Hearing loss etiology in patients referring to Isfahan cochlear implantation center

Seyed Hamidreza Abtahi[1], Mahsa Sepehrnejad[2], Shiva Dehghan[3], Mohammad Hossein Nilforoush[2*]

1. *Associate Professor, Department of Ear, Nose, and Throat, Faculty of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran.*
2. *Communication Disorders Research Center, Isfahan University of Medical Sciences, Isfahan, Iran.*
3. *Medical Student, Student Research Committee, Isfahan University of Medical Sciences, Isfahan, Iran.*

**Corresponding Author: Mohammad Hossein Nilforoush*
Communication Disorders Research Center, Isfahan University of Medical Sciences, Isfahan, Iran
Email: mhnilforoush@gmail.com

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Abstract

Introduction: Hearing loss is the most common congenital disorders occurring among newborn. Identifying the factors affecting it would reduce the incidence of this disorder. Therefore, the aim of this study was to examine the etiology of congenital hearing loss in patients referring to Isfahan Cochlear implantation center.

Materials and Method: This study was performed on 689 patients with cochlear implantation. Demographic data and relative frequency of different causes of congenital hearing loss (acquired and genetic) were determined and recorded. T-test, Chi-Square and Mann-Whitney tests were used to compare the variables studied.

Results: Our findings showed that 50 patients (7.7%) had history of drug use, 9 (1.3%) had history of taking ototoxic drugs, 99 (14.3%) had history of kernicterus, 157 patients (22.8%) had a history of hyperbilirubinemia, 15 (2.1%) history of meningitis, 57 (8.3%) had a history of seizure, one (0.1%) had a birth weight less than 1500 grams, 4 cases (0.06%) had history of hypoxia, 18 cases (2.6%) had history of trauma, one (0.1%) had CMV history, and one (0.1%) hearing loss due to syndromicity. On the other hand, it was determined that the probable causes of hearing loss were not related to the severity of hearing loss and age of the patients ($p < 0.05$)

Conclusion: Although the prevalence of congenital hearing loss is low, it is important to identify and screen for postnatal congenital hearing loss, especially in people with risk factors known in this study, including patients with kernicterus.

Keywords: Congenital hearing impairment, Cochlear implantation, kernicterus

Introduction:

Hearing loss is the most common congenital disorder developing among newborns (1-4). The incidence of congenital hearing loss is about 2-4 per 1000 births (2, 3). Also, one in every 1000 children suffers from hearing loss before the age of school (1). Neonatal screenings tests are widely used to recognize hearing impairment in infants that can provide early treatment interventions (5). Causes of bilateral sensory neural hearing loss can be due to genetic factors (syndromic, non syndromic), non-genetic factors (prenatal causes (measles/seizures, viral/alcohol/other), birth causes asphyxia, prematurity/other) and postnatal factors (meningitis /traumas/other) and unknown causes (6).

Genetic factors are among the main etiologies of severe to deep hearing loss and may play an important role in cochlear implantation (9-7). Up to now, genes that are related to forms of deafness have been cloned, but efforts to bring a particular genetic from with the results of cochlear implantation have failed (1). Researchers consider differences in spiral ganglion cells, hearing loss etiology/ and other factors as the causes of differences in the results of cochlear implantation (1).

Non syndromic genetic hearing loss includes an extremely heterogeneous group of disorders. So far 30 recessive genes and 39 dominant genes have been detected and some of these genes have been well identified (6).

Measles is now less known as hearing loss etiology, as this infection has decreased significantly because of the seriousness of antenatal care (5, 6).

Cytomegalovirus (CMV) is known as one of the causes of a changes in the white matter of the brain, causing a sensory-neural hearing loss by harming the cochlea; unfortunately the pathophysiology of this injury is not well known, though being natured of OAE indicates that a cochlea relater mechanism is involved in this process (10).

There is no clear limit for the amount of hypoxia which causes the risk of hearing impairment, and there are significant differences in the severity of hearing impairment among different individuals. in hypo poetic episodes (10).

