MODIFIED HIGH RISK REGISTER (HRR) FOR PROFESSIONALS AND NON-PROFESSIONALS —FORMULATION AND ITS EFFICACY

Register No.M2k03

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
Manasa Gangothri

Mysore

MAY 2001

Dedicated to

THE ETERNAL POWER

&

AMMA, DADDY, ANAND ANNA,

PRIYA MADHINI, ARUN ANNA & AACHI

CERTIFICATE

This is to certify that the Independent Project entitled : "MODIFIED HIGH RISK REGISTER (HRR) FOR PROFESSIONALS AND NON-PROFESSIONALS — FORMULATION AND ITS EFFICACY" is the bonafide work in part fulfillment for the degree of Master of Science (Speech and Hearing) of the student with Register **No.M2k03.**

Dr. M. Jayaram
Director
All India Institute of
Speech and Hearing
Mysore 570 006.

Mysore
May 2001

CERTIFICATE

This is to certify that this Independent Project entitled : "MODIFIED HIGH RISK REGISTER (HRR) FOR PROFESSIONALS AND NON-PROFESSIONALS — FORMULATION AND ITS EFFICACY " 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.

Asha Yathiraj

Dr. Asha Yathiraj
GUIDE
Reader and HOD
Department of Audiology
All India Institute of
Speech and Hearing
Mysore 570 006.

Mysore
May 2001

DECLARATION

I hereby declare that this Independent Project entitled "MODIFIED HIGH RISK REGISTER (HRR) FOR PROFESSIONALS AND NON-PROFESSIONALS — FORMULATION AND ITS EFFICACY" 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
May 2001.

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INTRODUCTION

Screening is the process of applying certain rapid and simple tests, examinations, or other procedures, to generally large number of persons, that will identify those persons with a high probability of a disorder from those persons who probably do not have the disorder (Northern and Downs, 1991). Since hearing-impairment is relatively invisible, hearing screening tests have been in use for at lest 60 years to identify children for further auditory evaluation. The Joint Committee on Infant Hearing (JCIH) formed in 1969 at that time itself addressed to the use of behavioral observation hearing screening tests that had been developed and described by Downs and Sterritt (1964) and Downs and Hemenway (1969). The JCIH also addressed attention to the development of a high-risk register (HRR) for hearing-impairment that identified five criteria. This five criteria HRR would identify newborn infants at risk for having severe hearing-impairment. The HRR thus formed may miss out the children with late onset hearing loss which may be of mild to severe category. Taking into consideration all these aspects and to increase the sensitivity and

specificity of HRR, the JCIH had made subsequent modifications in 1972, 1982, 1990, 1994 and recently 2000.

Most of the early prevalence of hearing loss estimates were determined from high-risk register studies of newborns. By the early 1990s, however evidence from numerous studies confirmed that use of the high-risk registry as the basis of infant hearing screening programs identified only 50% of infants with significant hearing loss (Elssman, Matkin and Sabo, 1987; Mauk, White, Montenson and Behrens, 1991; Pappas, 1983). Mahoney and Eichwald (1971) reported the sensitivity and specificity of HRR as 65 percent and 75 percent respectively. Though HRR has such sensitivity and specificity rates, JCIH specifies two purposes of risk indicators that often are associated with infant and childhood hearing loss. They are : (a) It help identify infants who should receive audiologic evaluation and who live in geographic locations (eg. developing nations, remote areas) where universal hearing screening is not yet available. (2) since normal hearing at birth does not pr eclude delayed onset or acquired hearing loss, risk indicators help identify infants who should receive on-going audiologic and medical monitoring and surveillance.

The need for and importance of early identification has been documented extensively (Calucci, 1999; Yoshinaga-Itano, 1998; Iskowitz, 1996; Ramey, 1996). The earlier the hearing loss is identified in the child, the easier it would be to bridge the gap between normal hearing and hearing-impaired children. One of the primary benefits of early identification being, early intervention and therefore early mainstreaming can be achieved by HRR. Thus, HRR has its own advantages.

Advantages of HRR over Universal Newborn Hearing Screening (UNHS)

In western countries, the current policy is of UNHS which uses a battery of tests including otoacoustic emissions and brainstem evoked response audiometry because of their greater sensitivity.

In India, universal screening in the form done in the west, is difficult to implement due to lack of healthcare facilities, trained professionals, economical realities and the sheer number of children

who would have to be screened by trained individuals. The HRR has its advantages as follows:

(1) It can be administered by any person like doctors, nurses, anganwadi workers etc. unlike other hearing screening procedures which needs technical and man-power expertise. Since a high risk register may be administered by medical or non-medical persons, it should be ensured that the terminologies used be easily understood by any individual who uses it.

(2) Time, effort and cost for administering the HRR is very low.

(3) The cost and time for training the health care worker to administer the HRR is minimum. Yathiraj, Sameer and Jayaram (2001) reported that the average cost of administering 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 or health workers. In a developing country like India where finance is a problem this would be the usual choice of testing.

(4) Certain risk indicators of late onset hearing loss can only be identified by HRR.

In view of all the above **the** present study aims at formulating the HRR specifically for children in the age groups of birth-28days and 29 days - 3 years that could be administered by professionals and non-professionals. The sensitivity of the HRRs will be checked by administering them on the parents or caregivers of children who have confirmed hearing loss.

Need for the Study

A HRR has to be modified many times in order to increase its sensitivity and specificity. It has been reported by professionals that the existing HRR in India is time consuming and also a lot of factors which may not be contributing to hearing loss were included. This leads to unnecessary over referral. Hence, there is a need to modify it. Also HRR is a tool which is time effective and cost effective unlike other tests used for UNHS. In a country like India where finance is a problem and the population is high, this is an essential factor,

The HRR that has been developed in western countries cannot be directly used for Indian population because some of the factors like

consanguinity which is more prevalent in India was not included in their HRR. The 1990 JC1H also encouraged the continued study and critical evaluation of known and unknown risk factors and recognized that, because the recommended protocols might not be appropriate for all institutions, modifications might be appropriate to meet the specific needs of any given facility. The above information indicate the need for formulating a HRR for Indian population which is time effective and sensitive.

Many medical and paramedical professionals reported that the HRR with specific medical conditions will be difficult for the grass root level workers to use. Also, it has been reported by Yathiraj, Sameer and Jayaram (2001) that the cost effectiveness of the HRR could be increased if the grass root level workers such as anganwadi workers, health workers, social workers and CBR workers can administer it. This reveals the need for formulating two separate high-risk registers for medical and non-medical persons.

The JCIH (1990, 1994 and 2000) recommended the high-risk factors for use with neonates from birth through 28 days and neonates

or infants from 29 days through 2 years. The risk indicators for infants or neonates of 29 days through 2 years place an infant at risk for progressive or delayed-onset hearing loss who has passed the birth screen should, nonetheless, receive audiologic monitoring every 6 months until age 3 years.

The JCIH recommends ongoing audiologic and medical monitoring of infants with unilateral, mild, or chronic conductive hearing loss. Infants and children with mild or unilateral hearing loss may also experience adverse speech, language, and communication skill development, as well as difficulties with social, emotional and educational development (Bess, Dodd-Murphy and Parker, 1998; Blair, Petterson and Viehweg, 1985; Davis, Elfenbein, Schum and Bentler, 1986). Infants with unilateral hearing loss are at risk for progressive and/or bilateral hearing loss (Brookhouser, Worthington and Kelly, 1994). Infants with frequent episodes of OME also require additional vigilance to address the potential adverse effects of fluctuating conductive hearing loss associated with persistent or recurrent OME (Friel-Patti and Finitzo, 1990; Gravel and Wallace, 1992; Jerger,

Jerger, Alford and Abrarns, 1983). This reveals the need for inclusion of late onset risk indicators.

Thus, it is evident that the HRR that is being currently being used by speech and hearing professionals in India, be modified and its sensitivity be checked.

REVIEW OF LITERATURE

Hearing loss is caused by various factors. High risk factors (HRFs) are those factors which may cause an increased likelihood of the development of a disorder or those factors which may indicate an increased likelihood of its presence. HRFs can be either causal (eg. virus or a teratogenic drug) or associative which merely indicate a risk factor (eg. congenital anomalies) or both. The high risk factors reported in literature are discussed in the following section:

Consanguinity

Human beings are all remotely related. In fact, it can be mathematically shown that most people are remote cousins. Marriage between two closely related persons or consanguinous matings, though a taboo in many places, has been an accepted system in certain communities and regions. It is commonly practiced among Muslims and in many communities in South India.

Shaw Nawaz (1974) surveyed the whole population of 4000 individuals in a village in Karnataka in South India. The inhabitants of the village, Gargeshwari were predominantly Muslims. It also had a minority Hindu community, in which system of consanguinous marriage was not commonly observed. In the Muslim community however, it was a rule more than just a custom. The whole Muslim population of Gargeshwari evolved out of two families. Almost everyone in that village was a first cousin to every body. Shah Nawaz (1974) found a greater incidence of speech and hearing problems in this community. The incidence figures were reported to be much higher than both the general incidence and incidence figures among communities and in regions observing this system of marriage (Ratna, 1980). The incidence figures of consanguinous marriages in certain parts of the country and among Muslims is reported to be very high. This contrasts with only 0.5 percent of all marriages in most of the western countries (Whittinghall, 1965).

