ORIGINAL ARTICLE

Frequency of Inner Ear Anomalies Among Cochlear Implant Candidates: A Case Study in the Northwest of Iran

Reza Javad-Rashid[1], Akbar Pirzadeh[2], Masoud Naderpour[3], Roya Mirhaji[4]

[1] Department of Radiology, Faculty of Medicine, Tabriz University of Medical Science, Tabriz, Iran.
[2] Department of Otorhinolaryngology, Faculty of Medicine, Ardabil University of Medical Science, Ardabil, Iran.
[3] Department of Otorhinolaryngology, Faculty of Medicine, Tabriz University of Medical Science, Tabriz, Iran.
[4] Faculty of Medicine, Tabriz University of Medical Science, Tabriz, Iran.

ABSTRACT

Introduction: Globally hearing impairment occurs in about 1 to 2 per 1000 live births. The etiology of severe sensorineural hearing loos (SNHL) is complex and multifactorial. Congenital inner ear abnormality is a major cause of hearing loss in children and cochlear implantation (CI) is a proper treatment in these children. Preoperative CT scan of the temporal bones are used to evaluate inner ear malformations in CI candidates. The frequency of internal ear malformations reported on temporal scan could varied from 20% to 30%. The aim of this study was to evaluate the frequency of inner ear malformations in CT scan of cochlear implant candidates . **Methods:** In a retrospective cross-sectional descriptive study, 201 infants (105 boys and 96 girls) with the mean age of 20.5 ± 14.01 month with severe SNHL who are CI candidates were examined by temporal bone imaging with multi-slice CT from March 2014 to March 2015 in CI center of Tabriz University. **Results :** CT revealed abnormalities of the inner ear in 26 (13%) of infants that most of them with 10 (38.5%) was enlarged vestibular aqueduct (EVA). Also, 31 infants (15.4%) had abnormalities in outer, middle ear or in mastoid aeration. **Conclusion:** Temporal bone imaging with CT is an essential method and could be suggested as a proper first step for evaluating inner ear malformations in CI candidates.

Keywords: Congenital inner ear malformation , Sensorineural hearing loss , Cochlear implantation , CT scan

Corresponding Author:
Akbar Pirzadeh, MD
Email: a.pirzadeh@arums.ac.ir
Tel: +984533534741

INTRODUCTION

The inner ear is a key member of the sense of hearing and balance (1 page.324). The inner ear is made up of a bony labyrinth which containing a membrane labyrinth . Bone labyrinth lasts up to week 23 and overall inner ear development up to week 26. Congenital inner ear malformations occur as a result of stopping or deviating from the growth path of the inner ear due to inheritance issues , gene mutations and other factors at each of these stages (2 page.2). ''Congenital inner ear malformation are a major cause of hearing loss in children . The etiology of congenital hearing disorders is divided into two major causes : 50% environmental and 50% genetic factors'' (3 page.859). ''Overall the prevalence of hearing disorders is two per thousand live births" (4 page.118). SNHL accounts for about 90% of reported hearing loss. SNHL is usually permanent and can be mild, moderate, severe, profound, or total (5). Early detection and treatment of SNHL in children is vital and critical , because it is well known that delays in identifying hearing disorders have adverse effects on language and speech development , academic education and the social and emotional growth of the child (6 page.1). The American Institute of Child Hearing Screening has reduced the age of hearing impairment from 24 to 30 months. ''CI is one of the greatest success of biomedical engineering and it allows many people with complete deafness to achieve very high postoperative hearing performance'' (7 page.323). For patients scheduled for CI, the surgeon in addition to identifies congenital anomalies (concurrent middle ear malformations, cochlear malformations, cochlear bone pattern and vestibular malformations) and anatomical information of variations (such as degree of mastoid and other internal ear canal and its form) needs a CT. This information helps surgeons to assess cochlear implantation and determine the place of implantation and predict complications. High-resolution computed tomography (HRCT) should be used to obtain this information (8 page.ws1).

