# ETIOLOGICAL FACTORS OF CONGENITAL HEARING LOSS IN CHILDREN – A HIGH RISK REGISTER BASED STUDY

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**Abstract**

*The study aimed to investigate the possible etiological factors of congenital severe to profound sensorineural hearing loss in children. The study also analyzed any difference in etiology, as a factor of geographic locations, socio-economic status, religious-community and gender. A survey-based study was carried out at the Department of Audiology, AIISH. A high-risk register was administered on parents of 70 children with congenital severe to profound sensorineural hearing loss. It was found that 73% of the participants had at least one causative factor associated with their condition whereas the remaining 27% had idiopathic onset of hearing loss. Parental consanguinity (31%) was the most common cause followed by maternal infections (13%), history of high fever (7%), NICU care more than a week (6%), family history of congenital hearing loss (4%), neonatal hyperbilirubinemia (4%) delayed birth cry (4%) medications in the first trimester (3%). Additionally, the etiological factors were analyzed in terms of geographic locations, socio-economic status, religious community and gender. Results revealed a significant difference in occurrence of consanguinity between the two geographical locations, religious community and gender. It can be concluded that consanguineous marriage as a major etiological factor that lead to congenital severe to profound hearing loss should be discouraged. Preventive measures should focus on geographic locations, socio-economic status, and religious community that exhibit higher rate of consanguineous marriages. Public education should be carried out regarding the adverse effect of interrelated marriages. Genetic counseling has to be carried out at least for those at risk of developing genetic diseases, including hearing impairment. Premarital and antenatal screening can also to be utilized.*

## Introduction

Congenital, severe to profound hearing impairment is one of the disabilities that have severe impact on speech and language abilities, academic achievement, social development and vocational preferences. In India, hearing impairment is the second most common disability after locomotor impairment (National Sample Survey, 2002). Furthermore, hearing impairment accounts for 10% of all disabilities in the rural population and 9% in the urban population. It was also revealed that nearly 7% of the people in India are born with hearing loss (NSS, 2002). High incidence of deafness insists the need for preventive research in this area.

Diverse etiological factors have been associated with congenital hearing loss. Medical conditions that occur during prenatal, perinatal and postnatal periods can results in pre-lingual deafness. Maternal rubella, cytomegalovirus, syphilis, ototoxic drugs during pregnancy are a few of the prenatal conditions leading to deafness. Hypoxia, neonatal hyperbilirubinemia, ototoxic drugs, infections etc. in the perinatal and postnatal periods can also lead to hearing loss (Northern & Downs, 2002). However, nearly 50% of the congenital hearing loss has been reported to be of genetic in origin (Fraser, 1976). Additionally, a large majority of genetic deafness has been reported to be due to autosomal recessive inheritance (Smith, 1986). Autosomal recessive inheritance is considered, when other causative factors such as infections, trauma, ototoxic drugs are not plausible. Such an inheritance is probable when the parents of those with hearing impairment are known to be relatives. In developing countries, consanguineous marriage is one of the major causes of hereditary sensorineural hearing loss. Zakzouk (2002) reported that prevalence of hearing impairment was significantly higher in children whose parents were related. Similarly, Bener, EIHakeem and Abdulhadi (2005) reported that parental consanguinity was significantly higher among infants with hearing loss compared to those with no hearing loss. Both the studies were carried out in developing countries of Middle East. In India, higher rate of consanguineous marriages have been found in the southern states than that of north and northeastern states (National Family and Health Survey, 1992-1993). Indian states of Andhra Pradesh, Karnataka and Tamil Nadu had higher rate of consanguineous marriages whereas, in Kerala it is the lowest (Bittles, 2002). In this context the present study attempts to compare the etiological factors of congenital hearing loss in two south Indian states of India, Kerala and Karnataka.

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There have been reports of different rate of consanguineous marriage in different religious communities. Further, those belong to poor socio economic status had higher prevalence of consanguineous marriages (Bittles, 2002). Higher rate of consanguineous marriages have been in the illiterate community than that of literate community (Zakzouk, 2002) Hence, the present study also investigates the association between geographic locations, socio-economic status, and religious community on etiological factors especially consanguineous marriage.

