RADIOLOGICAL FINDINGS IN COCHLEAR IMPLANT CASES IN PRELINGUAL DEAFNESS- A STUDY AND ANALYSIS OF MICHEL'S APLASIA AND ITS VARIATIONS

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ABSTRACT

BACKGROUND

The incidence of Prelingual deafness with bilateral profound sensory neural hearing loss varies among different regions with overall prevalence rate of one case per 1000 live births. Overall, there are more than 40 million such children all over the World. Hearing losses may be genetic or non-genetic. Genetic hearing losses (50%) may be syndromic (15%) or non-syndromic (35%). Genetic hearing loss may be Autosomal or X-linked, Dominant or Recessive, Syndromic or Non-Syndromic. Adequate radiological assessment and confirmation of select cases is utmost important before proceeding to cochlear implant surgery.

This study aims at a statistical analysis of various radiological presentation in bilateral profound sensory neural hearing loss in prelingual deafness children selected for cochlear implant in our Institution for a period of one year from January 2016 to January 2017, with study and analysis of Michel's Aplasia and its variations included in the study.

MATERIALS AND METHODS

The study is done as a retrospective study at Vellore Medical College at the ENT Department for a period of one year from Jan 2016- Jan 2017, based on the radiological presentation of 20 children included in the study group with other prior investigations. Interpretation was done based on the inner ear anomalies and individual variations are analysed. Study Design- Retrospective study.

RESULTS

Radiological analysis of 20 cases included in the study group of Prelingual deafness candidates with bilateral profound sensory neural hearing loss selected for cochlear implant showed that three cases had Michel's Aplasia with individual radiological variations which were analysed, while other cases had radiologically normal middle and inner ear.

CONCLUSION

Inner ear anomalies of children with bilateral profound sensory neural hearing loss are variable. Although, incomplete partition type and cochlear Hypoplasia type are common according to International studies, Michel's Aplasia (15%) is the most common anomaly in our study with some radiological variations.

KEYWORDS

Ear, Abnormalities, Sensory Neural Hearing Loss, Imaging, CT, MRI.

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BACKGROUND

Childhood deafness presents special problems in the assessment and rehabilitation, as it is usually present before language has been acquired (prelingual). Children do not complain of impaired hearing and even parents and carers are known to be unaware of the deficit in at least 30% of affected children. If a permanent effect upon language development is to be avoided, children with hearing loss must be found and treated promptly before the critical period for language acquisition has passed. Children born with sensorineural hearing loss may never acquire normal speech and language.[1]

Otitis media is also prevalent in disadvantaged and learning disabled children who are already at risk of

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language delay. Early intervention in chronic secretory otitis media has become an area of controversy, but hearing loss in the presence of language or development delay requires treatment. The management of hearing impairment in children is most effective if undertaken by a consultantled multidisciplinary team including audiology, paediatrics, genetics, otolaryngology, plastic surgery, education, speech therapy, mental health, social services and voluntary bodies.[1]

Congenital conductive hearing impairment is uncommon and is associated with congenital defects of the outer and middle ear. Middle ear defects such as congenital stapes fixation with or without the otobrachiorenal syndrome or osteogenesis imperfecta, are easily overlooked in the absence of visible stigmata. Hearing impairment in children with craniofacial abnormalities poses particular management problems because of the psychological effects upon the child and the parents. Sensorineural hearing impairment is present in 1-1.5/1000 children and can be congenital or of early onset.[1]

Early onset hearing loss can begin at any time during the first year and is progressive during the preschool years. Progressive deafness accounts for 15-20% of preschool children with sensorineural hearing loss. These children may not be detected by a neonatal hearing screen. The prevalence of hearing loss varies between populations. Those in which consanguineous marriages are common may have a prevalence of 12/1000 children, and graduates of special care baby units or neonatal intensive care have a prevalence of approximately 15/1000. Precise rates will be individual to a particular community.[1]