Hearing loss is divided into three major categories of conductive hearing loss, SNHL and mix of them. Central

auditory pathway disorders including disruption of the structures of the inner ear either in the auditory nerve pathway or in the central auditory pathway in the brainstem and thalamus cause SNHL. About 17 million Americans suffer from some degree of SNHL. The causes of SNHL are divided into two types primary (congenital) including: Bone labyrinth malformations (such as Michel and Mondini malformations) , membrane labyrinth malformations (complete or partial) , congenital Infections (TORCH), anoxy, birth trauma, exposure to auto toxic substances (streptomycin , qinine and thalidomide), Erythroblastosis Fetalis, Hyperbilirubinemia, cretinism, prematurity and all kinds of genetic syndromes and delayed inclusing: Neoplasms (Schwannoma, meningioma, etc), metastases, vasodilatation and endolymphatic sac, meningitis, Infectious labyrinthitis, trauma (temporal bone fracture), poisoning with autotoxic substances such as aminoglycosides, Munir's disease, vascular events and eventually autoimmune diseases (5). The aim of the present study was to evaluate the frequency of inner ear anomalies among CI candidates.

MATERIALS AND METHODS

Study design and participants

This retrospective cross-sectional descriptive study was performed on 201 children with severe SNHL who candidate for CI in Tabriz city hospital. The children firstly examined by an ENT specialist and then referred to the cochlear implant center. Information including parents' age, child's age, family history of hearing loss, history of previous childhood illnesses, history of trauma, birth weight, history of drug use, growth and speech status and age at which the first hearing diagnostic test was completed by interview. Then in all candidates the hearing assessment performed by underwent Auditory Brainstem Response (ABR) and SNHL severity were measured for them. At firstly time, hearing aids were used to enhance a child's hearing and functional abilities and these children were followed up frequently. Children with severe bilateral SNHL became candidates for CI after the medical commission was established and approved. All of children underwent temporal CT before CI surgery. The scan was performed with the Multi-Slice CT Scanner 64 (SOMATOM SENSATION SIMENS 64). The slices thickness was 0.6 mm . Images were recorded in axial , coronal and sagittal slices. Then the original images were reconstructed using MPR (Multiplanar Reformation) and if necessary by VRT (Volume-Rendering Technique). Finally the scans were carefully examined by two radiologists for normality of the atria and cochlea and their ducts, inner ear canal, presence of structural abnormalities and its types as well as associated findings. The result of the CT report was brought to the northwest CI center in Tabriz by parents.

Ethical approval

Written consent was obtained from the patients themselves and their parents. The results of this study was approved by Tabriz University of Medical Science ethical committee (IR.TBZMED.REC.1393.170).

Statistical analysis

Data were analyzed using descriptive and analytical statistical methods in SPSS version 16. Chi square test was used to determine relation between demographic variables and CT results. The P-value less than 0.05 was considered significant.

RESULTS

The mean age of patients was 20.24±14 month in the rage of one month to 6 years. Of all children, 57% were between the ages of 6 to 24 months. The average age of children with known abnormalities in CT was 23.5±15.9 month (3-72 months). Among the 26 children with reported abnormalities on CT, 14 (53.8%) were the first child of the family, 9 (34.6%) were the second and 3 (11.5%) were the third child of the family.

Of the 201 children studied, 7 (3.5%) weighed less than 1.5 kg, 189 (94%) between 1.5 and 4.5 and 5 (2.5%) more than 4 kg but all children with abnormalities reported on their CT scan results had weigh between 1.5 and 4.5 kg . Of all 201 children with SNHL, 105 (52.2%) were boy and 96 (47.8%) were girl. But among children with reported abnormalities of the inner ear on CT, 65.4% were girl.

Of all children with SNHL, the parents of 71 (35.3%) infants were non families. The parents of 103 children (51.2%) were classified as first degree families and the parents of 27 children (13.4%) were in second degree families group. Among the 26 children with abnormalities, 50% have parents with first degree family and 23.1% with second degree family.

