STATE OF THE ART REVIEW OF LITERATURE

NEONATAL AND INFANT HEARING SCREENING

2011

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CONTENTS

PREAMBLE

Deafness, if present from childhood has a significant impact on the social, economic and productive life of an individual. Our country faces a huge gap in resources and manpower available to meet the challenges faced by this health situation. The Government of India has retained primary health care as the strategy of choice for the provision and implementation of prevention of deafness and hearing. The Union Government initiated the National Programme for Prevention and Control of Deafness (NPPCD) in 2006 with the long term objective to prevent and control major causes of hearing impairment and deafness, so as to reduce the total disease burden by 25% of the existing burden by the end of eleventh five year plan.

India has a birth rate of 22.5 per 1000 population (SRS 2009), and it can be estimated from prevalence studies that out of every 1000 children born, there may be 5to 6 children who may not be able to hear properly. In a newborn and in early childhood, hearing impairment is difficult to detect since there are no visual indicators and most hearing-impaired children who are not screened at birth are not identified until between 1½ and 3 years of age. This is well beyond the critical period for healthy speech and language development.

However, with the help of newborn hearing screening modalities available, a hearing-impaired child can be identified and treated early. Reduced auditory input can also adversely affect the development of the central auditory nervous system, and can negatively impact speech perception that interferes with growth in behavioral, social and cognitive spheres, academic achievement, vocational options, employment opportunities and economic self sufficiency.

There has never been any attempt to screen the neonates or infants for hearing defects in large scale in India. The All India Institute of Speech and Hearing (AIISH) conducts infant screening for hearing disorder on regular basis in different hospitals attached to it using Behavioral Observational Audiometry (BOA), Otoacoustic Emissions (OAE) screening and administering High Risk Register (HRR). Literature reveals that almost 50% of hearing impaired children are likely to be missed by using a high risk register and NICU based approaches. Also, the institutional delivery is still less than 50% in our country, thus if we target only the infants born in institutions, we are expected to miss around half of the probably hearing impaired children. Thus, a need for a community based screening programme is altogether essential, which must undoubtedly, co-exist with an institutional protocol.

The instruments which are used for screening the auditory functions of newborns can be made available at each place where delivery occurs. The hearing test should be made mandatory before the discharge of the baby. Since the test is simple and interpretations easy to understand, it can be performed by the health workers as well, after an essential training.
ASHAs appointed in NRHM, Anganwadi Workers (AWWs), female health workers, all grass root level worker can be trained in new-born screening techniques under the National Program. The screening programme will lead to increase in demand of ENT specialists, audiologists, speech therapists and related man power. These qualified personnel needs to be produced or the available manpower can be trained additionally in hearing care. While targeting every effort to detect and manage hearing loss early with minimal social and academic consequences, there is a need to open special schools for rehabilitation. This will help the hearing impaired to achieve their due social status in the community and provide them with livelihood options which are commensurate with their intellectual capabilities

The screening programme for the country can be phasic, and can be started initially in the districts where the national programme (NPPCD) has been initiated. Gradually the programme should cover the whole country. Utilization of the existing infrastructure already developed under the programme for the screening will not create additional financial burden on the system except the cost of OAE/ABR machines.

Most essential is the creation of awareness in the community, so as to recognize hearing impairment as an important public health problem. For this, there is a need to develop and disseminate adequate and appropriate IEC materials regarding effective hearing screening programs to the community.

GLOSSARY OF ABBREVIATIONS

AAP	American Academy of Pediatrics
ABR	Auditory Brain Stem Response
A-ABR	Automated ABR
AN	Auditory Neuropathy
ANOVA	Analysis of Variance (statistical test)
ARC	Auditory Response Cradle
ASSR	Auditory Steady State Response
ASHA	Accredited Social Health Activist
ASHA	American Speech-Language-Hearing Association
AWW	Anganwadi Worker
BAHA	Bone Anchored Hearing Aids
BOA	Behavioral Observation Audiometry
CAPD	Central Auditory Processing Disorder
CDC	Center for Disease Control & Prevention (USA)
CDT	Connected Discourse Tracking
CHL	Conductive Hearing Loss
CI	Cochlear Implant, Confidence Interval
CMV	Cytomegalovirus
ECMO	Extra Corporeal Membrane Oxygenation
ECochG	Electrocochleography
EHDI	Early Hearing Detection & Intervention Services
FDA	Food & Drug Administration (USA)
GBD	Global Burden of Disease
HHI	Hereditary Hearing Impairment
HV	Health Visitor
JCIH	Joint Committee on Infant Hearing
LBW	Low Birth Weight
NHSP	National Hearing Screening Protocol (NHS, UK)
NICU	Neonatal Intensive Care Unit
NIDCD	National Institute on Deafness and Other Communication Disorders (USA)
NIH	National Institute of Health (USA)
NPPCD	National Program for Prevention and Control of Deafness (India)
NRHM	National Rural Health Mission
NSSO	National Sample Survey Organization (India)
OAE	Otoacoustic Emissions
PCHI	Permanent Childhood Hearing Impairment
PEHC	Primary Ear & Hearing Care
PCEHL	Permanent Congenital & Early onset Hearing Loss
PTA	Pure Tone Audiometry
SNHL	Sensorineural Hearing Loss
UNHS	Universal Newborn Hearing Screening
USPSTF	United States Preventive Services Task Force
VRA	Visual Reinforcement Audiometry
WHO	World Health Organization
YLD	Years Lived with Disability

Background

Significant hearing loss occurs in 1-2 per 1000 newborns and in 2 per 1000 young children. A congenital or an early acquired hearing loss is as a result of any damaging factor which affects the auditory organ. Delays in speech, language, and cognitive development may occur because of hearing loss in the early years of life[1] of which the speech and language delays secondary to hearing loss are often preventable.[2,3] Of course, the future behavioral and social consequences, including loss of self esteem and diminished quality of life as a result of such delays cannot be disregarded from a public health point of view. It is thus imperative, that an early diagnostic evaluation of any hearing impairment in children can be the key to a child's success with development of appropriate communication ability.

Definitions

Deafness means complete loss of the ability to hear from one or both ears; this is profound hearing impairment, 81 dB or greater hearing threshold, averaged at frequencies 0.5, 1, 2, 4 kHz.

According to the World Health Organization (WHO) **impairment** is any loss or abnormality of psychological, physiological or anatomical structure or function.

Hearing impairment means complete or partial loss of the ability to hear from one or both ears; this is mild or worse hearing impairment, 26 dB or greater hearing threshold, averaged at frequencies 0.5, 1, 2, 4 kHz.

Disability is any restriction or lack of ability to perform an activity in the manner or within the range considered normal for a human being.

Disabling hearing impairment means moderate or worse hearing impairment in the better ear; that is the permanent unaided hearing threshold level for the better ear of 41 or 31 dB or greater in age over 14 or under 15 years respectively, averaged at frequencies 0.5, 1, 2, 4 kHz.

Size of the problem

Moderate to profound hearing loss are disabling conditions which affects 278 million people worldwide according to the 2005 estimates of the WHO.[4] Of these, two thirds live in developing countries. Half of the hearing loss is preventable. India has the largest number of hearing impaired in the South East Asia region and houses the largest deaf (bilateral profoundly hearing impaired) population of the region.

Hearing loss is the second most common cause of years lived with disability (YLD) accounting for 4.7% of the total YLD.

As per the global burden of disease: 2004 update (WHO 2008), the prevalence of hearing loss by WHO regions is mentioned in table 1.

Prevalence (millions) of Hearing Loss by WHO Regions (GBD, 2004)

	World	**Africa**	**Americas**	**Eastern-Mediterranean**	**Europe**	**South East Asia**	**Western Pacific**
Moderate, or greater*	275.7	37.6	31.0	19.5	44.5	89.8	52.9
Mild**	360.8	18.6	45.7	25.2	75.8	88.5	106.3

*Hearing loss threshold in the better ear of 41 decibels or greater (measured average for 0.5, 1, 2, 4 kHz).

** Hearing loss threshold in the better ear of 26-40 decibels (measured average for 0.5, 1, 2, 4 kHz).

Estimated prevalence of moderate and severe disability* (millions) for hearing loss by age, for high-income and low- and middle-income countries, 2004

High income countries		**Low income countries**		**World**
0-59 years	**60 years and over**	**0-59 years**	**60 years and over**	**All ages**
7.4	18.5	54.3	43.9	124.2

**Includes adult-onset hearing loss, excluding that due to infectious causes; adjusted for availability of hearing aids*

Effects of deafness and hearing impairment on individuals and society

- Damage to the development of speech, language and cognitive skills in children especially if commencing at birth or during infancy.
- Slow progress in school.
- Difficulties in obtaining, performing and keeping an occupation.
- Social isolation and stigmatization in all ages and both sexes.
- Profound social and economic effects in communities and countries.

These difficulties are magnified in developing countries, where there are generally very few services or trained staff to deal with them.

Universal Newborn Hearing Screening (UNHS)

Universal newborn hearing screening is a method to identify hearing-impaired newborns with or without risk factors. Congenital hearing loss affects about one in every 1,000 newborns. Those newborns with positive screening tests are referred for definitive testing and intervention services.

The aim of UNHS Is for 'early identification of newborns with hearing loss so that they can access timely and appropriate interventions, thereby reducing inequalities and the outcomes for these children, their parents and families/communities and society can improve.

Rationale for Universal Screening

Screening for hearing loss in newborns is based on two concepts:

1. A critical period exists for optimal language skills to develop, and earlier intervention produces better outcomes.[2,5]
2. Treatment of hearing defects has been shown to improve communication.

Data from cohort studies indicate that diagnosis and intervention before six months of age can improve language and speech acquisition in hearing-impaired children. In 1993 the National Institutes of Health Consensus Development Conference on Early identification of Hearing

Impairment in Infants and Children recommended universal newborn screening. The Joint Committee on Infant Hearing (JCIH) issued similar guidelines in 1995 and again in 2000.[6,7]

Screening Tests: The goal of any infant hearing screening program is to achieve a high level of both sensitivity and specificity. The ideal program should permit the identification of as many newborns as possible that do have a hearing loss (high sensitivity) and also exclude as many newborns as possible who do not have a hearing loss (high specificity).

Indian Scenario:

The referred prevalence data for India shows that 6.3% of Indians suffer from significant auditory loss.[8] Rural areas in India have a higher prevalence of hearing loss than urban areas.[9] As per 58th round of NSSO survey in 2002, currently there are 291 persons per one lakh population who are suffering from severe to profound hearing loss. Of these, a large percentage is children between the ages of 0 to 14 years. The survey results revealed that about 7% of people have congenital hearing loss.

While retaining primary health care as the strategy of choice for the provision and implementation of prevention of deafness and hearing, the Union Government of India has initiated the National Programme for Prevention and Control of Deafness (NPPCD) in 2006.[10] Initially started as a pilot project in 25 districts in 10 states and 1 union territory, it has been scaled up to cover 203 districts in all states and union territories of the country. The long term objective of the programme was reducing the total disease burden of hearing impairment and deafness by 25% at the end of the eleventh 5-year plan (2007-12). The major components of the programme are capacity building and manpower development wherein each level of health care provider is sensitized to the hearing and ear care, ear health promotion and prevention where the grassroots workers and health personnel delivers IEC messages on a continuous basis to community members, early detection of ear problems and management by house-to-house surveys to ascertain hearing problems in all age groups by the AWWs and ASHAs, organizing community screening camps regularly at PHC/CHC/district level to screen the population for deafness and hearing impairment, referral of complicated cases to state medical colleges,

rehabilitation and hearing aid provision. A proper system of monitoring and supervision is in place to constantly evaluate the programme with pre-defined indicators.

Behavioral techniques to detect hearing loss usually give correct result in infants more than 6 months of age. These tests relying on operant conditioning involves testing an infant's response to specific tones projected within a sound-proof room from different directions. When performed correctly, these tests can yield accurate audiometric thresholds in children as young as six months of age who have normal neurologic development.

The instruments used for screening the auditory functions of new-born can be supplied at each place where delivery occurs. The hearing test should be made mandatory before the discharge of the baby. Since the test is simple and interpretations easy to understand, it can be performed by the nurses at the maternity centers. However, since the institutional delivery is still less than 50% in India, targeting only the infants born in an institution will miss around half of the probably hearing impaired child. The grass root level workers including ASHAs appointed in NRHM, Anganwadi Workers (AWWs) and Female Health Workers, etc. involved in providing health care services to the community, eg. Immunization etc, can be trained and utilized for new-born screening techniques under the NPPCD. The infrastructure already developed under the programme can be utilized for the screening, thus not create additional financial burden on the system except the cost of OAE/ABR machines where required.

Under the NPPCD and with support of the WHO, an expert committee meeting was held in January, 2009 at Maulana Azad Medical College, New Delhi wherein guidelines were developed for infant hearing screening in India. The main objectives of the meeting, apart from discussing the existing programme for neonatal and infant hearing screening in India and the world, were to (1) establish the most effective strategy for detection of deafness in infants at the community level and the hospital level; and (2) prepare a plan for implementation of Infant Hearing Screening under the NPPCD in India.

Gaps and needs: According to the WHO[11]

- Awareness is often lacking about hearing and hearing loss in society, including amongst decision makers, health workers, teachers and parents.
- National plans and programmes for prevention and rehabilitation of hearing loss are lacking in most developing countries.
- Primary ear and hearing Care (PEHC) programmes have not been implemented in most developing countries.
- Secondary and tertiary care programmes need developing or strengthening in most developing countries.
- Coordinated programmes to provide affordable hearing aids and services need to be set up in developing countries.
- Many more health and rehabilitation personnel need to be trained in ear and hearing care at the primary and secondary levels and for specialist care in developing countries

Future challenges

- Numbers and prevalence of persons with deafness and hearing impairment are rising because of increasing world population and lengthening life expectancy.
- More information is needed to measure the size of these problems and determine their individual, social, and economic effects.
- Decisive public health action is needed to reverse these increases, especially in developing countries

Methodology Followed for the State of the Art Review of Literature

The present state of the art review of literature intended to thoroughly scrutinize and examine all available literature and research in scientific journals accessible through the library resources and research database, including systematic reviews and meta analysis studies, where available. Literature search was carried out in Pubmed using keywords : Epidemiology of Hearing Loss, Causes of Hearing Loss, Modalities of early identification of Hearing Loss, Behavioral observation testing in Hearing loss, Behavioral observation audiometry, Hearing screening

programs, Deafness, Childhood deafness, Management of hearing impaired, Rehabilitation of hearing impaired, modalities for treatment of hearing impaired etc. while compiling relevant articles in reputed journals published in English language. Review articles of Uptodate™ were also referred for this task at some places with permission, and appropriate acknowledgement and referencing of original work.

A thorough search and compilation of articles was followed by an in-depth analysis and review of the available evidence in following broad domains :-

- Epidemiology of Congenital Hearing Loss
- Causes of Hearing Loss in neonates and infants
- Ideal and actual age of identification
- Modalities for Early Identification of Hearing Loss : A-ABR/BERA etc.
- Behavioral observational testing for the assessment of hearing loss in neonates
- Hearing screening programs and protocols in different countries, with a focus on comparison of feasibility of an Institutional protocol viz. a. viz Community based protocol in field conditions for a country like India
- Modalities for management of Hearing Impaired children including educational rehabilitation.

Need/Importance

A comprehensive review of literature and compilation of evidence is expected to facilitate the development of a scientific, i.e. evidence based, economical and feasible protocol(s) for the neonatal screening for hearing loss by the senior level policy makers and stakeholders.

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Chapter 2: Epidemiology of Congenital Hearing Loss

Congenital hearing loss is any hearing loss that occurs at, or shortly after birth and in the child, may be the reason for the hearing defect, whether conductive or sensorineural.

Epidemiology

Hearing impairment is one of the commonest congenital disabilities in the world. Incidence of hearing impairment in a standardized population of neonates at risk and not at risk to develop hearing impairment ranges from 6 to 60 per 1000 neonates with an average of 4 per 1000 neonates.[1]

According to the National Institute on Deafness and Other Communication Disorders (NIDCD, USA), hearing loss affects approximately 28 million Americans and approximately 17 in 1000 children and adolescents younger than 18 years. The average incidence of hearing loss in neonates in the United States is 1.1 per 1000 with variability among states ranging between 0.22 and 3.61 according to Mehra et al. In this study the prevalence of childhood and adolescent hearing loss was 3.1%, with higher rates in Hispanic Americans and in families with lower incomes.[2]

Congenital hereditary hearing loss must be differentiated from acquired hearing loss. Most cases of pre-lingual deafness are genetic. The remaining 40-50% of all cases of congenital hearing loss are due to non-genetic effects, such as prematurity, postnatal infections, ototoxic drugs, or maternal infection (with cytomegalovirus [CMV] or rubella). Most cases of genetic hearing loss are autosomal recessive and nonsyndromic. Hearing loss that results from abnormalities in connexin 26 and connexin 30 proteins likely account for 50% of cases of autosomal recessive nonsyndromic deafness in American children.

