



Alberta College of
Speech-Language Pathologists
and Audiologists

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Position Statement

Endorsement of Universal Newborn Hearing Screening (UNHS) in Alberta

April 2008

ENDORSEMENT OF UNIVERSAL NEWBORN HEARING SCREENING (UNHS) IN ALBERTA

An ACSLPA Position Statement sets out the official position or stand of the College on an issue or matter that is significant not only to the professions of speech-language pathology and/or audiology, but also to outside agencies or groups. It is recommended that the practice of College members will be consistent with Position Statements.

EXECUTIVE SUMMARY

Hearing loss is one of the most common major conditions present at birth and occurs more frequently than any other condition that can be, or is currently, screened at birth. The Alberta Health and Wellness newborn metabolic screening program was recently expanded to screen for 17 metabolic conditions in newborns, yet none of these conditions has a prevalence exceeding one in 3000 births. The prevalence of hearing loss in newborns and infants ranges from one to six per 1000 live births.

Left undetected, hearing impairments in infants can negatively affect speech and language acquisition, development of reading and writing skills and academic achievement, as well as social and emotional development. Early identification of hearing loss followed by timely and appropriate intervention can significantly reduce and even eliminate the negative consequences of hearing loss for the individual, the family and society.

Universal newborn hearing screening (UNHS) is a recommended strategy for identifying infants born with a hearing loss soon after birth. Early identification allows for earlier intervention, which can enhance a hearing impaired child's speech, language and social development. An accurate and objective hearing screening test on a newborn can be completed in less than five minutes; the typical quoted cost is about \$35 CAD per screened infant. Every case of unidentified hearing loss has been estimated to cost taxpayers \$1 million CAD. The costs of screening are directly offset by reduced expenditures on special education and support programs.

An increasing number of European countries have UNHS or are in the process of implementing UNHS as a standard practice for newborn health care. In the United States, 40 states have legislated newborn hearing screening programs and five states have voluntary programs with a targeted screening rate of 95%. Unfortunately, there is not a systematic approach to early identification, diagnosis and management of hearing loss in children in every province in Canada. Although some routine hearing screening is practiced in every province, it has not been universally established.

A 2001–2004 pilot UNHS program in four Alberta health regions (Mistihia/Peace Country, Palliser, Chinook and Calgary) indicated that four in every 1000 screened infants were identified with a potential hearing loss. Government funding for the UNHS program did not continue beyond the pilot project period. However, based on the success of the program, some health regions have voluntarily continued the program.

Recognizing the benefits of UNHS to all Albertans, the Alberta College of Speech-Language Pathologists and Audiologists (ACSLPA) strongly recommends the establishment and maintenance of an integrated and consistent UNHS program in Alberta. ACSLPA supports the recommendations of the Canadian Association of Audiologists (CAA), the Canadian Association of Speech-Language Pathologists and Audiologists (CASLPA), the Hearing Foundation of Canada, the American Joint Committee on Infant Hearing, the American

Academy of Pediatrics and the National Institutes of Health in developing and maintaining UNHS programs that enable confirmation of hearing loss by three months of age, and enrolment in a family-centered intervention program by six months of age.

NEWBORN AND INFANT HEARING: THE ISSUE

INTRODUCTION

Hearing impairment is defined as a permanent bilateral or unilateral hearing loss of any degree that is sensory, neural or conductive in origin.¹ Permanent bilateral hearing impairment in early childhood often negatively influences the development of speech, language, cognitive and psychosocial skills and, subsequently, literacy and academic achievement. The adverse affects of hearing loss on language and cognitive development, as well as psychosocial behaviour, are well established.^{2,3,4,5} Hearing impairment places severe limitations on educational and economic pursuits essential to achieving a positive quality of life.⁶

In addition to the loss of localization abilities and poor communication skills in the presence of background noise, studies have suggested that early-onset severe unilateral (i.e. hearing loss in one ear) sensorineural hearing impairment (USNHI) in children is also associated with significant deficits in auditory and psycholinguistic skills and school performance.^{7,8,9} According to Lieu,¹⁰ a literature search from 1966 to 2003 revealed a 22-35% failure rate of one grade in school for children with USNHI; 12-41% of these children received educational assistance.

RATIONALE

The urgency of securing language input for an infant born with any degree of hearing loss is the impetus for hearing screening programs.¹¹ Screening infants identified as high risk for hearing loss is an important preliminary step; however, two-thirds of all children born with permanent congenital hearing impairment (PCHI) are in the well-baby population¹² and do not have any other handicapping conditions.

Newborn and infant hearing screening programs provide the opportunity for timely intervention and early auditory stimulation. Hearing screening programs lower the age of hearing loss identification, reduce the age of initiation of intervention, and produce significantly improved outcomes for both the child and family.^{3,13,14} Universal newborn and infant screening is best complemented by a system of ongoing surveillance throughout infancy and early childhood^{1,15} to ensure that progressive, late onset and acquired hearing losses are also identified as early as possible.¹⁶

Although identification of hearing loss through screening is only the first step toward delivering services to infants with hearing loss, it provides the thrust for the implementation and maintenance of diagnostic, intervention and management components of early hearing detection and intervention programs. "Only through comprehensive identification will the need for early intervention programs be realized."¹⁷ Overall, the literature indicates that in the absence of systematic screening, the detection, confirmation, diagnosis and management of hearing impairment is significantly delayed. The impetus to screen while in hospital is intended to prevent the loss of infants to follow-up after discharge due to compliance (e.g. low priority or parental concern) and other factors (e.g. travel in rural areas).