## Method

*Participants:* The high-risk register (Anitha & Yathiraj, 2001) was administered on parents of children with hearing impairment (APPENDIX -A). There were 70 children (37 Male and 33 Female) in the age range of 2 to 9 years with the mean age of 4.3 years and standard deviation of 1.4. All the children were diagnosed at the Department of Audiology, AIISH. All the children had congenital, bilateral, severe-to- profound sensorineural hearing loss. All of them were fitted binaurally with hearing aids and attending listening training. Hearing loss was the major compliant in all the children with no associated conditions such as mental retardation, cerebral palsy or autism. Participants from two different geographic locations (33 Kerala & 37 Karnataka) were chosen, as majority of the patients at AIISH belong to these locations. In addition to the questions in the HRR, information regarding religious community (27 Muslim & 43 Hindu) and socio-economic status (59 Slab I & 11 Slab II) were also collected (Table 1). Slab I consisted of patients with monthly income less than 10,000. Those with monthly income between 10,000 to 20,000 were grouped into slab II.

*Table 1: Number of participants from to two geographic locations having different religious community, socio-economic status, gender*

| | Kerala | Karnataka |
|---|---|---|
| **Religious community** | | |
| Hindu | 7(21%) | 36 (97%) |
| Muslim | 26 (79%) | 1 (3%) |
| **Economic status** | | |
| Slab 1 | 25 (76%) | 34 (91%) |
| Slab 2 | 8 (24%) | 3 (9%) |
| **Gender** | | |
| Male | 20 (60%) | 17 (46%) |
| Female | 13(40%) | 20 (54%) |

The HRR consists of two questionnaires; one can be administered by a medical professional and the other one by a non-medical professional. Each of the questionnaires had two sub divisions; one consists of risk factors that can occur during birth to 28 days and the other one between 28 days to 3 years. Both the questionnaires were administered on each parent. After collecting information, the data was analyzed descriptively.

*Procedure:* As the HRR was developed in English, each question was translated to respective mother tongue of the participants. The questions were explained orally to each participant in person. . The data was collected from each parent in a silent room within the premises of the institute. Information given by each parent was verified with their case history that was collected earlier. The responses were collected in the form of yes or no format. All the parents were assured that the information collected would be used for research purpose and an informed consent was obtained.

## Results

It was observed that 73% of the participants had one or the other etiological factor associated with hearing loss. However, remaining 27% had no relevant clinical history. Among the etiological factors noticed, consanguineous marriage, maternal infections during pregnancy, high fever, NICU care more than a week, neonatal hyperbilirubinemia, family history of congenital hearing loss, delayed birth cry, medications in the first trimester were found. It can be noted in Figure 1 that parental consanguinity (31%) was the most common cause followed by maternal infections (13%). There were patients with a history of high fever (7%), NICU care more than a week (6%), family history of congenital hearing loss (4%), neonatal hyperbilirubinemia (4%) delayed birth cry (4%) medications in the first trimester (3%). Premature birth was associated with the above etiological factors in 15% of the participants. However, none of them who had parental consanguinity exhibited any other associated etiological factors indicating a hereditary hearing loss.

The above etiological factors were further grouped in terms of geographic locations, socio-economic status, religious community and gender.

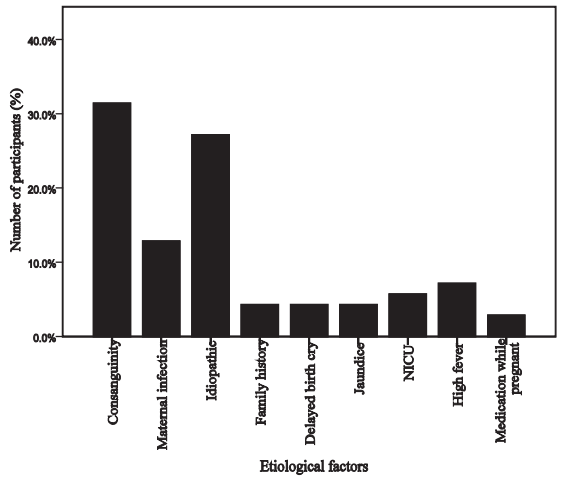


*Figure 1: Number of participants (in percentage) with different etiological factors*

Geographic locations: Among the 70 patients, 47% were from Kerala and 53% were from Karnataka. It can be found in Figure 2 that, consanguineous marriage was noted in 49% of the participants from Karnataka whereas, only 12% of the participants from Kerala had history of parental consanguinity. Chi square test revealed that there was a significant difference in consanguinity between the two geographical locations ($p < 0.05$). Maternal infections during pregnancy were found in 24% of the participants from Kerala. However, there were only 3% of the participants from Karnataka had maternal infections as an etiological factor. Similar number of participants from Kerala (24%) and Karnataka (29%) had idiopathic onset of hearing loss. Other etiological factors also exhibited similar percentage of occurrence between the two geographical locations.