Genetic sensorineural hearing loss (SNHL) appears to occur twice as often in developed countries as in underdeveloped countries. In addition to ancestry and race, the proportions of hereditary versus acquired and syndromic versus non-syndromic hearing losses across populations is highly variable and is heavily influenced by multiple factors, some likely not yet identified, including drift of populations, frequency of consanguinity, and health status.[3]

Estimating the prevalence of hereditary hearing loss in populations across the world is very difficult because access to health care, poor health conditions, and a low level of awareness of hearing loss is compounded by a higher frequency of complicating risk factors such as neonatal distress, prematurity, high fever, otitis media, meningitis, ototoxic medications, and illnesses such as rubella.[4]

An estimated 30,000 infants are born with sensorineural hearing loss each year in China, which has a population of about 1.3 billion, but the percentage of these hearing losses attributable to heredity is not known.[5] Saunders et al demonstrated a prevalence of significant hearing loss of 18% in a group of school-aged children in rural Nicaragua with a familial history of hearing loss in 24% of the children with hearing loss.[4] Large-scale epidemiologic studies are needed and will become more feasible as molecular testing is made available to the world's populations.

Population-based studies in Europe and North America have identified a consistent prevalence of approximately 0.1% of children having a hearing loss of more than 40 decibels (dB) through review of health or education records, or both. Other international studies using different methods or criteria (such as screenings, questionnaires, and less severe decibel thresholds) have reported higher estimates.[6] See Table 1.[6]

Table 1: Published literature on prevalence of Hearing Loss in Children (International)[6]

Published Literature on Prevalence of Hearing Loss in Children

Author	Published Year	Country	Study Period	Age at identification	Cases (n)	Population (N)	Decibel Threshold	Case Identification Method(s)	Prevalence/1000 (95% CI)
Kankkunen	1982	Sweden	1964-1969#	<6 years	50	40963 live births	>40 dB	Medical Records	1.2 (0.9, 1.6)
			1970-1979#	6 years	179	31286 live births	>40 dB	Medical Records/Questionnaire	1.3 (1.0, 1.8)
Martin	1982	EEC	1969#	8 years	3462	4126268 live births	≥50 dB	Questionnaire	0.84 (0.81, 0.87)
Upfold	1982	Australia	1949-1980#	<18 years	16199	7231000 live births	~60-90 dB	Medical Records	2.24 (2.20, 2.28)
Parving	1983	Denmark	1970-1979#	2-12 years	117	82265 live births	≥35 dB	Medical Records	1.4 (1.2, 1.7)
McPherson	1985	Gambia	1983	2-10 years	433	130776 children	≥30 dB	Screening	3.3 (3.0, 3.6)
Newton	1985	United Kingdom	1977-1980#	<6 years	111	136720 live births	>25 dB*	Medical Records	0.8 (0.7, 1.0)[1]
Sorri	1985	Finland	1966#	15 years	64	11748 children	>25 dB	Screening	5.4 (4.3, 6.7)
Sehlin	1990	Sweden	1964-1983#	≤20 years	160	63463 live births	≥30 dB	Registry	7.5 (2.2, 2.9)
Davis	1992	United Kingdom	1983-1986#	3-6 years	41	29317 live births	≥50 dB	Medical Records	1.4 (1.0, 1.9)
Parving	1993	Denmark	1970-1980#	2-12 years	117	82265 live births	≥25 dB	Registry	1.4 (1.2, 1.7)
			1980-1990#	2-12 years	94	60985 live births	≥25 dB	Registry	1.5 (1.3, 1.9)
Karikoski	1994	Finland	1973-1990#	≤18 years	353	270726 children	≥30 dB	Medical Records	1.3 (1.2, 1.5)
Davis	1995	United Kingdom	1983-1988#	3-5 years	200	164929 live births	≥40 dB*	Screening/Registry	1.2 (1.1, 1.4)[1]
Baille	1996	France	1976-1985#	<9 years	217	327095 live births	>70 dB	Education Records	0.7 (0.6, 0.8)[1]
Darin	1997	Sweden	1980-1984#	6-10 years	55	27881 live births	>20 dB	Medical Records	2.0 (1.5, 2.6)
Fortnum	1997	United Kingdom	1985-1993#	2-10 years	653	552558 live births	≥40 dB	Medical Records	1.2 (1.1, 1.3)
Vartianen	1997	Finland	1974-1987#	<9 years	98	46240 live births	>25 dB*	Screening	1.7 (1.4, 2.1)[1]
Maki-Torkko	1998	Finland	1973-1992#	<14 years	253	212328 live births	≥40 dB	Medical Records	1.2 (1.1, 1.3)
Streppel	1998	Germany	1992-1993#	2-19 years	314	738500 live births	>30 dB	Screening	0.4 (0.38, 0.47)
Varitanen	1998	Finland	1972-1986#	<10 years	84	49090 live births	≥40 dB*	Screening	2.1 (1.7, 2.6)[2]
Mencher	2000	Costa Rica	1988	8 years	122	81780 live births	>20 dB*	Screening	1.5 (1.3, 1.8)
Nekahm	2000	Austria	1980-1994#	6-20	165	124809 live births	>40 dB	Medical Records	1.3 (1.1, 1.5)
Uus	2000	Estonia	1985-1990#	8-13 years	240	139535 live births	≥40 dB	Screening	1.7 (1.5,1.9)
Fortnum	2001	United Kingdom	1995#	3 years	NR	NR	>40 dB	Education/Medical Records	1.1 (1.0, 1.1)
			1982-1989#	9-16 years	NR	NR	>40 dB	Screening/Medical records	2.1 (2.0, 2.1)
Liu	2001	China	1986-1989	<15 years	227	34157 children	>27 dB	Screening/Questionnaire	6.6 (5.8, 7.6)
Levi	2004	Israel	1968-1977#	Children**	82	63950 live births	≥56 dB*	Screening	1.3 (1.0, 1.6)[1]
			1978-1991#	Children**	150	141720 live births	≥56 dB*	Screening	1.1 (0.9, 1.2)[1]
Russ	2007	Australia	1993#	6 years	141	64116 live births	>40 dB	Screening	2.2 (1.9, 2.6)

*sensorineural hearing loss only

**age unknown

live Births

[1] bilateral cases only

[2] unilateral cases only

[3] Survey Questionnaire Item-'deaf/a lot of trouble hearing'

Abbreviations: dB=decibels; EEC=European Economic Community; n=Frequency; NR=Not Reported.

Risk factors for Congenital Hearing Loss

High Risk Factors associated with congenital hearing loss	Family history of hereditary childhood sensorineural hearing loss Hyperbilirubinemia Ototoxic medications Bacterial meningitis Low birth weight In utero infections (cytomegalovirus, rubella, syphilis, herpes, and toxoplasmosis) Craniofacial anomalies (including pinna and ear canal) Low Apgar scores at birth Mechanical ventilation lasting 5 days or longer Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss
Disorders Causing Hearing Loss at Birth	RH-Incompatibility Congenital Syphilis Anoxia or asphyxia at birth Persistent Fetal Circulation (pulmonary hypertension) Low Birth Weight High Forceps Delivery Violent Uterine Contractions

As per WHO estimates in India, there are approximately 63 million people, who are suffering from significant auditory impairment; this places the estimated prevalence at 6.3% in Indian population.[7]

National Sample Survey Organization, an agency of government of India responsible for documentation of census in its 58th Round (2003) estimated the population of persons with disability to be 18.49 million that equivalents to 1.8 percent of the total population of the country where 10 percent of this figure are likely to have hearing disability of moderate to profound degree.[8] Moreover, this number is likely to go up if we add lower degree of hearing disability. Considering such estimates, still there seems to be a dearth of any large scale incidence studies among the neonates in the Indian context and the documentation of the various newborn hearing screening programs being conducted in the country. A recent study done in India reported the incidence of congenital permanent hearing loss at 5.6 per 1000 live birth.[9]

Genetic hearing loss does have significant ethnic links. Angeli SI recently reviewed the ethnic variability of DGNB1 and showed greater allelic variability in Hispanics.[10] Schimmenti et al showed a lower prevalence of connexin-related hearing loss in Hispanic infants.[11]

Before universal hearing screening for newborns, less than 50% of children who had hearing impairment were identified before the age of 3 years. Detection of risk factors (eg, prematurity, low birth weight, low Apgar scores) helps in identification of less than 50% of infants who have or who are at risk for hearing loss. In one study, 78% of infants identified with hearing loss were in the well-baby nursery and not the neonatal intensive care nursery.[12] This finding emphasized the ineffectiveness of screening on the basis of risk identification alone. Hereditary hearing loss may also be progressive or adult in onset.

Social and economic burden

WHO states that Hearing impairment can impose a heavy social and economic burden on individuals, families, communities and countries. Hearing impairment in children may delay development of language and cognitive skills, which may hinder progress in school. In adults, hearing impairment often makes it difficult to obtain, perform, and keep jobs. Hearing impaired children and adults are often stigmatized and socially isolated. The poor suffer more from hearing impairment because they cannot afford the preventive and routine care to avoid hearing loss nor the hearing aids to make the disability manageable. Hearing impairment also makes it more difficult for them to escape poverty by hindering progress in school or in the workplace and by isolating them socially. For countries, the cost of special education and lost employment due to hearing impairment can burden the economy.[13]

The 350,000 individuals who are profoundly deaf in the United States earn approximately 30% less than the general population. Among school-aged children with hearing loss, approximately 52,000 attend schools or programs for the deaf, 100,000 are enrolled in special deaf-education classes, and 250,000 participate in standard public school settings.

Economic Cost

- During the 1999 - 2000 school year, the total cost in the United States for special education programs for children who were deaf or hard of hearing was $652 million, or $11,006 per child.[14]
- The lifetime educational cost (year 2007 value) of hearing loss (more than 40 dB permanent loss without other disabilities) has been estimated at $115,600 per child.[15]

It is expected that the lifetime costs for all people with hearing loss who were born in 2000 will total $2.1 billion (in 2003 dollars).[16]

    - Direct medical costs, such as doctor visits, prescription drugs, and inpatient hospital stays, will make up 6% of these costs.

    - Direct nonmedical expenses, such as home modifications and special education, will make up 30% of these costs.

- Indirect costs, which include the value of lost wages when a person cannot work or is limited in the amount or type of work he or she can do, will make up 63% of the costs.

  Note: These estimates do not include other expenses, such as hospital outpatient visits, sign language interpreters, and family out-of-pocket expenses. The actual economic costs of hearing loss, therefore, will be even higher than what is reported here.

ROLE OF WHO IN EPIDEMIOLOGICAL AND ECONOMIC ANALYSIS

WHO is encouraging countries with technical support, to conduct random sample, population-based prevalence and cause surveys of deafness and hearing impairment. Surveys are based on a specially developed WHO Survey Package (protocol, data entry form, coding instructions, dedicated software). The data will be used in planning, determination of priorities, economic analysis and raising awareness. Results will be used at local and national levels, and collated for regional and global levels. A meeting of all principal investigators for WHO surveys was recently held in Geneva. A global data bank on deafness and hearing impairment is being developed using data from the WHO Protocol Surveys and other published and unpublished sources.[17]

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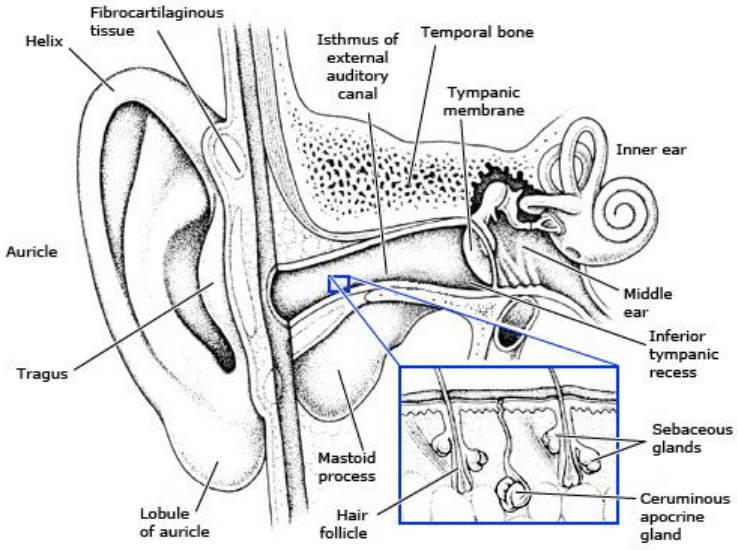
Chapter 3: Causes of Hearing Loss in Neonates and Infants

Keeping prevention as our goal, a timely knowledge of the cause of the hearing loss would incite the development of an appropriate management of the patient to avoid compromising the educational progress. Even if the cause is genetic, we can impact eugenically at the level of family planning through appropriate risk communication and counseling, and thus possibly prevent any future impairment.

THE ANATOMY AND PHYSIOLOGY OF THE HUMAN EAR

The human ear is anatomically divided into three segments:

- The outer ear: i.e. the auricle and ear canal
- The middle ear: comprising the tympanic membrane, ossicles, and the middle ear space
- The inner ear: with the cochlea, semicircular canals, and internal auditory canals



Ref: Cantor RM, Emerg Med 1999; 31:40. Copyright Quadrant HealthCom, Inc, 1999.

The unique structure of the auricle allows the capturing of incoming sound waves from all directions which then converge down the external auditory canal to the ear drum or the tympanic membrane. This vibration leads to the motion of the ossicles in the middle ear and a unique piston-like effect on the stapes bone. The stapedial motion is transmitted as a traveling wave into the inner ear fluid. These waves then propagate around the cochlear turns. Frequency-specific movement of the organ of Corti bends the stereocilia, causing depolarization of the inner hair cells and creating electrical impulses transmitted via the auditory nerve to the brain which organizes the information into what we perceive as complex sounds.

CLASSIFICATION OF HEARING LOSS: TYPES AND CAUSES

❖ TYPES OF HEARING LOSS

The etiology of hearing loss can be multifactorial and various known abnormalities that can causes hearing loss of each type in neonates and children are as follows:

Table: Types of Hearing Loss in Neonates and Infants

<table>
<tr><td colspan="3">TYPE OF HEARING LOSS</td></tr>
<tr><td colspan="3">I. CONGENITAL</td></tr>
<tr><td rowspan="2"></td><td rowspan="2">I(a) HEREDITARY</td><td>SYNDROMIC</td></tr>
<tr><td>NON-SYNDROMIC</td></tr>
<tr><td></td><td colspan="2">I(b) NON-HEREDITARY</td></tr>
<tr><td colspan="3">II. NON-CONGENITAL</td></tr>
</table>

❖ CAUSES OF HEARING LOSS

I. CONGENITAL

Congenital type of hearing loss which is genetic can be either associated with certain syndromes or can be non-syndromic.

Microtia can cause mild to moderate CHL, and atresia or significant stenosis of the EAC can cause moderate to maximal (60dB) CHL. Atretic malformations of the EAC occur in approximately 1 per 10,000 births, unilateral more common and these are usually associated with other craniofacial abnormalities such as Treacher-Collins syndrome, Robin sequence, or Crouzon syndrome.

I (a) HERIDITARY

Hereditary causes account for up to 50 percent of cases of SNHL and include SNHL with (one-third) and without (two-thirds) associated abnormalities. Approximately 80 percent of cases of Hereditary Hearing Impairment (HHI) are inherited in an autosomal recessive pattern, 18 percent are autosomal dominant, and 2 percent X-linked recessive.[1] In rare patients, HHI reflects a defect in the mitochondrial rather than nuclear DNA.[2]

Hereditary bilateral SNHL occurs from 1 in 2000 to 1 in 6000 births.[3] It may be present at birth (congenital), progressive from birth, or it may develop when the child is older

HERIDITARY-SYNDROMIC:

Various syndromes which are associated with such hearing loss include which are autosomal recessive, autosomal dominant, X linked or mitochondrial.

Autosomal recessive - common syndromic autosomal recessive disorders are Usher syndrome, Pendred syndrome, Alport syndrome, and Jervell-Lange-Nielsen syndrome (long QT syndrome with deafness).

Autosomal dominant —Waardenburg syndrome types I and II, neurofibromatosis I and II, and branchio-oto-renal syndrome. Hearing impairment, including SNHL, conductive loss, or both, is also common in patients with velocardiofacial and Williams syndromes.[4]

X-linked - Hunter syndrome (mucopolysaccharidosis 2), Alport syndrome, X-linked congenital SNHL, and early onset progressive sensorineural hearing loss.[3, 5, 6]

Mitochondrial — less than 1 % of all HHI. Hearing loss may be isolated or associated with other features of mitochondrial disorders: lactic acidosis, encephalopathy, myopathy, seizures, ophthalmoplegia, diabetes mellitus, cardiomyopathy, stroke-like episodes, ataxia, and optic atrophy.5

HERIDITARY-NON-SYNDROMIC:

Genes responsible for Nonsyndromic causes are-

a. DFNB1 disorder, caused by mutations in the GJB2 gene (which connect the protein *connexin* 26)
b. The GJB6 gene (which encodes protein *connexin* 30), accounts for 50% of AR Nonsyndromic hearing loss.

