INFANT SCREENING WITH THE HIGH RISK CHECKLIST

REG NO.M92I8

An Independent Project submitted as part fulfilment for the First Year M.sc.(Speech and Hearing) to the University of Mysore.

ALL INDIA INSTITUTE OF SPEECH AND HEARING: MYSORE-6

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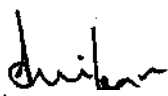
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CERTIFICATE

This is to certify that the Independent Project entitled: INFANT SCREENING WITH THE HIGH RISK CHECKLIST is a bonafide work, done In part fulfilment for the first year Degree of Master of Science (Speech and Hearing) of the student with Reg.No.M92

Mysore
May 1993

Director
All India Institute
 of Speech & Hearing
Mysore - 570 006.

CERTIFICATE

This is to certify that this Independent Project entitled: INFANT SCREENING WITH THE HIGH RISK CHECKLIST has been prepared under my supervision and guidance.

Mysore

May 1993

Dr.(Miss) S.Nikam

GUIDE

DECLARATION

I hereby declare that this Independent Project entitled: INFANT SCREENING WITH THE HIGH RISK CHECKLIST is the result of my own study under the guidance of Dr. (Miss) S.Nikam, Director, All India Institute of Speech and Hearing, Mysore, has not been submitted earlier at any University for any other Diploma or Degree.

Mysore
May 1993.   Reg.No.M9218

ACKNOWLEDGEMENTS

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CHAPTER -I
INTRODUCTION

1.1:Hearing   Speech and Language:

Hearing is a late development in evolution but it haS become the sentinel of our senses, always on the alert.

Hearing does more. The ear and the brain analyse these sound waves and their patterns in time, and thus we can discriminate between two sounds, that we hear. What; is more, we can locate the position of the carriage, and tell the direction in which it is moving.

Bats and some marine animals, living where light is poor or waters are murky, have learned to hear objects as well as events. They send out their own sounds and listen for the echoes. They thus learn the direction, the size and possibly even the texture of objects around them.

Many animals and birds have also learned to signal to one another by their voices, both for warning and for recognition. But we humans, with good ears and also mobile tongues and throats, and above all, our large complex brains, have learned to talk. We attach arbitrary and abstract meanings to sounds, and we have language. We communicate

oar experiences of the past and also our ideas and plans for future action. For human beings, then the loss of hearing brings special problem even and a special tragedy. Butt human society creates a special problem even for those with perfect hearing - the problem of unwanted sound, of noise, which is as much a hazard of our environment as disease germs or air pollution.

When hearing fails

Defective hearing is a common physical impairment in our country today.

From time to time, famous next have turned hearing pro-blems into advantages. The late Bernard Baruch America's great elder statesman, shielded himself from bores by switching off his hearing aid when the conversation degene-rated into prattle, Thomas Edison attributed his great powers of concentration to his deafness. For the vast majority, however, the lack of this critical sense is a constant burden.

The burden is greatest for those who are completely deaf, for total deafness has devastating effects upon psychological and social life. "I am just as deaf as I an blind", wrote Helen Keller, "The problems of deafness are

deeper and more complex, if not more important, than those of blindness. Deafness is a much worse misfortune. For it means the loss of the most vital stimulus - the sound of the voice that brings language, sets thoughts a stir, and keeps us in the intellectual company of man". These poignant words describe the frustration of the child who was either born deaf or became deaf at a very early age, and cannot recall ever hearing at all.

For such a child, learning is an unbelievable struggle. The normal child moves smoothly from hearing words to saying then, then goes on to recognize their representations on the printed page. Each successive step he takes is made easier by the preceding one. But the deaf child can never take the first step unaided. To overcome his initial handicap requires a Herculean effort.

Less of a practical disadvantage, but a severe psycho-logical blow, is the loss of hearing once known and relied upon. Just as speech provides a bridge between people, so the everyday sounds of life - from the hums of traffic to the ticking of a clock - provide a bridge between the indivi-dual and his environment. Most of the time, these sounds do not impinge on consciousness; they are taken for granted

as a background to life. But when they are absent, the world itself is altered. It seems unreal and even dead. The cerie sense of isolation that sets in teas profoundly disturbing effects.

Whether language develops as a result of imate capability or whether it is learnt or whether it is acquired along with general cognitive development, the acquisition or development of language is directly related to the kind and extent of sensory input the child receives. For speech, the input is and must be auditory (Schuell, 1974) and there must be plenty of it. Anything that interferes with the input severely geopardizes acquisition itself. The functional patency and the innateness of the hearing mechanism is therefore a must.

1.2:Early Identification:

First of all, it is difficult to define "how early is early". If we accept the premise that the development of language begins at birth, with the child's first cry, or atleast accept Menyuk's (1977) proposition that bubbling period enables the child to make both perceptual and pro-ductive categorization of the speech signal which nay be

crucial for later language development, then "Early" turns
out to be very early indeed. Downs (1978) puts it more
emphatically. "It is important to identify hearing loss
by three months of age". But Mencher (1980) goes further.
"When wo say early identification wo mean at birth; when
we say early diagnosis, we mean within the first few weeks
and when we say early management, we mean as early as
possible in life, even beginning within the first month.
Infact . . . .if he is over three months of age - he is a
geriatric".

why early identification of hearing loss is necessar?

     If hearing loss is identified as early as possible,
then early intervention (or mamtgemsnt) is possible which
may include s
1) putting a hearing aid on the child
2) and giving him speech and langaage training.

Downs (1973) proposed that "if the onset of hearing loss
is after six years of age, the child will have good
language. If the onset is around three years of age, the
problem is not much as the loss occurs at birth. If it is
congenital then the child will have maximum problem". So,
according to Downs (1978) "If a child with congenital

hearing loss is given training as early as possible, his/her speech and language can be brought to the level of child, who has acquired hearing less.

Critical Age Concept t

Lenneberg (1967) -gave the critical age concept. According to this concept "language is biologically innate and there is an optimum tine for its development. Language cannot be learnt once the time is passed critical age is within the puberty age."

Greenstain, et al (1976)- Compared two groups of deaf children and the upper age limit was 40 months, Group-I was given training before they were 16 months of age and Group-II was given training after 16 months of age. The Group-I had superior speech and language skills when compared to Grotqp-II So this proved that early identification by early training helps to develop speech and language skills.

Northern and Downs (1974) - After the puberty age even with training the hearing-impaired would have difficulty in repeating speech sounds. Several reasons have bees given why they are not able to acquire speech and language.

(1) After puberty cerebral plasticity is lost

(2) Physiological maturation of the brain is complete

(3) Cerebral organisation is complete.

Ebbin and Griffths (1973) - They studied children until the age of 24 months. Those children Who received training before eight months of age, a high percentage of theia regained their hearing ability. So 67% of children who were given training before eighth months of ago regained their hearing ability over a period of five months, but if the child had pre-natal maternal rubella then the hearing did not return to normalcy. For those who were given training after eighth months of age, their hearing did not return to normalcy. They found no correlation between the age at which they started therapy and the amount of remission (returning to normalcy). Also they found no correlation between the degree of loss and time taken to get back their normalcy.

   The remission occured due to several reasons:

(1) Due to immature hearing mechanism and due to this they did not receive adequate sound stimulation to the contex. With amplification hearing mechanism starts to function.

(2) Myelinization of the neural pathway is not complete, due to a lag in maturation.   With stimulation to the auditory mechanism the myelinization gets complete.

By eighth months of age myelinisation becomes static ie. after giving amplification also the child does net get back his hearing.   Amplification device used had a wide frequency range.   So this reports that there was no error in evaluation.   So this is another study supporting the critical age concept.

Ebbin (1974)- Says that instead of calling it a critical age, it must be called as "sensitivity period" ie. at this particular time a person can learn something more efficiently more quickly with less training.

Williams (1970) (A contradictory study) - Those children who were diagnosed to have bearing loss early had lesser amount of speech than those who were diagnosed to have hearing loss after two years.   He studied children between the ago of 5 years - 14 years.   Total 51 of them.   Those who were diagnosed to have hearing loss before two years of age only 27% of them got speech.   And those who were diagnosed after two years of ago - 80% of there learned speech.

It is possible that the children who were diagnosed as having hearing less early could have had also a central disorder or those children who were diagnosed early were not fitted with suitable hearing aids or did not receive adequate training.

Bench (1971) - says "Early identification is important but diagnosis is not so important".

The earlier the hearing loss is identified in a child, the easier it would be to bridge the gap between normal bearing and the hearing- impaired children.

And if hearing loss is identified early and followed-up with early intervention chances of the hearing-impaired child developing psychological problem will be less.

Implications: The above discussion makes it imperative for us, especially audiologists to take up the challenge to identify a child with hearing loss at the earliest possible time. There are two possibilities: one to test and evaluate every child born thoroughly, which by its sheet weight is not possible, or two, to screen all children or atleast a selected population of children whenever and wherever they are accessible soon after birth.

1. 3: <u>The screening method;</u>

Screening as accepted by World Health Organization (WHO) is defined as "the presumptive recognition of un-recognised disease of defects by the application of tests, examinations and other procedures which can be applied rapidly" (Roberts, 1979), Screening tests sort out apparently well persons who probably do not have a disease from those who probably do have the disease. They are not intended to be diagnostic. Persons with positive or suspicious findings must be referred to specialists for diagnosis and necessary treatment (Wilson and Lungnci, 1968).

<u>Types of screening:</u>

There are five types of screening which can be employed (modified from Roberts, 1977) viz:

1. Mass scre<u>ening</u> - Where an entire population may be screened by mass screening techniques. Eg. Newborn screening for phenyl ketonuria disease (PKU).

2. Selective or prescriptive screening - 'Which can be applied to a given group of people who are more suspect than the general population eg. screening of only Jewish population for Tay Sech' s disease.

3. Multiple screening - Which extends the number of screening measures used on a given individual from the two or three used in multiple screening to a battery of as many as ten screening tests.

4· Surveillance - Used to periodically follow as an individual or a group and to monitor their present state of well being.

SCREENING CRITERIA:

Public health experts have fixed certain for a successful screening program (Frakenberg, 1971, 1973). They are :

i) Occurence of the condition frequent enough or con-sequence serious enough to warrant screening.

il) Amenability to treatment or prevention that will foretell or change the expected outcome,

ill) Availability of facilities for diagnosis, follow-up and treatment, and referral,

iv) Cost of screening reasonably commensurate with benefits to the individual.

v) A screening tool or test that validity differentiates a disease from non-disease.

vi) Acceptable to the public.

(Frekenberg (1971) also prescribes the following

specific criteria for an efficient screening tool.

(i) Sensistivity- accuracy in correctly differen-

tiating an individual with the disease from the

general population. its cost should be reasonable.)

(ii) Specificity- accuracy in correctly differen-

tiating an individual with the disease from the
those having. It

(ill) Standardisation - The test should be well esta-

blished as compared with a standard - either

another test or a diagnostic test.

(iv) VALIDITY- *it should* measure what it is supposed

to measure.

(v)   Reliability   - Screening results should be consis-

tent each time the tool is used,

(vii) Acceptability - to the patient, the family,

society and the tester.

(vii) Its cost should be reasonable.

There   are also certain factors that one has to consider

in e screening program (Roberts, 1977): Screener' s skills,

population to be screened, the cost, the time factor for

the screener, the patient, and the family, screening place,

where and whom to refer failures or suspects and so on.

Considering all these, hearing, screening, in its present state-of-the art meets all these criteria at every level-newborn level, pre-school age level and school-age level. It also meets specific goals - goals that are different for each level (Downs, 1973).

<u>Principles of a hearing screening test</u>:

The following set of principles and/or(goals are given by Barley (1961) and Mencher (1977):

(1) The fundamental concern is the maintenance of an optimum state of health.

(2) The ultimate goal is conservation of human resources or the optimum functioning of the individual, acceptance by his peers and maximum use of his skins, regardless of severity of the hearing handicap.

(3) Need to be established on the broadcast possible base to reach the largest possible number of children.

(4) A compromise with the ideal program of hearing evaluation.

(5) It is not an end in itself nor does it stand alone. Proper referrals must be made when and where needed.

(6) It will not be effective unless high standards are established, implemented and maintained. It cannot fee expected to give 100% identification. False positives and false negatives are pert of the pictures and are to be expected .... without them the procedure is not screening.

(?) A longitudinal approach to hearing screening is needed, The ideal is to conduct a reliable test as early in life as possible and to provide follow-up screenings.

