Alberta STE Report

The Safety and Efficacy/Effectiveness of Using Automated Testing Devices for Universal Newborn Hearing Screening: An Update

October 2012

IHE
INSTITUTE OF HEALTH ECONOMICS
ALBERTA CANADA

ACKNOWLEDGMENTS

The Institute of Health Economics is grateful to the Expert Advisory Group for provision of information and comments on the draft report.

The views expressed in the final report are those of the Institute of Health Economics.

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Funding

This report was supported by a financial contribution from Alberta Health (AH) through the Alberta Health Technologies Decision Process: the Alberta model for health technology assessment and policy analysis. The completed report was submitted to AH in September 2011.

The views expressed herein do not necessarily represent the official policy of Alberta Health.

Declared Competing Interest of Authors

The authors of this publication claim no competing interests.

Suggested Citation (ICMJE or Vancouver Style)

Institute of Health Economics. *The Safety and Efficacy/Effectiveness of Using Automated Testing Devices for Universal Newborn Hearing Screening: An Update*. Edmonton AB: Institute of Health Economics. 2012.

Web Address

This publication is available for free download from the IHE website at: http://www.ihe.ca.

EXECUTIVE SUMMARY

Technology Effects and Effectiveness

Background

Permanent congenital hearing impairment/loss (PCHI/PCHL) is one of the most common congenital anomalies found at birth; it can be expected to lead to delays and deficits in the development of speech, language, cognition and learning, as well as secondary effects on the child and the family. Recently published evidence suggests that early identification and subsequent appropriate intervention (within the first 6 months) in infants with PCHL can minimize these effects. As a result, there has been a growing interest for universal newborn hearing screening (UNHS), in the attempt to diagnose PCHL as early as possible. The rapid expansion of UNHS programs during the last decade has brought into focus questions about the most appropriate screening technology for this indication.

Objectives

To update the 2007 review[1] of the published evidence on the efficacy/effectiveness and safety of using automated testing devices to detect PCHL in UNHS.

Results

According to results reported by three systematic reviews:

- Automated evoked otoacoustic emissions (AOAE) and automated auditory brainstem response (AABR) appear to be equally accurate screening tests for detecting moderate to profound PCHL. However, the available evidence is inadequate to support either AABR or AEOAE as the preferred testing method for one-stage protocols.
- There is good evidence to suggest that a two-stage protocol, using AOAE (transient evoked otoacoustic emissions, TEOAE) testing followed by AABR testing, is effective in increasing early identification of moderate to profound PCHL and may lead to early intervention in diagnosed infants. This protocol may achieve specificity of 98.5% and sensitivity of 91.7%, with a positive predictive value of 6.5%. Referral for confirmatory diagnostic testing and management of PCHL commonly occurs earlier and more frequently with a UNHS using TEOAE followed by AABR than without UNHS.
- The efficacy/effectiveness of using AOAE and/or AABR in UNHS programs in terms of longer-term outcomes (such as development of speech and language, cognitive ability, and communication skills as well as quality of life, mental health, satisfaction, and educational and professional status) may be difficult to establish because the impact on developmental outcomes and other patient-relevant outcomes relates to many other factors than just accuracy of screening technologies. Loss to follow-up is a major limiting factor for overall program effectiveness.
- The impact of a UNHS program on patient-relevant outcomes, such as language and communication development in an infant with PCHL, is still not clear. Receptive language acquisition improvements are reported with UNHS, but unclear findings have been reported from expressive language.

- The impact of a UNHS program on other long-term patient-relevant outcomes (such as quality of life, educational development, and employment status) has yet to be established.
- No safety issues or concerns have been reported associated with applying the AABR and/or AOAE technology in newborns. Limited data on psychosocial harms of UNHS indicated no significant differences in measures of concern, anxiety, and parental attitudes for families with newborns who pass versus those with newborns who do not pass the newborn screening test. Likewise, no differences in anxiety levels were found between parents of unscreened newborns or of screened newborns, regardless of whether the screening outcome was positive or negative.
- No definitive data exists to determine which is the best of the AOAE and/or AABR devices currently available on the market in Canada.

Conclusions

Based on the results reported by three systematic reviews, this review confirms previous findings that UNHS using AOAE (TEOAE) followed by AABR testing in a two-stage protocol is effective in increasing early identification of moderate to profound PCHL and early intervention in diagnosed infants. The available evidence indicates that referral for confirmatory diagnostic testing and PCHL management commonly occurs earlier and more frequently with a UNHS using this protocol than without UNHS. It also indicates that early identification and treatment of infants with hearing loss may be associated with advantages in language development. The risks and harms of UNHS seem slight. However, the data on early detection of hearing loss in newborns and infants are not very robust. Further investigation is warranted, particularly as to the effect on longer-term patient-relevant outcomes such as quality of life and educational development.

Currently, no definitive data exists to determine which is the best of the AOAE and/or AABR devices currently available on the market in Canada. These devices still await prospective validation against an accepted gold standard.

UNHS using the automated testing devices represents only one component of a well-integrated and structured system of early identification and management for all infants who have hearing loss that enables confirmation of hearing loss by 3 months of age, and enrolment in a family-centered intervention program by 6 months of age. Resources need to be available for diagnosis and intervention before UNHS can be considered. An important measure for the practical realization of early detection of hearing impairments in newborns and infants seems to be the installation of a functioning system for registering and tracking both non-screened children and screened children with a conspicuous screening result.

Methodology

Research studies reporting on the on the safety and efficacy/effectiveness of using automated testing devices to screen for PCHL in asymptomatic newborns were identified through a comprehensive, systematic search of the literature published in English between 2006 and February 2012. The search included: The Cochrane Library NHS Centre for Reviews and Dissemination Databases (EED, HTA, DARE), PubMed, EMASE, and CINAHL. Also searched were the web sites of various health technology assessment (HTA) agencies, evidence-based resources, and clinical practice guidelines

For the purpose of this review, only published reports of systematic reviews and HTA studies were selected to formulate the evidence base on the safety and efficacy/effectiveness of using automated testing devices for UNHS. Not included were individual primary research studies (of any design) published subsequent to the selected systematic reviews and HTA studies. One reviewer performed the study selection and extracted the data from the selected studies. The evidence was qualitatively synthesized and presented in summary tables. An informal quality assessment for all selected research studies was completed during the final study selection process.

Economic Analysis

Objective and method

The objective is to compare the cost effectiveness of various strategies used in UNHS. A review was conducted of the published economic literature on the cost effectiveness of alternative strategies for UNHS. The review was an update to the previous IHE report[1] on UNHS, published in 2007.

Results

Four studies met the final inclusion/exclusion criteria; however, only one of the four studies was of acceptable quality. Uus et al.[2] found that the cost per additional case detected was £12,500 for NHSP (TEOAE + AABR soon after birth) compared with IDTS (responses to low level sounds conducted at 8 months of age). The cost per case detected indicates that NHSP is not cost savings (that is, greater than 0) but it should be mentioned that the time horizon adopted in their analysis did not examine longer term outcomes and therefore would not have captured downstream cost impacts from earlier detection and intervention.

The 2007 IHE report did conduct an economic evaluation of one-stage (AABR) and two-stage (AABR and AOAE) protocols that included downstream costs associated with untreated hearing loss. The analysis showed that that the one-stage AABR screening protocol is cost effective compared to the one-stage AOAE protocol, because the one-stage AOAE protocol is less accurate and more costly. The two-stage protocol was found to be more effective in comparison to the one-stage AABR, but it was also associated with higher costs, with an incremental cost-effectiveness ratio of $7574.78 ($CAD 2003) per infant detected.

Conclusion

Limited published economic evidence informing the cost effectiveness of UNHS strategies is available. However, based on the economic evaluation conducted for the 2007 IHE report, one-stage AABR screening is less costly and more effective than one-stage AOAE screening, while the cost effectiveness of two-stage screening of AABR and AOAE is dependent on whether the additional effectiveness is worth the additional cost.

ABBREVIATIONS

AABR	automated auditory brainstem response
ABR	auditory brainstem response
ACSLPA	Alberta College of Speech–Language Pathologists and Audiologists
AHRQ	Agency for Healthcare Research and Quality
AN	auditory neuropathy
AOAE	automated evoked otoacoustic emissions
CASLPA	Canadian Association of Speech Language Pathologists and Audiologists
CI_{95}	95% confidence interval
CPS	Canadian Pediatric Society
dB	decibel(s)
EHDI	early hearing detection and intervention
HL	hearing Loss
HVDT	Health Visitor Distraction Test
Hz	hertz
IDTS	Infant Distraction Test Screen
IQWiG	Institut für Qualität und Wirtschaftlichkeit im Gesundheitswesen (Institute for Quality and Efficiency in Health Care)
JCIH	Joint Committee of Infant Hearing
mo	month(s)
MSAC	Medical Services Advisory Committee
NHS	National Health Service
NICU	Neonatal Intensive Care Unit
NNS	number needed to screen
NPV	negative predictive value
OAE	evoked otoacoustic emissions
PCHI	permanent congenital hearing impairment/loss
PCHL	permanent congenital hearing loss
PPV	positive predictive value
QUADAS	quality assessment of diagnostic accuracy studies
RCT	randomized controlled trial

ROC	receiver operating characteristic(s)
SCBU	special care baby unit
Sn	sensitivity
SNHL	sensorineural hearing loss
Sp	specificity
TEOAE	transient evoked otoacoustic emissions
UK	United Kingdom
UNHS	universal newborn hearing screening
US(A)	United States (of America)
USPTF	US Preventive Services Task Force
VRA	visual reinforcement audiometry
WBN	well-baby nursery
wk	week(s)

GLOSSARY

The glossary terms listed below were obtained and adapted from:[1]

www.phsa.ca/AgenciesAndServices/Services/BCEarlyHearing/ForFamilies/Glossary.htm

Auditory brainstem response (ABR): A measure used to predict hearing sensitivity and to assess the integrity of the eighth cranial nerve, or hearing nerve, and brainstem structures.

ABR test: A hearing test that measures an electrical response of the inner ear and auditory nerve. Young children are usually sleeping or sedated for this test. Small disk-shaped sensors are placed behind each ear and on the child's forehead. Sounds are then directed to the ears using miniature earphones.

AABR or automated ABR: A measure in which the recording is under computer control and detection of a response is determined automatically by the computer (see "**screening**").

Atresia: The lack of or narrowing of an ear canal opening where sound normally travels by air waves to the middle ear. Complete atresia results in a conductive hearing loss.

Audiogram: A chart or graph that shows how well a person hears. An audiogram can be thought of as a picture of your child's hearing. The audiogram shows the quietest level of sound your child can hear at each frequency (pitch) in each ear. Go to the page on how to read an audiogram for more information.

Audiologist: A health professional who identifies people who have hearing problems, and works with these people to help to improve their communication. Their role includes diagnosing hearing loss and fitting hearing aids. Audiologists in Canada have a minimum of a master's degree and are certified by the Canadian Association of Speech–Language Pathologists and Audiologists.

Audiometer: An electronic equipment used to test hearing.

Auditory nerve: The hearing nerve that connects the inner ear (cochlea) to the brain and sends messages from the ear to the brain.

Bilateral hearing loss: Hearing loss in both ears.

Bone-conduction testing: A test in which a bone vibrator placed behind a child's ear stimulates the inner ear directly and bypasses the middle ear to determine the location of the child's hearing problem. A problem located in the inner ear (cochlea) indicates a permanent (sensorineural) hearing loss.

Bone oscillator: The vibrating headphone used in bone-conduction testing.

Cochlea: A spiral-shaped, bony casing that forms the inner ear with many nerve endings inside of it.

Conductive hearing loss: A type of hearing loss characterized by problems with the outer or middle ear. An example of a problem with the outer ear is atresia, in which there is no opening to the ear canal. Problems with the middle ear can be the result of fluid in the middle ear, or there can be something wrong with the three little bones in the middle ear. Sometimes a conductive hearing loss is temporary, when it is the kind of problem that can be medically treated (such as fluid in the middle ear).

Congenital hearing loss: A hearing loss that is present at birth or associated with the birth process or which develops within the first few days of life.

Decibels (dB): Intensity (loudness) of sound is measured in decibels. For example, a sound that measures 100dB is a very loud sound, and one that measures 10dB is a very quiet sound.

ENT: An ear, nose, and throat specialist (that is, an otolaryngologist).

Evoked potential (EP): The electrical activity of the brain (and different structures; brainstem, cortex, etc.) in response to sensory (for example, auditory) stimulation.

False positive: A test outcome that indicates the presence of a disease or condition when, in fact, that disease or condition is not present.

Hearing threshold: The quietest sound that a person can hear.

Impedance/immittance testing: A hearing test during which a small probe is placed in the ear to determine if there is a problem in the middle ear.

Inner ear: The part of the ear that contains the cochlea and the auditory nerve, as well as the balance organ.

Latency: The time interval between two events (for example, between a stimulus and a response).

Longitudinal: Pertaining to research design in which same subjects are observed repeatedly over a period of time.

Loss of infants to follow up: in reference to when an individual or child is not seen for follow-up procedures due to factors such as low compliance from parent, moved to another province, lack of services available, and lack of tracking systems in place.

Middle ear: The middle section of the ear that contains three tiny bones through which sound is conducted from the eardrum to the inner ear.

Mild hearing loss: A degree of hearing loss in which a person is unable to detect sounds until they are in the loudness range of 26dB to 40dB.

Moderate hearing loss: A degree of hearing loss in which a person is unable to detect sounds until they are in the loudness range of 41dB to 55dB.

Moderately severe hearing loss: A degree of hearing loss in which a person is unable to detect sounds until they are in the loudness range of 56dB to 70dB.

Otoacoustic emissions (OAEs): Low level sound emitted by the cochlea, evoked by an auditory stimulus or echo; related to the functioning of normal outer hair cells of the cochlea.

Otoacoustic emissions (OAEs) test: A test in which a sensitive microphone is placed in the ear while the audiologist presents several soft clicks or tones. If the inner ear (cochlea) is normal, it makes sounds back (called otoacoustic emissions) that are picked up by the microphone. When these responses are present it usually means hearing is normal. If these responses are absent, it may indicate hearing loss. Responses may also be absent due to things such as wax in the ear canal or the presence of fluid in the middle ear.

Outer ear: The visible part of the ear that we can see, as well as the ear canal, which channels sound from outside through to the eardrum.

Physiological (physiology): Refers to the function of living organisms and their components.

Profound hearing loss: A degree of hearing loss in which a person is unable to detect sounds until they are in the loudness range of 90dB or louder.

Prospective (of future): The strategy of maintaining a watch over a suspected population after an event.

Screening: The application of rapid and simple tests to a large population, consisting of individuals who are undiagnosed and typically asymptomatic, in order to identify those who require additional diagnostic procedures. Screening typically results in either a "pass" or a "refer" outcome.

Sensitivity: The ability of a test to detect the disorder it was designed to detect; expressed as the percentage of positive results in those patients having the disorder.

Sensorineural hearing loss: Hearing loss that occurs because of a problem in the cochlea (inner ear).

Severe hearing loss: A degree of hearing loss in which a person is unable to detect sounds until they are in the loudness range of 71dB to 90dB.

Single-sided deafness: Hearing loss in one ear only, with the degree of loss being in the severe to profound range. Single-sided deafness is sensorineural, that is, either the inner ear (cochlea) and/or the auditory nerve are affected. Another term for this is unilateral deafness.

Slight hearing loss: A degree of hearing loss in which a person is unable to detect sounds until they are in the loudness level of 16 – 25dB.

Sloping hearing loss: A child's hearing loss is not the same across all frequencies (pitch). In most cases, a child with a sloping hearing loss has better hearing in the lower frequencies than in the higher frequencies.

Specificity: The ability of a test to differentiate a normal condition from the disorder the test was designed to detect. Specificity is expressed as the percentage of negative results in patients who do not have the disorder.

Tympanogram: A graph or chart that records the results of tympanometry testing.

Tympanometry testing: A hearing test in which a small probe is placed in the ear while the movement of the eardrum is measured to determine whether a problem exists in the middle ear.

Unilateral hearing loss: Hearing loss in only one ear.

Universal: Available and applicable to all, without discrimination.

Visual reinforced audiometry (VRA): A hearing test typically used for infants over 6 months of age, up to about 2 or 3 years of age. It involves teaching a child to turn toward sounds, using toys that light up as a reward. Sounds are presented through headphones and/or speakers in order to determine, for different kinds of sounds, the softest sound to which the child will respond.

Well-baby: Refers to babies who are not admitted to special care units.

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SECTION ONE: TECHNOLOGY EFFECTIVENESS/EFFICACY

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INTRODUCTION

Hearing impairment/loss, whether congenital or acquired during the first year of life, represents a barrier to speech and language acquisition and can interfere with a child's overall development.[1–11] According to recently published evidence, early identification and subsequent appropriate intervention (within the first 6 months) in children with permanent hearing loss can minimize these effects. As a result, interest has been growing in universal newborn hearing screening (UNHS), in the attempt to diagnose permanent hearing impairment in childhood as early as possible. The rapid expansion of UNHS programs during the last decade has brought into focus questions about the most appropriate screening technology for this indication.

This review updates a prior review[12] conducted in response to a request for information from Alberta Health about the safety and efficacy/effectiveness of using automated testing devices to detect permanent congenital hearing impairment/congenital permanent childhood hearing impairment/loss (PCHI/PCHL) in asymptomatic newborns. This review aimed specifically to answer the following questions:

1. What is the accuracy of the automated testing devices for differentiating newborns with normal hearing from those who need to be referred for diagnostic confirmation of PCHL and for appropriate intervention within their first 6 months of life?
2. Does the use of automated testing devices affect the detection rate for PCHL in infants within their first 6 months of life?
3. Does the use of automated testing devices affect the age at diagnosis of PCHL in infants?
4. Does the use of automated testing devices affect the age at start of treatment for PCHL in infants?
5. Does the use of automated testing devices affect the treatment decisions for PCHL in infants within their first 6 months of life?
6. Does the use of automated testing devices affect the developmental milestones (such as speech and language development), in infants and children diagnosed with PCHL?
7. Are there any side effects and complications to the newborn/infant and/or the screener due to performing the automated testing itself for UNHS?

The scope of the report was defined as follows:

Population: Asymptomatic newborns *(newborns defined: birth through 3 months; asymptomatic newborns defined: not necessarily considered at risk for congenital hearing impairment/loss)* at urban or rural settings (birthing hospitals).

Intervention: Automated testing devices approved/licensed in Canada to detect PCHL in asymptomatic newborns in UNHS programs.

Comparators: Screening devices(s) considered as reference standard for this indication, or with other tests considered for this indication, or with no testing.

Outcomes: Sensitivity, specificity, and predictive value; impact on age at diagnosis of PCHL; impact on the number of infants diagnosed with PCHL; impact on usage of diagnostic tests; impact on age at start of treatment for PCHL; impact on treatment decisions (such as type of treatment); impact on usage of interventions to treat PCHL; impact on speech and language acquisition and development in children diagnosed with PCHL; impact on social and emotional development and other developmental milestones (such as scholastic achievement) in children with PCHL; risks and complications to the newborns and/or screeners from performing the test itself; and adverse effects of false positive and false negative test results.

This review also aimed to determine the best practice for conducting UNHS and included the following elements of assessment:

- a systematic search for scientific literature published about the use of the automated testing devices of interest
- a summary of findings reported by published systematic reviews reporting about the safety and efficacy/effectiveness of using the automated testing devices of interest
- a summary of recommendations from relevant evidence-based clinical practice guidelines (CPGs), position statements, and consensus documents about best practice for UNHS and/or the use of the automated testing devices of interest
- clinical input from Expert Advisory Group members

The literature search strategy, data sources, and methods used for screening and reviewing the retrieved literature are described in more detail in Appendices A and B.

CLINICAL CONDITION: HEARING IMPAIRMENT/LOSS IN CHILDHOOD

Definition of Condition

Hearing impairment and hearing loss in childhood are terms used to describe a child's complete or partial loss of the ability to hear in one (unilateral) or both ears (bilateral) resulting from an abnormality or disorder anywhere in the auditory system (for example, auricle, external auditory canal, middle ear, inner ear, auditory nerve, central auditory pathways, and/or auditory cortex).[1–6,8–11,13,14] Hearing impairment/loss may be central, sensorineural, conductive, or a combination of sensorineural and conductive etiologies (mixed) with additive effects, and may be congenital, acquired, transient, fluctuating, recurrent, progressive, or permanent. Sensorineural hearing loss (SNHL) involves a problem with the inner ear (for example, insult to the cochlea or cochlear dysfunction) or a problem with the auditory nerve going from the cochlea to the brain. SNHL may be genetic or non-genetic; it is usually permanent and requires (re)habilitation. Conductive hearing loss is due to a problem in the outer (external) or middle ear; it is often medically or surgically treatable. Central hearing losses are rare and can be caused by problems along the auditory pathway or in the brain. Congenital hearing loss is present at birth or arises shortly thereafter as a consequence of progressive loss. Acquired hearing impairment occurs later in a child's life.

Hearing loss is classified according to the degree—or severity—of hearing loss and is described in decibels (dB), a unit of intensity or loudness at various hearing frequencies.[2–6,9,15] This is defined on the basis of a hearing threshold, the sound pressure level from which the hearing still detects an acoustic stimulus. Several different classification schemes have been used to describe degrees of hearing loss; currently, no system is universally accepted. The World Health Organization (WHO) classifies hearing loss as slight/mild (26–40dB, in better ear), moderate (41–60dB, in better ear), severe (61–80dB, in better ear), and profound, including deafness (≥81dB, in better ear).[15] However, this classification is not used consistently.

Known risk factors for hearing loss in newborns and infants include:[1,3–8,10,11,13,16,17]

- family history of permanent hearing loss
- craniofacial abnormalities, including those involving the external ear
- congenital infections (such as bacterial meningitis, cytomegalovirus, toxoplasmosis, rubella, herpes, and syphilis)
- neonatal intensive care unit (NICU) or special care baby unit (SCBU) length of stay of more than two days
- NICU/SCBU care (regardless of length of stay) for presence of extracorporeal membrane oxygenation or assisted ventilation, ototoxic drug use, or hyperbilirubinemia requiring exchange transfusion (with admission to NICU being an established risk factor particularly for auditory neuropathy)
- syndromes associated with hearing loss
- physical findings consistent with an underlying syndrome associated with hearing loss
- neurodegenerative or neurodevelopmental disorders.

Overall, known risk factors are present in only 50% of infants born with hearing loss.[1,3–8,10,11,16–18] The relative contributions of these factors are likely to vary from country to country. Preventing hearing loss in newborns and infants by eliminating these factors is unlikely to be achieved in any country.[10]

The determination of the prevalence of hearing loss in childhood depends on what is included in the target disorder.[3,19] Usually the target disorder is described by impairment severity, frequency range, laterality (one or both ears), and permanence, as well as by the site of the disorder in the auditory system and the associated categories of impairment type.

Definition of target disorder

For the purposes of this document, the target disorder is hearing impairment in childhood that is congenital and is stable or progressive, which is referred to as permanent childhood hearing impairment/loss or permanent congenital hearing impairment/loss (PCHI/PCHL). Most PCHL is sensorineural and irreversible.[1–7,9–11,13,20] Structural conductive impairments are usually included because, unless treated, they impose long-standing dysfunction. This condition has a wide range of severity, which may fluctuate over time. Severity may not be symmetrical (that is, may not be the same in both ears).

Prevalence and incidence of PCHL

PCHL is one of the most common anomalies found at birth and its prevalence increases throughout school-age.[1,3–5,7,9–11,16,20–22] Such increase is attributable to acquired, late-onset, and progressive impairment, the prevalence and time course of which are still unclear. The incidence of PCHL varies with race, gender, birth weight, intra-uterine infection, and other risk factors, including family history of hearing impairment or chromosomal abnormality.

