ORIGINAL RESEARCH PAPER

VOLUME-6 | ISSUE-5 | MAY - 2017 | ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

IMAGING IN CONGENITAL DEAFNESS AND RADIOLOGICAL CORRELATION OF OUTCOME OF COCHLEAR IMPLANTS

Radiology

Dr. Aryasree P.M	MBBS, Junior Resident, Department of Radiodiagnosis, Government Medical College, Kozhikode, Kerala
Dr. Gomathy Subramaniam	MBBS, MD,DMRD, MS . Professor, Department of Radiodiagnosis, Government Medical College, Kozhikode. Kerala.
Dr. Jineesh .T	MBBS , MD, DMRD , FRCR, EDIR. Associate Professor, Department of Radiodiagnosis, Government Medical College, Kozhikode. Kerala
Dr . V.R Rajendran	MBBS , MD , DMRD. Professor & HOD, Department of Radiodiagnosis, Government Medical College, Kozhikode. Kerala
Dr. Saanida M.P	MBBS , MD , DMRD,DNB, FRCR. Associate Professor, Department of Radiodiagnosis, Government Medical College, Kozhikode. Kerala
Dr.Swathilal S.A	MBBS , M.S . Additional Professor, Department of ENT , Government Medical College, Kozhikode. Kerala

ABSTRACT

Congenital deafness can hinder speech and language acquisition with significant negative consequences on a child's educational and psychosocial development. Cochlear implantation has become an accepted treatment in children with profound sensorineural hearing loss.

Aims and objectives: The study aimed at correlating the outcome of cochlear implantation surgery in the form of intra operative neural response telemetry (NRT), functional auditory performance indicators (FAPI) and assessment of language development (ALD) with the pre operative imaging findings.

Materials and methods: A cohort study was conducted during January 2015 to May 2016 in the Department of Radiodiagnosis, Government Medical college Kozhikode. 84 subjects under 5 years underwent preop imaging . Intra operative monitoring of neural response was done using neural response telemetry. Functional auditory performance indicators and assessment of language development were measured at 12th month after the surgery and auditory verbal therapy. The data were entered in the Microsoft excel and the analysis was done using SPSS software version 18.

Results:.Cochlear nerve deficiency and narrow bony canal for cochlear nerve were the two important findings that consistently correlated with poor post post-operative outcome; and the association was found to be statistically significant (p value < 0.001).No significant association was obtained between dimensions of cochlea or cochlear nerve thickness with intra operative NRT.

Conclusion: HRCT of the temporal bone and MRI play an important role in the evaluation and in the management of congenital sensorineural hearing loss. A thorough knowledge of the imaging anatomy of the middle and inner ear enables the radiologist to provide a structured report, which help the clinician in the proper pre-operative counseling, uneventful surgery and in the prediction of a realistic post-operative outcome.

KEYWORDS:

Cochlear implantation; Cochlear nerve deficiency; Bony canal for cochlear nerve; Neural response telemetry.

INTRODUCTION

Sensorineural hearing loss (SNHL) is the most common congenital sensory deficit, with an incidence of one to three per 1000 live births. Cochlear implantation (CI) has become an accepted treatment for profound deafness in patients who derive only minimal benefit from conventional amplification.(1) Improved hearing through cochlear implantation has been demonstrated to enhance the rate of language acquisition, enable development of spoken language, and advance literacy in deaf children.

Imaging has an important role in deciding candidacy, providing realistic preoperative counselling, and predicting postoperative outcomes. Imaging also provides information about the potential difficulties a surgeon may encounter during the implantation. High resolution computed tomography and high-resolution magnetic resonance imaging complement each other in assessing different aspects of the temporal bone and the auditory pathway in such patients.(2) High resolution computed tomography (HRCT) depicts the minute details of osseous structures, and magnetic resonance (MR) imaging allows visualization of the fluid-filled spaces and the vestibulocochlear nerve. Together, these complementary modalities can aid decision making about the best management strategy by facilitating the identification and characterization of inner ear malformations and any associated neurologic abnormalities. It is important that the radiologist be familiar with the key imaging features when interpreting HRCT and MR images obtained in this patient group.A multidisciplinary approach including early audiological and radiological evaluation of congenital hearing loss, intervention with cochlear implant, and hearing and speech habilitation will surely ensure the optimum outcome.

1. AIMS AND OBJECTIVES

To correlate the outcome of cochlear implantation surgery in the form of intra operative neural response telemetry (NRT), auditory performance and language improvement with the pre operative imaging findings .

2. MATERIALS AND METHODS

2.1 Study design: Cohort study

2.2 Study setting: Department of Radiodiagnosis, Govt. Medical College, Kozhikode.

2.3. Study period: January 2015 – May 2016

2.4 Study subjects:

Children <5yrs who had undergone imaging before cochlear implantation surgery from Department of Radiodiagnosis, Govt.

Medical college, Kozhikode from 2012 to 2016.

2.5.Sample size Sample size was calculated using the formulae, Sample size (N) = 4PQ/ d^2, where P= prevalence, Q= 100-P, d = precision of the study. Assuming the precision of the study being 10 %, the sample size needed (N) = 4PQ/ d^2 =4x30x70/10x10= 84.

2.6 .Inclusion criteria:

Children <5yrs who underwent imaging(CT/MRI/both) for the evaluation of congenital hearing loss and underwent cochlear implantation surgery and AVH (auditory verbal habilitation) who were willing to participate in the study

2.7 Exclusion criteria:

Those who lost to follow up because undergoing auditory verbal habilitation in other centres.

