THE SPECIFICITY OF A HIGH RISK REGISTER (HRR) DEVELOPED IN INDIA

Register No. 02SH0015

An Independent Project submitted in part fulfillment for the
First year **M.Sc, (Speech and Hearing)**
University of Mysore, Mysore.

ALL INDIA INSTITUTE OF SPEECH AND HEARING
Manasagangothri
Mysore

JUNE 2003

Dedicated to

TO ALL MIGHTY GOD

&

AMMA, APPAJI,
SMITHA,
SINDHU &
DEAR SWAROOP

CERTIGICATE

This is to certify that the Independent Project entitled : **"THE SPECIFICITY OF A HIGH RISK REGISTER (HRR) DEVELOPED IN INDIA"** is the bonafide work in part fulfillment for the degree of Master of Science (Speech and Hearing) of the student with Register No.02SH0015

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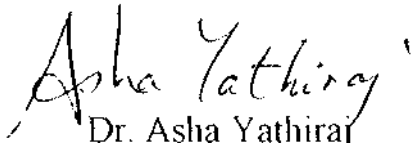
CERTIGICATE

This is to certify that the Independent Project entitled : **"THE SPECIFICITY OF A HIGH RISK REGISTER (HRR) DEVELOPED IN INDIA"** has been prepared under my supervision and guidance. It is also certified that this has not been submitted earlier in any other University for the award of any Diploma or Degree.



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DECLARATION

I hereby declare that this Independent Project entitled : **"THE SPECIFICITY OF A HIGH RISK REGISTER (HRR) DEVELOPED IN INDIA"** is the result of my own study under the guidance of Dr. Asha Yathiraj, Reader and HOD, Department of Audiology, All India Institute of Speech and Hearing, Mysore, and has not been submitted earlier at any other University for the award of any Diploma or Degree.

Mysore
June, 2003.

Reg. No. 02SH0015

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INTRODUCTION

Hearing loss in children is a hidden handicap, if undetected and untreated, hearing loss in pre-lingual children can lead to delayed speech and language development, social problem, emotional problems, and academic failure (Northern and Downs, 1991). The first three years of life are the most important for speech and language acquisition. To lessen the impact of hearing loss in the various area of development, hearing loss must be identified as early in life as possible and the child must be provided with habilitation in a timely manner (Camay and Moller, 1997, cited in Prieve and Stevens, 2000). Yoshinaga-Itano (1999), reported that if hearing loss in children is identified at the age of 2 month they will have better speech and language development compare to children identified at a later age.

The identification of hearing loss in new bom babies and infants is a challenging pursuit. According to Mahoney (1989, cited in Mauk and White. 1995; Northern and Hayes, cited in Mauk and White, 1995). over the past two decades, advances in technology have provided ever-improving opportunities to identify hearing impairments in infants soon after birth. The advantages of early intervention can only be attained when appropriate services are available and accessible to these children and their families. In recognition of the need to

identify hearing impairment as early in life as possible, auditory screening program should be implemented. This can be carried out in two ways :

- Universal new born hearing screening
- Screening infants at risk for hearing impairment

Universal new born hearing screening programmes would involve testing every child who is born with either a behavioral test or an objective test or a combination of both. Bess and Paradis (1994, cited in Northern and Downs, 2002) characterized universal screening for infant hearing impairments as "not simple, not risk free, not necessarily beneficial and not presently justifiable". Studies done in Israel in 1960's by (Feinmesser and Tell, (1975, 1976, 1982 cited in Gustason, 1989) indicated that the use of mass hearing screening of all infants at birth was too costly, impractical and undependable. Kirkwood (1999) reported of Dr Vohr's finding that screening each child costs about 30 to 40 US $. With the cost of screening all children being so high it would be impractical to carryout a universal screening program in India.

For the past 30 years or so, considerable effort has been devoted to the development of high-risk registers as a means for screening infants identified as being at high risk for a hearing impairment. The purpose of these screening has not been to specify the infants' hearing acuity, but rather to determine whether the possibility of hearing loss exists (Ruth, Dey-Sigman & Mills, 1985).

The term "high risk register" refers to a list of questions answerable at birth by parents or professionals and intended to increase the probability of finding a deaf infant (Jaffe, 1977). HRRs have the following advantages :

- They enable early identification of hearing impairment.
- They can be administered by any person such as Doctor's, Nurse's and Anganwadi workers.
- Time, effort and cost of administering the HRR is very low. Yathiraj, Sameer & Jayaram, (2001) reported that the average, cost of administering a HRR per child is Rs. 1.01/- and it could be brought down to Rs. 0.50/- with the use of allied professionals such as Anganwadi workers.
- Certain risk indicators of late onset hearing loss can only be identified by a HRR and not by hearing screening tests administered soon after the birth of a baby.

Most of the early prevalence of hearing loss estimates were determined from high-risk register studies of new born. By the early 1990's, however evidence from numerous studies confirmed that the use of high-risk registers as the basis of infant hearing screening programs identified only 50% of infants with significant hearing loss (Elssman, Matkin & Sabo, 1987; Mauk, White, Montesson & Beherens, 1991; Pappas, 1983). Mahoney and Eichwald (1979) reported the sensitivity and specificity of HRR as 65 percent and 75 percent respectively. Earlier Mencher (1974, cited in Gerber, 1977) showed that the use of the high risk

register leads to much higher correct detection in the newborn nursery that does the use of various screening methods. In addition, the risk factor information may be used to determine infants who are at risk for late onset hearing impairment and would therefore, need audiologic monitoring and follow up, despite a normal screen in the neonatal period (Brook-Houser, Worthington, & Kelly, 1994, cited in Vohr et al., 2000; Meyerhoff, Cass, Schwaber, Sculerati & Slattery, 1974) cited in Vohr, et al., 2000.

Any test should have a high sensitivity and high specificity (Jacobson and Jacobson, 1987, as cited in Hayes and Northern, 1996). This would apply to a HRR also. Information about the sensitivity and specificity can provide information about the number of persons correctly identified as hearing impaired or normal hearing as measures against predetermined pass-fail criteria (Northern and Downs, 1991).

Specificity : Refers to the test's accuracy in correctly identifying person who do not have the condition. It is the rate of correct classification for unaffected individuals (Hayes and Northern, 1996; Roush, 2001).

Sensitivity : Refers to the number of people with a given disorder who test positive, i.e., the rate of correct classification for affected individuals (Hayes and Northern, 1996; Roush, 2001).

The HRR developed by Anitha (2001) was evaluated by her to check its sensitivity. It was found by her that the HRR could identify 75.5% of the children with hearing impairment. The present study aims at establishing the specificity of the HRR developed by Anitha (2001). It also aims at noting its sensitivity using a general population instead of a group of hearing impaired individuals as done by Anitha (2001).