Deafness as a phenotype of human should be considered with caution because of its multiple etiological factors. In a study of all deaf children born in Belgium Anterwp, it was found that nearly forty-five

of a total of 111 were of recessively transmitted types. The proportion of first cousin marriages among their parents was too high to go with a recessive gene frequency (Whittinghall, 1965). Most traits are recessive and are therefore most likely to appear in consanguinous offsprings. If non-relatives have a 11/2 percent chance of their child having an abnormality, consanguinous parents would have a 3 percent chance of their child being affected.

Consanguinity has been considered as a HRF in an unpublished dissertation by Ashok Kumar in 1981. This was not considered in any of the western registers. Taking into consideration all the above studies, consanguinity as a HRF should be given due importance especially in India

Hereditary Hearing Loss

Hereditary deafness is a fairly common disease entity, occurring between 1 in 2000 and 1 in 6000 live births. A high percentage of congenital deafness is hereditary, according to Konigsmark (1972). Like other traits, hearing loss is transmitted either by the ordinary

paired chromosomes (autosomal) or by the sex chromosomes (X-linked). Inherited deafness can take many forms, including those which are not congenital. The pattern of inheritance varies according to the type and extent of gene(s) involvement. Konigsmark (1969) reported that there are over 60 types of hereditary hearing loss which can be separated from one another by the type of hearing loss, age of onset, severity of hearing loss, mode of transmission and the abnormalities caused in other systems by the abnormal gene or pair of genes. There are 12 types of hereditary hearing loss with no associated abnormalities. Studies have shown that about 40 percent of profound childhood deafness is autosomal recessive in origin, 10 percent is by dominant transmission and about 3 percent is caused by a sex-linked gene (Fraser, 1964).

In autosomal dominant inheritance, typically for each pregnancy, the chances for the child to be deaf are about 50 percent. Males and females are equally affected and the trait is carried vertically from one generation to the next. Some of these dominant disease types are well known, e.g. Wardenberg's syndrome, Treacher Collins syndrome, Apert syndrome and Crouzon syndrome. Some other forms

are characterized by the late onset of the trait in the offspring. Frequently these children are born with normal or near normal hearing. Without a HRR, they are liable to be lost during screening programs (Gerber, 1977).

In the X-linked inheritance, the mother carries the gene for hearing loss on one of her two X-chromosomes. Since X-linked traits are often recessive, the matching gene on the other X-chromosome usually allows for the expression of normal hearing. Thus a daughter would escape whereas each son has a 50 percent chance of inheriting the loss. An affected male can transmit the X-linked trait for hearing loss to all of his daughters, making them carriers, but, none to his sons because he can contribute only Y-chromosomes to them (Mencher, Gerber, and McCombe, 1997).

Apart from the gene transmission, chromosome abnormalities like trisomy, are known to cause hearing-impairment. In trisomy, an additional chromosome is found within a pair. Such conditions are not necessary hereditary. Multiple anomalies, malformed ears and hearing loss are frequently seen in such conditions.

Maki-Torkko, Lindholm, Vayrynen, Leisti and Sorri (1998) reported that among the generally known risk indicators, family history seems to be the most common which was present in 15 percent in the older cohort and in 24 percent in the younger cohort. Sutton and Rowe (1997) studying the risk factors for sensori-neural hearing-impairment in the Oxford region found a positive family history in 26 percent of cases.

All the above informations reveals that it is very essential to determine the family history correctly. Due to its high incidence it is perhaps the most crucial factor in a HRR. Thus factor has been included in HRRs of Joint Committee on Infant Hearing 1972, 1982, 1990, 1994 and 2000. It has also been given importance in Halifax Project (1977), Utah High Risk Program (1967) and Santa Barbara Plan (1978).

Maternal Illnesses

In general, the fetus is well protected and insulated against adverse physical, chemical and biological insults. Yet, a few maternal conditions, some peculiar to the mother may jeopardize the fetal safety (Singh, 1979). Though they may not directly enhance the risk of the infant developing a congenital condition like deafness, they may however, led to conditions which are high risk themselves. Also, drugs given to the mothers for their management may also adversely increase the risk of the infant developing a congenital condition.

Toxoplasmosis : The most common example of congenital deafness caused by protozoal infection is toxoplasmosis. Children affected by toxoplasmo gondii, which is acquired transplacentally are usually multiply involved. It has even been said that 10 to 20 percent of profound childhood deafness is caused by this infection (Theissing and Kittel, 1962). Stein and Boyer (1994) suggested the number may be even higher than that. The disease may occur more often than recognized, but the number and extent of hearing losses associated with it has yet to be confirmed. Suffice it to say, toxoplasmosis does cause

hearing loss and other neonatal disorders. The disease apparently develops as calcium deposits on the stria vascularis and the spiral ligament. The resultant hearing loss is therefore slowly progressive. The hearing-impairment may begin in early childhood as a mild loss and will advance toward profound deafness with time.

Rubella : Rubella embryopathy is a most important prenatally acquired cause of profound childhood deafness. Infants with congenital rubella virus infections have a variety of defects of varying severity depending on the embryonic stage during which the infection occurred. Hearing-impairment can result from fetal rubella not only during the first trimester of pregnancy but also in the second and even third trimester (Bordley, Brookhouser, Hardy, 1968). Diagnostic features include transient neonatal manifestations of low birth weight, hepatosplenomegaly, purpura, bulging anterior suture, corneal clouding, and jaundice. Anemia, pneumonia, meningitis, and encephalitis may develop. Major associated problems include hearing loss (50 percent), heart disease (50 percent), cataract or glaucoma (40 percent), and psychomotor and mental retardation (40 percent) (Cooper, Ziring and Ockerse, 1969).

Rubella deafness is commonly sensory in nature, often severe to profound in degree. Anvar, Mencher and Keet (1984) reported essentially flat audiograms, saucer-shaped audiograms with the greatest loss between 500 and 200 Hz and audiograms gently sloping from low to high frequencies. Thus, variation in audiometric configuration is common. Evidence of central auditory deafness due to central nervous system lesion has also been reported. The pathology of rubella deafness includes a variety of inner ear abnormalities and middle ear and external ear anomalies. Rubella virus is generally acquired by air borne distribution and enters the maternal respiratory tract (Vernon and Klein, 1982). The gestational age of the embryo or fetus is the critical factoring determining the outcome.

CytomegaloviTus : Cytomegalovirus (CMV) is far more common today than prenatal rubella. It is estimated that 2-3 percent of pregnant women excrete this virus and that it may be found in 1-1.5 percent of newborns. The disease resulting from CMV is silent in an many as 98 percent of the infants who have it (Marx, 1975). Furthermore, Marx estimated that by the age of 30 years, 80 percent of the population of

the United States at sometime will have carried a live CMV. Thus, die disease is extremely common and apparently easily transmitted. Transmission may occur during pregnancy by the transplacental route. Intrapartum infection is possible during the passage of the fetus through the birth canal in women with cervical CMV, but this is usually not associated with disease.

Classically, cytomegalic inclusion disease is characterized by enlarged liver and spleen, jaundice, blood and skin abnormalities, a variable involvement of the central nervous system, including cerebral classifications, microcephaly, chorioretinitis, deafness, and psychomotor retardation (Weller and Hanshaw, 1962). There are several variations and combinations of these findings. Overt congenital CMV, which occurs in 2 percent of the cases, is usually fatal (Aballi and Korones, 1963).

According to Dahie, McCollister, Hamner. Reynolds and Stagno (1974), a severe to profound high-frequency progressive sensory hearing loss is often associated with congenital CMV infection. Harris, Ahlfors, Ivarsson, Lernmark and Svanberg (1984) reported that the

composite audiogram could be interpreted as showing three relatively distinct sub-groups of hearing-impairment; subjects with a loss primarily at 8000 Hz, subjects with losses that involve two or more of the higher frequencies, and subjects with the more severe losses which involve all frequencies to some extent.

Herpes Simplex. Virus : Herpes simplex virus (HS V) is one of the most commonly transmitted sexual diseases. According to Northern and Downs (1991), HSV infects the genital tracts of an estimated 20 to 25 percent of the world's population. If the mother is infected, the disease will be transmitted to the baby during the birth process, and it has been suggested that as many as 50 percent of those infected will die. Hearing loss is usually present at birth for these who are infected. As most cases of congenital HSV are also symptomatic of the entire disease process at birth (including rash and other associated sequelae) it is not hard to recognize (Northern and Downs, 1991).

Syphilis : Congenital syphilis is still one of the most important contributors to peri-natal mortality and morbidity in many parts of the world. A seriously involved neonate will exhibit a rash over the entire

body, anemia, dental anomalies, central nervous system deficits, cardiac anomalies and hearing loss. Congenital syphilis may demonstrate a multitude of central nervous system abnormalities including vestibular dysfunction, sensori-neural hearing loss and occasionally, aortic valvulitis (Northern and Downs, 1991). Auditory impairment may not be present at birth. Onset of hearing loss is generally in early childhood, usually sudden bilaterally symmetrical hearing loss causing severe to profound impairment. Sensori-neural hearing-impairment of slow progressive nature is seen. Pattern of deafness shows variation dependent upon time of onset and rapidity of progression.

Other Illness

Toxemia of pregnancy is a disorder in which the mother presents with high blood pressure, edema or protein in urine. Babies of such mothers are at much risk for hearing-impairment. Similarly, babies born to diabetic mothers due to their large size, are often delivered pre-term or by caesarian section, and are susceptible to trauma, bruises, birth asphyxia and run a greater risk of hyaline

membrane disease. Incidence of congenital malformations in diabetic mothers is almost double the general incidence (Singh, 1979). Malaria, which is quite virulent in India, can indirectly indicate a risk because of the ototoxic drugs given for malaria.

