Universal newborn hearing screening

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, Community Paediatrics Committee
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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 1996 to the third week of August 2009. 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 nonrandomized 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.

Key Words: Deafness; Early intervention; Hearing screening; Newborns

Background
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 – definitions
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) \((\text{Table 1})\). 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 [8]. Overall, known risk factors are present in only 50% of infants born with hearing loss (Table 2) [7][8]. Because a substantial proportion of infants have no risk factors, universal screening has replaced selective screening in most developed nations.

### TABLE 1

<table>
<thead>
<tr>
<th>Hearing loss severity</th>
<th>Hearing threshold (dB)</th>
</tr>
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<tbody>
<tr>
<td>Normal hearing</td>
<td>0–20</td>
</tr>
<tr>
<td>Mild</td>
<td>20–40</td>
</tr>
<tr>
<td>Moderate</td>
<td>40–60</td>
</tr>
<tr>
<td>Severe</td>
<td>60–80</td>
</tr>
<tr>
<td>Profound</td>
<td>&gt;80</td>
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Data from reference [37]

### TABLE 2

<table>
<thead>
<tr>
<th>Risk factors for neonatal sensorineural hearing loss</th>
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<tr>
<td>Family history of permanent hearing loss</td>
</tr>
<tr>
<td>Craniofacial abnormalities including those involving the external ear</td>
</tr>
<tr>
<td>Congenital infections including bacterial meningitis, cytomegalovirus, toxoplasmosis, rubella, herpes and syphilis</td>
</tr>
<tr>
<td>Physical findings consistent with an underlying syndrome associated with hearing loss</td>
</tr>
<tr>
<td>Neonatal intensive care unit stay &gt;2 days OR with any of the following regardless of the duration of stay:</td>
</tr>
<tr>
<td>• Extracorporeal membrane oxygenation</td>
</tr>
<tr>
<td>• Assisted ventilation</td>
</tr>
<tr>
<td>• Ototoxic drug use</td>
</tr>
<tr>
<td>• Hyperbilirubinemia requiring exchange transfusion</td>
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</tbody>
</table>

Adapted from references [7][8]

### Diagnosis is significantly delayed without screening

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].

### Functional outcomes of delayed diagnosis and intervention

Without early intervention, children with hearing loss demonstrate predictable irreversible deficits in communication and psychosocial skills, cognition and literacy [9][12]. There is clear evidence that auditory deprivation in early infancy leads to structural and functional reorganization at a cortical level [13][14] – similar to amblyopia in an infant with visual deprivation. The impact on the child’s speech and language is directly proportional to the severity of hearing loss and the time delay in diagnosis and intervention [1]. In a detailed review of the psychological and functional outcomes of deafness on the child and adolescent, Mason and Mason [15] reported that this group demonstrates impaired socioemotional development including low academic achievement, underemployment, increased social maladaptation and psychological distress. In unscreened populations, children with severe to profound hearing loss typically graduate from high school with the language and reading level of a nine- to 10-year-old child [16]. These limitations in literacy, with the associated impact on socioeconomic and vocational status [17], are reflective of the critical importance of timely sensory input to the developing brain of young children and also to the inter-relatedness of hearing, speech, reading and writing at a neurocortical level.
Screening

Historically, clinical screening for hearing loss in infants and young children was limited to observation of the behavioural response to a sound, such as a ringing bell, introduced out of direct vision of the child. No studies were found to assess this method; however, the diagnosis of hearing loss in populations relying on this method typically occur only when the child demonstrates a significant and irreversible language delay. The presence of parental concern about their child’s hearing is predictive of true hearing loss; however, the absence of such concern is not an effective screening tool \[^{[18]}\].

As with all screening programs, the assessment of UNHS requires careful analysis. Based on the World Health Organization (WHO) screening guidelines \[^{[19]}\], successful screening would include the following components: availability of accurate, reliable screening tool(s); demonstration of earlier diagnosis; consideration for adverse effects of screening; evaluation of the availability and effectiveness of earlier intervention following diagnosis; consideration of the adverse effects of earlier intervention; and evaluation of the longer-term outcomes from earlier diagnosis and intervention.

**What tests are used to screen newborns for hearing loss? Do these tests accurately identify moderate to profound sensorineural hearing loss?**

Currently, hearing screening in newborns is performed via otoacoustic emission (OAE) and automated auditory brainstem response (AABR) testing. A summary of the test characteristics is provided in Table 3. These physiological, noninvasive, automated screening tests can be performed at the bedside in term and preterm 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.

**TABLE 3**

<table>
<thead>
<tr>
<th>Characteristics of the otoacoustic emission (OAE) and automated auditory brainstem response (AABR) screening tests</th>
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<tr>
<td><strong>OAE</strong></td>
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<tr>
<td>Performed by a trained technician; 10–15 min; portable equipment brought to the bedside</td>
</tr>
<tr>
<td>Screening best performed in infants older than 24 h, with a minimum 34 weeks' corrected gestational age</td>
</tr>
<tr>
<td>Results may be affected by the infant's movements, environmental noise or dysfunction in the middle or external ear (eg, debris in the external ear canal)</td>
</tr>
<tr>
<td>Noninvasive; ear probe placed in outer ear canal</td>
</tr>
<tr>
<td>Ear-specific testing; both ears can be tested simultaneously</td>
</tr>
<tr>
<td>Identifies conductive and cochlear hearing loss from the level of the external ear to the level of the outer hair cells in the cochlea</td>
</tr>
<tr>
<td>Screening thresholds set to detect at least moderate hearing loss (30–40 dB)</td>
</tr>
</tbody>
</table>

Data from references \[^{[1]}\][\[^{[2]}\][\[^{[7]}\]]

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 \cite{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 \cite{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 \cite{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 Table 2, particularly 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 \cite{23}.