PREVALENCE

Hearing loss is one of the most common major conditions present at birth and occurs more frequently than any other condition (e.g. phenylketonuria, hypothyroidism) that can be, or is

currently being screened.¹⁸ The prevalence of hearing loss in newborns and infants ranges from one to six per 1000 live births,^{19,20,21} depending on the threshold of permanent hearing impairment used.^{22,23,24,25} In children 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.^{26,27} The neonatal intensive care unit (NICU) population has a 10 to 20 times higher risk of having permanent congenital hearing impairment than the well-baby population.²⁸ It is estimated that there are up to 1100 new cases of hearing loss in newborns annually in Canada,²⁴ with some researchers placing the figure closer to 2000.²⁹

The Alberta Health and Wellness newborn metabolic screening program was recently expanded to screen for 17 metabolic conditions in newborns.³⁰ None of these conditions has a prevalence exceeding one in 3000 births, compared to the prevalence of hearing loss in infants, which is two to three in 1000 births. The Alberta UNHS Pilot Project Program found prevalence for hearing loss in Alberta infants of four in 1000 screened.³¹ Given an annual birth rate of 44,661 in Alberta,³² an estimated 88 to 179 newborns with hearing impairment would be identified per year, based on prevalence rates of two to four per 1000 live births.

Alberta became the first province in Canada to provide universal screening for the identification of cystic fibrosis, which occurs in one in 3300 live births in Alberta. Cystic fibrosis does not have a cure, but early identification will ultimately provide the affected individual with a better quality of life.^{33,30} Early identification of congenital or chronic hearing impairment is similarly a quality of life issue. It is identifiable at an early asymptomatic stage, and can be medically treated and/or habilitated with amplification and behavioural therapy.

BENEFITS OF EARLY IDENTIFICATION

There is growing evidence-based consensus that UNHS and appropriate follow-up services will improve hearing and communication development in children.^{5,24}

Early identification of hearing loss can significantly reduce the negative consequences of hearing loss for the individual, the family and society.^{4,34,35,36} Theoretical research on auditory and cognitive plasticity has suggested that early auditory stimulation is most effective for developing a child's auditory and cognitive potential.² Research found that the most significant factor in ultimate levels of language achievement is the timing of intervention. Intervention was found to be much more effective when initiated in the first six months of life for Colorado infants born in hospitals with UNHS compared to those without UNHS.^{4,25} Recent international research from the United Kingdom also confirms the significant impact of early identification (i.e. by nine months of age) on language skills at age eight.⁵ UNHS results in a median age of diagnosis of hearing loss of less than three months.²³

Hearing loss will affect a child's understanding and use of language, depending on the severity and the timing of the onset of the impairment. Longitudinal studies are starting to be published on language development in early-identified infants. Studies investigating speech (articulation) development have not found significant differences between early- and later-diagnosed hearing loss in school-age children, but the language development differences continue to be noted into school age,⁵ indicating more impact on language development than speech development in children identified through UNHS programs.

Early language development provides the foundation for the development of reading and writing skills.³⁷ Children who are deaf or hard of hearing not only do not have adequate access to the spoken word or phonological code, which is necessary for sound-letter association in reading the written word at an early age, but they also may not participate in the same literacy experiences as those children who have normal hearing. For example, a parent may not read to the child due to lack of positive feedback from the child, may not be comfortable signing while

reading, or may not be able to establish visual contact with the child while reading.³⁸

Between 40-75% of preschoolers with early language impairment will have reading difficulties later.³⁹ According to the Edmonton Public School Board, many children who are deaf or hard of hearing often learn to read and write while simultaneously learning their first language, and do not have the same foundation established by the time they start school as children with normal hearing. They suggested that children who are deaf or hard of hearing might not develop independent reading strategies (e.g. self-questioning, summarizing the main idea, predicting what text will follow, constructing representational images, etc.). Gallaudet University followed children over a 30-year period, and found that half of the children with hearing loss graduated from high school with a fourth grade reading level or less.⁴⁰ Early identification of hearing-impaired children and early intervention to begin teaching them symbolic language can be paramount for later achievement.⁴¹

The benefit of early identification of hearing loss to hearing impaired children, their families and our society has been resolved in professional, academic and research communities. Research has demonstrated that early identification and intervention for hearing impairment positively influences auditory cortical development. Benefits to speech, language, social, emotional, literacy and cognitive development have also been measured. Like all modern research endeavors involving human participants, research in this area continues to progress within the appropriate limitations of ethical practice. The prospective randomized controlled trials, considered a research “gold standard”, are often difficult or impossible to complete in this context given the ethical implications. Their absence in this area does not negate the significant research that has been done.

Auditory research supports evidence that suggests that a shorter duration of deafness prior to cochlear implantation improves the likelihood of normal cortical development and, consequently, the normal development of language skills in children. Researchers found that auditory stimulation is required in order for the auditory system to mature in children with cochlear implants. The P1 peak of the cortical-evoked potentials showed latency changes shortening to the adult values with increased exposure to sound for cochlear-implanted children. In a cross-sectional group study, auditory cortical-evoked potentials matured following the same rate as normal hearing children, even after a long period of auditory deprivation.⁴² However, longitudinal examination of a few cochlear implant children found that P1 latencies for implanted children tended to be prolonged and did not approach normal values at their maturational completion. Therefore, the impact of cochlear implantation may be limited by the age of onset and duration of deafness.⁴³ These auditory cortical plasticity findings support early identification of hearing loss.

SUPPORT FOR NEWBORN HEARING SCREENING PROGRAMS

Technological advances enabling easier and more cost-effective identification, and growing research evidence of the importance and benefits of early intervention for hearing loss, have resulted in a substantial increase in newborn hearing screening in Canada,²⁴ the United States⁴⁴ and the United Kingdom.²⁰ Approximately 1,700 newborns are screened each day in the United Kingdom,⁴⁵ and approximately 600 per year are identified as having a bilateral hearing loss.