Sensori-neural deafness has been reported occasionally in infants whose mothers had chickenpox during pregnancy. However, Seigel (1973) in a controlled cohort study found deafness in one control group, hence it is uncertain that chickenpox causes hearing loss.

In maternal illnesses, rubella has been considered as a HRF in registers like Joint Committee on Infant Hearing (1972), Colorado project (1972), Halifax project (1977) and Utah high risk program (1967). Only in later years all the TORCHES infections have been considered in Santa Barbara Plan (1978), position statements of 1982, 1990, 1994 and 2000 given by JCIH

Ototoxicity

The word "ototoxicity" means literally ear poisoning. More specifically, it is defined as the partial or total reduction of cochleo-vestibular function as a result of chemical interaction with drugs (commonly therapeutic agents) or other toxic substances or procedures. Although over 200 ototoxic substances have been described, there is a small group of regularly used agents consistently associated with toxicity. There are seven groups of substances and chemicals that are known to affect hearing and/or the vestibular system (Mencher, Gerber and McCombe, 1997). They are:

1. Antibiotics
2. Loop diuretics
3. Analgesics and antipyretics
4. Anti-malarial agents
5. Anti-neoplastic or chemotherapeutic agents.
6. Miscellaneous drugs
    a) Anti-heparinizing agents
    b) Anti-convulsive drugs
    c) Beta-blocking agents
7. Chemicals and general toxins (mercury, lead, alcohol, etc).

Among the currently used ototoxic drags are the aminoglycoside antibiotics such as amikacin, gentamicin, kanamycin, neomycin, netilmicin, streptomycin and tobramycin. Jung and Nissen (1991) noted cochlear damage to be greatest with amikacin and gentamicin, with an incidence of about 10 percent, followed by tobramycin and netilmicin (2 percent). Vestibulo toxicity is less common with an incidence of about 3.5 percent for amikacin and gentamicin and 1.5 percent for netilmicin. Probably the best known of all ototoxic drugs, or at least the one that has been studied the longest, is streptomycin. It was the first aminoglycoside to be discovered. Streptomycin has been and still is a drug of choice for the treatment of tuberculosis. Bergstrom and Thompson (1984) observed that vestibular damage due to streptomycin is more frequent than hearing loss. Szekely and Draskovich (1965) reported that when they examined eighty-four children between the ages of six and eight who had received streptomycin in infancy, forty-three had measurable high-frequency losses, although only fourteen showed evidence of residual vestibular damage.

Ingestion of ototoxic drugs by pregnant women can result in a multitude of congenital abnormalities, including hearing loss, from passage of the drugs across the placenta (Siegel and McCracken, 1981). Apparently, renal failure, concomitant use of diuretics such as ethacrynic acid and furosemide and a prolonged course of drug therapy are the most important factors in the development of fetal ototoxicity. The evaluation of aminoglycoside ototoxicity in infants is a complex problem, since these babies are receiving medical therapy for severe problems, including systemic infection, that may accompany low birth weight, jaundice or other health disorders that are of themselves associated with deafness.

An exhaustive search by Leach (1962) brought to light a total of nine antibiotics, most of them aminoglycosides, which were 'known or suspected to be toxic to the labyrinth to a greater or lesser degree when administered parenterally. These were : dihydrostreptomycin, framyeetin, kanamycin. neomycin. polymyxin B, ristocetin, streptomycin, vancomycin and viomycin. To these must be added gentamycin. Streptomycin may also cross the placental barrier insufficient concentration to damage the fetal ear, and Robinson and

Cambon (1964) found two instances (out of a total 300 hearing-impaired pre-school children) in which the loss could be clearly attributed to treatment of mothers with this drug during pregnancy. Dihydrostreptomycin may also cross the placenta (Kern, 1962). The most cochleotoxic of the basic streptomyces antibiotics are kanamycin and neomycin and many reports have appeared in the international literature about the toxic effects of both these drugs.

The incidence of ototoxicity in general figures or for specific drugs has not been accurately established. Thompson and Northern (1981) identify a number of risk factors that may enhance the potential risk of ototoxicity, including increased drug serum level and decreased renal function. The use of more than one ototoxic drug simultaneously or in increased daily doses or for an extended period of time also increased the risk of ototoxicity. Other factors that enhance ototoxicity are age, health, heredity, concurrent noise exposure or in the presence of pre-existing problems including severe visual impairment or blindness, or drugs administered in the presence of ear symptoms such as tinnitus, hearing loss and/or dizziness.

Ototoxicity as a HRF has been mentioned in 1990 and 1994 position statement and not in any other high risk programs.

Maternal Irradiation During Pregnancy

The effects of maternal irradiation on the fetus have been reported extensively. But the effects on the hearing of fetus is less and highly variable. MacMahon (1962) compared the children of 70000 mothers who had received pelvic X-rays during pregnancy with the children of mothers who had not been X-rayed. He concluded that the human tetus was far more vulnerable to miscarriage, malformations and cancer from X-rays than anyone had previously believed possible.

Department of Health, Education and Welfare (1970) had reported that the unborn face greater risk of radiation damage than adults receiving the same amount of exposure. The stage of pregnancy determines in large measure the type of fetal damage. During the first trimester risks of accidental miscarriage, congenital malformation and brain damage predominate. From the ninth day through the sixth week of pregnancy, organogenesis - the period of organ and limb

development occurs. The greatest radiation induced deformities can be produced because of the specialized rapid development and division of cells and tissues. Ear, nose, eye and structural brain deformities can result.

Taking into consideration all the detrimental effects of maternal irradiation during pregnancy, Morgan (1980) had fought for years to pass a recommendation that women in the child bearing age should not be given X-rays in the pelvic and abdominal region except during emergency situations.

Manifestations of in-utero irradiation in humans are microcephaly, mental retardation and other CNS defects and growth retardation (Mole, 1991). Microcephaly is the most common malformation reported after exposure to a high dose of radiation during pregnancy. Similar results were reported by Dekabon (1968) who revealed that mental retardation and microcephaly occur in fetuses exposed to X-ray especially during the 7^{th} to 15^{th} week of gestation. He did not report of deafness in his study. Exposure of the mother to X-rays has not been mentioned in any of the HRRs given by JC1H.

Prematurity and Low Birth Weight (LBW):

The weight of the newborn is dependent upon the quantity of subcutaneous fat which is accumulated mostly during the last trimester of pregnancy (Achar, 1969). Usually, babies with a weight of less than 2500 grams irrespective of the period of gestation are classified as LBW (Singh, 1979). Prematurity and LWB are usually seen together, particularly in infants weighting 1500 gms or less and are associated with greater mortality and morbidity incidence (Lansford, 1977).

There is a higher incidence of both neurological and mental handicaps in LBW babies. The incidence of congenital anomalies in small-for-dates is 10 to 20 times higher than in normal population (Singh, 1979). Funaki (1978) reported a higher incidence of hearing loss in LBW babies than in premature babies and concluded that physical under development of the hearing organ may be a crucial factor.

Earlier classifications of prematurity were based on the WHO 1948 definition, that any infant with a birth weight of 2500 grams or

less should be termed premature. In 1961, the Expert Committee on Maternal and Child Health appointed by WHO recommended that the concept of prematurity in the earlier definition should give way to that of "LBW". Since then the practice has developed of differentiating between children born before 37 full weeks of gestation (i.e. preterm) and those born at 37 weeks or more but weighing less than 2500 grams (i.e. small-for-dates) (Behrman, 1975).

About 30 percent to 40 percent of babies born in India are LBW as opposed to 6 to 7 percent in the western countries (Singh, 1979). Babies weighing less than 2000 grams, who are more vulnerable to disease and distress, account for nearly 10 percent of total babies born in India (Singh, 1969). 8 to 10 percent of newborns are born preterm in India as compared to 5 to 7 percent in west (Singh, 1979). Prematurity with or without other factors, carries a higher risk for central nervous system problems (Catlin, 1977). The incidence of congenital anomalies is twice as much as in term babies (Singh, 1979). Campanelli, Pollack and Henner (1958) found that 16 percent of their group of fourty-four premature infants showed moderate to severe bilateral sensori-neural losses.

Feinmesser and Tell (1976) noted that of the twenty-six infants they found to have a hearing impairment, one would not have been classified as LBW with the 1500 grams criterion. However, the National Joint Committee has reaffirmed 1500 grams as the criterion opining that redefinition would lead to as unduly large follow-up population.

McDonald (1967) noted that there was a higher incidence of deafness in premature children when it occurred along with other conditions. These included a history of jaundice or generalized edema and convulsions, or who had been given oxygen for more than 10 days or who had been given streptomycin. Matkin (1968), at Northwestern University, examined the records of 900 deaf children and found that 128 had severe high-frequency losses which must be considered potential sequelae of prematurity, anoxia, postnatal respiratory distress and Rh-incompatibility.

Galambos and Galambos (1975) recorded brainstem auditory-evoked responses in 24 infants ranging in age from 6 weeks premature to term. The data reveal that for these premature infants wave V

increases in latency at a rate of about 0.4 msec, with each 10 dB attenuation in stimulus level . Further, this latency at a given stimulus level decreases systematically with increasing gestational age. Cox, Hack and Metz (1984) reported that the ABR abnormalities seen in very low birth infants were localized to the peripheral auditory structures although some were associated with the central auditory study.

Vernon (1967) examined the medical records of 1468 children in a retrospective study. Of these children, 17 percent had birth weights less than 2500 grams and more than two-thirds of the premature group were multiply handicapped. In contrast, McDonald (1967) surveyed 1128 children with birth weights of 1800 grams or less and found only 19 children (1.7 percent) with moderate or severe sensori-neural deafness. Whereas Harper and Weiner (1965) found impaired hearing in 3.6 percent of their children of LBW.