Need for the Study :

It is essential that the HRR should have a high specificity so that over referral of the clients does not take place (i.e. false positive). The false positive would result in unnecessarily having to test the hearing levels of these children. This would increase the cost involved in testing. False positive finding are known to cause undue psychological stress among the family members. The proficiency of the professional in identifying hearing impairment would also be evaluated based on the sensitivity as well as specificity of the test they utilize. A test with a high false positive would result in its face validity being reduced.

REVIEW OF LITERATURE

Hearing loss is caused by various factors. Many known risk factors have been reported as a possible cause of hearing impairment in newborn and young children (Jariengprasert, Sriwanyong, Kasemsuwan & Supapannachait, 2002).

The Joint Committee on Infant Hearing JCIH (2000) has identified specific risk factors that are often associated with infant and childhood hearing loss. This high risk register (HRR) has been developed after being revised several times.

The following review highlights the various HRRs that have been developed since the first reported HRR by Feinmesser and Bauberger-Tell in 1971 (cited in Diefendorf, 1982). The review also reports of the utility of these HRRs in infant screening programs as determined by a host of studies conducted over the years incorporating various versions of HRRs. The HRRs reviewed are given chronologically.

Feinmesser and Bauberger-Tell, 1971 (cited in Diefendorf, 1982) developed a high-risk register, which contained a large number of factors that might be associated with an increased probability of deafness. This HRR, was used to screen 17,731 newborns for hearing impairment. These infants also received a behavioral screen at birth and then at several intervals until they were

three years of age. By the end of the program twenty three deaf children were identified ; seventeen of them had been on the HRR and only six had been identified by behavioral screening.

The results of the study led Feinmesser and Bauberger-Tell (cited in Diefendorf, 1982) to conclude that the conventional behavioral observation methods was not sensitive enough to detect deafness in neonates. The authors also expressed some concern about the HRR containing so many factors since a large number of children need follow-up (20% or 3,546 children) evaluation. Through modification of the register, they reduced the number of neonates identified as high-risk to six to seven percent of the population and still retained a large percentage of high-risk hearing impaired infants. Their restricted HRR included the following items :

1. Familial deafness
2. Rubella during pregnancy
3. Birth weight of 1500 grams are less.
4. Congenital craniofacial malformations.
5. Apgar score of 1-4.
6. Hyperbilirubinemia : 20mg / 100ml and over.
7. Severe neonatal infections.

The first HRR developed by JCIH in 1972, had a list of five factors that Could identify a satisfactoiy number of deaf infants. These categories were referred to as the ABCDs of deafness (Downs and Silver, 1972, cited in Gerkin, 1984). The risk factors included were :

1. *Affected family :* The presence of any form of hearing loss (other than presbycusis-hearing loss that beings in older age in a family member.
2. *Bilirubin levels :* Any free or indirect concentration judged to be potentially toxic.
3. *Congenital rubella syndrome :* Rubella at any time during pregnancy, sometimes hearing loss is the sole symptom.
4. *Defects of the ears, nose or throat:* A malformed, low set, or absent pinna; a cleft palate or lip (including sub-mucous cleft); any residual abnormalities of the first arch; any other anatomic abnormality of the otorhinolaryngeal system.
5. *Small at birth :* Infants weighing less than 1500 grams at birth.

Hirsch and Kankkunen in 1974 sought to establish a method for early detection of hearing impairment using high-risk criteria. The children bom in two Maternity Hospitals (MHA and MHB) in Gothenburg in 1970 were evaluated based on the presence of risk factors for hearing impairment. MHA used a fixed list of high-risk criteria as the basis for request for audiological examination.

The high-risk criteria used by Maternity hospital A (MHA) was :

1. Family history of hearing impairment.
2. Rubella or some other viral disease during the first half of pregnancy.
3. Multiple malformations particularly of the face and ears.
4. Immaturity (birth weight less than 2500g).
5. Serum bilirabin more than 20 mg% for newboms, not diagnosed as immaturity.
6. Serum bilirubin more than 15 mg% for newborns with the diagnosis of immaturity.
7. Anoxia or neurological symptoms of cerebral origin for more than 24 hours.
8. Congenital infections.
9. Diabetes mellitus in the mother.
10. Hypoglycemia.

MHB had no fixed routine but requested an examination after a general evaluation of each baby. A total of 362 children were examined because of the presence of high-risk history or suspicion of deafness, of which ten children were found to be hearing impaired by the end of 1973. Four of these children were bom in MHA and six in MHB. All the children bom in MHA were identified by the age of nine months. However, three of the six children bom in MHB were not identified until they were 23 to 33 months of age even through they had serve

hearing loss. Based on the proportion of hearing impaired children identified with various risk indicators, the authors concluded that, nine of the ten children would have been diagnosed by the high-risk criteria if it had been used by both Maternity Hospitals. However this would have resulted in a large number of over referrals. Hence, they recommended alteration in the risk criteria by omitting diabetes mellitus in the mother and hypoglycemia as possible risk indicators. This would reduce the number of over referrals without diminishing the deafness identification efficiency.

Ashok Kumar (1981) developed a high-risk register relevant to an Indian Population. The high risk register developed by him was based on risk histories presented by mothers of a group of deaf children as compared to histories presented by the mothers of a randomly selected group of non-deaf children.

The high-risk factors considered in this study were :

1. Family History of hearing loss.
2. Consanguinity.
3. Maternal viral infection
4. Any pregnancy complications
5. Threatened abortion
6. Maternal medication
7. Delivery complications

8. Birth asphyxia
9. Cyanosis
10. Smallness at birth
11. Jaundice soon after birth
12. Blood transfusion soon after birth
13. Any Rh or blood group in compatibility
14. Birth deformities of head, ear, nose and throat.
15. Any neonatal illness
16. Seizures
17. Unconscious episodes.
18. Any injections given to the neonates
19. Parental concern about hearing
20. Parents evaluation of their child's hearing
21. Parents evaluation of their child's speech and language.

Information on the risk factors was collected from 369 randomly selected children and 83 confirmed deaf children. Based on the results of the comparative analysis of deaf and non-deaf children, a list of five questions were selected to make-up their high-risk register. The questions tackled the following factors :

1. Family history of hearing loss.
2. Consanguineous parentage, primarily involving uncle-niece marriages.
3. History of rashes and fever during pregnancy, irrespective of the trimester.

4. Report of Rh / blood group incompatibility
5. Parental concern about their child's hearing.

Their results indicated that by using this register at birth or soon after birth, 53.5% of the deaf children and with 21.22% of non-deaf children could be classified as at-risk. By applying the same register at 1 year of age, 75.72% of the deaf children were identified as at-risk without any corresponding increase in the number of non-deaf children classified at-risk.

The JCIH position statement (1982, cited in Hayes & Northern, 1996), recommended the modification of the risk criteria of the 1972 HRR to include :

- Bacterial meningitis , especially H influenza; and
- Severe asphyxia, which may include infants with Apgar scores of 0 to 3 who fail to institute spontaneous respiration by 10 minutes and these with hypotonia persisting to two hours of age.