An increasing number of European countries have UNHS or are in the process of implementing UNHS as standard practice for newborn health-care.^{20,46,47} Development and implementation of sophisticated physiological hearing screening techniques has contributed to the feasibility of UNHS in many different countries.¹⁵

Support for UNHS, early diagnosis and early intervention is growing steadily as the identification of hearing loss in newborns improves through the development of rapid, valid, reliable and cost-

effective technology based on the use of objective physiological measures.²³ An accurate and objective hearing screening test on a newborn can be completed in less than five minutes. The instrumentation is portable, simple to operate, and the screening “pass” or “fail” determination is made automatically.

False positive outcomes have been reported as less than 2% in successful UNHS programs in the United States.^{23,48} Forty states with legislated newborn hearing screening programs and five states with voluntary programs targeted a screening rate of 95%.⁴⁹ In 2005, 92.8% of these newborns had newborn hearing screening.⁵⁰ However, almost half of the newborns who fail the screen do not have appropriate follow-up to confirm the presence of hearing loss and/or initiate early intervention services.²⁷

Canadian researchers support the importance of identification of hearing impairment and early intervention as the goal of UNHS. Hyde reported that universal screening lowers the age of diagnosis, which leads to earlier intervention and earlier access to hearing.²⁴ The Canadian Working Group on Childhood Hearing (CWGCH) has recommended that the link between screening and improved speech and language development should not be the only justification for UNHS, as all children should have the basic right to hear and access resources to facilitate hearing.²³

Durieux-Smith and Whittingham demonstrated that Ontario children systematically screened in infancy are diagnosed by six months of age.⁵¹ American research has shown that children with bilateral hearing loss who are not screened are generally identified by 2.5 years of age. Children with USNHI are not typically identified until they reach school age. The average age of identification of a USNHI of any degree (e.g. mild to profound) ranges from four years-11 months⁵² to 8.78 years.⁹ “Twenty percent of USNHI are identified at three to four years of age; 50% at five to six years of age; and another 20% at seven to eight years of age⁸.” These statistics are likely similar for Canada. “Newborn hearing screening accounted for 26.7% of all children identified before six years of age and 10% of all identifications. The percentage of children identified this way is likely to increase with implementation of universal newborn hearing screening programs.”⁵²

Canadian federal funding allocated to early childhood development was reported in the First Ministers Communiqué, dated September 11, 2000.⁵³ The Early Childhood Development Agreement earmarked a large portion of the social transfer of federal monies to the provinces specifically for the purpose of early childhood development in four key areas: services to expectant parents, new parents and infants; parenting supports and parenting skills programs; quality programming in childcare settings; and community capacity-building to effectively plan with government and non-government partners. The federal government pledged to continue that agreement to 2013. In 2007, Ontario received just under \$200 million in funding and Alberta received more than \$50 million. None of this money was earmarked for UNHS in either province.

COST

Based on recent extensive data from the United States, direct costs per diagnosed case are comparable to those for other screened congenital anomalies. The typically-quoted cost of about \$35 CAD per screened infant is higher than for blood tests, but because of the much higher incidence rate for hearing loss, the typical cost per identified case (\$14,400 CAD) is much lower than for screening for phenylketonuria (\$60,750 CAD)¹⁶ and cystic fibrosis (\$15,435 US).⁵⁴ In addition, the costs of screening are directly offset by reduced expenditures on special education and support programs.^{16,18} Other studies have investigated the cost of one- and two-stage screenings using auditory brainstem response (ABR) and otoacoustic emission (OAE) testing.²³ According to the Alberta Health and Wellness Synthesis Report, “health economists

... found that the one-stage automated auditory brainstem response (AABR) is the cost effective alternative to the one-stage automated otoacoustic emissions (AOAE) protocol. The two-stage protocol (involving initial screen with AOAE and repeat screen with AABR) was more effective (better specificity >97%), but at a higher expected cost compared to the one-stage AABR.⁵⁵

From a financial point of view, every case of unidentified hearing loss has been estimated to cost taxpayers \$1 million dollars CAD.⁵⁶ While a UNHS program is not considered cost-effective in the first year, net savings are projected by the fourth year, which would ultimately save taxpayers \$7 billion in one generation.⁵⁷

NEWBORN AND INFANT HEARING SCREENING IN CANADA

Hyde estimated that two to three in 1000 infants have congenital hearing loss that merits early detection.²⁴ Based on the 352,848 babies born in one year in Canada from July 1, 2006 to June 30, 2007,³² it can be estimated that anywhere from 1000 to 1400 newborns will have some degree of congenital hearing loss. According to Statistics Canada, preliminary figures show that 44,661 babies were born in Alberta in the same time period noted above, accounting for almost 13% of all the babies born in Canada.

Brown reported that Canada lagged behind Europe and the United States in UNHS programs.²⁹ The Canadian Academy of Audiology (CAA) and the Canadian Association of Speech-Language Pathologists and Audiologists (CASLPA) developed a position statement¹⁶ which stated that universal infant hearing programs are an important step in ensuring the hearing health of Canadians.

Unfortunately, there is no systematic approach to early identification, diagnosis and management of hearing loss in children in all provinces in Canada. A Canadian research survey of birthing hospitals in Canada in 2000 indicated that only 10% of the hospitals that responded reported some kind of hearing screening activity.⁵⁸ The majority of the hearing screening programs screened only infants who fit certain high-risk criteria; however, studies have found that approximately 50% of newborns with hearing loss do not have high risk factors.²⁹ Since Health Canada established CWGCH in 2000, infant hearing screening programs are on the rise in Canada. Loss to follow-up is the largest limiting factor of UNHS in Canada.²³

The Newborn Screening in Canada Status Report through the Canadian Organization for Rare Disorders (CORD) synthesized provincial activity and legal requirements for all types of newborn screening including hearing.⁵⁹ CORD's findings illustrated an array of various screening activities depending on the province. Although there is some routine hearing screening practiced in every province, it is not universally established. The current status of infant hearing screening in all other provinces and territories in Canada is summarized in the following chart (see next page).