Clarke and Conry (1974) carried out audiometric evaluations on a population of 104 LBW children and 123 controls. In the LBW group, children (3.3 percent) had a bilateral loss and 5 (2.5 percent) had

a unilateral sensori-neural high frequency hearing loss. There were 13 (6.4 percent) cases of conductive loss among the controls.

The above studies shows the importance of considering prematurity and low birth weight as HRFs in the registers. Also most of the HRRs have recommended LBW less than 1500 grams as a HRF. But prematurity is not mentioned as a separate HRFs in any of the HRRs given by JCIH. It has been mentioned by Ashok Kumar (1981), Halpern, Hosford-Dunn and Malachowski (1987).

Birth Asphyxia and Cyanosis

In utero the placenta serves to transfer nutrition and oxygen to the fetus. After the separation from the mother, the baby must breathe immediately or must be made to breathe because within two minutes of tying the cord the child's arterial oxygen tension reduces drastically resulting in asphyxia (Singh, 1979). Asphyxia occurs when the process of respiration fails. The infant is particularly vulnerable to asphyxia during labor and delivery and immediately after birth. One frequent cause of birth asphyxia is the compression of the umbilical cord

between the pressing part and the pelvic tissues during the process of labor and delivery. It may also be caused by inadequate maternal blood oxygenization, low arterial blood pressure, inadequate relaxation of the uterus to permit placental filling, inadequate attachment of the placenta or placental inadequacy as in toxemia and post-maturity (Lansford, 1977).

Birth asphyxia or hypoxia is the leading cause of peril-natal death and permanent damage to CNS. Cerebral depression or seizures may occur due to cerebral edema, anoxic brain damage and intra-cranial bleeding. Birth asphyxia is the commonest medical emergency among newborns and is one of the leading causes of neonatal mortality in India (Singh, 1979).

If the baby stays in an hypoxic state for long, its heart slows, blood pressure falls and then it lapses into terminal apnea. The baby appears pale rather than blue (Singh, 1979). This condition is known as cyanosis. The hypoxic infant has long been known to be at risk for neurosensory deficits including hearing-impairment. Reported

incidence of neuro-sensory deficits ranges from 0.9 percent to 10 percent (Marcus, 1976).

Limited evidence indicated that intra-partum and post-partum asphyxia and anoxia, as well as trauma, may be a predisposing factor to haemorrhage. Chessells and Wigglesworth (1971) have shown, in addition, that the hemostatic mechanism is unfavourably influenced by birth asphyxia. It is also possible that intra-partum asphyxia and anoxia may lead to hearing loss through toxic damage to the cochlea nuclei, as well as through the production of hemorrhages into the inner ear (Fisch, 1955; Hall, 1964). Such anoxia episodes may continue into the neonatal period and be manifested as cyanotic attacks, forming part of the respiratory distress syndrome. Subsequent deafness in such children damaged by birth injuries and/or anoxia is often accompanied by other evidence of neurological damage of the type of cerebral palsy, optic atrophy or epilepsy.

Scheiner (1980) suggests the use of multiple measures in determining clinically significant asphyxia, including the length, degree, frequency, and severity of the episodes. The occurrence of

asphyxia is commonly related to a number of other medical conditions, so it is difficult to absolutely establish asphyxia as the single cause of deafness. MacDonald (1980) points out that a pre-term infant is likely to experience anoxic episodes than a full-term infant.

The Apgar score taken at 1 minute is an index of asphyxia and shows the need for assisted ventilation. Apgar score of less than 4 has been used as a HRF (Feinmesser and Tell, 1974). Infants with anoxia may also have low Apgar scores at 1 minute and 5 minutes following birth, low arterial pH level, coma, seizures, or the need for resuscitation with oxygen by mask or intubation. D'Souza, McCartney and Nolan (1981) conducted hearing, speech and language studies in 26 children who survived severe peri-natal asphyxia. Although only one child had sensori-neural hearing loss, nearly one-third of the children in their sample had deficits of speech and/or language. Simmons (1980) implicates anoxia as the single high-risk factor that dominates all others in the medical histories of hearing-impaired babies.

Because of its importance it has been considered as a high risk factor in all the registers given by JCIH in 1972, 1982, 1990, 1994 and 2000 position statements.

Hyperbilirubinemia

Jaundice is the commonest abnormal clinical finding during the first week of life. Clinical jaundice manifests at a serum bilirubin level of 4 mg percent. Yellow colouration is first evident in the eyes, on the skin of the face, nasolabial folds and probe tip. As the extent of bilirubin accumulation in the skin increases the trunk becomes yellow first followed by abdomen, extremities, palms and soles. It is said that yellow staining of the trunk indicates a level of 10-15 mg percent and when soles and palms are distinctly yellow stained the accumulation is said to be more than 15 mg percent (Singh, 1979). The extent of colouration however, is in no way related to the level of bilirubin concentration (Behrman, 1975).

Many conditions lead to jaundice. The most common causes of jaundice, in their order of frequency of occurrence in India are

physiologic jaundice (functional immaturity of the liver), pre-maturity, blood group incompatibility between the mother and the fetus (both Rh and ABO group incompatibility) G-6-PD iso-immuno enzyme deficiency, pre and post-natal infections, drugs and breast-milk jaundice (Singh, 1979).

Bilirubin accumulation in the order of 15 mg percent or more is considered potentially neurotoxic depending upon the state of the infant (in pre-mature and LBW babies, a lesser level is indicated). Only few conditions, however, can result in such a high level of bilirubin accumulation like the Hemolytic disease of the new bom due to Rh or blood group incompatibility. Incidentally, ABO incompatibility, though less severe in its effect than the Rh incompatibility is said to be more prevalent than the latter in India. This has been ascribed to the low incidence of Rh negative women in India - only 2 to 7 percent as against 15 percent in white races (Achar, 1969). Rarely infections such as toxoplasmosis, CMV, syphilis, rubella, etc., or other conditions result in such high serum bilirubin accumulation (Behrman, 1975).

The day of onset of jaundice gives an important clue to the possible etiology and differential diagnosis of the conditions (Singh, 1979; Behrman, 1975; Achar, 1969). Onset on the first day is always suggestive of a serious disease process like hemolytic disease which is more likely to cause bilirubin toxicity or encephalopathy. If it appears on the second or the third day it is usually physiologic jaundice but may also be due to hyper-bilirubinemia. Jaundice appearing after the 3rd day and within the first week should suggest septicemias the most likely cause (Behrman, 1975).

If the serum bilirubin reaches a high level (usually around 20 mg% in a full term infant) and exchange transfusion is indicated, which replaces about 50 percent of the baby's blood. Any child whose bilirubin level has risen high enough to have necessitated an exchange transfusion is at-risk of having bilirubin encephalopathy. Clarke and Corny (1979) found that all the known bilirubin levels were high among children with sensori-neural losses.

It has been known for many years that a serumbilirubin level above 20 mg/100 ml may lead to damage of the cochlear nuclei (Fisch,

1955). Support is supplied by the Keaster, Hyman and Harris (1969) study of 405 patients who had hemolytic disease or hyperbilirubinemia as neonates. 17(4.2 %) of those patients were found to have sensori-neural hearing losses which varied from mild to profound and the audiogram also showed hearing losses most marked in the high frequencies.

Because of its common occurrence and serious sequelae most of the HRRs had mentioned hyperbilirabinemia as a HRF. Halifax project (1977), Utah high risk program (1967), Colorado screening project (1972) and Joint Committee on infant hearing (1982, 1990, 1994 and 2000) had mentioned hyperbilirabinemia requiring exchange transfusion as a HRF whereas JCIH (1972) had mentioned only hyperbilirabinemia as a HRF.

Incubation and/or Intensive Care Nursery (ICN) Admission

Babies under treatment in an intensive care nursery (ICN) are known to be at risk for a wide variety of sensory, motor and mental disorders. The results of Galambos, Hicks and Wilson (1982) study

shows that at least 10 percent of the babies discharged from ICN as healthy, hear poorly through one or both ears and that at least 2 percent of them suffer irreversible bilateral sensori-neural hearing losses so severe as to require correction with hearing aids. Jacobson, Seitz, Mencher and Parrott (1981) screened eighty-three ICN graduates. They found that twelve (14%) babies failed the ABR test at 60 dB HL.

Halpern, Hosford-Dunn and Malachowski (1987) reported that most of the items mentioned in a seven-point HRR were significantly associated with hearing loss in 820 ICN graduates who were judged at risk for hearing loss. This seven point HRR includes family history of hearing loss, craniofacial anomalies, asphyxia/low Apgar score, bacterial meningitis, congenital perinatal infection in the TORCH complex, hyperbilirubinemia and birth weight < 1500 gm. They have also reviewed the medical records of all subjects for variables that might be associated with hearing loss. These variables included the HRR items and other descriptors of the ICN stay. The results revealed that craniofacial anomalies and TORCH infections are two of the variables in HRR and length of stay in ICN and gestational age

predicted hearing loss with 98 percent sensitivity and also reduced the initial risk group by one-third.

Stein, Clark and Kraus (1983) reported that for ICN babies, birth weight <1500 gm was the most frequently reported risk factor. Bacterial meningitis, congenital perinatal infection and family history of childhood hearing-impairment were the most frequently reported risk factor. Cone-Wesson, Vohr, Sininger, Widen, Folson, Gorga and Norton (in press) analyzed the prevalence of risk indicators for infants identified with hearing loss. 3134 infants evaluated during their initial birth hospitalization were reevaluated for the presence of hearing loss between 8 and 12 months of age. The majority of these infants were ICN graduates (2847).