(3) Ongoing hearing screening programs not only identify disorders but also awaken awareness and interest on the part of the citizens in the prevention and treat. ment of hearing losses.

(9) They can alert conmunities as to future needs, how to utilise existing resources, the personal services and facilities needed.

(10) Money spent for the prevention of hearing loss on early identification and treatment of a problem in money and time saved.

(11) Extensive research must continue to investigate "why", "how often"; and "in whom" hearing impairment occurs and what can be done with it.

(12) Hearing screening programs are not the single
      province of otology, paediatrics, public health
      nursing or audiology. While one of these disci-
      plines must coordinate at any given site under
      any given circumstances, the process must be a
      joint effort of sister professions with appro-
      priate referral and ultimate management predicted
      on the etiology, prognosis and types of treatment
      required.

Once the goals and objectives of an identification
program are clear, it is much easier to determine the
appropriate procedure(s). In essence, an otherwise
formidable task is reduced to simple. Comparison shopp-
ing based primarily on the test operating characteristics,
developmental range, intensity range, frequency specifi-
city, and measurement construct (content) of each instru-
ment or procedure. To assist in this regard Table-1
summarises several common procedures relative to certain
of these various selection criteria. Additional factors
related to expenses, such as equipment costs, test admi-
nistration time, maintenance. Charges and so on, should
also be considered.

Summary of screening measures and their respective test operating characteristics based on data from the source indicated

Application/ method	Source	Age range	Test operating characteristics				Cut off criterion
			Sensitivity	Specificity	Under referral	Over referral	
Birth related							
High risk register	Mahoney & Eichwald (1970)	Birth	65	75	32	68	Positive on one or more factors
Behavioral arousal	Downs and Sterritt (1967)	0-4	63	29	25	75	50-60 dB HL
Auditory Brain stem	Fria(1985)	Birth-adult	98	90-95	2	50	30-& 60-dBs HL
School related							
Puretone individual	Wilson & Walton (1978)	36	63	97	2	99	20 dB HL
	Melhikek etal(1964)	85	85	98	3	10	20 dB HL
Primary care							
Behavioral VRA	Kenworthy etal(1986)	6	80	86	14	20	30 dB HL Fail any item at the 90% point at emergence.
	Coplan(1983)	0-36	90	93			
	Kenworthy (1980)		74	90	1	48	

<u>Early mass screening studies:</u>

In 1964 Downs and sterritt described a hearing screening method based on observing behavioral changes in the new born in response to a 90 or 100 dB signal (while noise and/or narrow band noise). In 1965, Downs noted "experience in observing auditory behaviour of over 5000 new borns infants leads us to believe that it is feasible to screen for peripheral hearing deficits at birth. In 1909, Downs Hemenway reported the results of their screening program involving 17,000 new borns. They found that they could identify one deaf child in every 1000 new borns.

Though their techniques were sound in principle, their specific testing procedures were not as sensitive as intended (Gerter, 1971), At a result those who did not understand the preliminary nature of the project instituted their own new born screening programs. Data published subsequently showed that they could detect any where from one deaf infant in 1000 to none in 14,000 in all, out of 61,000 babies of incidence of one in 2,800 was reported (Downs, 1971).

These screening procedures did not prove to be very accurate. There were too many misses and false hits. Also,

the low morbidity of hearing loss, very high false posi-
tive rate (usually in the order of 12 to 20%) made the
cost of screening too high for general use (Davins, 1978).
Some of the later projects also had limited successes.
Bordley and Hardy (1972) screened 1182 new borns and found
that his program misses 98% of the true positives.
Shapiro (1974) screened another 4000 new borns and could
not find any baby with a confirmed hearing loss (he was
unable to follow up most of his hearing test failures).
Nikam and Dharmaraj (1971) screened 941 infants and found
that their test failed 31.2% of them.   They too faced the
problem of follow-up.

Advantages of mass screening:

Goldstein and Tait (1971) list the following advan-
tages:

(1) Routine screening in a hospital is desirable because
    it is the only situation or time when all babies
    (except those born outside the hospital facilities)
    are available for testing.

(2) It provides an opportunity to discover the few deaf
    infants who might have escaped detection at birth
    solely on the basis of suspicion.

(3) It may provide information regarding adequacy of hear-
ing at birth in children who may later lose their
hearing.

(4) It can help alert the physician to the presence of a
more general or more pervasive disorder.

(5) It could, if carefully controlled, provide valuable
information about normal development of auditory
responsivity.

(6) Provides an important stimulus to the physician and
particularly, pediatrician to become more conscions
of, and, knowledgeable in auditory disorders in
children.

(7) The cost of screening can counter-balance cost of
training one deaf child (Downs, 1967).

Criticisms against pass screening:

The criticisms against new born screening have come
mainly from Goldstein and Tait (1971); Elsenberg (1971);
Ling (1976) and also Downs and Sterritt (1967). However,
the most comprehensive of all is tee review by Goldstein
and Tait (1971) who discussed them under four headings:

(A) Magnitude of the problem: They argue that the magni-
tude of the problem is not at all that bleak. Most deaf

children are seen before the age of two and that it is improper to blame the parents and physicians for it since the onset may be delayed one. They also point out that such delayed onset cases are most likely missal by a new born screen. They also feel that 90 to 100 dB level of test signal may be more and that unilateral hearing loss cases are not detected though they may have listening problems. They also point out the dangers of misdiagnosis and subsequent mismanagement of the child which may further compound the difficulty.

(B) Effectiveness of screening procedures: They point to the fast that Downs and Henenway could detect only four deaf out of 10,000 and the high false positive rate (150 initially suspected). They feel that rapid testing often resorted to allow no room for undothing the babies besides reducing its reliability.

(C) Effectiveness of follw-up procedures: They feel that clear answers to various questions concerning follow-up procedures are sot apparent and that they maintain that routine neonatal screening as proposed can lead to parental and professional confusion and to mismanagement.

(D) Limitation of emotional appeal:They question the validity of various arguements and appeal for encouraging neonatal screening. They argue that concern may lead to unnatural treatment of the deaf child and that parental interest and involvement cannot be taken for the deaf child and that parental interest and involvement cannot be taken for granted. Arguing about economic aspects, they feel that one year gained by early identification can be useless (unless that one year gained makes a quali- tatively important difference to the child) unless it eliminates atleast two years of special education at a later date and that evidence to support this is not available. They also point out that cost of screening is not really negligible as claimed.

   Arguing that comparison to PKU is not justifiable since it iS a reversible process Whereas deafness is not, they point out, that no follow-up studies have been done to confirm the expectations and benefits claimed. Finally, they quote Downs herself, who felt that original enthusiasm about the effectiveness of the screening had not been justified (Downs, 1970).

Eisenberg (1971), however, points out that new bom is not a suitable subject for volunteers or other un-trained personnel because the new bora hearing is a function of CHS maturity. She points the in-built danger of falacy in pass or fail procedure. She also points out that such inflexible test ess say nothing about the integrity of the 8th nerve or any other system. She emphasises the lack of basic research as its glaring drawback.

Thus in the face of proliferating new born screening programs, the peer showing, and waring consensus on the usefulness of the screening itself, a need for joint control and coordination of screening procedures was realised. The result was the appointment of a National Joint Committee on new born hearing screening whose main objective was to control and guide the research in this field. This perhaps changed the whole outlook of new born hearing screening.

THE PRESENT STATE - OF - THE- ART

Recommended screening procedures:

The joint committee reviewed the results of various programs and sought to halt all the mass screening programs. Following a conference on new born hearing screening hold

in San Francisco in 1971, it put-forth a set of recommen-
dations. In effect. it recommended selective screening
of those babies who may have a greater risk of developing
a hearing handicap.

The recommended program attacks the problem of iden-
tification from three aspects (1) The application of a
high risk register of all those babies at risk of having
or developing a hearing loss at birth of any time there-
after. (2) Application of behavioural screening method
or test, if perfected, as a supplement to the high risk
register, and (3) Follow-up screening of all those infants
in the high risk register.

In the present state-of-the-art, the high rial:
register is very well established, well supported by
research data and recognised as being effective in identi-
fying approximately 65 to 70% of those born deaf (Mencher,
1976, Northern ted Downs, 1974). In addition, a protocol
for behavioural screening has also been evolved. Beha-
vioural screening recommded is either an Arousal Test
(Mencher, 1974) or a semi-objective mechanical procedure

like the crib-o-gram (Simons and Rows, 1974; Simmons, 1976). However, both these methods are recommended only as a supplement to the high risk register. When children failing a behavioural test is added, the sensitivity of high risk register increases to nearly 80% (Mencher, 1977).

(THE HIGH RISK REGISTER:

The concept of high risk register was introduced to new born hearing screening by a paediatrician.(Hardy), The concept utilises history and/or evidence of physical abnormality to anticipate the likelihood for a hearing loss to occur or develop in any given child, its basic assumption is that deafness has a suggestive history or is accompanied by other demonstrable abnormalities. Thus any child who has a suggestive history or by his physical appearance suggests an abnormality, is at a risk. He is high risk infant.

In Mardy's concept a high risk register is an idea of registering every baby who is at risk, and carrying-out systematic follow-up every few months. Thus, it is a list of infants at risk. For the purposes of screening the concept assumes that "one can identify a small group of children whose history or physical condition identifies them as possessing a high chance of having the handicap searched for (Downs, 1978).

In the course of time, however, high risk register has assumed another meaning (Davis, 1978). The second meaning is the list of conditions that places the infant at risk. In case of hearing loss, there are a large number of factors that hare been associated with the handicap. However some thorough studies have shown that the greatest number of hearing-impaired children fall into only five or six categories of risk (The National joint committee has endorsed only these conditions for an effective high risk register (Gerber and Mencher, 1978). Presently, the high risk register consists of-

A) History of childhood hereditary imp airment.

B) Rubella or other non-bacterial intrauterine fetal infectious (Cytomegalorius infection, herpes infection)

C) Defects of ear, nose, throat, malformed, low set or absent pinnae cleft lip of palate (including submucous cleft) any residual abnormality of the otorhinolaryngeal system.

D) Birth-weight less than 1500 grams.

E) Billurubin level greater than 20 mg/100 ml serum.

F) Significant asphyxia associated with acidosia.

The high risk register should not be exhaustive, if it is to be effective. Longer the list higher will be the follow-up population and consequently less efficient it will be.Though a longer list can identify a higher number of deaf children it will enhance the cost and work load for the subsequent follow-up work.

According to public health specialists a high risk register, to be effective, must have a prevelance of the condition 14 times greater than that found in the general population (Richards and Roberts, 1967). Some of the programs have found a prevelance of one in 40 as compared with one in 700 in the general population, easily 14 times greater. Thus, the yield makesit a statistically accep-table approach (Downs, 1978).

Generally, the implementation of a high risk register requires some one to collect information required for high risk classification from various sources like hospital records oral or written interview of the mother using high risk checklists physical observation of the child, etc. such risk information is then classified and those children

categorised as at risk are followed op after a behavioral test or without it. In various places it has been conducted through trained volunteers and/or through public agencies and/or through private or community agencies (Downs, 1978).

OBJECTIVES OF THIS PROJECT:

Functionally, information required for high risk categorisation comes from three sources - history, medical records and physical observation or examination of the child either by an investigator or a physician. Historic information is collected from the mother by a querry, mostly about family history and rubella exposure. Rest of the information is gathered from the hospital records. Thus in most new born screening programs conducted elsewhere medical records form the chief source of risk information.

Conditions in our country are very different. Only five to ten percent of deliveries is India are medically supervised,mostly in big hospitals confined to cities and townships. Even in these hospitals there barely exists any system of maintaining detailed. Case records on every

birth. In many primary health centres babies are not
even weighed. Clearly, we cannot depend on medical
records for obtaining risk information *in* India.

Thus, we are left with only one source - history,
as given by the mother. History is potentially a very
important source. Most physicians to India agree that
history forms a very important source of information
for a functional diagnosis (Shetty, 1988). Moreover
most physical abnormalities found at birth associated
with deafness are quite evident even to a laymen. Thus
the mother earn very well report these abnormalities.
As far other conditions like maternal infections, asphyxia
and conditions resulting in the accumulation of bilurrubin
at birth makes themselves evident through their own
symptoms and signs. Hence, it is quite probable that the
mother can relate these signs and symptoms reliably, as
she does to a physician.

Thus, it appears that the mother could be the only
source of dependable if mot accurate information. But,
the validity of relying solely on the mother as the sources
of risk information is open for investigation.