Province/Territory	Newborn Hearing Screening Status
British Columbia	British Columbia implemented screening for babies in the NICU with stays over 48 hours in 2007, and started a phased implementation of screening well babies in October 2007. It was planned that by the end of 2008, all babies in the province will be screened.
Saskatchewan	There is a pilot proposal for UNHS for the Saskatoon health region only. Some high-risk screening is currently conducted in the Saskatoon health region in coordination with NICU neonatologists, but there is no consistent special care nursery screening, or UNHS in Saskatchewan.
Manitoba	Manitoba does not have a provincial-wide program in place. There is UNHS with limited funding based out of only two regional health authorities, which started as pilot projects. A home-based preschool aural rehabilitation program is offered through one regional health authority.
Ontario	Since implementation in 2002, the Ontario Infant Hearing Program (IHP) has offered hearing screening to newborns in all birthing hospitals. The IHP provides universal newborn hearing screening, surveillance for those at risk for developing hearing impairment in early childhood, audiology assessment, hearing aid selection and follow-up audiology visits and communication/language development services for children identified with permanent hearing impairment until Grade 1 entry. Implementation of a remote computer system has begun that will enable an audiologist to perform diagnostic ABR, from an urban centre, in locations where there is no audiologist available.
Quebec	<p>The Ministry of Health mandated the Public Health Institute of Quebec to draft a recommendation concerning newborn hearing screening. The Expert committee filed a recommendation for UNHS and early intervention to the Minister and will be reported on the L'Institut National de Santé Publique du Québec (INSQP) website (www.inspq.qc.ca) in the near future. The Minister's response is unknown at present time. Other statistics reported were as follows:</p> <ul style="list-style-type: none"> • 57% of births occur in centres offering some form of newborn hearing screening. • 36% of birthing centres offer newborn hearing screening. • 9.1% of births occur in birthing centres offering universal screening. • 47.9% of births occur in birthing centres offering selective screening.
New Brunswick	UNHS has operated in each of the New Brunswick health authorities since 2002.
Nova Scotia	Nova Scotia's "A Sound Start Campaign" program was to be implemented in 2007 in all 10 birthing centres. Programs in eight hospitals were expanded in 2007, and the remainder targeted for early 2008.
Prince Edward Island	The two hospitals that provide obstetrical service have provided UNHS programs since 2005.
Newfoundland Labrador	The Canadian Hard of Hearing Association in Newfoundland and Labrador raised funds for screening equipment to run a UNHS program without support from their provincial government. This program began approximately six to seven years ago. UNHS now exists in eight of the 11 birthing hospitals in the province, and 3 other hospitals perform high-risk screenings with the intention of implementing UNHS in the near future.
Northwest Territories	UNHS has been provided since 2004 for babies in Yellowknife and Inuvik with a screening goal of 95%.
Yukon	UNHS has 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	Since there are no birthing hospitals in Nunavut, expectant mothers are flown to the nearest province or territory. Therefore, it is dependent on the birth hospital as to whether or not a baby's hearing is screened.

NEWBORN AND INFANT HEARING SCREENING IN ALBERTA

Hearing loss in half of affected infants is of unknown etiology and is not identified by the use of a comprehensive list or high risk register.¹⁶ Targeting “high risk” populations has also proven ineffective in the province of Alberta. The Alberta UNHS Project reported that, in 1998, only eight out of a potential 227 hearing impaired children were identified between birth and three years of age.⁶⁰ Prior to Spring 2001, only two of the province's birthing hospitals offered screening programs and only infants identified as high risk were screened.

In Spring 2000, a team of researchers from the University of Calgary were awarded a grant through the Alberta Health and Wellness Innovation Fund to implement and assess the efficacy of UNHS in Alberta. The Palliser, Mistahia (which became Peace Country), Chinook and Calgary health regions participated in the project. Screening began in Spring 2001. The Alberta UNHS Project used a two-stage, two-technology approach. Stage one involved OAE screening of all infants with a hand-held device. If normal hearing was not confirmed, the infant was flagged for follow-up screening. This second screening was completed with the same equipment. If normal hearing was not confirmed on this second screening, the infant was tested with AABR either in the hospital or in a regional outpatient facility. When responses were not obtained within the normal limits, the infant was then referred for further evaluation. This technique was intended to provide the lowest or most accurate “refer” rate.

Statistics to March 31, 2003 indicated that 14,348 newborns were screened in the pilot project, with 82 referred for diagnostic testing and 49 confirmed as having hearing loss.⁶⁰ All 49 newborns received early intervention and/or medical treatment, with one receiving a cochlear implant at less than one year of age.⁶¹ Preliminary results demonstrated that UNHS can be delivered in a timely and cost-effective manner.²⁹ The program ended in December 2004 with a total of 19,930 newborns screened. Alberta did not continue funding for newborn hearing screening beyond the pilot project period.

Currently, many Alberta health regions do not have the infrastructure or expertise to operate an infant screening program in any capacity. Others have the necessary equipment and/or adequate staff, but still do not have a universal screening protocol, and rely on high-risk data or parental request. The regions involved in the pilot project continue to screen babies included on the high-risk register without further funding support.