Cox (1980) reported that the frequency of serious hearing deficit in ICN population is approximately 2 percent in contrast to approximately 0,2 percent in general population. Swigonski, Shallop, Bull and Lemons (1987) supported previous observations concerning a higher incidence of severe sensori-neural hearing loss in the population of infants who graduate from intensive care nursery setting.

The high incidence of hearing handicaps in ICN babies certainly justifies screening this population of babies. Mauk, White, Mortensen and Behrens (1991) reported that 63 percent of the children would have exhibited at least one risk factor if admission to neonatal intensive care unit had been considered as a risk factor which is otherwise only 50 percent.

There have been reports about the noise levels and their effects on infants admitted in neonatal intensive care unit. Falk and Farmer (1973) analyzed noise from functioning infant incubators and revealed a 74.5 dB noise level with a frequency distribution mainly below 500 Hz and also suggested that incubator noise should be considered as a possible contributing factor in causing hearing loss. Long, Lucey and Philip (1980) recorded 2-hour polygraphic tracings from infants' heart rate, respiratory rate, transcutaneous oxygen tension and intracranial pressure during the routine ICN schedule. They found that sudden loud noises usually caused agitation and crying in the infants, which led to decreases in transcutaneous oxygen tension and an increase in intracranial pressure, as well as increases in heart and respiratory rate.

Kryter (1966) examined damage risk contours which would indicate that incubator noise would not be a potential risk to hearing. He stated, however, that criteria were established with adults and the contours were based on intermittent sound which provides an opportunity for recovery. He further warned that a premature infant who is confined to an incubator for continuous periods may be at additional risk.

Taking into consideration the high incidence of hearing handicaps, the Joint Committee on Infant Hearing Year 2000 Position Statement had included this as a HRF which was not mentioned in any of the previous high risk programs including the one used in AIISH except Santa Barbara Plan, 1978.

Congenital Orofacial Anomalies

Most orofacial anomalies are sufficiently bizarre to be immediately obvious at birth. It does not require a specialist or a questionnaire to determine if an infant is born with one ear completely closed off. Orofacial anomalies frequently incorporate hearing-

impairment among their stigmata (Gerber, 1977). Stool (1975) observed that any child with an unusual appearance should be assumed to have a hearing-impairment until it can be demonstrated otherwise. Sometimes it is limited to the conductive apparatus, as in Treacher Collins syndrome or to cochlear, as in Wardenberg syndrome and sometimes to both, as in Hurler's syndrome. Other congenital defects are often seen accompanying deafness (Gerber, 1977).

Brown (1967) reported that Pendred's syndrome is the most common syndrome which included deafness and various enzyme defects are also associated with deafness. Fraser (1976) found that Usher's syndrome, which is characterized by progressive blindness associated with retinitis pigmentosa with congenital severe deafness is the second most commonly occurring identifiable syndrome. Similarly, cardiac defects (eg. Jarvell and Lange Nelson syndromes), skeletal anomalies (eg. Apert's), pigmentary abnormalities (like partial albinism) are associated with deafness.

Deformities of the lip and palate are among the most common major congenital malformations, occurring once in 900 newborns

(Northern and Downs, 1991). A substantial number of articles have been published concerning the otologic and audiologie problems of children with overt cleft palate. The incidence of recurrent otitis media in such children is quite high and has been reported to be from 50 to 90 percent by various investigators. Paradise and Bluestone (1974) reported the "universality" of otitis media findings in 50 infants with cleft palate.

Hearing loss as a secondary problem to the middle ear disorder related to cleft palate is also very common and may exist in 90 percent of such patients (Yules, 1970). The incidence and severity of middle ear problems related to the cleft palate decrease as the patient grows older. Otologic and hearing problems associated with submucous cleft palate have been reviewed by Bergstrom and Hemenway (1971). They reported an incidence of 39 percent with recurrent or chronic disease of the middle ear ranging in severity between serous otitis media to cholesteatoma. Conductive hearing loss was demonstrated by 34 percent of the group, while an additional 25 percent had either pure sensori-neural or mixed type hearing loss.

Although numerous ideas have been offered to explain the high incidence of hearing problems associated with overt and submucous cleft palate children, most clinicians agree that the deficiency of palatal musculature is the probable cause of poor eustachian tube function. This results in inadequate middle ear ventilation, effusion of fluid, tympanic membrane retraction and hearing loss. Such disease of the middle ear is most common in children between 3 and 8 years of age, which also corresponds to the increased exposure and susceptibility to upper respiratory infection found in this age group (Bluestone and Klein, 1988). The diagnosis of ear disease and hearing loss in the infant with cleft palate is difficult because of the small structure of the external ear and the infant's neurologic immaturity (Stool, 1971). The overt cleft palate is diagnosed within the first few days of life, although the submucous cleft palate may not be diagnosed until years later.

Cone-Wesson et al. (2000) reported a prevalence of 4.7 percent for congenital orofacial anomalies. Given the current data by Cone-Wesson et al, the 2000 year position statement had emphasized the importance of this HRF in addition to all the previous high risk programs. Fortnum and Davis (1997) also had emphasized the

importance of craniofacial anomalies as a HRF in 2000 position statement.

Childhood Illnesses

Bacterial and viral diseases have long been recognized as causes of deafness. Congenital deafness has also been attributed to meningo-encephalitis, chicken pox and other viruses. Bacterial postnatal infectious known to result in deafness due to meningogenic spread include streptococcus, pneumococcus and staphylococcus. Acute otitis media and upper respiratory infection are commonly associated with bacterial meningitis.

Common viruses of the later post-natal period known to or suspected to cause deafness and/or vestibular symptoms include mumps, measles, chickenpox, influenza and viruses of the common cold. Deafness results from damage to the inner ear as a result of direct infiltration via the internal meatus. Disease may be limited to the endolymphatic system with the inflammatory process beginning in the vascular beds (Lindsay, 1967).

The viral diseases may cause mild to profound sensori-neural hearing loss. Histopathologic effects of viral infections reported have included extensive destruction of organ of corti, degeneration of saccule, damage or complete destruction of stria vascularis and tectorial membrane, damage or obliteration of vestibular system, and atrophy or destruction of neural pathways (Lindsay, 1967). Residuals of post-natal viral infections include nerve atrophy, notably the optic nerve, cerebral palsy, mental retardation, disturbances of respiration, muscular atrophy or paralysis, convulsions, disturbances of autonomic system and disturbances of metabolism.

Infections meningitis causes deafness as the result of bacterial labyrinthitis due to an extension of the infection from meninges. The infecting virus has been traced from the meninges to the inner ear through the cochlear aqueduct and along vessels and nerves of the internal auditory meatus (Paparella and Suguira, 1967). Sensori-neural hearing-impairment is the most common complication of bacterial meningitis in infants and young children. The micro-organisms associated with hearing loss include Haemophilus influenzae, Neisseria meningitidis and streptococous pneumoniae.

Although the severity of the sensori-neural configuration may range between mild and profound hearing loss, the audiometric pattern is typically bilateral, symmetrical and irreversible. Conductive hearing loss components are often noted as acute otitis media and upper respiratory infection are commonly associated with bacterial meningitis. Estimates of the frequency of hearing loss following bacterial meningitis, based almost entirely on retrospective studies, have been reported to be 2.4 - 29 percent of children who survive this illness (Kaplan, Catlin, Weaver and Feigin, 1984). Despite the significant reduction in overall mortality due to the use of antibiotics, nearly 50 percent of children with bacterial meningitis still have significant handicaps. In a prospective evaluation of acute bacterial meningitis in children, Dodge, Davis and Feigin (1984) tested the hearing of 185 infants and young children older than 1 month of age. Of this population, 19 children (10.3 percent) showed permanent sensori-neural hearing loss. Transient conductive hearing loss was identified in 16 percent of the sample of patients.

Bacterial meningitis has been mentioned in the 1982, 1990, 1994, 2000 position statements and Colorado screening project (1972).

Neonatal infections like viral encephalitis, labyrinthitis, chronic lung disease were included in addition to bacterial meningitis in the Utah high risk program (1967) and National Institute on Deafness and other communication Disorders (NIDCD) consensus conference (1993).

Head Trauma

Head injuries, as the result of as the result of accidents involving traffic or falls at home, are common in the young. The child's skull is more deformable than that of an adult and will often dent without fracture. The sutures have not united and fissure fractures may persist as separated sutures. As in adults, fractures of the temporal bone are classified relative to the axis of the petrous portion (Schuknecht, 1993). The fracture line usually spares the cochlea and the deafness tends to be conductive in nature but may be sensori-neural or mixed.

Approximately 80 percent of temporal bone fractures are longitudinal and 20 percent are transverse, although they may occur together (Pansier, 1983). Electronystagmographic disturbances have been reported to persist as long as eight years after blunt head injury in

children (Vartianen, Karjalainen and Karji, 1985). Skull fracture involving the occipital or squamous portion of the temporal bone and involve the otic capsule. Should the fracture line cross the external auditory canal, laceration of the skin and bleeding of the external canal may occur with little permanent loss of hearing. More medial fractures may produce bleeding in the middle ear or disruption of the ossicular chain, which would create a maximal 60 dB conductive type hearing loss. Healing loss due to concussion may be totally or partially reversible, while hearing loss due to fracture of the cochlea is irreversible (Barber, 1969).