They also modified the questions dealing with family histoiy of hearing impairment to include only those with a histoiy of childhood hearing impairment.

Fitzzaland (1985) reported on the results of a screening programme instituted in British Columbia using the JCIH (1972, cited in Gerkin, 1984) high-risk criteria 52, 858 infants were screened in the period from 1975 to 1982. Of

that number 5.1% infants were judged to be at risk out of which 21.2% were eventually found to have a hearing impairment. Thus the screening false positive rate was 78.8%. The percentage of referrals for each risk indicator along with the false positive rate and percentage of hearing impaired infants identified were also reported.

The largest number of referrals, 72% was due to a positive response for family histoiy. The false positive rate for this risk factor was also veiy high. However, 59.1% of hearing impaired children were correctly identified with this criterion as well. Therefore, the author concluded that since the percentage of infants correctly identified was high, the high referral rate is well worth the cost. These findings of high false positive rate was also supported by Gerber and Mencher, (1978, cited in Mencher and Mencher, 1985 ; Mencher and Gerber, 1981 cited in Mencher and Mencher, 1985).

Halpern, Hosford-Dunn & Malachowski (1987) examined risk factors including those mentioned in the JCIH (1982, cited in Hayes and Northern, 1996), in a large group (975) of NICU infants. The babies selected had one or more of the following risk factors.

1. Birth weight < 1500g
2. > 24 hours respirator support
3. < 38 weeks gestational age

4. Suspected or confirmed sepsis that was treated with aminoglycosides for > 6 days
5. "TORCH" complex congenital prenatal viral infections (Toxoplasmosis, Rubella, Cytomegalovirus, Herpes), and
6. Craniofacial anomalies

These selection criteria were patterned on the JCIH HRR, with the exception of hyperbilimbinemia, family histoiy of hearing less and low Apgar scores. The subjects were tested in intervals from birth through 3 years of age using a Crib-O-Gram for neonatal testing and behavioural sound field testing for babies older than 6 months. Hearing diagnosis was made for 820 babies of which 50 had confirmed cochlear loss and one baby had permanent conductive loss.

The medical records of these 820 babies were reviewed for variables that might be associated with hearing loss. These variables included all seven JCIH HRR items and other descriptors of ICN stay, as shown below.

1. Days in ICN (admission to discharge)
2. Respiratory distress
3. Hours of constant positive pressure ventilation
4. Hours of constant positive airway pressure
5. Intubation
6. Gestational age
7. Birth weight

8. Bacterial meningitis (neonatal)
9. Bilirubinemia level
10. Cytomegalo virus
11. Toxoplasmosis
12. Rubella
13. Herpes
14. Microcephaly
15. Retrolental fibroplasia
16. Craniofacial anomalies
17. Perinatal asphyxia
18. Meconium aspiration and meconium aspiration syndrome
19. Apgar (5 minute score)
20. Days of aminoglycoside therapy

Most of the HRR items were significantly associated with hearing loss in these 820 babies. However, the four variables-craniofacial anomalies, TORCH infections, length of stay in ICN and gestational age-predicted hearing loss with 98% sensitivity and reduced the initial risk group by one-third. Of these four variables two (craniofacial anomalies and TORCH infections) are from the HRR and the other two (length of ICN stay and gestational age) are variables related to ICN stay. If the prediction of hearing loss was based on HRR alone the sensitivity was about 76% and the number of subjects who required testing remained the same.

Stein, Jabaley, Spitz, Stoakley & McGee (1990) examined the occurrence of risk factors and the patterns of identification and habilitation, in a group of hearing impaired infants, over the period of 1983-1988. They also compared their data with that of a previous study (Stein, Clark & Kraus, 1983) that reported results of a screening program over the period from 1980-1982. The subjects in both studies included babies from well baby nursery and N1CU. The subjects were screened with the JCIH 1982 HRR. Follow-up evaluation of the high-risk babies were done using ABR, immittance and behavioral audiometry. Only subjects with bilateral severe to profound hearing losses were considered in the analysis.

The major findings of this study was that only one-third of the identified hearing impaired infants were NICU graduates. Similar findings have been reported by Elssmann, Matkin & Sabo (1987) and Stein, Clark & Kraus (1983). Alberti, Hyde, Riko, Corbin & Fitzhardinge (1985). They also reported a substantial incidence of hearing impairment in the non-NICU group which was higher than in the NICU group. Family histoiy of hearing loss and ENT defects as in the case of cleft lip / palate are found to be the most common risk factors in the non ICU group (Alberti, Hyde, Riko, Corbin & Fitzhardinge, 1985). Infants with such risk factors may not require specialized care in an NICU. Since audiological screening is usually concentrated in NICU settings where the yield of true positive identification is projected to be higher, only one in three hearing impaired children

can be expected to be identified. Therefore, it is recommended to screen babies from well baby nursery as well.

Mahoney and Eichwald (1986, 1987) reported that one of the most successful methods of collecting information about the presence of the high-risk factors was to incorporate the relevant information into the legally required birth certificate.

Mauk, White, Mortensen & Behrens (1991) evaluated the efficiency of the birth-certificate based screening programs incorporating the JCIH (1982, cited in Hayes and Northern, 1996) HRR. 70 parents and guardians of six to nine year old children with significant sensori-neural losses were surveyed regarding their child's identification history. Results indicated that only 50% of the children with sensori-neural hearing loss exhibited any of the risk factors recommended by the JCIH (1982, cited in Hayes and Northern, 1996). 33f admittance to an NICU was included as a risk factor then 63% of the children with sensori-neural losses would have been identified as at-risk. However, almost 40% of hearing impaired children who did not exhibit any of the risk factors would not have been referred for detection assessment. These findings are in agreement with studies by Stein, Clark & Kraus (1983) and Elssman, Matkin & Sabo (1987) who reported that 30% and 50% of hearing impaired children respectively do not exhibit high-risk factors. Therefore, Mauk, White, Mortensen & Behrens (1991) suggested the need for

continued attention of regular hearing screenings upto and including the first years of formal education. Thus according to the findings of these studies the JCIH (1982, cited in Hayes and Northern, 1996), HRR seemed to have a poor sensitivity in identifying hearing impairment.

The JCIH further expanded the risk criteria in (1990, cited in Hayes and Northern, 1996). Here the risk criteria were divided into two categories. Those present during the neonatal period and those that develop within the first 2 years of life.

A. Risk Criteria : Neonates (birth-28 days)

The risk factors that identify those neonates who are at-risk for sensori-neural hearing impairment include the following :

1. Family history of congenital or delayed onset childhood sensori-neural impairment.
2. Congenital infection known or suspected to be associated with sensori-neural hearing impairment such as toxoplasmosis. syphilis, rubella, cytomegalovirus and herpes.
3. Craniofacial anomalies including morphologic abnormalities of the pinna and ear canal, absent philtrum, low hairline, etcetera.
4. Birth weight less than 1500 grams (~ 3.3 lbs.).

