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ASSESSMENT OF NEONATAL HEARING SCREENING

Dr sonal kala[1*], Dr Ravindra Singh Bisht[2], Dr Ashish gangwar[3]

[1*]Senior resident, department of ENT, VCSG Government Institute of Medical Sciences and Research, HNB Teaching and Base Hospital, Srinagar, Uttarakhand
[2]Professor, Department of ENT, VCSG Government Institute of Medical Sciences and Research, HNB Teaching and Base Hospital, Srinagar, Uttarakhand
[3]Junior resident, Department of ENT, VCSG Government Institute of Medical Sciences and Research, HNB Teaching and Base Hospital, Srinagar, Uttarakhand

***Corresponding author**: Dr sonal kala
Senior resident , department of ENT, VCSG Government Institute of Medical Sciences and Research, HNB Teaching and Base Hospital, Srinagar, Uttarakhand

Abstract :
The present statement reviews the evidence for universal newborn hearing screening (UNHS). A systematic review of the literature was conducted using Medline and using search dates from 2022 to 2024. The following search terms were used: neonatal screening AND hearing loss AND hearing disorders. The key phrase “universal newborn hearing screening” was also searched. The Cochrane Central Register of Controlled Trials and systematic reviews was searched. Three systematic reviews, one controlled non-randomized trial and multiple cohort studies were found. It was determined that there was satisfactory evidence to support UNHS. The results of the available literature are consistent and indicate clear evidence that without UNHS, delayed diagnosis leads to significant harm for children and their families; with UNHS, diagnosis and intervention occur earlier; earlier intervention translates to improved language outcomes; and in well-run programs, there is negligible harm from screening.

Keywords: Deafness, Early intervention, Hearing screening, Newborns

Introduction :

Permanent hearing loss is one of the most common congenital disorders, with an estimated incidence of one to three per thousand live births (1,2) – far exceeding the combined incidence of conditions for which newborns are routinely screened such as congenital hypothyroidism, phenylketonuria and other inborn errors of metabolism (3). In the past decade, universal newborn hearing screening (UNHS) has been widely adopted throughout North America, Europe and in most other developed regions, primarily as a result of technological advances in screening and intervention modalities. Based on available evidence, the American Academy of Pediatrics endorsed UNHS in 1994 (4) and 1999 (5), as has the US Preventive Services Task Force (USPSTF [2]).

Nonetheless, in 2011, many Canadian infants are still not offered UNHS. Ontario and British Columbia have fully funded provincial programs. Other provinces have partial programs, primarily targeting infants in the neonatal intensive care unit. Quebec confirmed funding for UNHS in July 2009, but has not yet implemented UNHS. This secondary prevention strategy is based on evidence

that early diagnosis and intervention allow for improved outcomes in hearing-impaired children. Most UNHS programs aim for screening by one month of age, confirmation of the diagnosis by three months, with intervention by six months.

Hearing loss is defined based on the degree of loss, measured in logarithmic decibels, at frequencies between 125 Hz (low-pitch sounds) and 8000 Hz (high-pitch sounds) . Hearing loss is further categorized based on etiology (sensorineural, conductive or mixed), and may be fixed or progressive. Most neonatal hearing loss is sensorineural; a known genetic cause is found in 50% of children. Of these children, approximately 70% have nonsyndromic deafness, most often related to cochlear hair cell dysfunction because of errors in production of the gap junction protein connexin 26. The remaining causes of neonatal sensorineural hearing loss include congenital infections, hyperbilirubinemia and ototoxic medications. Admission to a neonatal intensive care unit is an established risk factor for hearing loss in infants, particularly for auditory neuropathy (6). Overall, known risk factors are present in only 50% of infants born with hearing loss (7,8). Because a substantial proportion of infants have no risk factors, universal screening has replaced selective screening in most developed nations.

Without UNHS, infants with hearing loss are typically identified with an established language delay. For both caregivers and physicians, the symptoms and signs of hearing loss are subtle because infants with hearing loss often demonstrate a high degree of environmental vigilance. Thus, a deaf infant may appear to turn his or her head to the sound of a ringing bell, but may really be responding to a perception of the bell's movement via visual or tactile sensory input. Vocalizations, such as babbling, may also appear to develop normally. Historically, significant expressive language deficit, noted well beyond one year of age, has been the primary diagnostic feature in young children with hearing loss. Thus, in unscreened children, as is the current situation in many parts of Canada (9), the average age at diagnosis is approximately 24 months. Mild and moderate hearing losses are often undetected until school age (10). In sharp contrast, the median age of diagnosis in screened populations is three months of age or younger, with intervention by six months of age (2,10).

Currently, hearing screening in newborns is performed via otoacoustic emission (OAE) and automated auditory brainstem response (AABR) testing. These physiological, noninvasive, automated screening tests can be performed at the bedside in term and pre-term infants. Depending on the screening protocol, they may be performed singly (OEA or AABR) or sequentially. Both the OAE and AABR tests are automated screening adaptations of more detailed diagnostic tests for hearing loss.