Hypoxia has a powerful companion with hearing loss and sufficient oxygenation and perfusion is essential for the normal functioning of the cochlear the companion of birth weight of less than 1500 (ULBW) and hearing loss were detected long ago. Over the past two decades, babies' survival has incased due to many advances in childbirth care and new born care (10).

The most common bilateral etiology (SNHL) is the risk of unknown factors (6). In a study on patients with unknown SNHC were examined to identify the unidentified etiology involved in the onset of the disease. In one of these studies, 64 patients were subjected to serological tests (for toxoplasma, cytomegalovirus, measles, HSV1), CT-scan an ophthalmology, and in this study they identified an etiology related to the disease in 44% of cases. The remaining 56% also referred to non-syndromic hearing loss (in the population studied, the percentage of family marriage was high (more than 60%) (6).

Retrograde cohort studies in determining SNHL, had a bias in selection, for example patient who have been estimated to have hearing difficulties with mild hearing loss or mentally impaired patients, have been excluded from this study. Prospective studies and studies in populations are less proves to this error. Furthermore, population studies have an extra advantage, and that's a vast geographical area that causes children with different degrees of hearing loss and children with various types of defection to be involved in the study (6).

These kind of studies are better representative the total condition of the society (6). Currently, genetic tests and the pre-natal infections treatment have altered the statistics of hearing loss etiologies, and there is no study that examines these changes.

Material and Methods:

This study is an expository, prospective and cross sectional study. Patients in this study included all the patients with hearing impairments referring to the Isfahan hearing aid center. To do so, all the subjects with entry criteria and no outlet criteria were included in the study. The total number of 689 patients who referred to Isfahan cochlear implant center were included in the study and the sampling was done as a census.

The entry criteria included patients referred to Isfahan cochlear implant center and were included in cochlear implantation list. The

files should be exhaustive and in the case of occurring any defects in patient's file, the patient should be available through given numbers. The exclusion criteria were defects in patient's files, the case of absence in subsequent referrals in order to complete the file information or the patient not being available in order to complete the file information.

Following the necessary coordination with hospital management and medical records, the list of all the patients in the cochlear implantation was purveying to the executor of the study. Then by reviewing the files and inclusion/exclusion criteria, qualified patients entered the study and supplementary information was extracted from their records. After entering the study, demographic data like age, sex, disease records, hearing loss records in families are extracted and listed. By referring to the patient's file, other variables like hearing loss (genetic or acquired) were extracted and listed. It should be noted that during the study the patient's personal information were only available to the executor and the information were protected.

Data analysis:

All patient information includes demographic factors, and paraclinic symptoms were recorded in check lists and entered in SPSS 22 software. Data analysis was done in two descriptive and analytical parts.

In descriptive part, the frequency of hearing loss causes stays as the main variable. Demographic and clinical features are reported based on descriptive criteria. In analytical part appropriate parametric and nonparametric tests was employed. For analysis qualitative data, CHI- SQUARE test is used and for quantitative data comparison T-test was employed. In cases, which were not confirmed in our basic variables, Mann-Whitney non parametric test were used to compare the variables studied. All the exams were studied in $p<0.05$.