Keeping in mind the importance of early identification
and easier modes of screening the population for hearing
loss. This project aims at screening the infant population
using a easier mode that is the high risk checklist. And
also to supplement the checklist findings with a hearing
screening test ie, the behaviour observation testing.

INTRODUCTION TO THE METHODOLOGY ADOPTED :

A list of high risk factors were compiled from lite-
rature, authorities active in the field and local medical
and allied specialists. A review of high risk programs
and factors appear in Chapter-II. Based on these factors
checklist had been developed already (See Chapter-IV).

Mothers attending the local medical college hospital,
immunization clinics, and well baby clinics were inter-
viewed. Data was collected through the checklist given
to the mothers there.

The infants were also subjected to behavioural screen-
Ing testing (subjective informal method). This was just
used as a supplement procedure for confirmatory purposes.

The data collected and the responses obtained were subjected to statistical analysis. Chapter-III describes the Methodology, while Chapter-V discusses the Results, Summary and Conclusions follow in Chapter-VI.

CHAPTER - II

REVIEW OF LITERATURE

' Screening' as accepted by World Health Organization is defined as "the presumptive recognition of unrecognized disease or defects by the application of tests, examinations and other procedures which can be applied rapidly" (Roberts, 1977). They are not intended to be diagnostic. Persons with positive or suspicions findings must be referred to specialists for diagnosis and necessary treatment (Wilson and Laugner, 1968).

The concept of high risk register was introduced to screen the hearing in the new born by Hardy, a pediatrician. Thus any child who has a suggestive history or by its physical appearance suggests an abnormality, is at risk. Such a child is considered as a high risk infant. (In Hardy[1]s concept, high risk register is an idea of registering every baby who is at risk, and carrying out systematic follow-up every few months (Downs, 1973) for the purpose of early identification.)

In the course of time, however, the high risk register has assumed another meaning, that is, it is a list of conditions that places the infant at risk (Davis, 1978).

A review of the high risk programs:
The beginnings of high risk concept

It was during the Toronto conference on the "Identification and Management of the Young Deaf Child" that the concept of "picking up children at risk" of hearing-impairment and to test them soon-after birth was introduced. During the discussions, Febritus of Norway mentioned of a new birth registration form which was about to be introduced in his country that could make possible such a procedure.

During the same conference, Hardy, a paediatrician pointed out that most of the eases of impaired hearing are found in particular groups of children who can be identified in advance on the basis of family background, the mother's pregnancy, conditions of delivery and events of immediate post natal period. The high risk concept was well received and subsequently the panel recommended in effect, that "A high risk register should be instituted listing those babies with a substantially higher risk than those in the general population and they should be followed closely and tested frequently during the first two years".

It was also pointed out that success of such a program will depend on the education of the physician, public health personnel and above all, the parents. Active involvement of pediatricians and obstructions, among other specialists was sought (Davis, 1964).

However, there has been a few efforts to mass screen children for hearing loss before. The John Hopkin's collaborative screening project screened nearly 400 babies, but the results were disappointing (Hardy, 1974). Meanwhile, the 1964 rubella epidemic in the United States gave a spurt to many mass screening programs throughout that country. Unfortunately, many of there studies overlooked the Toronto conference recommendations and ultimately were found passing some hard-of-hearing children (false negatives) and failing a significant number of normal children (false positives) (Gerber, 1971). With neonates they employed both the high risk register (items are not known) and a pure tone screen. Similar procedure were used with older children but the screening was done at 60 dB rather than at 90 dB. By the end of 1974, 10,000 new borns had been screened of whom 600 were not cleared (high risk?) This figure seems to be consistent with these reported elsewhere for the size of follow-up population. Among those children referred to public health agencies. Fifteen of 383 were not cleared (Hearing-impaired).

Bordley and Hardy (1972) study: An ancillary study of the NINCDS collaborative perinatal project conducted at Johns Hopkins Medical Centre by Bordley and Hardy (1972), it assessed hearing of 1182 children born of high risk mothers. They found that 98% of children failing at eight year test had given normal responses to the neonatal. In addition, they found that 5% of their sample had sensori-neural loss, 11.6% had conductive loss and 3.6% had mixed loss. They suggested that these high percentages may be a function of their high risk inner city sample.

THE NATIONAL JOINT COMMITTEE RECOMMENDATIONS:

Downs, the Chairman of the Committee, carefully analysed the available data wad very cleverly came up with a simple and a very efficient five point high risk register. She gave a monemic devise which she called the A.B.C.D.S. of new born nursery (Downs, 1972), which is gives below:

A. Affected family (congenital sensori-neural hearing loss in first cousins or closer).

B. Serum Billurubia level of 20 mg or more.

C. Congenital rubella (regardless of trimester).

D. Any observable defects of ENT (any first arch syndromes)

E. Small at birth (1500 gms or less).

Would have had identified 15 of them. That could have
reduced the follow-up population to only seven percent
saving much time, money and efforts (Mencher, 1974).

The Newzealand study: Started with the assistance of
Rational Audiology Centre, Audiland in 1972, this program
knows as the national Women's Hospital Program screened
17,250 children between 1972 and 1976. It employed a
hearing test and a nine month at risk screening program.
All children were tested within 1-2 days after birth or
before being discharged, by two technicians with no speci-
fic training in audiometry. The criteria of risk are not
clear (Graville and Keith, 1978), but, they presumably
constitute a broad tilt.

   Those who failed twice to respond to a warble tone
of 90 dB and 100 dB and also those at risk were followed
up at nine months. Of the 29 failed, only three were deaf.
73% were thus over referrals. Among ten deaf children
born in that hospital during that period and who were
followed up retrospectively, only one had been placed on
the high risk register though eight of them should have
been. Among the 1400 high risk infants 1000 were followed
up and only two were found to be deaf (Greville and Keith,
1978). This is a poor performance in view of the reported
efficiency of high risk register.

THE ELKS-PURPLE CROSS PROJECT:

The Canadian Benevolent and protective order of Elke and their auxiliary, the order of Royal Purple, both non-profit service organisation had implemented a project called a deaf detection of development program for early identification of hearing-impairment at Halifax, Canada. Children were examined in three age groups; 48 hours to one week; three months to one year; and nine months to one year. A high risk register was maintained and older children were seen in public health facilities or in coop-erating audiologic facilities (Alexander, coulling and Coulling, 1974).

This trend continued despite the findings of many studies. Downs (1968) recommended that only high risk babies should be screened, Eisenberg, Coursin and Rupp (1966) and Feld et al. (1967) had noted that differential responses can be observed if the new borns could be cate-gorized on the basis of risk. The fact that most of such programs were unco-ordinated made the matter more murky.

Finally, as a result of proliferation of such programs the ASHA invited the American Academy of paediatrics to

to form a National Joint Committee on infant hearing screen-
ing in 1969. The committee formed in 1970 was critical of
testing programs at that time and sought to halt such un-
coordinated projects. It formulated some guidelines after
a thorough review of available data.

Subsequently in 1971 San Francisco conference on new
born screening the National Joint Committee recommended a
screening protocol which actually id furcated early identifi-
cation into two distinct but not necessarily independent
areas: The use of high risk register and behavioural audi-
tory screening of the new boras (Mencher, 1974). Conse-
quently many high risk registers were devised for the pur-
pose of predicting those infants who have auditory and/or
other neurosensory deficits (Gerber, 1977).

THE EARLY PROJECTS;

Around the sane time of forming of National Joint
Committee , the maternal and child health services division
of United States public service department funded two
longitudinal research projects in Israel. Another project,
the Nebraska Neonatal project, founded by the National

Foundation (March of the Dimes) began in 1970. these early projects later were to contribute much to the refinement of the high risk concept.

*The M*aifa study : Between 1965 and 1967 this study screened nearly 10,000 babies with a very broad high risk register consisting of 25 high risk factors. It included such factors as first cousin matings, family history of deafness, imminent abortion, prematurity and jaundice, On extensive follow-up they could identify 13 deaf children but, only a fell into their high risk register. Deafness was two to three times common in the high risk population than in the general population.

The Jerusalem study: This longitudinal study screened 17,731 new borns between 1967 and 1970 with a broad high risk register consisting of early 16 items. It included many items used in the Haifa study. All children were also screened with the Apriton test of Hawns and Sterritt (1967). Those included in the high risk register and as well as those filing the Apriton test were again tested at 5-7 Months by stycar test, a modified form of Swing test. Both these tests were administered by trained nurses in the new born nursery or the baby clinics.

Children failing the sty car test twice within a month were later evaluated thoroughly at an Audiology Centre. Rest were screened again at 18-24 months using communication and verbal skill tests by trained nurses, A fourth and a last screening test assessing hearing communication ability in children was administered at around three years of age. these failing were thoroughly evaluated in both the instances.

By the end of 1974 this study turned up 23 profoundly or partially deaf children. Feinmesser and Tell (1974) concluded that a broad high risk register which covered about *20%* of entire new born population did not prove to be economical and practical. A much restricted register recommended by the National Joint Committee. With an addition of two items via, Apnea and cyanosis (Apgar score 1-4) and neonatal severe infection. Downs also pointed out that this restricted list would increase the sensitivity of the screening nearly tenfold. In view of the accumulating evidence from various projects, the National Joint Committee in 1973 further recommended the application of

high risk register and endored, with a few modifications, the Down's manifest as its criteria for high risk classification.   Supplementary statement of Joint Committee of Infant Screening (July, 1972).

The committee recommends that, since no satisfactory technique is yet established that will permit hearing screening of all new borns, infants AT RISK for hearing-impairment should be identified by means of history and physical examination.These children should be tested and followed up as hereafter described.

I. The criterion for identifying a new born as AT RISK for helping impairment is the presence of one or more of the following:

A. History of hereditary childhood hearing-impairment.

B. Rubella or other nonbacterial infranterine fetal infectious (eg. cytomegalovirus infection, herpes infection).

C. Defects of ear, nose or throat. Malformed, low set or absent pinnae, cleft lip or palate (including submucous cleft); any residual abnormality of otorhino laryngeal system.

D. Birth weight less than 1500 gms.

E. Bilurubin level greater than 20 mg/100 ml serum.

II. Infants falling in this category should be referral
for an in depth audiological evaluation of hearing during
their first two months of life and, even if hearing
appears to be normal, should receive regular hearing
evaluations thereafter at office or well baby clinics.
Regular evaluation is important since familial hearing
impairment is not necessarily present at birth but nay
develop at an uncertain period of time later.

These recommendations clearly reflect the growing
awareness of the need for a compromise between the effec-
tiveness of the high risk register and the cost of realis-
ing that effectiveness in terms of the size of follow-up
population and testing time. It also recognized the
importance of frequent follow-up checks, especially in
those children in whom hearing loss need not necessarily
be present at birth but may develop any time thereafter.

The Nova Scotia Conference (1974);

At about the same time the National Joint Committee
was providing structure for the direction of research
programs, the U.S. Government, the Elks-Purple cross and

other Government and private foundations were founding planned programs necessary to further research and to develop and refine early identification technique. Since there programs were conducted in many parts of the world communication between then was essential.

In order to bring all those engaged actively in such programs together and to arrive as a consensus, a conference was convened at Nova Scotia, Halifax, Canada with the assistance of Elks-Purple Cross Foundation. It brought together representatives from six nations who met for four days during September, 1974. During the deliberations in public and in closed door meetings the conference reviewed the accumulated data involving more than 150,000 babies.

The conference confirmed the effectiveness of the high risk register and recommended that it be universally implemented and urged the WHO, National and local Governments and health agencies to adopt this stage, if necessary by legal mandate. While re-affirming the role of follow-up checks, it also recommended the use of suitable behavioral screening tests as a supplement to high risk register. It also noted that those who fall is the high

risk register often suffer from other communication dis-
orders Which can further the usefulness of the high risk
concept.

The University of Colorado Screening Preject:

Supported by a national Foundation Grant this program
starting from 1972 began to apply a high risk register
using a core of trained volunteers. About 50 volunteers,
most of whom had been involved in several years of testing
of new boms and observation of responses joined the pro-
gran. The program followed a procedure which had three
parts viz.

(1) Maternal interview with questions concentrating on
    family history of hearing loss and rubella infection
    or exposure daring pregnancy. A specific question-
    naire was used.

(2) Review of hospital charts to collect data on birth
    weight, hyperbilirubinemia, neonatal infectious, ENT
    anomaties, etc.

(3) Continued screening of infants using the Vicon Apriton
    Test. The criteria for a pass was arousal or startle
    response.