5. Hyperbilirubinemia at a level exceeding indication for exchange transfusion.
6. Ototoxic medications including but not limited to the aminoglycosides used for more than 5 days (e.g., gentamicin, tobramycin, kanamycin, streptomycin) and loop diuretics used in combination with aminoglycosides.
7. Bacterial meningitis.
8. Severe depression at birth, which may include infants with Apgar scores of 0-3 at 5 minutes or those who fail to initiate spontaneous respiration by 10 minutes or those with hypotonia persisting to 2 hours of age.
9. Prolonged mechanical ventilation for a duration equal to or greater than 10 days (e.g., persistent pulmonary hypertension).
10. Stigmata or other findings associated with a syndrome known to include sensori-neural hearing loss (e.g., Waardenburg or Usher's Syndrome).

B. Risk Criteria : Infants (29** days-2 **years)

The factors that identify those infants who are at-risk for sensori-neural hearing impairment include the following :

1. Parent / caregiver concern regarding hearing, speech, language and / or developmental delay.
2. Bacterial meningitis.

3. Neonatal risk factors that may be associated with progressive sensori-neural hearing loss (e.g., cytomegalovirus, prolonged mechanical ventilation and inherited disorders).
4. Head trauma especially with either longitudinal or transverse fracture of the temporal bone.
5. Stigmata or other findings associated with syndromes known to include sensori-neural hearing loss (e.g., Waardenburg or Usher's Syndrome).
6. Ototoxic medications including but not limited to the aminoglycosides used for more than 5 days (e.g., gentamicin, tobramycin, kanamycin, streptomycin) and loop diuretics used in combination with aminoglycosides.
7. Children with neurodegenerative disorders such as neurofibromatosis, myoclonic epilepsy, Werding-Hoffman disease, Tay-Sach's disease, infantile Gaucherie's disease, Nieman-Pick disease, any metachromatic leukodystrophy, or any infantile demyelintaing neuropathy.
8. Childhood infectious diseases known to be associated with sensori-neural hearing loss (e.g., mumps, measles).

The committee also recommended a specific hearing screening protocol using behavioral methods and ABR.

In 1994 the JCIH (cited in Hayes and Northern, 1996) addressed the need to identify all infants with hearing loss. Since high risk factor screening identifies

only 50% of infants with significant hearing loss (Elssman, Matkin & Sabo, 1987 ; Mauk, White, Mortensen & Behrens, 1991), the committee advocated universal neonatal hearing screening wherever and whenever possible. However it maintained a role for the high-risk factors described in the 1990 position statement, with modification of the list of indicators associated with sensori-neural and / or conductive hearing losses in newborns and infants. It also recommended an additional category of risk factors associated with late-onset hearing loss. A further modification was the addition of persistent / recurrent otitis media with effusion as a risk indicator. Given below is the position statement given by JCIH in (1994, cited in Hayes and Northern, 1996).

Indicators Associated with Sensori-Neural and/or Conductive Hearing Loss :

A. For use with neonates (birth through age 28 days) when universal screening is not available.

1. Family history of hereditary childhood sensori-neural hearing loss.
2. In utero infection, such as cytomegalovirus, rubella, syphillis, herpes, and toxoplasmosis.
3. Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
4. Birth weight less than 1,500 grams (3.3 lbs).
5. Hyperbilirubinemia at a serum level requiring change transfusion.

6. Ototoxic medications, including but not limited to the aminoglycosides, used in multiple courses or in combination with loop diuretics.
7. Bacterial meningitis.
8. Apgar scores of 0-4 at 1 minute or 0-6 at 5 minutes.
9. Mechanical ventilation lasting 5 days or longer.
10. Stigmata or other findings associated with a syndrome known to include a sensori-neural and / or conductive hearing loss.

B. For use with infants (age 29 days through 2 years) when certain health conditions develop that require rescreening.

1. Parent / Caregiver concern regarding hearing, speech, language, and / or developmental delay.
2. Bacterial meningitis and other infections associated with sensori-neural hearing loss.
3. Head trauma associated with loss of consciousness or skull fracture.
4. Stigmata or other findings associated with a syndrome known to include a sensori-neural and / or conductive hearing loss.
5. Ototoxic medications, including but not limited to chemotherapeutic agents or aminoglycosides, used in multiple courses or in combination with loop diuretics.
6. Recurrent or persistent otitis media with effusion for at least 3 months.

C. For use with infants (age 29 days through 3 years) who require periodic monitoring of hearing.

Some newborns and infants may pass initial hearing screening but require periodic monitoring of hearing to detect delayed-onset sensori-neural and / or conductive hearing loss. Infants with these indicators require hearing evaluation at least every 6 months until age 3 years, and at appropriate intervals thereafter.

Indicators associated with delayed-onset sensori-neural hearing loss include :

1. Family history of hereditary childhood hearing loss.
2. In utero infection, such as cytomegaloviurs, rubella, syphilis, herpes, or toxoplasmosis.
3. Neurofibromatosis type II and neurodegenerative disorders.

Indicators associated with conductive hearing loss include :

1. Recurrent or persistent otitis media with effusion.
2. Anatomic deformities and other disorders that affect eustachian tube function.
3. Neurodegenerative disorders.

These risk indicators have been revised further in the year 2000 based on the data from more recent research (Fortnum and Davis, 1997; Norton et al., 2000). Vohr et al., (2000) analysed the prevalence of risk indicators for infants

identified with hearing loss in the study of Norton et a1, (2000). 3,134 infants evaluated during their initial birth hospitalization were re-evaluated for the presence of hearing loss between eight and twelve months of age. The majority of these infants were NICU graduates (2,847) and the remaining (287) had risk indicators for hearing loss that did not require intensive care, such as family history or craniofacial anomalies. Infants with history or evidence of transient middle ear dysfunction were excluded from the final analysis, revealing 56 with permanent hearing loss.

An examination of the prevalence of the various risk indicators in the hearing impaired children revealed that syndromes had a prevalence of 11.7%, family history of hearing loss 6.6%, meningitis 5.5% and craniofacial abnormalities 4.7%. In contrast infants treated with aminoglycoside antibiotics had a prevalence of hearing loss of only 1.5%. Based on these findings the JCIH risk indicators were modified in 2000 (Joint Committee on Infant Hearing 2000 - Position statement). The indicators are :

A. Risk indicators for use in neonates (birth through 28 days) where universal hearing screening is not available :

1 An illness or condition requiring admission of 48 hours or greater to a NICU.

2. Stigmata or other findings associated with a syndrome known to include a sensori-neural and or conductive hearing loss.
3. Family history of permanent childhood sensori-neural hearing loss.
4. Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
5. In-utero infection such as cytomegalovirus, herpes, toxoplasmosis, or rubella.

Risk indicators for infants (29 days through 2 years).