2.8 Study method

Base line characteristics and patient information were collected using a proforma.

Study subjects were evaluated with HRCT, MRI or both using standard protocol.

HRCT temporal bone protocol

16 slice SIEMENS SOMATOM EMOTION CT machine was used to obtain submillimetre (0.625mm) spiral HRCT sections in axial plane with bone windows setting and edge bone enhancement, using the smallest pixel size with reformation of images in coronal and sagittal planes.HRCT evaluated the status of mastoid pneumatisation, thickness of the cortical bone, middle ear aeration, bony labyrinth, internal auditory canal,cochlear and vestibular aqueducts, bony infratemporal facial nerve canal, petrous internal carotid artery canal and jugular fossa, bone based disease of posterior labyrinth.

MRI protocol

WIPRO GE - SIGNA HDx 1.5 T MR machine protocol includes brain T2WI and focused inner ear T2W gradient echo images and heavily T2 weighted images delineated the endolymphatic and peri lymphatic fluid; each part of the membranous labyrinth and each nerve in the internal auditory meatus was delineated by axial or sagittal T2 sequences. MRI is useful in assessing the membranous labyrinth, status of fluid in inner ear, inner ear anatomy (Scala, utricle, endolymphatic duct and sac), inner ear disease, evaluation of the VII & VIII cranial nerves, auditory pathway and cortex.

After the pre-operative radiological and audiology evaluation, the subjects underwent cochlear implant surgery. Intra operative neural response was recorded using neural response telemetry (NRT). Post-operative imaging was done in selected cases. After surgery, the subjects underwent auditory verbal therapy (AVT).

Sequential assessment of language and auditory performance (outcome) were assessed at 3rd, 6th, 9th, 12th month after the auditory verbal therapy. The auditory performance was assessed by functional auditory performance indicators [3] (FAPI) score at 12th month after surgery and auditory verbal therapy. The improvement in language skills was assessed by change in RLA and change in ELA from pre-op status to 12th month after surgery and auditory verbal therapy.

Study variables

Variables studied in HRCT temporal bone

HRCT temporal bone studied the following - External ear, external auditory canal, tympanic membrane, ear ossicles, type of mastoid pneumatisation, middle ear, variations in normal ear anatomy (dehiscent facial nerve canal, high jugular bulb, aberrant carotid artery, mastoid emissary vein), round window, oval window, scala tympani, scala vestibuli, semi-circular canal, epitympanum, hypotympanum, bony labyrinth, cochlea (length, turns, thickness of turns), cochlear nerve canal (CNC) measurement, cochlear and vestibular aqueduct measurement, internal acoustic meatus (IAC- width, height, length), facial nerve course. Cochlear nerve canal less than 1.4 mm was referred as narrow CNC [4]. The vestibular aqueduct was considered to be dilated if it measures >1.5 mm in width at the midpoint between the common crus and its external aperture [5]. The cochlear aqueduct was considered to be dilated if it measures >3 mm [6]. The IAC was defined as normal if the vertical or transverse diameter was 4 mm or more [7], narrow if the diameter was 2 to 3 mm [8] and stenotic if less than 2mm [9]

Variables studied in MRI

In addition to the above variables, MRI evaluated the cochlea, presence and type of inner ear anomalies, endolymphatic sac, measurement of cochlear, vestibular and facial nerves, structural abnormalities of brain, brain stem, vascular anomalies. Inner ear anomalies were classified according to the Sennaroglu classification [5]. A deficient cochlear nerve was defined as a cochlear nerve that is smaller in diameter when compared with the adjacent facial nerve in the midportion of the internal auditory canal [10].

Outcome of the cochlear implant surgery was studied using the following variables.

Outcome of the cochlear implant surgery was studied using intra op neural response telemetry (NRT), functional auditory performance indicators (FAPI), assessment of language development (ALD). NRT indicates the evoked compound action potentials in response to electrical stimuli at the spiral ganglion of cochlea.

FAPI and ALD were assessed in cochlear implant recipient subjects after a minimum 3 months of auditory verbal therapy after surgery. And further evaluations were done at 6th, 9th, 12th months. Evaluation with FAPI generated a child's functional auditory skills profile, which lists auditory skills in a hierarchical order. It had seven hierarchical categories. Each skill was given scores ranging from 0-3, and were added yielding a sum with the minimal score of 0 and maximum of 21. Assessment of Language Development is a diagnostic tool to measure language development in children from birth to 7years and 11months. ALD assessed both Receptive Language Age (RLA) as well as Expressive Language Age (ELA). For statistical purposes, ALD needed to be graded and the score indicates the language age.

Table 1. shows the grading of ALD

Language age (according to ALD)	Score
Birth- 5 months	1
6 – 11 months	2
12-17 months	3
18-23 months	4
24-29 months	5
30-35 months	6
3 years - 3 years 11 months	7
4 years – 4 years 11 months	8
5 years -5 years 11 months	9
6 years – 6 years 11 months	10
7 years – 7 years 11 months	11

Analysis of data

The data was entered in the Microsoft excel and the analysis was done using SPSS software version 18.

Qualitative variables were described in percentages and quantitative variables are described in mean and standard deviation.