Congenital hearing losses may not be associated with obvious physical findings. Nonsyndromic hearing loss constitutes most cases of congenital sensorineural hearing loss. Autosomal dominant delayed onset progressive sensorineural hearing impairment is one such type, the gene for which is located at chromosome 5.

X-Linked recessive mixed hearing loss with stapes fixation and perilymphatic gusher is another entity which is non-syndromic, but would be evident if other family members had been affecte. The hearing impairment might be mild to severe, and any attempt to remove or mobilize the fixed stapes would lead to a sudden rush of perilymphatic fluid leaking out of the vestibule of the inner ear, thus leading to a risk of profound hearing impairment.

A congenital ossicular chain atresia, malformation, fracture or dislocation can cause congenital hearing loss by affecting the middle ear. Typically, absence or malalignment of the cura of the stapes, an abnormality of the incus or malleoincudal joint, a fixation of the malleoincudal joint or one of the ossicles to the scutal or attic ridge are seen.

Congenital malformations (dysplasias) of the inner ear cause SNHL. These can include a complete lack of inner ear, incomplete development and malformation, membranous

cochleosaccular degeneration and malformation of the cochlear membranous system. Genetic counseling is recommended for the parents, as well as for the child, since inner ear abnormalities may be associated with other abnormalities or may be hereditary

I(b) NON-HERIDITARY:

Nonhereditary etiologies involve an insult to the developing cochlea, such as intrauterine infection, medications, or toxins that have a teratogenic effect on the developing ear of the fetus (eg, alcohol, methyl mercury, quinine, trimethadione, retinoic acid, and maternal thyroid peroxidase autoantibodies).[7, 8]

Commonly seen non hereditary causes of congenital hereditary hearing loss include congenital infection caused by cytomegalovirus (CMV), toxoplasmosis, rubella, or syphilis which are associated with SNHL.

Important Congenital Infectious Causes of SNHL in neonates and children

- Congenital cytomegalovirus infection — Congenital CMV is the most common intrauterine infection in humans as well as the leading infectious cause of sensorineural deafness.[9, 10, 11] Even among asymptomatic children with documented CMV infection in one study, 7 % developed SNHL.[12] The hearing loss was progressive in 50 percent and delayed in onset (median age 27 months) in 18 percent. It can be either unilateral or bilateral.
- Congenital toxoplasmosis — Congenital toxoplasmosis occurs in one in 1000 to 3000 births. Ninety percent of the cases are asymptomatic at birth. SNHL occurs in 10 to 15 percent of infected children[13] and is usually delayed and progressive.[14]
- Congenital rubella — Although the incidence of congenital rubella has declined by more than 99 percent since the introduction of the rubella vaccine in 1969[15], approximately 5 to 25 percent of women of child-bearing age remain susceptible to rubella infection.[16] Hearing loss affects 68 to 93 percent of children with congenital rubella; it is usually profound and bilateral and sometimes progressive[7].

- Congenital syphilis —One-third to two-thirds of infected infants are asymptomatic at birth [7, 16]. SNHL, which occurs in 30 to 40 percent of these infants, is a late manifestation of congenital syphilis and appears after two years of age.

The hearing loss frequently is progressive or delayed in onset, emphasizing the need for universal newborn hearing screening and continued monitoring of children with known congenital infections.

Low birth weight of the baby, an important consideration for a country like India, is seen to be associated with congenital hearing loss. The prevalence of congenital SNHL among babies with low birth weight (less than 1500 g) is 51 per 10,000.[17] The high rate of hearing loss among these children has been linked to several factors, including administration of ototoxic drugs such as aminoglycoside antibiotics, ambient noise produced by the incubator[18], and perinatal complications (eg, hypoxia and acidosis).[19, 20]

A high uncontrolled level of bilirubin is also associated with congenital hearing loss. *Hyperbilirubinemia* in infants is frequently caused by hemolysis. Causes of hemolysis include Rh or ABO blood group incompatibility and glucose-6-phosphate-dehydrogenase deficiency.

II. NON-CONGENITAL OR ACQUIRED

Important acquired causes of hearing loss in this age group would include drug toxicity, meningitis, trauma, noise induced HL, infection with mumps, measles, AIDS and chronic otitis media.

Ototoxic drugs such as aminoglycosides, high dose intravenous loop diuretics, and chemotherapeutic agents such as cisplatin can cause significant hearing loss[21, 22].

- Aminoglycosides —The relative order of cochleotoxicity is gentamicin>tobramycin>amikacin>neomycin. There also appears to be a genetic predisposition to the development of ototoxicity with aminoglycosides[23, 24].
- Other antibiotics —erythromycin, vancomycin, and tetracycline.
- Chemotherapeutic agents —cisplatin, 5-fluorouracil (5-FU), bleomycin, and nitrogen mustard. The worst ototoxicity occurs with cisplatin, which causes hearing impairment in 10 to 26 percent of children[25, 26].
- Antimalarial medications such as quinine and chloroquine

Bacterial meningitis is the most common cause of acquired deafness in childhood[27, 28].The reported frequency of persistent hearing impairment varies from 2.5 to 18 percent in survivors[27, 28, 29, 30]. Another 10 percent of children have transient hearing impairment[30].

Sensorineural hearing loss occurs early in the course of bacterial meningitis (within the first 48 hours), with possible recovery or worsening during the first two weeks of illness[31, 32].Permanent hearing loss may be caused by damage to the cochlea, labyrinth, or eighth cranial nerve from direct bacterial invasion or the inflammatory response elicited by the infection[29, 33].

A short blast of loud noise also can cause severe to profound SNHL, this reaction usually involves exposure to noise greater than 120 to 155 dB.

Trauma to the temporal bone usually causes sensorineural or mixed hearing loss. Both penetrating and blunt trauma can result in transverse temporal bone fractures. Penetrating trauma is typically due to gunshot wounds and blunt trauma to blows to the occipital or frontal regions. Blunt trauma also causes sensorineural hearing loss when concussive forces are exerted on the inner ear fluids, causing a shearing effect on the cochlear organ of Corti.

Chronic otitis media, a common cause of ear morbidity in Indian children, causes conductive type of hearing loss by affecting the middle ear. By the age of three years, 83 percent of children will experience at least one episode of OM, and 46 percent will have at least three episodes[34]. About 3 to 20 percent of children have six episodes of OM in a year[35].

Small, retrospective, case-control, and longitudinal cohort studies have noted that children who have prolonged middle ear effusion have lower scores on tests of speech, language, and cognitive abilities[36, 37, 38].

However, a study in which children with persistent middle ear effusion were randomly assigned to prompt or delayed insertion of tympanostomy tubes found that developmental outcome at 3, 4, 6, and 9 to 11 years of age did not differ between the groups[39, 40, 41, 42, 43].

Less common causes of acquired hearing loss in this age group can be obstruction due to cerumen in the ear canal, which causes hearing loss of the conductive type and any infection causing otitis externa, which can lead to blockage of the EAC caused by the accumulation of debris, edema, or inflammation.

Most cases of hearing loss in this age group are of Sensorineural type (SNHL), conductive type being less common. These are due to an affected inner ear and imply problems at the level of the cochlea, internal auditory canal, eighth cranial nerve, or brain (CNS).

Sensorineural Deafness can be-

i). Prelingual (before the child acquires the ability to speak)
ii). Postlingual (after the child acquires the ability to speak)

While the Prelingual group can be identified with the universal newborn hearing screening the post lingual group can be missed out. In developing world – infection is the most common cause of hearing loss. This includes congenital conditions, such as rubella and cytomegalovirus and acquired infections such as mumps, measles, meningitis and chronic otitis media. In the developed countries about half of children with permanent congenital hearing loss have a genetic cause for their deafness. (Scott-Brown)

In general AR forms Prelingual deafness and AD tends to present later and result in progressive Postlingual deafness.[44]

CENTRAL TYPE OF HEARING LOSS

Central processing of sound occurs after the sensory reception of auditory signals through the auditory apparatus, but prior to language processing or comprehension. A Central type of hearing loss or a Central auditory processing disorder (CAPD) is described as a deficit in the perception or complete analysis of auditory information that is associated with dysfunction of the central auditory nervous system, most commonly the cerebral cortex[45, 46].

Risk factors would include in-utero infections, bacterial infection, hyperbilirubinemia, asphyxia, head trauma, and other neurological insults during infancy. Where regular audiologic testing in children cannot explain the auditory perceptual problems, a differential diagnosis of CAPD should be considered.

SUMMARY

The hearing loss in neonates and infants can be congenital or non-congenital. Congenital can be hereditary (i.e. cause of a disorder involves abnormal genes) or non-heriditary. Hereditary causes may present with other associated abnormalities (syndromic) or may be non-syndromic. Non-hereditary or non-inherited causes commonly are Rubella, CMV infection, etc. Non-congenital causes include ototoxicity due to drugs, mumps, meningitis, any trauma or noise induced hearing loss, etc. It is essential that cause of hearing loss need to be understood in its presentation for it to be suspected, and identified at the earliest possible stages in life. Early detection, diagnosis and rehabilitation of hearing loss are necessary for the development of appropriate speech, language and cognitive abilities. Success will depend on the early and precise detection and management of disability, thus a timely knowledge of the cause of the hearing loss would incite the development of an appropriate management of the patient to avoid compromising the educational progress.

Advances in molecular biology in investigating hereditary hearing impairment are occurring rapidly, thus the relative importance of these causes will increase. This becomes all the more important, as with time the prevalence of infectious causes of hearing impairment are expected to decline in developing countries.

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Chapter 4: Ideal and actual age of identification of Hearing Loss

An appropriate age of hearing loss screening compatible with the health systems of a country is an evidence based approach to determine a national clinical and public health practice. Epidemiologists and public health specialists always recommend an earlier screening for any disease that allows such an assessment, and in pure economic terms, for resource constrained countries where such measures decrease the need for medical intervention may also lead to financial savings.

The world-wide incidence of profound hearing loss is estimated to be approximately 1 in 1000 live births. Prevalence of deafness in India is more likely to be of significant public health concern, considering its large 1.21 billion population and the large prevalence of consanguineous marriages.

Universal Newborn Hearing Screening (UNHS) is mandatory in the United States of America. The Joint Committee on Infant Hearing (JCIH, 1994) has also recommended that, all infants with hearing loss should be identified by 3 months of age, and receives intervention by the age of six months. Unfortunately, there is no policy or regulation mandating the hearing screening of newborns in India, thus the identification of hearing loss is mostly at later ages.

Development of the auditory system begins early in fetal life, with the first sign of expansion of auditory brain-stem pathway occurring by the 16^{th} week of gestational period. Auditory brainstem responses can be actually recorded by the end of 28^{th} week of gestation which corresponds to the age of fetal hearing with the appearance of synapses. Few weeks before birth, the inner ear and functional auditory brainstem pathway develops, as a result a normal newborn baby can process the sound waves and accurately analyze the loudness and the pitch of sound. Localization of sound and speech discrimination occurs in early infancy, thus normal infants with hearing ability learn to process speech and understand the language by the age of one. Morphological and functional properties of neurons are affected adversely if there is any deprivation in the sensory input, which can be ameliorated by the re-introduction of sensory stimulation during this period.[1]

Hearing impairment goes undetected until as late as two years of age without a specialized screening test.[1, 2] It is seen that most cases are then identified by the parents of these children, indirectly, since the children do not start achieving language milestone that correspond to their chronological age. The mean age of presentation of hearing impairment without screening is approximately thirty months and the critical period of the language development starts before six months of age[2]. This time lag of the detection of hearing impairment in the population can be reduced by an early screening protocol.

RATIONALE FOR SCREENING —

The incidence of congenital permanent hearing loss is 1.5 to 6 per 1000 live births[3]. Recent study done in India reported 5.6 per 1000 live birth[4]. Genetic factors contribute to 50% of the childhood hearing impairment. Prenatal, perinatal and postnatal environmental factors account for 20-25% and etiology is uncertain in 25-30%of cases[5]. Unless screened, hearing impairment remains undetected during infancy.

Screening newborns for hearing loss leads to earlier detection and intervention in patients with congenital hearing impairment. Early intervention appears to improve speech and language development, and educational achievement in affected patients[6].

The incidence of hearing loss is much higher than the usually screened conditions like congenital hypothyroidism (0.25 in 1000 live births) and phenylketonuria (0.08 in 1000 live births)[7]. The critical period of speech and language development is 0-3 years[2, 5, 8, 9, 10, 11, 12].

Cost effective screening tools like Otoacoustic emission (OAE) and auditory brain stem response (ABR) are readily available. These tests have high sensitivity (92%) and specificity (98%) in early neonatal period itself.[5] There is easy availability of cost effective intervention which, if utilized appropriately, can correct the disability.

Earlier diagnosis — Screening newborns detects patients with hearing impairment at an earlier age than relying solely on identifying clinical signs of hearing loss.[13, 14] Parents, caregivers and or clinicians do not usually detect hearing loss until there is a delayed speech and language milestone in the child.

A controlled trial of 53,781 newborn infants who were born in four English hospitals from 1993 to 1996, that alternated periods of time between universal newborn hearing screening (UNHS) and no screening was carried out by the Wessex Universal Neonantal Hearing Screening Trial Group.[13] Overall, 106 cases of bilateral permanent hearing impairment per 100,000 target population were identified. After adjustment for severity of hearing loss, infants with hearing loss born during periods of screening compared to those without screening were more likely to be detected at an earlier age (odds ratio [OR] = 5, 95% C.I. 1.0-23.0) and to receive earlier intervention (OR= 8, 95% C.I. 1.5-41.0). In the follow-up study, published consequently in the same journal, it was seen that the proportion of seven- to nine-year-old children with permanent hearing impairment who were identified before six months of age was higher during periods of UNHS compared to no screening (74% versus 31 %).[15]

Intervention and outcome — in literature, there are few controlled studies of long-term outcome, but available evidence clearly shows that early intervention in hearing impaired infants is promising and that it can definitely improve the language and developmental outcome of these children. [16, 17, 18, 19, 20, 21, 22, 23]

Kennedy CR et al in a controlled study of 120 children with bilateral permanent hearing loss who were from the birth cohort of the previously mentioned trial comparing universal newborn hearing screening (UNHS) to no screening, demonstrated the beneficial effect of early detection and intervention on language development.[22]

The following findings were noted when children who were diagnosed with hearing loss by nine months of age were compared to those diagnosed after nine months of age at eight years of age:

- All the patients who were detected by nine months of age had been identified by UNHS.
- Children whose hearing loss was confirmed by nine months of age compared to those with confirmation of permanent hearing loss after nine months of age had better receptive and language abilities.
- There was no difference in speech development between the two groups.
- There were no differences in the severity of hearing loss, the frequency of other disabilities (such as cerebral palsy, visual impairment and learning disability), or parental socioeconomic status between the two groups.

In a subsequent report by Mc Cann DC et al, 2009, it was observed that patients who were identified before nine months of age compared to those diagnosed later also had better reading and communication skills when they were evaluated at primary school age.[24] Compared with birth during periods without UNS, birth during periods with UNS was associated with better reading scores (inter-group difference 0.39 SDs, 95% CI 0.02 to 0.76, p = 0.042) and communication skills scores (difference 0.51 SDs, 95% CI 0.06 to 0.95, p = 0.026). Compared with later confirmation, confirmation of PCHI by age 9 months was also associated with better reading (difference 0.51 SDs, 95% CI 0.15 to 0.87, p = 0.006) and communication skills (difference 0.56 SDs, 95% CI 0.12 to 1.00, p = 0.013). In the children with PCHI, reading, communication and language ability were highly correlated (r = 0.62-0.84, p<0.001). Thus, in conclusion birth during periods with UNS and early confirmation of PCHI predicted better reading and communication abilities at primary school age. These benefits represent functional gains of sufficient magnitude to be important in children with PCHI.

Similar results were reported by Korver AM et al from a population-based study from the Netherlands that compared outcome at three to five years of age of children with permanent hearing impairment identified by newborn screening with those diagnosed by distraction screening at nine months of age between 2002 and 2006.[23] Multivariate analysis showed the newborn screening group had higher scores for developmental, social development, gross motor development and quality of life testing when evaluated at age three to five years of age. The authors concluded that compared with distraction hearing screening, a newborn hearing screening program was associated with better developmental outcomes at age 3 to 5 years among children with permanent childhood hearing impairment.