1. Parental or caregiver concern regarding hearing, speech, language, and / or developmental delay.
2. Family history of permanent childhood healing loss.
3. Stigmata or other findings associated with a syndrome known to include a sensori-neural or conductive hearing loss or Eustachian tube dysfunction.
4. Postnatal infections associated with sensori-neural hearing loss including bacterial meningitis.
5. In-utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis.
6. Neonatal indicators, specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation.

7. Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher's syndrome.
8. Neurodegenerative disorders, such as Hunter's syndrome, or sensory motor neuropathies, such as friedreich's ataxia and Charcot-Marie-Tooth syndrome.
9. Head trauma.
10. Recurrent or persistent otitis media with effusion for at least 3 months.

The committee recommended that any infant with these risk indicators for progressive or delayed-onset hearing loss, who has passed the birth screen should receive audiologic monitoring every 6 months until the age of 3 years.

Sunil (1993), used a check-list containing 18 questions on 90 subjects. These questions were :

1. Is any one in the (child's) family, on the father's side or mother's side, having a severe hearing problem since childhood?
2. Is any one in the (child's father's family or mother's) family having a speech problem?
3. Is any one in the (child's father's family or mother's) who has a cleft lip and / or cleft palate?
4. Does the child have ears which look different i.e., abnormal (too small, rather big, slightly away from where ears are normally found).

5. Does the child have a cleft lip or cleft palate?
6. Is the child's jaw or tongue different i.e., abnormal?
7. Did the (child's) mother take any drugs during pregnancy?
8. Did the (child's) mother have illness such as measles, mumps, chicken pox etc, during pregnancy?
9. Did the (child's) mother require treatment for conditions such as blood pressure during pregnancy?
10. Did the (child's) mother notice bleeding during pregnancy?
11. Was the (child's) mother exposed to radiations, such as X-rays, during pregnancy?
12. Was the (child's) mother hospitalized for long prior to delivery of the child?
13. Did the child weigh much less than normal at the time of birth?
14. Was the child bom prematurely? By how many weeks? If yes, say the number.
15. Was the child's appearance blue at the time of birth?
16. Did the child not cry immediately after birth but did so after some time?
17. Was the child given blood transfusion soon after birth?
18. Was the child's appearance yellow at the time of birth?

The presence of a hearing loss was confirmed using a behavioural observation audiometry (BOA). It was found that 20% of newborn population

were at-risk. The risk factor that had the maximum prevalence was "low birth weight[7]. The factors that had the least prevalence was "X-ray exposure by the mother" and "blood transfusion postnatally". Only 50% of the at-risk group had a suspected hearing loss as per the BOA test.

Anitha (2001) formulated the HRR specifically for children in the age groups of birth-28 days and 29 days-3 years for medical and non-medical persons. She also checked the sensitivity of the HRRs developed by her, by administering them on the parents or caregivers of 200 children who had confirmed hearing loss. She developed four questionnaires, with two ment for medical professionals and two for non-medical professionals. For each of these professionals two questionnaires, each for different age groups was included. The four questionnaire developed by her were as follows :

***HRR FOR MEDICAL PERSONS (Birth** - 28 days)*

1. Was the marriage of the child's parents consanguineous?
2. Was there any family histoiy of permanent early childhood sensori-neural hearing loss?
3. Was the child's mother exposed to radiations such as X-rays during pregnancy?

4. Did the child's mother have any conditions during pregnancy such as measles, mumps, chickenpox, herpes, syphilis, cytomegalovirus, rubella or toxoplasmosis?
5. Was the child's mother hospitalized for long prior to delivery of the child?
6. Did the child's mother take any Ototoxic medications for illness during pregnancy?
7. Was the child born prematurely?
8. Was the child's birth cry delayed?
9. Did the child weigh less than 1500 grams at birth?
10. Did the child have hyperbilirubinemia at a serum level requiring exchange transfusion soon after birth?
11. Did the child have Apgar scores of 0-4 at 1 minute or 0-6 at 5 minutes?
12. Was there any craniofacial anomalies including those with structural abnormalities of the pinna and ear canal?

HRR FOR MEDICAL PERSONS (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 early childhood sensori-neural hearing loss?

3. Did the child's mother have any infections such as herpes, cytomegalovirus, toxoplasmosis, syphilis or rubella during pregnancy?
4. Did the child have any craniofacial anomalies, including those with structural abnormalities of the pinna and ear canal?
5. Did the child have hyperbilimbinemia at a serum level requiring exchange transfusion?
6. Did the child have any of the conditions known to be associated with sensori-neural hearing loss such as measles, mumps, bacterial meningitis, viral encephalitis or labyrinthitis?
7. Did the child have any head 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 effusion for at least 3 months?.

HRR FOR NON-MEDICAL PERSONS (Birth - 28 days)

1. Are the parents of the child blood relatives?
2. Did any one in the child's family have hearing loss in early childhood?
3. Was the child's mother exposed to X-rays during pregnancy?
4. Did the child's mother have any serious illness during-pregnancy?____
5. Did the child's mother take any medicines for illness during pregnancy?
6. Was the baby born before the due date given by the doctor (before 37 weeks from last menstrual period)?

7. Did the child appear yellow or blue at birth?
8. Did the child cry immediately after birth?
9. Was the child's weight low at birth (less than 1.5 kg)?
10. Was there any defects of the head and face when the child was born?
11. Was the child kept in hospital for treatment after birth?

HRR FOR NON-MEDICAL PERSONS (29 days - 3 years)

1. Was there parental or caregiver concern regarding the child's hearing, speech or developmental milestones?
2. Did any one 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 defects of the head and face when the child was bom?
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?

Result of her study showed that the sensitivity of HRR was 75.5% (i.e., 151 out of 200 children were at risk for hearing impairment). It was found by her that parental concern had the highest percentage of occurrence followed by

consanguinity. Maternal irradiation during pregnancy and head trauma to the child had the least percentage.

From the review, it is evident that HRR can be used as a tool for hearing screening as it is time effective and cost effective unlike a Universal Hearing Screening Programme.

TECHNIQUES FOR NEONATAL HEARING SCREENING :

A child's hearing can be tested in a variety of ways. The selection of a measurement technique is dependent on a number of variables including the age of the child, his / her ability to participate in the test procedure. Screening for hearing loss in infants / young children can be categorized under the following four headings :

I. Subjective techniques

II. Semi-objective techniques

III. Objective techniques

IV. Speech tests for hearing screening

/. Subjective Techniques:

a. Behavioural observation audiometry (Ewing & Ewing, 1994).

b. Visual Reinforcement Audiometry (Liden and Kankkunen, 1969).

c. Tangible Reinforcement Operant Conditioning Audiometiy (Lloyd, Spadlin & Reid, 1968).

d. Conditioned play audiometry (Utley, 1949, cited in Lloyd, 1969).

e. Pure Tone Hearing Screening (Newhart, 1948).