Of the 201 children with hearing loss, 88 (43.8%) had a history of previous illnesses including: ikter, meningitis, seizures, hypoxia at birth, immaturity, trauma and antibiotic use by the mother. In 26 children with malformations, 77% had a family history of hearing loss. Among 201 children, the first audiometric test was performed in 21 children (10.4%) under one month and 106 children (52.7%) between one month and one year of age. In 74 children (36.8%), the first hearing test was performed at the first one year of life. Of the 26 children with abnormalities, only one child had a hearing test at the birth time.

Of the 201 CT studied, 144 infants (71.6%) had normal result and 57 (28.4%) abnormal results. Of 57 abnormal scans, 26 (45.6%) had internal ear malformations

with no accompanying middle and external ear disorders and 31 (45.6%) had external and middle ear disorders in associated with other disorders such as abnormalities in mastoid cells. Among 26 children with inner ear abnormalities on CT, 10 infants (38.5%) had atrial dilatation and 7 infants (26.9%) had cochlear dilatation (Figure 1).

In the age group below 12 months 8 cases (30.8%) had internal ear anomalies of which 5 cases (62.5%) had atrial duct dilatation. There was no significant relationship between age and the frequency of inner ear disorders among children (Figure 2).

Of all malformations, 34.6% were reported in males and 65.4% in female. The most common malformations among boys was atria and cochlear ducts each with three cases (11.5%). Also among girls, most of the children had atrial duct dilatation in seven cases (26.9%). There was no significant relation between gender and frequency of abnormalities. There was no significant relation between familial history and frequency of inner ear malformations. Of the 201 children, 88 (43.8%) had a previous history of the disease which of them, 8 cases (9.1%) reported abnormalities that of them, 3 (37.5%) had atrial dilatation and 3 (37.5%) had both atrial and cochlear dilatation. There was no significant relation between the history of the disease and the frequency of abnormalities . Of the studied children, 6 (23.1%) had a positive history of the disease in their family. There was no significant relation between the positive history of the disease in the family and the frequency of the abnormalities (Table I). In our study due to Michel deformity indications in cochlear implant surgery, one patient was excluded from the list of implant candidates.

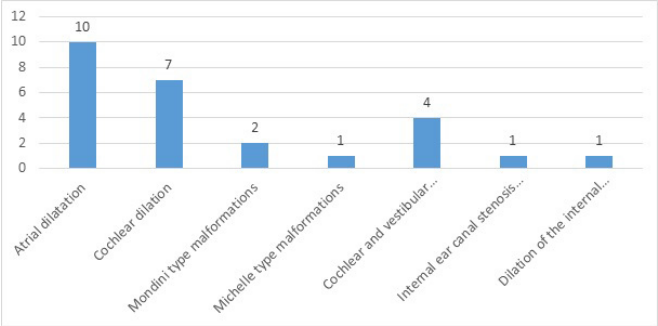

Figure 1: Types of inner ear malformations in CT results of patients

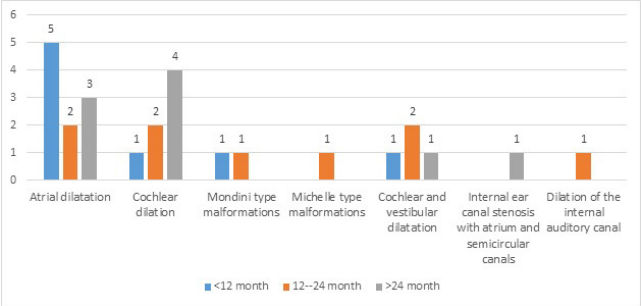

Figure 2: Types of inner ear malformations in CT results of patients by age groups

DISCUSSION

In the study of Nikolopoulos et al, meningitis was the etiology of hearing loss in 40.8% of cases and congenital in 47.2% of cases (9). In the study of Bamiou et al, 44% of deafness cases were due to genetic problems (10). In the Kadom et al study, 20-40% of patients with SNHL had a history of TORCH meningitis or trauma (8). In the Nair et al study, meningitis was the cause of cochlear bone in 44% of cases (11). In our study, the history of diseases such as ikter, meningitis, seizure, trauma, low birth weight, immaturity, anorexia and history of aminoglycoside was investigated and 43% of the children had a positive history of these diseases which was in line with the above studies. But their deafness cannot be attributed to these diseases and it should be compared with the healthy children in a large statistical group. Also, no significant relationship was found between the reported abnormalities in the scan and the history of previous child diseases .