Paired t test, one way ANOVA and Bonferroni tests (post hoc analysis) were used to find out the association between the outcome and quantitative variables.

Association between neural response telemetry (NRT), with imaging findings was calculated by Chi square test.

The correlation between FAPI, change in RLA, change in ELA and cochlear length, number of turns, width of inner, middle and outer turns was found using Pearson correlation. p value less than or equal

to 0.05 was taken as statistically significant.
Ethical concerns

The study protocol was submitted to institutional ethics committee, Government medical college, Kozhikode and got approved before starting the study. The parents of the subjects were explained about the study and informed written consent were obtained from both the parents.

3. RESULTS

3.1.1 Age of subjects at the time of surgery

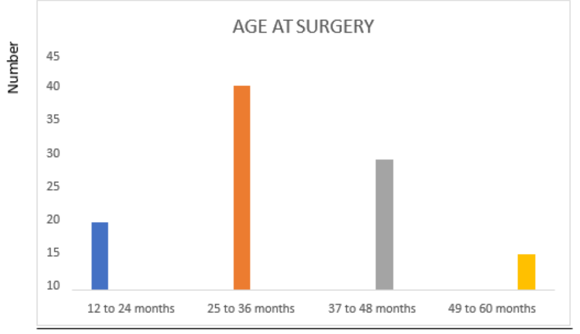

Figure 3. 1. shows distribution of age at the time of surgery

Out of 84 subjects, majority (n= 39, 46.4 %), were between the age of 25 to 36 months followed by 25 (29.8 %) subjects who were between 37 to 48 months.

3.1.2 . Sex distribution

Out of 84 subjects, 44 (52.4 %) were females and 40(47.6 %) were males.

3.2. Intra op NRT

Table 3.1 shows distribution of Intra op NRT

	FREQUENCY	PERCENTAGE
PRESENT	75	89.3
ABSENT	9	10.7

Intra op NRT was obtained in 75 (89.3 %) of the subjects, whereas intra op NRT was not obtained in 9 (10.7 %) subjects.

3.3 ASSOCIATION OF VARIOUS IMAGING VARIABLES WITH INTRA OP NRT

3.3.1 Association of cochlear nerve deficiency with intra op NRT

Table 3.2 shows association of cochlear nerve deficiency with intra op NRT

Cochlear nerve deficiency	Intra op NRT	
	PRESENT	ABSENT
ABSENT	17 (89.2 %)	2 (10.5 %)
PRESENT	58 (89.2 %)	7 (10.8 %)

χ^2 0.001 df 1 p value 0.976

17 subjects who got intra op NRT had cochlear nerve deficiency. 58 subjects who got intra op NRT had normal cochlear nerve thickness. And the association is between cochlear nerve deficiency and intra op NRT is not statistically significant. (p value 0.976).

3.3.2 Association of cochlear nerve canal (CNC) thickness with intra op NRT

Table 3.3 : Shows association of cochlear nerve canal thickness with intra op NRT

CNC THICKNESS	INTRA OP NRT	MEAN	SD	P value
	Present (75)	1.84	0.24	0.593
	Absent (9)	1.798	0.26	

Paired t-test 0.537 df 82 p value 0.593

Though the mean cochlear nerve canal thickness (1.84 ±0.24) is more in subjects (n=75) who got intra op NRT, compared to those who (n= 9) didn't get intra op NRT (mean 1.798 ±0.26), the association of cochlear nerve canal thickness with intra op NRT is not statistically significant (p value 0.593)

3.3.3 Association of dimensions of cochlea with intra op NRT

Table 3.4 : Shows association of dimensions of cochlea with intra op NRT

DIMENSIONS OF COCHLEA	NRT	MEAN	STANDARD DEVIATION	P value
COCHLEAR DUCT LENGTH	ABSENT (9)	25.02	2.22	0.054
	PRESENT (75)	26.36	1.91	
NUMBER OF COCHLEAR TURN	ABSENT (9)	2.57	0.10	0.523
	PRESENT (75)	2.61	0.20	
WIDTH OF INNER TURN	ABSENT (9)	0.79	0.08	0.747
	PRESENT (75)	0.78	0.13	
WIDTH OF MIDDLE TURN	ABSENT (9)	1.73	0.09	0.488
	PRESENT (75)	1.77	015	
WIDTH OF BASAL TURN	ABSENT (9)	1.88	0.21	0.372
	PRESENT (75)	1.96	0.29	

Though the mean cochlear duct length (26.36 ±1.91) is more in subjects (n=75) who got intra op NRT, compared to those who (n= 9) didn't get intra op NRT (mean 25.02±2.22), the association of cochlear duct length with intra op NRT is not statistically significant (p value 0.054)

Though the mean number of cochlear turns (2.61± 0.20) is more in subjects (n=75) who got intra op NRT, compared to those who (n= 9) didn't get intra op NRT (mean 2.57 ±0.10), the association of number of cochlear turns with intra op NRT is not statistically significant (p value 0.0523)

The association of width of inner, middle and basal cochlear turns with intra op NRT is not statistically significant (p value 0.372)

3.3.4 Association of post op imaging with intra op NRT

Table 3.5 : Shows association of post op imaging with intra op NRT

INTRA OP NRT	POST OP IMAGING	
	NOT DONE	DONE
PRESENT	72(96 %)	3(4%)
ABSENT	1(11.1%)	8(88.9 %)

χ^2 50.883 df 1 p value < 0.001

72 subjects who had no post op imaging got NRT intra operatively and 8 subjects had post op imaging who didn't get intra op NRT. And the association of post op imaging with intra op NRT is statistically significant. (p value <0.001)

3.4. IMPROVEMENT IN AUDITORY PERFORMANCE AND LANGUAGE AFTER CI SURGERY AND AUDITORY VERBAL THERAPY

The FAPI score after surgery and auditory verbal therapy at 12th month is (13.04 ±2.8)

The mean change in RLA from pre-op to post surgery and auditory verbal therapy at 12 months is (2.7± 1.28).