ACSLPA ENDORSEMENT OF UNIVERSAL NEWBORN HEARING SCREENING

ACSLPA strongly supports the establishment and maintenance of an integrated and consistent UNHS program for Alberta. The goal of this program is for all children 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.

The program should include:

- universal screening (using physiological methods) of all newborns born in the province of Alberta.
- appropriate, accessible services for diagnosis, hearing and communication development options.
- a seamless transition for infants and families through the process of screening, confirmed diagnosis by three months and early intervention by six months.
- ongoing surveillance throughout infancy and early childhood of those children at risk for developing hearing loss.
- education for parents, primary caregivers and health care providers on the early signs of hearing-impairment and risk factors associated with a hearing loss.
- early intervention with an assigned point of entry and intervention options.
- multidisciplinary teams of professionals that work closely with families.
- continuing education opportunities for audiologists, educators and health professionals to achieve and maintain expertise in the fitting of amplification in infants and in parent-infant habilitation strategies.
- implementation of a uniform Alberta provincial registry that could be integrated with inter-provincial and territorial registries, and a national program database. The data management aspect of the system is critical to assess and monitor the quality and efficacy of the processes (e.g. screening, evaluation and intervention), and to ensure that the program is stable and sustainable.

In summary, ACSLPA supports the recommendations of the Canadian Association of Audiologists (CAA),¹⁶ the Canadian Association of Speech-Language Pathologists and Audiologists (CASLPA),¹⁶ The Hearing Foundation of Canada,⁶² the American Joint Committee on Infant Hearing,^{1,27} the American Academy of Pediatrics⁶³ and the National Institutes of Health¹⁵ in developing and maintaining UNHS programs that enable confirmation of hearing loss by three months of age, and enrollment in a family-centered intervention program by six months of age. This can only be achieved through the establishment of a well-integrated and structured system of early identification and management for all infants who have hearing loss. ACSLPA supports continued research in the development of more efficient, simple, reliable and accurate methods for detecting and managing hearing loss in newborns and infants.

GLOSSARY^{64,65,66}

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

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

Behavioural: pertaining to externally observable activity of a person. Used here in context of therapy or intervention/rehabilitation to promote normal development of speech and language.

Cochlea: auditory portion of inner ear (as opposed to balance portion); consisting of fluid filled channels; location of other structures (e.g. outer hair cells, etc. - see otoacoustic emissions or OAEs)

Cochlear implant (CI): device enabling persons with profound hearing loss to perceive sound; consisting of an electrode array surgically implanted in cochlea and an external amplifier, which activates the electrode; delivers electrical signals to CN VIII or hearing nerve.

Cognitive or cognition: the process involved in knowing, including perceiving, recognizing, conceiving, judging, sensing and reasoning.

Congenital: present at birth.

Cortex: outer layer (e.g. the brain).

Cortical: of or pertaining to the cerebral cortex.

Etiology: the study of the causes of a disease or condition.

Evoked Potential (EP): electrical activity of the brain (and different structures; brainstem, cortex, etc.) in response to sensory (e.g. auditory) stimulation (see ABR, P1, etc.)

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

Latency: time interval between two events (e.g. 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 (once being identified with or at risk of hearing impairment) due to factors such as low compliance from parent, moved to another province, lack of services available, no tracking systems in place, etc.

Metabolic (metabolism): in reference to a sum of physical and chemical processes that maintain a living organism. Abnormal conditions may include: cystic fibrosis, PKU, congenital hypothyroidism, etc. Alberta screens for 17 “metabolic disorders”.

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.

P1: auditory evoked potential recorded after stimulus onset; originating in the auditory cortex.

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

Plasticity: the capacity to be formed, molded or influenced (e.g. brain development).

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

Psycholinguistic: or **psychology of language** is the study of the [psychological](#) and [neurobiological](#) factors that enable [humans](#) to acquire, use, and understand [language](#).

Randomized Controlled Study/Trial: pertaining to research design with an unbiased selection of subjects randomly assigned to different treatment or non-treatment groups with controlled methods and outcome measures; used to establish efficacy of treatment (e.g. health care).

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

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

Specificity: the ability of a test to differentiate a normal condition from the disorder that the test was designed to detect; expressed as the percentage of negative results in patients without the disorder.

Universal: available and applicable to all without discrimination.

Well-baby: in reference to babies not admitted to special care units.

RISK INDICATORS FOR HEARING LOSS

These risk indicators were specified in the year 2007 position statement of the Joint Committee on Infant Hearing (JCIH). Presence of any of these indicators places an infant at risk for progressive or delayed-onset sensorineural hearing loss and/or conductive hearing loss.

The following risk indicators apply to neonates (birth through age 28 days):

- An illness or condition requiring admission of 48 hours or greater to a NICU
- Stigmata of other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss
- Family history of permanent childhood sensorineural hearing loss
- Craniofacial anomalies, including those with morphologic abnormalities of the pinna and ear canal, and preauricular tags or pits
- In-utero infection such as cytomegalovirus (CMV), syphilis, herpes, toxoplasmosis, or rubella

The following risk indicators apply to neonates or infants (29 days through two years):

- Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay
- Family history of permanent childhood hearing loss
- Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or Eustachian tube dysfunction
- Postnatal infections associated with sensorineural hearing loss including bacterial meningitis
- In-utero infections such as cytomegalovirus (CMV), herpes, rubella, syphilis, and toxoplasmosis
- Neonatal indicators – specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECMO)
- Syndromes associated with progressive hearing loss such as neurofibromatosis type II, osteopetrosis, and Usher's Syndrome
- Neurodegenerative disorders, such as Hunter Syndrome, or sensory motor neuropathies, such as Friedreich's Ataxia and Charcot-Marie-Tooth Syndrome
- Head trauma
- Recurrent or persistent otitis media with effusion (OME) for at least three months
- Chemotherapy

Important Note: The JCIH (2007) risk indicators¹ were the most current and widely accepted at the time that the *ACSLPA Professional Recommendation Statement on Endorsement of Universal Newborn Hearing Screening (UNHS) in Alberta* [ACSLPA 2008] was published. Individuals making use of this information are encouraged to contact a clinical audiologist to ensure that these risk indicators remain current before utilizing them.