Table I: Relation between results of CT and demographic data

Results of CT Variables n		Atrial duct dilatation		Cochler duct dilatation		Mondini type anomaly		Simultaneous opening of the cochlear and atrial ducts		others		p-value
		%	n	%	n	%	n	%	n	%		
Sex	Female	7	26.9	4	15.4	1	3.8	3	11.5	2	7.7	0.53
	Male	3	11.5	3	11.5	2	7.7	1	3.8	-	-	
History of disease	+	3	11.5	-	-	-	-	3	11.5	-	-	0.16
	-	7	26.9	6	23	-	-	-	-	-	-	
Age (month)	12-24	2	7.7	2	7.7	1	3.8	2	7.7	1	3.8	0.27
	>24	3	11.5	4	15.4	-	-	1	3.8	1	3.8	
Relative of parents	Degree 1	7	26.9	4	15.4	-	-	1	3.8	1	3.8	0.25
	Degree 2	4	15.4	3	11.5	-	-	1	3.8	1	3.8	
	none	3	11.5	3	11.5	-	-	1	3.8	-	-	

In the study of Bamiou et al, the average age at diagnosis time was 3.2 year (10). In the Park et al study, the average age at diagnosis time was 25.7 months (12). In this study, the average age at diagnosis time was 8 months and in 52.7% of children in Tabriz CI center, the first diagnostic test for hearing loss was performed in one month to one year old .

In the Bamiou et al study, 6.4% of parents of children with SNHL were related. 59.8% of children with SNHL were male with a positive family history in 29.3%. In this study gender of the parents and positive family history were not correlated with CT results (10). In the Ahmad et al study, 64% of children were boy and had SNHL (13). In the Park et al study, 12.8% of children with SNHL had a positive family history (12). In the present study, 52.2% of the children with SNHL were boy and 47.8% were girl and consistent with the above studies. The rate of SNHL in the male is slightly higher than female. There was no significant relation between gender and abnormalities found in CT.

In the present study, 64.6% of parents were first and second degree relative, it is much more than above statistics and this could be due to the high prevalence of familial marriage in Iran or the high prevalence of SNHL in children with related parents . An extensive comparative study is needed to distinguish the results. In our study, there was no significant relation between scan results and familial history and consistent with the above studies . In our study, 66.2% of children had no positive family history of hearing loss and no meaningful relationship was found between the results of the scan and the family history of hearing loss which is consistent with the above studies.

In a study by Kadom et al, they concluded that in patients undergoing CI, Surgeons in addition to identify congenital ear malformations (such as middle ear malformations, type of cochlear malformations and atrial duct malformations) also need to a lot of CT scan information about the degree of mastoid cell as well as the size and shape of the inner ear canal (8). In another study by Masoud et al, they concluded that previously unrecognized abnormalities such as the large atrioventricular duct, the narrow or inner ear canal as well as the cochlear nerve canal stenosis have been identified by new CT techniques and so this technique is of great help to the genetic counseling physician (14).

In a study by Huo et al, they concluded that CT are preferred in cases when the decision is to exclude middle and external ear malformations. MRI is preferred in examining delicate changes in the membrane labyrinth or neuromuscular abnormalities in the inner ear canal (9). In the Nair et al study, the accuracy of HRCT in detecting bone cochlea was 94.6% (with 100% specificity and 71% sensitivity). This study showed that, HRCT had high accuracy in detecting cochlear malformations (15).