Factors that increase the prevalence of hearing impairment are a family history of deafness, congenital abnormalities of the head and neck, prenatal, perinatal infection, prematurity, low birth weight, anoxia, high bilirubin concentrations and parents who are first cousins. The mumps, measles and rubella vaccination programme has reduced the prevalence of congenital deafness due to these infections, but cytomegalovirus, herpes and meningitis from any organism are also associated with congenital and perinatal deafness.[1]

To improve detection neonatal hearing screens are being established, whereas infant surveillance and screening aims to detect those not found at birth. Neonatal hearing screening is either universal or if resources are more limited selective. A selective screen is targeted at those neonates with the highest risk of congenital deafness. A neonatal screen is insufficient on its own as 15-20% of children with prelingual hearing loss will develop that loss in the first 2 years of life. All children should be screened in the neonatal period using transient-evoked otoacoustic emissions (TEOAEs).[1]

Another area of development is in hearing preservation surgery, allowing combined acoustical and electrical speech processing following implantation. The preservation of low frequency hearing leads to improved word understanding in noise and better music appreciation. Essentially, themain criterion for candidacy for CI has been audiological performance in both the aided and unaided state. Current agreed audiological criteria include failure to achieve aided scores higher than 30%.[2]

Imaging is an essential element in the workup of a patient for CI, the two modalities available being High Resolution Computed Tomography (HRCT) and Magnetic Resonance Imaging (MRI). Both have their proponents. HRCT demonstrates the bony architecture of the temporal bone. This supports the accuracy in detecting ossification ranged from 86 to 95%. Loppenen et al have demonstrated a method of producing a three-dimensional (3D) model of the temporal bone before cochlear implantation using helical scanning to acquire high-resolution data from the middle and inner ear. The data are then processed to a form suitable for creating a high accuracy 3D model.[2]

MRI scanning is used increasingly in assessment for CI, with centres using techniques like T2-weighted imaging. MRI is helpful in CI candidates for identifying soft tissue abnormalities and areas of residual cochlear patency in cases of labyrinthitis ossificans. In 4 of 13 cases (31%), the MRI provided information not available on HRCT alone. MRI can also show abnormalities in the internal auditory meatus such as vestibular schwannoma, cochlear nerve aplasia and other abnormalities in the central auditory pathways. In summary, both MRI and HRCT are used in the assessment of these patients prior to surgery with the choice of modality often dependant on the facilities available. MRI is more accurate at identifying cochlear dysplasia, LVA and the presence of the cochlear nerve.[2]

CT technique- In conventional tomography, the image of a thin section of any part of the body is obtained by blurring out the information from unwanted adjacent areas. In Computerised Tomography (CT) also the image of a thin section is obtained, but here a computer mathematically constructs the image of a very thin section of the body by using data arising only from the section of interest, leaving out totally the data obtained from the adjacent anatomical areas. Images are generated from cross-sections of the anatomy that are oriented perpendicular to the axial dimensional of the body. The data obtained from the detectors is computed by the computer and the image of a thin slice of the part of the body to be studied is generated.[3]

The temporal bone is a complex structure of dense bone and air-filled space; hence, High Resolution CT (HRCT) with thin sections (1-2 mm) and special bone algorithm are essential requisites for visualising its intricate anatomy. It must be appreciated that routine CT scans with thicker slices and routine algorithms as used for the brain are very unsuitable if not totally useless in evaluating temporal bone anatomy. Contrast enhancement is normally not required for pathology or bones or air spaces; it is essential in vascular lesion, disruption of Blood Brain Barriers (BBB) or soft tissue changes contrast enhanced CT (CECT) is done. Multiplanar CT reformations can display the complex anatomy of the temporal bone almost in all projections.[3]

Usually during CT scan, only axial and direct coronal sections are taken, but with the advent of improved technology, reconstruction/reformation of images at various planes is also possible. This means that we can have not only coronal and axial cuts during CT, but also curved or sinusoidal reconstruction of the CT image with improved software application. Air CT meatography is helpful for identifying small acoustic neuromas. In this process, 2 to 4 mL of air is injected into subarachnoid space through lumbar puncture. Oxygen or carbondioxide instead of air is preferred, because they are absorbed earlier than air and also cause less post-procedural headache.[3]