The mean change in ELA score from pre-op to post surgery and auditory verbal therapy at 12 months is (2.38 ± 1.11)

3.4.1. Improvement in RLA after surgery and auditory verbal therapy

TABLE 3.6 - Shows improvement in receptive language age (RLA)

OUTCOME RLA	Pre-op RLA	RLA at 3rd month	RLA at 6th month	RLA at 9th month	RLAat 12th month
Mean Age	1.98	3.01	3.57	4.13	4.68
Standard deviation	0.969	1.322	1.347	1.220	1.416

Paired t- test df = 83 p value < 0.001

Paired t test was done to compare the pre RLA before surgery and RLA at 3rd, 6th,9th ,12th month after surgery and auditory verbal therapy. We observed that the mean receptive language age was improved from 1.98 to 4.68 at 12 months. The association was found to be statistically significant (p value < 0.001).

3.4.2. Improvement in ELA after surgery and auditory verbal therapy

Table 3.7 : show improvement in expressive language age (ELA)

OUTCOME ELA	Pre-op ELA	ELA at 3rd month	ELA at 6th month	ELA at 9th month	ELAat 12th month
Mean Age	**1.90**	**2.76**	**3.39**	**3.81**	**4.29**
Standard deviation	0.913	1.137	1.162	1.135	1.168

Paired t test df = 83 p value < 0.001

Paired t - test was done to compare the pre-op ELA and ELA at 3rd, 6th ,9th ,12th month after surgery and auditory verbal therapy. We observed that the mean expressive language age was improved from 1.90 to 4.29 at 12th month. The association was found to be statistically significant (p value < 0.001).

3.5. ASSOCIATION OF IMAGING FINDINGS WITH THE OUTCOME

3.5.1. Association between cochlear nerve deficiency and outcome

Table 3.8: shows association between cochlear nerve deficiency and the outcome

OUTCOME	COCHLEAR NERVE DEFICIENCY	MEAN	SD	p value *
FAPI score	Present (19) Absent (65)	9.53 14.06	2.37 2.04	**0.001**
Change in RLA	Present (19) Absent (65)	1.63 3.01	0.76 1.23	**0.001**
Change in ELA	Present (19) Absent (65)	1.47 2.64	0.96 1.02	**0.001**

* p value was found using paired t test

The mean FAPI score was 14.06 ± 2.04 in subjects who didn't have cochlear nerve deficiency, compared to those who had cochlear nerve deficiency (mean FAPI score was 9.53 ± 2.37).

Similarly, change in RLA is high (mean change 3.01 ± 1.23) among the subjects who had normal cochlear nerve, compared to those with cochlear nerve deficiency (mean change 1.63 ± 0.76).

Similarly, change in ELA is high (mean change 2.64 ± 1.02) among the subjects had normal cochlear nerve, compared to those with cochlear nerve deficiency (mean change 1.47± 0.96).

And the association between cochlear nerve thickness and FAPI score, change in RLA, change in ELA is statistically significant (p value <0.001)

3.5.2. Association between cochlear nerve canal thickness and outcome

Table 3.9 : shows association between cochlear nerve canal thickness and outcome

OUTCOME	COCHLEAR NERVE CANAL THICKNESS	MEAN	SD	p value *
FAPI score	Narrow (4) Normal (80)	10.25 13.18	3.69 2.75	**0.044**
Change in RLA	Narrow (4) Normal (80)	1.75 2.75	1.50 1.25	0.127
Change in ELA	Narrow (4) Normal (80)	1.25 2.44	0.50 1.11	**0.037**

* p value was found using paired t test

The mean FAPI score among those subjects (n=80) who had normal cochlear nerve canal is 13.18 ±2.75, which was higher, compared to those (n=4) who had narrow cochlear nerve canal.

And the association between cochlear nerve canal thickness and FAPI score is statistically significant (p value 0.044).

Mean change in ELA is high (2.44 ± 1.11) among the subjects (n= 80) had normal cochlear nerve canal, compared to those who(n=4) had narrow cochlear nerve canal. And the association between change in ELA and cochlear nerve canal thickness is statistically significant. (p value 0.037)

Mean change in RLA is high (2.75± 1.25) among the subjects had normal cochlear nerve, compared to those who had abnormal normal cochlear nerve canal thickness. And the association between change in RLA and cochlear nerve canal thickness is not statistically significant (P value 0.127).