Information on every new born was collected and a
risk category was assigned. Parents and physicians were
informed when a child fell into high risk group and
follow-up appointments were made. The following consti-
tuted the criteria for high risk classifications

1. Positive family history of hearing loss (before the
    age of five years) in parents and/or siblings.

2. Maternal rubella or rubella exposure

3. Congenital anomaly of the head or neck (cleft palate,
    microtia grossly abnormal pinnae, cleft lip) •

4. Neonatal meningitis

5. Birth weight of less than 1500 gms.

6. Unconjugated Bilurubin level of over 20 mg or an exchange
    transfusion.

As en 1977, the results showed (Gerkin, 1977) that out of
a total number of 10,727 births, 1,144 were classified as
high risk (one in nine or 10.7%) and 17 were identified of
having loss (one in 67 or 15%) four subjects suspected
hearing loss were lost to follow-up. Significantly all the
confirmed eases were classified as high risk and though, six
of them passed the Apriton test they were identified on basis
of high risk register. On an average, they were suspected
at 4.4 months and confirmed at nine months. The mean

suspected and confirmation age were 3,6 months and 6.5
months if those who did not turn up at advised time were
excluded. Gerkin (1977) sums the five year experience
with the following statement:

(i)   "Volunteers can do the required work in the nerseries.
       But, one needs some one to assume the primary
       responsibility and to coordinate the work.

(ii)   No attempt has been made to contact those not at risk
       and therefore little is known about missed deaf
       children in that population. Only one not-at-risk
       child has been referred back with a hearing loss. The
       incidence of confirmed hearing loss of all types
       significant for language development of 1:600 high
       risk sensitivity.is 1:80.

(iii) The follow-up response has been poor with only 30%
       keeping appointments, even after the repeat tests
       were made free of cost. This is probably because of
       the type of the population the hospital serves.
       Another private hospital in Denver with a similar
       program has been averaging a 98% return for repeat
       tests.

(iv)   The ideal time to screen infants for hearing loss
       is probably at the age of six months, at well baby
       clinics.

(v)   Letters and public education pamphlets have conside-
       rable educational value."

THE HALIFAX PROJECT:

A mass infant screening program was initiated in the
Grace Maternity Hospital, Halifax, Nova Scotia in Canada
in 1977. The program (diagram) incorporated the recommen-
dations of the Nova Scotia conference and utilised the
high risk register proposed by the Rational Joint Committee
and a behavioral test. All children listed on/the high
risk register as well as any child whose parent requested
a hearing screening evaluation were behaviourally screened.
Children op for adoption and some children falling under
specific investigation categories were also behaviourally
screened as part of ongoing research.

Every mother admitted to the hospital received a
packet of material containing, among other things, a letter
from Nova Scotia hearing and speech clinic which informed
her about the alms and procedures of the program. She was
asked to fill in a simple questionnaire and to provide
additional information regarding the family and the baby.
The questionnaires wore collected, answers verified and
medical record checked for birth weight, first arch syndromes
and bilirubin count by a part time staff person.

All children considered for behavioral screening were tested according to a set protocol. Ho child below one day in age was tested. Any child failing this test was re-tested within 24 hours. Failure on the second test meant immediate and automatic referral for a full audiological and otologic evaluation and follow-up. This examination was considered a part of routine hospital care, very much like the investigatory x-ray and was covered by the initial blanket permission signed by the parent. To avoid unnece-ssary trauma to the families the parents were not even involved in the program until after the full audiological test. Counselling and follow-up appointment were deferred till then.

The family doctor was then posted with the details of results and placement on the high risk register and was requested to provide specific follow-up on high risk children. The visiting nurse from Nova Scotia public health department was also provided with all information and they in turn, provided additional screening at hone and assisted in follow-up as and when needed.

The accumulated results of the program are not yet available. However, according to the yearly report

(Mencher,et al. 1930) in 1979 the centre screened 491© babies of whom 669 were high risk. (The high risk register was essentially the same as national Joint Committee has recommended with as-phyvia included on the recommendation of the saskatoon conference, 2-6 below). They constituted 13.6% of the new born population. In addition 373 babies in the intensive care unit (ICU), 119 children up for adoption and 325 babies meeting other special research needs, were tested with the Arousal test (Mencher, 1974)

(Diagram - see in next page).

   The testing was done as outlined by the Nova Scotia protocol. Eventually 110 infants were referred for detailed evaluation. 70 of them were cleared after the initial visit. Of the remaining 31, 8 were definite failures while 23 were still questionables. Subsequently 15 of the 23 have been cleared and eight were still pending.

   Among the eight definite failures, I had confirmed sensorineural loss and five conductive hearing loss. However, it WAS not sure if any of these conductive hearing loss eases had a sensorineural component as well. As Meneher et al. (1980) noted "It is quite possible that any or all seven of

SCHEMATIC DIAG. OF NEWBORN HEARING SCREENING AND DEAFNESS DETECTION PROGRAM

**High risk screening
(identification)**

Joint Committee	SMCH	Adoption	Special request

Behaviourally screened

Fail	Pass	Discard

Rescreen

Family Doctor
& Health Dept.
notified   Fail   Pass   Family Doctor
& Health Dept.
notified

Audiology-Otology
(Diagnostic)

Fail/? Pass   Pass

Additional Audiological testing   Pass

Follow-up   Fail

Confirmed   Hearing loss

Rehabilitative perform-
ing (diagnostic)

Medical   Audiological   Family/Child

them may develop a sensorineural hearing loss later on,
something which has been reported to occur with children
exposed to rubella and other viral infections. However,
it should be noted that all three of the confirmed hear-
ing loss cases were on the high risk register, one being
a case of severe Asphyxia and the other two being low
birth weight babies.

As part of the ongoing research, the centre also
screened all children admitted to ICU at another hospital
using a crib-o-gram. However, no high risk register was
considered. It picked up four deaf children among 158
tested and 23 failed initially. When the loss was con-
firmed all the four were less than three months old. Inte-
restingly, all the four could have been placed on the high
risk register. That means that all the seven deaf babies
identified in Halifax last year were on the high risk
register (Mencher, et al. 1930).

THE UTAH HIGH RISK PROGRAM:

This project actually began in 1967. If followed a
model which facilitated data collection with minimum
hospital and/or professional participation and at a time

when it was easily obtainable on the majority of the target population. The goal was to screen all the babies born in Utah hospitals Which comprised of 98.9% of the total number of births in that state. (Mahoney and Eichwald, 1979).

The seven item questionnaire incorporated the following factors: hereditary deafness, rubella exposure, birth weight, ENT defects, Rh factor requiring blood transfusion, severe neonatal illness and parental concern. Since the respondent was the mother it was so designed as to make it easily understood by all. It also included a question on neonatal severe illness and one on parental concern. The program protocol consisted of eight basic steps (Mahoney and Eichwald, 1979), viz.

(1) High risk questionnaires were sent to the hospitals from the Speech Pathology and Audiology section of the state division of health.

(2) Questionnaires given to mothers for completion along with birth certificate. Also included a covering letter explaining the program and an information leaflet that outlined the normal auditory development.

(3) The questionnaires were accumulated and returned to the section at regular intervals, by the hospital staff.

(4) the returned questionnaires wore immediately dichotomized into high risk or not high risk.   A positive response to one or more, items constituted a high risk determination, as did failure to complete any item.

(5) When the high risk child was between six and eight months of age, the mother was sent a follow-up questionnaire that included the original questions plus two additional questions regarding her child 's auditory behaviour; "When your child is in light sleep in a quiet roomdoes he move and begin to wake up when there is a sudden noise?" and "Does your child turn towards an interesting sound or when his name is called?.

(6) When auditory behaviour reported by the mother was found questionable or when parental concern did exist, either an audiological evarluation was arranged or educational literature was mailed to parents followed by another telephone.

(7) Parents who desired an audiological evaluation were asked to bring their children to one of the three regional clinics that had sound isolation test environment. When found necessary the initial screening was accomplished at one of the state-wide in ten er ant clinics. In both cases hearing and middle ear assessment was accomplished by

certified audiologists. Periodic follow-up procedures were per formed as advised by the National Joint Committee. Brain stem evoked response evaluation was also arranged for the difficult to test.

(8) Hearing aid evaluation, medical consaltations and family physician contact was initiated with infants found to be hearing-impaired. Referrals for habilitation was made preferably before or by the time the baby was one year old. The parent infant program (PIP) of the Utah School for the deaf usually became involved at this time. Parent advisors visited hone on a regular basis and trained parents in hearing aid management and in methods to develop language skills in their children.

As reported (Mahoney and Eichwald, 1979) the results show that of the 50,7OO births between January 1, 1976, and December, 30,1976, 26,352 (52%) completed questionnaires were recevied. 4,591 (17.4%) were classified as high risk on the first inspection, ie. one or store of the seven items were marked positive or left blank. Of there 181 (3.9%, of the high risk) remained at risk after the follow-up contact and 54 infants (29,4% of those at risk) were found to be

hearing-impaired by audiological evaluation. There were in all 711 false positive questionnaires consisting of inaccurate responses that mistakenly identified the baby as high risk. Typically such responses involved a presby-cusic relative in the family history category.

The program relied heavily as not only the accuracy of the parents response to questions but also on this ability to assess their baby's early auditory behaviour,

THE UTAH STATEWIDE INFANT HIGH RISK HEARING PROGRAM:

This pilot program was instituted in 1978 after much search for an alternative to hospital material and staff as the source of high risk data. This utilized the birth certificate as a means of obtaining information about high risk hearing factors.

The Utah Live Birth Certificate has two sections, designed for health and medical use - one to be completed by the parents and the other by the physician supervising the birth. (Mahoney, 1980). It contains the following items pertaining to high risk register; complications of pregnancy, current illness. Or condition affecting pregnancy Apgar score, birth weight, and congenital malformations.

Since all items are computerized it was relatively easy
to generate a computer program for the project, the
speech pathology and audiology section receives a monthly
readout from the state bureau of vital statistics contain-
ing the names and addresses of all infants with one or
more high risk factors and an item analysis of each risk
category. The program has established a set protocol
(see fig.)

Master Birth Tape

|

High risk

|

Questionnaire No response
Response
|

| |

At risk Not at risk

|

Testing

| |

Hearing impaired   Normal

|

Habilitation
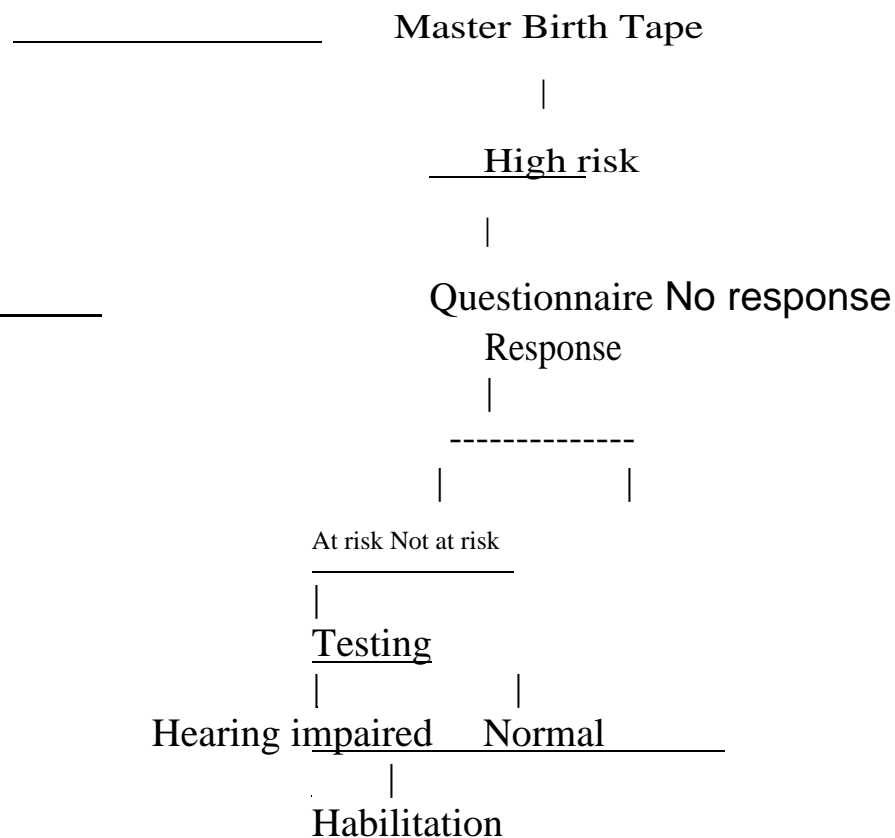
When the high risk infant is six to eight months of
age, a questionnaire is mailed to the parents, which contains
two questions concerning normal auditory development viz.