//. ***Semi-Objective Techniques :***

a. Crib-O-Gram (Simmons & Russ, 1975).

b. Auditory Response Cradle (Bennett, 1975^ cited in Wharrad, 1994).

c. Accelerometer Recording System (Altman, Shenhav & Schandinzchky, 1975).

///. ***Objective Techniques :***

a. Oto Acoustic Emissions (Kemp, 1978)

b. Auditory Brainstem Response (Jewett and Williston, 1971, cited in Erenberg, 1999).

c. Immittance Audiometry (Keith, 1973, 1975).

d. Reflectometry (Teele and Teele, 1984, cited in Northern, 1988).

IV. Speech Tests for Hearing Screening :

a. The Ling's 5 Sound Test (Ling, 1978, cited in Markides, 1987).

b. Ling's 7 Sound Test (Ling, 1996).

c. The Co-operative Test (McCormick, 1994)

d. Four-Toy Eye Pointing Test (McCormick, 1988)

e. Toy Discrimination Test (McCormick, 1977, cited in McCormik, 1994)

f. Reed Screening Hearing Test (Reed, 1959, cited in Markides, 1987)

g. The Kendall Toy Test (Kendall, 1953, cited in Markides, 1987)

h. Verbal Auditory Screening Test for Pre-school Children (Griffing, Simonton & Hedgcock, 1967, cited in Martin, 1987).

Each of these tests are used for hearing screening of children of different age groups. The sensitivity and specificity of these tests vary. The sensitivity and specificity of the screening measure are given in Tables I, II and III.

Table-I: Sensitivity and specificity of behavioral test for newborn screening as given by different ,

Application / Method	Sources	Age **range**	**Test operating characteristics (%)**				
			Sensi-tivity	**Speci-ficity**	**Under referral**	**Over referral**	**Cut off criterion**
High Risk Register	Mahoney and Eichwald(1979. cited in Alpiner & McCarthy. 1987).	Birth	65	**75**	32	68	Positive on one or more factors
High Risk Register	Anitha(2001)	Birth	75.5				Positive on one or more factors
Behavioral arousal	Downs and Sterritt (1967)	0-4 months	63	**29**	25	75	50 dBHL
Distraction Test (Noise makers)	Davis (1998)	12 to 20 months	Varies from 18-88				
Behavioral Visual Reinforcement Audiometry (VRA)	Kenworthy. Bess and Wright (1986 cited in Northern. 1988)	>6 months	80	**86**	14	20	30 dBHL
Audioscope screening	Bienvenue, Michael, Chaffinch and Zeigler (1985)	3 years (First screening)	75	**50**	25	50	500 Hz, 1kHz, 2 kHz, and 4 kHz, at 25 dBHL
		3 years (Second screening)	75	**50**	25	50	
		4 years (First screening)	80	**63**	20	37	
		4 years (Second screening)	80	**75**	20	25	
		5 years (First screening)	78	**73**	22	27	
		5 years (Second screening)	87	**80**	13	20	
pure-tone	Wilson and Walton (1978, cited in Alpiner & McCarthy. 1987)	>36 months	63	97	2	49	20 dBHL

Table-II : Sensitivity and specificity of Semi-objective Test for newborn screening as given by different authors.

Application / Method	**Sources**	Age **range**	**Test operating characteristics** (%)				
			Sensi-tivity	**Speci-ficity**	**Under referral**	**Over referral**	**Cut off criterion**
Crib-O-Gram	McFarland, Simmons and Jones(1973—1979)	Infants (well baby nursery)	100	90	**0**	**7**	92dBA
Crib-O-Gram	McFarland, Simmons and Jones (1973-1979)	Infants (Intensive Care nursery)	91	77	0.01	15	92dBA
Crib-O-Gram (Sensitivity and specificity for different BERA pass criteria)	Durieux-Smith, Picton, Edwards, Goodman and McMurray (1985)	31-37 weeks	47.1	66.3	52.9	33.7	30 dBnHL
		J I-J7 weeks	25.0	64	75	36	40 dBnHL
		31-37 weeks	0	64.1	100	35.9	50 dBnHL
		38-57 weeks	64.3	66.9	35.7	33	30dBnHL
		38-57 weeks	92.3	66.3	7.7	33.7	40 dBnHL
		38-57 weeks	100	65.4	**0**	34.6	50 dBnHL

Table-III : Sensitivity and specificity of Objective Technique for newborn screening as given by different authors.

Application / Method	Sources	Age range	Test operating characteristics (%)				
			Sensi-tivity	Speci-ficity	Under referral	Over referral	Cut off criterion
Immitance (Tympanometry in OME subjects)	Paradise and Colleagues (1976)	10 days to 5 Years 11 months	86	75	~	-	-
Immitance (Gradient)	Paradise and Colleagues (1976)	10 days to 5 Years 11 months	95	96		-	-
Autoscopy and Tympanometry	Paradise and Colleagues (1976)	10 days to 5 Years 11 months	97	90	-	-	-
Immitance (Traditional procedure based on TPP <-200	Roush, Dralce, & Sexlon (1992, cited in Margolis & Hunter, 1999)	3-4 years	95	65			TTP <-200 da Pa
Aroustic Immitance	Nozza(1992, cited in Margolis & Hunter, 1999)	1 -8 years	90	86			
Auditory Brainstum Responses (ABR)	Fria(1985, cited in Alpiner and McCarthy, 1987)	Birth to adult	98	90 to 95	2	50	30 and 60 dBnHL
Automated (ABR)	Hall, Kinleny & Ruth (1978, cited in Erenberg, 1999)	Infants	100	96 to 98	-	-	35 dBHL
	Jacobson, Sacobson & Spahr(1990, cited in Hayes & Northern, 1996)						

Transient Otoacoustic Emissions (TEOAEs) compared with ABR	Bonfils and Uzied(1989)	New born SNHL	95				> 40 dBHL
TEOAEs compare with ABR	Stevens Webb, Hutchiwon Connell, Smith and Buffin(1989).	Infants	95	84			30 dBHL
TEOAEs / ABR	Stevens(1990, cited in Davis, Bamford & Stevens, 2001)	Newton intensive care unit	94/ 84	84			
Programmable Otoacoustic Emissions Measurement System (POEMS)	Meredith, Stephens, Hogan, Cartlidge and Drayton(1994)	5 years	100	72.3			> 40 dBHL
OAE + ABR	Steven et. al., (1991, 1997, in Davis, Bamford & Stevens, 2001)	Infants	OAE =86 ABR =97				43 dBHL
Narrow Band TEOAEs against commercially available broad band TEOAEs test	Brass, Watkins, Kemp (1994)	Infants	100	92			
ABR + OAE	Psaramatis, Isakanilcos, Diamantopoulow Douniadakis & Apostolopoulos (2001)	6-14 years	93	91			Signal-to-noise ratio of 3 dB click rate 80 / sec.