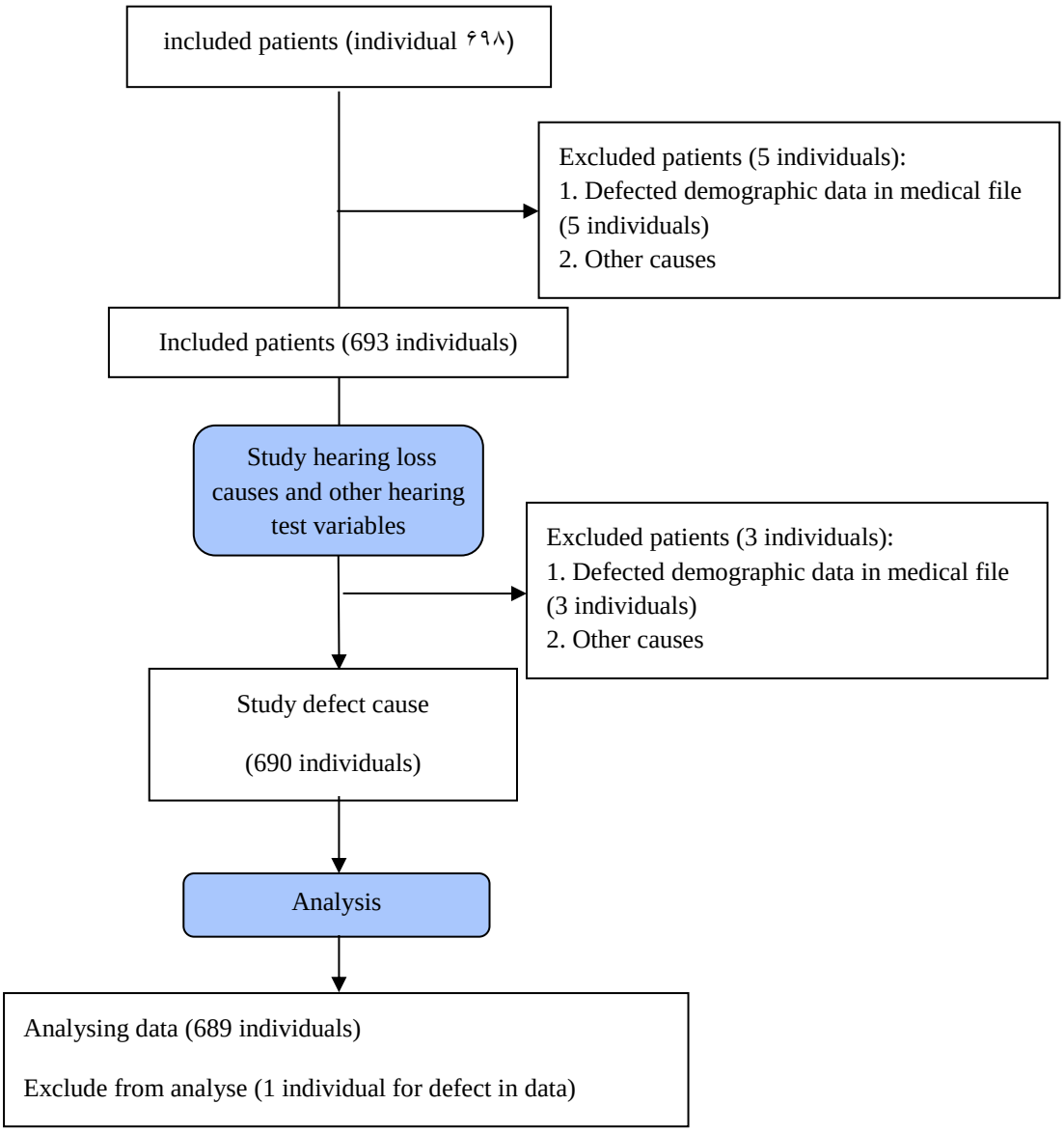

Figure 1-3: Flowchart

Results:

This study was performed on 689 patients who had a mean age of 69.38 ± 12.16 ± 36 months (1-840 months), and the mean age of the patients diagnosed with the disease was 95 ± 6.18 / 24 months (10 days to 693 months). Of the 689 patients enrolled in the study, 423 (61.4%) were the outcome of familial marriage. It was also found that 193 patients (28%) had a family history of hearing loss.

By assessing the amount of hearing loss, 291 (42.3%) had severe hearing impairment, 216 (31.4%) had severe to severe hearing loss, 59 (8.6%) had severe hearing loss, 101 People (14.7%) had moderate to severe hearing loss and 12 (1.7%) had moderate hearing loss.

The findings of the study showed that 50 (7.3%) patients had history of drug use and 9 (1.3%) had a history of taking ototoxic drugs. The study found that 99 (14.3%) patients had history of kernicterus, 157 (22.8%) had a history of hyperbilirubinemia, 15 (2.2%) had history of meningitis, 57 (8.3%) had a history of seizure, one person (0.1%) had a weight loss of less than 1500 grams at birth, 4 cases (0.6%) had a history of hypoxia, 18 (2.6%) had history of trauma, one (0.1%) had a history of CMV, one person (0.1%) had hearing loss due to syndromicity.