In the study of Bamiou et al on patients with bilateral SNHL, they concluded that the percentage of reported abnormalities in the scan varied from 6.8% to 12.8% and may be abnormal even up to 30% in patients candidate implantation. In this study, 28% of people with SNHL had internal ear malformations (10).

In the Shim et al study, 20%-30% of people with SNHL had internal ear malformations (12). In the study of Ahmad et al in 2016, 12.8% of people with SNHL had internal ear malformations . The study also revealed abnormalities such as calcification of the inner ear and chronic purulent otitis in addition to the inner ear malformations (13). In the Furmanek et al study, 55% of people with SNHL had internal ear malformations (16). In the study of Huo et al, 30% of people with SHNL had internal ear malformations (15). In the study of Hui et al, 43% of people with SNHL had internal ear malformations (17). In our study, 57 out of 201 children with SNHL had abnormal scans. Of them, 26 were internal ear malformations without accompanying middle and outer ear disorders and 31 were external and middle ear disorders as well as other disorders such as mastoid cell. In our study 13% of children with SNHL had internal ear malformations which is consistent with the above studies.

In a study conducted by Bamiou et al, 28 patients with cochlear implantation with malformations recognized by temporal HRCT were retrospectively studied. They concluded that different types of inner ear malformations after CI have a completely different prognosis for excellent hearing improvement and preoperative HRCT is a necessary action (10). In the study of Shim et al, the most common malformation of the inner ear was atrial ventricular dilatation (18). In the study of Ma H et al, the most common abnormality reported was vestibular dilatation with 44% (17). In the study of Kadom et al, the most common reported abnormality was atrial duct dilation (8). In the study of Huo et al, atrial malformations accounted for 30% of malformations (15). In the study by Nair et al, atrial dilatation appears in 18% of cases as single and in 36% of cases associated with other types of abnormalities. Overall the most common finding was atrial malformations. In this study the atrial duct was clearly associated with progressive SHNL (11). In the study of Park et al, atrial dilatation was seen in 58% of cases and in fact was the most common radiographic findings in these patients. In this study, atrial cochlear and atrial dilatation were the most reported abnormalities in patients with SNHL (12).

In the study of Ahmad et al in 2016 on 72 patients

with SNHL, results showed that 63% of the reported abnormalities were vestibular malformations with 9% and cochlear malformations with 18% (13). Finally in the study of Wang et al, 55% of the reported abnormalities were in the form of Mondini type and 28% in the form of cochlear dilatation (19).

In the present study among 26 children with internal ear malformations, CT results showed that 10 cases (38.5%) had atrial duct dilatation, 7 cases (26.9%) had loose cochlea, 2 cases (7.7%) had Mondini type malformations and other seven cases had another type of malformations. Finally in our study the most common abnormality reported in the scan of CI patients candidates was atrial duct dilatation which is consistent with the above studies. In this study for performing hearing assessment we used the Auditory Brainstem Response (ABR) and for detection of abnormalities in the bony labyrinth (OTIC Capsule) we only used the CT-scan without MRI results.

CONCLUSION

There was no significant relationship between the results of the scan with child gender, positive family history, history of child disease, familial disease and birth weight. In this study we concluded that the identification interval for children with SNHL in northwestern of Iran was between one month and one year of age and this is a good time compared to global statistics. Another result of the present study was that CT is the first and most necessary step in the evaluation of patients with SNHL (to identify the frequency of various types of inner ear malformations, to identify concurrent middle and outer ear malformations and to guide the surgeon during different stages of CI). Finally the identification of atrial dilatation as the most common internal ear malformations in CT of patients candidates for CI was the most important result of the present study. It is recommended that all infants be screened for SNHL at birth. Extensive genetic studies and doing temporal CT scan should also be performed on all children with SNHL.

ACKNOWLEDGEMENT

The authors wish to thank all infants for participated in the study.