Immediately after the subarachnoid injection of the air or gas, the patient is asked to raise the head and trunk supported by the elbow such that the gas enters the CP angle. As soon as this occurs, the patient complains of pain in the ear. The head is lowered and cuts are taken at 1 to 1.5 mm interval, preferably by a high resolution CT scanner. If there is no tumour in the internal auditory meatus, then its contours are depicted as a black area and only 7th and 8th cranial nerves are seen as white streaks. If a small intracanalicular tumour is present, the air or gas outlines the contours of the swelling on the nerve which appears as a white area on a black background. If the tumour completely fills the internal auditory meatus then the air fails to enter the meatus and no black area is seen in the internal auditory meatus.[3]

MRI is the modality of imaging on the phenomenon of chemical magnetic resonance. The MR image shows a high contrast between various tissues due to the difference, at which magnetised nuclei of the different tissues resume their original states, thereby emitting different radiofrequency signals. MRI studies can be carried out using many different techniques depending on the suspected abnormality. T1 weighted spin-echo coronal or axial images are usually done without contrast. T2 weighted axial spin-echo images accurately display the CSF spaces and many pathologies at high signal intensity. MRI study is valuable for evaluation of blood vessel related disorders with many gradient echotechniques; flowing blood can be seen as high signal regions and phase imaging can be used to quantify velocity and flow volumes.[3]

Otologic sections of patients for cochlear implants-
It is helpful to classify potential cochlear implant subjects into the following groups-

i. Acquired post-lingually deaf adults.
ii. Acquired post-lingually deaf children.
iii. Acquired pre-lingually deaf children.
iv. Acquired pre-lingually deaf adults.
v. Congenital deaf adults.[4]

Management of the child with hearing impairment is a monumental challenge to the clinical audiologist. In learning to understand the spoken language of others and to speak it there is no adequate substitute for an intact auditory system. Without normal or near-normal hearing, it is extremely difficult to acquire an adequate oral communication system. Because so much of the language-learning process occurs within the first few years of life, there has been considerable emphasis on early identification and intervention for young hearing-impaired children.[5]

Impaired children who exhibit the best spoken language and show the most satisfactory progress in school are those who have had the benefit of early identification and intervention. Early intervention is essential to the successful development of speech and language. The intervention must include adequate parent-infant management, wearable amplification, speech and language training and development of perceptual and cognitive skills.[5]

Hereditary hearing loss and deafness may be conductive, sensory neural or combination of both; syndromic (associated with malformation of external ear canal) or non-syndromic (not associated with visible anomalies) and prelingual (before language develops) or post-lingual (after language develops). The genetic causes of hearing loss are diagnosed by otologic, audiologic, physical examination, family history, radiological assessment (ex: CT and MRI of temporal bone) and molecular genetic testing.

Sensory neural hearing loss is a type of hearing loss or deafness where aetiology lies in inner ear either sensory organ (cochlea) or neural type. Sensory neural hearing loss may be mild, moderate, severe, profound or total. Sensory hearing loss occurs as a consequence of damaged or deficient cochlear hair cell function. Neural or retrocochlear hearing loss occurs because of damage to the cochlear nerve.

Congenital and acquired hearing impairment in children may not be noticed until speech and language fails to develop. The average age for the detection of congenital deafness was found to be around 2-3 years according to International standards. The age of detection is gradually improving. Neonatal screening tests, surveillances and hospital visit detection are some of the standard methods for detection of bilateral profound sensory neural hearing loss, especially in cases with prelingual deafness.

Cochlear implantation in cases with bilateral profound sensory neural hearing loss of prelingual deafness has become established over past 15 years or more.