It was found that the causes of hearing loss were only one of the causes of syndromic causes, which were also due to familial marriage, but the sample size was not significant (p = 1). By examining the amount of hearing loss based on the underlying causes, there was no significant relationship between the severity of hearing loss and the causes of hearing loss (p> 0.05) (Table 1). By examining age and its relation to the underlying causes of hearing loss, it was found that there was no significant relationship between these two variables (p> 0.05).

Chart 1: possible hearing loss causes in studied patients

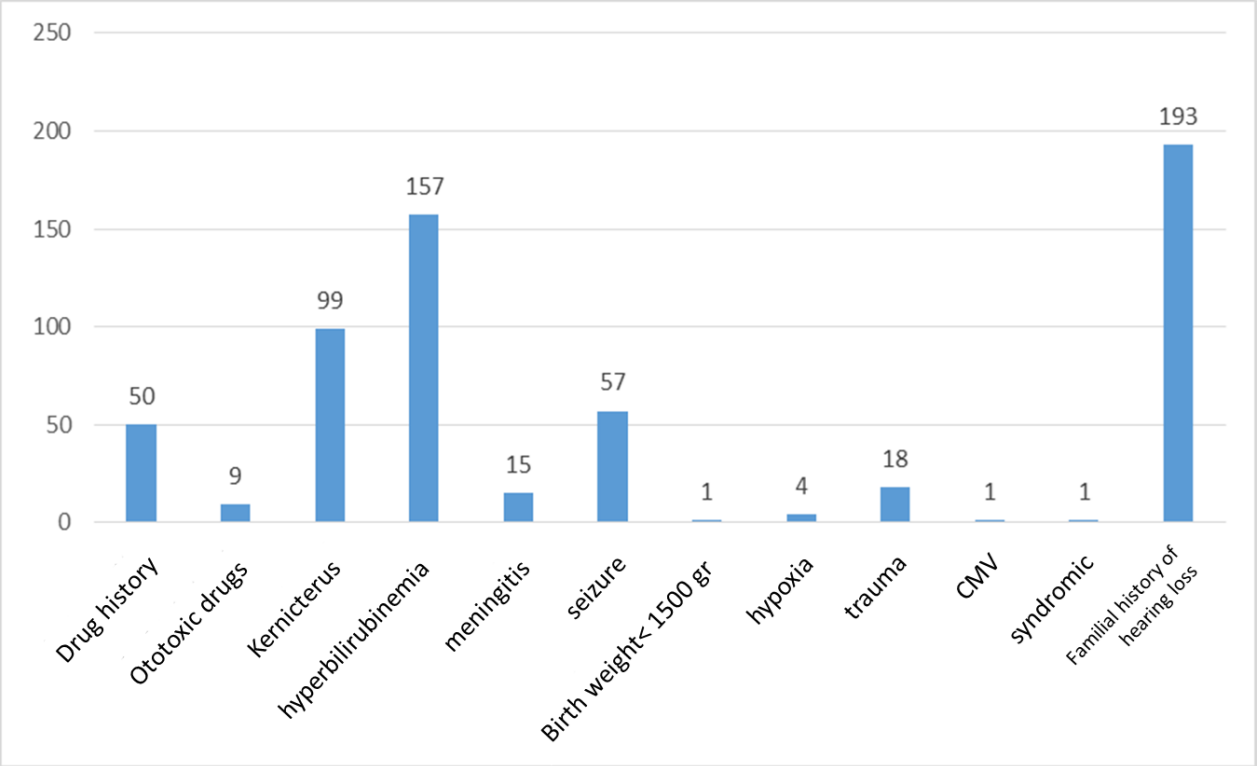

Table 1: Frequency of hearing loss intensity due to its basic cause

p-value	moderate	moderate to severe	severe	severe to profound	profound	Hearing loss intensity cause
0/065	1 (2.0%)	11 (22.0%)	2 (4.0%)	8 (16.0%)	28 (56.0%)	Drug history
0/594	0	3 (33.3%)	1 (11.1%)	1 (11.1%)	4 (44.4%)	ototoxic
0/843	2 (1.0%)	25 (13.1%)	15 (7.9%)	60 (31.4%)	87 (45.5%)	Family history
0/397	2 (2.0%)	21 (21.2%)	7 (7.1%)	25 (25.3%)	43 (43.4%)	kernicterus
0/546	3 (1.9%)	29 (18.6%)	15 (9.6%)	50 (32.1%)	57 (36.5%)	Hyperbili-rubinemia
0/282	0	1 (6.7%)	1 (6.7%)	2 (13.3%)	11 (73.3%)	meningitis
0/355	1 (1.8%)	4 (7.0%)	8 (14.0%)	18 (31.6%)	26 (45.6%)	seizure
1	0	0	0	1 (100.0%)	0	Birth weight <1500 gr
0/054	0	0	2 (50.0%)	2 (50.0%)	0	hypoxia
0/374	0	3 (16.7%)	0	5 (27.8%)	9 (50.0%)	trauma
1	0	0	0	1 (100.0%)	0	CMV
0/325	0	1 (100.0%)	0	0	0	syndromic

Discussion:

In our study, it was found that the most common causes of hearing loss are hyperbilirubinemia and kernicterus, and in the next steps, drug use, trauma and meningitis, and only one genetically recognized cause were found. In general, familial marriage was high in the parents of patients and more genetic causes are needed to study in patients, and it was found that the possible causes of hearing loss in both groups are not related to the severity of hearing loss and the age of the patients. In general, it is possible to find out that important causes of hearing loss are acquired causes and also the history of marital marriage is high in these patients, which needs further investigation. The most common bilateral SNHL etiology is an unknown risk factor (6). Two studies of patients with unknown SNHL were studied to identify an unidentified etiology involved in the onset of this disease. In one of these studies, 64 patients were subjected to serological tests (for toxoplasma, cytomegalovirus, measles, HSV1), CT scan, and ophthalmology, and in this study they identified an etiology related to the disease in 44% of cases. The remaining 56% also referred to non-syndromic hearing loss (in the population studied, the percentage of marriages was high (more than 60%). A total of 32 patients with unknown etiology were included in this study. Blood sampling, ECG, scan CT, and ophthalmic examinations were performed. Data of family history of genetic hearing loss were collected and cases were examined by Bekesy Auditory Test, and 34% had non syndromic genetic hearing loss (6).