In a prospective study (Vohr B et al) from the Rhode Island UNHS program of infants with hearing loss, enrollment in an early intervention group before three months of age compared to enrollment after three months of age appeared to have beneficial effects on language when patients were tested at 12 to 16 months of age.[25] However, this study was limited by the small number of patients with hearing loss in each group, especially those with moderate to profound early loss. This was a prospective longitudinal study of the outcomes of a cohort of 30 infants identified in the Rhode Island universal newborn hearing screening program and 96 hearing

control subjects. Child language skills were assessed by using the MacArthur-Bates Communicative Development Inventory, Words and Gestures, at 12 to 16 months. It was seen that children with moderate/profound hearing loss had significantly lower numbers of phrases understood, words understood, and early, later, and total gestures, compared with children with mild/minimal hearing loss and hearing control subjects. Furthermore, children with hearing loss who were enrolled in early intervention at</=3 months had significantly higher percentile scores for number of words understood, words produced, and early, later, and total gestures, compared with those enrolled at>3 months. Regression analyses to test the independent effects on language skills of children with hearing loss identified enrollment in early intervention at</=3 months as an independent predictor of percentile scores for word and early gesture production. The authors concluded that very early enrollment (</=3 months) in early intervention has beneficial effects on early language for children with hearing loss. Nevertheless, 12- to 16-month-old children with moderate/profound hearing loss exhibit delayed receptive and expressive language skills in oral and signed English modes, compared with peers with either mild/minimal hearing loss or typical hearing sensitivity

One study evaluated the language and speech outcome of infants identified at 13 to 30 months of age who had mild to profound hearing loss.[18] Intervention programs had been initiated in all patients once hearing loss was identified. Patients who were diagnosed before six months of age compared to those diagnosed later had better outcomes. Adjusted mean Minnesota Child Developmental Inventory scores were significantly higher for those identified before six months for receptive language (79.6 versus 64.6), expressive language (78.3 versus 63.1) and total language quotients (79 versus 64), compared to those identified after 13 months of age. For children with normal cognitive abilities, this language advantage was found across all test ages, communication modes, degrees of hearing loss, and socioeconomic strata. It also was independent of gender, minority status, and the presence or absence of additional disabilities. Methodological problems that may have influenced the results included differences between groups in maternal education, severity of hearing loss, and cognitive impairment, as well as no information on subjects lost to follow-up.

The effect of screening on outcome was evaluated in a report by Yoshinaga-Itano C et al that compared language performance in 25 children with hearing impairment born in hospitals with

UNHS to 25 matched patients born at sites without UNHS.[26] Average scores for expressive, receptive, and total language were 18 to 21 points higher (approximately one standard deviation) in the screened compared to the unscreened group. More children in the screened group had normal language development (56 versus 24 percent). Newborn screening programs for hearing loss are positively related to scores in expressive and receptive language ($p<0.001$) and vocabulary production ($p<0.001$) on standardized inventories; speech intelligibility ($p = 0.015$) from independent ratings; number of different simple consonants ($p<0.01$) and consonant blends ($p = 0.026$) from phonological transcripts; and total number of intelligible words ($p<0.01$) and number of different words produced ($p<0.01$) from computer analysis of videotaped language samples. In conclusion it was stated that Hospital-based newborn hearing screening programs are positively related to language and speech performance for children in early intervention programs who are deaf and hard of hearing. Limitations of this study included unmasked assessment of outcome, selection of subjects through their enrollment in the CHIP program, and no information on subjects who were excluded.

SUMMARY

Newborn screening leads to the detection and treatment of hearing loss at an earlier age, and there is evidence that earlier intervention for a positive screen improves language development and communication. The evidence may be limited by the quality of the data, but one controlled trial reported improved language development in children identified before 10 months of age,[22] which is supported by evidence from cohort and case-control studies. [18, 26,27]

Screening needs to be done with minimal false positivity and minimum delay, as any delay in diagnosis has consequences on the timely and preventive management to prevent any irreversible damage to the child's development.

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Chapter 5: Modalities for Early Identification of Hearing Loss

Newborn screening for hearing loss is not a novel method, and has evolved over past many decades. Objective tests used in the past for screening were not so advanced or with good predictive values, thus they with medical practice have seen through an evolution. Thomas Barr in late 1890's introduced sound bells and sharp voice for evaluation of hearing. Though these tests were in use till 1940's they had high false positivity rates and a diagnostic drawback that mild and moderate hearing loss could not be detected. During the period of 1940-1950, electro-dermal response audiometry was used for screening. In 1974, Blair Simmons developed an automated technique of behavioral screening, known as the Crib-o-gram. This test utilized motion sensitive transducers attached underneath the infant's bassinets, an automatic timer, a loudspeaker and a strip that recorded motion. Baby's movements were charted after a high frequency signal of 92dB was presented in scheduled interval. The incidence of hearing loss with Crib-o-gram that could be identified was about 1 in 1000 in well-baby nursery and 1 in 52 in intensive care problems. The results depended upon the behavioral state of the baby, optimal stimulus, and the ambient noise. It had a high false positivity and negativity with poor reliability.[1, 2] Only later did the two tests namely Oto-acoustic Emission (OAE) and Auditory Brainstem response (ABR) were used and found to be most useful for screening of hearing defects in infants.

Who should be screened?

In early 1970s, the National Joint Committee on Infant Hearing Screening (JCIH), composed of representatives of Academy of Pediatrics, Academy of Otolaryngology and American Speech and Hearing Association (ASHA) recommended the high risk register for hearing evaluation.[1]

The five categories included were:

1. A history of hereditary childhood hearing impairment.
2. Rubella or other nonbacterial intrauterine fetal infections
3. Defects of ear, nose or throat.
4. Birth weight < 1500gms
5. Any free or indirect serum bilirubin concentration judged to be potentially neurotoxic.

Prevalence of hearing impairment in high risk groups was seen to be 14 times higher than the general population. This concept, being a high risk approach towards hearing evaluation by screening assumed that one can identify small group of children possessing high chance of having the handicap. Although there was an observed (and expected) increased incidence of hearing impairment in this group, nevertheless, it was also seen that almost 50% of the children with significant hearing loss did not have risk factors in question.

Consequently, in 1993, the National Institute of Deafness recognized the above issue and recommended a Universal hearing screening within first three months of birth. The recommendation consisted of a more robust two stage screening protocol, with Otoacoustic emission (OAE) followed by an Auditory brainstem response (ABR) for infants who fail OAE.

In the year 1994 the JCIH recommended identification of the impairment by three months and intervention by six months.[1]

In 2000, JCIH recommended the following:[3]

1. All babies should be screened before one month and all babies who required admission in NICU should receive screening before discharge.
2. All infants who do not pass birth admission screen should be screened before three months of age.
3. All infants with confirmed permanent hearing loss should receive services before six months of age.
4. All infants who pass screening and have risk factors should receive ongoing audiological and medical surveillance and monitoring for communication development.
5. Infants and family rights are guaranteed through informed choice, decision-making, and consent.
6. Infant hearing screening and evaluation results are afforded the same protection as all other health care and educational information.
7. Information system used to measure and report the effectiveness of early hearing detection and intervention (EHDI) services.

In 2007 the JCIH updated their year 2000 statement. These included definition of targeted hearing loss, hearing screening and re-screening protocol, diagnostic audiological evaluation,

medical evaluation, early intervention, surveillance and screening in medical home, communication and information infrastructure. The definition was expanded to include neural hearing loss (auditory dys-synchrony) in infants admitted in NICU.

Recently, in July 2008, United State preventive service and task force made following recommendations (B recommendation):[4]

1. Screening for all newborns within one month with two step screening process, first with OAE and followed by ABR who fails the first test.
2. Newborn delivered at home and hospitals without hearing screening facilities, should have some mechanism for referral
3. Early intervention should be designed to meet the individual needs of the infant and the family
4. Babies with risk factors should undergo periodic screening evaluation till three years.

Techniques for identification of hearing loss may be:

- Subjective methods - Behavioral Observation Audiometry (BOA), the Distraction test, Visual Reinforcement Audiometry (VRA), Pure Tone Audiometry (PTA)
- Objective methods: electrophysiological tests include the Otoacoustic Emission (OAE), Auditory Steady State Response(ASSR), Auditory Brain Stem Response(ABR).

The various available methods such as BOA, OAEs, AABR and BERA and ASSR form the bed-rock of early diagnosis of hearing impaired infants.

Behavioral Response Assessment by Distraction testing (BOA)

The baby is observed in response to sound stimuli.

I. response behavior up to 4 months of age, to sound stimuli include-
   a. eye widening,
   b. eye blink (auropalpebral reflex)
   c. arousal from sleep
   d. startle or shudder of the body of definite movement of the arms , legs or body.

II. From 4-7months
   Lateral inclination of the head towards the sound or listening attitude.

III. From 6-12months-
   Definite eye movement/listening/turning of the head towards the sound.

If all of these are absent the result is considered as Refer.

The test is performed with the child cradled on the mother's lap, his/her attention is lightly engaged and a sound is presented for < 2seconds in a horizontal plane behind the ear approximately 15 cm out of the child's visual field. The distracter observes for the response.
BOA under 6 months of age has largely been superseded by the availability of electrophysiological techniques which are objective in nature.

Automated Behavioral method

The Auditory Response Cradle (ARC) was developed for screening of normal term infants. It has a pressure sensitive mattress and head-rest which monitor head movements, body activity and respiratory activity. Sounds with high intensity of 85dB are presented to the baby via headphones. The ARC compares reactions to sound with reactions when no sound is being produced. The behavior of the baby is analyzed and a pass or refer outcome is given.
Advantages of this technique is that the whole auditory pathway is screened, it is non-invasive, no application of electrodes required, relatively simple, easy and quick taking only few minutes.

It's disadvantages are it cannot detect mild hearing loss, is not suitable for preterm or sick neonates and results are somewhat subjective. However it is reported to have a high detection performance for the serious hearing impairment and relatively low false-positive results (1.3%).[5] Another automated behavioral screening method similar to but less sophisticated then the ARC is the Crib-o-Gram. Its sensitivity (75%) and specificity (71%) are too low when compared to ABR and hence not recommended by the Joint Committee on Infant Hearing 1994 statement.

The Distraction test: This test is performed for babies between 6-18months of age. When the child can sit erect unsupported and able to perform head turns in a horizontal plane. Normal children in this age group will response to sound presented to their ear by turning their head toward the source of the stimuli. Habituation is more likely after 12 months. It remains valuable when used by a trained, experienced tester in an appropriate setting. The test is performed by two testers in a suitably sized room like in the BOA with ambient noise < 30dB and a suprathreshold stimulus is presented to the baby's ear. The distracter observes the child's response. Loss of interest suprathreshold responses, inaccurate frequency and stimulus estimations are some of its pitfalls.[6]

Visual Reinforcement Audiometry (VRA): Key developmental age is 6-36 months. Children can be trained by Operant condition to produce a localizing turn to a visual stimulus in response to a sound stimulus. This is the principle of VRA. The test is performed in a sound treated room with the child on the parent's lap in front of a low table. A second tester may sit on the other side of the table or adjacent to the child to provide low level play activity. The speaker is placed at 45, 60 or 90 degree from the child at the same height of at least 1m from the ear, well calibrated. Frequency specific sound stimuli is presented to the ear (soundfield testing). Alternatively the sound can be delivered via headphones using foam tips or ear moulds if available. First 2000Hz if conductive hearing(CHL)loss is suspected and then proceeds to 500Hz or start with 500Hz if sensorineural hearing loss(SNHL) is suspected and proceed on to 2000Hz directly. Since most patients with CHL hears better at higher frequencies and those of SNHL hears better in low frequencies. The reinforce acts as a reward and therefore increasing the appeal to the child to reduce habituation and sustain interest. Further development and modification of the technique have been described by Liden and Kankkunen.[7, 8]

How should we screen the infants?

The American Academy of Pediatrics (AAP) Task Force on Newborn and Infant Hearing defined an effective neonatal hearing screening test as one that detects hearing loss of ≥35 dB in the better ear and is reliable in infants ≤3 months of age.[9]

Two electrophysiologic techniques meet these criteria:

- Otoacoustic emissions (OAE)
- Automated Auditory brainstem responses (AABR)

Evoked OAE and AABR are thus the two screening tests recommended for hearing assessment.[1, 10]

Other forms of hearing evaluation include electrocochleography, visual reinforcement audiometry (VRA), play audiometry and sound field audiometry. However, only OAE and ABR are suitable for neonatal screening.[11, 12]

Otoacoustic emissions —

David Kemp discovered OAEs in 1978. Normally "echoes" are generated by the cochlear hair cells when presented with sound waves. Otoacoustic emissions are acoustic energy originating in the normal inner ear, which can be detected by acoustic probe connected in the external ear canal. These are produced by the action of cochlear outer hair cells. A microphone at the external ear canal detects these low-intensity OAEs. There are two types of OAE. One is spontaneous, present in the absence of external stimulation and the second is evoked OAE. Evoked can be further divided into transiently evoked OAE (TEOAE) which are evoked by acoustic transient such as click or tone burst, stimulus frequency OAE (SFOAE), elicited by single continuous tone, and distortion product OAE (DPOAE), which are generated by two continuous pure tone separated by specific frequency. Emissions are not measurable if conductive or sensorineural hearing loss exceeds 30dB to 35dB HL at 0.5 kHz, 2 kHz and 4 kHz and 40dB at 1 kHz. Background noise in nursery does not interfere with the procedure if the probe is well fixed, but internally generated noise, such as sucking or swallowing may interfere in estimating low frequency emissions.[11, 12, 13, 14]

Technique — The apparatus for OAE screening consists of a miniature microphone placed into the infant's outer ear canal. The microphone produces a stimulus (clicks or tones) and detects sound waves as they arise from the cochlea. The device also measures the signal-to-noise ratio to ensure accuracy.

The sound waves occur spontaneously in approximately 40%-60% of general population and can be evoked by various stimuli in 92-100% of a normal hearing population. They are non-linear acoustic signals produced by the outer hair cells of the Organ of Corti. Most intensity of OAEs is between -10 and 20dB. Sound produced by the outer hair cells travels in a reverse direction from:

Outer hair cells → *Basilar membrane* → *Perilymph* → *Oval window* → *Ossicles* → *Tympanic membrane* → *Ear canal* which is then pick up by the microphone.

The tests most commonly used for clinical purposes are the distortion product OAEs (DPOAEs) and the transient OAEs (TOAEs). They are classified by the stimuli used to produce the cochlear basal membrane vibrations.[15, 16]

- DPOAEs are detected when the cochlea is stimulated with two specific frequencies, F1 (65 dB) and F2 (55 dB) that result in a single predictable frequency response. These frequencies travel through the middle ear to the cochlea, where a third tone is generated at the outer hair cell level. Normal cochlear stimulation produces DPOAEs at a specific frequency that is predicted by the formula, (2 x F1) - F2.
- TOAEs utilize a click stimulus that results in the emission of several frequencies at the same time.

These measurements are valid provided that the patient's middle ear function is normal, can be used to assess cochlear function for the 500-6000 Hz frequency range. The presence of evoked OAE responses indicates hearing sensitivity in the normal to near-normal range. Depending on the stimulus used OAEs can be detected in up to 98% of humans with normal hearing and are absent when there is a hearing impairment of more than 30dB

OAE testing generally requires approximately four to eight minutes. Approximately 5 to 21 percent of infants screened with this technique are referred for further audiologic evaluation.[17]

Jacobson and Jacobson recorded the time for OAE testing < 3min for both ears. But the actual mean time to obtain to obtain a result due to noise, myogenic activity, relocation of probes was 16.6 min (range 7-45 min) under these conditions the specificity and sensitivity were 52% and 50% respectively.[18]

Edward L. McNellis et al found that among 50 low risk infants screened by ABR there was 98% initial passing rate whereas the OAEs was 61% of 100 ears, which improved with each successive retest and approximated the ABR rate by the 4th test.[19]

A study on Rhode Island on more than 4000 newborns reports an overall OAEs passing rate of 93percent.[20]

Thus, use of automated OAEs is a convenient screening method. The technique is quick, non-invasive, and objective in nature and gives specific information of cochlear function. The inbuilt software keeps noise level rejection limit and gives the result as pass or refer. This does not require electrode placement or the audiologist to interpret it. However, the effectiveness of the test is reduced by contamination with low-frequency ambient noise in a busy nursery, vernix in the ear canal, or any middle ear pathology.

Auditory Brainstem Response (ABR)

Auditory brainstem response is the second screening test used if babies fail in the EOAE. The auditory stimuli produce electrical activity in the brain stem that can be detected by surface scalp electrodes. ABR is used for estimating thresholds, absolute latencies, and inter-wave latencies. There are seven positive waves, the presumed origins of which are as follows:

Wave I – VIIIth nerve
Wave II – cochlear nucleus
Wave III – superior olivary complex
Wave IV – lateral lemniscus
Wave V – inferior colliculus
Wave VI – medial geniculate body
Wave VII – auditory cortex

The auditory brainstem response (ABR) has been used for auditory assessment in infants since 1974.[21] A modified screening version of the ABR test has been available for screening since

approximately 1987. The screening ABR measures the summation of action potentials from the eighth cranial nerve (cochlear nerve) to the inferior colliculus of the midbrain in response to a click stimulus. Other names for this test include the screening ABR (SABR), brainstem auditory evoked response (BAER), and automated auditory brainstem response (AABR). Approximately 4 percent of infants screened with ABRs are referred for further audiologic evaluation.[22, 23] ABR is the screening test needed to detect auditory neuropathy.