Nominal rationale for cochlear implantation are-

- Failure to obtain adequate benefit from conventional acoustic amplification.
- Tonotopic organisation of cochlea.
- Variable spiral ganglion cells.
- The ability to gain surgical access to cochlea with suitable device.
- Availability of speech processing strategies, etc.

The study aims at the analysis of various radiological anomalies and its variations in bilateral profound sensory

neural hearing loss of prelingual deafness candidates selected for cochlear implant surgery.

Aim

A statistical analysis of radiological findings in prelingual deafness candidates selected for cochlear implant in our Institution during the period of one year and analysis of Michel's Aplasia using radiological variations.

Objective

Primary objectives are to study the incidence of radiological anomalies in prelingual deafness children evaluated for cochlear implant surgery to determine the commonest anomaly, to study the commonest anomaly and its variations among the study cases.

Investigations

Investigate them with Otoscopic findings, Pure tone audiometry, Behavioural observation audiometry, Impedance audiometry, Otoacoustic Emission (OAE) analysis and Brain stem Evoked Response Audiometry (BERA), Computed Tomography (CT) and Magnetic Resonance Imaging (MRI).

MATERIALS AND METHODS

- To identify children between the age group of 1-6 years with bilateral profound sensory neural hearing loss.
- Investigate them with Otoscopic findings, Pure tone audiometry, Behavioural observation audiometry, Impedance audiometry, Otoacoustic Emission (OAE) analysis and Brain stem Evoked Response Audiometry (BERA).
- After primary investigations they are subjected to hearing aid fitting trail for 6 months with subsequent auditory verbal habilitation.
- Subsequent failed hearing aid fitting and habilitation, they undergo complete radiological assessment for the feasibility of cochlear implant surgery including Computed Tomography (CT) and Magnetic Resonance Imaging (MRI).
- Such radiological findings are statistically analysed and interpreted in the study.

Inclusion Criteria

1. All children with congenital hearing loss with prelingual deafness are included in the study.
2. All children with bilateral profound sensory neural hearing loss between the age group of 1 to 6 are included in the study.
3. Both male and female children between the age group of 1 to 6 are included in the study.
4. All children with bilateral profound sensory neural hearing loss who failed hearing aid fitting and auditory verbal rehabilitation are included in the study.

Exclusion Criteria

1. All children below age 1 and above age 6 are not included in the study.
2. All children with post-lingual deafness are not included in the study.
3. All children with other severe mental disability are not included.
4. All children who respond well with hearing aid fitting and vocal rehabilitation are not included in the study.
5. All children with chronic infection of middle ear and mastoid cavity are not included.

RESULTS

Total number of cases included in our study are 20(Twenty). Among the 20 cases of bilateral profound sensory neural hearing loss of prelingual type selected for cochlear implant, 17 cases(85%) had radiologically normal inner and middle ear without any anomalies which is not discussed.

Three cases (15%) had radiologically identifiable anomaly. All three cases had Michel's type of Aplasia with some individual variations, which is analysed and discussed as case 1, case 2 and case 3.