The results of these studies are in line with the findings of our study. In our study, it was also found that the ratio of marital marriages was high in patients, and on the other hand, kernicterus and hyperbilirubinemia in these patients were higher than the normal population and were considered as the major causes of hearing loss. In a study done by

Zamani et al,. that evaluated the frequency of hearing impaired high risk neonates in neonatal wards of Tehran University of Medical Sciences hospitals, multivariate logistic regression analysis revealed that substitution, low birth weight and low Apgar score were provided the highest independently risk factors for infants. At the end of the study, despite the low prevalence of hearing loss, the screening was of great importance for early detection of cases.(12) Since the most common cause of blood transfusion is hyperbilirubinemia and kernicterus, the results of this study are in line with our findings. However, in our study, low birth weight was seen in only one case. Another study by Russ SA et al,. found that 54 (40%) of patients with congenital hearing loss had a mild hearing loss (20 to 40 dB HL). The prevalence of moderate and severe hearing loss (40 dB HL) was 1.12 / 1,000. The cause of hearing loss was known in 57 (43%) congenital cases, and the genetic causes of non-syndrome were 21 (37%). (13) Another study by Shahnaz Pourarian et al. In 2015 study found that among the 124 neonates involved in the study, 17 (13.7%) were short-term inferior to hearing loss. There was a significant relationship between gestational ages less than 36 weeks, antibiotic therapy, oxygen therapy and hearing loss. Conversely, there is no meaningful relationship between hearing loss and the use of ventilator, sepsis, hyperbilirubinemia, congenital heart disease, transient infantile tachycardia, congenital pneumonia, or respiratory distress syndrome. (14) The results of this study are not in line with the findings of our study, because in the recent study, kernicterus and hyperbilirubinemia were significantly higher in individuals with congenital hearing loss, and this difference may be due to differences in the sample size, the difference The type of study performed is the difference in the type of sampling and the difference in the type of data comparison with each other.