3.5.3 ASSOCIATION OF DIMENSIONS OF COCHLEA WITH OUTCOME

Table 3.10 : shows association of dimensions of cochlea with outcome

DIMENSIONS OF COCHLEA	FAPI score	P value	CHANGE IN RLA	P value	CHANGE IN ELA	p value*
COCHLEAR LENGTH	-0.008	0.94	-0.120	0.28	-0.127	0.25
COCHLEAR TURN	0.079	0.48	0.026	0.81	-0.035	0.76
INNER TURN	-0.001	0.99	0.077	0.49	0.049	0.66

MIDDLE TURN	0.058	0.60	-0.043	0.70	-0.014	0.90
BASAL TURN	-0.215	0.05	-0.109	0.33	-0.081	0.46

* p value was found using Pearson correlation

The correlation between FAPI score, change in RLA, change in ELA and cochlear length, number of turns, width of inner, middle and basal turns was found using Pearson correlation (If the correlation coefficient is less than -0.2, there is no correlation; if it is -0.2 to -0.4, mild correlation; if -0.42 to -0.6, moderate correlation). And correlation is significant at the 0.05 level. There was a mild negative correlation between FAPI score and width of basal turn. And the association between width of basal turn and the FAPI score is statistically significant (p value 0.05)

3.5.4. Association between width of internal acoustic meatus (IAC) and outcome

Table 3.11 : shows association between width of internal acoustic meatus and outcome

OUTCOME	WIDTH OF IAC	MEAN	SD	p value*
FAPI score	Narrow (9)	11.33	3.31	0.057
	Normal (75)	13.24	2.74	
Change in RLA	Narrow (9)	2.00	1.12	0.081
	Normal (75)	2.79	1.28	
Change in ELA	Narrow (9)	1.78	1.20	0.087
	Normal (75)	2.45	1.09	

* p value was found using one way ANOVA test

One way ANOVA test was done to find out the association between the outcomes (FAPI, change in RLA, change in ELA) and IAC width. (IAC width was divided into three groups, (>4 mm – normal, 3-4mm is narrow, less than 3mm - stenotic). Though there was better outcome in subjects who had normal IAC width, the association between IAC width and the outcome is not statistically significant (p value 0.057).

3.5.5. Association between abnormality in brain parenchyma and outcome

Table 3.12 : shows association between abnormality in brain parenchyma and outcome

OUTCOME	BRAIN PARENCHYMA	MEAN	SD	p value *
FAPI score	Abnormal (18)	11.94	3.28	0.066
	Normal (66)	13.33	2.66	
Change in RLA	Abnormal (18)	2.72	1.31	0.941
	Normal (66)	2.69	1.28	
Change in ELA	Abnormal (18)	2.33	1.33	0.840
	Normal (66)	2.39	1.07	

* p value was found using paired t test

The mean FAPI score among those subjects who had normal brain parenchyma is 13.33 ±2.66, which was higher, compared to those who had abnormal brain parenchyma (mean score 11.94 ± 3.28)

And the association between abnormality in brain parenchyma and FAPI score is not statistically significant (p value 0.066).

The association between change in ELA and abnormality in brain parenchyma is not statistically significant (p value 0.941).

The association between change in ELA and abnormality in brain parenchyma is not statistically significant (p value 0.840).

REPRESENTATIVE CASES

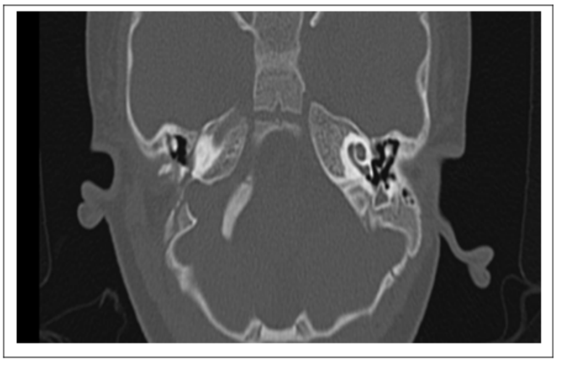

Figure 1:HRCT temporal bone axial image showing microtia and hypo plastic external auditory canal on the right and overhanging pyramidal eminence obscuring round window on left.

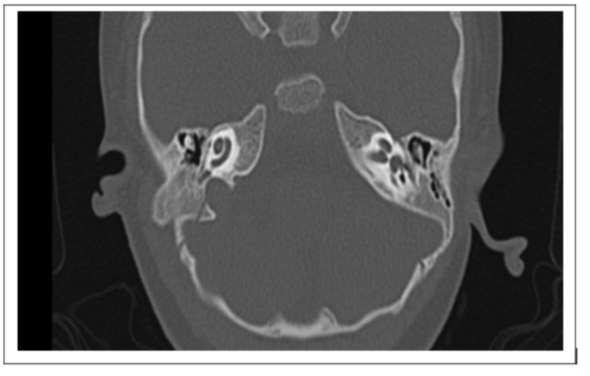

Figure 2 : HRCT temporal bone axial image showing hypoplastic cochlea on the right side

Figure 3. Volume rendered image showing Mondini's dysplasia with 1.5 cochlear turns , cystic apex and dilated vestibule

Figure 4: HRCT temporal bone axial image showing narrow cochlear aperture on the right side .