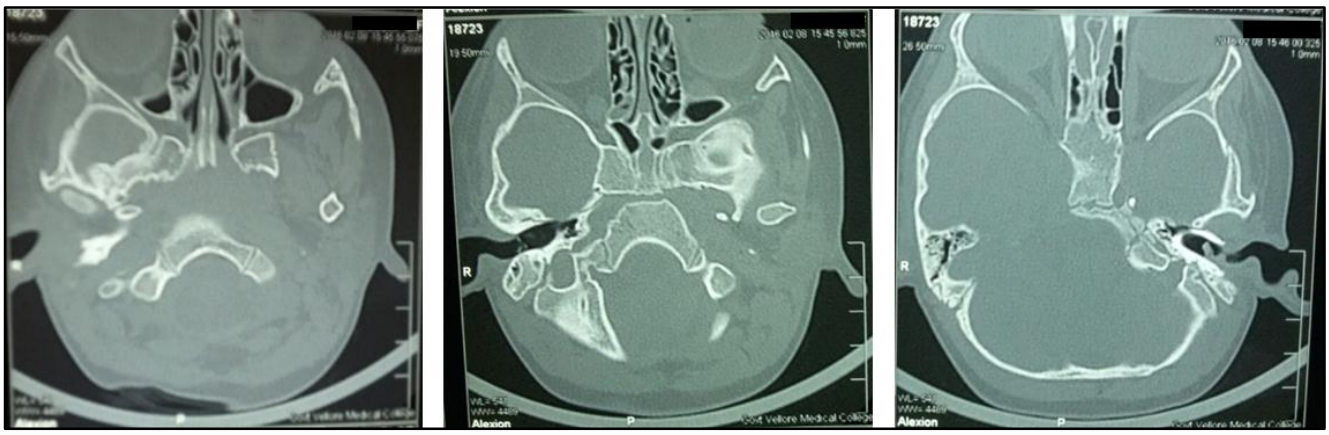
Most common inner ear anomalies are cochlear aplasia, cochlear hypoplasia, anomalies of vestibule and vestibular aqueduct, anomalies of Internal Acoustic Canal (IAC), incomplete partition type 1-lacking entire modiolus and cribriform area resulting in cystic appearance, incomplete partition type 2(Mondini's deformity) with cochlea consisting of 1.5 turns with middle and apical turns coalesce to form cystic apex with dilated vestibule and vestibular aqueduct, common cochlea deformity and Michel's deformity.

Michel's Aplasia is a complete failure of development of otic placode or Complete Labyrinthine Aplasia (CLA) with or without other anomalies such as-

- Hypoplasia of petrous part of temporal bone.
- Platybasiaof the skull base.
- Abnormal course of facial nerve and absent 2nd part of geniculate ganglion.
- Persistent stapedial Artery (or) Aberrant Internal carotid artery.
- Non-differentiation of stapes with absent round and oval window with normal incus and malleus.
- Aberrant course of jugular vein.
- Absent or narrow (stenosed) internal acoustic meatus.
- Absent cochleovestibular nerve.
- Flat medial wall of middle ear cavity.
- Due to failure of development of otic placode and other ectodermal constituents at 3rd week of generation.
- In our study, three such cases of Michel's Aplasia were identified and its variations analysed.
- Out of three cases of Michel's Aplasia,

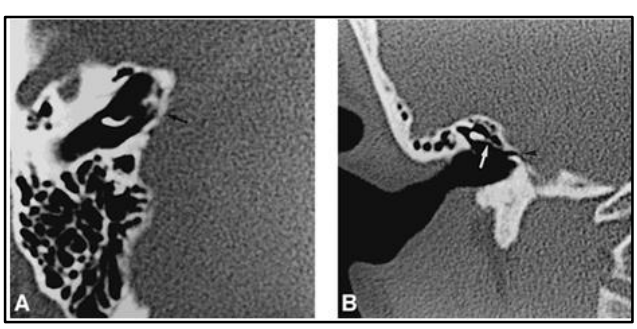
Case 1– A 3yrs. old female child for whom both parents had bilateral profound sensory neural hearing loss, the radiological findings are-

- Complete absence of inner ear structures– both cochlea and semi-circular canals.
- Hypoplastic internal acoustic meatus and internal auditory canal.
- Hypoplastic petrous apex.
- Normal skull base and Jugular vein.
- Presence of both cochleovestibular and facial nerve at CP angle cistern.