Based on data reported in the international literature, recently estimated prevalence rates for bilateral PCHL at hearing loss > 35dB, > 40dB, and > 50dB range from 0.7 to 1.8 per 1000 infants and from 0.2 to 1.5 infants per 1000 infants for unilateral PCHL at all three levels of hearing loss.[3] The median prevalence of moderate to profound (> 35dB) bilateral and unilateral PCHL is estimated to be 1.3 and 0.6 per 1000 infants. The median prevalence of bilateral and unilateral PCHL > 40dB is estimated to be 1.3 and 1.2 per 1000 infants, respectively. The prevalence of bilateral and unilateral PCHL > 50dB is estimated to be:1.2 per 1000 infants. If audiometric criteria for PCHL are broadened to include lesser severities (down to > 25dB) and unilateral losses, the prevalence in early infancy increases to two to three per 1000 infants.[12]

Up to 50% of PCHL cases are thought to be due to environmental factors and the reminder to genetic causes.[6,10,14,23,24] In infants with high risk factors, such as prematurity, severe hyperbilirubinemia, or congenital craniofacial defects, the prevalence of hearing loss can be as high as 10 per 1000 live births.[1,8,10] The prevalence of hearing loss in newborns with specific risk indicators (NICU/SCBU population) is 10 to 20 times higher than in the general population of newborns (well-baby population).[1,7,8,10,11,25]

The prevalence of congenital hearing loss in newborns has long been thought to range from one to over three infants per 1000, or approximately 13,000 infants are born in the United States each year with some degree of permanent hearing loss.[2,9,21] Information published in 2009 indicates that the average incidence of neonatal hearing loss in the US is 1.1 per 1000 infants.[9,26,27]

The prevalence rate in Canada is not readily available,[1,11,28–31] but estimates from the province of Ontario indicate that three in 1000 infants suffer from permanent hearing loss, either at birth or developing early in childhood.[28] Of the 352,848 infants born in Canada between July 1, 2006 and June 30, 2007, it was estimated that 1000 to 1400 newborns have some degree of congenital hearing loss.[1]

A pilot UNHS program project, initiated in four former Alberta health regions (Mistihia/Peace Country, Palliser, Chinook, and Calgary) between 2001 and 2004 and funded by the Alberta Health Innovation Fund, provided the only Alberta-specific prevalence estimates.[1,12] The estimated prevalence was 4.02 per 1000 screened infants. Given an annual birth rate of 44,661 in Alberta (born between July 1, 2006 and June 30, 2007), an estimated 88 to 179 newborns with hearing loss would be identified per year, based on prevalence rates of two to four per 1000 live births.[1]

Burden of PCHL

Hearing loss can impose a heavy burden on the affected individuals, their families, and society.[1-11,13,16,20,21,32,33] PCHL can be expected to lead to delays and deficits in the development of speech, language, cognition and learning, social/emotional development, and vocational development as well as to secondary effects on the child and the family. The principal factors that may decide how PCHL

affects a child's overall development are the degree of hearing loss and the age at which it is diagnosed. Hearing disorders have also been associated with increased behaviour problems, decreased psychosocial well-being, and poor adaptive skills, affecting the child's quality of life. 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. Societal costs attributable to hearing loss include expensive special education services, a less productive subgroup of the work force (resulting in fewer tax dollars contributed over a lifetime), and individual costs that are both monetary and personal.

Early identification of PCHL

Consensus is growing that early identification of hearing loss in childhood can reduce the negative consequences of hearing loss for the child, the family, and society.[1–11,11,13,14,16,18,20,32] According to the Joint Committee on Infant Hearing (JCIH) in the United States, evidence indicates that when identification and appropriate intervention occur at no later than 6 months of age for newborn infants who are deaf or hard of hearing, they "perform as much as 20 to 40 percentile points higher on school-related measures (vocabulary, articulation, intelligibility, social adjustment, and behavior)".[8]

Age of identification of hearing loss is the underlying premise of newborn and infant hearing screening initiatives.[1–10,13,14,20,32] The age at which hearing loss is detected without a screening program depends on the severity of the hearing loss and is found later in those with less severe deficits. In unscreened children, the reported average age at diagnosis ranges between 1 and 3 years.[2,3,6,13,14,34]

A growing body of evidence documents that early detection of PCHL has been improved through newborn and infant hearing screening.[1–11,14,20,26,32,35,36] With screening, the median age of PCHL diagnosis ranges from 2 to 6 months. However, children not screened at birth, those lost to follow-up after failing newborn screening, and children who present with later-onset hearing loss may be identified too late to prevent serious developmental problems associated with untreated hearing loss.

Treatment of PCHL

Children with hearing loss are best managed by a coordinated team including family physicians, pediatricians, audiologists, otolaryngologists, and speech–language pathologists/educational specialists.[2–4,6,8,10,13,33,37] Management of hearing loss is dependent on the etiology and early intervention strategies and may be placed into the following broad categories:

- audiological, medical/surgical management
- communication and educational (re)habilitation methods
- child and family support

UNIVERSAL NEWBORN HEARING SCREENING (UNHS)

It is widely believed that early identification (at 3 to 6 months) and administration of appropriate intervention at or before 6 months of age provides a child with PCHL the opportunity to develop normal speech and language.[1–11,14,20,26,32,37–39] The logic that underlies these beliefs is sketched below:

- PCHL → impairment in language abilities → lifelong burden
- Habilitation → improvement in language ability → decreased burden
- UNHS → earlier diagnosis
- Earlier diagnosis → earlier (re)habilitation
- Earlier habilitation → greater improvement in language ability

On the basis of this reasoning, many countries have implemented UNHS as part of early identification and intervention programs.[1–6,8,10,11,20,23,40] Important characteristics of developing and implementing early detection and intervention programs include a family-centered approach, culturally responsive practices, collaborative professional–family relationships and strong family involvement, developmentally appropriate practice, interdisciplinary assessment, and community-based provision of services.

Prior to the implementation of UNHS, only newborns identified as being at high risk for PCHL, such as those in NICU, were routinely screened using a risk assessment tool (high-risk registry).[1–6,8,10–13,20,40,41] In comparison to the resources required for UNHS, substantially fewer resources are required to screen only the high-risk group. However, a major limitation of selective or targeted screening (in at-risk populations) is that as many as 50% of infants with PCHL have no known risk factors. The relatively high incidence of PCHL in infants without known risk factors, and the introduction of new and automated screening technology over the last 20 years, has led prestigious bodies in many countries worldwide (including the JCIH in the United States and the Canadian Paediatric Society) to recommend UNHS as an alternative to selective/targeted screening.[1–8,10,11,40]

Since 2007, the development and implementation of sophisticated physiological hearing screening techniques has contributed to the feasibility of UNHS worldwide. An increasing number of countries with different healthcare systems and different economic and social circumstances have implemented or are in the process of implementing UNHS programs.[1,4–6,8,10,11,16,20,23,38–40] UNHS is either endorsed and recommended or already practiced and legally regulated (nationally or regionally) in many European countries (for example, Germany, Austria, Great Britain/United Kingdom, Italy, Spain, the Netherlands, Belgium, and France), as well as in the United States, Canada, Australia, and various Asian countries.

Measurements Used for Detecting PCHL in UNHS

UNHS uses electrophysiologic measurements of evoked otoacoustic emissions (OAE) and/or auditory brainstem responses (ABR) to detect permanent sensory or conductive hearing loss in newborns and infants.[2–8,13,16,20,37] OAEs are forms of low intensity energy, measured as sound, generated by the outer hair cells of the human cochlea in response to controlled acoustical stimulation (sound/noise, either clicks or tones). In response to noise, vibrations of the outer hair cells in the healthy inner ear (normal cochlea) generate electrical responses (faint sounds) that are radiated back through the middle ear to the external canal. OAE presence or absence reflects normal or abnormal hearing sensitivity up to and including the cochlea. Although most normal ears yield OAE, the likelihood of obtaining a response decreases rapidly in the presence of a PCHI of 30dB or greater.

The ABR are auditory evoked potentials (originating from the auditory nerve) generated in the brainstem in response to controlled auditory signals (sound/noise) composed of either clicks or tones.[3–5,6,8,13,16,20,42] It consists of a series of peaks corresponding to the neural response to an auditory stimulus along the pathway between the auditory nerve and the brainstem. ABR measurements are obtained from surface electrodes that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli delivered via an earphone. Electrodes are placed on the head, and brain wave activity in response to sound is recorded. ABR testing can detect damage to the cochlea, the auditory nerve, and the auditory pathways in the brain stem.

Currently, the ability to screen large numbers of newborns in UNHS programs relies on the use of automated ABR (AABR) testing devices and automated OAE (AOAE) testing devices specifically designed for this purpose.[3–6,8,14,16,20,32,38,42–44]

AABR and AOAE as UNHS Screening Tools

AABR screening is an adaptation of conventional ABR testing, which is a widely accepted proxy gold standard measure of hearing sensitivity in newborns and infants.[3,4,6,8,12,14,16,45] The AABR device delivers a rapid series of low-intensity clicks (usually at about 35 dB hearing level) through an insert or supra-aural earphone and records electrical activity from the scalp via electrodes/sensors. Average electrical waveforms are computed and automated statistical response detection algorithms evaluate the presence or absence of the ABR. AABR systems compare an infant's waveform with that of a template developed from normative ABR infant data to determine a pass/refer ("fail") result. The test may take approximately ten minutes per baby.

AOAE screening measures either transient-evoked OAE (TEOAE) or distortion-product OAE (DPOAE) (www.infanthearing.org; www.otoemissions.org).[3,4,6,8,14] Both are frequency-specific measurements of peripheral auditory sensitivity. A transducer placed in the ear delivers the stimuli and records OAE for immediate computer processing. Multiple responses are averaged to get a specific repeatable waveform and a pass/refer result. The test may take less than five minutes per baby. The fact that a newborn infant has acceptable OAE (a pass case) at the tested frequencies does not imply that the newborn can hear (www.otoemissions.org/index_1024.html).

Both AABR and AOAE are noninvasive, rapid screening tests that are rapid and can be performed at the bedside in inpatient or outpatient settings, in term and preterm newborns.[2–4,6,8,16,38] The tests are performed on any newborn who is asleep, or at least at quiet rest, in a moderately quiet test environment. Both provide measurements of physiologic activities that are easily recorded in newborns and that correlate highly with the degree of peripheral hearing sensitivity. They are not mutually exclusive, but complementary. Neither method needs voluntary responses and either can be carried out on newborns and infants without sedation. However, there are important differences between the two measurements:

- AOAE measurements reflect the status of the peripheral auditory system extending to the cochlear outer hair cells.
- AABR measurements reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway.

The automation of OAE and ABR measurements and interpretation of results reduces the knowledge and skill required for performing these tests (www.infanthearing.org; www.otoemissions.org).[3–6,10,16,44] However, from the practical clinical viewpoint, neither AOAE nor AABR is simple to perform. Although the equipment has become much easier to use, it is still advisable for the tests to be conducted by well-trained and experienced screeners and to keep the referral rate reasonably low. The screeners must understand the techniques' limitations and some skill is required, especially in identifying an appropriate behavioural state of the newborn during testing, and in correctly placing the recording electrodes and earphone/ear probe. Both technologies allow a variety of personnel to be trained as screeners, including audiologists, nurses, midwives, physicians, speech–language pathologists, screening technicians, and volunteers/health visitors. Ongoing quality control is essential for accurate, consistent test results.

Although the use of AABR and AOAE testing has emerged as an integral part of newborn and infant hearing screening, neither provides a direct measure of hearing (www.infanthearing.org; www.otoemissions.org).[6,8,16] Both methods test the structural integrity of the auditory pathway. They are not considered true screening tests of hearing, because they don't assess cortical processing of sound. Even if a newborn or infant passes screening with these tests, hearing cannot be definitively considered normal until the child is mature enough for a reliable behavioural audiogram to be obtained.

AABR can screen for hearing loss due to auditory neuropathy (AN) in newborns, whereas AOAE does not screen for neural auditory pathology or dysfunction (www.infanthearing.org; www.otoemissions.org).[6,8,13,16,37,44,46] AN may comprise up to 10% of all PCHI in infants and the majority of newborns/infants with AN are likely to have been in NICU. Therefore, the use of AABR screening in all NICU graduates is recommended in order to identify most cases of AN.[8]

AABR lacks frequency-specific information and cannot be used to determine the degree or nature of hearing loss (www.infanthearing.org; www.otoemissions.org).[3,4,6,16] AOAE devices have potential for providing frequency specific information. However, AOAE screening may require interpretation by the screener while AABR screening does not. Although TEOAE technology has been used since the early 1990s for newborn hearing screening, many different pass/refer criteria are still being used in TEOAE-based programs (www.otoemissions.org). The screening criteria for DPOAE are protocol dependent.

False positive results from AOAE screening can be caused by any mechanism that interferes with sound transmission from the earphone to the cochlea and back to the recording microphone.[3,6,8,16] Common problems include debris or fluid in the middle ear or in the external canal; the latter is more common when AOAE screening is done within 24 hours of birth. AABR results are less affected by middle or external ear debris.

Screening with AABR devices takes longer in comparison to screening with AOAE devices (www.infanthearing.org; www.otoemissions.org).[6,16] However, due to improvements in AABR algorithms, the time differences between AABR and AOAE testing are decreasing (www.otoemissions.org/index_1024.html) Fourth generation AOAE equipment incorporates AABR modules, thus both technologies are being developed in parallel.

Some newborns/infants who pass hearing screening will later demonstrate permanent hearing loss.[6,8,16,19] Although this loss may reflect delayed-onset or later acquired hearing loss,[8] both AABR

and AOAE technologies will miss some hearing loss (such as mild or isolated frequency region losses).[6,8,12,16,19]

(See Table T.1 for a summary of the characteristics of AABR and AOAE technologies.)

Table T.1: Characteristics of AABR and AOAE

Technology	Advantages	Limitations
AABR (measures activity of auditory nerve and brainstem pathways)	Non-invasive Quick, simple operation Screening best performed in infants older than 24 h, with a minimum 34 wk corrected GA Provides ear-specific results; response not dependent on infant cooperation Can be carried out on infants without sedation Can screen for HL due to AN May be administered at bedside Requires no interpretation by screener Average referral rates for HL of ≤ 4% may be achievable using AABR alone Pass/refer results are immediately available Results are less affected by middle ear or external ear debris than are the results of AOAE A variety of personnel can be trained as screeners	Does not directly measure hearing and is not considered a true screening test of hearing Requires the infant to be sleeping or quiet May not detect infants with reverse slope loss, or those with risk factors for hearing deficits May not detect infants with mild and very mild HL and those with low frequency HL (screening threshold set to detect at least moderate HL) More susceptible to electrical interference Cannot provide frequency-specific information May be less acceptable to parents because of the need to apply electrodes to the infant
AOAE (measures cochlear response to controlled acoustic stimulus; provides information about ear structures up to and including the cochlea)	Non-invasive Quick, simple operation Screening best performed in infants older than 24 h, with a minimum 34 wk corrected GA Provides ear-specific results; response not dependent on infant cooperation Can be carried out on infants without sedation May be administered at bedside Average referral rates for HL of <4% may be achievable using AOAE alone (especially if screened after 24 h of age) Can provide frequency-specific information Motion artefacts interfere less with results Pass/refer results are immediately available A variety of personnel can be trained as screeners	Does not directly measure hearing and is not considered a true screening test of hearing Requires the infant to be sleeping or quiet Does not detect nerve or auditory brainstem pathway dysfunction May not detect infants with reverse slope loss, or those with risk factors for hearing deficits May not detect infants with mild and very mild HL and those with low frequency HL (screening threshold set to detect at least moderate HL) Debris or fluid in the external and middle ear can affect results Screeners may need to decide what constitutes a pass/refer response

AABR – automated auditory brainstem responses; AN – auditory neuropathy; AOAE – automated otoacoustic emissions; dB – decibel(s); GA – gestational age; h – hour(s); HL – hearing loss; wk – week(s)

No standards exist for the calibration of AOAE or AABR instrumentation and there is a lack of uniform performance standards.[8,35,45] Manufacturers of hearing–screening devices do not always provide sufficient supporting evidence to validate the specific pass/refer ("fail") criteria and/or the

automated algorithms used in their instruments. In the absence of national standards, audiologists must obtain normative data for the instruments and protocols they use.

AABR and AOAE in UNHS protocols

Depending on the UNHS protocol, AABR and AOAE testing may be performed alone (either AABR or AOAE in one-stage protocols) or combined in multi-stage (two- or three-stage) protocols, which use more than one test sequentially (www.otoemissions.org/index_1024.html; www.infanthearing.org).[3,4,6,8,10,18,20,25,37] The most common UNHS protocol is a two-stage screening process in which AOAE testing (either TEOAE or DPOAE) is performed first, followed by AABR in those newborns who do not pass the AOAE. The multi-stage protocols aim to achieve very low overall false positive rates.

Different approaches have been taken in the well-baby and NICU/SCBU nurseries.[3,4,6,8,20,25,38] Important issues differentiate the screening performed in the well-infant nursery from that performed in the NICU. Although the goals in each nursery are the same, numerous methodological and technological issues must be considered in program design and pass/refer criteria.

AABR and AOAE devices available in North America

Table T.2 lists the companies that currently offer automated testing devices for newborn and infant hearing screening in North America. Information sources for Table T.2 are:

- Health Canada website (see Appendix T.D)
- website of the National Center for Hearing Assessment and Management (NCHAM) at Utah State University (www.infanthearing.org)
- manufacturers' websites

These technologies include various models of portable or handheld (battery operated) devices, which can be stand-alone (TEOAE, DPOAE, or AABR only) and/or combination units. Combination units offer multiple configurations of TEOAE, DPOAE, and AABR technology in a single device, allowing AOAE/AABR or AABR/AOAE testing sequences in a single screening session. Under optimal screening conditions, testing can take 10 to 30 seconds per ear for measuring OAE and 1.5 to 2 minutes per ear for measuring ABR.[12] Test results can be stored in memory, for review on the display, or printed (a printer can be connected to the device).

Table T.2: Automated testing devices available in North America for newborn hearing screening

Manufacturer/ Distributor	Device Name	Technology		
		AABR	DPOAE	TEOAE
Grason-Stadler Inc (GSI) http://grason-stadler.us/	GSI 70 http://grason-stadler.us/index.php?option=com_content&view=article&id=9&Itemid=18		X	
	GSI Audioscreener http://grason-stadler.us/index.php?option=com_content&view=article&id=11&Itemid=13	X	X	X

Intelligent Hearing Systems www.ihsys.com/site/	SmartScreener–Plus 2 www.ihsys.com/site/SmartScreenerPlus.asp?tab=0	X	X	X
	SmartDPOAE www.ihsys.com/site/SmartDPOAE.asp?tab=2		X	
	SmartTrOAE www.ihsys.com/site/SmartTrOAE.asp?tab=2			X
Interacoustics A/S www.interacoustics.com/com_en/Pages/Frontpage.htm	Otoread www.interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=60998		X	X
	TEOAE25 www.interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=61028			X
	DPOAE20 www.interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=60864&pane=1		X	
	DPOAE440 (Titan Software) www.interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=71112		X	
	Abris www.interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=60808	X		
Otometrics (Madsen) www.otometrics.com/	Accuscreen www.otometrics.com/hearing-assessment/newborn-hearing-screening/MadsenAccuScreen.aspx	X	X	X
Maico Diagnostics www.maico-diagnostic.com/com_en/Menus/Pages/frontpage.htm	Ero-Scan www.maico-diagnostic.com/com_en/Menus/ProductByUser/3-Newborn-Screening/1-Otoacoustic-Emissions		X	X
	MB 11 Classic www.maico-diagnostic.com/com_en/Menus/ProductByUser/3-Newborn-Screening/2-ABR-BERA	X		
	AABR Screener MB 11 BERAphone www.maico-diagnostic.com/com_en/Menus/ProductByUser/3-Newborn-Screening/2-ABR-BERA	X		
Natus Medical Inc. www.natus.com	ALGO 5 www.natus.com/index.cfm?page=products_1&crid=176	X		

	ALGO 3i www.natus.com/index.cfm?page=products_1&crid=176	X		
	Echo-Screen www.natus.com/index.cfm?page=products_1&crid=183	X	X	X
	ABaer www.natus.com/index.cfm?page=products_1&crid=181	X	X	X
	AuDX (AuDX I, AuDX Pro, AuDX II, AuDX Pro II, AuDX Pro Plus, AuDX III) www.natus.com/index.cfm?page=products_1&crid=184		X	X
Otodynamics Ltd. www.otodynamics.com/ www.otodynamics.com/screening.asp	Echocheck		…	X
	Echoport (IL0288) USB-I and USB-II www.otodynamics.com/productSplash.asp?ID=530&sub=14	…	X	X
	Otoport Screener www.otodynamics.com/Product_info.asp?id=215&DisMode=&sub=1&lm=0&pp=0			X
	Otocheck Screener www.otodynamics.com/Otocheck-intro.asp?sub=48		X	X
	Otoport DP&TE www.otodynamics.com/Otoport-DPTE-intro.asp?sub=47		X	X
	Otoport Advanced www.otodynamics.com/Product_info.asp?id=246&sub=49		X	X
Vivosonic, Inc. www.vivosonic.com/	Aurix www.vivosonic.com/en/aurix/index.html	X		
Welch Allyn® www.welchallyn.com/	Welch Allyn OAE Hearing Screener www.welchallyn.com/apps/products/product.jsp?id=11-ac-100-0000000001094		X	

AABR – Automated Auditory Brainstem Response; DPOAE – Distortion Product Otoacoustic Emissions; TEOAE – Transient Evoked Otoacoustic Emissions

Safety

According to some manufacturers and distributors of the AABR and AOAE technologies currently available in North America, these devices are designed, tested, and manufactured to meet North American, European, and/or International Standards for medical electrical equipment, and comply with the Medical Device Directive.[12] The stimuli generated by either technology are not harmful and no issues or concerns have been raised about the safety of either AOAE (DPOAE or TEOAE) or AABR.[12]

Risks associated with newborn hearing screening using any of the AABR and/or AOAE devices available on the market include parental/caregiver anxiety due to false positive results and possible delay of diagnosis and appropriate treatment due to false negative results.[2–4,12,24,32,43] False positive results may also lead to disease labelling, iatrogenesis from unnecessary testing, and increased costs in terms of time and money.

Expert opinion suggests that the newborn/infant, the screener, the equipment, and the environment can all be sources of error when conducting newborn hearing screening.[3] Invalid test results can occur when:

- the newborn/infant is not asleep or settled, is not positioned optimally, or moves during the test
- the screener is unfamiliar with the test equipment and inexperienced at determining whether the test result is valid, is inexperienced at handling newborns/infants, positions the insert probe or couplers inadequately, places electrodes poorly, or allows insufficient time for testing
- the equipment malfunctions, is calibrated incorrectly, or has an occluded probe tip
- the environment has too much ambient noise or causes electrical interference (for example, monitors affect both AABR and AOAE tests)

Manufacturers/distributors of AABR and AOAE technologies emphasize the importance of correct assembly and placement of probes and electrodes/sensors, which is deemed as crucial to the success of screening newborns for hearing loss.[12] If screeners follow the guidelines for sensor and probe assembly and placement, and if they take normal care in handling newborns/infants, no risks should be associated with performing the test itself. Failure to follow guidelines can lead to cross-infection of newborn/screener, and poor probe fit can lead to unnecessarily long testing and the possibility of overly high stimulation. Extreme care is recommended regarding the preparation of the skin for sensor placement.

Regulatory status in Canada

In Canada, the following companies are licensed to market automated testing devices for newborn hearing screening (http://webprod3.hc-sc.gc.ca/mdll-limh/index-eng.jsp, accessed January 10, 2012):

- Grason-Stadler Inc.
- Maico Diagnostics
- Otodynamics Ltd.
- Interacoustics
- Otometrics
- Natus Medical Inc.
- Natus Europe GMBH

- Vivosonic Inc

See Appendix T.D for a list of these companies' devices.