Because head trauma, both penetrating and non-penetrating, may occur to any part of the head and with any degree of severity, the auditory consequences are equally variable. Such a patient may have a conductive impairment, a sensor neural impairment, or a mixed impairment. Transverse fractures are more critical for hearing losses and frequent severe vertigo. Transverse fracture of the temporal bone represents a severe and long-term injury to the auditory structures. Hearing loss among these patients is primarily sensori-neural in type and frequently total in extent (Tos, 1973). Williams, Ghoraye and

Yeakley (1992) evaluated twenty-seven temporal bone fractures in 25 pediatric patients and found hearing loss in 24 patients, the most common of which was conductive hearing loss. He also reported a bimodal distribution in the incidence of temporal bone fractures in children. There is an early peak at 2 to 3 years, and a second peak at 13 and 16 years. The early peak is due to auto/pedestrian and motor vehicle accidents, while the latter peak is due to falls and motor vehicle accidents.

Hearing loss and vertigo may appear with longitudinal fractures but they are less common. A conductive component is found more often with longitudinal fractures. Relatively moderate trauma to the occiput of the skull can cause a permanent sensori-neural hearing loss. Schuknecht (1974) states that a blow to the head creates a pressure wave in the skull that is transmitted through bone to the cochlea just as a pressure wave in air is carried by the conducting mechanism and the injury must be attributed to intense acoustic stimulation.

Koike (1972) noted that 15 percent of a group of skull trauma patients with facial paralysis had total loss of hearing and vestibular

function. Only 6 percent of tills group had normal hearing following the injury. Barber's (1969) data revealed that high frequency hearing loss is an audiometric pattern rarely observed in functional patients who exaggerate the hearing loss. Most patients who sustain true hearing loss from concussion injuries will have pure sensori-neural losses or loses with sensori-neural components.

The above studies show the importance of head trauma as a cause of hearing loss. It has been considered as a HRF in the 1990, 1994 and 2000 position statement but not in the previous high risk registers.

Persistent Otitis Media with Effusion (OME)

OME is a common illness in young children, by some estimates affecting two-thirds of infants in their first years of life (Klein, 1983). The incidence statistics concerning otitis media are impressive : 76-95 percent of all children have had at least one episode of otitis media by age 6 years (Howie, Ploussard and Sloyer, 1995), approximately 50 percent of all children have had one episode of OM by age 1 year; and

by age 2 years, this later incidence increases to 75 percent. In Pittsburg, a careful study of preschool children was carried out by Casselbrant, Brostoff and Cantekin (1985). They confirmed that nearly 60 percent of the children examined monthly for 2 years had documented OME that lasted approximately 2 months in duration. The prevalence of this disorder showed strong seasonal variation associated with the presence of upper respiratory tract infections and often resolved spontaneously with a high recurrence rate.

In addition to the discomfort experienced by those suffering from the disease, it is a frequent cause of hearing-impairment in children who do not have sensori-neural hearing loss. For instance, Bess (1983) estimates that 26 - 55 percent of children with OME have mild to moderate hearing losses in the speech frequency range. There has even been the suggestion that because the hearing loss in children with otitis media is intermittent, the impact upon language development may be even greater than in those with a stable hearing loss (Menyuk, 1980).

Children who have experienced peri-natal insults such as pre-maturity asphyxia and respiratory distress syndrome have also been noted to be at risk for developmental difficulties, including delays in acquiring language. Language impairments have been documented as early as 1 year of age and persist through the early school years (Hubatch, Johnson, Kistler, Burnum and Moneka, 1985). Similar results have been reported by Wallace, Gravel, McCarton and Ruben, 1988; Gravel and Wallace, 1992.

Although the pathogenesis of otitis media during the first 2 to 3 years of life has been studied extensively there are few data available on the specifics of the hearing loss that frequently accompanies the condition in infants and young children. The hearing loss associated with OME is temporary, but sometimes persistent or recurrent, and fluctuating in degree (Fria, Cantekin and Eichler, 1985) and symmetry (Hall and Grose, 1993). In theory, such a reduced and fluctuating input signal could make learning the auditory-linguistic code challenging for some children (Friel-Patti and Finitzo, 1990; Gravel and Wallace, 1995; Nozza, 1994). Gravel and Wallace (2000) reported that children who were classified as bilaterally OME positive in years 1, 2 and 3 had

significantly poorer hearing than children classified as bilaterally OME free in each of these time periods. Similar results have been reported by Fria, Cantekin and Eichler (1985).

High frequency (HF) hearing loss has been consistently reported in retrospective studies of children and adults with otitis media (Lopponen, Sorri, Pekkala and Penna, 1992). In report of HF hearing in children who had recovered from otitis media (OM), Margolis, Hunter, Saupe and Giebink (1993) reported that : (1) Children who had persistent mechanical middle ear dysfunction had the poorest HF hearing; (2) Children who recovered from OM had poorer HF thresholds than children without OM histories; (3) Children with more severe OM histories had poorer HF hearing than those with less severe disease; and (4) HF hearing losses in children with OM histories increased with frequency over the HF range (8 to 20 kHz). Similar results were reported by Hunter, Maugolis, Ryyken, Le, Daly and Giebink (1996). Both middle ear and inner ear pathology were hypothesized to contribute to HF hearing losses in children with OM histories. Welsh, Welsh and Healy (1983) demonstrated that 75 percent of the children with a past history of middle ear disease failed

at least one segment of a battery of central auditory tests introduced by Willeford. This shows that the auditory processing abilities of children with persistent OME are also affected along with language and academic abilities.

In view of the incidence and prevalence of OME and then-effects on the child's hearing and language abilities this has been considered as a HRF in the position statements of JCIH (1994 and 2000).

Parental Concern

McAree (1970), a mother of a deaf child stated "when he was 7 days old ... our car door was faulty and to close it we had to bang it several times. The driver scolded us saying that we would upset the baby, but Janie lay in his carry-cot staring at the ceiling of the car ... No blink, No jerk .., No response at all.... After 6 weeks my husband began to worry and he devised his own tests" (P.314). This shows the concern of the parents regarding their child's hearing ability. Because

of its importance many people started investigating about parental concern regarding the child's speech and hearing abilities.

Barringer, Strong, Blair, Clark and Watkins (1993) reported that informal hearing screening procedures (parental suspicion and referral) were the procedure of identification for 80 percent of the children. Whereas Rosenfeld, Goldsmith, Tetlus and Balzano (1997) reported that caregiver concern was the highest rated item and more than 50 percent of caregivers reported spending at least a good part of then-time worried about their child's ear problems.

There are variations regarding the age at which the parental concern starts. Mindel and Vernon (1971) suggest that hearing-impairment can be found as early as two weeks after birth. But, the discovery may be a slow process, related to the nature of the parents personality, parents relationship with the other members of the family, ordinal position of the deaf child and the importance of verbal language to the family and culture. Thus, parental concern has caught the attention of many as an important indicator of the risk. On the other hand, Northern and Downs (1974) reported 11 months as the average

age naive parents suspect deafness in their children. Whereas Mahoney and Eichwald (1979) found that parents became better reporters with the advancing age of the infant, regardless of educational efforts they undertook to increase the awareness. Maki-Torkko, Lindholm, Vayrnen, Leisti and Sorri (1998) reported that parents were the first to suspect a hearing-impairment in half of the cases that they studied.

Parental concern as the HRF has been considered in the Utah project and position statements of 1990, 1994 and 2000 given by JCIH. Thus, utilization of parents in the screening of their own children seems a significant step towards the better utilization of the concept of high risk register.

From the review of literature, it is evident that there are several causes of hearing-impairment. The cause could occur at different stages of the child's development. At times it may not be possible to determine the cause. In such time it would be the concern of the parents that lead to the suspicion of a hearing loss. Based on the causes

of hearing impairment several HRRs have been developed. Most of the HRRs have included all or some of the following factors :

1. Consanguinity
2. Family history of childhood sensori-neural hearing loss.
3. Maternal illness during pregnancy.
4. Drug intake by mother during pregnancy.
5. Maternal irradiation during pregnancy.
6. Pre-maturity
7. Low birth weight.
8. Delayed birth cry
9. Birth asphyxia and cyanosis
10. Hyperbilirubinemia
11. Incubation or ICN admission
12. Congenital orofacial anomalies
13. Illness to the child.
14. Head trauma
15. Persistent otitis media with effusion
16. Parental concern.

METHODOLOGY

The objectives of the study:

(i) To formulate two separate high risk registers (HRR) for children in the age groups of birth - 28 days and 29 days - 3 years to be used by professionals and non-professionals,

(ii) To check the efficacy of the formulated high risk register in identification of hearing loss.

The study was conducted in two stages. Stage one involved the development of high risk registers for medical and non-medical persons. Stage two involved checking the efficacy of the developed high risk register.

Materials used:

The study was done using four questionnaires which are given below:

(i) For medical persons two questionnaires were developed which consisted of high risk indicators of high risk indicators of hearing loss for children in the following age groups:

(a) Birth - 28 days (Appendix A)

(b) 29 days - 3 years (Appendix B)

(ii) For non-medical persons two questionnaires were developed which consisted of high risk indicators of hearing loss for children in the following age groups:

(a) Birth - 28 days (Appendix C)

(b) 29 days - 3 years (Appendix D)

Procedure:

Stage-1: Development of questionnaire

In stage one, the questionnaires were formulated which were distributed to medical and non-medical persons for pilot study.