The procedure is not affected by sleep or sedation. Advantages of this test include objective measurement of auditory system, provide ear specific information and are independent of subject state.[11, 12, 13, 14]

Technique — The screening ABR utilizes click stimuli presented at 35 dB. Three surface electrodes placed on the forehead, nape, and mastoid detect waveform recordings generated by the auditory brainstem response to the click stimuli. The morphology and latency of the waveforms are compared to normal and a pass or fail reading is generated. Delayed or absent waves suggest a neurologic or cochlear deficit.[22] Screening ABR requires 4 to 15 minutes for testing.

Comparison of OAE and AABR — The following is a comparison between the two neonatal screening methods:

Feature	**OAE**	**AABR**	**Comments**
Test time	less patient preparation time and a shorter test time than AABR[24]	Longer preparation and test time than OAE	OAE can be performed when the infant is awake, feeding, or sucking on a pacifier. AABR may also present time constraints because infants need to be asleep when tested

Interference	OAE is sensitive to background noise and noise generated by the baby.[15] This noise interference is greater when the recorded frequency is below 1500 Hz.	AABR, is not subject to movement artifact[16]	screening with OAE can be improved by recording higher frequencies, which are more important for understanding speech[15, 25]
False positive results	More infants appear to have hearing loss by OAE than by AABR during the first three days of life, in 19 to 25 percent of newborns screened by OAE during the first three days after birth[26, 27, 28, 29]	Less false positivity	most likely a false positive result that is due to occlusion of the external ear canal by vernix[16, 26, 27, 30] Cleaning of vernix can increase the pass rates.[27]
Tympanic membrane mobility	OAE requires a normal middle ear. Thus, decreased tympanic membrane mobility can reduce screening pass rates with this technique	Not so much affected	The magnitude of this problem was illustrated in a series of 200 infants, in which the pass rate in the 23 percent of infants with decreased tympanic membrane mobility was lower with OAE than AABR screening (33 versus 95 percent, respectively).[27]
Auditory neuropathy	OAE will not detect the hearing loss in	AABR will detect the hearing loss in infants	AABR should always be used to screen hearing in

	infants with auditory neuropathy, which may lead to a false negative result[31, 32]	with auditory neuropathy	infants who are at risk for auditory neuropathy (eg, infants with hypoxia, prematurity, hyperbilirubinemia, or neurologic impairment).
Relative costs	actual screening cost is lower for OAE	the overall cost of screening and audiologic evaluation may be lower with AABR because of the lower referral rate for audiologic assessment.	One study evaluated screening programs initiated at two sites, one using automated ABR administered by neonatal nurses and the other using OAE performed by master's level audiologists.[33] Less time was needed for testing with OAE than with ABR (5 versus 13 minutes), but the rate of referral for further testing was higher with OAE (15 versus 4 percent). Although the costs before discharge were similar for the two programs, the increased referral rate with OAE increased the overall cost per infant screened.

Both AABR and OAE techniques are inexpensive, portable, reproducible, and automated. Thus, they are well suited to newborn screening.[15] They evaluate the peripheral auditory system and the cochlea, but cannot assess activity in the highest levels of the central auditory system. These tests alone are not sufficient to diagnose hearing loss; thus, any child who fails one of these screening tests requires further audiologic evaluation. Also, AABR should be included in any screening program to detect hearing loss in the newborn, especially in populations (eg, premature infants) at risk for auditory neuropathy.

Auditory steady-state response (ASSR) Audiometry

Auditory steady-state response (ASSR) audiometry is a new, clinical electrophysiological tool that measures far-field electrical potentials in response to amplitude/frequency-modulated and frequency-specific auditory stimuli. This can give us a frequency specific response curve of the auditory system in normal individuals.

The Auditory Steady-State Response (ASSR) is similar to auditory brainstem response (ABR) in many ways. However, ASSR potentially evaluates four frequencies (500, 1000, 2000, and 4000 Hz) in both ears simultaneously, and ASSR can help differentiate severe from profound hearing loss—a distinction that is clearly meaningful and significant with regard to aural (re)habilitation and the selection of powerful hearing aids versus cochlear implants.

ASSR v/s ABR : ASSR and ABR use many of the same basic tools and protocols, The *similarities* between ABR and ASSR include:

- Both deliver an auditory stimulus to stimulate the auditory system via insert earphone,
- Both records electrical responses from the auditory system via electrodes
- Both are objective test method.
- Like ABR both the ears are tested simultaneously using insert earphones.

The *differences* between ABR and ASSR include:

- ABR stimulus is usually a click or a tone burst presented at a slower rate, whereas ASSR uses amplitude or frequency modulated sounds. presented rapidly to excite the auditory system while stimulating four

- ABR is highly dependent on a relatively subjective analysis of amplitude versus latency.
- ASSR is dependent on statistical analysis of the probability of a response, usually at 95% confidence limit. ASSR uses an objective, sophisticated statistics-based mathematical detection algorithm to detect and define hearing thresholds.
- The ABR response is measured in millionths of a volt (microvolts), and the ASSR is measured in billionths of a volt (nanovolts). ABR protocols typically use clicks or tone-bursts while ASSR can be used for evaluating broad bands or four frequencies (500 Hz, 1,000 Hz, 2,000 Hz, and 4,000 Hz) binaurally simultaneously.

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Chapter 6: Behavioral observational testing for the assessment of hearing loss in neonates

It is undoubtedly accepted that in any hearing disorder, however mild it may, it is essential to identify and accurately delineate it for further appropriate management. This is more true for the critical period of acquiring language skills in early part of life. However, it was commonly believed by many physicians, otolaryngologists and pediatricians that only gross behavioral assessment of hearing function can be obtained from infants younger than two years of age. This belief precluded the accurate delineation of audiometric configuration and degree of hearing loss for each ear and the determination of the type of impairment (conductive Vs. sensorineural). Such specific audiometric information is however extremely important for the medical, surgical and audiological management of this young pediatric population. Also, the ability to accurately assess the infant's hearing sensitivity using behavioral methods has expectedly improved over past decades.[1]

Pure tone or speech audiometry testing may not receive the cooperation of all children. In such cases, hearing can be assessed using behavioral methods in conjunction with other tests such as acoustic impedance testing, OAE, ABR, etc.

Behavioral observation audiometry — Behavioral observation audiometry (BOA) can be used to examine auditory function in infants younger than six to eight months, children with multiple handicaps, or adults who are not able to cooperate for other types of testing. It is a passive type of assessment in which the audiologist basically observes changes in behavior when various auditory stimuli are presented. Speech, live voice, warbled tones, noise makers, music, or narrow-band noises etc are presented in a sound field environment to elicit and observe any reflexive and orienting responses to auditory stimuli. The responses can include head or limb reflex, whole-body startle, sucking, eye blinking, raising of the eyebrows, or cessation of certain behaviors, such as movement or sucking. The responses to stimuli are not reinforced.[2, 3]

Behavioral observation techniques were first used in hearing screening of infants in the United States in 1964 when Downs and Sterritt[4] used the auropalpebral response, startle response, and lim and head movements to judge a response to auditory stimuli. Efforts were made to perfect these techniques that typically used high-frequency (3000 Hz warble tone or narrowband noise), very high intensity stimuli (90 to 100 dB SPL) and required trained observers.[5]

Behavioral observation is a subjective measure of hearing ability and does not provide ear-specific or frequency-specific information. It is best used in conjunction with objective methods. Since these responses are unconditioned, they can be highly variable, imprecise, habituate rapidly, influenced by the type of auditory stimulus used, and most often obtained using observers that may be biased.[6, 7]

BOA evaluations are usually completed in the sound field and thus are non-specific with regard to individual ear an bone conducted response system.[1]

A test technique that is better over the BOA, is called Visual reinforcement audiometry (VRA). It relies upon the principle of operant conditioning (stimulus, response, reward).

Visual reinforcement audiometry — Visual reinforcement audiometry (VRA) is a visually reinforced, operant test procedure in which an infant is taught to make a behavioral response contingent upon the presentation of an auditory stimulus. VRA can be used to evaluate the hearing of infants and young children from about six months of age through the second year. Studies have demonstrated that hearing thresholds obtained using the operant, head turn procedure are both accurate and reliable in infants as young as 5-6 months of age.[8] Sound stimuli (live voice and tones) are presented in the sound field and via insert earphones. The child is visually rewarded with lighted and animated toys for turning his or her head toward the sound source, so as to reinforce the behavioral response. The child is conditioned to perform this task repeatedly. An experienced audiologist will use several lighted and animated toys and an intermittent reinforcement schedule to maintain the child's attention. Once the contingent response is learned, the intensity if the auditory signal may be lowered and raised until a threshold is determined.[9] Under ideal circumstances, complete ear-specific information for speech stimuli and interactive frequencies from 250 through 8000 Hz can be obtained.

Play audiometry — Not the evaluation method in case of neonates, play audiometry can be used for hearing evaluation in children between the ages of 30 months and 5 years. Pure tones and speech information are presented via insert earphones. The child is taught to perform a simple task such as placing a block in a bucket or a peg on a pegboard each time a sound is heard. They are expected to perform the task over and over again when they hear the sound. The detection of speech sounds is determined by the child's ability to point to simple pictures when instructed by the audiologist.

Speech understanding or speech discrimination scores are obtained in children who are able to perform the tests. Such tests use picture-pointing tasks for younger children and require reading skills for older children. Speech-understanding testing is used to evaluate the child's ability to hear and understand speech in quiet and noisy listening environments.

Behavioral Audiometry: The Delaroche Protocol[10]

The *Delaroche Protocol* makes it possible from the age of 6 months to perform measurements with both a vibrator and headphones. The protocol has also been adapted for babies under 6 months of age, following on from neonatal screening.

Method/Principle of the Protocol

First, the vibrator is used to measure hearing thresholds by bone conduction. Here the function of the auditory nervous system can be examined directly without using the middle ear. Any pathology or infection of the middle ear would mask the real extent of the hearing impairment. Owing to the transcranial passage, only one bone conduction is needed to confirm the integrity of the auditory nervous system of at least one ear or the deterioration of both. Therefore, despite limitation in the intensity (45 dB at 250 Hz, 60 dB at 500 Hz and 70 dB at 1000, 2000, and 4000 Hz), measurement of hearing loss by bone conduction is the key stage of the diagnosis.

Secondly, the protocol allows the hearing thresholds to be measured in each ear by using headphones over the whole range of hearing frequencies (from 250 to 8000 Hz). Moreover, by using headphones, it is possible to deliver stimulations of up to 120/130 dB, values unattainable for sound field stimulations.

Thirdly, the method can be used to compare bone and air conduction (BC/AC) in order to specify the type of hearing impairment and, if necessary to quantify an air-bone gap.

Finally, the protocol is complemented by tympanometry and, in some cases for the study of stapedial thresholds.

Behavioral Screening in India: Role of a Community Health Worker

A formal screening at the subcentre level can be undertaken by the Multi-purpose worker. A questionnaire based assessment undertaken using a suitable, simply worded, pertinent questionnaire that is based on the high risk register and history regarding parental observation. If a child has a positive factor on history, irrespective of the result of behavioral testing, the child ought to be subjected to hearing screening. Those who are without any positive factor on history, but have failed on behavioral testing, should be reassessed at the next visit. If the baby's response still remains inadequate, the health worker should refer the baby to the district hospital for further assessment.

In all referred cases the health care worker must provide suitable guidance to parents regarding:-

- Need to get the child's hearing tested
- The fact that getting the hearing tested is a painless and easy procedure
- Need to do so at the earliest possibility
- Where and how the hearing tests can be done
- Need to follow the advice given by the doctor or audiologist

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Chapter 7: Hearing screening programs and protocols

Infant screening is defined as the use of a preliminary method for early detection of infants at increased risk of a serious morbidity or condition, with the expectation that a positive screen result will be followed by appropriate diagnostic tests and intervention. Early diagnosis followed by intervention or treatment is expected to either prevent or diminish the severity of the condition.[1]

Historical Evolution of Screening

Screening programs for hearing impairment have been both 'total population' and 'high risk population' (or selective) based. A successful total population, universal approach to diagnosing hearing loss within the first few months of life through a reliable and valid screening protocol was first reported in 1961 in the United Kingdom.[2] As a component of the national health system, "health visitors" screened hearing in the home, using a behavioral observation technique. Although it was subjective, less reliable, being limited by type of stimuli, this method still allowed all infants to be screened within the first year of life. Infants who seemed to have impaired hearing were further afforded an audiological evaluation to determine their hearing status at an early age. A similar national health program was also effective in Israel.[3]

In an attempt to develop a cost effective approach for infant hearing screening in United States, the Joint Committee on Infant Hearing (JCIH) in 1972 had recommended identifying infants "at risk" of hearing impairment. Initially they recognized five, and then seven criteria present at birth for which a high risk audiologic evaluation and follow up were recommended.[4, 5] In 1990, as a result of follow up data on the incidence of sensorineural loss from the neonatal intensive care unit, the JCIH guidelines were expanded to include 10 risk factors for neonates.[6] But it was observed that, on the basis of these criteria, only 2.5-5% of neonates with at least one risk factor had a sensorineural hearing loss, while, it was estimated that approximately 10-12% of neonates have an established risk factor.[7] Therefore, an infant hearing screen program that used the JCIH risk criteria was expected to substantially reduce the number of infants screened at birth. Restricting the number of infants screened at birth would obviously limit the number of children who are identified with an abnormality at an early age.

It was reported by Mauk et al[8] during this time that approximately half of the children who are ultimately identified with sensorineural hearing loss did not exhibit any risk factor at birth. Thus this evidence was clearly in support of the concept of a universal hearing screening.

Hearing Screening in Present Scenario

SELECTIVE or HIGH RISK SCREENING

This approach is directed towards newborns who are at increased risk for hearing loss. Major risk factors for neonatal sensorineural hearing loss (SNHL) include:[9]

- Admission to a NICU for at least two days.
- Syndromes associated with SNHL (eg, Waardenburg syndrome).
- Family history of hereditary childhood SNHL.
- Craniofacial anomalies (eg, anomalies of the pinna or ear canal).
- Congenital infection (eg, cytomegalovirus, toxoplasmosis, rubella, syphilis, herpes) or bacterial meningitis.
- Severe hyperbilirubinemia requiring exchange transfusion.

It is estimated that approximately 10 to 30 % of newborns would have one or more of these risk factors. A targeted or selective screening program towards the high risk neonates can identify 50 to 75 percent of all cases of moderate to profound bilateral hearing loss.[9, 10, 11] In a retrospective study of permanent childhood hearing impairment in the United Kingdom, screening newborns with a NICU admission, positive family history, or craniofacial abnormality detected more than 59 percent of infants with hearing impairment.[10] Based on evidence of the yield from hearing screens the authors suggest a wider implementation of neonatal screening and further consideration of the role of the health visitor distraction test in the identification of children with Permanent Childhood Hearing Impairment (PCHI).

However, as described before, selective screening alone would obviously delay the detection of hearing loss in a substantial number of children with congenital hearing loss.[9, 12, 13, 14, 15, 16, 17] Health care organizations, professional societies, and the United States Preventive Task Force recommend universal screening for newborn infants.[18]

In the Netherlands, the prevalence of hearing loss in infants admitted to the NICU was 1.8 percent and ranged from 0.7 to 3.7% between NICUs, in infants based upon data from the NICU hearing screening and the national neonatology databases for 2002 to 2005.[19] Risk factors associated with hearing loss included central nervous system disorders, cardiopulmonary compromise, intrauterine infection, and craniofacial, chromosomal and syndromal anomalies.

Infants who are admitted to the Neonatal ICU are at increased risk for hearing loss including auditory neuropathy (AN) with about a 2 percent rate of hearing loss. Xoinis K et al, in a review of 4250 infants screened by ABR for hearing loss in a single NICU from 1999 to 2003, 2.1 percent (95 neonates) failed the hearing screening. All patients had sensorineural hearing loss (SNHL), including 71 with cochlear abnormalities and 24 with auditor neuropathy.[20] With a SNHL prevalence of 16.7/1000, the rate for AN was 5.6/1000 NICU infants. Compared to infants with SNHL, infants with AN were significantly younger (GA 28.3+/-4.8 AN vs 32.9+/-5.2 weeks SNHL, P<0.0001) and smaller (BW 1318+/-894 AN vs 1968+/-1006 g SNHL). Nearly two-thirds of the AN infants were ELBW and had significantly longer hospital stays compared to SNHL infants of the same birth weight group. In conclusion it was seen that low birth weight NICU infants are at significant risk for AN. ELBW infants are at significantly higher risk for both AN and SNHL. Infants admitted to the NICU should be routinely screened by automated ABR and if abnormal, and further evaluation should be started before hospital discharge. Early identification of AN will result in better understanding of this disorder and lead to the development of appropriate intervention strategies.

In a nested case-control Norwegian study, for example, children with birth weights less than 1500 g had a six-fold greater risk for hearing loss compared to children with birth weights between 3500 and 3999 g.[21] Birth weight<1500 g, as compared with 3500-3999 g, gave an adjusted odds ratio for sensorineural hearing loss of 6.3 [95% CI 2.4, 16.4], controlled for gestational age, gender, parity, maternal age and concurrent birth defects. The risk of hearing loss decreased with increasing birth weights, with adjusted odds ratios of 4.4, 3.8, 1.7 and 1.4 for the birth weights 1500-1999, 2000-2499, 2500-2999 and 3000-3499 g respectively. The risk of both mild to moderate (MHL 35-70 dBHL) and severe/profound hearing losses (MHL>70 dBHL) were influenced by birth weight.