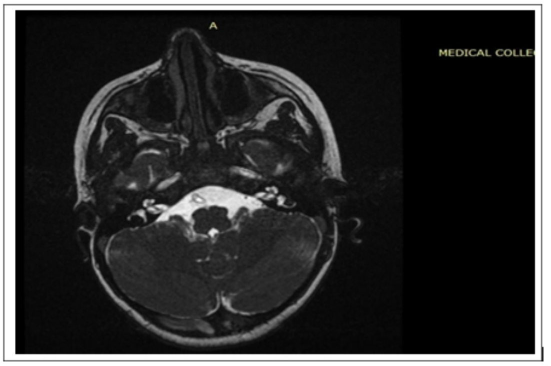

Figure 5: Axial MRI FIESTA image showing normal Cochlear nerve, which is thicker than the vestibular nerve within the right IAC

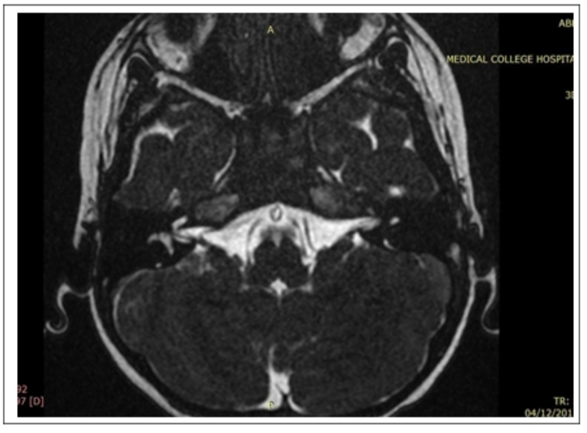

Figure 6: Axial MR FIESTA image showing deficient cochlear nerve, which is smaller than the ipsilateral facial nerve ,on the right side

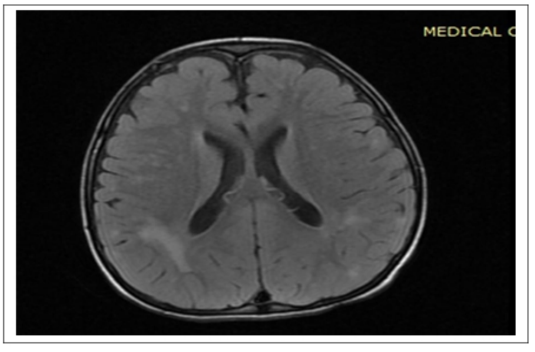

Figure 7: MRI brain T2 FLAIR sequence showing the subcortical and periventricular white matter hyper intensities bilaterally.

Figure 8. MRI brain FIESTA sequence showing the AICA loop over the vestibulocochlear nerve complex in the left IAC .

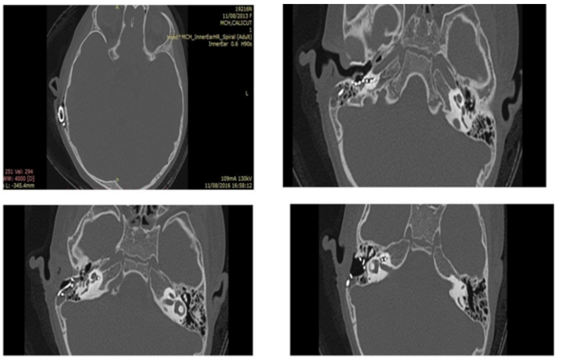

Figure 9 : HRCT temporal bone axial image showing cochlear implant with electrodes within the basal and apical turns of right cochlea

DISCUSSION

A cohort study was conducted to study the various imaging findings which influenced the outcome after cochlear implant surgery and auditory verbal therapy. A total of 84 subjects less than 5 years of age who underwent imaging, cochlear implantation and auditory verbal therapy were included.

In the present study, majority of the children were between the age of 25 to 36 months at the time of cochlear implantation, 52.4 % were females and 47.6 % were males. In a study by May Mederake , Birjit et al[11], the mean age at implantation was 14.3 ± 5.5 months. According to a study by Kang DH et al [12], the mean age at diagnosis of deafness in patients with delayed CI was 33.1±14.9 months in the poor performance group and 27.2±15.3 months in the good performance group. The association between age at surgery and the post-operativeoutcome was found to be statistically significant with p value 0.02. Earlier auditory stimulation enables children to understand the spoken language better and to use spoken language themselves.

In the present study, the patients underwent HRCT and MRI examinations. Out of the 84 subjects, one subject (1.2 %) had microtia (hypo plastic external ear), hypo plastic external auditory canal, ear ossicles and tympanic cavity on right. The patient was operated on normal ear on left side and so there is no significance in comparing these imaging pathologies with the outcome.

The present study showed the mean width of IAC on right side was 4.44 ± 0.606 and on left was 4.60 ± 0.668. The IAC was defined as stenotic if less than 2mm[9]. Though there was better outcome in subjects who had normal IAC width, the association between IAC width and the outcome was not statistically significant. But , a study by Walton et al (13) demonstrated that the children with AN and normal cochlear nerves are likely to obtain greater benefit from a cochlear implant than those with abnormal cochlear nerves or IAC. The present study evaluated the inner ear anomalies as described by Sennaroglu [5] and had 2 patients with cochlear anomalies and dilated vestibular aqueduct (2.4 %), out of which 1 had bilateral cochlear dysplasia of Type II incomplete partition(Mondini dysplasia). 1 subject had unilateral hypo plasticcochlea on right with 1.5 turns, who underwent CI on left. The subject who had Mondini dysplasia, had partial insertion of electrode, but obtained intra op NRT.