TEOAE	Jariengprasent, Soiwanyong, Nasemsuwan, Suapannachart (2002)	< 6 mt and 6 mt	100	100	0	0	Wave form reproducibility criterion of nore than 60% and the overall single-to-noise ratio of more than 3 dB.
TEOAEs and ABR (High risk subject)	Jariengprasent, Soiwanyong, Nasemsuwan, Suapannachart (2002)	6 days- 7 years	94.9	96	5	5	Wave form reproducibility criterion of more than 60% and the overall single-to-noise ratio of more than 3 dB.
TEOAEs and ABR	Jariengprasent, Soiwanyong, Nasemsuwan, Suapannachart (2002)	6 days- 7 years	95.9	98.2	8	8	Wave form reproducibility criterion of more than 60% and the overall single-to-noise ratio of more than 3 dB.

The result of these studies shows that physiological methods have a much higher sensitivity and specificity, compared to behavioural measures. Physiological measures enable the clinician to assess neonates and younger infants in ways that cannot be done by behavioural audiometry. This is on account of them not requiring the child's co-operation and enabling testing of each individual ear. They also directly measure the physiological integrity at the lower portions of the auditory system.

Physiological test are often useful even with children who can be tested with behavioral methods, because they serve as a cross check on the behavioral results (Jerger and Hayes, 1976, cited in Gelfand, 1997 ; ASHA, 1991, cited in Gelfand, 1997) and also provide additional differential diagnostic information.

Through the behavioural tests have relatively lower sensitivity and specificity they too do identify a large number of individuals having a hearing problem. This is especially true with pure-tone audiometry. Thus to detect the presence or absence of a hearing problem, it would be best to use a combination of a physiological measure and a behavioural measure.

METHOD

The objective of the study was to determine the specificity of the high risk register (HRR) developed by Anitha (2001).

The study was done in the following two stages:

1. Stage-I involved administering the HRR on parents.
2. Stage-II involved the audiological testing of children's in the age range of 3 to 5 years.

Stage I: Administering the HRR on Parents

Materials used:

The study was done using two of the questionnaires developed by Anitha (2001), for non-medical professionals. The two questionnaires consisted of high risk indicators of hearing loss for children in the age groups of 0 to 28 days and 29 days to 3 years. Each questionnaire included questions regarding pre-, peri and postnatal high risk indicators of hearing impaired. Details of the HRR are given in Appendix- A and B.

Subjects :

The questions were administered on each parent / caregiver (including the grandmothers and aunts) of 320 children in the age range of 3 to 5 years who knew

the early childhood history of the child. All the 320 children did not have any history of an acquired hearing loss or any otological abnormalities after the age of three years and medical conditions (Eg. Cold, Cough) during the time of testing.

Procedure :

A face-to-face interview was conducted for subject. The HRR were administered to 288 mothers, 15 fathers and 17 caregivers (12 grandmothers and 5 aunts). The interview was conducted in the subject's native language or English. The respondents were instructed to indicate as to whether the risk factor occurred between birth to 28 days or 29 days to 3 years.

Response recording:

The risk factors and the age at which it occurred were noted by the investigator. The responses obtained were analyzed and the children were categorized as either at risk or not at risk as per the criteria suggested by Anitha (2001). The child was considered at risk if they failed on even one question.

Stage II: Audiological testing of children

Subjects :

Children in the age range of 3 to 5 years, who met the criteria mentioned in Stage I were subjected to audiological tests. The audiological tests were carried out to confirm the presence / absence of a healing loss.

Materials Used:

1. An otoscope was used to rule out the presence of any external ear or tympanic membrane problem.
2. Interacoustic MT-10 was utilized for pure tone screening and immittance screening.

Procedure :

Initially an otoscopic examination was carried out on each child before the audiological test to rule out any otological abnormalities such as wax in the ear. The ear was cleaned using cotton buds before administration of the audiological tests. Only children with no evidence of wax or any other external auditory problems were subjected to further audiological evaluation.

The hearing ability was tested in a quiet environment using a portable audiometer (Interacoustic MT-10). Both the ears were screened at 500 KHz, 1 kHz, 2 kHz, and 4 kHz at 30 dBHL. Biological calibration was done before testing the children. Prior to testing at the specified intensity, training was given to each child at of 50 dBHL. The subject's were instructed to indicate either by finger raising or through verbal response that they heard the stimuli.

Tympanometry and reflex were measured at 110 dB using the same instrument (Interacoustic MT-10). During this test the subjects were instructed to sit quietly.

Response recorded :

Response were recorded in terms of pass or fail at each frequency (500 Hz, 1 kHz, 2 kHz and 4 kHz) at the specified intensity during the pure tone testing. The tympanogram type (A type, As type, Ad type, B type and C type) and the presence or absences of reflexes at 110 dB were noted.

The results of the screening audiological testing (pure tone and Immittance) and HRR were tabulated. The specificity of the HRR was computed using the formula:

$$\text{Specificity} = \frac{\text{True negative}}{\text{True negative} + \text{False positive}} \times 100\%$$

Similarly sensitivity of the HRR was computed using the formula.

$$\text{Sensitivity} = \frac{\text{True positive}}{\text{True positive} + \text{False negative}} \times 100\%$$

RESULTS AND DISCUSSION

Parents and caregivers of 320 children in the age range of 3 to 5 years were administered the questionnaire developed by Anitha (2001), for non-medical professional. Information regarding the presence of risk factors in the children as well as any other relevant information were recorded. The number of children having a particular risk factor was noted and this was converted into percentage. Using the data collected, the sensitivity and specificity of the HRR has been established.

Of the 320 subjects evaluated in the study, it was found that 250 children passed the HRR while 70 of them failed it (Table-IV). These children failed on one or more of the risk factors.

Table-IV : Number of children who passed and failed the HRR and audiological hearing screening.

	HRR	Audiological Hearing Screening
Pass	250	247
Fail	70	5

Specificity of the HRR :

Of the 250 children who passed the HRR, 247 passed both the audiological hearing screening tests (i.e. pure tone and immittance). Only 3 of these children

failed both the screening tests. Five of the 70 children who failed the HRR also failed both the hearing screening tests. Using this information, the specificity of the test was found to be 79%.

The following formula was used for the calculation of the specificity of the HRR :

$$\text{Specificity} = \frac{\text{True negative}}{\text{True negative + False positive}} \text{X } 100\%$$

(True negative = 247 children who passed the HRR and the audiological hearing screening.

False positive = 65 children who failed the HRR and passed the audiological hearing screening.)

Thus, the result indicates that out of every 100 normal hearing children who are administered the HRR, 79 would be identified as not having a problem. However, 21 would wrongly be identified as having a hearing problem.

Risk factors failed in the false positive subject

Table-V : Number and the percentage of false positives subjects who failed **the HRR but** passed auduiological hearing screening.