Guidelines and Position Statements on UNHS

The literature search identified no formal guidelines specifically developed on the use of AOAE and/or AABR (alone or in combination) for newborn or infant hearing screening. Neither did it yield any evidence-based clinical practice guidelines identifying best practice for UNHS.

The following section summarizes the recommendations provided by recent position papers.

Recommendations in North America

In the **United States,** multiple professional societies, advocacy groups, and government agencies participating on the Joint Committee on Infant Hearing (JCIH) and other organizations and professional bodies endorsed UNHS (using electrophysiologic measures) as an important component of early detection and intervention for newborns and infants with congenital hearing loss.[7–9,16,47–49] It is recommended that all newborns be screened for congenital hearing loss before 1 month of age. Newborns who do not pass the screen should undergo confirmatory diagnostic testing before 3 months of age. Those with confirmed hearing loss should receive appropriate intervention at no later than 6 months of age from health care and education professionals with expertise in treating hearing loss and deafness in infants and young children. UNHS is widely recognized as the path to earlier identification and the first step in the process to improve developmental outcomes in children with hearing loss, and its effectiveness is dependent on the timely provision of appropriate diagnostic and rehabilitative services.

In 2007 the JCIH issued its updated benchmarks for UNHS in developed countries:[8]

- *Screening coverage.* At least 95% of newborn infants should complete their hearing screening by 1 month of age (age correction for preterm newborns is acceptable).
- *Referral rate.* Less than 4% of newborns should fail their initial screening and any subsequent rescreening before being referred for comprehensive audiological evaluation.
- *Follow-up rate.* Of newborns who fail initial screening and any subsequent rescreening, at least 90% should complete a comprehensive audiological evaluation by 3 months of age.

In its 2000 and 2007 position statements, the JCIH endorsed integrated, interdisciplinary state and national systems of UNHS, evaluation, and family-centered intervention.[8] It is recommended that screening is recommended to be performed during the postpartum hospitalization for most newborns (within 1 month for birth outside of hospitals). It includes AOAE or AABR followed by a repeated or second test for those who do not pass the first test. The 2007 JCIH position statement recommends separate hearing screening protocols for NICU and well-infant nurseries, to identify infants with congenital permanent sensory, conductive, and neural (auditory neuropathy/auditory dyssynchrony) hearing loss. High risk NICU newborns (those admitted for more than 5 days) are to have AABR included as part of their hearing screening so that neural hearing loss will not be missed. For newborns who do not pass AABR testing in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, for comprehensive evaluation, including diagnostic ABR testing, rather than for general outpatient rescreening.

In 2008, the United States Preventive Services Task Force (USPSTF) revised its UNHS recommendations.[7] USPSTF found good evidence that screening newborns for hearing loss is highly accurate and leads to earlier identification and treatment of infants with hearing loss, and recommended UNHS. All newborns should undergo hearing screening before 1 month of age, and those who do not pass the screening should undergo audiologic and medical evaluation before 3 months of age.

According to USPSTF, screening programs should be conducted using a one-step or two-step validated protocol.[7] A frequently used two-step screening process involves AOAE testing followed by AABR testing in newborns who do not pass the first test. Infants with positive screening test results should receive appropriate audiologic evaluation and follow-up after discharge. Equipment should be well maintained, staff should be thoroughly trained, and quality-control programs should be in place to reduce avoidable false positive test results. For newborns delivered at home, at birthing centers, or in hospitals without hearing screening facilities, a referral mechanism should be put in place for hearing screening, including follow-up tracking.

According to the JCIH, hearing screening should identify newborns at risk for specifically defined hearing loss that interferes with development.[8] The aim for UNHS programs is detection of permanent sensory or conductive hearing loss averaging 30 to 40dB or more in the frequency region important for speech recognition (approximately 500–4000 Hz).[7,8] The focus of UNHS is on congenital as opposed to acquired or progressive hearing loss that may not be detected in the newborn period.

The American Academy of Pediatrics Task Force on Newborn and Infant Hearing, the National Institute on Deafness and Other Communication Disorders, and the Centers for Disease Control and Prevention Early Hearing Detection and Intervention Program support the JCIH recommendations (www.cdc.gov/newbornscreening).[7]

The American Speech-Language-Hearing Association (ASHA) recommends that all hearing screening programs be conducted under the supervision of an audiologist holding the ASHA Certificate of Clinical Competence.[12] The American Academy of Audiology Task Force on the Early Identification of Hearing Loss agrees that the use of support personnel in newborn hearing screening programs is an appropriate and often necessary strategy for achieving universal detection of congenital hearing loss.[7] The supervising audiologist should be experienced in both the development and maintenance of a UNHS program, including an understanding of technology options.

In **Canada,** national and provincial organizations support the JCIH position statements and recommendations.[1,6,11,30,31] These organizations also recommend the establishment of a well-integrated and structured system of early identification and management for all infants with hearing loss, tailored to the unique geographic, demographic, cultural, and political features of Canada. None recommend a specific screening test or protocol.

In their 2008 position statement on UNHS, the Alberta College of Speech–Language Pathologists and Audiologists (ACSLPA) endorses UNHS and strongly supports the establishment and maintenance of well-integrated and structured systems for the early identification and management for all newborns/infants with hearing loss in Alberta.[1] The goal is for all infants with a permanent

bilateral or unilateral sensory or conductive hearing loss to be identified, diagnosed, and provided with adequate audiological, medical, technological, and behavioural follow-up as early as possible.

In 2010, the Canadian Association of Speech–Language Pathologists and Audiologists (CASLPA) submitted a position paper on UNHS as part of a national campaign to improve early identification of hearing loss in children in Canada.[11] CASLPA endorses UNHS as a strategy for identifying children with PCHI and for initiating family-centred audiological and communication intervention. According to CASLPA, the UNHS, using non-invasive testing devices, would ideally be performed before a newborn leaves the hospital. The organization called for a Canada-wide adoption of UNHS programs in all provinces and territories. The goal of these programs is for all children with PCHI to be identified and provided with comprehensive, family-centred, early intervention.

According to these position papers, a well-integrated and structured system of early identification and management for all newborns/infants with hearing loss, which enables identification of hearing loss by 1 month of age, confirmation of hearing loss by 3 months of age, and enrolment in a family-centered intervention program by 6 months of age should include:[1,11]

- universal screening (using electrophysiological methods) of all newborns
- appropriate, accessible services for diagnosis, hearing, and communication development options
- a seamless transition for infants and families through the process of screening by 1 month of age, a confirmed diagnosis by 3 months of age, and initiation of early intervention by 6 months of age
- ongoing surveillance throughout infancy and early childhood of those children at risk for developing hearing loss
- education for parents, primary caregivers and healthcare providers about the early signs of hearing impairment and the risk factors associated with a hearing loss
- early intervention with an assigned point of entry and intervention options
- multidisciplinary teams of professionals who work closely with families
- continuing education opportunities for multidisciplinary/interprofessional teams to achieve and maintain expertise in screening, assessment, the fitting of amplification in infants, and parent-infant habilitation strategies
- implementation of provincial/territorial registries for each program, which could be integrated with interprovincial and territorial registries and a national program database; this data management aspect of the system is critical for assessing and monitoring the quality, efficiency, and effectiveness of the screening, evaluation, and intervention processes and to ensure the program is stable, sustainable, and conforms to established program benchmarks and quality indicators

In 2011, the Canadian Paediatric Society (CPS) published a position statement on UNHS that was based on a systematic review of the literature.[6] Based on the available evidence, CPS 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. According to CPS, several Canadian provinces, including Ontario and British Columbia, offer excellent examples of integrated systems.

Recommendations in other countries

The Newborn Hearing Screening Programme (NHSP) in the United Kingdom has recently developed standard care pathways, which summarize its guidelines.[25] According to the care pathways, the goal is to screen for hearing loss in newborns; the target condition for the screen is bilateral, permanent hearing loss (sensorineural or permanent conductive) averaging 40 dB or more in the better ear. Screening may be attempted up to age 3 months (corrected age); infants older than 3 months (corrected age) should be considered for referral to an audiology at an appropriate age.

A multistage screening protocol is used, with AOAE and AABR.[25] There are two versions of the protocol: one for well newborns and the other for newborns who have been in NICU/SCBU. The protocols are different because NICU/SCBU babies have a higher risk of auditory neuropathy, auditory dssynchrony, or other neurological problems that are more likely to be picked up by AABR rather than AOAE testing. NHSP uses the terms "clear response" and "no clear response" rather than "pass" and "fail", as the former are more family friendly.

The well-baby and NICU/SCBU protocols cover both hearing screening in hospitals and community-based services.[25] Well-baby screening in hospital-based services is usually carried out by screeners specifically trained and employed to carry out hearing screening, with the aim of completing screening by the age of 4 weeks or prior to discharge from hospital. If the process is not completed in hospital, an outpatient appointment, clinic appointment, or home visit is required to complete the process, usually within one visit. It is recommended that screening not be performed on newborns with a gestational age of less than 34 weeks. Community-based screening is usually carried out by health visitors. The first screening test usually takes place during the visit of the primary health visitor when the infant is approximately ten days old. Any subsequent testing should be completed by the time the infant is 5 weeks old.

NICU/SCBU screening in hospital-based services is performed by one of the hearing screeners. In community-based services it may be performed by various people, including trained NICU nurses.[25] The aim is to complete screening by 44 weeks gestational age (4 weeks corrected age), as close to discharge as possible while the newborn is in hospital. If the process is not completed in hospital, an outpatient or clinic appointment or home visit is required to complete the process, usually within one visit. Screening is not recommended for babies less than 34 weeks gestational.

The tests used for both well-baby and NICU/SCBU screening are AOAE and AABR performed on both ears (unless considered inappropriate).[25] AOAE is usually performed first, followed by AABR where indicated by the pathway. AABR may be considered inappropriate if a baby has a skin condition that makes it medically inadvisable to attach electrodes. For well-baby screening, two AOAE attempts can be carried out if necessary, followed by AABR performed on both ears regardless of AOAE results. An AABR "no clear response" result in one or both ears is referred to audiology for early audiological assessment. For NICU/SCBU only one AOAE attempt is carried out.

NHSP policy for missed and incomplete screens is that:[25]

- infants under the age of 3 months (corrected age) should be offered an appointment to complete the screen from whichever stage (AOAE or AABR) had been previously reached
- infants over the age of 3 months should be considered for referral to audiology at an appropriate age
- in most cases, the referral for behavioural testing will be at the age of 7 to 12 months
- in the event of parental or professional concern, an earlier appointment may be required, using whatever methods are appropriate and possible

The guidelines developed recently by the Commission for the Early Detection of Hypoacusis (CODEPEH) in Spain provide recommendations for early hearing detection and intervention (EHDI) programs.[38] According to COPEDEH recommendations, screening should be performed using either TEOAE and/or AABR before the first month, diagnosis at 3 months, and treatment at 6 months. Separate protocols are recommended for infants from NICU and those from maternity. Infants with no history or risk of retrocochlear hearing loss may be tested in the screening phase by either TEOAE or AABR. If the AABR screen is not passed, a second test would not be needed and the infant could be referred for diagnostic confirmation. However, if the TEOAE is used, especially if it's performed before the child is more than 72 hours old, the test should be repeated at least once before referral to the diagnostic phase.

In TEOAE-based screening programs, infants with risk factors for retrocochlear hearing loss must be subjected to a complementary test by AABR or by Auditory Brainstem Evoked Potentials (ABEP) for diagnosis even if they have passed the TEOAE, in order to avoid the false negatives associated with the existence of auditory neuropathy.[38] For the same reason, in infants screened by AABR, the TEOAE should be applied jointly in cases where the first test is not passed, to document the existence of a possible auditory neuropathy. Ongoing monitoring of children is important, even if they have passed the screen in the neonatal period. For infants with risk factors associated with hearing loss, the timing and number of auditory re-evaluations should be adapted and individualised depending on the factor identified.

Newborn Hearing Screening in Canada

Since 2007, interest has been growing in the early detection, diagnosis, and management of PCHI, which has been identified as an important public health issue in Canada.[1,6,10,11,30,31,34] Currently, however, there is no systematic approach to this issue in every Canadian province. No coordinated national approach to UNHS is in place, and in many cases no dedicated funding exists.

UNHS is now offered in British Columbia, Yukon, Nunavut, the Northwest Territories, Manitoba, Ontario, Quebec, New Brunswick, Nova Scotia, Saskatchewan, and Prince Edward Island. (www.albertahealthservices.ca/facilities.asp?pid=saf&rid=1047787; www.nshsc.ns.ca/babyhearing.html; web2.gov.mb.ca/bills/39-5/b215e.php; www.children.gov.on.ca/htdocs/English/topics/earlychildhood/hearing/brochure_hear.aspx).[1,6,10,30,34,50-54] Some provinces—such as Ontario, British Columbia, Nova Scotia, and New Brunswick—have offered newborn hearing screening for some time, while other provinces, such as Alberta, still

have no universal programs. Ontario and British Columbia have fully funded provincial programs, while other provinces have partial programs, primarily targeting infants in the NICU.[6] In some areas without UNHS programs, no equipment or expertise is available to perform newborn hearing screening at birthing hospitals.[34]

It has been reported that, as of 2009, over two-thirds of Canada has implemented early detection of hearing loss programs.[10] The various programs mirror the JCIH recommendations and aim to screen all neonates by 1 month, confirm diagnosis by 3 months, and initiate appropriate intervention by 6 months.[6,10,50-52] Reported problems have focused on the need to provide screening coverage 7 days a week (because some newborns leave the hospital over the weekend), the placing of UNHS into an overall program that includes screening for other congenital problems, and the need for central oversight at the provincial level as opposed to fragmented control by local hospitals and clinics.[10,51] Loss to follow-up is the largest limiting factor of UNHS in Canada.[1]

The British Columbia Early Hearing Program (BCEHP) is a province-wide program for early hearing screening and intervention.[51,52,54] By early 2009, implementation of screening in birthing hospitals and community sites was complete in all health authorities with the exception of Northern Health. Rural and remote communities may have other screening access arranged for smaller birthing hospitals. Most newborn hearing screens are done before the infant leaves hospital. If babies have not had screening completed by the time of discharge, the family will be offered a follow-up appointment, usually at the local public health audiology clinic. Screening may be offered at other community sites in each health authority. Screening is a two-stage process (AOAE testing first, followed by AABR testing) that is completed by BCEHP-trained personnel using standardized equipment and following provincial protocols.

Since its implementation in 2002, the Ontario Infant Hearing Program (IHP) has offered hearing screening to newborns in all birthing hospitals (www.children.gov.on.ca/htdocs/English/topics/earlychildhood/hearing/brochure_hear.aspx).[1,50] The IHP provides UNHS, surveillance for those at risk for developing hearing impairment in early childhood, audiology assessment, hearing aid selection, follow-up audiology visits, and communication/language development services for children identified with permanent hearing loss, until Grade 1 entry. All newborns in Ontario can have their hearing screened, either in the hospital when they are born or at a community screening clinic. There is no charge for the screening. The first screening, using a DPOAE device, is followed by AABR testing. Newborns with a "refer" result from the AABR screen will be offered further AABR testing 1 to 2 weeks after they go home from the hospital (to allow any fluid or wax in the ears to clear).

Information regarding the status of newborn hearing screening in the other Canadian provinces and territories was obtained from the 2008 ACSLPA position statement[1] and the 2011 CPS position statement.[6] Although Quebec confirmed funding for UNHS in July 2009, as of 2011 UNHS had not been implemented.[6] UNHS has operated in each of the New Brunswick health authorities since 2002.[1] The provincial strategy in Nova Scotia includes access to UNHS for all newborns in birthing hospitals. The two hospitals that provide obstetrical service in Prince Edward Island have provided UNHS programs since 2005. As of 2008, UNHS existed in eight of the 11 birthing hospitals in Newfoundland and Labrador, and three other hospitals performed high-risk screenings with the intention of implementing UNHS in the near future. UNHS has been provided since 2004 for newborns in Yellowknife and Inuvik, with a screening goal of 95%. UNHS has also been provided

in Whitehorse since 2002; the program is a joint project by Health and Social Services Hearing Services and the Whitehorse General Hospital. Nunavut expectant mothers are flown to the nearest province or territory.

Two of the four UNHS programs (in former Palliser and Peace Country Health Regions in Alberta) that participated in a 2001–2004 pilot project were maintained after the cessation of project funding in 2004.[1] UNHS services are also provided to all newborns in the Medicine Hat, Brooks, Bow Island, and Oyen areas (www.albertahealthservices.ca/facilities.asp?pid=saf&rid=1047787). Audiology assistants are notified of hospital births and provide hearing screens while the newborn is in hospital. A registered audiologist provides a more thorough assessment if the newborn does not pass the hearing screening. If the newborn is discharged before undergoing hearing screening, the family is called to see whether they wish to book an outpatient hearing screening at any of the Community Health Office sites (for example, Medicine Hat Regional Hospital audiology department, Brooks Community Health office, Bow Island Community Health and Oyen Community Health). Approximately 80% to 85% of newborns are screened prior to discharge from the hospital.

There is no provincial registry of PCHI cases in Alberta, nor is there a national registry (Howarth, personal communication, January 13, 2012). No standardized protocol is used for newborn hearing screening across Alberta, nor do national guidelines exist in Canada. Ontario and British Columbia have well developed, standardized, evidenced-based practices and protocols for universal newborn and infant hearing screening. The various sites in Alberta that conduct newborn hearing screening have purchased equipment independently, and consequently the equipment is not standard and the testing protocols employed vary. For hearing screening of newborns and infants (under the age of 6 months) any one of the following might be employed in Alberta: diagnostic ABR equipment with a "screening protocol", AABR equipment, or AOAE (TEOAE or DPOAE) using either a true screening AOAE device or a diagnostic AOAE device set up with a "screening" protocol. The screening may be a one- or two-stage process (with variations in the definitions of what the one and two stages are). The equipment in use in the province has not undergone a rigorous examination as to its merits for screening hearing. While the equipment that is in use has been purchased under the expert guidance of local public health audiologists at each of the sites—and for this reason is appropriate equipment—a coordinated, systematic, standardized approach has not occurred to evaluate which equipment and which parameters are optimal for use in newborn and infant hearing screening.

Available Research Evidence

A comprehensive literature search was conducted to identify the most recently conducted systematic reviews (SRs) or health technology assessment (HTA) studies that examined the research evidence about the safety and efficacy/effectiveness of using automated testing devices in UNHS programs for the detection of PCHL. Only published reports of SRs and HTA studies that, by virtue of design and quality of reporting were most likely to provide the best level of evidence, were selected for data extraction. Individual primary research studies (of any design) published subsequent to the selected SRs and HTA studies are not included. See Appendix T.A for a detailed description of the literature search strategy and study selection process.

The literature search conducted for this review identified 1369 citations. After screening of titles and abstracts, 1264 citations were excluded from the final selection process. The full text of 105 potentially relevant articles was retrieved and further evaluated for inclusion in the review. The application of the selection criteria to 13 full-text articles retrieved as potentially relevant research studies resulted in eight being excluded (see Table T.B.1, Appendix T.B for the main reasons for their exclusions).

Figure 1 in Appendix T.A outlines the research study selection process for this review.

Three SRs[3,32,43] were selected for the review. Two other SRs[2,4] were identified as multiple publications of the two selected SRs.[32,43] Although the multiple publications[2,4] were not included as unique studies, any relevant information that the authors provided was included when appropriate.

All selected studies[2-4,32,43] addressed questions on the benefits and harms of UNHS (programs) for PCHL detection, as compared to selective/targeted screening or no screening in terms of patient-relevant outcomes. Two systematic reviews also addressed the diagnostic accuracy of the automated testing devices, used alone or in combination (AOAE and/or AABR), in UNHS.[3,4,32]

All selected studies[2-4,32,43] fulfilled the criteria for a systematic review by:

- posing a clear question *a priori*
- identifying the relevant literature
- extracting the data and assessing the methodological quality or risk of bias of the primary research in a reproducible fashion
- qualitatively or quantitatively summarizing and analyzing the reviewed evidence
- exploring the sources of variation in the results from study to study

Objectives and selection criteria varied across the three SRs and there was little overlap among their included primary research studies. The only papers included in all selected SRs were those reporting results from the large, non-randomized controlled trial of UNHS for early identification of PCHL conducted by the Wessex Universal Neonatal Screening Trial Group (published in 1998, 1999, and 2000),[55-57] an 8-year follow-up of this Wessex study (published in 2005),[58] and a community-based cohort study conducted by Kennedy and colleagues (published in 2006).[59] These studies were considered by all SRs to be of good methodological quality.

The following commentary summarizes the findings about the safety and efficacy/effectiveness of using automated testing devices to detect PCHL in UNHS as reported by the selected SRs. See Appendix T.C (Tables T.C.1 and T.C.2) for details from the selected systematic reviews.

Efficacy/effectiveness

Wolff et al.[32] reported the results from an update of a systematic review commissioned by the German Institute for Quality and Efficiency in Health Care (IQWiG)[4] to evaluate the benefits and harms of UNHS programs in the detection of hearing impairment in childhood. In the absence of randomized controlled trials (RCTs) evaluating screening programs, the IQWiG study and its update[4,32] consisted of three systematic reviews of non-randomized, controlled studies of diagnostic

accuracy of screening tests, benefits and harms of screening versus no screening, and the therapeutic effect of early versus later treatment (Table T.C.2).

The IQWiG systematic review and its update included nine studies (reported in 12 publications) that looked at the diagnostic accuracy of automated testing used in UNHS (see Table T.C.1).[4,32] Eight cross-sectional studies (seven of which were published before 2000 and one of which was published in 2000) investigated the diagnostic accuracy of TEOAE as compared with AABR. One controlled study evaluated a two-stage screening UNHS protocol (TEOAE followed by AABR). The age at screening in these nine studies varied between 15 and 120 hours from birth. Most studies providing information on acoustic conditions reported performing the test in a quiet room or a room with sound insulation. The threshold in all studies was 35 to 40dB. Study quality was generally poor for items such as the appropriate sample size, blinded assessment of outcome parameters, the consideration of confounding factors, and the documentation of uninterpretable tests or tests that were not performed. From the available information, it appears that none of these studies evaluated any of the AOAE or AABR devices that are currently licensed in Canada.

The eight cross-sectional studies comparing TEOAE with AABR (using various older models of TEOAE and AABR devices) reported sensitivities of OAE measurement ranging from 50% to 100% and specificities ranging from 49% to 97%.[4,32] With one exception, between 105 and 500 neonates were tested as inpatients in a hospital, usually a university hospital. In only half of these studies were sensitivity and specificity calculated on the basis of the number of neonates investigated. In the other studies, results were reported relative to the number of ears investigated. A specific problem with these studies was that AABR was used as reference standard, although this is not suitable as the definite "gold standard".

Data on the diagnostic accuracy of a two-stage UNHS protocol was obtained from the Wessex study and its 8-year follow-up study.[4,32] The Wessex study included newborns (average- and high-risk) from four hospitals in the United Kingdom (25,609 born during periods of UNHS and 28,172 not screened as newborns). TEOAE testing (using the ILO88 device) was followed, if this test was failed, by AABR testing (no information was available about the model of AABR device, who performed the testing/qualification, or the environmental or acoustic conditions). Most infants were screened within 48 hours of birth; infants in NICU/SCBU were screened at the end of their hospital stays. Comprehensive audiological clarification was planned for weeks 6 to 12 of life for the children who still had abnormal results. The actual comparison for the question of screening was performed at the age of 8 months with the Health Visitor Distraction Test (HVDT) also known as the Infant Distraction Test Screen (IDTS). In addition, diagnostic audiological testing was performed after about 8 years (results were published in the 8-year follow-up article) at all institutions in the region treating children with hearing impairment. The 8-year follow-up focused on the effect of UNHS on the proportion of all true cases of PCHL ≥ 40dB HL that had early referral.