The prevalence of inner ear anomaly is too low in the present study, compared to the study of Dagkiran et al [14] in 300 subjects, where they got a prevalence of 20.26 % . Since the prevalence was too low, the post-operative hearing outcome could not be compared. This may be attributed to the fact that, the subjects with cochlear anomalies were not operated as frequent as the other group, as our study subjects included subjects having pre-op imaging, who underwent surgery subsequently. According to study by Kang et al (12), it is universally accepted that children with more severe inner ear anomalies have poorer hearing outcomes after CI than those with less severe anomalies.

In the present study, there was no significant correlation with the change in RLA and change in ELA with the dimensions of cochlea including cochlear duct length, number of turns and width of turns. In a study by Finley et al [15], no statistically-significant correlation between cochlear size and apical-most insertion depth or outcome measures were found, similar to the present study.

Estimation of cochlear nerve integrity may be of great importance while selecting the candidates for surgery, as the absolute contraindication for CI surgery include complete aplasia of cochlea / absence of cochlear nerve. According to the study by Govaerts et al [16], the cochlear nerve was described as normal if it was the same size as or larger than the facial nerve. The nerve was deficient if smaller than the facial nerve, rudimentary when the vestibulocochlear nerve [VCN] complex unbranching. Higher mean FAPI score (14.06 ± 2.04) was obtained in subjects who didn't have cochlear nerve deficiency, compared to those who had cochlear nerve deficiency (mean FAPI score was 9.53 ± 2.37). Similarly, the change in RLA was higher (mean change 3.01 ± 1.23) among the subjects who had normal cochlear nerve, compared to those with cochlear nerve deficiency (mean change 1.63 ± 0.76). Similarly, change in ELA was higher as well (mean change 2.64 ± 1.02) among the subjects who had normal cochlear nerve, compared to those with cochlear nerve deficiency (mean change 1.47± 0.96). On analysis, the association between cochlear nerve thickness and the FAPI score, the change in RLA, the change in ELA is statistically significant (p value <0.001). This finding is consistent with the study by Walton et al [13], who found out that children with auditory neuropathy and cochlear nerve deficiency have worse cochlear implant outcomes when measured by speech perception score than those with auditory neuropathy and normal cochlear nerves.

The present study found out no significant association between cochlear nerve deficiency and intra op NRT .The study by Walton et al [13] had found out the association between implant evoked electric auditory brainstem response waveform score (EABR) which was employed instead of intra op NRT, to assess the integrity of the auditory pathway up to the brainstem and quantify brainstem response thresholds for programming purposes. In a study by Ji et al [17], patients with auditory neuropathy (AN), waveforms of neural response telemetry (NRT) could be present, showing characteristics of low incidence, low differentiation, and large variation. However the current study evaluated the presence or absence of NRT , rather than its various characteristics, as studied by Ji et al [17]. The presence of valid ECAP waveforms measured by NRT usually indicated that the electrode has been activated and the neurons have a good response to electrical stimuli. This could be an objective indicator of hearing reconstruction. The ECAP having significant correlation with postoperative hearing and speech performance has been studied by Kim et al [18]. But in the current study , the association between intra op NRT and the FAPI score was not statistically significant in contrast to the study of Kim et al [18] who studied the slope of ECAP rather than the mere presence or absence of neural response .

In the present study, the mean cochlear nerve canal measurement on the right was 1.84 ± 0.242 and on the left, was 1.87 ±0.230. Cochlear nerve canal measurement less than 1.4 mm was considered as narrow [7]. In the present study, the association between cochlear nerve canal measurement and the FAPI score and change in expressive language age was found to be statistically significant. Thus the present study is in concordance with study by Wilkins et al [19] who described narrow cochlear nerve canal as the indicator of cochlear nerve deficiency. Though the mean cochlear nerve canal thickness (1.84 ±0.24) was more in subjects in whom intra op NRT was present, the association of cochlear nerve canal thickness with intra op NRT was not statistically significant in the present study.

In the present study, 18 subjects (21.4 %) had abnormal T2 and FLAIR hyper intense signals in the periventricular and subcortical white matter. The mean FAPI score among those subjects who had normal brain parenchyma is 13.33 ±2.66, which was higher, compared to those who had abnormal brain parenchyma (mean score 11.94 ± 3.28). But we couldn't find any significant association between brain parenchymal abnormality and the FAPI score. In a study by Jonas et al [20], the most common abnormality detected was white matter changes (70%) and this was found in 22% of all patients investigated ; similar to what we observed in our study. In another study by Hong et al [21], abnormal MRI with varying degrees of white matter changes was noted in 10 (18%) children. Two of the 10 patients (20 %) demonstrated significant delay and difficulties with postoperative CI performance.

CONCLUSION

A cohort study was conducted to study the various imaging findings which influenced the outcome after cochlear implant surgery and auditory verbal therapy A total of 84 subjects less than 5 years of age were examined during the course of the study.

Majority of the children were between the age of 25- 36 months, the earlier the age at cochlear implantation, the better was the outcome found to be.. No association was found between intra op NRT with cochlear nerve thickness , dimensions of cochlea , cochlear nerve canal width and post-operative outcome.

There was an improvement in auditory performance and language during the follow up period. Cochlear nerve deficiency, narrow bony canal for cochlear nerve were the imaging characteristics that were found to be associated with the poor post- operative outcome. Hence, pre-operative evaluation with HRCT complimented by MRI plays a main role in the management of congenital sensorineural hearing loss with a greater impact on the surgical approach, the intra operative challenges and the anticipated post-operative outcome. Thus, the present study assumes its significance in that, it addresses a relatively newer imaging methodology, filtering from the very large pool of imaging data, the relevant ones which can influence the post-operative hearing and language improvement.

