2-4% reported in one study²⁷ and 1.05% in another study²⁸.

Postoperative nausea and vomiting (PONV) was present in 4 (10%) cases compare to 1.05% reported by Darlong V. *et al.*²⁹.

Conclusion:

CI is an effective and reliable to restore auditory sensation in profoundly deafened patients. The result of this survey was find out the risk factor of congenital hearing loss. With a thorough preoperative evaluation, we can select proper candidates for CI which is a reliable and safe procedure with a low percentage of severe complication. However, the patients should receive life time follow up.

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