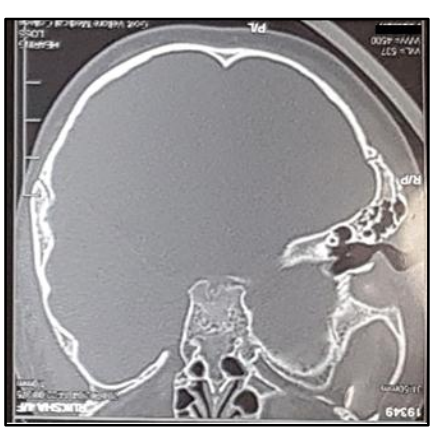
Case 2– A 1-½yrs. old female child for whom both parents had normal hearing, the radiological findings are-

- Complete absence of inner ear structures– both cochlea and semicircular canals.
- Non-differentiation of stapes with absent round and oval window.
- Complete stenosis of internal acoustic meatus and internal auditory canal.
- Absence of cochleovestibular nerve- Only single nerve is visualised in both CP angle cistern– likely to be facial nerve.
- Normal petrous apex.
- Normal skull base and Jugular vein.



Case 3- A 3yrs. old female child for whom both parents had normal hearing, the radiological findings are-

- Complete absence of inner ear structures– both cochlea and semicircular canals.
- Complete stenosis of internal acoustic meatus and internal auditory canal.
- Non-differentiation of stapes with absent round and oval window.
- Absence of cochleovestibular nerve- Only single nerve is visualised in both CP angle cistern– likely to be facial nerve.
- Hypoplastic petrous apex.
- Normal skull base and Jugular vein.



	Absence of Cochlea and Semicircular Canals	Non-Differentiation of Stapes	Absence of Cochleo-Vestibular Nerve	Hypoplastic Petrous Apex	Stenosis of Internal Acoustic Meatus	Platybasia of Skull Base	Intracranial Anomalies
Case– 1	+	-	-	+	-	-	-
Case– II	+	+	+	-	+	-	-
Case– III	+	+	+	+	+	-	-

Table 1. Cases of Michel's Aplasia with other Variations

DISCUSSION

In the past few years the incidence of acquired sensory neural hearing loss in children, especially in developing countries has reduced as a result of advanced neonatal care, widespread implementation of immunisation, awareness and early intervention. But congenital bilateral profound sensory neural hearing loss undergoes only slow downslope probably due to genetic contribution, consanguineous marriage, intrauterine infections and other unknown sporadic causes.

Prevalence of consanguinity is high in many areas, especially in this region with both genetic and acquired forms of sensory neural hearing loss are more common, particularly in children who live in poverty.

Adequate genetic counselling and health education was one of the reason for decreased incidence and decreased prevalence rates.

Neonatal screening programmes enable early detections and early intervention, which will prevent delay in speech and language development leading to better emotional, social living and improved quality of life.

Congenital infections, particularly cytomegalovirus infection is one of the commonest reason for profound sensory neural hearing loss.

Institutions with all the adequate infrastructure, counselling, surgical and non-surgical availabilities, neonatal screening programmes and habilitation with both hearing aids and implantable hearing devices aid in the reduction of children with bilateral profound sensory neural hearing loss with respect to morbidity better survival and reduced social burden to the society.

In a study by Derak D. Mafong et al, radiological abnormalities of inner ear malformations are common. Identification of inner ear malformations has direct impact on management of these children, suggesting that all children should undergo radiological imaging as integral component of evaluation of SNHL.[6]

In a study by L. Sennaroglu et al- on new classifications of cochleovestibular malformations-incomplete partition type (IP 1) is the commonest type. The type 1 malformation is less differentiated than type 2(Mondini), so the amount of dysplasia is much less than type 2.[7]

In a study by Kathlyn Marsor et al,Michel's Aplasia is a rare congenital inner ear anomaly defined by the absence of inner ear structures. Associated skull base anomalies should be identified, as they lead to potential complications, especially if surgical procedure is considered. Finally, the occurrence of bilateral inner ear aplasia in siblings suggest a genetic origin due to new mutations inherited as autosomal dominant type.[8]

Improved understanding of pathophysiology and molecular mechanisms of underlying hearing loss, recent advances and genetic testing will promote the development of the new treatment and screening strategies.

The commonest anomaly in our study group of 20 during the period of one year was Michel's Anomaly, which is complete failure of development of inner ear structure due to failure of development of otic placode during the 3rd week of intrauterine life leading to complete labyrinthine aplasia.