LIMITATIONS OF THE STUDY

We followed up the subjects for 1 year after cochlear implantation surgery and auditory verbal therapy. Long term improvement in speech and language was not studied as both these are evolving over a long period of time.

RECOMMENDATIONS

Radiologists should be able to provide a structured report based on combined HRCT and MRI check lists to ensure the clinician, aware of all relevant information prior to the cochlear implantation surgery. And this will help to provide proper pre-operative counselling, uneventful surgery, and realistic post-operative outcome. It is recommended to conduct similar study in a larger population where more children with inner anomalies are getting operated and being followed up for longer periods

REFERENCES

1. Witte RJ, Lane JI, Driscoll CLW, Lundy LB, Bernstein MA, Kotsenas AL, et al. Pediatric and Adult Cochlear Implantation. RadioGraphics. 2003 Sep;23(5):1185– 200.
2. Vaid S, Vaid N. Imaging for cochlear implantation: Structuring a clinically relevant report. Clin Radiol. 2014 Jul;69(7):e307–22.
3. FERREIRA K, MORET ALM, BEVILACQUA MC, JACOB R de ST. Translation and adaptation of functional auditory performance indicators (FAPI). J Appl Oral Sci. 2011;19(6):586–98.
4. Fatterpekar GM, Mukherji SK, Alley J, Lin Y, Castillo M. Hypoplasia of the bony canal for the cochlear nerve in patients with congenital sensorineural hearing loss: initial observations. Radiology. 2000 Apr;215(1):243–6.
5. Sennaroglu L. Cochlear Implantation in Inner Ear Malformations — A Review Article. Cochlear Implants Int. 2010 Mar;11(1):4–41.
6. Swartz JD, Loevner LA. Imaging of the Temporal Bone. Thieme; 2011. 622 p.
7. Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kertesz TR, Shelton. C.Imaging findings of cochlear nerve deficiency. AJNR Am J. Neuroradiol.2002 Apr;23(4):635-43
8. Casselman JW, Kuhweide R, Deimling M, Ampe W, Dehaene I, Meeus L. Constructive interference in steady state-3DFT MR imaging of the inner ear and cerebellopontine angle. AJNR Am J Neuroradiol. 1993 Feb;14(1):47–57.

9. Valvassori GE, Pierce RH. THE NORMAL INTERNAL AUDITORY CANAL. Am J Roentgenol Radium Ther Nucl Med. 1964 Dec;92:1232–41.
10. Kutz JW, Lee KH, Isaacson B, Booth TN, Sweeney MH, Roland PS. Cochlear implantation in children with cochlear nerve absence or deficiency. Otol Neurotol Off Publ Am Otol Soc Am Neurotol Soc Eur Acad Otol Neurotol. 2011 Aug;32(6):956–61.
11. May-Mederake B. Early intervention and assessment of speech and language development in young children with cochlear implants. Int J Pediatr Otorhinolaryngol. 2012 Jul;76(7):939–46.
12. FERREIRA K, MORET ALM, BEVILACQUA MC, JACOB R de ST. Translation and adaptation of functional auditory performance indicators (FAPI). J Appl Oral Sci. 2011;19(6):586–98.
13. Walton J, Gibson WPR, Sanli H, Prelog K. Predicting cochlear implant outcomes in children with auditory neuropathy. Otol Neurotol. 2008;29(3):302– 309.
14. Dagkiran M, Dagkiran N, Surmelioglu O, Balli T, Tuncer U, Akgul E, et al. Radiological Imaging Findings of Patients with Congenital Totally Hearing Loss. J Int Adv Otol. 2016 Jun 20;12(1):43–8.
15. Finley CC, Holden TA, Holden LK, Whiting BR, Chole RA, Neely GJ, et al. Role of electrode placement as a contributor to variability in cochlear implant outcomes. Otol Neurotol Off Publ Am Otol Soc Am Neurotol Soc Eur Acad Otol Neurotol. 2008 Oct;29(7):920–8.
16. Govaerts PJ, Casselman J, Daemers K, De Beukelaer C, Yperman M, De Ceulaer
17. Ji F, Li J-N, Liu K, Jiao Q-S, Sun L, Hong M-D, et al. NRT test in auditory neuropathy patients with cochlear implants. Acta Otolaryngol (Stockh). 2014 Sep;134(9):930–42.
18. Kim J-R, Abbas PJ, Brown CJ, Etler CP, O'Brien S, Kim L-S. The relationship between electrically evoked compound action potential and speech
19. Wilkins A, Prabhu SP, Huang L, Ogando PB, Kenna MA. Frequent association of cochlear nerve canal stenosis with pediatric sensorineural hearing loss. Arch Otolaryngol Head Neck Surg. 2012 Apr;138(4):383–8.
20. Jonas NE, Ahmed J, Grainger J, Jephson CG, Wyatt ME, Hartley BE, et al. MRI brain abnormalities in cochlear implant candidates: how common and how important are they? Int J Pediatr Otorhinolaryngol. 2012 Jul;76(7):927–9.
21. Hong P, Jurkowski ZC, Carvalho DS. Preoperative cerebral magnetic resonance imaging and white matter changes in pediatric cochlear implant recipients. Int J Pediatr Otorhinolaryngol. 2010 Jun;74(6):658–60