Wolff et al.[32] mentioned that because there was no actual follow-up of the screen-negative children in the Wessex study, it was assumed that identification of at least a portion of children with a false negative test result was guaranteed.[32] Based on this assumption, the estimated sensitivity of the two-stage screening was 91.7% (95% confidence interval [CI] 74.2% to 97.7%), the specificity 98.5% (95% CI 98.3% to 98.7%). The positive predictive value was 22 out of 342 children (6.5%). If these results are transferred to 100,000 newborns, about 110 of 120 children with hearing impairment would be positively identified (sensitivity of 91.7%) and screening programs would lead to false

suspicions of hearing impairment in about 1500 children (specificity of 98.5%). If the children not participating in the screening were included (intention-to-screen), program sensitivity could be calculated as 71.0 (95% CI 52.0 to 85.8). This means that approximately 30% of the children with hearing impairment are not identified by the program. The risk of a hearing impairment is slightly increased for children who did not participate in the screening or whose parents rejected participation (1.6 per 1000 versus 1.1 per 1000; p = 0.344).

Based on the reviewed evidence, Wolff et al.[32] concluded that an overall reliable evaluation for the diagnostic accuracy of AOAEs and AABR as initial screening tests was not possible because there has been no evaluation of these tests in an adequately large group of children without risk factors. Although the results from the Wessex study and its 8-year follow-up study indicate that a two-stage protocol (first TEOAE and then, if the finding is abnormal, AABR) in practical use might achieve acceptable sensitivity of >90% with specificity of >98%. These estimates must be confirmed as they are based on a relatively small number of children with hearing impairments and the 95% CI for sensitivity extends from 74% to 98%. Wolff et al.[32] also stated that "it must be considered that the proportion of unidentified children markedly increases if the children not participating in screening are included in the evaluation (intention-to-screen analysis)".

The IQWiG systematic review and its update included two comparative studies (with 120 and 50 children with hearing impairment) in the evaluation of benefits and harms of screening versus no screening in terms of patient-relevant outcomes.[4,32] One is the cohort study conducted by Kennedy et al.[59] that prospectively compared alternating screening periods, with and without UNHS, (subpopulation I, including only children from the Wessex study), and also compared hospitals with UNHS versus those without UNHS programs (subpopulation II, including children from four districts in Greater London). The other study (conducted by Yoshinaga-Itano and colleagues)[60,61] retrospectively investigated 50 children with hearing impairment who had been born in hospitals either with or without UNHS. Both studies showed deficiencies regarding study and publication quality. The reviewers found no studies that compared UNHS with screening of only at-risk children (selective/targeted screening).[4]

Both selected comparative studies contained results for language development, general communicative ability, and spontaneous speech.[4,32] Taken together, the results indicate that children with hearing impairment identified by UNHS are at an advantage with regard to language development at (average) ages of 3 years and 8 years compared with children whose hearing impairment was identified outside a specific screening program or in a screening program performed at a later age. The chances of normal speech development appear to be higher for screened children, possibly due to earlier diagnostic clarification in these children. Data on other potential long-term patient-relevant outcomes were not available (for example, quality of life, mental health, satisfaction, educational and professional development). Based on the reviewed evidence, Wolff et al.[32] concluded that "[e]arly identification and early treatment of children with hearing impairments may be associated with advantages in language development. Other patient-relevant parameters, such as social aspects, quality of life, and educational development, have not been adequately investigated".

Nelson et al.[43] conducted a systematic review to update the 2001 USPSTF recommendations on UNHS used to detect moderate-to-severe, permanent, bilateral, congenital hearing loss.[2,43] The focus was on three key questions addressing the benefits and harms of UNHS when compared to targeted screening or no screening, in terms of patient-relevant outcomes (see Table T.C.2). Nelson et al. did

not update the additional key questions that were included in the previous review, such as sensitivity and specificity of testing procedures, because they considered that these were adequately addressed by existing evidence.[2,43] However, the reviewers included some of the diagnostic accuracy data reported for the screening protocol used in the Wessex study and its 8-year follow-up study, which had a false positive rate of 1.5% and a false negative rate of 4%. The yield of screening was estimated at 90 cases of PCHL at 40dB hearing level or more per 100,000 target population (equivalent to 80% of expected prevalence in the population).

Nelson et al.[2,43] found no randomized controlled trials or trials that directly compared targeted screening with UNHS, and reported data about the initiation of early treatment for infants at average risk or those at high risk. To address question # 1, the reviewers included the cohort study by Kennedy et al.[59] that evaluated the effect of UNHS on the speech and language outcomes of 120 children with hearing impairment. In this study, 67% of children undergoing UNHS had confirmation of hearing impairment by the age of 9 months compared to 27% of those not undergoing UNHS. The results indicated that those children who had early versus late confirmation, and those who had UNHS versus no screening, had better receptive language outcomes at 8 years of age but not better expressive language or speech (Table T.C.1). Children with hearing impairment confirmed by 9 months of age had significantly better age-adjusted scores than those confirmed later, on two tests of receptive language and one of two tests of expressive language but not on the speech scale. All of the aggregate scores for receptive and expressive language were significantly better for the early confirmation group. In contrast, a fair-quality, community-based cohort study of children with hearing impairment who did not undergo UNHS indicated no relationship between age at diagnosis and language, speech, and reading measures at age 7 to 8 years. However, few children were diagnosed by age of 6 months in this study (conducted in Australia).

To address question # 2, Nelson et al.[2,43] included the Wessex study and its 8-year follow-up study, as well as six descriptive studies that reported relevant follow-up data from UNHS programs (see Table T.C.1). Results indicate that average- and high-risk infants with PCHL born in hospitals with UNHS have earlier referral and initiation of treatment than those born in hospitals without UNHS. In the analysis from the 8-year follow-up study, one additional case of PCHL was referred before the age of 6 months for every 1969 (1011–12,896) infants in the UNHS population. More children with true PCHL were referred to audiology services prior to age of 6 months if they were born during periods with UNHS than during periods without (74% versus 31%; difference 43%; 95% CI 19 to 60%; p=0.001). Adjustment for the effect of severity of hearing impairment on age of referral increased the odds ratio between newborn screening and early referral from 6.3 to 6.9 (2.2–22.0; p=0.001). The percentage of all true cases referred was greater at any given age during the first 3 years for children screened as newborns versus not (percentages were similar after age of 3 years). The age at referral was lower for children undergoing UNHS when compared to those not screened (zero months versus 8 months; $p < 0.001$). It was noted that eight children with hearing impairment had screened negative in infancy and seven had had documented progression in severity after detection in infancy. The sum of these two figures represents 23% of all cases that might have had progressive losses if the eight negative screens in infancy had been an accurate reflection of the hearing status of the child at that time.

Although Nelson et al.[2,43] found no studies that directly compared the yields of universal versus targeted screening approaches, they determined some estimates by applying results from relevant studies. Assumptions for the model included the proportion of newborns considered high risk, the

prevalence of PCHL in high-risk and average-risk populations, the proportion not screened in the hospital, the sensitivity of two-stage screening, compliance with follow up testing (estimated), accuracy of diagnostic tests, and the proportion of average-risk newborns diagnosed with PCHL by age 3 months (estimated). Using these assumptions, if 10,000 newborns underwent UNHS, there would be 11 to 12 diagnosed cases by the age of 3 months, 86 false positive screening tests, and possibly one missed case. The number needed to screen (NNS) in order to diagnose one case would be 878. If only high-risk newborns underwent screening, there would be four or five diagnosed cases, six false positive screening tests, and eight or nine missed cases. The NNS in order to diagnose one case would be 178.

According to Nelson et al.,[2,43] their findings "indicate that infants identified with PCHL through UNHS have significantly earlier referral, diagnosis, and treatment than those identified in other ways. Although the clinical community has acknowledged the significance of early treatment for many years, evidence of its effect on long-term functional outcomes has been limited. New data on improved language outcomes at school age strengthen the case for UNHS, but are also dependent on effective methods of referral, follow-up, and treatment. As these needs are being addressed with ongoing projects, further research will be required to demonstrate effectiveness for the entire process that UNHS initiates".

Recently, the Adelaide Health Technology Assessment Program was commissioned by the Medical Services Advisory Committee (MSAC) to conduct an update of a systematic review conducted in 2003 on the safety, effectiveness, and cost effectiveness of UNHS.[3] The research questions addressed the diagnostic accuracy of the screening tests used to detect PCHL in neonates or infants, the impact of UNHS on the clinical management or treatment options available to permanently hearing-impaired infants, and the impact of UNHS on the adverse outcomes associated with PCHL in terms of patient-relevant outcomes (see Table T.C.2).

The MSAC assessment included five comparative studies (evaluated as level III-2 diagnostic evidence, average quality) to address the question on the diagnostic accuracy of screening tests.[3] Three of these studies compared TEOAE with a conventional ABR test (no information is provided on what devices were used). One study compared the accuracy of TEOAE to tympanometry (no information is provided on what TEOAE device was used) and one study compared the AABR test (using the "earliest" model of AABR test) with a conventional ABR. Reviewers found no studies that compared the DPOAE test with a relevant reference standard. From the available information, it appears that none of these studies (four of which were published before 2000 and one of which was published in 2002) evaluated any of the AOAE or AABR devices currently licensed in Canada.

The ability of initial TEOAE testing in a one-stage screening protocol to accurately identify PCHL in neonates and infants varied widely in the included studies, with sensitivity ranging from 50% to 100% when compared to conventional ABR.[3] In terms of identifying conductive hearing loss, TEOAE testing was found to have 100% sensitivity and specificity, as compared to tympanometry in one study that included infants with no cerumen occlusion in their ears. The accuracy of TEOAE appears to depend on the level of local ambient noise (and therefore ear–probe fit and the testing environment), as well as the condition of the infant's ears (for example, whether occluded by vernix or wax) at testing. If these factors are addressed adequately, diagnostic accuracy of TEOAE is very good (up to 100% sensitivity), although even under quiet conditions the rate of false positives can still be quite high (8%). The positive predictive value of an initial TEOAE testing performed under

quiet conditions is very low (1.5%). This is probably a consequence of the frequency of transient hearing losses (ear occlusion) in newborns, as well as the low prevalence of PCHL in the general population.

The ability of AABR testing (using the "earliest" model of AABR devices in one-stage screening protocols) to accurately identify PCHI in neonates and infants was compared to conventional ABR testing in one study (published in 1998) that cross-classified infants on the two tests.[3] The reported specificity and sensitivity values for AABR testing were 96% and 80%, respectively. In this study on the earliest version of AABR test, the positive predictive value is still very low (2.2%), although marginally better than TEOAE testing conducted under quiet conditions.

The MSAC assessment included five controlled studies to address the question on the impact of a UNHS program on clinical management of hearing-impaired infants.[3] Two of these studies were the 8-year follow-up of the Wessex study and the community-based cohort study conducted by Kennedy et al.[59] The remaining four studies analyzed, retrospectively, cohorts of children with hearing impairment, and assessed whether UNHS affected the time of PCHL diagnosis and the age at which management was initiated. Given the paucity of controlled trials available, the MSAC assessment[3] also collated information from uncontrolled trials of screening programs to provide descriptive supplementary data.

Based on the evidence provided by these studies, the MSAC assessment[3] determined that referral for confirmatory diagnostic testing and management of PCHL commonly occurs earlier and more frequently with UNHS than without. According to the MSAC assessment, the 8-year follow up in the Wessex study provided the highest level of evidence available (level III-1 screening evidence) indicating that referring an infant for diagnostic testing before the age of 6 months is nearly three times more likely [RR=2.9, 95% CI 1.4 to 6.3] (19 times when controlling for the severity of hearing impairment) with than without UNHS. Infants born during periods of UNHS availability are twice as likely to receive a diagnosis of PCHL than infants born in periods without UNHS [RR=2.3, 95% CI 1.1 to 4.7]. The absolute increase in benefit is small, however (an extra five children identified per 10,000) because of the low prevalence of the condition. There is also an indication that screening may increase the likelihood of PCHL management before the age of 10 months by nearly two-and-a-half times [RR=2.4, 95% CI 1.0 to 5.8] (eight times when controlling for the severity of PCHL). Similar results were reported in studies with a lower level of evidence (level III-2 screening evidence).

According to the MSAC review,[3] descriptive data indicated that the majority of UNHS programs manage to screen over 90% of infants in their catchment areas. These programs are largely hospital-based, with initial screening occurring prior to discharge. Community-based studies also obtain very good coverage when screening is "piggy-backed" onto other health or immunization checks at the health clinic or when it occurs in the home. Losses to follow-up commonly occur when there is a long delay prior to re-screening or diagnostic testing of the infant, or when infants and mothers are discharged early from the hospital. Data from uncontrolled studies suggest that given the higher referral rate from TEOAE screening protocols, the number of false alarms associated with these programs is higher (up to approximately 10%) than with programs using AABR screening protocols (up to approximately 6%).

The MSAC review[3] found that there is limited information available about the effect of UNHS on primary or patient-relevant outcomes. Two good quality cohort studies (level III-2 screening

evidence) were identified that assessed the impact of screening on language acquisition and communication ability. These were the prospective cohort study by Kennedy et al.[59] and the retrospective cohort study conducted by Yoshinaga-Itano and colleagues.[60] As with the other two systematic reviews,[2,4,32,43] the MSAC assessment[3] did not find information on impact of UNHS programs on the longer-term outcomes (such as educational and employment status) and stated that "it is unlikely to be reported in the peer-reviewed literature for another decade or so".

Based on the reviewed evidence, the MSAC report concluded that "at this time the effect of UNHS on primary or patient-relevant outcomes is not entirely clear. Two cohort studies (level III-2 screening evidence; one cohort in this study used a two-stage protocol TEOAE followed by AABR) were available that measured linguistic and communicative abilities quite differently. From the evidence, language acquisition improvements are seen with UNHS for receptive language but unclear findings have been reported for expressive language. Children identified with PCHI through UNHS appear to have improved communicative abilities compared to those identified without UNHS, according to the small study which used the most sensitive form of measurement (direct and blinded observation of children)".

Safety

None of the selected systematic reviews[2-4,32,43] reported on safety issues or concerns associated with using AABR and/or AOAE technology for UNHS in terms of side effects and complications to the newborn and/or to the screener due to performing the test itself (see Table T.C.1). The primary research studies they included provided no indications of direct negative consequences or physical adverse effects from performing the screening test. Because both the AOAE and AABR test procedures are non-invasive, reviewers considered that direct harm seems to be limited.[3,4,32]

The frequency and severity of the possible psychosocial (indirect) harms from newborn hearing screening were not systematically investigated in the studies included in the IQWiG systematic review and its update.[4,32] According to the reviewers, the "very limited" available data about the development of hearing ability, the mother's anxiety, and the effects on the mother–child relationship could hardly be interpreted because of the unclear selection mechanisms and the lack of control groups (without screening). They noted however, that the potential of indirect harms from screening findings exists, particularly from false positive findings, which can increase parental anxiety. The frequency of these harms depends primarily on the quality regulations and quality assurance measures in a screening program. The extent of parental anxiety caused by false results depends on the type of education and support, as well the quality of the program. In unfavourable cases, a false positive result could lead to the "over-treatment" of a child with normal hearing.

To address the question of the adverse effects of UNHS, Nelson et al.[2,43] included two fair-quality cohort studies (one of which is a subset of the Wessex study), one poor-quality case-control study, and five survey studies with greater than 40% response rates that provided relevant information on the adverse effects of newborn hearing screening. The limited evidence from these studies indicates that usual parental reactions to an initial non-pass on a hearing screen include worry, questioning, and distress. These negative emotions resolve for most parents when a diagnostic test is provided with a normal result. No studies addressed the adverse effects of delaying screening.

The MSAC assessment[3] included five controlled studies of poor to average quality to address the question regarding the safety of UNHS (see Table T.C.1). The main outcomes reported were

parental anxiety concerning the screening, a false positive result (with a large consideration given to the high false alarm rate), and a positive result. Levels of moderate to severe anxiety were predominantly low in all three groups of outcome. There were no clinically significant differences in anxiety levels between parents of babies with positive screens and parents of babies with negative results, or between parents of babies with positive results and parents of unscreened babies (level III-2 interventional evidence). No clinically significant differences were found between levels of anxiety or worry in mothers of infants screened by UNHS compared to mothers of infants aged 6 months or older screened by a behavioural test. More satisfaction was expressed after UNHS than after the distraction test. No studies reported on the psychosocial effects of false reassurance or of a true positive diagnosis.

DISCUSSION

During the last 5 years, the ability to screen infants using UNHS programs has progressed at different levels in various countries. The development and maintenance of UNHS programs that enable early identification and intervention for all infants who have hearing loss as part of a well-integrated and structured system is currently supported by a broad consensus of professional opinion.[1-8,10,11,16,20,25,38,40,47-49,62] The main goal of an ideal UNHS program is that all newborns should have been screened for PCHL before 1 month of age, preferably before hospital discharge. All infants whose screening tests are positive for hearing impairment should undergo confirmatory diagnostic testing as soon as possible (recommended before 3 months of age) and then receive the appropriate family-centered intervention (recommended before 6 months of age). To be effective, UNHS programs must have a high coverage rate, high sensitivity and specificity, and proper tracking, with a low rate of loss to follow-up.

Although newborn hearing screening is generally well accepted and tolerated by parents, coverage and follow-up rates are the major inhibitors to the success of UNHS programs in many countries.[3-5,8,10,13,20,24,36,39,40,44,62,63] Loss to follow-up is still a major limiting factor for program sensitivity. According to the available evidence, UNHS safety and clinical efficacy has yet to be established by well-designed clinical trials as required by current standards for evidence-based health care.[2-4,32,43] Data are lacking to directly compare the short- and long-term benefits and harms of UNHS versus those associated with selective screening.

Whether a UNHS program is successful depends on how reliable the screening test results are. UNHS is currently based on electrophysiological screening using AOAE (DPOAE or TEOAE) and/or AABR technologies, and many different UNHS programs use devices in each of these categories, in single- or multiple-stage protocols.[2-8,13,14,16,20,38,40] In North America, various companies currently offer stand-alone (TEOAE, DPOAE, or AABR only) and/or combination units (configuring TEOAE, DPOAE, and AABR technologies within a single device). Most of these companies have received marketing approval from Health Canada. Screening with these devices is non-invasive and can be performed at the bedside in inpatient or outpatient settings. The available devices are relatively safe for the newborn and the screeners, are easy to use, and do not require highly trained staff. The rapid expansion of UNHS programs has brought into focus questions about the most appropriate screening devices and protocols to be used.

Efficacy/effectiveness of AOAE and AABR Devices

According to the primary research studies included in the selected systematic reviews[2-4,32,43] UNHS using AABR and/or TEOAE (either alone or in combination in two-stage protocols) increases the early identification of moderate to profound PCHL and may lead to early intervention in diagnosed infants. Under quiet conditions, the TEOAE test (in a one-stage protocol) has a sensitivity of up to 100% and a specificity of up to 97% for detecting PCHL.[3,4,32] In comparison, the AABR test has a specificity of 96% and a sensitivity of 80%. The positive predictive value of both TEOAE and AABR is also very low (1.5% and 2.2%, respectively). When compared to conventional ABR testing, the accuracy of TEOAE testing in a one-stage screening protocol appears to depend on the level of local ambient noise, as well as on the condition of the infant's ears at testing. No good evidence exists to support either AABR or TEOAE as the preferred testing for one-stage newborn hearing screening protocols.

From the available information, it appears that none of AOAE or AABR devices currently licensed in Canada were evaluated by any of the primary research studies included in the selected systematic reviews.[2-4,32,43] Neither did any of these studies evaluate the diagnostic accuracy of DPOAE testing.

Eight-year follow-up results from one good quality non-randomized controlled trial (considered as the best level of evidence that is currently available) suggest that a two-stage protocol using TEOAE followed by AABR may achieve a specificity of 98.5%, a sensitivity of 91.7%, and a positive predictive value of 6.5%.[3,4,32] This screening protocol had a false positive rate of 1.5% and a false negative rate of 4%. Results from the same study suggest that referral for confirmatory diagnostic testing and management of PCHL commonly occurs earlier and more frequently with UNHS using TEOAE followed by AABR than without UNHS.[2-4,32,43] These findings are corroborated by several cohort studies and by multiple descriptive studies. The assumption is that earlier and more frequent referral, diagnosis, and management will impact on the long-term functional outcomes of infants with PCHL.

The other questions related to the efficacy/effectiveness of using AOAE and/or AABR for UNHS could not be answered based on the evidence available from the selected systematic reviews. The efficacy/effectiveness of AOAE and/or AABR in terms of longer-term outcomes (such as development of speech and language, cognitive ability, and communication skills, as well as quality of life, mental health, satisfaction, and educational and professional status) may be difficult to establish because the impact on developmental outcomes and other patient-relevant outcomes is related to many other factors than just the accuracy of the screening technologies.

The impact of a UNHS program on patient-relevant outcomes such as language and communication development is still not entirely clear.[2-4,32,43] From the evidence reported by two good quality cohort studies, receptive language acquisition improvements are seen with UNHS. However, findings are unclear for expressive language improvements.

A major limitation of the application of these findings to the Canadian context is that they were all conducted outside of North America. Although screening methods and maternity experiences are likely similar, the processes of referral, follow-up, and treatment are expected to differ. Currently, no standard method is in place in Canada to track children through these processes in order to ultimately obtain language outcomes from a birth cohort, as was done in the Wessex study

conducted in the United Kingdom. Factors influencing follow-up and treatment as well as exposure to UNHS need to be considered when determining impact in terms of patient-relevant outcomes.

The impact of a UNHS program using AABR and/or AOAE on long-term, patient-relevant outcomes such as quality of life, educational development, and employment status has yet to be established.[2-4,32,43] Further investigation is warranted.

Safety of UNHS using AOAE and AABR Devices

UNHS using AOAE and/or AABR devices always has the potential to cause direct or physical harm from performance of the testing itself, and indirect or psychosocial harm as a consequence of the screening findings. The consequences of false positive and false negative findings are of special relevance, as are the consequences of possible over-diagnosis and over-treatment of children that do not actually need treatment.

Neither of the selected systematic reviews[2-4,32,43] found studies that reported any safety issues or concerns associated with using AABR and/or AOAE technology for newborn hearing screening in terms of side effects and complications to the newborn and/or screener due to performing the test itself. Both the AOAE and AABR test procedures are non-invasive, so reviewers considered that direct harm seems to be limited.[3,4,32] Local transient hypersensitivity reactions to electrode gels are possible, but no cases were reported.[3]

Since the previous 2007 review, efforts have been made to study the psychosocial harms of UNHS, including negative emotions, parental worry and anxiety, and attitudes toward infants.[2-4,32,43] Limited data on these outcomes were obtained from poor- to average-quality studies. The available data indicated no significant differences in measures of concern, anxiety, and parental attitudes for families with newborns who pass versus those who do not pass the screen. Likewise, no differences in anxiety levels were found between parents of unscreened babies or screened babies, regardless of whether the screening outcome was positive or negative. No studies reported on the psychosocial effects of false reassurance. No studies addressed the adverse effects of a child with PCHL being screened or diagnosed late.