Michel's Aplasia may be different from Michel's hypoplasia or dysplasia where the components and sequelae vary.

The commonest anomaly associated with Michel's Aplasia are non-differentiation of the stapes, especially the foot plate with absent round window and oval window while the malleus and incus will be usually present.

There may be skull base anomalies, of which platybasia may be the commonest anomaly which may be well documented with enhanced CT.

Among the middle ear anomaly, persistent stapedial artery and aberrant internal carotid artery are the commonest anomalies which carries important surgical significance for the surgeons concerned at the time of surgery.

The next important anomaly may be the stenosed or hypoplastic internal acoustic meatus, which may be associated with presence or absence of cochleovestibular nerve or an abnormal course of facial nerve with absent geniculate ganglion or 2nd part of facial nerve or abnormal course of facial nerve directly entering into the mastoid again indicating the surgical significance for the surgeons operating on such cases.

The next important anomaly is the aberrant course of the jugular veins with or without presence of jugular bulb.

There may be flat medial wall of middle ear cavity with absent stapes, round window or oval window.

For neurosurgeons, abnormal positioning of the pons and posterior cerebellum may provide surgical difficulties.

In our study all the cases had normal intracranial structures, normal jugular veins and skull base.

Of the three cases, all the cases presented with complete absence of cochlea and semi-circular canals, hypoplastic or stenosed internal acoustic meatus and normal or hypoplastic petrous apex.

All the three cases of Michel's Aplasia were referred to Speciality Institution for Auditory Brain Stem Implant surgery and followed up.

CONCLUSION

Radiological analysis and exact interpretation of inner ear anomalies in candidates with bilateral profound sensory neural hearing loss becomes utmost important before selecting a candidate for a definitive surgery, i.e. cochlear implants.

- Regional variation of inner ear anomalies were under study among various developed and developing countries.
- The predominant inner ear anomaly in our study in our Institution for the period of 1 year is Michel's Aplasia or complete Labyrinthine Aplasia with some minor variations, which is documented.
- The incidence of the above-mentioned anomaly among with bilateral profound sensory neural hearing loss is 15% of the total cases studied.
- Such cases were referred for Auditory Brain Stem implants rather than cochlear implants.

- The radiological interpretation of such inner ear Anomaly carries regional as well as statistical significance.

REFERENCES

[1] Ludman H, Wright T. Childhood deafness. Mausen's disease of ear. 6thedn. London: Georgina Bentloff 2002:164-168.

[2] Gleason M, Browning GG, Burton MJ, et al. Cochlear implants. Scott-Brown's otorhinolaryngology head and neck surgery. 7thedn. Vol. 3. Great Britain: Edward Arnold2008:3650-3652.

[3] Biswas A. Imaging in neuro otology. Chapter 5. Anirban Biswas's an introduction to neuro otology. 2nd edn. Mumbai: Phalani2006:90-93.

[4] Hussain Z, Nair SK. Hearing aids. Chapter 59. Otorhinolaryngology. 2ndedn. Hyderabad/Delhi: Kothari 2008:662-663.

[5] Bess FH, Humes LH. Amplification and rehabilitation for the hearing impaired. Chapter 7. Audiology the fundamentals. 3rdedn. USA: Lippincott 2003:247-249.

[6] Mafong DD, Shin EJ, Lalwani AK, et al. Use of laboratory evaluation and radiologic imaging in the diagnostic evaluation of children with sensory neural hearing loss. The Laryngoscope 2002;112(1):1-7.

[7] Sennaroglu L, Saatchi's I. A new classification of cochleovestibular malformation. The Laryngoscope 2002;112(12):2230-2241.

[8] Marsot-Dupuch K, Dominguez-Brito A, Ghasli K, et al. CT and MRI findings of Michel anomaly- inner ear aplasia. American Journal of Neuro Radiology 1999;20(2):281-284.