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Version 2019.01

PROTOCOL FOR UNIVERSAL NEWBORN HEARING SCREENING IN ONTARIO

Ministry of Children, Community and Social Services
Ontario Infant Hearing Program
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CONTRIBUTORS
Marlene Bagatto
Susan Boehm
Rana El-Naji
Vanessa Martin
Marie Pigeon
Louise Tanaka
Martyn Hyde

CORRESPONDENCE
Vanessa Martin, MClSc, Aud (C), Reg. CASLPO
Program Consultant
Ministry of Children, Community and Social Services
101 Bloor Street West, 3rd Floor,
Toronto, ON M5S 2Z7
Tel: 416 327-4872
E-mail: vanessa.martin@ontario.ca

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SECTION 1: INTRODUCTION

This document describes the Ontario Infant Hearing Program's (IHP) protocol for universal newborn hearing screening (UNHS) of newborns and infants. It overrides all previous protocols on this subject provided by the IHP. The primary audience for this protocol is those who conduct newborn hearing screening within the IHP.
All newborn and infant hearing screening funded by the Ontario Ministry of Children, Community and Social Services (MCCSS) must be carried out in full accordance with this protocol. It is based on continuous review of the best available scientific and clinical evidence and expert consultation complemented by consultation and collaboration with other major Early Hearing Detection and Intervention (EHDI) programs in Canada and worldwide. Every child and family is entitled to the high quality of care that this protocol represents. The three major pillars of quality are effectiveness, equity, and efficiency. Departure from the protocol may result in: 1) failure to identify a child with genuine hearing loss; 2) unnecessary referral with resulting family anxiety and disruption; 3) corrective action as described by the IHP Screening Continuous Quality Improvement (CQI) Initiative, such as having to recall infants for re-screening; or 4) incurring medico-legal risk. If any portion of this protocol is not fully understood, it is imperative that IHP Hearing Screeners seek clarification from IHP Regional Trainers (RTs) or through IHP Regional Coordinators.

The scope of this document includes the details of the UNHS procedures as funded by the MCCSS for the IHP.

1.1 VERSION HISTORY

This version of the IHP UNHS protocol (2019.01) supersedes all previous documents relating to UNHS. It is substantially revised from previous documents related to Screening within the IHP.

VERSION DATE	DOCUMENT TITLE	PREVIOUS VERSION
JULY 2013	IHP Hearing Screening Protocol and Support Document	Infant Hearing Screening Assessment & Communication Development Local Implementation Support Document (2002)

Revisions to this version are largely due to changes to IHP Screening which now includes an additional risk factor screen using the dried blood spot (heel prick) sample collected by Newborn Screening Ontario (NSO). Additionally, changes to the risk indicator list which drives the choice of hearing screening technology, bypass of hearing screening, and Audiological Surveillance are included.

1.2 REVISION SUMMARY FOR VERSION 2019.01

Recent amendments to this Hearing Screening protocol are largely due to the introduction of the risk factor screen.

TOPIC	DESCRIPTION	SECTION
RISK FACTOR SCREEN	NSO will screen the blood spot from the infant's heel prick for congenital cytomegalovirus (cCMV) and several common genetic mutations in three genes associated with permanent childhood hearing loss.	2.4
SCREENING POPULATION	Infants over two months corrected age must not be screened.	2.10
CONTACT & CONSENT	Explicit informed consent is required for the risk factor screen. In IHP regions where sharing agreements are in place, consent is also needed to share	3.3

	demographic information within the IHP database.	
RISK INDICATORS	List of risk indicators has been updated.	3.7 – 3.10
GROUP 2 RISK INDICATORS	A single-point surveillance is targeted at 15 to 18 months of age and conducted by an IHP Audiologist for infants who have these risk indicators.	3.7.2
SURVEILLANCE	Due to the updated risk indicators, the surveillance protocol has been modified and follow-up is determined by the Group in which the infant's risk indicator falls.	3.7.2 & 3.7.3
NUMBER OF ADPOAE SCREENING ATTEMPTS	In any given ear, the maximum allowable number of Stage 1a ADPOAE attempts is *four*.	4.5
DOUBLE-REFER	A refer result on ADPOAE with acceptable infant and environmental conditions must be repeated where the attempt limit permits. If a double-refer is obtained in a given ear on ADPOAE, **there must be no further ADPOAE screening in that ear**. A double-refer with AABR in any given ear is preferred to obtain an overall refer for that ear. A double-refer is preferred to move to the next stage.	4.7 5.2
NUMBER OF AABR SCREENING ATTEMPTS	The maximum permissible number of AABR screening attempts pre-discharge or per scheduled community appointment for Stage 1b or Stage 2 AABR in any given ear is *two*.	4.12
AABR SCREENING OF NO RISK INFANTS	If acceptable test conditions cannot be achieved within **two scheduled appointments** for any infant with no risk for PHL, **further screening will not be scheduled.**	4.12