Because of the increased risk for SNHL, especially AN, in patients admitted to the NICU, the Joint Committee on Infant Hearing (JCIH), which encompasses seven national organizations including the American Academies of Pediatrics, Audiology, and Otolaryngology-Head and

Neck Surgery, recommends ABR screening for these patients since OAE screening will fail to detect those with AN.[22]

Post discharge follow-up— As a consequence of the evidence of risk of hearing loss for neonates admitted to the NICU, for any infant requiring more than five days of NICU care and one or more of the following risk factors:-

- Received extracorporeal membrane oxygenation (ECMO) therapy
- Received mechanical ventilation
- Exposed to nephrotoxic drugs such as aminoglycosides (eg, tobramycin and gentamicin) and loop diuretics (eg, furosemide)
- Hyperbilirubinemia that required exchange transfusion

The Joint Committee on Infant Hearing recommends at least one audiologic reassessment between 24 and 30 months of age.[22] This assessment is to be performed irrespective of the result of the newborn hearing screen prior to discharge.

As mentioned before, an association between birth weight<1500 g (very low birth weight (VLBW) and hearing loss has been long recognized. VLBW premature infants are at risk of experiencing progressive or delayed-onset hearing loss, and thus should continue to have serial hearing evaluations after discharge from the neonatal intensive care unit. As a result, follow-up monitoring with a diagnostic hearing test by 12 months adjusted chronologic age is recommended.

UNIVERSAL OR TOTAL POPULATION SCREENING

The goal of universal newborn hearing screening (UNHS) is early recognition and treatment of hearing impairment, which may maximize linguistic competence and literary development of children who are deaf or hard of hearing.[22, 23]

In the 1990's, the development of rapid, low-cost screening tests made it feasible for all newborns to be screened for hearing during birth hospitalization.

The impact of UNHS was demonstrated in an analysis by the Center of Disease Control and Prevention (CDC) based upon surveillance data from the Early Hearing Detection and Intervention Programs in all 50 states and United States territories.[24] Nearly 500 more infants with documented hearing loss were identified in 2007 compared to 2001 (1,736 identified in 2001 versus 2,212 in 2007) in the 21 states that had reported data for both years. As of 2007, 97 percent of infants were screened for hearing loss in the United States, which demonstrates progress toward achieving benchmarks for screening, evaluation, and intervention and document the continued need to ensure infants receive recommended services in a timely manner.

In a study from Belgium by Declau F et al,[25] a prospective analysis of 170 consecutive records of neonates referred to a tertiary center after universal neonatal hearing screening (UNHS) failure, between 1998 and 2006, was performed (from a database representing the equivalent of 87,000 screened newborns [0.2 percent]). Permanent hearing loss was confirmed in 116 children (68.2%). Bilateral hearing loss was diagnosed in 68 infants (58.6%) and unilateral hearing loss in 48 infants (41.4%). Median thresholds for the neonates with confirmed hearing loss were severe in both unilateral and bilateral cases, at 70 dB nHL and 80 dB nHL, respectively. In 55.8% of those cases, no risk factors for hearing loss were found. In 60.4%, the initial automated auditory brainstem response diagnosis was totally in agreement with the audiologic evaluation results. In 8.3% of the cases, however, a unilateral refer result was finally classified as bilateral hearing loss. An etiologic factor could be identified in 55.2% of the cases. Of the causes identified, a genetic mechanism was present in 60.4% of the cases, peripartal problems in 20.8%, and congenital cytomegalovirus infection in 18.8%. Thus in conclusion, an etiologic factor could be identified for nearly one half of the children with confirmed congenital hearing loss referred through a universal hearing screening program.

Guidelines for Universal Screening

The 2008 update of the 2001 US Preventive Services Task Force recommendation on universal newborn hearing screening weighed the benefits and harms of universal newborn hearing screening, incorporating new evidence addressing gaps identified in the 2001 US Preventive Services Task Force recommendation statement.

Nelson HD et al carried out a systematic review to update the 2001 US Preventive Services Task Force Recommendation on universal newborn hearing screening to detect moderate-to-severe permanent, bilateral congenital hearing loss. They focused on 3 key questions: (1) Among

infants identified by universal screening who would not be identified by targeted screening, does initiating treatment before 6 months of age improve language and communication outcomes? (2) Compared with targeted screening, does universal screening increase the chance that treatment will be initiated by 6 months of age for infants at average risk or for those at high risk? And (3) What are the adverse effects of screening and early treatment? Medline and Cochrane databases were searched by the authors to identify articles published since the 2002 recommendation. Data from studies that met inclusion criteria were abstracted, and studies were rated for quality with predetermined criteria. A good-quality retrospective study of children with hearing loss indicated that those who had early versus late confirmation and those who had undergone universal newborn screening versus none, had better receptive language at 8 years of age but not better expressive language or speech. A good-quality nonrandomized trial of a large birth cohort indicates that infants identified with hearing loss through universal newborn screening have earlier referral, diagnosis, and treatment than those not screened. These findings are corroborated by multiple descriptive studies of ages of referral, diagnosis, and treatment. Usual parental reactions to an initial non-pass on a hearing screen include worry, questioning, and distress that resolve for most parents. In conclusion, children with hearing loss who had universal newborn hearing screening have better language outcomes at school age than those not screened. Infants identified with hearing loss through universal screening have significantly earlier referral, diagnosis, and treatment than those identified in other ways.[26]

In the 2008 statement, the United States Preventive Services Task Force (USPSTF) recommended UNHS based upon its review of the evidence that demonstrated currently available inexpensive screening tests can accurately identify newborns with permanent hearing loss, and early detection improved language outcomes.[18, 26]

The USPSTF statement supports the recommendation of the Joint Committee on Infant Hearing (JCIH) in 2007 of implementing an integrated, interdisciplinary system of UNHS to detect and treat early hearing loss.[22] The JCIH includes seven professional societies; American Academies of Audiology, Pediatrics (AAP), Otolaryngology-Head and Neck Surgery, the American Speech-Language-Hearing Association (ASHA), the Council on Education of the Deaf, Directors of Speech and Hearing Programs in State Health and Welfare Agencies, and the Alexander Graham Bell Association for the Deaf and Hard of Hearing.

The JCIH and USPSTF recommendations include the following:[18, 22]

All newborns should be screened before they reach one month of age. Either otoacoustic emissions (OAE) or auditory brainstem response (AABR) can be used in a single stage or two stage UNHS protocol.
Audiologic assessment of all infants who fail their screening test by three months of age
Intervention for those infants with significant hearing impairment by six months of age designed to meet the individualized needs of the infant and family.

Attributes of an effective screening program include: [22, 27]

- Screening a minimum of 95 percent of infants before hospital discharge.
- Either OAE or ABR can be used for normal term infants but for infants at risk for auditory neuropathy (eg, infants admitted to the NICU), screening ABR should be used.
- A false-positive rate (infants with a positive screening test who do not have hearing loss) ≤3 percent and a rate of referral for audiologic testing after a positive screening result ≤4 percent.
- A false-negative rate (infants with significant hearing loss who are missed by the screening test) of zero.
- A follow-up rate of infants referred for audiologic assessment and for infants who were not screened in their birthing hospital (whose parents did not refuse screening) of at least 95 percent.
- Rescreening for infants who are readmitted within the first month of life for conditions associated with potential hearing loss (eg, hyperbilirubinemia).
- An effective communication system that ensures results of screening tests from the birth hospital are conveyed to the family and the designated primary care provider and referral of infants who fail the screening test for audiologic assessment.

PROTOCOLS OF UNIVERSAL NEONATAL HEARING SCREENING (UNHS)

The two types of universal screening protocols routinely used are single- or two-stage UNHS.

Single stage UNHS : It utilizes a single screening test, either OAE or AABR, which detects 80 to 95 percent of neonates with a hearing impairment. Because of high false positivity, with either single test, this results in a substantial number of referred infants with normal hearing for audiologic assessment, thereby increasing the overall cost of UNHS. Referral for audiologic evaluation is generally required for 4 percent of infants screened with ABR[28, 29] and between 5 to 21 percent of infants screened with OAE.[13] The prevalence of moderate to severe hearing loss is estimated to be one case for every 900 to 2500 newborn infants. Thus, one can estimate that for one case of significant hearing loss, the number of infants with normal hearing that would be referred for an audiologic evaluation can range from 36 to 525 patients.

Two stage UNHS : In this protocol, only patients who fail the initial first test require a second screening study, and only patients who fail both tests are referred for audiologic assessment.[30]
Two studies that utilized a two stage UNHS reported that approximately 900 to 1400 infants would need to be screened to identify one case of bilateral hearing loss.[31, 32]
In a Controlled trial of universal neonatal screening for early identification of permanent childhood hearing impairment by the Wessex Universal Neonatal Hearing Screening Trial Group, between 1993 and 1996, two teams of four part-time testers and equipment moved between two pairs of hospitals to achieve four periods with neonatal screening and four without neonatal screening, each of 4-6 months' duration. Babies did or did not undergo neonatal screening dependent on during which periods they were born. The authors used a transient evoked otoacoustic emissions (OAE) test and, in babies who failed this test, an automated auditory brainstem response (ABR) test on the same day. They only referred babies with positive results for audiological assessment. 53,781 babies were included in the trial, and 25,609 were born during periods with neonatal screening. Neonatal screening achieved 87% coverage of births, with a false-alarm rate of 1.5%, and an overall yield of 90 cases of bilateral PCHI of 40 dB or more relative to hearing threshold level per 100,000 target population, equivalent to 80% of the expected prevalence of the disorder in the population. 71 more babies with moderate or severe PCHI per 100,000 target population were referred before age 6 months during periods with neonatal screening than during periods without. Early confirmation and management of

PCHI were significantly increased. The rate of false-negative results from neonatal screening was significantly lower than that for the distraction test (4 vs 27% p=0.041).[31]

Prieve BA & Stevens F of Syracuse University, New York, USA attempted to determine the feasibility of universal newborn hearing screening, including intervention of identified infants, in the state of New York. Seven regional perinatal centers (eight hospitals) representing the various regions of the state were funded for 3 yr to implement universal newborn hearing screening and follow-up of identified infants. Detailed data analysis was performed for inpatient, outpatient, and intervention outcome measures and for the various protocols. Most of the outcome measures were analyzed in terms of year of program operation, nursery type, and geographic region of the state. In conclusion it was seen that Universal newborn hearing screening was feasible in regional perinatal centers across the state of New York. The average ages of identification of hearing loss, hearing aid fitting, and enrollment in early intervention were less than those reported in published studies where universal newborn hearing screening was not in place.[32]

It is estimated that one of every 45 infants from the well-baby nursery referred for audiologic evaluation by a two stage UNHS would have moderate to profound bilateral permanent hearing loss.[9]

Two-stage compared to single stage UNHS decreases the referral rate for audiologic assessment. In a study by Lin HC et al from Taiwan, a statistically significant decrease of referral rate was achieved in the group using OAE and ABR as screening tools when compared with OAE alone (1.8% vs. 5.8%).[33] Of the 340 newborns referred to a Dutch single center after initially failing the first stage of OAE screening, second (ABR) stage screening found 21 percent had normal hearing.[34]

Two-stage screening may miss infants with hearing loss, because it inaccurately assumes that all infants who fail the initial screen but pass the second have normal hearing.[30, 35] As an example, a study conducted in seven birthing centers demonstrated that two-stage UNHS (OAE and ABR) failed to detect a number of infants with hearing loss.[30] In this study, 86634 infants were screened for hearing loss at these sites. Of those infants who failed the OAE but passed the A-ABR in at least 1 ear, 1524 were enrolled in the study. When the infants were an average of 9.7 months of age, diagnostic audiologic evaluations were done for 64% of the enrolled infants

(1432 ears from 973 infants). Twenty-one infants (30 ears) who had failed the OAE but passed the A-ABR during the newborn hearing screening were identified with permanent bilateral or unilateral hearing loss. Twenty-three (77%) of the ears had mild hearing loss (average of 1 kHz, 2 kHz, and 4 kHz<or =40-decibel hearing level). Nine (43%) infants had bilateral as opposed to unilateral loss, and 18 (86%) infants had sensorineural as opposed to permanent conductive hearing loss. Thus, if all infants were screened for hearing loss using the 2-stage OAE/A-ABR newborn hearing screening protocol, then approximately 23% of those with PHL at approximately 9 months of age would have passed the A-ABR.

Recommended Approach for Institutions : In the well baby nursery setting, it would be appropriate to perform a two-stage UNHS primarily to decrease the number of infants with normal hearing (false positives) who would need referral for further audiologic assessment. The OAE is usually used as the initial test because it takes less time to administer, its cost is lower than AABR, it is not dependent on the infant being asleep (as is the case for AABR) and there is a low incidence of auditory neuropathy in infants admitted to the well nursery.[36] Infants who fail the OAE test are then screened using AABR.
In the case of infants admitted in the NICUs, AABR would be the recommended screen due to the increased risk of auditory neuropathy in this population.

If a program decides to utilize only a one-stage UNHS, then screening with AABR would be appropriate, since it results in a lower false positive rate and lower referral rate for audiologic assessment than OAE. In addition, there is evidence that AABR (but not OAE) can detect infants with auditory neuropathy (AN).[37, 38] This is especially important if the screening population includes infants who are at risk for AN such as premature infants or those with hyperbilirubinemia.

In the case of a community based setting, continued surveillance by the health visitor/primary care provider for hearing problems is recommended and includes assessment of developmental milestones, auditory skills through behavior observation testing, parental concerns, and the status of the middle-ear during routine well child visits. If behavior observation testing fails, then a referral for further assessment can be recommended, that may include the two step objective protocol of an OAE followed by an ABR testing. In addition, increased oversight by the primary

care provider can be suggested for infants who fail the initial screen but pass the second screening test as they are still at risk for hearing impairment.

Owen M et al did a prospective cohort study to test the feasibility of health visitors (HVs) performing neonatal otoacoustic emissions (OAE) hearing screening in the community (local health centres and babies' homes in urban and rural settings in West Gloucestershire) and found that HVs are able to perform OAE testing in the neonatal period at home and in local health centre clinics. They achieved high population coverage rates and low false positive rates. Universal neonatal hearing screening by HVs using OAE testing is feasible, well received, and could be less demanding of HV time than the current distraction testing.[39]

Follow-up

Infants who failed UNHS : The success of a UNHS program is dependent upon ensuring that every infant who fails the initial screening test be assessed by audiologic evaluation by three months of age.[22] However, infants with significant hearing loss can be missed if they cannot be located due to inadequate tracking procedures.[40]

In the previously mentioned CDC report, the failure rate of follow-up of infants who failed UNHS decreased from 64 to 46 percent from 2005 to 2007 in 44 states and territories.[24] In Massachusetts and Colorado the UNHS programs actively follow-up with families and providers, resulting in a failure rate of only about 6 percent.

Poor socioeconomic status contributes to a decreased rate of follow-up for audiologic evaluation after a positive screening test.

In a study of 39,153 newborns screened in Rhode Island, infants with traditional Medicaid insurance were significantly less likely to be rescreened than those with commercial health insurance or managed care Medicaid.[41]

Similar results were reported in a retrospective review with fewer NICU graduates with Medicaid coverage than those with private insurance returning for follow-up (51 versus 78 percent).[42] Infants with Medicaid coverage and hearing loss also obtained hearing aids at a later age (50 versus 28 weeks of age).

In a study from Massachusetts from 2002 to 2003, 11 percent of infants who did not pass the hearing screening test were lost to follow-up.[43] Children were at increased risk for not returning

to follow-up if their mothers were nonwhite, covered by public insurance, or smokers during pregnancy.

Targeting at-risk groups for failure of follow-up may improve return rates for further audiologic evaluation.

Reassessment of at-risk infants : Infants with risk factors for hearing loss and who have passed the neonatal screening, still are at-risk for hearing loss. As a result, the Joint Committee on Infant Hearing recommends audiologic reassessment between 24 and 30 months of age for any infant with the following risk factors:[22]

- Family history of permanent childhood hearing loss
- Congenital infections such as cytomegalovirus, herpes simplex, rubella, syphilis, and toxoplasmosis
- Craniofacial anomalies including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
- Syndromes associated with deafness including Waardenburg syndrome
- Culture-positive postnatal infections associated with sensorineural hearing loss including bacterial and viral meningitis
- Reassessment of hearing may also be needed in NICU survivors who are also at risk for hearing loss.

Cost of newborn screening programs: The cost of UNHS programs (USA) was observed to range from $7 to $30 per infant screened.[27, 40] AABR is associated with the lower costs as compared to OAE alone, achieves the lowest referral rates at hospital discharge, and has the quickest learning curve to achieve those rates.