Considerations on the Performance of AOAE and/or AABR in UNHS

The accuracy of AOAE and/or AABR as screening tools in UNHS programs depends on many factors, including the cut-off impairment levels (dB hearing level, frequency range), the age of the newborn at screening, the screening protocol used, and the environment in which the screening is performed.[2-6,8,14,16,20,32,35,43,45]

The use of reliable diagnostic behavioral audiological testing (i.e., visual reinforcement, play, or standard audiometry assessment techniques) in the entire population of newborns who receive AOAE and/or AABR is necessary to reliably estimate the test sensitivity for detecting PCHL. Only the Wessex study conducted such an evaluation in a large population, but the investigators used distraction or behavioural observation measures, the diagnostic quality of which is considered inadequate.[4] The less than perfect accuracy of the gold standard assessment itself might have biased the estimates of the evaluated screening tests.

The available evidence indicated that the performance of a screening test or protocol for UNHS should be interpreted in the context of:

- the prevalence of the target disorder
- the administration of the testing
- the extent to which newborns are successfully accessed for screening and are successfully followed up after a referral result
- the efficacy of the intervention(s) initiated following PCHL confirmatory diagnosis[3,4]

The benefits of implementing UNHS ultimately depend upon the availability of public services able to rise to the challenge of earlier diagnosis and the initiation of appropriate interventions.[5,8,10,14,20,26,33,36,40,41,62] Educational and support services also must be available to provide appropriate habilitation for newly identified and diagnosed infants and their families.

Technical Considerations

Both AOAE and AABR technologies emerged as integral parts of UNHS, although none provides a direct measure of hearing or is considered a true screening test of hearing.[5,6,8,16,19,20] They measure slightly different physiological mechanisms related to hearing and are most often used in multiple-stage screening protocols to reduce the number of false positive results and test all possible aspects of the structural integrity of the auditory pathway. However, even if an infant passes screening with these tests and protocols, hearing cannot be definitively considered normal until the child is mature enough for a reliable behavioural audiogram.

The AOAE and/or AABR devices perform better for infants with moderate to severe PCHL, for whom there is little debate that intervention needs to occur early in life to improve developmental outcomes.[2-6,8,16,20,23,24,35,42,45] Mild to moderate PCHL is usually missed by AOAE and/or AABR screening devices, regardless of the screening protocol involved, and is diagnosed later than more severe levels. Detection of auditory neuropathy typically needs both AOAE and AABR testing.

The use of AABR and /or AOAE devices for UNHS is still evolving (www.otoemissions.org).[44] No standards exist for the calibration of AOAE or AABR devices and uniform performance standards are lacking.[8,35,45] Manufacturers do not always provide sufficient supporting evidence to validate the screening criteria and/or automated algorithms used in their instruments.

The available evidence was obtained from studies evaluating earlier models of stand-alone TEOAE and AABR devices, and may underestimate the capabilities of the newer versions of these devices currently available on the Canadian market. However, whether the devices currently licensed in Canada (including stand-alone TEOAE, DPOAE, and AABR devices and combo units) would result in an improved discriminating ability between newborns with normal hearing and those with PCHI is yet to be determined.

LIMITATIONS OF THIS REVIEW/REPORT

The present review has several limitations. The literature search was limited to published reports of articles and documents that were written in English. Proprietary reports were excluded. Only full-text articles were included.

The review is limited because it summarizes only the results from three systematic reviews, and the results from subsequently published primary research studies (which may have addressed some of

the outstanding issues associated with the use of AOAE and/or AABR for UNHS) are not included. This review addresses the diagnostic screening accuracy of the various devices within a universal screening program. It does not address the question on what sequence of use of the various devices provides the best diagnostic screening accuracy within a universal program.

Also, the review only summarizes the recommendations from reports of relevant clinical practice guidelines and positions statements, and does not appraise their scientific foundations.

CONCLUSIONS

Based on the results reported by three systematic reviews, this review confirms previous findings that UNHS using AOAE (TEOAE) followed by AABR testing in a two-stage protocol is effective in terms of increasing early identification of moderate to profound PCHL and early intervention in diagnosed infants. The available evidence indicates that referral for confirmatory diagnostic testing and PCHL management commonly occurs earlier and more frequently with a UNHS using this protocol than without UNHS. It also indicates that early identification and treatment of infants with hearing impairments may be associated with advantages in language development. The risks and harms of UNHS seem slight. However, the data on early detection of hearing impairment in newborns and infants are not very robust. Further investigation, particularly as to the effect on longer-term, patient-relevant outcomes such as quality of life and educational development, is warranted.

Currently, no definitive data exists to determine which of the AOAE and/or AABR devices currently available on the Canadian market are the best. These devices still await prospective validation against an accepted gold standard.

UNHS using the automated testing devices represent only one component of a well-integrated and structured system of early identification and management for all infants who have hearing loss, which enables confirmation of hearing loss by 3 months of age and enrolment in a family-centered intervention program by 6 months of age. Resources need to be available for diagnosis and intervention before UNHS can be considered. An important component for the practical realization of early detection of hearing impairments in newborns and infants is the development and implementation of a functioning integrated system for registering and tracking both non-screened children and those with a conspicuous screening result.

APPENDICES

APPENDIX T.A: METHODOLOGY

Literature search

A research librarian from the Institute of Health Economics conducted a literature search between December 20, 2011 and February 2, 2012. The search was developed and carried out prior to the study selection process and was limited to English language publications and human studies published between 2006 and February 2012.

In addition to the search strategy outlined in Table T.A.1, the bibliographies and reference lists of all retrieved articles were examined and Internet searches were conducted to retrieve grey literature. Grey literature searches were conducted to identify literature from non-indexed sources, health technology assessment reports, guidelines, government documents, and regulatory status information (for example, National Guidelines Clearinghouse, Health Canada, Google).

Table T.A.1: Search strategy
See below for limits[†]

Database	Edition or date searched	Search Terms [††]
MEDLINE (includes in-process and other non-indexed citation) OVID Licensed Resource	2006–Dec. 20, 2011	1 hearing loss/ or deafness/ or hearing loss, bilateral/ or hearing loss, conductive/ or hearing loss, functional/ or hearing loss, high-frequency/ or hearing loss, mixed conductive-sensorineural/ or hearing loss, sensorineural/ or hearing loss, central/ or hearing loss, noise-induced/ or presbycusis/ or usher syndromes/ or hearing loss, sudden/ or hearing loss, unilateral/
		2 (PCHI or deaf* or auditory neuropathy).tw.
		3 (hearing adj2 (loss or impairment)).tw.
		4 1 or 2 or 3
		5 mass screening/ or neonatal screening/
		6 (screen* or diagnos* or test or tests or testing).ti.
		7 5 or 6
		8 4 and 7
		9 diagnostic techniques, otological/ or hearing tests/ or acoustic impedance tests/ or audiometry/ or audiometry, evoked response/ or audiometry, pure-tone/ or exp audiometry, speech/ or psychoacoustics/ or dichotic listening tests/ or recruitment detection, audiologic/ or otoscopy/ or vestibular function tests/ or caloric tests/ or electronystagmography/
		10 ((hearing or audiological) adj1 (assessment* or screening)).tw.
		11 Evoked Potentials, Auditory, Brain Stem/ or Otoacoustic Emissions, Spontaneous/
		12 (auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE).tw.
		13 9 or 10 or 11 or 12
		14 8 or 13
		15 (ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA).tw.

		16 4 and 15 17 14 or 16 18 limit 17 to (yr="2006 -Current" and "all infant (birth to 23 months)") 19 exp Infant/ 20 (neonate or infant or newborn).mp. 21 19 or 20 22 17 and 21 23 limit 22 to yr="2006 -Current" 24 18 or 23 25 (cochlear implant* or otitis media).ti. 26 24 not 25 **1345** results
Cochrane Library (including DARE, HTA)	2006–January 16, 2012	#1 MeSH descriptor Hearing Loss explode all trees #2 (PCHI or deaf* or auditory neuropathy):ti,ab,kw #3 (hearing NEAR/2 (loss or impairment)):ti,ab,kw #4 (#1 OR #2 OR #3) #5 MeSH descriptor Mass Screening, this term only #6 MeSH descriptor Neonatal Screening, this term only #7 (screen* or diagnos* or test or tests or testing):ti,ab,kw #8 (#5 OR #6 OR #7) #9 (#4 AND #8) #10 MeSH descriptor Diagnostic Techniques, Otological explode all trees #11 ((hearing or audiological) NEAR/1 (assessment or screening)):ti,ab,kw #12 MeSH descriptor Evoked Potentials, Auditory, Brain Stem, this term only #13 MeSH descriptor Otoacoustic Emissions, Spontaneous, this term only #14 (auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE):ti,ab,kw #15 (#10 OR #11 OR #12 OR #13 OR #14) #16 (#9 OR #15) #17 (ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA):ti,ab,kw #18 (#4 AND #17) #19 (#16 OR #18) #20 (neonate* or infant* or newborn*):ti,ab,kw, from 2006 to 2012 #21 (#19 AND #20) **14 results**
Web of Science	2006–January 16, 2011	# 18 #17 and #16 # 17 TS=(meta-analysis OR metaanalysis OR search OR pubmed OR medline OR cinahl OR HTA OR "technology assessment" OR (systematic* SAME review*)) # 16 #14 and #15 # 15 TS=((neonate* or infant* or newborn*)) # 14 #13 Databases=SCI-EXPANDED, CPCI-S Timespan=2006-2012 # 13 #10 or #12 # 12 #3 AND #11

		# 11 TS=(ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA) #10 #8 or #9 # 9 #5 OR #6 or #7 # 8 #4 AND #3 # 7 TS=((auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE)) # 6 TS=((hearing assessment* or audiological assesment* or hearing screening or audiological screening)) # 5 TS=(hearing test* or audiometry or psychoacoustics) # 4 TI=(screen* or diagnos* or test or tests or testing) # 3 #2 OR #1 # 2 TS=((hearing NEAR/2 (loss or impairment))) # 1 TS=((PCHI or deaf* or auditory neuropathy)) **31 results**
CINAHL	2006–January 16, 2011	S23 S20 or S22 S22 S19 and S21 S21 meta-analysis OR metaanalysis OR pubmed OR medline OR cinahl OR search* OR (systematic* AND review*) Limiters - Published Date from: 20060101-20121231 Narrow by SubjectAge: - Infant: 1-23 months S20 S19 Limiters - Published Date from: 20060101-20111231; Publication Type: Meta Analysis, Systematic Review Narrow by SubjectAge: - Infant: 1-23 months S19 S16 or S18 S18 S5 and S17 S17 ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA S16 S9 or S15 S15 S10 or S11 or S12 or S13 or S14 S14 auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE S13 (MH "Otoacoustic Emissions, Spontaneous") S12 (MH "Evoked Potentials, Auditory, Brainstem") S11 hearing assessment* or audiological assessment* or hearing screening or audiological screening S10 (MH "Diagnosis, Ear") OR (MH "Hearing Tests") OR (MH "Acoustic Impedance Tests") OR (MH "Audiometry+") OR (MH "Dichotic Listening Tests") OR (MH "Hearing Screening") OR (MH "Otoacoustic Emissions, Evoked") OR (MH "Otoscopy") OR (MH "Vestibular Function Tests+") S9 S5 and S8 S8 S6 or S7 S7 TI screen* or diagnos* or test or tests or testing S6 (MH "Health Screening") OR (MH "Neonatal Assessment") S5 S1 or S2 or S3 or S4 S4 (hearing loss or hearing impairment) S3 (MH "Auditory Neuropathy")

		S2 PCHI or deaf* or auditory neuropathy S1 (MH "Hearing Disorders") OR (MH "Deafness") OR (MH "Deaf-Blind Disorders") OR (MH "Usher's Syndrome") OR (MH "Hearing Loss, Partial") OR (MH "Hearing Loss, Conductive") OR (MH "Hearing Loss, Functional") OR (MH "Hearing Loss, High-Frequency") OR (MH "Hearing Loss, Sensorineural") OR (MH "Hearing Loss, Central") OR (MH "Hearing Loss, Noise-Induced") OR (MH "Pendred Syndrome") **8** results
Embase	2006–January 16, 2011	1 hearing impairment/ or branchiootorenal syndrome/ or conduction deafness/ or exp congenital deafness/ or deafblindness/ or hearing loss/ or hypoacusis/ or mixed hearing loss/ or monaural hearing/ or morquio syndrome/ or norrie disease/ or exp perception deafness/ or sudden deafness/ or unilateral hearing loss/ 2 exp vestibulocochlear nerve disease/ 3 (PCHI or deaf* or auditory neuropathy).tw. 4 (hearing adj2 (loss or impairment)).tw. 5 1 or 2 or 3 or 4 6 mass screening/ or auditory screening/ or developmental screening/ or newborn screening/ 7 (screen* or diagnos* or test or tests or testing).ti. 8 6 or 7 9 5 and 8 10 auditory system examination/ or otoscopy/ or tonotopy/ or tympanometry/ 11 hearing test/ or exp audiometry/ or auditory screening/ or dichotic listening/ 12 ((hearing or audiological) adj1 (assessment or screening)).tw. 13 evoked brain stem auditory response/ 14 exp otoacoustic emission/ 15 (auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE).tw. 16 10 or 11 or 12 or 13 or 14 or 15 17 9 or 16 18 (ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA).tw. 19 5 and 18 20 17 or 19 21 limit 20 to yr="2006 -Current" 22 (neonate or infant or newborn).mp. 23 21 and 22 24 limit 21 to infant 25 23 or 24 26 25 not (cochlear implant* or otitis media).ti. 27 meta analysis/ 28 "systematic review"/ 29 (search* or meta-analysis or medline or pubmed or psychinfo or psycinfo or

		(systematic* adj3 review*)).tw. 30 technology assessment.mp. or HTA.tw. 31 27 or 28 or 29 or 30 32 26 and 31 **40 results**
Grey Literature			
Guidelines			
AMA Clinical Practice Guidelines www.topalbertadoctors.org/TOP/CPG/		Dec. 20, 2011	Browsed list of topics **0 results**
NICE Guidance www.nice.org.uk/		Feb. 2, 2012	otoacoustic; "auditory brainstem"; "hearing screening" **0 results**
CALSPA www.caslpa.ca		Feb. 2, 2011	Browsed list **1 result**
ACSLPA www.acslpa.ab.ca/		Feb. 2, 2011	Browsed list **1 result**
CMA Infobase http://mdm.ca/cpgsnew/cpgs/index.asp		Dec. 20, 2011	Browsed list of publications **1 result**
National Guideline Clearinghouse www.ngc.gov		Dec. 20, 2011	(newborn* OR infant*) AND hearing AND screening **4 results**
Coverage/Regulatory/Licensing Agencies			
Alberta Health and Wellness www.health.gov.ab.ca		Jan.25, 2011	Infant +hearing +screening; newborn +hearing +screening **0 results**
Medical Devices Active Licence Listing www.mdall.ca/		Dec. 20, 2011	Newborn hearing or infant hearing Or OAE or ABR or otoacoustic or auditory brainstem **68 results**
Health Canada www.hc-sc.gc.ca		Dec. 20, 2011	Newborn hearing or infant hearing Or OAE or ABR or otoacoustic or auditory brainstem **0 results**
US Food and Drug Administration Databases www.accessdata.fda.gov/scripts/cdrh/devicesatfda/index.cfm		Jan. 25, 2012	ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA **18 results**
Aetna Clinical Policy Bulletins www.aetna.com/about/cov_det_policies.html		Jan 26, 2011	otoacoustic; "auditory brainstem response" or "newborn hearing screening" or "infant hearing screening" **0 results**

HTA resources		
INESS www.inesss.qc.ca/	Jan. 26, 2012	otoacoustic; "auditory brainstem"; "hearing screening" **0 results**
CADTH www.cadth.ca/index.php/en/	Jan. 26, 2012	otoacoustic; "auditory brainstem" "hearing screening" **4 results**
Institute for Clinical and Evaluative Sciences (ICES), Ontario www.ices.on.ca	Feb 2, 2012	Browsed list **0 results**
Health Technology Assessment Unit at McGill www.mcgill.ca/tau/	Feb 2, 2012	Browsed list **0 results**
Medical Advisory Secretariat www.health.gov.on.ca/english/providers/program/mas/mas_mn.html	Feb. 2, 2012	Browsed list **0 results**
Other Grey Literature Sources		
Proquest Dissertations and Theses	Feb 2, 2011	Otoacoustic or auditory brainstem or hearing screening AND (infant or newborn or neonate) **0 results**
Search Engines		
Google	Feb 2, 2011	infant OR newborn OR neonate OR otoacoustic OR "auditory brainstem" "hearing screening" –pubmed **7 results**
NHS Evidence	Feb 2, 2011	hearing screening AND (infant or newborn or neonate) OR otoacoustic OR auditory brainstem **13 results**

Note:

† **Limits:** Searches were limited to **publication dates** 2006–2012**; language**: English only; **studies**: human studies only. These limits are applied in databases where such functions are available.

†† "*", "# ", and "?" are truncation characters that retrieve all possible suffix variations of the root word, e.g., surg* retrieves surgery, surgical, surgeon, etc.

Search Strategy: # Searches Results

Selection of studies

One reviewer screened titles and abstracts. Full-text publications of relevant articles were retrieved. The same reviewer determined eligibility of studies according to predefined inclusion/exclusion criteria.

Inclusion criteria

Research studies were included if they met the following criteria:

Study design: systematic reviews (quantitative and/or qualitative) conducted to evaluate the efficacy/effectiveness and safety of automated testing devices used for UNHS.

Note: A review was considered to be systematic if it met the following five criteria from Cook et al.:[64]

- focused clinical question
- explicit search strategy
- use of explicit, reproducible, and uniformly applied criteria for article selection
- critical appraisal of the included studies
- qualitative or quantitative data synthesis

Population: asymptomatic newborns (*newborns defined: birth through 3 months; asymptomatic newborns defined: not necessarily considered at risk for congenital hearing impairment/loss*) at urban or rural settings (birthing hospitals).

Interventions: automated testing devices approved/licensed in Canada to detect PCHL in asymptomatic newborns in an UNHS program.

Comparators: screening devices(s) considered as reference standard for this indication, other tests considered for this indication, or no testing.

Outcomes: sensitivity, specificity, and predictive value; impact on age at diagnosis of PCHL; impact on the number of infants diagnosed with PCHL; impact on usage of diagnostic tests; impact on age at start of treatment for PCHL; impact on treatment decisions (such as type of treatment); impact on usage of interventions to treat PCHL; impact on speech and language acquisition and development in children diagnosed with PCHL; impact on social and emotional development and other developmental milestones (such as scholastic achievement) in children with PCHL; risks and complications to the newborns and/or screeners from performing the test itself; and adverse effects of false positive and false negative test results.

Time frame: published from 2006 onwards.

Only full, peer-reviewed articles were included because abstracts do not provide adequate detail on the review methodology. However, where appropriate, relevant information contained in abstracts of primary research studies was used to inform the "Available evidence" section.

Studies were included if the published report was publicly available. In the case of duplicate publications, the most recent and complete version was included.

Also considered for inclusion in this review were publicly available published reports of:

- evidence-based clinical practice guidelines (CPGs) for using the automated testing devices to detect PCHL in newborns in an UNHS program
- evidence-based CPGs, position statements, and/or consensus statements on performing UNHS
- clinical reviews, overview articles, commentaries and discussion papers presenting background information on congenital hearing impairment/loss, UNHS, and the use of the automated testing devices of interest

An article was deemed to be an evidence-based CPG if:

- it contained the word "guideline" or "recommendation" in its title or introduction, or contained specific guidance, in the form of advice or instructions, on how to conduct UNHS to detect PCHL in asymptomatic newborns and/or the use of the UNHS testing devices of interest in an UNHS program
- it was developed by at least two authors
- it used an evidence-based approach in the process of developing the guidance (recommendations, advice, or instructions were based on a systematic review of the literature, were graded based on the strength of the supporting evidence, and reflected the consensus of the experts involved in the development of the guidance)
- it described the evidence-based approach used for the development of recommendations, advice, or instructions

Only articles reporting on research/analyses conducted in countries with developed market economies were considered, since the health status and disease burden of individuals, cultural and legal norms, and access to health care in countries with another status are likely to be too different from those of Canada to be clinically relevant. Countries deemed to have developed market economies, as defined by the United Nations, include Australia, Canada, Japan, New Zealand, the United States, and European countries (except for countries with market economies in transition) (http://unpan1.un.org/intradoc/groups/public/documents/un/unpan008092.pdf).

Only those publicly available, evidence-based CPGs, positions statements, and/or consensus documents developed by national bodies in Canada and other countries with developed market economies were considered.

Exclusion criteria

Excluded were:

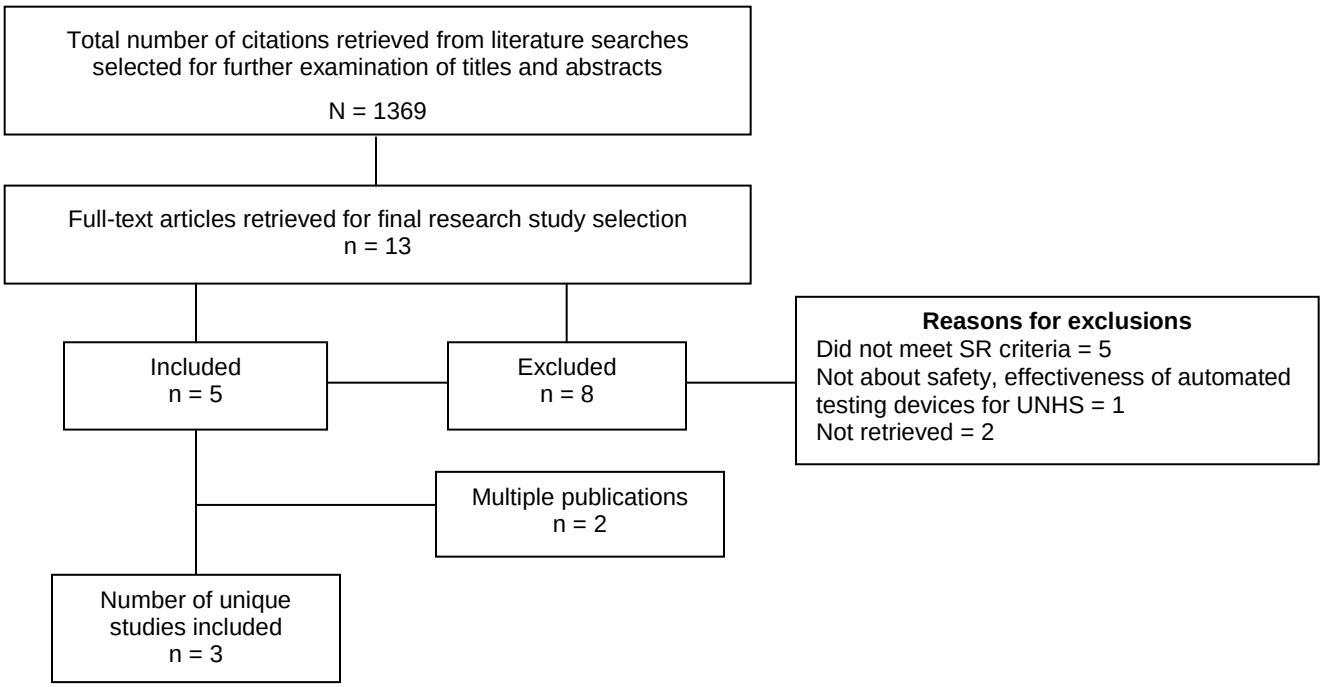
- published reports of systematic reviews reporting on the safety and efficacy/effectiveness of using the UNHS automated testing devices based only on results from primary research studies published before 2000
- published reports of primary research studies (screening accuracy studies, randomized and non-randomized controlled studies, comparative studies, cohort studies, case-control studies, cross-sectional studies, case series, or case reports) reporting on the safety and efficacy/effectiveness of using the automated testing devices for UNHS
- published reports of systematic reviews and primary research studies on the use of the testing devices of interest for detecting hearing impairment in infants *older than 3 months* and/or in young children in (universal) hearing screening programs
- published reports of systematic reviews and primary research studies that involved newborns, infants older than 3 months, and young children, but did not separately report on the use of the automated testing devices of interest for detecting PCHL in asymptomatic newborns in a UNHS program

- published reports of studies that evaluated the use of the automated testing devices of interest for other indications (for example to diagnose congenital hearing impairment/loss in screened newborns) and/or in other categories of newborns (for example in symptomatic newborns at risk for developing hearing impairment/loss)
- published reports of UNHS program evaluation studies
- published reports of animal studies
- conference abstracts, editorials, letters, technical reports, and book reviews

Also excluded were published reports of narrative and descriptive reviews, which summarized the research on the topic but lacked an explicit description of a systematic approach to the identification and interpretation of evidence. They were considered only as a source of background information, where appropriate.