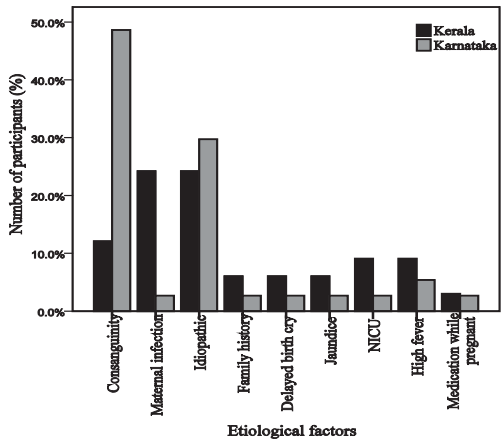


*Figure 2: Number of participants (in percentage) from two geographical locations with different etiological factors*

*Religious community:* Among the 70 patients, 61% belonged to Hindu community and 39% belonged to Muslim community. It can be found in Figure 3 that, consanguineous marriage was noted in 42% of the participants belonged to Hindu community whereas, only 15% of the participants belonged to Muslim community exhibited parental consanguinity. Chi square test revealed that there was a significant difference in consanguinity between the two religious communities ($p < 0.05$). Maternal infections during pregnancy were found in 26% of the participants from Muslim community nevertheless, there were only 5% of the participants belonged to Hindu community who had maternal infections as an etiological factor. Other etiological factors exhibited similar percentage of occurrence between the two religious communities.

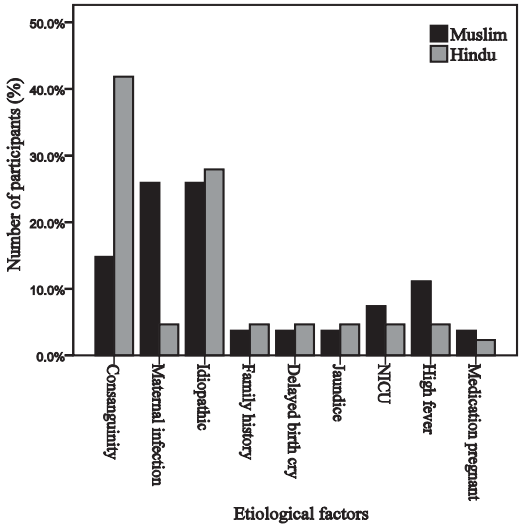


*Figure 3: Number of participants (in percentage) belonged to two religious communities with different etiological factors*

*Socio-economic status:* Out of the 70 participants, 84% belonged to slab I and 16% belonged to Slab II. It can be found in Figure 4 that, etiological factors such as consanguineous marriage, maternal infections during pregnancy, idiopathic onset, family history and history of NICU were found to be higher in slab I. In slab II conditions such as delayed birth cry, neonatal hyperbilirubinemia and high fever were not observed.

## Discussion

It was noted that 73% of the participants with congenital severe to profound hearing loss had at least one causative factor associated with their condition whereas the remaining 27% had idiopathic onset of hearing loss.

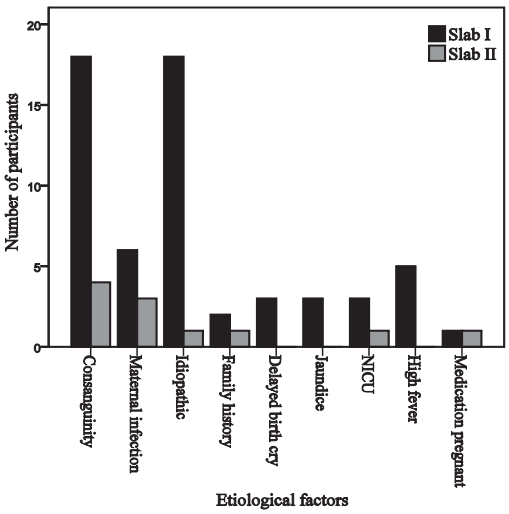


*Figure 4: Number of participants belonged to two socio-economic background with different etiological factors*

It was observed that parental consanguinity was the most commonly observed etiological factor (31%) followed by maternal infections (13%). Studies have reported higher rate of parental consanguinity in children with congenital hearing loss (Reddy et al. 2004). They reported that 58% of 138 children with syndromic hearing loss had history of parental consanguinity. Higher rate of consanguinity in the present study might be due to the geographic location where the study was carried out. It has been noted that compared to northern and northeastern states consanguineous marriages are highly common in southern states of India (Kapadia, 1958).