Formulation of the questionnaires:

Four questionnaires were developed which consisted of questions regarding pre-, peri- and postnatal high risk indicators of hearing impairment. High risk factors relevant to Western and Indian conditions were included in the questionnaires. These high risk indicators were related to both congenital and late onset hearing loss. Among the four questionnaires, two were meant for medical persons and the other two for non-medical persons. Each of the above HRR had two different set of questions for children in the age groups of birth - 28 days and 29 days - 3

years. The questionnaires meant for medical and non-medical persons consisted of the same risk indicators with differences in terminology and sentence structure. The questionnaires for non-medical persons were formulated with simple terminologies and sentences such that even a layman could understand them.

Pilot study :

The developed questionnaires were distributed to ten audiologists who had at least three years of experience. They were requested to give their suggestions regarding the adequacy of the questionnaires in terms of the following:

- ✧ Understandable
- ✧ Need to change sentence structure
- ✧ Need to change terminology
- ✧ Need to change both terminology and sentence structure

They were also requested to give their additional comments regarding inclusion or deletion of risk factors in the questionnaires. The HRR used in All India Institute of Speech and Hearing (AIISH), the position statement given by the Joint Committee on Infant Hearing in

1994 and 2000 were given along with the questionnaires for cross reference. Their suggestions regarding the terminologies, sentence rephrasing and combining the questions were incorporated in the questionnaires. The details of the number of questions in each questionnaire is given in Table-1. It indicates the difference in number of questions between the questionnaires for medical and non-medical persons. Questions for medical persons had specific medical condition whereas these were mentioned in a general way for non-medical persons. Hence the questionnaire for medical persons for each of the age groups had an additional question.

Table-1: Number of questions in the questionnaire

Questionnaire		Number of questions
I. Medical	- Birth - 28 days	12
	- 29 days - 3 years	9
II. Non-medical	- Birth - 28 days	11
	- 29 days - 3 years	8

After incorporating the suggestion by audiologists the questionnaires meant for medical persons were distributed to five doctors and five nurses. The modified versions meant for non-medical persons were distributed to ten parents/caregiver of the hearing-impaired children.

As with the audiologists they were requested to give their suggestions regarding the adequacy of the questionnaires in terms of the following :

- ✧ Understandable
- ✧ Need to change sentence structure
- ✧ Need to change terminology
- ✧ Need to change both terminology and sentence structure

Neither medical persons nqr non-medical persons suggested any modifications. Hence, the same questionnaires were used for the study.

Stage-2: Evaluating **the** efficacy **of the HRRs** :

Both the HRRs consisted of the same risk indicators. Hence only the questionnaires meant for non-medical persons were used to check their efficacy. The results obtained would be common to both the versions of the questionnaires.

/. Subject criteria:

To check the efficacy of the questionnaires, 200 subjects who met the following criteria were selected.

- Parents/caregiver of the hearing impaired children in the age range of 1-8 years who knew the birth history of the child.
  They should be from different parts of the country including southern and northern regions.
  They should be able to communicate through speech.

//. *Method of administration*

The questionnaires meant for both the age groups were administered to each parent/caregiver. Of the 200 subjects, 196 were the mothers of the hearing-impaired children, two were the fathers and the remaining two were the caregivers. A face-to-face interview was conducted by the investigator alone or with the help of a translator. The interview was conducted in the subject's native language or English. The subjects were instructed to indicate as to whether the risk factor occurred between birth-28 days or 29 days -3 years.

///. *Response recording:*

The risk factors and the age at which it occurred was recorded by the investigator. The responses obtained were analyzed and the children were categorized as either at-risk or not at-risk.

RESULTS AND DISCUSSION

Two hundred parents or caregivers of hearing-impaired children were administered the developed high-risk registers. This was done though face-to-face interviews. Information regarding the presence of risk factors in the children and as well as any other relevant information were recorded. Raw data was obtained and converted into percentage. The results thus obtained are discussed for the risk factors considered in all the four questionnaires that were developed.

It was found that 151 (75.5%) out of 200 children were at risk for hearing-impairment. They had one or more high risk factors (HRFs). Fourty-nine children (24.5%) did not have any of the HRFs. Table II shows the raw scores and percentage of occurrence of HRFs in the order of highest occurrence to the least occurrence.

Table-II: Raw data and percentage of occurrence of HRFs included in the four questionnaires.

High risk factors	%of occurrence	n
Parental concern	100.0	200
Consanguinity	35.5	71
Family history of childhood SN hearing loss	19	38
Delayed birth cry	16.5	33
Incubator/ICN admission	13	26
Pre-maturity	12.5	24
Maternal illness during pregnancy	9.5	19
Illness to the child.	7.5	15
Hyper-bilirubinemia in the child	6.5	13
Drug intake by mother during pregnancy	6	12
Birth asphyxia	5.5	11
Drug given for illness to child	4.5	9
LBW<1500gm	3	6
Congenital craniofacial anomalies	1	2
Persistent OME	0.5	1
Maternal irradiation	0	0
Head trauma	0	0

It is evident from table II 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. Each risk factor is discussed further in the following section.

Parental concern :

Parental concern regarding hearing loss was seen in all the children (n=200). This could be attributed to the reasons that the present study is retrospective which was done in confirmed hearing loss population where parental concern would be present. This occurrence is in agreement with Barringer, Strong, Blair, Clair and Watkins (1993) study which reported that informal hearing screening procedures (parental suspicion and referral) were the procedure of identification for 80% of their population.

In the present study, the mean age of suspicion is twenty-one months (ranging froml month - 43 months) which occurs at much later age when the speech had started developing. Only one child was identified at the age of 15 days.

Yoshinago-Itano (1999) had reported about an early intervention at an average age of two months of age. To achieve such an earlier

identification age, the parents/caregivers should be educated regarding the development of auditory behaviour.

Consanguinity

Consanguinity as the HRF was seen in seventy-one children (35.5%). In eighteen (9%) these seventy-one children, it was the only HRF, while in the others it coexisted along with other risk factors which were included in the questionnaire. Though it has not been considered as a HRF in the western registers it has been considered in the high risk questionnaire developed by Ashok Kumar (1980) for the Indian population. This is a support that consanguinity should be considered for the Indian population because of its high occurrence, as can be seen in the present study.

Family history of childhood sensori-neural hearing loss :

This HRF was seen in thirty-eight children (19%). In eleven of these thirty-eight children (5.5%), it was the only HRF, while in the others it coexisted along with other risk factors which were included in the questionnaire. This is in agreement with the study by Cone-Wesson, et al.

(2000) which reported a prevalence of 6.6%. Hence, it was considered essential to retain this risk factor in the final register.

Delayed birth cry and birth asphyxia :

Delayed birth cry was reported in thirty-three children (16.5%). It has occurred along with other HRFs such as low birth weight, pre-maturity, asphyxia or hyperbilirubinemia. Achar (1982) reported that most infants begin to breathe spontaneously within a few seconds of birth. Those infants who cry lustily, their lungs expand fully and quickly. But in others whose response is dulled by various factors (resulting in prenatal or peri-natal hypoxia), the breathing may be delayed (because of delayed birth cry) which may lead to irrepairable brain damage from asphyxia. In the present study, birth asphyxia was reported to be present in eleven children (5.5%) and it did not occur as a sole HRF. This is in agreement with the literature in which it is reported that the occurrence of asphyxia is commonly related to a number of other medical conditions so it is difficult to absolutely establish asphyxia as the single cause of a specific case of deafness. One such instance is reported by MacDonald (1980) who pointed out that a pre-term infant is more likely to experience anoxic episodes.

Though the occurrence of delayed birth cry was so high in the present study the parents were able to report it only if the doctors, during the delivery, had informed them about it. Hence, it is possible that if the baby is born without medical assistance or if the doctor does not report this to the parent, they would be unable to report about the occurrence of delayed birth cry. However, as it coexists with other HRFs this population can be identified by the occurrence of other HRFs. In spite of its co-existence with other HRFs this has been retained in the questionnaire because of its high occurrence in the present study. It is also possible that parents may not be able to report other high risk factors such as low birth weight or asphyxia which co-exists with delayed birth cry. In such cases, those children can be identified with this factor hence increasing the sensitivity of the questionnaire.

Incubation or Intensive Care for Neontes (ICN) Admission :

Twenty-six (13%) children were admitted in ICN or incubated immediately after birth. Most of these children also had other complications including high bilirubin level, intrauterine viral infection, pre-maturity, low birth weight and anoxia.

Similar observations were reported by Simmons (1980) who noted that most of the high risk register items were significantly associated with hearing loss in their ICN graduates, who were judged at risk for hearing loss. He found anoxic episodes at or shortly after birth in 73 percent of ICN babies while Halpern, Hosford-Dunn and Malachowski (1987) found peri-natal function as a factor in 10 percent hearing-impaired ICN subject. They have also found that the length of stay in ICN and gestational age predicted hearing loss with 98 percent sensitivity and also reduced the initial risk group by one-third. Simmons (1980) had reported an incidence of hearing loss of 1:52 in ICN babies. Hence, it can be concluded that children who were missed out through any of the co-occurring HRFs, can be identified based on them being admitted in the ICN or incubated.

Pre-maturity:

Pre-maturity was reported in twenty-five children (12.5%). It had co-occurred along with low birth weight (LBW), asphyxia, hyperbilirubinemia and ICN admission in most of the children. A number of parents were able to report about pre-maturity. They were able to report this more often than other factors such as LBW or birth asphyxia. The findings of the present study are in agreement with the findings of Barton, Court and Walker (1962) who reported a pre-maturity rate of 12.5 percent

with 18 percent of those children having a history of severe neonatal jaundice which is considered a HRF. In view of number of children who had pre-maturitity as a risk factor in the present study as well as in the literature, it was decided to retain this in the final register.