A higher rate of referral for audiologic assessment will increase the cost. This rate is comparable to that for metabolic screening, although the incidence of hearing impairment is higher. The cost of identifying one affected child is approximately $4000 to $22,000 for hearing loss compared to $10,000 for phenylketonuria and $40,000 for hypothyroidism.[44]

Mohr et al, USA found that severe to profound hearing loss is expected to cost the society $297,00 over the lifetime of an individual. Most of these losses (67%) would be due to reduced

work productivity, although the use of special education resources among children contributes an additional 21%. The life time costs for those with prelingual onset would exceed $ 1 million.[45]

Mezzano P et al, Italy, did a cost analysis of a regional UNHS programme was on 32,502 newborns. 31,992 (98.4%) were no audiological risk (NAR) subjects and 510 were babies with audiological risk (WAR) (1.6%). UNHS was performed on two levels, the first level involved otoacustic emission (OAE) testing on NAR subjects, while the second level involved auditory brainstem response (ABR) testing of WAR and of NAR with 2nd OAE Refer tests. The cost of screening was (Euro) €13.32 per screened NAR infant. The total cost (OAE+ABR) was €16.58, spreading the cost over the whole NAR population. The total cost per screened infant in the WAR population was €415.9. The average cost per detected case in the NAR population was €32,951, while in the WAR population it was €11,303.[46]

The costs and performance of hospital-based and community-based infant hearing screening models were evaluated in Lagos, Nigeria. The protocol consisted of two-stage screening with transient-evoked otoacoustic emissions and automated auditory brainstem response followed by diagnostic evaluation for all infants referred after the second-stage screening. Screening cost per child was lowest (US$7.62) under community-based universal screening and highest (US$73.24) under hospital-based targeted screening. Similarly, cost per child detected with permanent congenital and early-onset hearing loss PCEHL was lowest (US$602.49) for community-based universal screening and highest (US$4631.33) for hospital-based targeted screening. It was seen by the authors that community-based universal screening of infants during routine immunisation clinics appeared to be the most cost-effective model for early detection of PCEHL in low-income countries.[47]

Although Visual reinforcement audiometry (VRA) is the gold standard for hearing assessment for non-verbal children, but it cannot be performed reliably before the infant is eight to nine months of age.[48] Until VRA can be performed, infants who fail a screening test should have a diagnostic ABR performed.

BEHAVIORAL AUDIOMETRY PROTOCOL FOR MEASURING HEARING THRESHOLDS IN INFANTS

Although the widespread use of various neonatal screening methods like OAE or AABR have allowed for assessment screening for hearing loss of very young babies, and the electrophysiological techniques, electrocochleography (ECochG), ABR and ASSR are able to confirm or rule out hearing impairment, yet the measurement of thresholds over the whole hearing range can only be achieved by behavioral audiometry.

Through a sound exploration technique, following are not possible: measuring hearing thresholds by bone conduction; defining the air conduction of each ear individually; delivering stimulation at values greater than 90 dB because of reverberation phenomena; performing audiometric measurements in babies less than 6 months old or in older children, who because of their retardation or other handicap, cannot express perception through an orientation reflex.

The *Delaroche Protocol* makes it possible from the age of 6 months to perform measurements with both a vibrator and headphones. The protocol has also been adapted for babies under 6 months of age, following on from neonatal screening.[49]

SUSTAINABILITY AND QUALITY CONTROL: Despite the recommendation from medical societies and the United States Preventive Services, the implementation of developing an integrated, interdisciplinary system of UNHS that detects and treats early hearing loss is challenging as illustrated by the following studies:

A population-based review of UNHS in Colorado in 2006 and 2007 (covering 204,694 births) revealed 98 percent screening in the newborn period. However, of the newborns with abnormal screening results, only 82 percent received targeted follow-up. Newborns with normal Apgar scores were ten times more likely than infants with low Apgar scores to receive initial hearing screening; newborns with normal birth weights were four times more likely than newborns with low birth weights to receive initial hearing screening. Those infants lost to follow-up constituted a high risk group for hearing loss because they were more likely to have had low APGAR scores and/or low birthweight.[50]

All states and U.S. territories also have established Early Hearing Detection and Intervention (EHDI) programs, which embody evidence-based public health policy for addressing infant hearing loss. EHDI programs help ensure that newborns and infants are screened and receive recommended follow-up through data collection and outreach to hospitals, providers, and

families. To determine the status of efforts to identify newborns and infants with hearing loss, CDC analyzed EHDI surveillance data from 1999-2007. Differences in how data were reported and collected limit comparability between 1999-2004 and 2005-2007 data; however, available data indicated an increase in infants screened from 46.5% in 1999 to 97.0% in 2007, although only 85.4 percent were tested by 1 month of age.[24] Forty-six percent of infants with an abnormal initial screen were never documented as having had a follow-up diagnostic evaluation. Enrollment in early intervention by six months of age was documented in 60.8 percent of infants with abnormal hearing.

Quality Standards for the NHSP [National Hearing Screening Protocol, NHS (UK)][51]

The purpose of the quality standards is to outline the minimum family friendly standards expected to ensure that:

- Families are able to make informed choices about screening uptake
- Screening and diagnostic services are effective and carried out to a high standard
- Results are communicated to parents/guardians effectively
- Families are given comprehensive support post-diagnosis
- Responsibilities for recording and reporting performance are clear

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Chapter 8: Modalities for management of Hearing Impaired children including educational rehabilitation.

Young children who suffer from hearing loss may often appear to be normal as the impairment or handicap may not be obvious. Observational evidence has clearly shown the effect of undetected and untreated hearing loss, with an impact on the speech, language and consequentially cognitive delays. Early identification and effective treatment of hearing loss is thus the key approach in effective management of such cases to improve these outcomes.

The underlying etiology is important to be considered and the options for treatment depend on the cause(s) accordingly. At some places surgical intervention is warranted. Cochlear implantations may be required for some children with bilateral and profound hearing loss where other methods have failed. Amplification devices are appropriate where there is a bilateral sensorineural hearing loss or a long term conductive hearing loss. It is most appropriate the management of all children with a hearing loss should be a multidisciplinary approach with expertise from otolaryngologists, speech pathologists, audiologists, geneticists, and educational specialists. Referral to a pediatric ophthalmologist should also be considered, since these children obviously rely on sight for their communication and learning.[1]

The basic goal of early initiation of appropriate educational programs is the improvement of communication skills of the hearing-impaired children. Educational options vary according to the degree of hearing loss and cognitive ability of the child. The importance of multidisciplinary team lies here, in providing a collaborative effort by professionals to perhaps establish an individual treatment plan for each child.

HEARING AIDS AND ASSISTIVE DEVICES

Hearing amplification is usually the first step undertaken in the management of hearing impairment, once the underlying etiology is addressed. Yoshinaga-Itano C et al[2] compared the receptive and expressive language abilities of 72 deaf or hard-of-hearing children whose hearing losses were identified by 6 months of age with 78 children whose hearing losses were identified after the age of 6 months. All of the children received early intervention services within an

average of 2 months after identification. The participants' receptive and expressive language abilities were measured using the Minnesota Child Development Inventory. Their results showed that children whose hearing losses were identified by 6 months of age demonstrated significantly better language scores than children identified after 6 months of age. For children with normal cognitive abilities, this language advantage was found across all test ages, communication modes, degrees of hearing loss, and socioeconomic strata. It also was independent of gender, minority status, and the presence or absence of additional disabilities.

It is essential for the family members of the child to agree that the child would definitely benefit from a hearing aid, which may not necessarily restore the hearing to normal, but an improved hearing is definitely expected, which if done timely, is expected to improve the language outcome as well.

Hearing aids: There is a wide range of types, and costs in which hearing aids are available. The audiologic assessment and evaluation guides to the best hearing aid selection for the child. The final choice depends upon the age, the level and type of hearing loss and family preference and or affordability. Hearing aid fittings for children under the age of 1 year provide many challenges to the audiologist, since in young children it would be difficult to know whether a hearing aid is correctly adjusted and not too loud. A tiny microphone can be used to measure the sound intensity in the ear canal for this problem. There are computer programs available to help the audiologist determine whether a particular device is appropriate for a given child. These use real ear measurements or simulated real ear measurements. The desired sensation level (DSL) approach estimates a frequency gain target that amplifies speech to audible levels across a broad frequency range and is beneficial in fitting hearing aids for very young children.[3, 4] The different styles of hearing aids available include bone conduction, behind-the-ear, in-the-ear, and completely-in-the-canal instruments. Bone conduction hearing aids are used for children who have atretic ears or chronic otorrhea.

Commonly used hearing aids are behind the ear type as they are fit with the ear. The ear mold that is coupled to the ear is easily remade as the child grows. Although, the other types, i.e. In-the-ear and in-the-canal instruments are cosmetically better for the child, these devices are appropriate only for hearing loss less than 60 dB.

The electronic circuitry of a hearing aid can be analog, digital, or digitally programmable. The advantages of digital and programmable hearing aids over conventional analog hearing aids include better sound quality, increased precision, improved speech recognition,[5, 6] and flexibility

of settings; the disadvantage is higher cost. Most digital hearing aids are not appropriate for children with profound hearing loss; however, several programmable aids provide adequate gain for children with severe to profound losses.

In a study by Bamford J et al to investigate the efficacy of a 2-channel hearing aid with low-frequency compression and high-frequency linear amplification on a group of school-age hearing aid wearers, it was seen that 2-channel hearing aids appear to be an acceptable management option for audiometrically suitable children. The two-channel hearing aids showed significantly higher mean scores for speech perception in noise and significantly higher composite questionnaire scores (reflecting aspects of satisfaction and benefit). Final choice of hearing aids at the end of the study by parents and children also favored the 2-channel device.[5]

Kuk FK et al examined the efficacy of a digital hearing aid with a directional microphone in a school-aged population. Twenty children (9 with a mild-to-moderately-severe hearing loss and 11 with a moderate-to-severe hearing loss) between 7 1/2 and 13 2/3 years of age wore the study hearing aids binaurally for 30 days prior to the evaluation. Subjective rating of hearing aid efficacy in the classroom was examined using the Listening Inventory For Education (LIFE) questionnaire. Parental impression on hearing aid efficacy was also collected at the end of the study. The results showed improved speech recognition in noise with the digital directional hearing aid at all presentation levels. Preference for the digital directional hearing aids over the subjects' own omnidirectional analog hearing aids was also seen on the LIFE questionnaire and parental impression. The degree of hearing loss did not seem to have affected the benefits offered by the digital directional hearing aids.[6]

Assistive listening devices: These are designed to improve hearing perception in the patients, especially where there is a noisy environment. They consist of a microphone for the speaker, a frequency modulated (FM) transmitter, and a receiver worn by the listener. These are available as stand-alone units, or the FM receiver can be attached to a hearing aid.[7] These devices work on the concept of providing a gain (i.e. amplification) by eliminating background noise. This improves the signal-to-noise ratio, and thus a better perception of a more clear sound.

The whole exercise of wearing of fitted hearing aids and assistive devices, can be a stressful time for parents and a reason for apprehensions for their children's condition. They should be regularly and professionally provided with counseling and support. More educated parents can in

addition be provided with FAQs or patient information as written information on hearing loss, hearing aids/assistive devices, etc. and troubleshooting tips. Once fitted, a regular follow-up with the audiologist is necessary to establish the aided benefit and its sustainability, and to also check the fitting of the hearing aids and the growing ear mold of the child.

HEARING AIDS: BONE CONDUCTION TYPE

A bone conduction type of hearing device is different from a conventional hearing aid (air conduction type). It transmits the sounds directly through the skull, and have to be held against the skull, which can be more cumbersome and uncomfortable to use. They does not achieve a hearing quality comparable to conventional hearing aids, but it seen that some patients who do not benefit from the standard air conduction type of hearing devices, may benefit from this type of a device that has a different mechanism of functioning.

In contrast, an implantable bone conduction hearing aid has significant advantages.[8,9,10,11,12] The main implantable system available is a bone-anchored implantable hearing aid system, known as BAHA. A small titanium screw is inserted and osseointegrates with the bone of the skull over several months. An abutment is attached to the screw such that a small portion of the abutment sticks out through the skin and forms an attachment point for a removable bone-conduction hearing aid. The sound quality is far superior to that of traditional bone conduction hearing aids.

Lloyd S et al[8] did a retrospective review of children undergoing implantation with bone-anchored hearing aids (BAHAs) at the Great Ormond Street Hospital for Children, London, UK. Case notes of 71 children undergoing BAHA placement between December 1990 and August 2002 were reviewed. Outcome measures included hearing thresholds, incidence of fixture loss, skin reaction and need for revision. Quality of life outcomes were also measured. Among the 71 children (eighty-five ears) implanted, fifty-four per cent of children had experienced no complications, 42 per cent had required revision surgery and 26 per cent had experienced fixture loss at some point. Young age at implantation was associated with an adverse outcome. Trauma and failure of osseointegration had been the commonest reasons for failure. A skin reaction around the abutment had occurred at some point in 37 per cent of children but had persisted for longer than six months in only 9 per cent; this had been associated with fixture loss. The use of fixture site split skin grafts had reduced problems with skin hypertrophy and hair overgrowth. Hearing thresholds when using BAHAs had been comparable to those when using bone

conduction hearing aids. However, BAHAs had significant additional benefits in terms of sound quality, ease of use and overall quality of life.

Hol MK et al[10] from the Radboud University Nijmegen Medical Center, Nijmegen, Netherlands, performed an A prospective clinical follow-up study in a tertiary referral center. Evaluation of the audiological and subjective benefits of the bone-anchored hearing aid (BAHA) as a device for transcranial routing of sound (BAHA CROS) in 56 patients with unilateral inner ear deafness was done. Previously reported results of 29 patients were supplemented with a second series of 30 patients with unilateral inner ear deafness. Audiometric measurements were taken before and after BAHA CROS fitting. Subjective benefits were quantified with 4 different patient questionnaires. The sound localization results in a well-structured test setting were not differentiable from chance. The 5 patients with congenital hearing loss showed better scores in the unaided sound localization measurements. Overall, most patients reported some subjective improvement in their capacity to localize sounds with the BahaCROS in daily life. The main effect of the Baha CROS was to alleviate the head shadow effect during the speech-in-noise test.

Saliba I et al[11] from the Sainte-Justine University Hospital Center, Montreal University, Canada performed a prospective longitudinal study. Seventeen patients between the ages of 5 and 18 years old were included. All patients underwent a complete tonal and vocal evaluation at four pre-determined intervals between the pre-operative period and one-year of bone-anchored hearing aid (BAHA) use. Basic pure-tone average and speech reception threshold were measured in different sound environments. Speech discrimination improvement was tested with the voice originating from the side of the BAHA-fitted ear and with the voice originating from a source directly in front of the patient. These measures were repeated with confounding noise facing the patient then from the side of the affected ear. All tonal and vocal evaluations were performed pre-operatively, the day of processor insertion, 6 months and 12 months after processor insertion. The authors found that hearing gain with BAHA was clinically and statistically significant at all intervals. Conventional tonal evaluation revealed significantly improved hearing gain after BAHA insertion compared with pre-operative testing with BAHA (26.3 dB vs. 17.3 dB), and this improvement was maintained at one year (27.9 dB). Speech discrimination gain at one year was better than immediately post-insertion (21.9% vs. 11.7%). Maximal gain with BAHA was found with the voice originating from the side of the affected ear and with confounding noise facing the patient (27.1% at one year), whereas the least gain was found in a silent room with the voice

coming from straight ahead (11.9% at one year). Thus, the authors concluded that pure-tone average gain at one year post-insertion was similar to immediate post-insertion gain. BAHA aids speech discrimination most when the voice originates from the side of the affected ear with confounding noise facing the patient. Speech discrimination gain improves with time, suggesting an underlying learning process. The best BAHA gain in speech discrimination occurred with background noise.

Potential indications for an implantable system include:

- Congenital atresia of the ear canal[13, 14, 15]
- Chronic infection of the middle or outer ear which can be exacerbated by using a conventional hearing aid
- Allergic reactions to standard hearing aids
- Profound unilateral sensorineural hearing loss (USNHL) as may occur after removal of a vestibular schwannoma (acoustic neuroma), from trauma, or from a viral or vascular insult[16]

Mazita A et al[13] from Universiti Kebangsaan, Malaysia did a retrospective analysis of 16 patients who had BAHA implantation at Universiti Kebangsaan Malaysia Medical Centre. 11 patients had implantation of BAHA performed in two stages, while the other five patients had it as a single-staged procedure. The complications that occurred were failure of osseointegration (one patient), granulation tissue overgrowth into the abutment (two patients) and cellulitis surrounding the abutment (three patients). The average preoperative unaided air conduction threshold was 64.9 dB and the average postoperative aided hearing threshold was 29.7 dB. The overall mean functional gain was 35.2 dB. The authors concluded that BAHA has many advantages over the conventional hearing aid in the form of cosmesis, discomfort and hearing gain. It is a reliable hearing rehabilitation tool with good predictable hearing outcome in patients with bilateral canal atresia, especially those unsuitable for canalplasty. Despite its higher cost and the need for surgical implantation, its use is justifiable in properly selected patients

Ricci G et al[14] from the University of Perugia, Italy examined 31 patients with bilateral congenital aural atresia in whom a Baha system had been implanted found that Baha system to treat patients with bilateral aural atresia were extremely satisfactory compared both with those of

surgical reconstruction of the auditory canal and those of traditional bone-conduction hearing aids. Furthermore, great improvement was noted in quality of life of the patients.