REFERENCES

1. Eric G. Ekdale . Comparative Anatomy of the Bony Labyrinth (Inner Ear) of Placental Mammals. PLoS One. 2013; 8(6): e66624.
2. Varsha M, Shantanu K, Ravi G, Gitender K,Vinay E . CT and MR imaging of The Inner Ear and Brain in Children with Congenital Sensory Neural Hearing Loss .The journal of continuing medical education in radiology. 2012; 32: 683-698
3. Yiin R, TANG P, TAN T. Review of congenital inner ear abnormalities on CT temporal bone . The British Journal Of Radiology. 2011; 84: 859-863
4. KS Tse, KM Chu, LF Chiu, TW Fan. Congenital Inner Ear Malformations. Hong Kong J Radiol. 2011;14:118-25
5. Sensorineural hearing loss. Available form: https://en.wikipedia.org/wiki/Sensorineural_hearing_loss.
6. Huang B.Y, Danski C.Z, Castillo M. Pediatric sensorineural hearing loss, part 2: syndromic and acquired causes. AJNR Am J Neuroradiol. 2012;33(3):399-406.
7. Bruce RW, Timothy AH, Barry SB, Charles CF, Margaret WS . Use of Computed Tomography Scans for Cochlear Implants . J Digit Imaging. 2008; 21(3): 323–328.
8. Kadom N, Sze RW. Radiological Reasoning: Congenital Sensorineural Hearing Loss . AJR Am J Roentgenol. 2010;194:WS1-4.
9. Nikolopoulos TP, O'Donoghue GM , Robinson KL, Holland IM, Ludman C, Gibbin KP. Preoperative radiologic evaluation in cochlear implantation. American journal ofotology . 1997; 18(6):73-74.
10. Bamiou DE, Phelps P, Sirimanna T. Temporal bone computed tomography findings in bilateral sensorineural hearing loss .BMJ.2011; 82:257-260.
11. Nair SB, Abou–Elhamd KA , Hawthorne M. A retrospective analysis of high resolution computed tomography in the assessment of cochlear implant patients . Clin Otolaryngol allied Sci. 2000; 25 (1), 55- 61.
12. Park AH, Kou B, Hotaling A, Azar-Kia B, Leonetti J, Papsin B. Clinical course of pediatric congenital inner ear malformations .Laryngoscope. 2000; 110:1715-1719.
13. Ahmad M. Aldhafeeri, Abdulrahman A. Alsanosi. Prevalence of inner ear anomalies among cochlear implant candidates. Saudi Med J. 2016; 37(10): 1096–1100.
14. Masoud S , Usui S , Matsunaja T. High prevalence of inner ear and/ or internal auditory canal malformations in children with unilateral sensorineural hearing loss . International Journal of Pediatric Otorhinolarynology .2013;77:228-232.
15. Huo L , Wang H. Characteristics and application of inner ear CT in 20 cases of sensorneural hearing loss in children . Acta Otolaryngol.2012; 132(12):1261-5.
16. Furmanek MI, piotrowska A, Skarzynski H, Boguslawska-Walecka R, Walecki J. Value of multidetector computed tomography in detection of Congenital inner ear malformations and its usefulness in qualification for cochlear implantation. Otolaryngol Pol. 2007;61(4), 585 -590.

17. Ma H, Han P, Liang B, Tian ZL, Lei ZQ, Kong WJ, Feng GS. Multislice Spril Computed Tomography Imaging in Congenital Inner Ear Malformations . Journal Comput Assist Tomogr. 2008; 32(1): 146 – 150.
18. Shim HJ ,Shin JE , Chung JW , Lee KS . Inner Ear Anomalies in Cochlear Implants : Importance of Radiologic Measurements in the Classification .Otology & Neurology.2006;27: 831-837.
19. Wang J,Meng M , Huan Y , Zhang J. HRCT imaging characterized of cobgenital abnormalities of the inner ear in 45 cases. Department of Otolaryngology. Journal of NCBI.2003; 17(10):583-585.