(1) When year child is in a light sleep in a quiet room, does he move and begin to wake up when there is a sudden noise? and (ii) Does your child turn towards an interesting sound, or When his name is called?. A third question allows the parents to express their concern regarding their child's hearing. Along with the questionnaire an information leaflet on normal auditory development is also mailed.

If the questionnaire is not returned no further action is taken. From the returned questionnaires at risk determination is made on the basis of auditory or parental concern. The remaining procedure is the same as in the hospital program described earlier.

Initial data analysis has indicated that of a total population of 21.109 infants born in the first six months of 1978, 5647 infants (26.9%) were considered high risk by the present criteria. Item analysis revealed that 13.2% of population answered positively to "complications of pregnancy". A sample analysis of 500 high risk birth certificates was run and it was found that more than 98% of medical conditions listed under complication of pregnancy

were not pertinent to hearing risk according to the National Joint Committee criteria. It was then realized that the permanent inclusion of this item would weaken the sensitivity of the birth. The question is now eliminated as a high risk item. The revised data projected a high risk population of 13.7%. which is close to the 7% population. Sensitivity reported by Northern= and Downs (1974) (Mahoney and Eichwald, 1979).

It is now proposed that, if proven successful/ the BC should permanently replace the hospital questionnaire which should improve both initial screening rate and program efficacy, it would then realize the promise of screening nearly 100% of the state's new born population so ardently recommended by the National Joint Committee.

THE SANTA BARBARA PLAN :

The Senta Barbara Unit of Speech and Hearing Sciences had been involved in a pilot screening program till 1978. The mother was required to complete a questionnaire before the child is born with the help of a obstetrics nurse. It was then completed after birth by nurses in delivery room and nursery. The questionnaire were then verified by volunteer graduate students. The primary care physician

as a paediatrician was then posted with details and we entrusted the follow up work. After an initial family contact the infant was screened for an arousal. A second failure in this entailed the child to a detailed behavioural and electrophysiologic tests.

Meanwhile, a conference of infant auditory assessment was convened in Santha Barbara in February, 1979. Its overall objective was to assist the maternal and infant health section in formulating guidelines for auditory screening along the lines of those existing for visual, neurological and pulmonary disorders, the conference concerned with the consensus arrived at all the precious conferences that there is no universal auditory screening test what is both cost effective and diagnostically effective. Hence they reaffirmed the validity of the high risk register and recommended that a high risk registry be set-up in the state of California. They agreed that all infant should be risk rated as follows:

(1) All with a family history of childhood hearing-impairment.

(2) All with cranio-facial anomaly.

(3) All who have confirmed disease by TORCH (ie. Toyoplasmosis, Rubella, cytomegalovirus, and herpes), and

(4) All side enough to have been in a teritiary ICU and some of those discharged from a secondary intensive care nursery.

The conference also proposed that "all high risk infants, as defined above are to be sent for definitive diagnosis to centres specifically certified for that purpose.   Since the families of such infant also need ancillary services like public health nursing social services, nutritional and health education support etc. there services (hearing evaluation) should be built within the care program".

To ensure a definitive diagnosis, they recommended, "initial contact is to be established by too months post discharge or at three months post discharge or at two months adjusted postnatal age.   Definitive assessment on hearing sensitivity should come by five months after the date of definite diagnosis.   It is an unacceptable practice to defer a definitive beyond this time arrival, considering both lost benefits to the hearing-impaired infant and the state of art in establishing a diagnosis by this time. The procedure should include evoked potential screening or complete evoked response audiometry or total auditory evaluation (Gerber, 1979).

The conference also considered the possibility that
an automated behavioural test might be employed in all
teritiary ICUs and perhaps in secondary care nurseries.
In that care, all infants who fail in that test should
be referred to the centres for the definitive diagnosis
as just defined. The conference also recommended that
12 geographically distributed centres should be established
in the state of art of California to serve all the high
risk infant as defined above (Gerber, 1979).

HIGH RISK REGISTER JUS AN ADJUNCT TO BEHAVIOURAL SCREENING
AND RESEARCH:

The high risk register has come to be recognised as
a very useful tool in selecting the test population in
behavioural screening and machine aided diagnostic proce-
dures like BER procedures. Mencher (1977) and it is an
adjunct to validate crib-o-gram and found it a valid method
of differentiating infants with severe impairment from
normal children. Be also noted an abnormally high percent-
age of mental retardation, cerebral palsy, childhood
aphasia and other associated speech and language problem
in the group with normal hearing but which is high risk and
has failed on the Apriton behavioural screen (Mencher,1978).

Galambos (1978) suggested a protocol on "how to test almost every neonate with peripheral hearing loss". It proposes, in effect, selection of candidates from (1) ICUs (except those previously tested and cleared) (2) High risk register should be maintained ia every new born nursery (3) Those who fall behavioural test, and (4) Those suspected for any other reason. He concludes that "Only rarely will a hearing-impaired one is diagnosed as normal".

Mendel (1978) found middle evoked potential testing particularly useful in conjunction with a high risk register. He reported of a project in Santa Barbara where high risk questionnaire is employed in two of the local hospitals to determine the cases at risk to be tested by a behavioural screening method and if they fail in that test are scheduled for electro encephalic audiometry.

High risk registry has also served as an adjunct to the study of early vocal behaviour of deaf infants, One Pilot study in Memphis, has indicated the possibility of deaf infants being identified through cry-spectrographic prints.

Ashok (1980) carried out a feasibility study in Mysore population as as attempt at utilizing a questionnaire

to collect risk information Which can be used for risk categorisation for hearing loss children. The question which was developed was administered to mothers of children. And he found that the questionnaire could be effectively used to collect risk information.

REVISED HIGH RISK REGISTER (1982):

The Joint Committee on New Born Hearing met again in 1981 to revise and expand the original five-point high risk criteria. The 1981 ABCDs - of deafness are as follow:

I. IDENTIFICATION:

A. Risk criteria:

A - Asphyxia which may include infants with APGAR scores of 0-3 or those who fail to exhibit spontaneous respiration by 10 minutes and those with hypotonia persisting to two hours of age.

B - Bacterial meningitis, especially H-influenza.

C - Congenital perinatal infectious (eg. cytomegalovirus, rubella, herpes, toxoplasmosis, syphillis)

D - Defects of the head or neck (eg. craniofacial syndromal abnormalities, overt or submucous cleft palate, morpho-logic abnormalities of the pinna).

E - Elevated bilirubia exceeding indications for exchange transfusion.

F - Family history of childhood hearing-impairment.

O - Gram birthwetght less than 1,500.

Katherine Pike Gerkth(1982) has summarized the risk factors and their most common effect on *the* hearing mechanism. However, this categorization is merely a guideline and each risk factor should be viewed as any individual is viewed - unique, variable and with endless possibilities.

High risk factor	Most common manifestation of hearing loss					
	Condnc tive	SN	Mixed	Unila- teral	Bila- teral	Degree
Asphyxia					+	Mild-pro-
Bacterial Manin- getis		+			+	Sev-Pro
Toxo		+			+	Mod-pro
Syphillis		+			+	Sev-pro
Rubella		+			+	Pro.
CMV					+	Mild—pro
Herpes		+		*7*	?	*7*
Defects of head & neck	+		+	+	+	Mild-pro
Elevated bilmribin		+				Mild—pro
Family history	+	+	+	+	+	Mild-pro
Low birthweight					+	Mod-ser.

SN=sensorlneural ; Pre -profound; Sev-severe; Mod-moderately

But repeated follow-ups were recommended.

The Joint Committee on Infant Hearing position statement also makes recommendations for the evaluation and treatment of the hearing-impaired infant.

I. Screening procedure:The hearing of infants who manifest any item on the list of risk criteria should be screened, preferably under the supervision of an audiologist, optimally by 3 months of age but not later than six months of age. The initial screening should include the observation of behavioural or electrophysiologic response to sound. If consistent electrophysiologic or behavioural responses are detected at appropriate sound levels, then the screening process will be considered complete except in those eases in which there is a probability of a progressive hearing loss, eg, family history of delayed onset or degenerative disease, or history of intrauterine infection. If results of an initial screening of an infant manifesting any risk criteria are equivocal, then the infant should be referred for diagnostic testing.

II. Diagnosis for infants failing screening:

A. Diagnostic evaluation of an infant six months of age should includes

i) General physical examination and history including:

a) Examination of the head and neck

b) Otoscopy and otomicroscopy

c) Identification of relevant physical abnormalities

d) Laboratory tests such as urinalysis and diagnostic tests for perinatal infectious.

2) Comprehensive audiologic evaluation:

a) Behavioural history

b) Behavioural observation audiometry

c) Testing of auditory evoked potentials. if indicated.

B. After the age of six months, the following are also recommended:

1) Communication skills evaluation

2) Acoustic immittance (Impedance) measurements

3) Selected tests of development.

III. Management of hearing:

Habilitation of the hearing-impaired infant may begin while the diagnostic evaluation is in process. The Committee recommends, however, that whenever possible, the diagnostic process should be completed and habilitation begun by the age of six nonths. Services to the hearing-impaired infant less than six months of age includes

A) Medical management

    1) Reevaluation

   2) Treatment

   3) Genetic evaluation and counselling When indicated.

B) Audiologic management

   1) Ongoing audiologic assessment

   2) Selection of hearing aid(s)

   3) Family counselling

C) Psychoeducational management

   1) Formulation of individualized educational plan

   2) Information about implications of hearing-impairment.

Studies carried-out supplementing the BOA:

   The desirability of early identification of hearing loss
in infants led to experimentation with mass auditory screen-
ing of neonates. Large scale efforts to detect hearing loss
through neonatal audiometric screening got underway in the
1960s (Downs and Sterrit, 1964, 1967; Downs and Hemenway,
1969) and since that time the concept of neonatal hearing
screening programs has been very popular. Most screening
protocols were based on the format developed by Downs. The
screening procedures involved monitoring of changes in the
infant's state following presentation of high intensity
signals often in excess of 90 dB sound pressure level (SPL).
The expected response from there behavioural procedures
included reflexive activity, marked movement of arms. Or

legs, eye widening, eye blink, around from sleep, or any
combination of these responses. Clinicians and researchers
now agree that these procedures for screening neonates are
sensitive only to hearing losses of about 75 dB or greater
(Mencher,1974, Northern and Downs, 1974).

The Joint Committee addressed itself to the use of
behavioural tests of screening in the new born nursery,
which had been previously proposed. The committee issued
a statement which did not recommend mass behavioural screen-
ing, although it argued increased research efforts, Gene-
rally, the committee agreed that screening programs were not
identifying deaf infant very successfully, and that the
large number of false positives were not only time and cost
ineffective, but were also causing unnecessary parental
anxiety. The inefficiency of mess screening, coupled with
the fact that many of the infants identified by testing
would also havee been identified by some means of an "at-
risk" register, led to the suggestion that a high risk for
deafness be identified with a prenatal history and post-
natal physical examination.

Feinmesser and Tell (1971, 1976) initially used a high
risk segister which designated *20%* of the test population
as at risk. A total of 17,731 new boms were screened with
various methods over a period of 37 months. At birth the

Infants were given behavioural screening and were also subjected to a high risk categorization. Almost all of the children were again tested in public health clinics at the age of five to six months, again at eighteen to twenty four months, and finally at three years of age. Twenty-three deaf children were identified by the end of the program/ 17 of them had been on the high risk register; only six had been identified by behavioural screening.

The results of this study led to conclude that the conventional procedure of observing motor responses of awake infants was not sensitive enough to detect deafness in neonates. That is the high number of false positives (367) and false negatives (17) indicated that their behavioural screening procedures were ineffective and invalid.

Based on the high number of children needy follow-up care (20% or 3,546 children) from the high risk register, Feinmesser and Tell expressed some concern about their register containing such a large number of risk factors. Through modification of the register, they reduced the number of neonates included to six to seven percent *(6-7%)* of the population and still retained a large percentage of the deaf neonates within the register.