SI. No.	High Risk Register	N	% of occurrence
1.	Maternal illness during pregnancy.	35	53.8%.
2.	Drug in taken by mother during pregnancy for illness.	35	53.8%.
3.	Consanguinity.	19	29.2%.
4.	Family history of hearing loss.	6	**9.2%.**
5.	Delayed birth cry.	6	**9.2%.**
6.	Premature birth (< 37 weeks from last menstrual period).	3	**4.6%.**
7.	Neonatal jaundice	3	**4.6%.**
8.	Drug given for illness to the child	2	**3.0%**
9.	Low birth weight (<1.5 kg)	1	**1.5%**

N = Number of Subjects.

Table - V indicates the risk factor failed by the false positive children. It is evident from this table that "maternal illness during pregnancy" and "drug intake by mother during pregnancy for illness" had the highest percentage followed by "consanguinity". Low birth weight had the least percentage.

Maternal illness during pregnancy and drugs taken by the mother during pregnancy where the main two factors which reduced the specificity of the test.

By eliminating these two risk factors the specificity of the HRR could have been increased. However, these two factors have been found to be important in identifying the true positives. This is evident from the findings of the present study as well as that of Anitha (2001). Hence, elimination of these two risk factors is not recommended as it would compromise the sensitivity of the HRR.

Sensitivity of the HRR :

Among the 320 children who were tested, eight of them failed the audiological tests. The three children who passed the HRR and failed only in the audiological hearing screening tests did so in both the puretone and immittance tests. The presence of a "B" type tympanogram indicated the evidence of a middle ear problem which was probably acquired later in life and not detected by the parents.

Five children failed the HRR and also failed both the audiological hearing screening tests [i.e. pure tone and immittance test]. The presence of a "B" type tympanogram indicated the presence of a middle ear problem. Two of these children had a history of intermittent ear discharge during early childhood.

Thus, the sensitivity of the test was calculated and formatted to be 63% using the following formula:

$$\text{Sensitivity} = \frac{\text{True positive}}{\text{True positive + False negative}} \text{X } 100\%$$

(True positive = 5 children who failed the HRR and the audiological hearing screening.

False negative = 3 children who passed the HRR and failed the audiological hearing screening.)

Thus, the result indicates that by using the HRR, out of every hundred children who have a hearing loss, 63 of them would be identified while 37 of them would not be identified.

The three false positives subjects were found to have a "B" type tympanogram with reflexes absent. They failed the screening test at only one or two frequencies. All three subjects pass the screening when the intensity of the pure tones was increased from 30 dB to 40 dB. These children probably had a mild conductive hearing loss. Their problem was so minimal that the parents did not suspect the presence of any hearing problem. Further, these children had no deviant speech and language. If these three false positives subjects are eliminated, since it is highly likely that they acquires the problem later in life the sensitivity of the HRR would be much higher (100%). None of the false positive children had indication of a sensory neural hearing loss. Thus, it can be concluded that the

HRR was able to identify all children who had a significant hearing loss that would possibly effect their speech and language development.

Risk factors failed in the true positives subjects:

Table-VI : Number and the percentage of true positives who failed on a particular risk factor.

SI. No	High Risk Factors	N	% of occurrence
1.	Drug given for illness to the child	3	60%
2.	Maternal illness during pregnancy	2	40%
3.	Drug intake by mother during pregnancy for illness	2	40%
4.	Consanguinity	2	40%
5.	Persistent OME	2	40%
6.	Neonatal jaundice	1	20%

N - Number of Subjects

Table-VI indicates the risk factors failed by the five true positives subjects. Three of these subjects failed one or more than one risk factors. It is evident from table-VI, that drug given for illness to the child had the highest percentage of occurrence (60%) followed by maternal illness during pregnancy, drug intake by mother during pregnancy, consanguinity, persistent OME. These risk factors had an equal percentage of occurrence (40%). Neonatal jaundice had the least percentage (20%).

Thus, the result of the present study correlated with the results reported by Mahoney and Eichwald (1971). They reported that the sensitivity and specificity of HRR as 65% and 75% respectively.

However the sensitivity of HRR in the present study was found to be low when compare to the results obtained by Anitha (2001). She obtained a sensitivity of 75.5% while in the present study it has been found that the sensitivity and specificity of the HRR is 63% and 79% respectively. The sensitivity of the present study is been reduced due to the presence of three false positive children. These three children probably had an acquired problem later in age, since they had no speech and language problems. By the elimination of the three children the sensitivity of the test in the present study would be 100%. Thus, is can be concluded that the HRR developed by Anitha (2001) can be used without modification as its specificity and sensitivity is high.

SUMMARY AND CONCLUSION

Hearing impairment in infants interferes with normal speech and language development. Koop (1993, cited in Wall, 1995) emphasized that the tragic results of hearing impairment in infancy can be prevented or substantially lessened if intervention is initiated early enough. Thus, early identification, diagnosis and habilitation of hearing loss in young children are critical. The HRR concept is based on the premise that a subgroup of the general population can be selected for further study on the basis of certain criteria that indicate an increased risk for the target disorder or illness (Wall, 1995). It is however, important that the HRR that is used, should have a high sensitivity as well as a high specificity. Anitha (2001) developed a HRR and determined its sensitivity. The present study aimed at establishing the specificity of the HRR developed by her. It also aimed at noting its sensitivity using a general population instead of a group of hearing impaired individuals, as done by Anitha (2001). The study was done using two of the questionnaires developed by Anitha (2001) for non-medical professionals. One was ment for infants in the age group of 0 to 28 days and the other for children in the age group of 29 days to 3 year.

Parents and caregivers of 320 children in the age rage of 3 to 5 years were administered the questionnaire. Each of these children was also evaluated using two audiological screening procedures (i.e., pure tone screening and immittance

audiometry). The audiological tests were carried out to confirm to presence/absence of a hearing loss.

Responses were recorded in terms of pass of fail at each frequency at the specified intensity during the pure tone testing. The type of tympanogram and the presence \ absence of reflexes at 110 dB were noted.

Based on the audiological tests, the true positives, true negatives, false positives and false negatives were determined. Using this information the sensitivity and specificity of the HRR were found to be 63% and 79% respectively. However the sensitivity of HRR in the present study was found to be low when comparing to the results obtained by Anitha (2001). She obtained a sensitivity of 75.5%. The sensitivity of the present study is reduced due to the presence of three false positives children. These three children probably had an acquired problem later in age since they had a "B" type tynpanogram and failed only in one or two pure tone frequencies. They also had no speech and language problems. By the elimination of these three children, the sensitivity of the test in the present study could be raised to 100%.

Thus, it can be concluded that the HRR developed by Anitha (2001) can be used without modification, as its sensitivity and specificity are high. This would be a quick and cost effective method to identify hearing loss early.

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APPENDIX-A

***HRR FOR NON-MEDICAL PERSONS (Birth** - 28 days)*

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

APPENDIX-B

HRR FOR NON-MEDICAL PERSONS (29 days - 3 years)

1. Was there parental or caregiver concern regarding the child's hearing, speech or developmental milestones?
2. Did any one 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 defects 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?