SECTION 2: SCOPE

2.1 WHAT IS THE ONTARIO INFANT HEARING PROGRAM?

The Ontario Infant Hearing Program (IHP) is an example of what are known worldwide as EHDI programs. Such programs have become widespread worldwide over the last 20+ years. EHDI programs include universal newborn hearing screening (UNHS), diagnostic hearing assessments (audiological assessments) that confirm and describe any hearing loss that is present, as well as interventions that are aimed at improving a child's hearing and communication abilities. These interventions take many forms: medical (such as medications to prevent or treat ear disorders), surgical (such as cochlear implants), audiological (such as amplification devices – hearing aids or other assistive listening devices) and a range of counseling and supports, including spoken language and sign language support for language development. Data systems are an important infrastructural element that tracks

infants' progress through the EHDI process, triggers key required actions, records procedural outcomes and assembles program performance reports. Further information about the IHP can be found in the *IHP Guidance Document*.

Approximately one to three in 1000 infants are born with a permanent hearing loss (PHL), regardless of the presence or absence of a risk indicator (Fortnum et al 2002). Since the inception of the IHP in 2001, approximately 450 infants in Ontario are identified each year with PHL. The ultimate goal of the IHP is to minimize the impact of PHL on the individual child's development. It is well-established that very early detection of hearing loss, when coupled with appropriate follow-up, can allow a child with PHL to function well socially and educationally (Joint Committee on Infant Hearing (JCIH), 2007). Before EHDI programs were introduced, the *average* age of identification of PHL in children was about two to three years in most developed countries. Now, with a high-quality EHDI program, the average age of hearing loss confirmation can be under three months of age with intervention initiated by six months of age. This time frame is very important, because early development of the brain's auditory system depends on sensory stimulation, namely hearing ability. Access to speech through hearing allows the developing brain to organize the necessary networks for developing spoken language. The first two or three years of a child's life are especially important for neurological development of the auditory system as well as other areas of the brain involved in acquiring, understanding, and expressing spoken language. This time period is often referred to as the "critical period" for language development. The UNHS portion of an EHDI program is a crucial first step in engaging the child and family in the EHDI process.

Because the goal of the IHP is early access to effective interventions for hearing and language development, timeliness of the various steps in the overall process is extremely important. The most widely accepted benchmarks for EHDI programs reflect a "1-3-6" timeline (JCIH, 2007). These benchmarks specify that, to the fullest extent possible, the overall process of newborn hearing screening should be completed by one month corrected age, audiological assessment for confirmation and detailed description of hearing should be completed by three months corrected age, and intervention services, should a PHL be confirmed, should be initiated by six months corrected age. Achievement of these benchmarks is the crucial measure of program effectiveness.

A glossary of terms is provided in Appendix B to offer descriptions of key terms used throughout this Hearing Screening protocol.

2.2 IHP CORE PRINCIPLES

Newborn hearing screening using electrophysiological measures shall be provided in accordance with the IHP core principles of informed family/caregiver choice and consent, timely provision of unbiased information based on the best available scientific evidence, and sensitivity to family culture and values. Further details about the IHP can be found in the *IHP Guidance Document*.

2.3 WHAT IS UNIVERSAL NEWBORN HEARING SCREENING?

UNHS is the first step in the EHDI process whereby families consent to an electrophysiological test (hearing screening) as well as an additional risk factor screen conducted with their newborn to determine whether further hearing assessment is required. Screening is not diagnostic or therapeutic. It separates infants into two groups: those who are likely to have a PHL and those who are not. As such, hearing screening determines whether the infant needs a more comprehensive hearing test conducted by an IHP Audiologist.

Electrophysiological hearing screening technology includes two methods: automated distortion product otoacoustic emissions (ADPOAE) and automated auditory brainstem response (AABR). See Appendix B for an explanation. The "universal" aspect of UNHS means that all infants who live in Ontario are offered hearing screening. There are some conditions or factors, known as risk indicators, which mean the infant has a higher likelihood of being born with PHL. However, only 50% of infants born with PHL have a risk indicator (Mehl & Thomson, 1998). Therefore, population screening of all infants born, regardless of the presence of a risk indicator, is the goal of UNHS as part of a comprehensive EHDI program.