Behavioural Screening and the High Risk Register;

Mencher (1974a) detected bearing loss in neonates through a combination of a high risk register and a behavioural screening procedure. Initially, 10,000 infants were seen and 60% were followed up for a period of two years. The follow-up evaluations confirmed nine babies, seven Could nave been identified by screening only (use of narrow band noise to arouse the infant from light sleep), and five would have been detected by the High risk register. Two babies had passed both the behavioural screening and a high risk classification. Mencher concluded that both behavioral screening and a high-risk register are necessary for neonatal screening.

Downs (1976) reported results that demonstrated impressive agreement for detecting hearing loss in neonates through a combination of behavioral screening, five were found to have auditory impairment. Those five were identified by both behavioral screening and the high risk register. Downs used the register proposed by the Joint Committee on Infant Hearing Screening. Additionally, she used an arousal response from a light sleep.

It appears from the Down's study and the report by Mencher (1974b) that an arousal response from a light sleep

is a more valid response than reflexive behaviour from awake infants, and that the use of this criterion results in fewer false positives when identifying hearing loss in neonates. when motor responses are observed from awake infants, (Feinmesser and Tell, 1971). The false positive rate is increases tremendously and behavioural screening becomes inefficient as a screening procedure, Hodgson (1973) concluded from a review of literature that behavioural screening of neonates is valid and efficient only when the babies are tested in a quiet room and arousal from light sleep is the criterion response.

The international conference on early identification of hearing loss (Mencher, 1976) recommended the Joint Committee five point register for deafness be adopted. It also recommended that neonates at risk for deafness should receive individual behavioural screening. And behavioural screening was considered a supplement only when certain standards could be maintained. The conference recommended that the lnf art be asleep prior to testing. They also recommended that the test stimulus be a predominantly high frequency complex signal with a sharp rise time, a maximum SPL of 90 dB, and a durationof one half to two seconds with an interstiraulus interval of at least 15 seconds. The acceptable response is generalized body movement involving more

than one limb and accompanied by some form of eye movement.
Two or more responses out of eight signals represents a
passing score. One of two scoring criteria was suggested
for use -

1. The observer should not know when a signal is presented
   and thus has to make a "blind" decision; or

2. That two observers score the infants response indepen-
   dently. Ambient noise levels during testing should be
   reported and it is recommended that testing not be done
   in intensity levels exceeding 60 dB SPL (Northern and
   Downs, 1974).

When there guidelines are met, behavioural screening
and a high risk register go hand inhand in neonatal hearing
screening. If the guidelines of the International Confe-
rence cannot be met, the evidence suggests that the screen-
ing program will not be efficient or valid and the use of
a high risk register alone would be a more appropriate tool
for identifying possible hearing-impairment.

The 1982 position statement by the Joint Committee on
infant hearing recommends that infant at risk for hearing
impairment be screened by 3 months of age and that the
diagnostic process be completed and habilitation begun by
six months of age. Kraus (1983) studied how close to tine

ideal actual practice comes in an urban setting. Data on 88 infants referred to a hospital basal parent-infant program were retrospectively examined to determine the occurrence of risk factors and at What ages (1) hearing loss was first suspected, (2) hearing loss was diagnosed and (3) habilitation was initiated. Results indicate that over one-quarter of all hearing-impaired infants will not manifest any of the risk factors proposed in the 1982 position statement and that regardless of Whether the infant graduates from a neonatal intensive care unit or well baby nursery, the median age for enrollment in a parent-infant program is a year or more later than the 1982 recommendation.

Never   too   young   project   (1987):

A sub-committee of this project, made up of audiologists, neonatologists, and otolaryngologists, developed a question-naire that was distributed to 300 parents of hearing-impaired children who were then residing la Arizona. The questionnaire included questions pertaining to the identification and inter-vention process that the parents had experienced, and to the children* s birth and medical histories.

This projects was undertaken to develop uniform neonetal screening programs in Arizona. A survey of the 159 completed questionnaires yielded there finding:

1. Of the infants in tails survey, 79% hour had congenital
   hearing loss versus 21% with acquired hearing loss.

2. Approximately one half of the babies with congenital
   hearing loss would not have been detected by the
   current high risk register.

3. Average age of identification had been approximately
   19 months, regardless of whether the infant was high
   risk or not at risk.

4. An inverse relationship existed between degree of hear-
   ing-impairment and age of identification, that is, the
   more moderate the impairment, the greater the risk of
   delayed identification.

5. Hearing loss in the babies born since 1982 had been
   identified earlier than for those babies born in the
   1960s.   ,

   As a part of infant hearing screening Holly Hosford
Dunn et al (1937) found congenital and early onset hear-
ing losses in 6.1% of 975 intensive care nursery (ICN)
graduates. The method used were neonatal screening by
crib-o-gram (COG) and high risk/register, in combination
with repeated behavioural hearing tests at 1 to 3 years.
This 7 year longitudinal study had follow up hearing
evaluation for a remarkably high 84% of all subjects.
Significant losses that interferred with speech and language

development (1000 to 8000 Hz average too greater than
45 dB HL bilaterally) were found in 4.3% of infants.
Behavioural hearing screenings detected bilateral hearing
losses of even mild (greater than 20 dB HL) degree. Sensi-
tivity to significant hearing losses was 82.6% and would
have been improved if test frequencies greater than 3000 Hz
were included in the screen. Even if screening failure
occured at one year of age, the age of actual confirmation
of hearing loss depended on severity of the loss and ear
involvement.

Jerry Halpern, Holly Hosford-Dunn and Malachowski
Natalie (1987) studied the four factors that accurately
predict hearing loss in 'high risk' neonates. Findings are
based on univariate and mrultivariate analyses of a number
of variables that night be associated with permanent hear-
ing loss. Study variables included all seven high risk
register item and a number of other features of the inten-
sive care nursery history. They were examined in 799 ICN
graduates whose hearing had been monitored in their first
few years of life. These babies composed of 40% of the
ICN population and were selected because they had one or
more high risk factors in their neonatal history. The four
factors that predicted hearing loss with 98% sensitivity was -

- Craniofacial anomalies

- TORCH infectious

- Length of stay in the ICN

- Gestational age

Nancy Swigonsti at al (1987) did a prospective screen-
ing of an extremely high risk group of 137 infants cared
for 1B the new born. Intensive care unit of James Whitcomb
Riley Hospital for children was undertaken during 1983.
Auditory brain stem responses were obtained utilizing a
clinical evoked potential system (Madsen 2250) . Patients
were selected for screening prior to discharge or transfer
to the referring hospital on the basis of one or more of
the following criteria; birth weight less than 1250 grams,
birth weight less than 1500 grams and ventilatory support
significant depression at birth (Apgars less than 3 and *6*
at one and five minutes, respectively) . Seizures, meningetis,
and/or Seplis. Of the 187 infants tested, 82 passed the
initial auditory brain stem response, 22 conditionally
passes, and 34 failed. Eighty two infants had follow-up
behavioral and audiometric testing while 20 infant died and
35 were lost to follow-up. Four infants had severe sensori-
neural hearing loss, each of whom had failed the initial
auditory brain stem response. High risk factors for sensori-
neural hearing loss in the neonatal period included: intra-

ventricular/periventricular hemorrhage, apnea, family
history, major malformations of the head and neck, and
possibly hyper bilirubinemia and congenital infection.
No relationship of sensorineural hearing loss with very
low birth weight, hyponatremia, infection, seizures, or
medications was found. On the basis of these data, it
was suggested that electrophysiologic hearing screening
of a high risk population may be delayed until three to
six months of age to improve specificity of testing.

A Community based high risk register for hearing loss:

A high risk register was established cooperatively
by the Brescia College, Hearing impaired project and the
Owenshare-Davien country Hospital. The study was/taken
under
by Fitch, Williams, Etienne (1987). Follow-up testing of
children identified as being high risk for hearing loss
was accomplished through the Brescia college Speech and
Hearing Clinic (ie. tympanometry and observation of loca-
lization responses). The results for the first year of
operation, including a six months follow-up of all children
show that, Of 1973 infants screened, 166, or 8.3% were
found to be at risk. This percentage compares favourably
with other studies, Northern and Downs (1978) reported
6,9% and McFarland, Simmon and Jones (1980), 10%.

Eightynine of 166 children identified as high risk were seen for a screening test. It was found that 19 (21%) of the 89 children failed at least one screening in *the* first year.

Laszlo K Stein et al. (1990) did a study which was a follow-up of an 1980-1982 study that examined the occurence of risk factors and the patterns of identification and habilitation in a group of hearing-impaired infants from an urban setting. The findings indicate that only one out of three hearing-impaired infants can be expected to be identified through audiological screening programs in Neonatal ICUs and although the age at diagnosis for Neonatal ICU graduates is significantly earlier for well baby nursery (WBN) graduates, age at enrollment in a parent-infant program for both neonatal ICU and WBN infants is around 20 months. Over the eight year period covered by an two studies, the age hearing-impaired infant are enrolled in habilitation has remained a year or more later than the six month ideal recommended in 1932 by the Joint Committee on infant hearing.

Mac, Wallar, Whan, Stelmachowica (1991) did a study and examined factors which may affect early identification of hearing loss. The medical records of 123 children with

educationally significant hearing-impairment were examined. Information about each childs degree and type of hearing loss, etiology, referral source, birth end medical history, additional handicaps, age of suspicion of less, node of identification, age of identification and age at which aided was entered into a data base for further analysis. The age range for identification was seven weeks, to 10 years with a median age of 2.1 year. Children with a greater degree of hearing loss, an additional handicap,additional medical conditions, or an etiology strongly associated with hearing loss, were identified earlier than those without there factors. Unexpectedly, children with a history of middle ear dysfunction were identified no later than those without and children with a positive family history of hearing loss were identified later than those with a negative family history. There results agree with other studies which show that, in general, children are identified and habilitated at a later age than that recommended by both the American Speech Language Hearing Association Committee and Joint Committee on Infant hearing.

The Joint Committee on new born hearing position statement(1990)

It recommended that the linguistic process for hearing loss be completed and rehabilitation begun by the age of six months.

The utility and short-comings of high risk registry;

    In general, employment of the high risk register
has proved to be fairly productive. Reports of its
success have been shown by Hencher (1976). Stevert
(1977), Rossi and Guidoti (1976), Mahoney (1977)& (1979)
among others. Only Mayer and Wolfe (1975) have had
limited success as did Greville and Keith (1978).

    Downs (1976) reported of finding one deaf child
in 57 listed in her high risk register. Hencher (1974)
applied the five national Joint Committee items retro-
spectively to data from a number of sources and found
that the five item register would have correctly detected
about 66% of true positive eases. In general, it is
observed that the five item list include about 6 to 8%
of the new born population, and 2 to 4% of the high risk
population will prove to have a hearing loss and of these
perhaps half will be severely impaired cases (Gerber, 1977).

    The high risk register has succeeded when behavioural
methods fail. Mencher (1974) found that it leads to much
higher correct detection in the new born nursery than does
the use of various screening methods. One can recall that

in Jerusalem study 17 hearing-impaired passed the five stage behavioural screen. Findings of the Newzealand study further stresses the role of high risk register When correctly applied. So hearing-impaired child in the high risk register passed the 9th mouth behavioural screen.

Gerber (1972) and Meneher (1974) found that those who are at risk and those who fail to respond to intense acoustic stimuli frequently have neurosensory deficits, ether than deafness like mental retardation, cerebral palsy, childhood aphasia, an interesting side benefit of the high risk register those at risk and who are sot deaf form a very intruguing group who merit intensive study and follow-up.

However, the implementation of high risk registry is not without its own problems. The most often sited areas of difficulty are:

i) Continued professional contact with each hospital has proved to be time consuming and lumbersome procedure.

ii) Hospital staff changes adversely affect continuity of the program, especially, questionnaire delivery and retrieval.

ill) Heavy work load of most drawbacks is a major drawback.

iv) Since most of the programs are non-voluntary, preventive health programs, certain amount of compliancy on the part of the hospital staff has to be taken for granted.

v)   An equal amount of, if not greater than, complerency
on the part of the parents in returning -the question-
naires is also to be expected.

vi)   Limited hospital stay of the mother decreases the
population of mothers in terms of opportunity to
complete the questionnaire.

vii)   Initial has availability of certain groups of children
like those is the ICUs, most of whom are initially
lost to the high risk registry.

Till) Many columns in the returned questionnaires are either
left blank or contain false positive information.

ix)   The transient nature of the population in many places
makes follow-up difficult.

In spite of these difficulties the high risk registry
has proved its feasibility because -

1) it enhances the cost efficiency of the screening proce-
dure by virtually by-passing tine new for mass biologic
screening. This is very important asset since the
incidence of deafness in the general population is low
(about one in 1000 to one in 2000) (Gerkin, 1977).