REFERENCES

1. Joint Committee on Infant Hearing, American Academy of Audiology, American Academy of Pediatrics, American Speech-Language-Hearing Association, and Directors of Speech and Hearing Programs in State Health and Welfare Agencies. (2007). *Year 2007 position statement: Principles and guidelines for early hearing detection and intervention programs*. *American Academy of Pediatrics*, 120, 898-921. Retrieved November 5, 2007 from <http://pediatrics.aappublications.org/cgi/content/extract/120/4/898>
2. Davis, J., Efenbein, J., Schum, R. Bentler, R. (1986). Effects of mild and moderate hearing impairment on language, educational and psychosocial behavior of children. *Journal of Speech and Hearing Disorders*, 51, 53-62. Retrieved July 24, 2007 from <http://jshd.asha.org/cgi/content/abstract/51/1/53>.
3. Moeller, M.P. (2000). Early intervention and language development in children who are deaf and hard of hearing. *Pediatrics*, 106, 1-9. Retrieved July 24, 2007 from <http://pediatrics.aappublications.org/cgi/content/full/106/3/e43?maxtoshow=&HITS=10&hits=10&RESULTFORMAT=&fulltext=Moeller&andorexactfulltext=and&searchid=1&FIRSTINDEX=0&sortspec=relevance&resourcetype=HWCIT>.
4. Yoshinaga-Itano, C., Sedey, A.L., Coulter, D.K., Mehl, A.L. (1998). Language of early- and later-identified children with hearing loss. *Pediatrics*, 102, 1161-1171. Retrieved July 26, 2007 from <http://pediatrics.aappublications.org/cgi/reprint/102/5/1161>.
5. Kennedy, C.R., McCann, D.C., Campbell, J.J., Law, C.M., Mullee, M., Petrou, S., Watkins, Pl., Worsfold, S., Yen, H.M., Stevenson, J. (2006). Language ability after early detection of permanent childhood hearing impairment. *The New England Journal of Medicine*, 354, 20, 3131-2141. Retrieved September 30, 2007 from <http://content.nejm.org/cgi/content/abstract/354/20/2131>.
6. Holden-Pitt, L., Diaz, J. (1998). Thirty years of the annual survey of deaf and hard of hearing children & youth: A glance over the decades. 143(2), 72-76. *American Annals of the Deaf*, 142, 72-76.
7. Bess, F.H. (1982). Children with unilateral hearing loss. *Journal of the Academy of Rehabilitative Audiology*, 15, 206-216.
8. Bess, F.H., Tharpe, A. (1984). Unilateral hearing impairment in children. *Pediatrics*, 74, 206-216.
9. Brookhouser, P., Worthington, D., Kelly, W. (1991). Unilateral hearing loss in Children. *Laryngoscope*, 101, 1264-1272.
10. Lieu, J.E. (2004). *Archives of Otolaryngology and Head and Neck Surgery* 2004, 130, 524-530.
11. Northern, J.L., Downs, M.P. (1984). *Hearing in Children*; third edition. Baltimore MA: Lippincott Williams and Wilkens.
12. Yoshinaga-Itano, C. (2003). From screening to early identification and intervention: Discovering predictors to successful outcomes for children with significant hearing loss. Oxford University Press.
13. Yoshinaga-Itano, C. 2003). Universal newborn hearing screening programs and

- developmental outcomes. *Audiological Medicine*, 1(3), 199-206. Retrieved July 26, 2007 from <http://www.informaworld.com/smpp/content~content=a714025337~db=all>.
14. Yoshinaga-Itano, C. (2004). Levels of evidence: Universal newborn hearing screening (UNHS) and early hearing detection and intervention systems (EHDI). *Journal of Communication Disorders*, 37, 451-465. Retrieved July 26, 2007 from http://www.sciencedirect.com/science?_ob=ArticleURL&_udi=B6T85-4CDJGWK-2&_user=10&_coverDate=10%2F31%2F2004&_rdoc=1&_fmt=&_orig=search&_sort=d&_view=c&_acct=C000050221&_version=1&_urlVersion=0&_userid=10&md5=92d50e01ce0f02ecffe21f70f0449a88.
 15. National Institutes of Health. (1993). Early identification of hearing impairment in infants and young children. *NIH Consensus Statement*, 11, 1-24. Retrieved July 24, 2007 from <http://consensus.nih.gov/1993/1993HearingInfantsChildren092html.htm>.
 16. Durieux-Smith, A., Seewald, R., Hyde, M. (1999). CASLPA-CAA position on universal newborn and infant hearing screening in Canada. *Journal of Speech-Language Pathology and Audiology*, 24(3), 139-141. Retrieved July 24, 2007 from <http://www.casipa.ca/PDF/position%20papers/newborn%20infant%20hearing%20screening%20for%20pdf.pdf>.
 17. Kenworthy, O.T. (1990). Screening for hearing impairment in infants and young children. *Seminars in Hearing*, 11, 315-332.
 18. Mehl, A.L., Thompson, V. (1998). Newborn hearing screening. The great omission. *Pediatrics*, 101 (1), 9-29. Retrieved July 24, 2007 from <http://pediatrics.aappublications.org/cgi/content/full/101/1/e4>.
 19. Watkin, P., Baldwin, M., McEnery, G. (1991). Neonatal at risk screening and the identification of deafness. *Archives of Diseases in Childhood*, 66, 1130-1135. Retrieved July 24, 2007 from <http://adc.bmj.com/cgi/content/abstract/66/10/1130>.
 20. Parving, A. (2003). Guest Editorial. *Audiological Medicine*, 1, 154.
 21. White, K.R., Behrens, T.R. (Eds). (1993). The Rhode Island hearing assessment project: Implications for universal newborn hearing screening. *Seminars in Hearing*, 14, 1-119.
 22. Institute of Health Economics (IHE). (2007). IHE report: Screening newborns for hearing. Retrieved on November 26, 2007 from <http://www.ihe.ca/documents/IHE%202007%20Reports.pdf>.
 23. Canadian Working Group on Childhood Hearing. (2005). Early Hearing and Communication Development: Canadian Working Group on Childhood Hearing Resource Document. *Ottawa: Minister of Public Works and Government Services Canada*. Retrieved March 30, 2007 from <http://www.phac-aspc.gc.ca/publicat/eh-dp/index.html>.
 24. Hyde, M. (2005). Newborn hearing screening programs: Overview. *The Journal of Otolaryngology*, 34, s70-s78. Retrieved July 23, 2007 from <http://ihp.mtsinai.on.ca/english/documents/Hydearticle.pdf>.
 25. Yoshinaga-Itano, C., Coulter, D., Thomson, V. (2001). Developmental outcomes of children with hearing loss born in Colorado hospitals with and without universal newborn hearing screening programs. *Seminars in Neonatology*, 2001, 6, 521-529. Retrieved July 24, 2007 from <http://www.colorado.edu/slhs/mdnc/research/publications/itano19.html>.