It was found that those who had parental consanguinity did not exhibit any other associated etiological factors indicating a hereditary hearing loss. Hence, the marriages between people having common ancestor increase the risk of transmitting the detrimental recessive gene that they inherited from their common ancestor. Autosomal recessive inheritance was found to be the major cause of such hereditary hearing loss (Smith, 1986).

*Geographical locations:* It was found that consanguineous marriages were significantly higher among those from Karnataka (49%) than that of Kerala (12%). Further, maternal infection was found in 24% of the participants from Kerala whereas; only 3% of the participants from Karnataka had history of maternal infection. State wise difference in consanguinity found in the present study is in accordance with the reports of National Family and Health Survey (1992-1993). They reported, higher rate of consanguineous marriages in the south Indian states of Andhra Pradesh, Karnataka and Tamil Nadu whereas, in Kerala it is the lowest. Additionally, Bittles et al. (1991) reported 31.4% consanguinity all over Karnataka whereas in Kerala only 13% was observed (Ali, 1968). Though the number consanguineous marriages were found to be lesser in Kerala, the prevalence of hearing impairment was found to be higher than the other south Indian states (NSS 2002). This highlights the need for multicenter studies in different geographic locations to explore the etiological factors leading to congenital hearing loss.

*Religious community:* The results revealed that consanguineous marriages were significantly higher among those belonged to Hindu community (42%) than that of Muslim community (15%). Additionally, maternal infection was found in 26% of those belonged to Muslim community and 5% of the Hindu community. Reddy et al. (2004) had similar findings where 87% of their patients belong to Hindu community in which 58% had history of consanguinity. Increased rate of consanguinity among Hindu community is accounted to the custom followed in Hindu families of south India (Bittles, 2002). Further, Hindu marriage act of 1955 and Hindu code bill of 1984 recognized cross-cousin and uncle-niece marriages. This was reported to be the probable cause for increased rate of consanguineous unions in Hindu community. In contrast, consanguineous marriages in south Indian Muslim community (10%) were found to be lesser than that of northern states (43.3%).

Increased rate of maternal infections observed in the Muslim community (26%) than that of Hindu community (5%) is unclear. Similarly, 24% of the participants from Kerala had maternal infections whereas in Karnataka only 3% had history of maternal infections. Additionally, it was found that 80% of the participants from Kerala belong to Muslim community. However, it is hard to explain the association between increased rate of maternal infection and geographical location or religious community.

*Socio-economic status:* It was found that consanguineous marriages accounted 82% of the etiological factors in those belong to slab I. In contrast, in slab II consanguinity accounted only 18% of the etiological factors. Higher rate of consanguineous marriages have been reported among those belong to rural areas, low in socio economic status and poor literacy (Bittles, 2002). It was believed in rural areas that such a custom might reduce the financial and health uncertainties that arise due to marriage with other families. Further, consanguineous unions might

simplify the premarital arrangements and lead to better couple and in law relationships (Bittles, 2002).

## Conclusions

The survey based study revealed that a consanguineous marriage was found to be the most common etiology followed by maternal infections. Additionally, it was noted that there was a significant difference in rate of consanguinity between geographical locations as well as religious community. Hence, it can be concluded that consanguineous marriages as a major etiological factor that lead to congenital severe to profound hearing loss should be discouraged. As there was a significant difference in consanguinity between different geographical locations and religious community, preventive measures should be focused on these groups. Public education should be carried out regarding the adverse effect of interrelated marriages. Genetic counseling, to be carried out at least for those at risk of developing genetic diseases, including hearing impairment. Premarital and antenatal screening can also to be utilized. However, the results of the study should be interpreted with caution due to its small sample size. Further, large-scale epidemiological studies need to be carried out to confirm the results.

## References


Ali, S. G. M. (1968). Inbreeding and endogamy in Kerala (India). *Acta Genetica et Statistica Medica* 18, 369-379.

Anitha, T., & Yathiraj, A. (2001). *Modified High Risk Registers (HRR) for Professional and Non Professional Formulation and its Efficacy*. Unpublished Independent Project submitted to Univ. of Mysore, as a part fulfillment of M.Sc. (Sp. & Hg.)

Bener, A., EIHakeem, A. M., & Abdulhadi, K. (2005). Is there any association between consanguinity and hearing loss. *International Journal of Pediatric Otorhinolaryngology*, *69* (3), 327-333.

Bittles A. H, Mason W. M., Greene J., & Appaji Rao, N. (1991). Reproductive behavior and health in consanguineous marriages. *Science*. 252, 789-794.

Bittles, A. H. (2002). Endogamy, consanguinity and community genetics. *Journal of Genetics, 81,* 91-98.