Conclusion:

In our study, it was found that the most common cause of hearing loss is hyperbilirubinemia and kernicterus, and in the next stages of seizure, drug use, trauma and meningitis, and only one genetically recognized cause, it was found that familial marriage was high in the parents of patients And studying of genetic causes is more needed in patients, and it was found that the possible causes of bilateral hearing loss is not related to the severity of hearing loss and the age of the patients. Although the incidence of congenital hearing loss is low, it is important to identify and screen for postnatal congenital hearing loss, especially in people at risk, including those with kernicterus. Also, looking at the personal characteristics and factors associated with the occurrence of congenital hearing loss identified in this study, it is necessary to reduce the incidence of congenital hearing loss due to the underlying condition of the patients.

Conflict of interests

Authors declare no conflict of interests.

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References:

1. Lai, R., et al., Genetic diagnosis and cochlear implantation for patients with nonsyndromic hearing loss and enlarged vestibular aqueduct. J Laryngol Otol, 2012. 126(4): p. 349-55.
2. Deklerck, A.N., et al., Etiological approach in patients with unidentified hearing loss. International Journal of Pediatric Otorhinolaryngology, 2015. 79(2): p. 216-222.
3. Wonkam, A., et al., Aetiology of childhood hearing loss in Cameroon (sub-Saharan Africa). European Journal of Medical Genetics, 2013. 56(1): p. 20-25.
4. Espeso, A., D. Owens, and G. Williams, The diagnosis of hearing loss in children: Common presentations and investigations. Current Paediatrics, 2006. 16(7): p. 484-488.
5. Lammens, F., et al., Aetiology of congenital hearing loss: A cohort review of 5 ٦٩subjects. International Journal of Pediatric Otorhinolaryngology, 2013. 77(9): p. 1385-1391.

6. Morzaria, S., B.D. Westerberg, and F.K. Kozak, Systematic review of the etiology of bilateral sensorineural hearing loss in children. International Journal of Pediatric Otorhinolaryngology, 2004. 68(9): p. 1193-1198.
7. Janeschik, S., et al., Influence of etiologic factors on speech perception of cochlear-implanted children. Cochlear Implants Int, 2013. 14(4): p. 190-9.
8. Varga, L., et al., Is deafness etiology important for prediction of functional outcomes in pediatric cochlear implantation? Acta Otolaryngol, 2014. 134(6): p. 571-8.
9. Rajput, K., T. Brown, and D.-E. Bamiou, Aetiology of hearing loss and other related factors versus language outcome after cochlear implantation in children. International Journal of Pediatric Otorhinolaryngology, 2003. 67(5): p. 497-504
10. Cristobal, R. and J.S. Oghalai, Hearing loss in children with very low birth weight: current review of epidemiology and pathophysiology. Arch Dis Child Fetal Neonatal Ed, 2008. 93(6): p. F462-8.
11. Haupt, H., F. Scheibe, and C. Ludwig , Changes in cochlear oxygenation, microcirculation and auditory function during prolonged general hypoxia. Eur Arch Otorhinolaryngol, 1993. 250(7): p. 396-400.
12. Zamani A, Karimi A, Naseri M, Amini E, Milani M, Sazgar A A, et al . Prevalence of hearing loss among high risk newborns hospitalized in hospitals affiliated to Tehran University of Medical Sciences. Tehran Univ Med J. 2010; 68 (1) :64-70
13. Russ SA1, Poulakis Z, Barker M, Wake M, Rickards F, Saunders K, Oberklaid F. Epidemiology of congenital hearing loss in Victoria, Australia. Int J Audiol. 2003 Oct;42(7):385-90.
14. Shahnaz Pourarian, Bijan Khademi, Narjes Pishva, Ali Jamali. Prevalence of Hearing Loss in Newborns Admitted to Neonatal Intensive Care Unit. Iran J Otorhinolaryngol. 2012 Summer; 24(68): 129–134.