2) the population of mothers of new born is easily available
in hospitals, well baby clinics, etc.

3) 75 to *90%* of all children Who eventually incur
   hearing loss could be listed on a high risk register
   (Downs, 1969).

4) High risk information can be obtained through a simple
   questionnaire, and where possible it can be obtained
   relatively easily through legal documents like the
   birth certificates.

5) It can make possible *100%* screening rate, especially
   when it can be made mandatory without making it cumbersome.

6) Since high risk registers often include those children
   who would eventually suffer handicaps other than deafness,
   its value can be immense.

7) It has proved as a very useful adjunct to research involv-
   ing obtain diagnosis and management of not only hearing-
   impairment but also other handicapping conditions.

THE OUTLOOKS: Though high risk registers have been found to
be very effective. But the most effective way to comply with
the Joint Committee goals is to supplement screening measures
also (eg. otoacoustic emissions or ABRs) that can be used
to evaluate newborns before hospital discharge (Stevens, Webb,
Hutchinson, Connell, Smith and Buffin, 1990; Norton and
Wilder;, 1990).

<u>METHODOLOGY</u>

<u>DATA COLLECTION AND ANALYSIS;</u>

Data collection for the study was carried in two ways: Interview by the investigator; and written querry of mothers.

<u>Investigator interview:</u>

The investigator sought the permission of various hospital and nursing home authorities to interview the mothers. Finally three locations were selected. The local medical college hospital for women and children , a well baby clinic (private nursing home), and an immu-nization clinic (JSS Medical College).

In the Medical College three locations were made use of for tine purposes of interviewing viz.

(1)   The post-partum clinic - This also houses the well baby clinic where children are immunized. Bulk of the data was collected from this place.

(ii)   Pediatric OPD for fresh cases - The investigator made use of this location whenever the post-partum clinic was dosed or was too crowded. Doctors attending were requested to divert a random sample for interviewing.

(iii) Pediatric ward - This location was chosen because of three reasons:
a) Mothers were wore accessible here,
b) They were more free and were not in a hurry, and
c) It suited the investigators free time. Beds were chosen randomly and their present histories were discarded from the purview.

The well baby and the immunisation clinic cared for mostly mothers utilizing post-partum care and advise facilities. this location was selected mostly because the investigator could visit it in his free hours.

In all the three locations, mothers were told the purpose of the interview and tried to make the interview appear as part of the hospital procedure in order to gain acceptance and motivation on the part of the mothers.

written querry of mothers:

Before interviewing mothers were first asked if they were educated and if they were willing to answer questionnaire in writing. Whoever consented were given the questionnaire and a pen and were asked to fill it there itself. Questionnaires were also given to doctors attending the post-partum clinic so that they could get it filled in the absence of the interviewer.

The subjective behaviour observation testing:

All the infants who were screened using the high risk check list were also subjected to an informal behaviour observation testing. This was used to study the auditory responses of the infant.

Materials used in this testing:

A drum, a cowbell, and speech stimuli was also used.

PROCEDURE: This is done in a quiet room. And is done when the infant is aweke. And the infant is held in the mother's lap.

This testing was carried-out with the help of another experienced audiologist who presented the stimuli. And the observations were done by the investigator.

Responses: All behaviour responses like eye-blinking, startle, eye widening, localization responses, body movements were looked for.

recording of responses: The follwing mode of recording was used.

Type of stimuli	Levels			
	Mild	Moderate	Moderately loud	Loud
1. Drum				
2. Cowbell				
3. Speech				

Based on the responses given fay the infant, the hearing was screened to see whether it was normal or not.

Data analysis:

It was assumed that each pregnancy and delivery were unique in themselves and that the factors affecting them were also unique. Hence, for the purposes of this study a response concerning to one child has been considered as a unit of data.

Ninety (N=90) infants were screened using the high risk cheek list. With the data obtained from these infants the following were analysed.

1. The number of high risk babies vs. non-risk babies, (the percentage of high risk babies).
2. The percentage of each factor in the given infant population identified to be at risk.
3. Percentage of infants identified to have hearing loss using the informal behaviour observation testing.

CHAPTSR-IV

THE HIGH RISK CHECKLIST

1. High risk registration methodologies

    a) Medical records

    b) Querry interview method

    c) Legal document

2. Options in India

3. The questionnaire or checklist method

    a) Purposes of a checklist

    b) Uses of a checklist

    c) Criteria for an efficient checklist

    d) Types of questions

    e) Merits and limitations of a checklist

4. The checklist

    a) The respondent

    b) The language

5. The question in the checklist for the present study.

1. High risk registration methodologies:

    A high risk register earn be easily maintained by enter-
ing the names and risk information along with other details
of those babies suspected to be at risk of developing a
hearing loss. Various methods have been employed to collect
particulars for risk classification. Functionally the sources
of there information can be divided into the following three:

a) Medical Records: Invesrtigators or volunteers can rumage into case history forms and other medical records and identify conditions relevant to the high risk register. This has been successful where detailed records of / every birth are maintained. But this cannot serve as the sole source, however exhaustive or efficient the system of medical records may be, often, the records do not contain all the information needed for high risk classification. Interpretation of varied medical terminologies, abbrevations and even handwriting is often problematic. In many places legal complications concerning the confidentiality of medical records arise.

b) Ouerry Interview method: A written questionnaire is administered to mothers at some time after the body is born. This is usually followed by an interview to cross check the answers. By far, this has been the most frequently employed method because of its ease and effectiveness. Few programs, like the two Utah programs have employed the questionnaire alone. Low return rate, high rate of false positive answers, and /reliance on literacy, coupled with the drawbacks of the questionnaire) method itself seemingly reduce the efficacy of this method, when employed without as adjuctant interview. A personal interview, along with its own advantages, also allow, for a visual examination of the baby for any congenital malformations.

c) Legal documents: In many countries, where most births are in hospitals the birth certificate is a mandatory legal document. They are required to be filled by either the supervising physician on the parent or by both. Birth certificate employed in many places contain certain medical information which may be useful for risk/categorization. The Utah state-wide high risk program has been utilizing this source very effectively. The fact that this system of birth registration often employs computerisation data retrival and classification are made much easier. However, the birth certificates may not contain all the information needed for risk computersation. In such cases, modification or exten- sion of details entered into the birth certificate is nece- ssary Which involves legal procedures. If it is success- fully exploited, it is the only system that can ensure 100% screening rate.

2. Options in India:

India is a developing country and as such has not been able to afford the kind of health care benefits many of the developed countries have been providing. Unlike la countries like Sweden and Denmark where virtually, all deliveries are in hospitals, barely 3 to 5% of the deliveries in India are conducted in hospitals (Savitha, Rani et al. 1979). Possibly,

another 5% of deliveries way bo medically supervised. Except
in few, big, well equipped hospitals confined mostly to
metropolitan cities, there hardly exists a system of main-
taining a detailed ease history for every birth. As such,
risk information from case history or medical records seems
a distant proposition.

Though every live birth has to be legally registered
in our country, barely 20 to 30% are actually registered.
(Manorama year book (1979) Manorama, Kottayam). Our birth
registers hardly contain any medical information needed for
risk categorization. Thus, legal documents like birth
certificates are unlikely a choice as potential sources of
high risk data.

According to 1975 census only 18.7% of women in India,
are literate, meaning just able to read and write. Most of
the literate women liven in urban areas. Even if/we assume
that atleast high school enrollment as the level required
to enable the mother to read and answer a detailed question-
naire, only 9.12% of a total of 105.7 Million mothers could
be administered a written questionnaires. Moreover, unlike
in Westers countries this population of mothers available to
fill a questionnaire is not easily accessable. All these,

coupled with the in built drawback of the written question-
naires itself. Seemingly make it virtually impossible to
employ a written questionnaire as a source of high risk
data. However, it may not so bleak a picture, We can
utilise services of basic health workers (BHWs), Auxiliary
Nurse Medwifes (ANMs) and other social workers to/help
mothers fill the questionnaire. If this approach proves
feasible. it will supplement the additional advantages of
scheduled interview method to this method.

Presently a scheduled interview with the mother seems
to be most logical choice. Inspite of the projected unso-
phistication, illiteracy and social conservatism she seems
to be the only potential source of information relevant to
high risk registry. It is quite likely that she will
remember most details of events during her pregnancy, of
the delivery, the physical appearance of her child at birth
and events during early post natal life of her child. In
fact, the basic premise of this study, is that every mother,
if approaches in a manner acceptable to her, her family or
her community, can be a very useful source of information
relevant to a high risk register. This would mean that we
may have to interview 2500 mothers every year in Mysore Dist.
alone, which has a conservative population of 15,000,00 in
which 2500 children are born calculated at a rate of 35 per
1000.

3. The questionnaire or checklist method:

Among the data collection methods interviews and schedules have the distinction of being capable of collecting a great deal of information through fairly straight forward questions. Only in such eases as income, family problems, sexual matters etc, wherein reluctance, unwillingness. Or just inability of respondents, they may fall to collect the desired amount of information (Kerlinger, 1973).

Checklist is the tern used for almost any kind of instrument that has questions or items to which individuals respond. Usually they are of two types, namely - schedules (interviews set up a pre conceived schedule and self administered (written questionnaire). Few, however, consider the term 'Questionnaire as more applicable to a self administered (written) questionnaire (Kerlinger, 1973).

Types of questions :

Basically, there are two types of questions or schedule items (Kerlinger, 1973) viz.
(i) Fixed Alternative (closed) type: As the term suggest they force the respondent to respond in given alternatives. Usually, a dichotimized yes or no choice is given. Some add "undecided" or "not sure" and even a "does not know" alterna-

tive. They provide for greater uniformity of responses
and elicit desired responses to fit previously devised
categories and be thus more reliable. But, there is a
danger of superficiality and inaccurate alternatives. A
respondent may prefer an inappropriate alternative than
conceal ignorance. However, when judiciously used with
probes and cues and mixed open items they can be very
useful.

(ii) Open end type:They are flexible and allow for in
depth questionning, can clear up misunderstanding through
probing, detect ambiguity, encourage cooperation. Some
times they elicit unexpected answers which may be useful.
They are very useful in interviews.

a) <u>Purposes of a checklist</u>: It translates research objective
into specific questions with minimum distortion of the
response it elicits and secondly, it assists the respondent
to communicate the required information (Kerlinger, 1973).

b) <u>Uses of a checklist</u>: Its uses are many viz.(Kerlinger,
1973).
(1) It can be used to/study relations and to test hypothesis.
(2) Can be used as an exploratory device to identify varia-
    bles, relations,to suggest hypotheses and to guide other
    phases of research.

(3) Can be used as a main instrument of research rather than as were information gathering devices.

(4) Can be used to supplement other methods used in a research study follow up unexpected results, validate other methods, and to *go* deeper into motivations of respondents for responding as they do.

e) Criteria for an efficient checklist (Kerlinger, 1973):

(1) Interviewers must be trained, questions should not be ambiguous. It should be shown to be able to gather data in much easier and better way than other methods.

(II) It should be reliable. And free from interviewer bias.

e) Merits and limitations of a checklist:

The checklist method has the distinction of being the only method that can collect any kind of information needed la social research with relative ears (Festinger, and Katz, 1965). It enjoys many advantages over methods, vis.

1. It enables us to collect a large amount of data in a relatively short-time.

2. It reduces multiple meaning and ambiguity of responses.

3. It is economical in that, it does **not** require instruments.

4. It provides sharp and constant focus on the problem being tackled.

5. It has greater reliability.

Its major disadvantages is that it takes a long time, energy, money and skill to construct a reliable "checklist" problems of language, dialect time taken to administer are other disadvantages. In addition, it can be disadvantages by the kind of questions it employees, their arrangement, its social adaptability and various other factors relating to the interviewer or questionnaire, the respondent etc.

a) The Respondent:The mother was the respondent in this study. All those unable to read and write on those who expressed their inability to comprehend the written questionnaire/checklist were administered the oral questions. Many educated mothers were interviewed likewise. In most cases the mother was the sole respondent. In many instances, however, other family members volunteered information or had to be asked for clarification.

b) The language : The checklist had both the English and Kannada versions. And was administered according to **the** respondents convenience. And the Kannada dialect was that spoken in and around Mysore city.

5. The question in the checklist f or the present study:

The checklist consists of 18 questions with the high risk factors incorporated in it. And all the questions are fixed alternative/closed/ type of questions. A dichotimized 'yes' or 'no' choice is given.