Figure 1 provides a summary of the research study selection process.

Figure T.1: Research study selection process



Data extraction

One reviewer abstracted the data from the published reports of the selected systematic reviews. Main characteristics, findings, and conclusions from these studies, and details of their methodology are summarized in Table T.C.1 and Table T.C.2 (see Appendix T.C).

For studies in which the reporting of the methodology was unclear, the authors or agencies that produced the published reports were not contacted for further information. These studies were excluded from data extraction for not meeting all criteria for a systematic review (see Table T.B.1).

Methodological quality assessment

Due to time constraints, a formal critical appraisal of the methodological quality of the selected research studies was not performed. An informal methodological quality assessment of the selected research studies was conducted in the selection process by applying the five criteria from Cook et al.[64] However, no attempt was made to assess the validity of their findings.

No attempt was made to appraise the scientific foundations of the selected CPGs.

Data synthesis

Due to time constraints, a comprehensive qualitative analysis was not conducted.

External review

Members of the provincial Expert Advisory Group assembled for this project reviewed the draft report.

APPENDIX T.B: EXCLUDED STUDIES

The application of the selection criteria for research studies described in Appendix T.A resulted in eight full text articles being excluded from data extraction and synthesis. Table T.B.1 lists the excluded full-text reports of the retrieved research studies and the main reasons for their exclusion.

Table T.B.1: Excluded full text articles

Main reason for exclusion: *The study did not meet the SR criteria* (n = 5)
Kamdar S, McGarry BJ, and Roemheld-Hamm, B. Should we recommend routine newborn hearing screening? *Evidence-Based Practice* 2010;13(8):8-9.
Papacharalampous GX, Nikolopoulos TP, Davilis DI, Xenellis IE, and Korres SG. Universal newborn hearing screening, a revolutionary diagnosis of deafness: real benefits and limitations. *European Archives of Oto-Rhino-Laryngology* 2011;268(10):1399-1406.
Patel H, Feldman M, Amit M, Cummings C, Gander S, Grueger B, and Rowan-Legg A. Universal newborn hearing screening. *Paediatrics and Child Health* 2011;16(5):301-5.
Ptok M. Early detection of hearing impairment in newborns and infants. *Deutsches Arzteblatt* 2011;108(25):426-31.
Tann J, Wilson WJ, Bradley AP, and Wanless G. Progress towards universal neonatal hearing screening: a world review. *Australian & New Zealand Journal of Audiology* 2009;31(1):3-14.
Main reason for exclusion: *The study did not report on the safety and/or effectiveness of automated testing devices used in UNHS programs* (n = 1)
Canadian Agency for Drugs and Technologies in Health. Auditory screening and hearing loss prevention: a review of the clinical evidence and guidelines. Ottawa: Canadian Agency for Drugs and Technologies in Health (CADTH). 2011. Accessed online on January 06, 2012 at www.cadth.ca/media/pdf/htis/march-2011/L0252_Audio_screening_final.pdf.
Main reason for exclusion: *The full text of the study was not retrieved* (n = 2)
Schnell Inderst P, Kunze S, Hessel F, Grill E, Siebert U, Nickisch A, et al. Screening of the hearing of newborns – Update (Brief record). SO: Cologne: German Agency for Health Technology Assessment at the German Institute for Medical Documentation and Information (DAHTA DIMDI). 2006.
Haute Autorite de Sante. Assessment of systematic screening for permanent bilateral deafness in neonates (Structured abstract). *SO: Paris: Haute Autorite de Sante (French National Authority for Health) (HAS)*. 2007.

Multiple publications of studies included in the overview

From five included articles,[2-4,32,43] two were identified as multiple publications (Table T.B.2);[2,4] that is, cases in which the same study was published more than once, or part of the data from an original report was republished. The multiple publications were not considered to be unique studies; and any information that they provided was included with the data reported in the main study.

Table T.B.2: Multiple publications

Multiple publications of studies included in the review (n = 2)
Institut fuer Qualitaet und Wirtschaftlichkeit im Gesundheitswesen. Newborn hearing screening in the detection of hearing impairment (Structured abstract). *SO: Cologne: Institut fuer Qualitaet und Wirtschaftlichkeit im Gesundheitswesen (IQWiG).* 2007.[4] **Associated publication of Wolff et al.**[32]
Agency for Healthcare Research and Quality. Universal newborn hearing screening: systematic review to update the 2001 U.S. Preventive Services Task Force recommendation (Structured abstract). *SO: Rockville: Agency for Healthcare Research and Quality (AHRQ)*, 2008. Agency for Healthcare Research and Quality (AHRQ).[2] **Associated publication of Nelson et al.**[43]

APPENDIX T.C: RESULTS REPORTED BY SELECTED SYSTEMATIC REVIEWS

Abbreviations

AABR	automated auditory brainstem response
ABR	auditory brainstem response
AHRQ	Agency for Healthcare Research and Quality
AOAE	automatic evoked otoacoustic emissions
CI_{95}	95% confidence interval
IQWiG	Institut für Qualität und Wirtschaftlichkeit im Gesundheitswesen (Institute for Quality and Efficiency in Health Care)
mo	month(s)
MSAC	Medical Services Advisory Committee
NHS	newborn hearing screening
NNS	number needed to screen
OAE	evoked otoacoustic emissions
PCHL	permanent (bilateral) congenital hearing loss
PPV	positive predictive value
QUADAS	quality assessment of diagnostic accuracy studies
RCT	randomized controlled trial
ROC	receiver operating characteristic(s)
Sn	sensitivity
Sp	specificity
TEOAE	transient evoked otoacoustic emissions
UK	United Kingdom
UNHS	universal newborn hearing screening
US(A)	United States (of America)
USPTF	US Preventive Services Task Force
VRA	visual reinforcement audiometry
WBN	well-baby nursery
wk	week(s)

Table T.C.1: Selected systematic reviews (characteristics, search results, main findings, and conclusions)

Study	Study's characteristics*	Study's search results* main findings* and conclusions**
Wolff et al. (2010)[4,32] **Type:** report commissioned by IQWiG **Countries:** *Germany, UK, the Netherlands*	**Included studies:** non-RCTs (including a concurrent control group) of diagnostic accuracy of screening tests (application studies); RCTs; non-randomized screening studies; controlled cohort screening studies (as long as intervention and control groups were observed, at least approximately, concurrently) **Excluded studies:** duplicate publications without relevant additional information; no full-text publication available **Participants:** children up to 12 mo**;** unselected screening population **Intervention:** UNHS using automated OAE (AOAE) and/or automated ABR (AABR) **Comparator(s):** other tests used to detect hearing impairment; different screening strategies; no screening **Outcome(s) and outcome measures:** test accuracy; quality of life, hearing ability, language development, psychosocial development, emotional development, cognitive and educational development, "screening/diagnosis adverse effects" caused by false positive or false negative results, adverse effects of treatment	**Search results:*** Included nine studies (reported in 12 publications), of which eight were cross-sectional studies (one published in 1994, one in 1997, two in 1998, three in 1999, and one in 2000) investigating the diagnostic accuracy of AOAE compared to AABR; one was a good-quality non-RCT (reported in four articles published in 1998, 1999, 2000, and 2005) that evaluated a two-stage screening procedure (AOAE and AABR); two cohort studies—one retrospective study reported on in two articles (published in 2000 and 2001) and one prospective study reported on in eight articles (published in 1998, 1999, 2000, 2005, 2006)—included in the evaluation of benefits and harms of screening versus no screening **Main Findings:*** Efficacy/Effectiveness: Data from eight cross-sectional studies showed that compared to AABR, the AOAE Sn values vary between 0.50 and 1.0 and OAE Sp values vary between 0.49 and 0.97. Based on results from one non-RCT, estimated Sn of two-stage screening (combined AOAE and AABR) was 0.917 (95% CI 0.742 to 0.977) and Sp was 0.985 (95% CI 0.983 to 0.987). When children not participating in screening were included (intention-to-screen), program Sn was 0.710 (95% CI 0.520 to 0.858). PPV was 6.5%. Results from two comparative studies indicate a benefit for UNHS in terms of language development of children (average ages of 3 to 8 years) with hearing impairments. Safety Included studies provided no indications of direct negative consequences from the screening test. Because of lack of reliable studies, possible harms from neonatal hearing screening could not be evaluated. Potential of harm exists, particularly from false positive findings. **Conclusions**** "There are indications that children with hearing impairment identified in UNHS programs have advantages with respect to language development. Other patient-relevant outcomes, such as social aspects, quality of life, educational development and, finally, professional situation, have not been adequately investigated for evaluation." "There is a lack of high-quality evidence regarding all elements of newborn hearing screening." "No overall reliable evaluation is possible for the diagnostic accuracy of OAEs and ABR as initial screening tests, as there has been no evaluation in an adequately large group of children without risk factors. On the other hand, one study indicates that sequential screening (first OAE and then, if the finding is abnormal, ABR) in practical use might achieve acceptable sensitivity of >90%, with specificity of >98%. However, this estimate must be confirmed; as it is based on a relatively small number of children with hearing impairments, the 95% CI for sensitivity extends from 74% to 98%. In addition, it must be considered that the proportion of unidentified children markedly increases if the children not participating in screening are included in the evaluation (intention-to-screen analysis)."

* Search results and main findings regarding the use of automated testing devices for UNHS
** Conclusions stated by the author(s) and quoted directly from the published report

Table T.C.1: Selected systematic reviews (characteristics, search results, main findings, and conclusions) (cont'd)

Study	Study's characteristics	Study's search results*, main findings* and conclusions**
Nelson et al. (2008)[2,43] **Type:** AHRQ Evidence Synthesis Report **Country:** *USA*	**Included studies:** studies addressing key questions; published in English; conducted in the US or a comparable location; included infants screened before 6 months of age (for screening studies); descriptive and comparative studies (for question # 3) **Excluded studies:** editorials; letters; non-systematic reviews; non-comparative studies; case series; chapters; comment/opinion **Participants:** infants screened before 6 months of age **Intervention:** UNHS using AOAE and/or AABR **Comparator(s):** selective/targeted screening; no screening **Outcome(s) and outcome measures:** yield of screening; effects of screening or early detection and treatment on language and communication; effects of UNHS on treatment initiation before 6 months; adverse effects of screening and early treatment	**Search results*** Included two community-based cohort studies (one fair quality published in 2005 and one good quality published in 2006) for question #1; one good non-RCT and six descriptive studies of UNHS (one published in 1998, one in 2000, two in 2002, one in 2003, and one in 2006) for question #2; and two fair quality cohort studies (published in 1999 and 2001), one poor quality case-control study (published in 2003), and five survey studies (published in 1997, 2000, 2001, 2004, and 2006) for question #3. **Main Findings*** <u>Efficacy/Effectiveness:</u> In a good community-based cohort of both high- and average-risk children with PCHL, those who had their hearing loss confirmed by age of 9 months or younger had better scores at age 8 years on measures of receptive and expressive language, but not on speech, than did those confirmed later. Children with PCHL who underwent UNHS had better scores than those who did not on measures of receptive language, but not on expressive language and speech. More children undergoing UNHS had confirmation of impairment by age of 9 months than did those not screened as newborns (67% versus 27%; CI 24-56%; p<0.001). Data from a large, good-quality non-RCT and six descriptive studies indicate that average and high risk infants with PCHL born in hospitals with UNHS have earlier referral and initiation of treatment than those born in hospitals without UNHS. In the non-randomized trial, one additional case of PCHL was referred before the age of 6 months for every 1969 (1011-12,896) infants in the UNHS population. <u>Safety</u> No reporting on physical or direct harms of performing UNHS using automated testing devices. Two fair-quality cohort studies, one poor-quality case-control study, and five survey studies with >40% response rates indicate that usual parental reactions to an initial non-pass on a hearing screen include worry, questioning, and distress. These negative emotions resolve for most parents when a diagnostic test is provided with a normal result. Little information exists about the adverse effects of early interventions. **Conclusions**** "Children with hearing loss who had UNHS 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."

* Search results and main findings regarding the use of automated testing devices for UNHS
** Conclusions stated by the author(s) and quoted directly from the published report

Table T.C.1: Selected systematic reviews (characteristics, search results, main findings, and conclusions) (cont'd)

Study	Study's characteristics	Study's search results*, main findings* and conclusions**
Merlin et al, (2007)[3] **Type:** MSAC Assessment Report **Country:** *Australia*	**Included studies:** studies published after 1980; conducted in Australia or a comparable location; cross-sectional surveys (random sampling), case series (consecutive children, or cohort studies for question #1; RCTs and non-RCTs, cohort studies, registers or systematic review of these study designs, case-control or cross-sectional studies for questions on safety; cross-sectional studies and case-control diagnostic studies for question on and diagnostic accuracy **Excluded studies:** studies not addressing research question; not providing information on pre-specified target population; not including one of pre-specified interventions; not comparing results to a pre-specified comparator; not addressing one of pre-specified outcomes and/or not providing adequate numerator and/or denominator; not having appropriate study design **Participants:** neonates and infants (without particular diseases or traumas associated with hearing impairment for question #3) up to 6 months of age undergoing testing for PCHL; parents of neonates and infants up to 6 months of age undergoing testing for PCHL **Intervention:** universal, including targeted, neonatal hearing screening using either AOAE or AABR	**Search results*:** Included five controlled trials (level II-2 interventional evidence; one case-control study and four cohort studies; poor to average quality; published 1998, 1999, 2005, 2006, and 2007) to address safety of UNHS; five comparative studies (level III-2 diagnostic evidence; average quality; published in 1994, 1997, 1998, 1999, and 2002) to address diagnostic accuracy of tests; and five controlled studies (levelII-1 and III-2 screening evidence; good quality; reported on in seven articles published one in 1998, two in 2001, one in 2005, and three in 2006) addressed impact of UNHS on clinical management of PCHL and on PCHL-associated adverse effects. **Main Findings*** Efficacy/Effectiveness: Data from four comparative studies reported sensitivity of initial TEOAE in a one-stage protocol ranging from 50% to 100% when compared to conventional ABR. Under quiet conditions, the test can elicit sensitivity of 100% and specificity 92%, positive predictive value of 1.5%. Data from one comparative study reported specificity and sensitivity values for AABR testing of 96% and 80%, respectively, and positive predictive value of 2.2% (when compared to conventional ABR). Highest level of evidence available indicates that referring an infant for diagnostic testing before the age of 6 months is nearly three times more likely with UNHS than without. Infants born during periods of UNHS are twice as likely to receive a diagnosis of PCHL than infants born in periods without UNHS. Screening may increase the likelihood of PCHL management before the age of 10 months by nearly two-and-a-half times. Two good-quality cohort studies indicate that children with PCHL born in hospitals with UNHS have better receptive language abilities than children with PCHL born in hospitals without screening. Information on the impact of UNHS on longer-term outcomes (such as educational and employment status) is yet to be obtained. Safety Included studies did not report any physical harm resulting from UNHS using automated testing devices. Five controlled studies on psychosocial harms of UNHS reported, more frequently, higher parental anxiety levels when infants screened positive rather than negative, but no clinically important differences in anxiety levels were found. No differences in anxiety were found between parents of unscreened babies or screened babies regardless of whether the screening outcome was positive or negative. No studies reported on psychosocial effects of false reassurance or of a true positive diagnosis. **Conclusions**** "UNHS does not cause psychosocial harm, although no data were found on harms caused by false reassurance." "Under quiet conditions the TEOAE test (in a one stage protocol) posses excellent sensitivity (up to 100%) and good specificity (92%) for detecting PCHI. The positive predictive value of TEOAE is poor at 1.5%. In comparison, the AABR test has excellent specificity (96%) and good sensitivity (80%). The positive predictive value of AABR is also very low (2.2%)" "... findings from one good quality level III-1 study (two stage TEOAE followed by AABR) suggest that referral for definitive diagnostic testing, actual PCHI diagnosis, and management of PCHI commonly occurs earlier and more frequently with UNHS than without it. However, at this time the effect of UNHS on primary or patient-relevant outcomes in not entirely clear."

	Comparator(s): no UNHS; medical or behavioural assessment, tympanometry, steady state evoked potential testing, and/or conventional or diagnostic ABR testing at ≤ 6 months **Outcome(s) and outcome measures:** prevalence; adverse psychological, psychosocial, or physical health outcomes associated with testing procedure, diagnosis, and/or treatment options; sensitivity, specificity, positive and negative predictive values; screening yield; rate and quality of language acquisition; behaviour; family functioning; communication ability/social functioning; educational achievement; employment status; socioeconomic status; quality of life; age of referral for diagnostic testing; age of PCHL diagnosis; age of receiving therapeutic interventions	

* Search results and main findings regarding the use of automated testing devices for UNHS
** Conclusions stated by the author(s) and quoted directly from the published report

Table T.C.2: Selected systematic reviews (objective and methods)

Study	Study's objective and methods
Wolff et al. (2010)[4,32]	**Objective:** The main objective was to evaluate the benefits and harms of UNHS programs in the early detection of hearing impairment. In the absence of randomized controlled trials evaluating whole screening programs, the study divided the objective into three systematic reviews of non-randomized controlled studies of diagnostic accuracy of screening tests, screening versus no screening, and therapeutic effect of early versus later treatment. **Methods:** MEDLINE (Ovid), EMBASE, CINAHL, PsycINFO, PSYNDEX, ERIC Cochrane Library databases (Clinical Trials, systematic Cochrane Reviews), DARE, NHS EED, and health technology assessments (HTA) were searched. The search strategy in Medline (Ovid) used combinations of medical subject heading terms and text words and was not restricted to specific languages or years of publication. The search strategies for other databases were conducted following similar search strategies. Last search was carried out on 1 October 2007. Reference lists of included studies and identified reviews were searched for additional references. Their authors were contacted for additional information on included studies and enquiries were sent to hospitals and to manufacturers of screening instruments, hearing aids, and cochlear implants. Search findings were screened for potentially eligible studies. Abstracts and full articles were obtained for detailed evaluation, and eligible trials were included in the systematic reviews. To evaluate tests in screening populations the study assessed the diagnostic accuracy of automatic otoacoustic emissions and auditory brainstem response against any reference test. A quality evaluation tool of the Centre for Reviews and Dissemination was modified and used to evaluate screening and treatment studies. Particular attention was paid to aspects of sample size planning, blinding, comparability of groups in baseline characteristics, consideration of confounding factors, and transparency of patient flow. The QUADAS instrument was used for quality assessment of studies evaluating the diagnostic accuracy of screening tests. Two reviewers independently carried out all stages of study selection, data extraction, and quality assessment. Any disagreement during the selection, extraction, and assessment process was resolved by discussion and consensus. Based on limitations of included studies, no meta-analysis or sensitivity analysis could be performed. Graphs were generated using Version 5.0.17 of the Review Manager. The report was prepared in collaboration with external experts. The preliminary report was published on the Internet so interested parties could submit written comments. All written comments fulfilling formal criteria were discussed in a scientific debate before production of the final report.

Table T.C.2: Selected systematic reviews (objective and methods) (cont'd)

Study	Study's objective and methods
Nelson et al. (2008)[2,7,43]	**Objective:** This review is an update of the USPSTF recommendations on the use of UNHS to detect moderate-to-severe permanent, bilateral congenital hearing loss. The aim was to answer the following 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? (3) What are the adverse effects of screening and early treatment? **Methods:** Literature searches were conducted to systematically identify articles published since the 2002 USPSTF recommendation. Databases included the Cochrane Central Register of Controlled Trials, Cochrane Database of Systematic Reviews, Database of Abstracts of Reviews of Effects (through the fourth quarter of 2007), and Ovid Medline (2000 to November 2007 for key questions #1 and #2 and 1996 to November 2007 for key question #3). Additional articles were obtained from reference lists of related reviews, studies, editorials, reports, websites, and by consulting experts. Abstracts and selected full-text articles were screened based on inclusion and exclusion criteria specific to each key question. Data from studies that met inclusion criteria were abstracted in evidence tables (data included study, year, setting, patient population, inclusion/exclusion criteria, risk status, methods, and results.). Selected studies were rated for quality with predetermined criteria (design-specific criteria developed by the USPSTF). The overall rating of each study considers internal validity and applicability. Descriptive studies without quality criteria were summarized. An outcomes table, estimating the number needed to screen under various assumptions, was determined using estimates from the most relevant studies. The USPSTF advised the Oregon Evidence-based Practice Center in formulating and reporting this review. Additional experts provided comments on an earlier draft.

Table T.C.2: Selected systematic reviews (objective and methods) (cont'd)

Study	Study's objective and methods
Merlin et al. (2007)[3]	**Objective:** The objective of this MSAC assessment was to determine whether there is sufficient evidence to establish a program of universal neonatal hearing screening in Australia. Research questions included: 1. What is the prevalence of permanent hearing impairment in neonates and infants in Australia? 2. What is the diagnostic accuracy of the tests for permanent childhood hearing impairment when conducted on the neonate or infant? 3. Does universal neonatal hearing screening, and the finding of a positive and/or negative test, affect the clinical management or treatment options available to permanently hearing-impaired infants? 4. Does universal neonatal hearing screening, and therefore possible alterations in clinical management, have an impact on the adverse outcomes associated with permanent childhood hearing impairment? **Methods:** The medical literature was searched to identify relevant studies on UNHS for the period between 1996 and August 2007. Twelve electronic databases (including Cochrane Central Register of Controlled Trials, Cochrane Database of Systematic Reviews, PubMed, PsycInfo, Web of Science and Embase) were searched. Other sources included the Internet (to access websites of various agencies and organizations, Trip database, and the National Library of Medicine Locator Plus database), and hand searching of 12 journals (2006–2007). Expert clinicians (MSAC Advisory Panel) were contacted to identify studies other than those found in regular searches. All included articles had their reference lists searched for additional relevant source material. The searches used combinations of medical subject heading terms and text words and some were was restricted to English language or by years of publication (1966–2007). Two reviewers assessed citations for study selection independently. Any doubt was resolved by consensus between the two reviewers; a third reviewer was included where necessary. A study profile was developed for each included study outlining the level of evidence, study quality, authors, publication year, location, study population characteristics, type of intervention, testing or screening protocol, comparator or reference standard, and outcomes assessed. The evidence presented in the selected studies was assessed and classified using dimensions of evidence defined by the National Health and Medical Research Council (NHMRC 2000a). These dimensions consider important aspects of the evidence supporting a particular intervention and included three main domains: strength of evidence, size of the effect, and relevance of evidence. With respect to diagnostic evidence, the selected studies were graded according to pre-specified quality and applicability criteria (MSAC 2005). Study quality was assessed using NHMRC critical appraisal checklists designed for various study designs. The analysis of diagnostic accuracy, calculations of sensitivity, specificity, and positive predictive values of tests (with 95% confidence intervals) were undertaken where possible. Meta-analysis was not undertaken as the evidence-base was heterogeneous and there were only g=few controlled trials of screening. A narrative synthesis of the data was undertaken. An advisory panel with expertise in paediatrics, otorhynolaringology, audiology, deaf education, epidemiology, consumer issues, and neonatal hearing screening was established to evaluate the evidence produced by the assessment and to provide advice to the MSAC from clinical and client perspectives.