OAEs are forms of energy, measured as sound, generated by the outer hair cells of the human cochlea, in response to received auditory input. First described by a geophysicist in the mid 1940s, the screening test was developed in 1978 (20) by David Kemp. Based on the natural phenomenon of 'sound echoes', a sound stimulus is sent to the newborn's auditory system via ear-specific probes placed in the external ear canal. The probe simultaneously records emissions returning from the outer hair cells of the cochlea via the middle ear. OAEs can be recorded in 99% of normally hearing ears. The response is generally absent in ears with a hearing loss of 30 dB or greater (21).

The AABR test records brainstem electrical activity in response to sounds presented to the infant via earphones. In contrast to the OAE test, the AABR evaluates the auditory pathway from the external ear to the level of the brainstem, enabling diagnosis of auditory neuropathy, which is a less common cause of hearing impairment (22).

A two-step screening procedure has been implemented in most UNHS programs as a cost-effective and accurate approach. This includes the faster and less expensive OAE as the first test in newborns with no risk factors, followed by AABR in newborns who do not pass the OAE. The AABR is also recommended in infants with any of the risk factors summarized in, particulary in infants requiring neonatal intensive unit care because this population is at an increased risk of auditory neuropathy. There is strong evidence indicating that two-step screening is highly effective in identifying infants with hearing loss (23).

EFFECTIVENESS OF EARLIER INTERVENTION :

Studies of the effectiveness of screening have examined the differences in outcomes of children who received newborn screening (earlier intervention) versus normally hearing children and versus unscreened hearing-impaired children (later intervention; usually after 12 to 24 months of age). Using standardized methods, the USPSTF examined this issue in 2001 (8) and again in 2008 (2). The impact of long-term language outcomes was ranked as uncertain in the 2001 review, and a call for further study was made. A Cochrane review (26), originally published in 2005 and now withdrawn due to lack of revision, cited a similar conclusion. In the most recent USPSTF review (2), the authors concluded that there was adequate evidence demonstrating that children with an earlier diagnosis had improved expressive and receptive language scores. Updated evidence from multiple studies (2,27–29) now indicate that infants who are diagnosed and receive intervention before six months of age score 20 to 40 percentile points higher on school-related measures (language, social adjustment and behaviour) compared with hearing-impaired children who receive intervention later on (7).

Children with hearing loss are best managed by a coordinated team including family physicians, paediatricians, audiologists, otolaryngologists and speech pathologists/educational specialists. Management of hearing loss is dependent on the etiology. Early intervention strategies may be placed into the following broad categories: audiological, medical/surgical management; educational and (re)habilitation methods; and child and family support. Medical and surgical interventions focused on establishing functional access to sound have improved significantly as a result of technological advances during the past two decades. Depending on the etiology and severity of hearing loss, this may involve hearing aids, cochlear implants or bone-anchored hearing aids. Rarely, brainstem-implanted auditory devices may be used. Surgical options exist for many conductive disorders including ear malformations, ossicular chain abnormalities, tumours and cholesteatomas. Hearing aids, which offer sound amplification, are now widely available using advanced digital technology, and may be worn by very young infants. Environmental sound amplification devices, including FM and wireless devices, are also available for individuals of all ages.

Cochlear implants, used in children for the past 20 years, are electronic devices surgically placed in the cochlea to provide stimulation to the auditory nerve. A systematic study (30) showed clear effectiveness in hearing and language development. Cochlear implants, along with oral language habilitation, have transformed the hearing and language potentials of severely and profoundly deaf individuals, enabling highly functional language development. Current recommendations for eligible children are bilateral implantation between eight and 12 months of age, coupled with auditory oral therapy (30).

Habilitation strategies focus on the development of ‘linguistic competence and literacy development’ in children who are deaf or hard of hearing (7). This may take many forms including oral and gestural communication, or a combination of both. Families require clear, objective information on the interventional options and expected outcomes. With the advances in hearing aid and cochlear implant technology, along with early intervention, functional oral communication and mainstream education are realistic goals for many hearing-impaired children. Thus, in North America, the development of spoken language is the primary objective of almost all English-based programs for hearing-impaired children (31). Data from the Ontario newborn hearing screening program indicate that between 2001 and 2007, 91.8% of parents selected oral communication as the interventional objective of choice for their child (unpublished data).

Specialized auditory-verbal therapists, teachers of the deaf and speech therapists, who are trained to work with infants and young children and their families, are instrumental in the auditory habilitation process. It is also widely recognized that parental/caregiver involvement is essential. Therefore, child and family support is a key element of early intervention. Caregivers benefit from family-centred guidance, focusing on an enriched daily exposure to language. Family support groups and access to up-to-date information are also essential. For older children, liaisons with school services

are important. In the 2007 position statement (7), the JCIH summarized intervention strategies and provided recommendations.

RECOMMENDATIONS

Based on the available evidence, the Paediatric Society recommends hearing screening for all newborns. This should be provided universally to all newborns via a comprehensive and linked system of screening, diagnosis and intervention. Several provinces, offer excellent examples of integrated systems. Advocacy, at the provincial and federal levels, is required to ensure that all infants can benefit from the advantages of early hearing loss detection and intervention.

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