Low Birth Weight (LBW) :

Of the 200 children only six (3%) had low birth weight of < 1500 gms. This concurs with the finding of Wright et al. (1972) who examined seventy infants with birth weights of ≤ 1500 gm and found four children (5.7?/o) were deaf in one or both ears. In the present study, fifteen children (7.5%) had birth weight between 2000 and 2500 gm and eleven children (5.5%) had birth weight between 1500 and 200 gm. Though twenty-six children (13%) had birth weight in the range of 1500-2500 gm, they were not considered as high risk in order to reduce the unnecessary over referrals. All the children with a birth weight of 1500-2000 gm invariably had some other risk factor which was included in the present study. Hence, they would not have been missed out because of the presence of the other risk factors such as pre-maturity, hyper-bilirubinemia, asphyxia or incubation.

Maternal Illness During Pregnancy :

Nineteen mothers (9.5%) had the history of illness during pregnancy. Among the illnesses, bleeding during pregnancy and hypertension in the mother were the most frequently occurring illness (four in each condition). Though both these illnesses are not specifically mentioned in the questionnaire, the question for maternal illness has been worded such that it is broad enough to include other illnesses. Also, in the present study these two conditions have always occurred along with other HRFs and hence would not be missed out.

Malaria, chicken pox, cytomegalovirus (CMV), rubella and diabetes (one in each condition)were the other illnesses which did not occur with other HRFs. Nahmias (1974) estimated that although 20 percent of pregnant woman may have CMV, only 2 percent of the infants are infected at birth. This is in agreement with the present study where 1 percent showed occurrence of CMV in children.

Fitzzaland (1985) had reported 9.8 percent occurrence of rubella or other non-bacterial intrauterine fetal functions. Though the present study showed only 1 percent occurrence, the question was retained because Nahmias (1974) had estimated that 1-5 percent of all deliveries are

infected by one of the TORCH agents. Further, due importance has been given to maternal illness in all the high risk registers including the recent 2000 position statement.

Hyperbilirubinemia in the child

Hyperbilirubinemia in the child during the first 29 days after birth was reported in thirteen children (6.5%) which was in agreement with Fitzzaland's (1985) study who reported 4% occurrence of hyperbilirubinemia in which 3.1% of children exhibited hearing loss. In the present study, exchange transfusion was done only in two of these thirteen children,

Illness to the child :

Fifteen children (7.5%) had illnesses like meningitis (n=5), viral fever (n=4), measles (n=4), nephrogenic diabetes (n=l) and pneumonia (n=l). In three of these children the occurrence of illness was between 29 days and 3 years of age whereas in the remaining twelve it occurred before 29 days. Nine children (4.5%) were given medications for the management which may have also adversely increased the risk. The occurrence of the above mentioned illnesses is in agreement with Lindsay

(1967) who reported that mumps, measles, chicken pox, influenza, and viruses of the common cold are the common viruses of the later post-natal period known to cause deafness. Dodge, Davis and Feigin (1984) also found that acute bacterial meningitis resulted in a permanent sensor neural hearing loss in nineteen (10.3%) children.

Congenital craniofacial anomalies :

Congenital craniofacial anomalies were seen in two children (1%). Further it has been reported by Gerber (1977) that orofacial anomalies frequently incorporate hearing-impairment among their stigmata. The findings of this study is in agreement with Fraser (1964) who found congenital craniofacial syndrome in 21/2 of children and with Cone-Wesson et al. (2000) who reported a prevalence of 4.7 percent. It is to be noted that not all infants with orofacial anomalies will have hearing-impairment. The presence of such abnormalities, however, increases the risk of hearing problems in a child.

In the present study, though it had occurred in such a small percentage it was not deleted from the questionnaire because it is an obvious symptom which can be easily detected by professionals or lay-people. It's importance is also evident as it has been considered a relevant

factor in several high risk registers given by Joint Committee on Infant Hearing in the position statements of 1972, 1982, 1990, 1994 and 2000, Santa Barbara Plan (1978), Halifax Project (1977) and Utah State Wide Infant High Risk Hearing Program (1978).

Persistent Otitis Media With Effusion (OME) :

The occurrence of persistent OME was found in only one child (0.5%) in the present study. The reason for such a low percentage of occurrence can be attributed to the sample that was studied. Usually children having a hearing-impairment caused by persistent OME are placed in an integrated set-up, whereas the present study obtained information about children who were predominantly in a special school set-up. Population from different set-ups, including the integrated schools will give a clear picture about the occurrence of the persistent OME. The incidence of middle ear dysfunction is reported to be high in children, with percentages ranging from 33 to 74 percent (Mace, Wallace, Whan and Stelmachowicz (1991). In view of the above, despite it is occurrence being less in the present study, it was retained in the questionnaire.

Maternal Irradiation During Pregnancy :

Of the 200 children, none of them exhibited the HRF of maternal irradiation during pregnancy. This could be because the medical or paramedical professionals are knowledgeable about the detrimental effects of radiation during pregnancy. Hence, it has not been recommended for any of the expectant mother. It can be inferred that unless it is absolutely essential, it will not be recommended in any expectant mother. Recent literature on the causes of hearing-impairment do not include maternal irradiation. Taking into consideration this, it was decided to delete the factor of maternal irradiation from the questionnaire.

Head Trauma to the Child :

None of the parents/caregivers reported head trauma to the child in the postnatal period. Though it has not been reported in the present study it was retained in the questionnaire because Koike (1972) had reported that only 6% of his study group of skull trauma had normal hearing following the injury. Also, it has been reported that head injuries, as the result of accidents involving traffic or falls at home, are common in the young (Adams, 1997). In view of the above despite it is not occurring in the present study, it was retained.

Other risk factors:

A few of the parents/caregiver reported of the children having risk factors other that what is mentioned in the formulated questionnaires. These included postdated delivery ($\geq$ 7 days after due date) and fits to the child. Post-dated delivery was reported to be present in nine mothers (4.5%). In two (1%) of these mothers, this occurred as the only HRF. Since this HRF constitutes only 1% of the population it was not included in order to reduce the number of unnecessary over referrals. Similarly, fits to the child was seen in nine children (4.5) and only fits with no other HRFs was seen in three children (1.5%). Hence this was also not included in the questionnaire in order to reduce the large number of false positive rates.

Taking into consideration all the above mentioned results, the question of maternal irradiation during pregnancy (3^{rd} question in Appendix A and C) was deleted from the questionnaire. The rest of the questions for professionals and non-medical persons in the age ranges of birth - 28 days and 29 days -3 years were not altered.

SUMMARY AND CONCLUSION

High-risk register (HRR) is a questionnaire based method with which one can identify a small group of children whose history or physical condition identifies them as possessing a high chance of having the handicap being searched for (Northern and Downs, 1978). The aims of the present study were to formulate two separate HRRs for children in the age range of birth-28 days and 29 days -3 years and to check its efficacy.

Questionnaires consisting of risk factors relevant to western and Indian conditions were formulated. These high risk factors were related to both congenital and late onset hearing loss. Of the four questionnaires, two were meant for medical persons and the other two for non-medical persons. All the four questionnaires consisted of the same risk indicators with differences in terminology and sentence structure. The developed questionnaires were distributed to ten audiologists and they were requested to give their suggestions regarding the adequacy of the questionnaires. The suggestions of the audiologists were incorporated in the questionnaires. Following this, the questionnaires meant for medial persons were distributed to five doctors and five nurses. The modified versions meant for non-medical persons were distributed to ten parents/caregiver of the hearing-impaired children. Neither medical

persons nor non-medical persons suggested any modifications. Hence, the same questionnaires were used for the study.

Data was collected by interviewing 200 parents/caregiver of the hearing-impaired children. Information regarding the presence of risk factors in the children and as well as any other relevant information were recorded. Raw data thus obtained were converted into percentage.

The study showed that 75.5 percent (n=151) could be identified by these questionnaires whereas 24.5 percent were missed out. In the present study, parental concern was the most frequently reported high-risk factor followed by consanguinity. The other high risk factors in the order of occurrence are as follows:

- ✧ Family history of childhood sensori-neural hearing loss.
- ✧ Delayed birth cry
- ✧ Incubated or ICN admission
- ✧ Prematurity
- ✧ Maternal illness during pregnancy
- ✧ Illness to the child.
- ✧ Hyperbilirubinemia
- ✧ Drug intake by mother during pregnancy

- Birth asphyxia
- Drug given to the child for illness
- LBW<1500gm
- Congenital craniofacial anomalies
- Persistent OME.

Maternal irradiation during pregnancy and head trauma to the child were not reported as high risk factors. Based on the results, the HRF of maternal irradiation during pregnancy (question no. 3 in Appendix A and Appendix C) were deleted leaving other questions unaltered.

In view of these results it can be concluded that in a developing country like India where UNHS is not possible due to various reasons, HRR can be used as a tool for hearing screening. It is recommended that the specificity of the HRR can be studied in further research.

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APPENDIX-A

HRR FOR MEDICAL PERSONS
Birth - 28 days

1. Was the marriage of the child's parents consanguinous?
2. Was there any family history 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?

Note: In the finalized questionnaire only question No.3 was deleted.

APPENDIX B

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 hyperbilirubinemia 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?

APPENDIX-C

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 bom 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?

Note: In the finalized questionnaire only question No.3 was deleted.

APPENDIX-D

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 me 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?