APPENDIX T.D: NEWBORN HEARING SCREENING DEVICES APPROVED BY HEALTH CANADA

1. Manufacturer

Company ID: 116857

GN OTOMETRICS A/S
Hoerskaetten 9
Taastrup, DK, 2630

Licence No.: 61627

Type: Device Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2003-01-23	OAE ANALYZERS

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2003-01-23	ACCUSCREEN	2003-01-23	ACCUSCREEN

www.otometrics.com/hearing-assessment/newborn-hearing-screening

Licence No.: 83603

Type: Device Group Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2010-08-11	ACCUSCREEN (SCREENERS FOR HEARING LOSS)

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2010-08-11	ACCUSCREEN ABR	2010-08-11	8-04-13903
2010-08-11	ACCUSCREEN ABR/DP	2010-08-11	8-04-13905
2010-08-11	ACCUSCREEN ABR/TE	2010-08-11	8-04-13904
2010-08-11	ACCUSCREEN ABR/TE/DP	2010-08-11	8-04-13906
2010-08-11	ACCUSCREEN DP	2010-08-11	8-04-13901
2010-08-11	ACCUSCREEN LITE DP	2010-08-11	8-04-13908
2010-08-11	ACCUSCREEN LITE TE	2010-08-11	8-04-13907
2010-08-11	ACCUSCREEN TE	2010-08-11	8-04-13900
2010-08-11	ACCUSCREEN TE/DP	2010-08-11	8-04-13902

Licence No.: 78175

2. Manufacturer

Company ID: 105147

INTERACOUSTICS A/S
Drejervaenget 8
Assens, DK, 5610

Licence No.: 65725

Type: Device Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2004-08-31	ABR (AUDITORY BRAIN STEM RESPONSE TESTING) UNITS

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2004-08-31	ABR UNITS	2004-08-31	EP15
		2004-08-31	EP25

http://interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=60914

http://interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=71112

Licence No.: 66910

Type: Device Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2004-12-22	OAE (OTOACOUSTIC EMISSION) UNITS

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2004-12-22	OTOREAD	2004-12-22	CLINICAL
		2004-12-22	SCREENING
		2004-12-22	STANDARD

http://interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=60998

Licence No.: 67181

Type: Single Device

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2005-01-20	ABRIS

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2005-01-20	ABR UNITS - ABRIS	2005-01-20	ABRIS

http://interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=60808

Licence No.: 67184

Type: Single Device

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2005-01-20	TEOAE25

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2005-01-20	TEOAE SYSTEM	2005-01-20	TEOAE25

http://interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=61028

Licence No.: 86197

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2011-05-25	TITAN SOFTWARE DPOAE440

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2011-05-25	TITAN SOFTWARE DPOAE440	2011-05-25	DPOAE440

http://interacoustics.com/com_en/Pages/Product/Abr/_index.htm?prodid=71112 b

3. Manufacturer

NATUS MEDICAL INCORPORATED
One Bio-Logic Plaza
Mundelein, IL, US, 60060

Licence No.: 80595

Type: Single Device

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2009-09-02	ALGO 5 NEWBORN HEARING SCREENER

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2009-09-02	ALGO 5 NEWBORN HEARING SCREENER	2009-09-02	010130

www.natus.com/index.cfm?page=products_1&crid=176

Licence No.: 62352

Type: Single Device

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2003-04-29	ALGO 3I NEWBORN HEARING SCREENER

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2003-04-29	ALGO 3I NEWBORN HEARING SCREENER	2003-04-29	010072
		2003-04-29	010073
		2003-04-29	010074
		2003-04-29	010075
		2003-04-29	010076
		2003-04-29	010077

www.natus.com/index.cfm?page=products_1&crid=303&contentid=118

Licence No.: 37239

Type: System

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2002-04-05	ALGO 3 NEWBORN HEARING SCREENER

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2002-04-05	ALGO 3 SCREENER	2002-09-09	010067
2002-04-05	ALGO 3 SCREENER - CABLES	2002-04-05	040518
		2002-04-05	040520
		2005-10-25	030697
		2006-01-19	040740
2002-04-05	ALGO 3 SCREENER - PRINTER	2002-04-05	040556
2002-04-05	PREPPING GEL	2002-04-05	900191

Licence No.: 24091

Type: Device Group Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2000-10-12	OTOACOUSTIC EMISSIONS DEVICES

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2000-10-12	AUDX I	2006-09-13	AX-001
2006-09-13	AUDX PRO	2006-09-13	AX-005
2006-09-13	AUDX PRO II	2006-09-13	AX-006
2006-09-13	AUDX PRO PLUS	2006-09-13	AX-007
2007-01-04	AUDX II	2007-01-04	AX-002
2007-01-04	AUDX III	2007-01-04	AX-003

www.natus.com/index.cfm?page=products_1&crid=184

Licence No.: 24089

Type: System

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2000-10-12	ABAER EVOKED POTENTIAL SCREENING SYSTEM

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2000-10-12	ABAER EVOKED POTENTIAL SCREENING SYSTEM	2000-10-12	520-AMPS01
		2000-10-12	541-NAVC01
		2000-10-12	541-NAVC04
		2000-10-12	580-ABAER1
		2000-10-12	580-ABRC01
		2000-10-12	580-INSER1-125
		2000-10-12	580-MEPTDH-125
		2000-10-12	580-OAEER2

		2000-10-12	580-TSTBX1
		2003-03-14	580-NAVCUB
		2004-11-05	515-TSTCBL
		2004-11-05	520-PS6V4A
		2004-11-05	541-ABRC10
		2004-11-05	541-USB001
		2004-11-05	580-ABAER2
		2004-11-05	580-BINSER
		2004-11-05	580-PROAE3
		2004-11-05	580-SINSER-012

www.natus.com/index.cfm?page=products_clinical&crid=181

4. Manufacturer

Company ID: 132443

NATUS EUROPE GMBH
Baermannstrasse 38
Munich, 09, DE, 81245

Licence No.: 84638

Type: Device Group Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2010-12-06	ECHO SCREEN HEARING SCREENER

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2010-12-06	ECHO SCREEN DPOAE	2010-12-06	010109-D
2010-12-06	ECHO SCREEN DPOAE&AABR	2010-12-06	010109-DA
2010-12-06	ECHO SCREEN DS DPOAE	2010-12-06	010129
2010-12-06	ECHO SCREEN DS SET	2010-12-06	010127 DS
2010-12-06	ECHO SCREEN PLUS AABR	2010-12-06	010127-A
2010-12-06	ECHO SCREEN PLUS DPOAE	2010-12-06	010127-D
2010-12-06	ECHO SCREEN PLUS DPOAE&AABR	2010-12-06	010127-DA

2010-12-06	ECHO SCREEN PLUS TEOAE	2010-12-06	010127-T
2010-12-06	ECHO SCREEN PLUS TEOAE&AABR	2010-12-06	010127-TA
2010-12-06	ECHO SCREEN PLUS TEOAE&DPOAE	2010-12-06	010127-TD
2010-12-06	ECHO SCREEN PLUS TEOAE&DPOAE&AABR	2010-12-06	010127-TDA
2010-12-06	ECHO SCREEN TEOAE	2010-12-06	010109-T
2010-12-06	ECHO SCREEN TEOAE&AABR	2010-12-06	010109-TA
2010-12-06	ECHO SCREEN TEOAE&DPOAE	2010-12-06	010109-TD
2010-12-06	ECHO SCREEN TEOAE&DPOAE&AABR	2010-12-06	010109-TDA
2010-12-06	ECHO SCREEN TS SET	2010-12-06	010127 TS
2010-12-06	ECHO SCREEN TS TEOAE	2010-12-06	010128

www.natus.com/index.cfm?page=products_1&crid=183

5. Manufacturer

Company ID: 114955

VIVOSONIC INC.
5525 Eglinton Avenue West, Unit 120
Toronto, ON, CA, M9C 5K5

Licence No.: 81824

Type: System

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2010-01-21	AURIX NEWBORN HEARING SCREENING SYSTEM

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2010-01-21	AURIX - AMPLIFIER	2010-01-21	11003
2010-01-21	AURIX - BATTERY CHARGER	2010-01-21	100351
2010-01-21	AURIX - BATTERY PACK	2010-01-21	100020
2010-01-21	AURIX - EAR DOME	2010-01-21	100004
2010-01-21	AURIX - ELECTRODE	2010-01-21	100001

2010-01-21	AURIX - NEWBORN HEARING SCREENING SYSTEM	2010-01-21	AURIX-1
2010-01-21	AURIX - SOUND STIMULATOR	2010-01-21	11002
2010-01-21	AURIX - SYSTEM SOFTWARE	2010-01-21	100021

www.vivosonic.com/en/aurix/index.html

6. Manufacturer

Company ID: 121647

OTODYNAMICS LTD.
36-38 Beaconsfield Rd.
Hatfield, HT, GB, AL10 8BB

Licence No.: 70568

Type: Device Group Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2006-02-23	OTOACOUSTIC EMISSION INSTRUMENT

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2006-02-23	OTOACOUSTIC EMISSION INSTRUMENT - ECHOCHECK OAE SCREENER	2006-02-23	A10-14-0-0
2006-02-23	OTOACOUSTIC EMISSION INSTRUMENT - ECHOPORT ADVANCED CLINICAL OAE ANALYSER	2006-02-23	A51-01-0-0
2006-02-23	OTOACOUSTIC EMISSION INSTRUMENT - ECHOPORT CLINICAL OAE ANALYSER	2006-02-23	A31-01-0-0
		2006-02-23	A31-02-0-0
2006-02-23	OTOACOUSTIC EMISSION INSTRUMENT - ECHOPORT OAE SCREENER/ANALYSER	2006-02-23	A21-01-0-0
		2006-02-23	A21-02-0-0
2007-12-18	OTOPORT OAE SYSTEM	2007-12-18	A15-02-0-3
		2007-12-18	A15-02-1-3
		2007-12-18	A15-02-2-3
		2007-12-18	A15-02-3-3
		2007-12-18	A15-02-4-3
		2007-12-18	A15-02-5-3

		2007-12-18	A15-02-6-3
		2007-12-18	A15-02-7-3
		2007-12-18	A15-03-0-3
		2007-12-18	A15-04-0-3
		2007-12-18	A15-05-0-3

www.otodynamics.com/screening.asp

7. Manufacturer

Company ID: 114365

DIAGNOSTIC GROUP LLC DBA MAICO DIAGNOSTICS
7625 Golden Triangle Drive
Eden Prairie, MN, US, 55344

Licence No.: 17657

Type: Single Device

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2000-03-02	ERO-SCAN

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2000-03-02	ERO-SCAN OTOACOUSTIC INSTRUMENT	2000-03-02	586

www.maico-diagnostics.com/us_en/Menus/ProductByUser/3-Newborn-Screening/1-Otoacoustic-Emissions

Licence No.: 61168

Type: Device Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2002-12-10	GSI AUDIOSCREENER

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2002-12-10	GSI AUDIOSCREENER	2002-12-10	2002-9700
		2002-12-10	2002-9750
2007-03-14	GSI AUDIOSCREENER + ABR	2007-03-14	2205-9735

	ONLY		
2007-03-14	GSI AUDIOSCREENER + ABR UPGRADE	2007-03-14	2205-9667
2007-03-14	GSI AUDIOSCREENER + DP/TEOAE ONLY	2007-03-14	2205-9730
2007-03-14	GSI AUDIOSCREENER + DP/TEOAE UPGRADE	2007-03-14	2205-9668
2007-03-14	GSI AUDIOSCREENER + DP/TEOE AND ABR	2007-03-14	2205-9740

Licence No.: 61235

Type: System

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2002-12-18	GSI AUDERA

Device Section		Identifier Section	
First Issue Date	**Device Name**	**First Issue Date**	**Device Identifier**
2002-12-18	AUDERA AMPLIFIER	2002-12-18	2001-9635
2002-12-18	GSI AUDERA	2002-12-18	2001-97XX
		2007-03-28	2001-9400
		2007-03-28	2001-9700
		2007-03-28	2001-9705
		2007-03-28	2001-9710
		2007-03-28	2001-9715
		2007-03-28	2001-9720
		2007-03-28	2001-9725
		2007-03-28	2001-9730

Licence No.: 61275

Type: Device Family

Licence Section		
Device Class	**First Issue Date**	**Licence Name**
2	2002-12-20	OTOACOUSTIC DEVICES

Device Section		Identifier Section	
First Issue Date	Device Name	First Issue Date	Device Identifier
2002-12-20	GSI 60 DPOAE SYSTEM	2002-12-20	1760-9715
		2002-12-20	1760-9755
2002-12-20	GSI 70	2002-12-20	1770-9702
		2002-12-20	1770-9705
		2002-12-20	1770-9706
		2002-12-20	1770-9708

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SECTION TWO: ECONOMICS ANALYSIS

Charles Yan, PhD; Anderson (Andy) Chuck, PhD, MPH

OBJECTIVES AND SCOPE

The objective is to compare the cost effectiveness of various strategies used in the universal screening of newborn hearing (UNHS).

METHODS

A review was conducted of the published economic literature on the cost effectiveness of alternative strategies for UNHS. The review was an update to the previous IHE report[1] on UNHS, published in 2007.

Search Strategy

Selected databases were searched for economic evaluation studies of UNHS. Databases searched included Medline, EMBASE, CINAHL, Cochrane Database of Systematic Reviews, Web of Science and grey literature. To supplement the electronic searches, reference lists of retrieved articles were also reviewed to find further studies. See Appendix E.1 for the literature search summary.

Selection Criteria

The search was limited to human studies and English-language publications. Eligible studies met the following predefined inclusion/exclusion criteria:

Inclusion Criteria:

- Study design: health technology assessment reports, systematic reviews and economic evaluation studies including studies of cost effectiveness, cost-utility, or cost-benefit
- Population: new-born children
- Interventions and comparators: various UNHS strategies
- Language: English
- Search period: from 2006 onward

Exclusion Criteria:

- Abstracts, case studies, narrative reviews, letters, and editorials
- Studies that reported the cost and outcomes of only one UNHS strategy (without a comparator)

Outcomes of Interest

- Number of correctly detected cases referred for follow-up/confirmatory testing
- Number of correctly identified non-cases not referred for follow-up/confirmatory testing

- Proportion of infants whose deafness was diagnosed within a follow-up period (for example, 6 months)
- Referral rate
- Cost per infant screened
- Cost per case detected
- Additional cost per health outcome gained

Quality Assessment

A formal quality assessment of economic studies was conducted with the Quality of Health Economic Studies (QHES) instrument.[3] The QHES instrument was designed to evaluate health economic analyses, including the analysis of cost minimization, cost effectiveness, and cost utility. It includes a weighting system to score and aggregate across individual criteria, thereby providing a summative quality index. The quality index ranges from 0 to 100, with a score of 75 or greater indicating acceptable quality.

Data Extraction

Data extracted from studies included study objective, UNHS strategies under investigation, cost components, health outcome measures, results, and conclusions.

RESULTS

Search Results

The literature search identified 150 references. After reviewing the titles and abstracts/summaries, 33 were retrieved for further review. Of the 33 studies, four studies met the final inclusion/exclusion criteria. See Appendix E.2 for data extraction from included studies and Appendix E.3 for quality assessment scores of included studies.

Evidence from Economic Literature

Uus et al.[2] examined the costs and cost effectiveness of the Newborn Hearing Screening Programme (NHSP) as compared to the Infant Distraction Test Screen (IDTS) in England. The NHSP takes place soon after birth and is based on transient evoked otoacoustic emissions (TEOAE) followed by automated auditory brainstem response (AABR) testing if there is no clear response at the TEOAE. The IDTS is performed when the child is approximately 8 months old and consists of localization responses to low-level sounds presented to child by a tester while a second tester manipulates the child's attention. Note that these testers were not audiologists but public health nurses or other health visitors who were part of the child's health check-up at 8 months of age. The primary outcome measure was the number of cases detected. The cost categories considered in the study included the costs of staffing, equipment, overhead, staff training and travel, and audiology. The results showed that NHSP and IDTS detected 1.06 and 0.36 cases per 1000 infants screened, respectively. For every 1000 infants screened, the costs associated with the NHSP and IDTS were £34,315 and £25,171, respectively. The cost per case detected was £31,410 for NHSP and £69,919 for IDTS. The marginal analysis indicated that compared to IDTS, NHSP was associated with an

additional £12,500 per additional case detected. The authors concluded that the study supports replacing IDTS with NHSP. The study had a quality score of 81.5.

Uilenburg et al.[4] examined the clinical efficacy, costs, and cost effectiveness of UNHS using TEOAE in three different settings in the Netherlands. The settings were:

1) screening at home in combination with screening for metabolic diseases, within 4 to 7 days of birth
2) screening at home in combination with the first home visit, within 3 weeks of birth
3) screening in baby clinics in a special screening session, within 4 weeks of birth

The primary outcome measures were the participation rate, the referral rate, and the number of cases detected. Cost components considered in the study were not reported. The results showed that the participation and referral rates were:

- 88.9% and 1.4% respectively for screening at home combined with metabolic diseases screening
- 86.9% and 2.7% respectively, for screening at home during an intake visit
- 75.2% and 2.3% respectively, for screening in baby clinics

The costs per infant screened and per detected case were:

- €28.3 and € 41,500, respectively, for screening at home combined with metabolic diseases screening
- €41.7 and € 61,800, respectively, for screening at home during an intake visit
- €20.5 and € 35,100, respectively, for screening in baby clinics

The authors stated that the most cost-effective setting was at-home screening combined with metabolic diseases screening, although a marginal analysis was not conducted. The study concluded that the TEOAE UNHS at home combined with the screening for metabolic diseases is a preferred option. The study was assessed with a quality score of 67.5.

Lin et al.[5] compared the efficacy, initial referral rate, cost, and cost effectiveness of three newborn hearing screening protocols in Taiwan. The three protocols were TEOAE alone, TEOAE plus AABR, and AABR alone. The primary outcome measures in the study were the referral rate and rate of congenital deafness cases detected. The direct medical costs were the equipment, salary, initial screening, and diagnostic follow-up tests. The study also included "intangible costs," including parental anxiety, transportation fees, and days off during follow-up tests.

The results indicated that the referral rates were:

- 5.8% for TEOAE alone
- 1.6% for TEOAE plus AABR
- 0.8% for AABR alone

AABR alone was associated with a significant decrease in referral rate compared to the other two strategies.

The accurate identification rate of congenital hearing loss was:

- 0.45% for TEOAE alone
- 0.25% for TEOAE plus AABR
- 0.42% for AABR alone

The total direct medical cost per screened infant was:

- $10.04 for TEOAE alone
- $8.60 for TEOAE plus AABR
- $7.33 for AABR alone

Results for intangible costs were not reported, but the authors stated that the intangible costs were large with TEOAE, medium with AABR, and small with AABR. The authors did not conduct a marginal analysis but stated that AABR alone was the most cost-effective. The study concluded that AABR alone significantly reduces the referral rate and is therefore less costly than the alternatives. The study was assessed with a quality score of 64.5.

Olusanya et al.[6] examined the efficacy and costs of hospital- and community-based infant hearing screening programs in Nigeria, with a focus on universal and targeted screening for newborn infants. Both the hospital- and the community-based programs consisted of a two-stage screening strategy with TEOAE and AABR, followed by diagnostic evaluation. The targeted newborn infants were defined as high risk if they were admitted into the special care baby unit in hospital or born outside hospital facilities, or if they had a history of neonatal jaundice. The primary outcome measures in the study were the referral rate and detected number of permanent, congenital, and early-onset hearing loss (PCEHL). The cost components included the costs of staff salary, equipment, consumables, transportation to screening sites, and diagnostic tests.

First-stage referral rates for universal and targeted infants were:

- 32.2% and 31.7%, respectively, for hospital-based screening
- 14.3% and 15.2%, respectively, for community-based screening

Second-stage referral rates for universal and targeted infants were:

- 3.3% and 4.4%, respectively, for hospital-based screening
- 4.1% and 4.9%, respectively, for community-based screening

The detected number of PCEHL per 1000 universal and targeted infants were:

- 5.3 and 16.7, respectively, for hospital-based screening
- 22.5 and 27.4, respectively, for community-based screening

The costs per infant screened for universal and targeted screening were:

- $13.30 and $73.24, respectively, for hospital-based screening
- $7.62 and $12.27, respectively, for community-based screening

The costs per infant detected with PCEHL for universal and targeted screening were:

- $2764.86 and $4631.33, respectively, for hospital-based screening
- $602.49 and $714.53, respectively, for community-based screening

The authors concluded that community-based screening was the most cost-effective option, although a marginal analysis was not conducted. The study was assessed with a quality score of 71.5.

DISCUSSION

The 2007 report[1] reviewed eight studies assessing the cost effectiveness of UNHS strategies published between January 2001 and August 2006. Results across the eight studies were mixed and greatly limited by their low quality. As a result, the report concluded that the cost effectiveness of UNHS could not be determined from the literature review. Subsequent to the 2007 report, four studies have been published assessing the cost effectiveness of UNHS. However, only one of the four studies was of acceptable quality based on the QHES instrument.[3]

According to the study conducted by Uus et al.,[2] the additional cost case detected was £12,500 for NHSP (TEOAE + AABR soon after birth) compared with IDTS (responses to low level sounds conducted at approximately 8 months of age). The study concluded that NHSP is cost effective and should replace IDTS. The cost per case detected indicates that NHSP does not provide cost savings (that is, greater than 0), but it should be mentioned that the time horizon adopted in their analysis did not examine longer-term outcomes and therefore would not have captured downstream cost impacts from earlier detection and intervention. Another limitation in their study was the use of the number of cases detected as their primary measure of effectiveness. Using the number of cases detected may not capture the full economic value because it focuses on sensitivity and not specificity of the screening test. From an economic perspective, the most cost-effective screening strategy should not only detect infants who are suitable for further diagnostic testing but also those who do not require further testing, thereby minimizing unnecessary resource use and other intangible costs such as parental anxiety.

The 2007 report did conduct an economic evaluation of one-stage (AABR) and two-stage (AABR and automatic otoacoustic emissions (AOAE)) protocols that included downstream costs associated with untreated hearing loss. The analysis showed that that the one-stage AABR screening protocol is cost effective compared to the one-stage AOAE protocol because the one-stage AOAE protocol is less accurate and more costly. The two-stage protocol was found to be more effective than the one-stage AABR but was also associated with higher costs, with an incremental cost-effectiveness ratio of $7574.78 ($CAD 2003) per infant detected, so the cost effectiveness of the two-stage protocol compared to the one-stage protocol depends on whether the additional benefit is worth the increased cost to the health system.

In conclusion, limited economic evidence has been published about the cost effectiveness of UNHS strategies. However, based on the economic evaluation conducted in the 2007 IHE report, one-stage AABR screening is less costly and more effective than one-stage AOAE, while the cost effectiveness of two-stage screening of AABR and AOAE depends on whether the additional effectiveness is worth the increased cost.

APPENDICES

APPENDIX E.1: LITERATURE SEARCH SUMMARY: HEARING SCREENING SCOPING SEARCH- ECONOMICS

General Information

The IHE research librarian conducted the literature search, which was limited to English language publications and was developed and carried out prior to the study selection process. In addition to the strategy outlined below, reference lists of retrieved articles were reviewed for potential studies.