The BAHA fitting can be considered effective and beneficial in children with bilateral or unilateral hearing loss.[17] Adequate bone thickness is a prerequisite for successful, long-lasting osseointegration of titanium fixtures for bone-anchored hearing aids . Children are typically about six years of age before BAHA is feasible, because 3 to 4 mm of bone is needed to ensure osseointegration.[18]

COCHLEAR IMPLANTS:

Cochlear implants are surgically implanted prosthetic devices that electrically stimulate the cochlear nerve to provide hearing. The device consists of a battery-powered external processor (that looks like a hearing aid), a receiver coil implanted below the scalp, and an electrode inserted directly into the cochlea through a surgical opening.

Children younger than 2 years of age were initially excluded from cochlear implant candidacy for a variety of reasons. Reasons ranged from concerns about the reliability of the diagnosis of a profound hearing loss in very young children, to concerns about surgical safety and long-term durability of the device in a growing child. Results from several recent studies have shown that children younger than 2 years of age can safely and successfully be implanted.[19]

A nonfederal, nonadvocate, 14-member consensus panel representing the fields of otolaryngology, audiology, speech-language pathology, pediatrics, psychology, and education, and including a public representative, in addition, 24 experts in auditory anatomy and physiology, otolaryngology, audiology, aural rehabilitation, education, speech-language pathology, and bioengineering presented data to the consensus panel in NIH consensus conference on Cochlear implants in adults and children.[20] They concluded that cochlear implantation improves communication ability in most adults with severe-to-profound deafness and frequently leads to positive psychological and social benefits as well. Currently, children at least 2 years old and adults with profound deafness are candidates for implantation. Cochlear implant candidacy should be extended to adults with severe hearing impairment and open-set sentence discrimination that is less than or equal to 30% in the best-aided condition. Access to optimal education and rehabilitation services is important for adults and is critical for children to maximize the benefits available from cochlear implantation.

The criteria for selecting cochlear implantation include profound bilateral sensorineural hearing loss and little or no benefit from hearing aid use after six months.[19, 20, 21]

Worldwide trends in Cochlear Implantation:

Peters BR et al[21], from Dallas, Texas, USA did an electronic survey consisting of 59 mainly multiple-choice questions for online completion. It examined the implant experience and clinical opinion of expert cochlear implant (CI) centers worldwide on the indications, motivations, and contraindications for adult and pediatric, simultaneous and sequential BCI candidacy. Centers were chosen to complete the survey based on their known reputation as a center of excellence. Patient demographics were queried for two time periods to elucidate trends: 2006 and prior, and for the year 2007. Seventy-one percent (25/35) of the CI clinics approached completed the survey. Collectively, these 25 clinics represent experience with approximately 23,200 CI users globally, representing 15% of the total estimated CI population worldwide. It was seen as a result of this survey that the total number of BCI surgeries reflected in their experience (2,880) represents 36% of the estimated number worldwide as of December 2007. Cumulatively to the end of 2007, 70% of all BCI surgeries have occurred in children, with the 3- to 10-year-old age group having the highest representation (33% of all BCIs), followed in order by adults (30%), children under 3 years of age (26%), and children between 11 and 18 years of age (11%). Seventy-two percent of all BCI surgeries were performed sequentially (70% of children, 76% of adults). Children<3 years of age represent the only age group of all patients in which simultaneous surgeries predominate (58% simultaneous). For all other age groups, sequential surgeries far outnumber simultaneous (3-10 years, 84% sequential; 11-18 years, 94% sequential; adults, 76% sequential). Prior to January 2007, 68% of BCIs were performed in children. This increased to 79% for the year 2007 (P<.001). With regard to children only, a change is apparent over time in terms of the age group making up the majority of pediatric BCI surgeries performed. Prior to 2007, children 3 to 10 years of age made up 50% of the children undergoing BCI, whereas those<3 years made up only 33%. In 2007 this shifted more toward the younger age group (47% for those<3 years and 40% for 3-10-year-olds; P<.001). United States clinics had a higher proportion of adult BCI patients (59% children, 41% adults) than the non-United States clinics (78% children, 22% adults; P<.001). The majority of responders do not hold to a minimum or maximum age by which they limit BCI.

Cochlear implant devices in USA are approved by the US Food and Drug Administration (FDA) for use in children as young as 12 months. Vlastarakos PV et al[22] carried out a meta analysis by reviewing the current knowledge on cochlear implantation in infancy, regarding auditory perception/speech production outcomes. The authors suggests that the role of cochlear implantation in children less than 12 months of age remains unclear. Evidence of these children's outperformance regarding auditory perception/speech production outcomes is limited. Wide-range comparisons between infant implantees and under 2-year-old implanted children are lacking. The literature is lacking in prospective studies or long-term follow-up on this issue.[22] We have to balance the advantage of an early auditory stimulation during the critical period of hearing development against the risks of the implantation procedure. Early bilateral simultaneous implantation increases the likelihood of binaural hearing and ensures implantation of the better ear in this population of children whose course is often complicated by formation of scar tissue and ossification within the cochlea. As an example, pneumococcal meningitis may cause labyrinthitis ossificans, and may necessitate consideration for earlier cochlear implantation.[23]
Currently, bilateral CI is widely accepted as a safe and effective means of bilateral auditory stimulation.[24]
Papsin BC and Gordon KA[25] from the University of Toronto, Canada based on their review summarized that both the interval between onset of deafness and cochlear implantation and the interval between implantation of the first and second ears should be narrow in children. They recommended that simultaneous bilateral implantation be provided when possible and, if not, the inter-stage interval should be limited. There is a need Continued exploration of outcomes in children with longer inter-stage intervals with a view to defining a point at which bilateral cochlear implantation provides so little benefit that it is not cost-effective
Bilateral cochlear implants have been advocated as they allow enhanced speech perception under noisy conditions and improved sound localization to hear sound coming from either side without having to turn the head.[24, 25, 26]

Zeitler DM et al[27] from the New York University School of Medicine, USA did a retrospective analysis to examine speech perception outcomes and determine the impact of length of deafness and time between implants on performance in the sequentially bilateral implanted population at a tertiary academic referral center. Forty-three children (age <18 yr) and 22 adults underwent sequential bilateral implantation with at least 6 months between surgeries. Speech perception tests were performed preoperatively before the second implantation and at 3 months

postoperatively. Results revealed significant improvement in the second implanted ear and in the bilateral condition, despite time between implantations or length of deafness; however, age of first-side implantation was a contributing factor to second ear outcome in the pediatric population. Thus they concluded that sequential bilateral implantation leads to significantly better speech understanding. On average, patients improved, despite length of deafness, time between implants, or age at implantation.

The disadvantage of bilateral implantation would include increased surgical and anesthetic risk, risk to residual hearing, and saving one ear for future technologies.

Outcome — Extensive auditory, speech, educational, and psychologic testing has been performed before and after implantation. Results show that the cochlear implant provides auditory detection over much of the speech signal. Compared with the preimplant period, there is significant improvement in auditory discrimination and speech production skills.[28]

Tomblin JB et al[29] from the University of Iowa, USA did a comparison of language achievement in children with cochlear implants and children using hearing aids. English language achievement of 29 pre-lingually deaf children with 3 or more years of cochlear implant (CI) experience was compared to the achievement levels of pre-lingually deaf children who did not have such CI experience. Language achievement was measured by the Rhode Island Test of Language Structure (RITLS), a measure of signed and spoken sentence comprehension, and the Index of Productive Syntax (IPSyn), a measure of expressive (signed and spoken) English grammar. When the CI users were compared with their deaf age mates who contributed to the norms of the RITLS, it was found that CI users achieved significantly better scores. Likewise, we found that CI users performed better than 29 deaf children who used hearing aids (HAs) with respect to English grammar achievement as indexed by the IPSyn. Additionally, the authors found that chronological age highly correlated with IPSyn levels only among the non-CI users, whereas length of CI experience was significantly correlated with IPSyn scores for CI users. Clear differences between those with and without CI experience were found by 2 years of post-implant experience. Thus children who receive CIs benefit in the form of improved language comprehension and production.

Manrique M et al[30] from the University of Navarra Hospital and Medical School, Spain did a prospective cohort single-subject, repeated-measures study compare the auditory abilities and speech performance of children with a profound pre-lingual bilateral hearing-impairment when subjected to a cochlear implant (CI) before or after 2 years of age. The study analyzed 130 children subjected to multichannel CI for profound pre-lingual bilateral hearing-impairment in two age groups: 0 to 2 (n = 36) and 2 to 6 years of age (n = 94). The children were evaluated before, and each year after, the intervention (for up to 5 years) with both closed-set and open-set auditory and speech perception tests. Their speech ability was evaluated according to the Peabody Picture Vocabulary and Reynell general oral expression scales. The results showed that auditory and speech perception tests improved significantly in all children after CI, regardless of the follow-up time. The infant's performance was better the earlier the implant was performed. Speech tests showed that the development of children treated before 2 years of age was similar to normal children, and no additional complications were observed when compared with CI in older children. Thus they concluded that when performed before 2 years of age, CI offers a quicker and better improvement of performance without augmenting the complications associated with such an intervention.

Niparko JK et al[31] from the Johns Hopkins University School of Medicine, Baltimore, USA did a prospective, longitudinal, and multidimensional assessment of spoken language development over a 3-year period in children who underwent cochlear implantation before 5 years of age (n = 188) from 6 US centers and hearing children of similar ages (n = 97) from 2 preschools recruited between November 2002 and December 2004. Follow-up was completed between November 2005 and May 2008. The main outcome measure was performance on measures of spoken language comprehension and expression (Reynell Developmental Language Scales). In their study it was seen that children undergoing cochlear implantation showed greater improvement in spoken language performance (10.4; 95% confidence interval [CI], 9.6-11.2 points per year in comprehension; 8.4; 95% CI, 7.8-9.0 in expression) than would be predicted by their preimplantation baseline scores (5.4; 95% CI, 4.1-6.7, comprehension; 5.8; 95% CI, 4.6-7.0, expression), although mean scores were not restored to age-appropriate levels after 3 years. Younger age at cochlear implantation was associated with significantly steeper rate increases in comprehension (1.1; 95% CI, 0.5-1.7 points per year younger) and expression (1.0; 95% CI, 0.6-1.5 points per year younger). Similarly, each 1-year shorter history of hearing deficit was associated with steeper rate increases in comprehension (0.8; 95% CI, 0.2-1.2 points per year

shorter) and expression (0.6; 95% CI, 0.2-1.0 points per year shorter). In multivariable analyses, greater residual hearing prior to cochlear implantation, higher ratings of parent-child interactions, and higher socioeconomic status were associated with greater rates of improvement in comprehension and expression. Thus, the use of cochlear implants in young children was associated with better spoken language learning than would be predicted from their pre-implantation scores.

Mitchell TE et al[32] from the Children's Cochlear Implant Centre (New South Wales), Sydney, Australia did a comparison of children deafened by meningitis, and those congenitally deaf. Their speech perception and speech production performance following cochlear implantation were analyzed. Three groups consisting of 70 congenitally deaf children, 22 children deafened by meningitis before two years of age and 14 children deafened by meningitis after two years of age were compared. The group deafened by meningitis after two years of age demonstrated significantly better speech perception than the other two groups. Their speech production appeared better but did not achieve statistical significance compared with the other two groups. There was no significant difference in either speech perception or speech production between the congenitally deaf group and the group deafened by meningitis before two years of age.

Thus, children whose deafness occurs after age two years, who are deaf for short periods of time, and/or who are implanted at an earlier age have the best language outcome.[28, 30, 31]

Young age at intervention and oral communication (rather than total (eg, including signing))mode are the most important known determinants of later speech perception in young children after cochlear implantation. Early identification of candidate children necessitates implementation of universal neonatal screening programmes for hearing impairment. This was evidential from a study by O'Donoghue GM et al[33] of the University Hospital, Nottingham, UK. They examined 40 children, with mean age at implantation of 52 months who were either born deaf or became deaf before 3 years. All patients received the same multichannel implant system and were followed up for 5 years. The authors used connected discourse tracking (CDT) as the measure of speech perception. The effect of five potential predictors (age at implantation, number of inserted electrodes, origin of deafness, mode of communication, and socioeconomic group) on speech perception was analyzed. The authors found that the mean number of words per minute perceived increased from 0 before implantation to 44.8 (SD 24.3) 5 years after implantation (27, 35, and 45 mean words per minute at three, four, and five years of follow-up, respectively). Repeated-measures ANOVA showed that children significantly progressed over

time (p=0.001). Age at implantation was a significant covariate (p=0.01) and mode of communication was a significant between-individuals factor (p=0.04).

SURGICAL INTERVENTION: Some conditions associated with hearing loss are amenable to surgical intervention:

- Pressure equalization tubes for the tympanic membrane may improve hearing for children with middle ear effusion, eustachian tube dysfunction, or recurrent acute otitis media.
- Tumors and cholesteatomas require surgical excision or mastoidectomy.
- Children with fluctuating or progressive sensorineural hearing loss may need surgical exploration for and repair of a perilymph fistula.
- Some conditions that cause conductive hearing loss can be treated with either surgery or amplification
- Otosclerosis or other ossicular chain abnormalities
- Stenosis of the external auditory canal
- Some cases of atresia of the external auditory canal[34]

Educational Rehabilitation Social Rehabilitation and Employment Opportunities

Venail F et al[35] from the Centre Hospitalier Universitaire Gui de Chauliac, Montpellier, France in their Prospective study examined Educational and employment achievements in a series of 100 prelingually deaf children who received cochlear implants before six years of age and were followed for at least four years after implantation. Mainstream schooling, regardless of educational level, was the standard experience for children without additional disabilities (16 of 24 [67%]in the 12- to 15-year age group to 20 of 24 [83%]in the 8- to 11-year age group). Four of 8 participants older than 18 years (50%) had a university-level education; the remainder had vocational training. Delayed reading and writing skills were experienced by 19 of 74 participants (26%) and, depending on the age group, 42% to 61% of participants (10 of 24 in the 8- to 11-year age group to 11 of 18 in the 16- to 18-year age group) had failed a grade. The number of grade failures was associated with communication mode at the time of the survey. Age at implantation, preoperative communication mode, and educational support influenced the final

communication mode. In children with additional disabilities, the level of academic achievement and employment status varied.

This study concluded that despite significant differences in the grade failure rate between the children with cochlear implants and the general population, the participants ultimately achieved educational and employment levels similar to those of their normal-hearing peers. To minimize these delays and improve academic success in mainstream education, early oral education and early cochlear implantation are important.

The main goal of a cochlear implant program is to improve communication possibilities and the quality of life of hearing-impaired individuals by providing them optimal hearing performance. Cochlear device systems can help the profoundly deaf and hard of hearing avoid social isolation and transform them into socially active individuals.

Cochlear implant equipment has improved considerably over the last few years. The devices have become more esthetic, convenient for daily use, highly reliable, technologically advanced and, as a result, more widely used.[36]

Migirov L et al[36] studied how well-or poorly, individuals who underwent cochlear implantation as children integrated into the general Israeli hearing community. They surveyed the 30 subjects > or = 18 years old who underwent cochlear implants in their institution from 1990 to 2004**,** when they were < 18 years of age and had used their device for at least 3 years before replying. Eighteen implant users responded (14 males), yielding a 60% response rate. Their mean age was 13.3 +/- 7.0 years (range 6-17) at implantation and 21.1 +/- 3.6 years (range 18-34) when they filled in the questionnaire. Five were attending rabbinical school (yeshiva students), four were in regular military service, five were university students (three also held jobs), two were attending high school, one was employed (and had a university degree), and one had left the yeshiva and was unemployed when he returned the questionnaire. Fourteen respondents use the oral communication mode for conversation and the other 4 use both oral and sign languages. Longer daily implant use was significantly associated with coping with the difficulties in the setting in which they were currently active, with a higher level of satisfaction with their current lifestyle and with recognition of the implant's contribution to this satisfaction (P = 0.037, P = 0.019 and P = 0.001, respectively). The authors concluded that advances in cochlear implant technologies enable profoundly deaf implanted children to integrate well into the Israeli hearing society, albeit with a large intersubject variability.

Fazel and Gray[37] found that a cochlear implant is associated with an improved chance of being employed among adult recipients, that it helps them with career prospects, and improves their job satisfaction. Other authors demonstrated the benefits of a cochlear implant in speech perception and production and in language and literary skills, all resulting in positive changes in quality of life and opportunities of employment.[38, 39, 40]

There are few reports, however, on the long-term social and occupational outcome of patients who received cochlear devices in their childhood.[41, 42, 43]

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