**Effectiveness of UNHS for earlier diagnosis and intervention in infants with hearing impairment**

In two systematic reviews \cite{24,7}, there was sufficient evidence to conclude that infants who are screened are identified earlier and receive intervention earlier. This conclusion is best supported by the results of a large controlled trial from Wessex, England \cite{23}, and multiple subsequent cohort studies. Based on the 2001 USPSTF statement \cite{8}, the 2008 update \cite{2} and multiple subsequent studies, it is clear that newborn screening significantly lowers the age of diagnosis of moderate to severe hearing loss in children, with the greatest reductions occurring in children with moderate hearing loss \cite{8,23}. For example, in the Champagne-Ardenne region of France \cite{24}, the median age at diagnosis of hearing impairment decreased from 17 months to 10 weeks with UNHS implementation. The American Academy of Pediatrics’ Joint Committee on Infant Hearing (JCIH) recommends that UNHS programs aim for a hearing impairment diagnosis by 12 weeks of age or younger as a reasonable and expected outcome parameter \cite{7}.

The actual diagnosis of hearing loss in infants is both precise and reliable. Systematic reviews \cite{25} have shown that frequency-specific auditory brainstem responses can predict, with a high degree of confidence, the configuration, severity and nature of the hearing loss in infants. This test, as well as high-frequency tympanometry (middle ear function analyzer) and diagnostic OAEs, are performed by trained audiologists and are available in most Canadian regions.

**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 \cite{8} and again in 2008 \cite{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 \cite{26}, originally published in 2005 and now withdrawn due to lack of revision, cited a similar conclusion. In the most recent USPSTF review \cite{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 \cite{22,7,25,26} 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 \cite{7}.

**Intervention strategies**

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.

What are the adverse effects of screening?

This question was systematically evaluated by Nelson et al [2] in 2008. Data were extracted from two fair-quality cohort studies and multiple survey studies. In summary, it appears that screening is well accepted by the vast majority of parents, with rates of refusal estimated to be 0.08% [2]. Studies show some anxiety in parents, particularly in those whose infants require follow-up testing. Anxiety was found to be highest in parents whose infants had confirmed hearing loss. Integrated parental information and counselling is recommended as part of a high-quality UNHS program.

The false-positive rates, indicating the proportion of normally hearing children who are referred for diagnostic testing, are reported to be between 2% and 4% in most UNHS programs [2], with well-established programs reporting rates of 0.5% to 1.0%. Comparatively, the false-positive rates for newborn thyroid screening are approximately 2%.

Limitations of UNHS

While the overall benefits of UNHS are increasingly clear, limitations exist. Implementation requires a comprehensive and organized approach that includes screening, diagnosis, intervention and follow-up. Less severe congenital hearing loss (less than 30 dB to 40 dB) is not detected in most UNHS programs. Progressive or late-onset hearing impairment, for example, as seen with congenital cytomegaloviral infection or in some inherited conditions, is also not detected by a newborn screening program. In two-step screening, low-risk infants with auditory neuropathy may not be detected by the OAE test alone. Health care providers, educators and parents must remain attentive to the developmental progress of children, especially in expressive and receptive language domains. A hearing (re)assessment is recommended.
for all children experiencing developmental or learning difficulties.

**Cost effectiveness of UNHS**

Beyond the quality of life and psychosocial benefits of improved language, communication and learning, there are increasing data on the cost effectiveness of UNHS. The actual costs of screening vary according to region. In general, there is agreement that the lifetime costs of deafness, particularly prelingual, are very high [32][33]. Costs of UNHS are comparable with other newborn screening programs [34] and, even with wide modelling parameters, the benefits of UNHS outweigh the costs [35]. In February 2008, the Institut national de santé publique du Québec published a detailed report [17] on the costs and benefits of UNHS. They reported that provincial implementation of UNHS, costing approximately $5.3 million in 2001, would result in a net benefit of $1.7 million per year to taxpayers, primarily through educational and vocational savings.

**Role of the physician**

In their role as health care advocates for children, physicians and, particularly, paediatricians should be aware of UNHS and whether it is available in their practice region. For infants with positive or equivocal test results, timely follow-up and parental compliance with the UNHS program recommendations are critical. In regions where UNHS has not been implemented, parents of newborns and young children need to be aware that clinical screening is ineffective in early diagnosis and that late diagnosis is associated with irreversible long-term language and cognitive deficits. Advocacy for UNHS throughout Canada is needed.

In a child with confirmed hearing loss, the etiology requires clarification. A detailed family history should be included. The medical evaluation, including history and physical examination, will be instrumental in determining whether the child has associated comorbidities, and/or syndromic or nonsyndromic hearing loss. Consultation with a paediatric otolaryngologist, ophthalmologist and geneticist is indicated. Prompt vision assessment is important in maximizing sensory input and determining whether there is an underlying genetic condition (eg, Usher syndrome). Further evaluations, including neuroimaging studies, specific genetic testing, and renal and cardiac evaluation require consideration on a case-by-case basis.

Physicians can help facilitate timely referral for medical, educational or surgical interventions. Knowledge of these strategies and expected outcomes will help inform and support parents. Health care providers should also be aware of the increased risk of complicated otitis media and meningitis in the general population of children with hearing loss [36]. Children with cochlear implants have a multifactorial increased risk of meningitis; specific recommendations for preventive vaccination have been made [36].

In the evaluation of all children, paediatricians are expected to be familiar with normal patterns of language development and to provide ongoing developmental surveillance. Children with known risk factors require close monitoring. Parental concerns should be seriously considered, with prompt referral for a hearing evaluation. More detailed guidelines on the role of the paediatrician, from expert consensus, are available [7][25].

**Recommendations**

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

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**References**


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