Database	Edition or date searched	Search Terms ††
MEDLINE (includes in-process and other non-indexed citation) OVID Licensed Resource	2002–Jan 4, 2012	1 hearing loss/ or deafness/ or hearing loss, bilateral/ or hearing loss, conductive/ or hearing loss, functional/ or hearing loss, high-frequency/or hearing loss, mixed conductive-sensorineural/ or hearing loss, sensorineural/ or hearing loss, central/ or hearing loss, noise-induced/ or presbycusis/ or usher syndromes/ or hearing loss, sudden/ or hearing loss, unilateral/ 2 (PCHI or deaf* or auditory neuropathy).tw. 3 (hearing adj2 (loss or impairment)).tw. 4 1 or 2 or 3 5 mass screening/ or neonatal screening/ 6 (screen* or diagnos* or test or tests or testing).ti. 7 5 or 6 8 4 and 7 9 diagnostic techniques, otological/ or hearing tests/ or acoustic impedance tests/ or audiometry/ or audiometry, evoked response/ or audiometry, pure-tone/ or exp audiometry, speech/ or psychoacoustics/ or dichotic listening tests/ or recruitment detection, audiologic/ or otoscopy/ or vestibular function tests/ or caloric tests/ or electronystagmography/ 10 ((hearing or audiological) adj1 (assessment or screening)).tw. 11 exp Evoked Potentials, Auditory, Brain Stem/ or exp Otoacoustic Emissions, Spontaneous/ 12 (auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE).tw. 13 9 or 10 or 11 or 12 14 8 or 13 15 (accuscreen or capella or chartr or otoread or abris or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA).tw. 16 4 and 15 17 14 or 16 18 limit 17 to (yr="2006 -Current" and "all infant (birth to 23 months)") 19 exp Infant/

		20 (neonate or infant or newborn).mp. 21 19 or 20 22 17 and 21 23 limit 22 to yr="2006 -Current" 24 18 or 23 25 (cochlear implant* or otitis media).ti. 26 24 not 25 27 hearing loss/ or deafness/ or hearing loss, bilateral/ or hearing loss, conductive/ or hearing loss, functional/ or hearing loss, high-frequency/ or hearing loss, mixed conductive-sensorineural/ or hearing loss, sensorineural/ or hearing loss, central/ or hearing loss, noise-induced/ or presbycusis/ or usher syndromes/ or hearing loss, sudden/ or hearing loss, unilateral/ 28 (PCHI or deaf* or auditory neuropathy).tw. 29 (hearing adj2 (loss or impairment)).tw. 30 27 or 28 or 29 31 mass screening/ 32 (screen* or diagnos* or test or tests or testing).ti. 33 31 or 32 34 30 and 33 35 diagnostic techniques, otological/ or hearing tests/ or acoustic impedance tests/ or audiometry/ or audiometry, evoked response/ or audiometry, pure-tone/ or exp audiometry, speech/ or psychoacoustics/ or dichotic listening tests/ or recruitment detection, audiologic/ or otoscopy/ or vestibular function tests/ or caloric tests/ or electronystagmography/ 36 ((hearing or audiological) adj1 (assessment or screening)).tw. 37 exp Evoked Potentials, Auditory, Brain Stem/ or exp Otoacoustic Emissions, Spontaneous/ 38 (auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE).tw. 39 (conditioned oriented response* or COR or VRA or audiometry or behavi?ral audiogram*).tw. 40 35 or 36 or 37 or 38 or 39 41 40 and 30 42 34 or 41 43 limit 42 to yr="2002 -Current" 44 limit 43 to "preschool child (2 to 5 years)" 45 44 not (cochlear implant* or otitis media).ti. 46 26 or 45 47 exp "Costs and Cost Analysis"/ 48 (economic adj1 (evaluat* or analys* or study or studies or assess* or consequence*)).tw. 49 (cost-benefit or benefit-cost or cost effectiv* or cost utility).tw. 50 (cost minimization or cost minimisation or cost consequence* or cost offset*).tw. 51 ((cost or costs) adj2 analys*).tw. 52 "cost of illness".tw. 53 (cost* or economic* or expenditures or fiscal or pharmacoeconomic or

		spending).ti. 54 47 or 48 or 49 or 50 or 51 or 52 or 53 55 46 and 54 **59 results**
Embase	2002–January 23, 2012	1 hearing impairment/ or branchiootorenal syndrome/ or conduction deafness/ or exp congenital deafness/ or deafblindness/ or hearing loss/ or hypoacusis/ or mixed hearing loss/ or monaural hearing/ or morquio syndrome/ or norrie disease/ or exp perception deafness/ or sudden deafness/ or unilateral hearing loss/ 2 exp vestibulocochlear nerve disease/ 3 (PCHI or deaf* or auditory neuropathy).tw. 4 (hearing adj2 (loss or impairment)).tw. 5 1 or 2 or 3 or 4 6 mass screening/ or auditory screening/ or developmental screening/ or newborn screening/ 7 (screen* or diagnos* or test or tests or testing).ti. 8 6 or 7 9 5 and 8 10 auditory system examination/ or otoscopy/ or tonotopy/ or tympanometry/ 11 hearing test/ or exp audiometry/ or auditory screening/ or dichotic listening/ 12 ((hearing or audiological) adj1 (assessment or screening)).tw. 13 evoked brain stem auditory response/ 14 exp otoacoustic emission/ 15 (auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE).tw. 16 10 or 11 or 12 or 13 or 14 or 15 17 9 or 16 18 (ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA).tw. 19 5 and 18 20 17 or 19 21 limit 20 to yr="2006 -Current" 22 (neonate or infant or newborn).mp. 23 21 and 22 24 limit 21 to infant 25 23 or 24 26 25 not (cochlear implant* or otitis media).ti. 27 "COST"/ 28 exp Economic Evaluation/ 29 health economics/ 30 ((economic or cost*) adj2 (evaluat* or analys* or study or studies or assess* or consequence*)).tw. 31 ((cost-benefit or benefit-cost or cost effectiv* or cost utility) adj2 (analys* or evaluat* or assess* or study or studies)).tw.

		32 "cost of illness".tw. 33 27 or 28 or 29 or 30 or 31 or 32 34 26 and 33 35 limit 34 to english language **45 results**
Cochrane	2006–January 23, 2011	#1 MeSH descriptor Hearing Loss explode all trees #2 (PCHI or deaf* or auditory neuropathy):ti,ab,kw #3 (hearing NEAR/2 (loss or impairment)):ti,ab,kw #4 (#1 OR #2 OR #3) #5 MeSH descriptor Mass Screening, this term only #6 MeSH descriptor Neonatal Screening, this term only #7 (screen* or diagnos* or test or tests or testing):ti,ab,kw #8 (#5 OR #6 OR #7) #9 (#4 AND #8) #10 MeSH descriptor Diagnostic Techniques, Otological explode all trees #11 ((hearing or audiological) NEAR/1 (assessment or screening)):ti,ab,kw #12 MeSH descriptor Evoked Potentials, Auditory, Brain Stem, this term only #13 MeSH descriptor Otoacoustic Emissions, Spontaneous, this term only #14 (auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE):ti,ab,kw #15 (#10 OR #11 OR #12 OR #13 OR #14) #16 (#9 OR #15) #17 (ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA): ti,ab,kw #18 (#4 AND #17) #19 (#16 OR #18) #20 (neonate* or infant* or newborn*):ti,ab,kw #21 (#19 AND #20) #22 (#19 AND #20), from 2006 to 2012 **18 results**
Web of Science		# 17 (#16 AND #15) AND Language=(English) Databases=SCI-EXPANDED, SSCI, A&HCI, CPCI-S, CPCI-SSH, BKCI-S, BKCI-SSH Timespan=2006-2012 # 16 TI=(cost* OR economic* OR expenditures OR price OR fiscal OR pharmacoeconomic OR spending) # 15 #14 AND #13 # 14 TS=((neonate* or infant* or newborn*)) # 13 #10 or #12 # 12 #3 AND #11 # 11 TS=(ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA) # 10 #8 or #9 # 9 #5 OR #6 or #7 # 8 #4 AND #3 # 7 TS=((auditory brainstem response or ABR or AABR or otoacoustic

		emission* or OAE or AOAE or TEOAE or DPOAE)) # 6 TS=((hearing assessment* or audiological assesment* or hearing screening or audiological screening)) # 5 TS=(hearing test* or audiometry or psychoacoustics) # 4 TI=(screen* or diagnos* or test or tests or testing) # 3 #2 OR #1 # 2 TS=((hearing NEAR/2 (loss or impairment))) # 1 TS=((PCHI or deaf* or auditory neuropathy)) **14 results**
CINAHL		S28 S20 and S27 S27 S21 or S22 or S23 or S24 or S25 or S26 TI cost* OR economic* OR expenditures OR fiscal OR pharmacoeconomic Limiters - Published Date from: 20060101 20121231 S25 (MH "Economics") or (MH "Economics, Pharmaceutical") Limiters – Published Date from: 20060101-20121231 S24 (MH "Health Care Costs") Limiters - Published Date from: 20060101 20121231 S23 (MH "Economic Aspects of Illness") Limiters - Published Date from: 20060101-20121231 S22 (MH "Cost Benefit Analysis") Limiters - Published Date from: 20060101-20121231 S21 (MH "Costs and Cost Analysis") Limiters - Published Date from: 20060101-20121231 S20 S16 or S18 Limiters - Published Date from: 20060101-20121231 Narrow by SubjectAge: - Infant: 1-23 months Narrow by SubjectAge: - Infant, Newborn: birth-1 month S19 S16 or S18 S18 S5 and S17 S17 ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or ECHO or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA S16 S9 or S15 S15 S10 or S11 or S12 or S13 or S14 S14 auditory brainstem response or ABR or AABR or otoacoustic emission* or OAE or AOAE or TEOAE or DPOAE S13 (MH "Otoacoustic Emissions, Spontaneous") S12 (MH "Evoked Potentials, Auditory, Brainstem") S11 hearing assessment* or audiological assessment* or hearing screening or audiological S10 (MH "Diagnosis, Ear") OR (MH "Hearing Tests") OR (MH "Acoustic Impedance Tests") OR (MH "Audiometry+") OR (MH "Dichotic Listening Tests") OR (MH "Hearing Screening") OR (MH "Otoacoustic Emissions, Evoked") OR (MH "Otoscopy") OR (MH "Vestibular Function Tests+") S9 S5 and S8 S8 S6 or S7 S7 TI screen* or diagnos* or test or tests or testing S6 (MH "Health Screening") OR (MH "Neonatal Assessment")

		S5 S1 or S2 or S3 or S4 S4 (hearing loss or hearing impairment) S3 (MH "Auditory Neuropathy") S2 PCHI or deaf* or auditory neuropathy S1 (MH "Hearing Disorders") OR (MH "Deafness") OR (MH "Deaf-Blind Disorders") OR (MH "Usher's Syndrome") OR (MH "Hearing Loss, Partial") OR (MH "Hearing Loss, Conductive") OR (MH "Hearing Loss, Functional") OR (MH "Hearing Loss, High-Frequency") OR (MH "Hearing Loss, Sensorineural") OR (MH "Hearing Loss, Central") OR (MH "Hearing Loss, Noise-Induced") OR (MH "Pendred Syndrome") **78 results**
Grey Literature		
Guidelines		
AMA Clinical Practice Guidelines www.topalbertadoctors.org/TOP/CPG/	Dec. 20, 2011	Browsed list of topics **0 results**
NICE Guidance www.nice.org.uk/	Feb. 2, 2012	otoacoustic; "auditory brainstem"; "hearing screening" **0 results**
CALSPA www.caslpa.ca	Feb. 2, 2011	Browsed list **1 result**
ACSLPA www.acslpa.ab.ca/	Feb. 2, 2011	Browsed list **1 result**
CMA Infobase http://mdm.ca/cpgsnew/cpgs/index.asp	Dec. 20, 2011	Browsed list of publications **1 result**
National Guideline Clearinghouse www.ngc.gov	Dec. 20, 2011	(newborn* OR infant*) AND hearing AND screening **4 results**
Coverage/Regulatory/Licensing Agencies		
Alberta Health and Wellness www.health.gov.ab.ca	Jan.25, 2011	Infant +hearing +screening; newborn +hearing +screening **0 results**
Medical Devices Active Licence Listing www.mdall.ca/	Dec. 20, 2011	Newborn hearing or infant hearing Or AOE or ABR or otoacoustic or auditory brainstem **68 results**
Health Canada www.hc-sc.gc.ca	Dec. 20, 2011	Newborn hearing or infant hearing Or OAE or ABR or otoacoustic or auditory brainstem **0 results**
US Food and Drug Administration Databases www.accessdata.fda.gov/scripts/cdrh/devicesatfda/index.cfm	Jan. 25, 2012	ACCUSCREEN or CAPELLA or CHARTR or OTOREAD or ABRIS or TEOAE25 or DPOAE440 or ALGO or AUDX or ABAER or AURIX or ECHOCHECK or ECHOPORT or OTOPORT or ERO SCAN or GSI or AUDERA **18 results**

Aetna Clinical Policy Bulletins www.aetna.com/about/cov_det_policies.html	Jan 26, 2011	"otoacoustic emissions"; "auditory brainstem response" or "newborn hearing screening" or "infant hearing screening" **0 results**
HTA resources		
INESS www.inesss.qc.ca/	Jan. 26, 2012	otoacoustic; "auditory brainstem"; "hearing screening" **0 results**
CADTH www.cadth.ca/index.php/en/	Jan. 26, 2012	otoacoustic; "auditory brainstem"; "hearing screening" **4 results**
Institute for Clinical and Evaluative Sciences (ICES), Ontario www.ices.on.ca/	Feb 2, 2012	Browsed list **0 results**
Health Technology Assessment Unit At McGill www.mcgill.ca/tau/	Feb 2, 2012	Browsed list **0 results**
Medical Advisory Secretariat www.health.gov.on.ca/english/providers/program/mas/mas_mn.html	Feb. 2, 2012	Browsed list **0 results**
Other Grey Literature Sources		
Proquest Dissertations and Theses	Feb 2, 2011	Otoacoustic or auditory brainstem or hearing screening AND (infant or newborn or neonate) **0 results**
Internet Search Engines		
Google		"hearing screening" cost OR economic OR expenditures OR fiscal OR spending infant OR newborn OR neonate OR otoacoustic "auditory brainstem" –pubmed **5 results**
NHS Evidence	Feb 2, 2011	hearing screening AND (infant or newborn or neonate) otoacoustic auditory brainstem **13 results**

Note:

†† "*", "# ", and "?" are truncation characters that retrieve all possible suffix variations of the root word, for example, surg* retrieves surgery, surgical, surgeon, etc.

Search Strategy:

Searches Results

APPENDIX E.2: SUMMARIZED EVIDENCE FROM SELECTED STUDIES

#	Item	Description
1	Study[2]	Authors/publish year: Uus et al., 2006; country: UK; study type: CEA; Setting: the community and hospital; study perspective: society
	Objective	The study examined the costs and cost effectiveness of the Newborn Hearing Screening Programme (NHSP), compared with the Infant Distraction Test Screen (IDTS)
	Population	Newborn babies
	Intervention	NHSP, which takes place either in the maternity hospital or in the community very soon after birth, is based on transient evoked otoacoustic emissions (TEOAE), followed by automated auditory brainstem response (AABR) if no clear response from TEOAE. IDTS, performed at around 8 months of age, consists of localization responses to low-level sounds presented to child by a tester while a second tester suitably manipulates the child's attention.
	Time Horizon/discount rate	Short time/NA
	Currency/ price year	£/2003
	Outcomes measure	Cases detected
	Cost components	Staffing, equipment, overheads, staff training and travel, and audiology costs
	Results	
	Outcomes	For 1000 infants screened, 1.06 and 0.36 cases detected with NHSP and IDTS, respectively
	Costs	For 1000 infants screened, the healthcare cost and total cost (including family cost) were £34,315 and £37,383 with NHSP, and £25,171 and £42,459 with IDTS. The healthcare cost and total cost (including family cost) per case detected were £31,410 and £34,826, respectively, with NHSP, and £69,919 and £117,942, respectively, with IDTS.
	Marginal Analysis	NHSP costs the health system an additional £12,500 for each case detected compared with IDTS.
	Conclusion	NHSP is less costly, and the study supports replacing IDTS with NHSP.
2	Study[4]	Authors/publish year: Uilenburg et al., 2009; country: Netherlands; study type: CEA; Setting: baby clinic and community; study perspective: NA
	Objective	This study examined the clinical efficacy, costs, and cost effectiveness of UNHS in three different settings.
	Population	Healthy newborn babies
	Intervention	The UNHS was a three-stage transient evoked otoacoustic emissions screening, with a time interval of about 1 week between stages. The three settings under investigation were: 1) at home in combination with metabolic diseases screening, within 4 to 7 days of birth; 2) at home in combination with the first home visit, within 3 weeks of birth and (3) in the baby clinics in a special screening session, within 4 weeks of birth.
	Time Horizon/discount rate	Short time/NA
	Currency/ price year	€/not stated
	Outcomes measure	Rate of participation and refer rate
	Cost components	Not reported

	Results	
	Outcomes	The participation rate and refer rate were 88.9% and 1.4% for at-home screening combined with metabolic diseases screening, 86.9% and 2.7% for at-home screening during an intake visit, and 75.2% and 2.3% for screening in baby clinics. Cases detected were not reported.
	Costs	The costs per baby screened were €20.5 for screening in baby clinics, €28.3 for at home screening combined with metabolic diseases screening, and €41.7 for at-home screening during an intake visit. Costs per detected baby were €35,100, €41,500 and €61,800, respectively, in the three settings.
	Marginal Analysis	The authors did not report the extra costs per baby detected, but stated that the most cost-effective setting was the at-home screening combined with the metabolic diseases screening.
	Conclusion	The UNHS combined with the screening for metabolic diseases is a preferred option.
3	Study[5]	Authors/publish year: Lin et al, 2007; country: Taiwan; study type: CEA; Setting: hospital; study perspective: NA
	Objective	The study compared the efficacy, initial referral rate, costs, and cost effectiveness of three newborn hearing screening protocols.
	Population	Newborn babies
	Intervention	The three protocols were transient evoked otoacoustic emission (TEOAE) alone, TEOAE plus automated auditory brainstem response (AABR), and AABR alone.
	Time Horizon/discount rate	Short time/NA
	currency/ price year	US$/not stated
	Outcomes measure	Referral rate, identification rate of congenital deafness
	Cost components	Equipment, salary, initial screening, and diagnostic follow-up tests
	Results	
	Outcomes	The referral rates were 5.8%, 1.6% and 0.8% with TEOAE alone, TEOAE plus AABR, and AABR alone, respectively. AABR alone was associated with a significant decrease in referral rate compared to the others. The accurate identification rate of congenital hearing loss was 0.45%, 0.25% and 0.42% with TEOAE alone, TEOAE plus AABR, and AABR alone, respectively.
	Costs	The total direct costs per screening were $10.04, $8.60, and $7.33 with TEOAE alone, TEOAE plus AABR, and AABR alone, respectively. The intangible costs, including parental anxiety, transportation fees, and days off during follow-up tests, were lowest with AABR alone.
	Marginal Analysis	The authors did not report the extra costs per baby detected, but stated that AABR alone was the most cost effective.
	Conclusion	AABR alone significantly reduces the referral rate, and is therefore much less costly.
4	Study[6]	Authors/publish year: Olusanya et al., 2009; country: Nigeria; study type: CEA; Setting: hospital and community; study perspective: NA
	Objective	The study examined the efficacy and costs of hospital- and community-based infant hearing screening programs, with a focus on universal and targeted newborn infants.
	Population	Universal newborn and targeted newborn infants. The infants were defined as high risk if they were admitted into the special care baby unit, born outside hospital facilities, or had a history of neonatal jaundice.

Intervention	A hospital-based screening program was compared with a community-based program. Both programs consisted of two-stage screening with transient evoked otoacoustic emissions (TEOAE) and automated auditory brainstem response (AABR), followed by diagnostic evaluation.
Time Horizon/discount rate	Short time/NA
Currency/ price year	UN$/ not stated
Outcomes measure	The referral rate and detected number of instances of permanent congenital and early-onset hearing loss (PCEHL)
Cost components	The cost components included staff salary, equipment, consumables, transportation to screening sites, and diagnostic tests.
Results	
Outcomes	The first-stage referral rates for universal and targeted infants were 32.2% and 31.7%, respectively, for hospital-based screening, and 14.3% and 15.2%, respectively, for community-based screening. The second-stage referral rates for universal and targeted infants were 3.3% and 4.4%, respectively, for hospital-based screening, and 4.1% and 4.9%, respectively, for community-based screening. The detected number of PCEHL per 1000 universal and targeted infants was 5.3 and 16.7, respectively, for hospital-based screening, and 22.5 and 27.4, respectively, for community-based screening.
Costs	The screening costs per infant were $13.30 and $73.24, respectively, for hospital-based universal infant and targeted screening, and $7.62 and $12.27, respectively, for community-based universal infant and targeted screening. The costs per infant detected with PCEHL were $2,764.86 and $4,631.33, respectively, for hospital-based universal infant and targeted screening, and $602.49 and $714.53, respectively, for community-based universal and targeted screening.
Marginal Analysis	Not performed
Conclusion	Community-based screening for universal newborn infants was identified as the most cost-effective option.

APPENDIX E.3: QHES INSTRUMENT

#	Questions	QHES Scores			
		Uus et al., 2006	Uilenburg et al., 2009	Lin et al., 2007	Olusanya et al., 2009
1	Was the study objective presented in a clear, specific, and measurable manner?	7	7	7	7
2	Were the perspective of the analysis (societal, third-party payer, etc.) and the reasons for its selection stated?	4	0	0	0
3	Were variable estimates, used in the analysis from the best available source (i.e., randomized control trial—best; expert opinion—worst)?	4	4	4	4
4	If estimates came from a subgroup analysis, were the groups pre-specified at the beginning of the study?	1	1	1	1
5	Was uncertainty handled by: (1) statistical analysis to address random events; and (2) sensitivity analysis to cover a range of assumptions?	4.5	4.5	4.5	4.5
6	Was incremental analysis performed between alternatives for resources and costs?	6	0	0	0
7	Was the methodology stated for data abstraction (including the value of health states and other benefits)?	5	5	5	5
8	Did the analytic horizon allow time for all relevant and important outcomes? Were benefits and costs that went beyond 1 year discounted (3% to 5%), with justification given for the discount rate?	7	7	7	7
9	Was the measurement of costs appropriate and the methodology for the estimation of quantities and unit costs clearly described?	4	0	4	4
10	Were the primary outcome measure(s) for the economic evaluation clearly stated and did they include the major short-term was justification given for the measures/scales used?	6	6	6	6
11	Were the health outcomes measures/scales valid and reliable? If previously tested valid and reliable measures were not available, was justification given for the measures/scales used?	7	7	7	7
12	Were the economic model (including structure), study methods and analysis, and components of the numerator and denominator displayed in a clear, transparent manner?	8	8	8	8
13	Were the choice of economic model, main assumptions, and limitations of the study	7	7	0	7

	stated and justified?				
14	Did the author(s) explicitly discuss direction and magnitude of potential biases?	0	0	0	0
15	Were the conclusions/recommendations of the study justified and based on the study results?	8	8	8	8
16	Was there a statement disclosing the source of funding for the study?	3	3	3	3
	TOTAL POINTS	**81.5**	**67.5**	**64.5**	**71.5**

REFERENCES

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Author Contribution Statements

Paula Corabian contributed to study conception and design, data analysis and interpretation, and approved the final version for publication.

Christa Harstall contributed to study conception and design, revision of manuscript for critical content, and approved the final version for publication.

Charles Yan contributed to study conception and design, statistical analysis, economic expert review of the literature, revision of manuscript for critical content, and approved the final version for publication.

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Dagmara Chojecki developed and executed the literature search.

This report is an update of a 2007 report. Permanent congenital hearing impairment/loss (PCHI/PCHL) is one of the most common congenital anomalies found at birth which can lead to delays and deficits in the development of speech, language, cognition and learning, as well as secondary effects on the child and the family. Early identification and subsequent appropriate intervention (within the first 6 months) in infants with PCHL can minimize these effects. The report also compares the cost effectiveness of various strategies used in the universal screening of newborn hearing (UNHS)

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ISBN 978-1-926929-11-8 (on-line)