Following are the questions included in the checklist:

1. Is any one in the (child's) family, on the father's side or mother's side, having a severe hearing problem since childhood?

2. Is anyone in the (child's father's family or mother's) family having a speech problem?

3. Is any one in the (child's father's family or mother's) family who has a cleft lip and/or cleft palate?

4. Does the child have ears Which look different ie. abnormal (too small, rather big, slightly away from where ears are normally found)?

5. Does the child have a cleft lip or cleft palate?

6. Is the child's jaw or tongue different is abnormal?

7. Did the (child's) mother take drugs during pregnancy?

8. Did the (child's) mother have illness such as measles, mumps, chicken pox etc. during pregnancy?

9. Did the (child's) mother require treatment for conditions such as blood pressure during pregnancy?

10. Did the (child's) mother notice bleeding during pregnancy?

11. Was the (child's) mother exposed to radiations such as x-rays, during pregnancy.

12. Was the (child's) mother hospitalized for long prior to delivery of the child?

13. Did the child weigh much less than normal at the time of birth?

14. Was the child born prematurely? By how many weeks?

15. Was the child's appearance blue at the time of Birth?

16. Did the child not cry immediately after birth but did so after some time?

17. Was the child given blood transfusion soon-after birth?

18. Was the child's appearance yellow at the time of birth?

CHAPTER V

RESULTS AND DISCUSSION

The present study aimed/at screening the infant population using a easier mode that is the high risk checklist (See Chapter on High Risk Checklist).

It also aimed at supplementing the checklist finding with a hearing screening test ie, the behaviour observation testing,

As revealed by the review of literature the following are the percentages of high risk babies reported in the new borns screened:

i)   Feinrnesser and Tell (20%), Jerusalem study,

ii)   University of Colorado screening project (Gerkin, 1977) - (10.7%)

iii)   Utah high risk program (Mahoney and Eichwald, 1979) - (17.4%)

lv)   Utah state wide infant high risk hearing project (Mahoney and Eichwald, 1980) - (13.7%).

The results of the present study show that of the 90 infants screened using the high risk checklist, 18 (20%) were identified to be at risk. And this percentage values was close to the percentage values reported by the previous studies.

When looking at the high risk factors (or question in the checklist) eight out of 90 mothers have answered "Yes" for the question 13 ie. low birth weight. And the percentage of this factor in the population was 11.1% (See " Table I). Next to this factor, delayed birth cry and prematurely born had percentages of 7.7% And none of the mothers answered "yes" to question 17 and 11 (ie., blood transfusion soon-after birth, and x-ray exposure during pregnancy respectively). Those results show a high prevalence of the factor low birth weight among the population that was screened.

After screening the infants with the checklist BOA was also done as a supplementary procedure to obtain details about the auditory behaviour of the infants screened.

The review of literature revealed that when a high risk register was supplemented with an objective hearing test it yielded better results of early identifiation of hearing loss. Feinmesser and Bauberger-Tell (1971) 20% of the population at risk, 23 were identified to have bearing loss, and 17 were identified by the high risk program. And only six by the BOA (after a follow-up for three years).

Sub. No.	High risk +ve	High risk -ve	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18
31		-																		
32		-																		
33	+ve			+											+	+				
34		-																		
35		-																		
36		-																		
37		-																		
38	+ve			+					+							+				
39		-																		
40		-																		
41		-																		
42		-																		
43		-																		
44		-																		
45		-																		
46		-																		
47		-																		
48		-																		
49	+ve								+						+		+			
50		-																		
51	+ve						+									+				+
52	+ve														+		+	+		
53		-																		
54		-																		
55		-																		
56		-																		
57		-																		
58		-																		
59		-																		
60		-																		
61		-																		
62	+ve		+							+							+			
63		-																		
64		-																		
65		-																		

Table-1: Showing the analysis of each high risk factor.

Sub. No.	High risk +ve	-ve	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18
1		–																		
2		–																		
3	+ve									+					+		+	+		
4		–																		
5	+ve														+	+	+			
6		–																		
7		–																		
8		–																		
9	+ve		+												+			+		
10	+ve										+				+					
11		–																		
12		–																		
13		–																		
14		–																		
15	+ve		+							+					+					
16		–																		
17		–																		
18	+ve																+			
19		–																		
20		–																		
21		–																		
22		–																		
23		–																		
24	+ve								+						+		+	+		
25		–																		
26		–																		
27		–																		
28		–																		
29		–																		
30		–																		

Sub. No.	High risk +ve	-ve	| Analysis of each factor																
			1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18
66		-																		
67		-																		
68		-																		
69	+ve					+				+							+			
70		-																		
71	+ve				+									+		+				
72		-																		
73	+ve					+					+				+			+		
74		-																		
75		-																		
76		-																		
77		-																		
78		-																		
79	+ve				+										+			+		
80		-																		
81		-																		
82		-																		
83		-																		
84		-																		
85		-																		
86		-																		
87		-																		
88	+ve					+									+			+		
89		-																		
90		-																		

Total No.90
No.of high risk babies - 18
Not at risk   - 72
Percentage of high risk- 20%

| Percentage of each factor. | | | 3.3 | 2.2 | 2.2 | 2.2 | 1.1 | 1.1 | 3.3 | 4.4 | 1.1 | 1.1 | 6 | 1.1 | 11.1 | 7.7 | 7.7 | 8.8 | 0 | 1.1 |

Mencher (1974) - 10,000 infants seen, 80% wore followed up for two years. The results confirmed nine babies, seven identifies by screening only, and five by high risk register. Two babies passed both.

In this particular study when the BOA was supplemented, 44.4% were suspected to have hearing loss and 55.6% had normal hearing (See Table - 2 and 3)

The reasons for the equal distribution of percentages may be the followings

1) The behavioural observation testing was done is a crude manner ie. in the open wards sad not in an acoustical treated room,

2) Only one observer was present to observe the behaviour of the child.

3) The external noise stimuli that would have made the child to respond.

4) Only once the child was tested using the BOA, Actually many follow-ups are required to confirm the results.

So, it is important that the infant be followed-up regularly. And the mothers are adviced to observe the auditory behaviour and the speech development of the child. They must be adviced for a regular follow-up.

Table-2: Results of the behaviour observation audiometry

Subject 1.	High risk/ or not 2.	BOA done or not 3.	Bearing loss suspected 4.	Normal hearing 5,
1	Not at risk	Yes		
2	Mot at risk	Yes		
3	Hot at risk	Yes		
4	Hot at risk	Yes		
5	At risk	Yes	+	
6	Not at risk	Yes		
7	Hot at risk	Yes		
8	Hot at risk	Yes		
9	At risk	Yes		
10	At risk	Yes	+	
11	Hot at risk	Yes		
12	Hot at risk	Yes		
13	Hot at risk	Yes		
14	Hot at risk	Yes		
15	Hot at risk	Yes	+	
16	Hot at risk	Yes		
17	Hot at risk	Yes		
18	At risk	Yes		
19	Hot at risk	Yes		
20	Hot at risk	Yes		
21	Hot at risk	Yes		
22	Hot at risk	Yes		
23	Hot at risk	Yes		
24	At risk	Yes	+-	
25	Hot at risk	Yes		
26	Hot at risk	Yes	(+)	
27	Hot at risk	Yes		
28	Hot at risk	Yes		
29	Hot at risk	Yes		
30	Hot at risk	Yes		
31	Hot at risk	Yes		
32	Hot at risk			
33	At risk	Yes	+	
34	Hot at risk	Yes		
35	Hot at risk	Yes		
36	Hot at risk	Yes		
37	Hot at risk	Yes		
38	At risk	Yes		
39	Hot at risk	Yes		
40	Hot at risk	Yes		

1.	2.	3.	4.	5.
41	Not at risk	Yes		
42	Not at risk	Yes		
43	Not at risk	Yes		
44	Not at risk	Yes		
45	Not at risk	Yes		
46	Not at risk	Yes		
47	Not at risk	Yes		
48	Not at risk	Yes		
49	At risk	Yes		
50	Not at risk	Yes		
51	At risk	Yes		
52	At risk	Yes	+	
53	Not at risk	Yes		
54	Not at risk	Yes		
55	Not at risk	Yes		
56	Not at risk	Yes		
57	Not at risk	Yes		
58	Not at risk	Yes		
59	Not at risk	Yes		
60	Not at risk	Yes		
61	Not at risk	Yes		
62	At risk	Yes		
63	Not at risk	Yes		
64	Not at risk	Yes	(+)	
65	Not at risk	Yes		
66	Not at risk	Yes		
67	Not at risk	Yes		
68	Not at risk	Yes		
69	At risk	Yes		
70	Not at risk	Yes		
71	At risk	Yes		
72	Not at risk	Yes		
73	At risk	Yes	+	
74	Not at risk	Yes		
75	Not at risk	Yes		
76	Not at risk	Yes		
77	Not at risk	Yes		
78	Not at risk	Yes		
79	At risk	Yes		
80	Not at risk	Yes		

1.	2.	3.	4.
81.	Not at risk		
82	Not at risk	Yes	(+)
83	Not at risk	Yes	
84	Not at risk	Yes	
85	Not at risk	Yes	
86	Not at risk	Yss	
87	Not at risk	Yes	
88	At risk	Yes	+
89	Not at risk	Yes	
90	Not at risk	Yes	

Percentage of infants
having hearing loss   -   14.4%
in the total population

Table-31 Showing percentage of high risk infants
having hearing loss when BOA was done.

Sub.No.	BOA done/or not	Hearing loss suspected	Normal hearing
1	Yes		+
2	Yes	+	-
3	Yes	-	+
4	Yes		-
5	Yes		-
6	Yes	-	+
7	Yes		-
8	Yes		-
9	Yes	-	
10	Yes	-	
11	Yes	-	+
12		+	-
13	Yes	-	+
14	Yes	-	+
15	Yes	-	•f
16	Yes		-
17	Yes	-	+
18	Yes	+	-

N = 18

Percentages   44.4%

Percentage of high risk infants
suspected to having hearing loss   =   44.4%

CHAPTER-VI

SUMMARY AND CONCLUSION

The aim of the present study was to screen the infant population using the high risk check list and supplement this with the behaviour observation audiometry.

Data was collected by interviewing mothers of infants attending the local medical college hospitals, immunization clinics and well baby chinics. The infants **were** also subjected to behaviour observation audiometry.

The percentage of high risk infants in the population was drawn. And the prevelance of the factors were also studied. Finally, the percentage of infants suspected of hearing loss was also deducted.

The study showed that out of 90 infant screened 20% of the new born population was at risk. Low birth weight was the factor that was found to be more prevelant, x-ray exposure by the mother and blood transfusion postnatally to the child were the two factors that had least prevelance.

Looking at. the percentage of .infants suspected of hearing loss. It was found that there was almost **equal**

distribution of this percentage values (ie. 44.4% was suspected to have hearing loss, and 55.6% had normal hearing).

However, to confirm about the hearing loss the high risk program has to be supplemented with other objective procedures like BSERA etc. And a regular follow-up to Monitor the auditory behaviour of the infants.

CHAPTER - VII

SUGGESTIONS FOR FURTHER STUDY

(1)   The infants can be followed tip further for a con-siderable period of time to monitor his auditory behaviour and for a confirmatory diagnosis of hearing loss.

(ii)   The sane study can be carried out by supplementing the high risk program with other objective tests such as ABR etc.

(iii) During the follow-up the mothers can be advised to look for any abnormal auditory behaviour or any delayed speech development and can report this to the investigator.

a) Auditory behavioural responses that the parents are supported to look for -

1) Eye blink or eye lid activity.

2) Violent startle reaction, consisting of a Jerking of the entire body, with arms and legs drawn towards the midline.

3) Cessation of activity.

4) Limb movements.

5) Head turn away from sound or toward sound

6) Grimacing

7) Arousal

8) Widening of the eyes.

b) Prelinguistic behaviour to be monitored:

1) Using sounds in a repetitive manner

2) At two to four months should be using vowel sounds

3) By five months the consonant vowel sequences should begin (eg. /ka/ ka/,/ki/ /ki/)

4) At five to six months, labial (/pa/, /ba/) should be obtained.

5) Mine to ten months look for production of alveolars.

So, it is important that the mothers report about the auditory behaviour and speech behaviour also in this follow-up.

(iv) Those high risk factors that have a high degree of predicting hearing lose can be studied by correlation methods between the factors and the behaviour observation findings.

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