26. Vohr, B.R., Oh, W., Stewart, E.J., Bentkover, J.D., Gabbard, S., Lemons, J., Papile, L.A., Pye, R. (2001). Comparison of costs and referral rates of 3 universal newborn hearing screening protocols. *The Journal of Pediatrics*, 139(2), 238-244. Retrieved July 24, 2007 from http://www.ncbi.nlm.nih.gov/sites/entrez?cmd=Retrieve&db=PubMed&list_uids=11487750&dopt=Abstract.
27. Joint Committee on Infant Hearing (JCIH), American Academy of Audiology, American Academy of Pediatrics, American Speech-Language-Hearing Association, and Directors of Speech and Hearing Programs in State Health and Welfare Agencies. (2000). Year 2000 position statement: Principles and guidelines for early hearing detection and intervention programs. *American Journal of Audiology*, 9, 9-29. Retrieved July 24, 2007 from <http://www.jcih.org/jcih2000.pdf>.
28. US Preventative Services Task Force (USPSTF). 2001. Screening for Newborn Hearing. Retrieved July 24, 2007 from <http://www.ahrq.gov/clinic/uspstf/uspstfnbhr.htm>.
29. Dort J.C., Tobolski C.J., Brown D.K. (2000). Screening strategies for neonatal hearing loss: Which test is best? *The Journal of Otolaryngology*, 29(4), 206-210. Retrieved July 24, 2007 from http://www.auditoryresearch.ca/Publications/Screening_strategies_for.pdf.
30. Government of Alberta. (2007). Information on Cystic Fibrosis Newborn Screening Program. Retrieved July 25, 2007 from <http://www.health.gov.ab.ca/key/devscreensvc.html#Newborn>
31. Corabian, P., Eng, K., Lier, D., Schopflocher, D. (2007). The use of the automated auditory brainstem response and otoacoustic emissions tests for newborn hearing screening. Institute of Health Economics, Alberta.
32. Statistics Canada. (2007). Birth and birth rate by province and territory. Retrieved November 8, 2007 from <http://www40.statcan.ca/l01/cst01/demo04a.htm?sdi=births>.
33. Grosse, D., Boyle, C.A., Botkin, J.R., Comeau, A.M., Kharrazi, M., Rosenfeld, M., Wilfond, B.S. (2003). Newborn screening for cystic fibrosis: Evaluation of benefits and risks and recommendations for state newborn screening programs. Centre for Disease Control and Prevention. Retrieved July 25, 2007 from <http://www.cdc.gov/mmwr/preview/mmwrhtml/rr5313a1.htm>.
34. Bamford, J., Davis, A. (1998). Neonatal hearing screening: A step towards better services for children and families. *British Journal of Audiology*, 32, 1-6.
35. Diefendorf, A.O. (1999). Screening for hearing loss in infants. *The Volta Review*, 99, 43-61.
36. Yoshinaga-Itano, C., Gravel, J. (2001). The evidence for universal newborn hearing screening. *American Journal of Audiology*, 10, 62-74.
37. American Speech-Language and Hearing Association. (2006). Communication Facts - Special Populations: Literacy; 2006 edition. Retrieved January 2, 2008 from <http://asha.org/members/research/reports/literacy.htm>.
38. Edmonton Public School Board. (2007). District review: Students who are deaf and hard of hearing. Phase 1: Summary (Special Education Programs, February).
39. Snow, C.E., Burns, S.M., Griffin, P. (1998). Preventing difficulties in young children.