Fraser, G. R. (1976). *The causes of profound deafness in childhood*. Baltimore; The Johns Hopkins University press.

Kapadia, K. M. (1958). *Marriage and Family in India (2nd edition)*. Oxford University Press, Kolkata.

National family and health survey report (1992-1993), Report No. 1

National Sample Survey Organization (NSSO) report. Disabled Persons in India. Report No. 485, 2002, 58/26/1.

Northern, J. L & Downs, M. P. (2002). Medical aspects of hearing loss (5th edition). Hearing in children. Lippincott Williams and Wilkins. Pp. 91-124.

Reddy, M. V. V, Sathyanarayana, V. V. V, Sailakshmi, V, Hemabindu, L., Usha Rani, P., & Reddy, P. P. (2004). An epidemiological study on children with syndromic hearing loss. *Indian Journal of Otolaryngology and Head and Neck Surgery, 56,* 208-213.

Smith, R. J. H. (1986). Medical diagnosis and treatment of hearing loss in children. In C. W. Cummings (Ed.) *Otolaryngology Head and Neck Surgery, 4*, 3225-46. St. Louis: CV Mosby Co.

Zakzouk, S. (2002). Consanguinity and hearing impairment in developing countries: a custom to be discouraged. *The Journal of Laryngology and Otology, 116*, 811-816.


## APPENDIX-A

HIGH RISK REGISTER FOR NON-MEDICAL PERSONS

Birth – 28 days

1. Are the parents of the child blood relatives?
2. Did anyone in the child's family have hearing loss in early childhood?
3. Did the child's mother have any serious illness during pregnancy?
4. Did the child's mother take any medicines for illness during pregnancy?
5. Was the baby born before the due date given by the doctor (before 37 weeks from last menstrual period)?
6. Did the child appear yellow or blue at birth?
7. Did the child cry immediately after birth?
8. Was the child's weight low at birth (less than 1.5 kg)?
9. Was there any defect of the head and face when the child was born?
10. Was the child kept in hospital for treatment after birth?

29 days – 3 years

1. Was there parental or caregiver concern regarding the child's hearing, speech or developmental milestones?
2. Did anyone in the child's family have hearing loss in early childhood?
3. Did the child's mother have any infections during pregnancy?
4. Was there any defect of the head and face when the child was born?
5. Did the child's skin appear yellow?
6. Did the child have brain fever, measles or mumps?
7. Did the child have head injury associated with loss of consciousness, skull fracture, bleeding or discharge from ear following injury?
8. Did the child have ear discharge for at least 3 months?

NOTE: I f the answer to any of the questions is 'YES', get the child's hearing evaluated by a qualified Audiologist.

HIGH RISK REGISTER FOR MEDICAL PROFESSIONALS

Birth – 28 days

1. Was the marriage of the child's parents consanguineous?
2. Was there any family history of permanent early childhood sensorineural hearing loss?
3. Did the child's mother have any conditions during pregnancy such as measles, mumps, chickenpox, herpes, syphilis, cytomegalovirus, rubella or toxoplasmosis?
4. Was the child's mother hospitalized for long prior to delivery of the child?
5. Did the child's mother take any ototoxic medication for illness during pregnancy?
6. Was the child born prematurely?
7. Was the child's birth cry delayed?
8. Did the child weight less than1500 grams at birth?
9. Did the child have hyperbilirubinemia at a serum level requiring exchange transfusion soon after birth?
10. Did the child have Apgar scores of 0-4 at 1 minute or 0-6 at 5 minutes?
11. Was there any craniofacial anomalies including those with structural abnormalities of the pinna and ear canal?

29 days– 3 years

1. Was there parental or caregiver concern regarding the child's hearing, speech or developmental milestones?
2. Was there any family history of permanent childhood sensori-neural hearing loss.
3. Did the child's mother have any infections such as herpes, cytomegalovirus, toxmoplasmosis, syphilis or rubella during pregnancy.
4. Did the chili have any craniofacial anomalies, including those with structural abnormalities of the pinna and ear canal?
5. Did the child have hyperbilirubinemia at a serum level requiring exchange transfusion soon after birth?
6. Did the child have any of the conditions known to be associated with sensori-neural hearing loss such as mumps, measles, bacterial meningitis, viral encephalitis or labyrinthitis?
7. Did the child have any trauma associated with loss of consciousness, skull fracture, bleeding or discharge from ear following trauma?
8. Did the child have recurrent or persistent otitis media with middle ear effusion for at least 3 months?

NOTE: I f the answer to any of the questions is 'YES', get the child's hearing evaluated by a qualified Audiologist.