Chapter 4: Predictors for success and failure in reading. National Research Council, National Academy of Sciences. Courtesy of National Academy Press (reprinted with permission). Retrieved on January 2, 2008 from http://www.pbs.org/launchingreaders/chancetoread/helpfularticles_2.html

40. Gallaudet Research Institute. (1996). Stanford achievement test, ninth edition, Form S, Norms Booklet for Deaf and Hard of Hearing Students; Washington, DC. Gallaudet University. Retrieved on January 2, 2008 from <http://gri.gallaudet.edu/Literacy>.
41. Robinshaw, H.M. (1994). Deaf infants, early intervention and language acquisition. *Early Child Development and Care* 99, 1-22.
42. Ponton, C.W., Don, M., Eggermont, J.J., Waring, M.D., Kwong, B., Masuda, A. (1996). Auditory system plasticity in children after long periods of complete deafness. *NeuroReport*, 8, 61-65.
43. Ponton, C.W., Moore, J.K., Eggermont, J.J. (1999). Prolonged deafness limits auditory system developmental plasticity: Evidence from an evoked potentials study in children with cochlear implants. *Scandinavian Audiology*, 28 (Suppl. 51), 13-22.
44. White, K.R. (2003). The current status of EHDI programs in the United States. *Mental Retardation and Developmental Disabilities Research Reviews*, 9, 79-88. Retrieved July 24, 2007 from <http://www.infanthearing.org/ncham/publications/MRDDRR%20Paper.pdf>.
45. Newborn Hearing Screening Programme (NHS). (2007). Questions and answers. Retrieved September 22, 2007 from <http://hearing.screening.nhs.uk/cms.php?folder=1203>.
46. Davis, A., Hind, S. (2003). The newborn hearing screening programme in England. *International Journal of Pediatric Otorhinolaryngology*, 67, 193-196.
47. NHS Quality Improvement Scotland. (2005). Pregnancy and newborn screening. Retrieved September 22, 2007 from <http://www.nhshealthquality.org/nhsqis/2761.html>.
48. Hyde, M., Picton, N. (2004). Family anxiety & UNHS: A review of current evidence. Retrieved on July 24, 2007 from http://childhearinggroup.isib.cnr.it/docs/unhs_and_family_anxiety.pdf.
49. Shafer, D.N. (2007). Infant screening gains media spotlight. *The ASHA Leader*, 12(8), 1, 7. Retrieved July 25, 2007 from <http://www.asha.org/about/publications/leader-online/archives/2007/070619/070619a.htm>.
50. National Center for Hearing Assessment and Management (NCHAM). (2007). State summary statistics: Universal newborn hearing screening. Retrieved September 22, 2007 from <http://www.infanthearing.org/status/unhsstate.html>.
51. Durieux-Smith, A., Whittingham, J. (2000). The rationale for neonatal hearing screening. *Journal of Speech-Language Pathology and Audiology*, 24, 59-67.
52. Ruscetta, M.N., Arjmand, E.M. (2003). Unilateral Hearing Impairment in Children: Age of Diagnosis. Retrieved from www.audiologyonline.com/articles.
53. Early Childhood Development Agreement; First Ministers Communiqué. (2001). Retrieved on April 2, 2008 from http://www.socialunion.gc.ca/ecd/toc_e.html.
- 54.

- Connecticut Department of Public Health. (2006). Universal newborn screening for cystic fibrosis in Connecticut. Retrieved July 25, 2007 from http://www.dph.state.ct.us/genomics/Documents/CF%20in%20CT%207_06.pdf.
55. Alberta Health Technologies Decision Process. (2007). Synthesis Report 07-01S-2: Review of newborn hearing screening, AHTDP #05-02.
 56. Northern, J.L., Downs, M.P. (1991). *Hearing in Children*; fourth edition. Baltimore MA: Lippincott Williams and Wilkens.
 57. Gorga, M.P., Neely, S.T. (2003). Cost-effectiveness and test-performance factors in relation to universal newborn hearing screening. *Mental Retardation and Developmental Disabilities Research Reviews*, 9, 103-108. Retrieved July 24, 2007 from <http://www3.interscience.wiley.com/cgi-bin/abstract/104535565/ABSTRACT?CRETRY=1&SRETRY=0>
 58. Brown, D.K., Dort, J.C., Sauve R. (2000). Newborn Hearing Screening Programs – A truly Canadian Perspective. *Journal of Speech-Language Pathology and Audiology*, 24(2), 48-58. Retrieved July 24, 2007 from http://www.auditoryresearch.ca/Publications/Newborn_hearing_screening.pdf.
 59. Canadian Organization for Rare Disorders (CORD). (2007). Newborn screening in Canada status report; May 2007. Retrieved September 22, 2007 from http://www.raredisorders.ca/index.php/site/resources/newborn_screening
 60. University of Calgary. (2004). Screening methods of the Alberta UNHS project. Retrieved September 22, 2007 from <http://www.babyhear.ucalgary.ca/>.
 61. Alberta Public Health Association. (2003). Resolution 3: Alberta Universal Newborn Hearing Screening Program. Calgary: Annual General Meeting, May 13, 2003. Retrieved July 24, 2007 from <http://www.cms.apha.ab.ca/modules.php?name=Contentdocs&pa=showpage&pid=25>.
 62. The Hearing Foundation of Canada. Universal Newborn Hearing Screening Program: One Simple Test Could Change Your Child's Future. Retrieved September 22, 2007 from http://www.thfc.ca/hearing_publicFoundation.asp
 63. American Academy of Pediatrics. (1999). Newborn and Infant Hearing Loss: Detection and Intervention. *American Academy of Pediatrics Task Force on Newborn and Infant Hearing*. Retrieved July 25, 2007 from <http://aappolicy.aappublications.org/cgi/reprint/pediatrics;103/2/527.pdf>.
 64. Stach, B.A. (2003). *Comprehensive Dictionary of Audiology*; Illustrated; Second edition. Thomson Learning (Delmar).
 65. Wikipedia. (2008). Some definitions retrieved on April 1, 2008 from http://en.wikipedia.org/wiki/Main_Page.
 66. Some definitions retrieved on April 2, 2008 from www.dictionary.com .