Although PHL can be present at birth (that is, *congenitally*), there is an increase in PHL prevalence after birth due to *acquired* PHL. It has numerous causes (infections, injuries, noise exposure, genetic disorders, etc.), most of which cannot be detected within the IHP during the newborn period. However, some conditions or events that are recognized in the neonate (a newborn up to one month of age) are associated with development of non-congenital PHL, referred to as *delayed*- or *late-onset* PHL. These associated conditions or events are risk indicators that predict an increased likelihood of late-onset PHL. The IHP detects some of these affected children relatively early by *audiological surveillance,* which involves recall of at-risk infants for audiological assessment at specific times or ages (see *IHP Audiological Surveillance Protocol*).

The ages referred to in 1-3-6 are *corrected age*, which is the chronological age of the infant adjusted for a 37-week full term gestation period. Infants are considered premature if they are born before 37 weeks of pregnancy are completed (World Health Organization, 2015 [fact sheet 363]). Therefore, corrected age is to be calculated using **37 weeks** as full term. For example, an infant born 6 weeks ago at an estimated 34 completed weeks of gestation has a corrected age of: 6 – (37 – 34) = 6 - 3 = 3 weeks. Except where otherwise stated, any age specified in this document is a corrected age.

2.4 RISK FACTOR SCREEN

The IHP and NSO offer a risk factor screen that uses the blood spot obtained from a heel prick that is already used to detect various rare but treatable diseases in the newborn. The risk factor screen looks for several common genetic mutations in three genes known to cause PHL in infants as well as congenital cytomegalovirus (cCMV) which is the leading cause of non-genetic PHL.

The goal of offering families the additional risk factor screen is to improve the IHP's risk assessment process. It allows for earlier and more accurate identification of infants with specific risk factors for PHL and their subsequent assessment or surveillance monitoring.

2.5 WHO CAN CONDUCT NEWBORN HEARING SCREENING WITHIN THE IHP?

Only individuals who are trained and authorized by the IHP to conduct this protocol may provide newborn hearing screening services with IHP funding. Specific qualifications of Hearing Screeners can be found in the *IHP Guidance Document* and are at the discretion of the Lead Agency IHP Coordinators. If the IHP Hearing Screener has been inactive in this protocol for six months or more, the re-training review procedures in the *IHP Guidance Document* will apply. Regional Trainers (RTs) are highly qualified IHP Hearing Screeners that meet specific criteria (see Appendix C). Their role is to train individuals new to IHP Hearing Screening and retrain IHP Hearing Screeners as identified through MCCSS' CQI initiative.

Authorization for Screening may be withdrawn at the discretion of the MCCSS.

2.6 PROTOCOL ADHERENCE IS A REQUIREMENT

All IHP Hearing Screening must be conducted in adherence to this protocol. Such adherence is an expectation for continued authorization to provide IHP services. Hearing screening forms, including all notes, must be completed and submitted in a timely fashion so that the IHP Lead Agency may follow up on CQI activities.

2.7 LEGITIMATE DEPARTURE FROM PROTOCOL

It is acknowledged that case-specific situations that justify departure from mandatory protocol elements can arise. Such departures must be noted in the infant's records with a brief explanation. All such notes must be accessible by the IHP Lead Agency to support IHP standard practice review or case audits.

2.8 PROCEDURAL CONCERNS

Prior approval by MCCSS is required in order to change substantively any element of this protocol. Program-wide changes can occur only through MCCSS directive or by a systematic process that may include survey of Screeners' experiences or concerns, evidence review, and recommendation by an IHP Designated Training Centre (DTC). IHP Protocols are evidence-based to the extent possible. Evidence is reviewed by the DTCs on an ongoing basis. This may result in specification of procedures that differ from opinions in published journals. Substantive issues will be addressed by new evidence review, re-examination of existing evidence, and/or provincial consensus development. Changes to IHP protocols are outside the mandate of regional management and shall be authorized *only* by modification of the relevant IHP protocol document (such as this document), which shall govern IHP Screening services throughout Ontario.

2.9 NON-IHP SERVICES

Hearing Screening services conducted by any person who is not authorized by the IHP shall not be funded by the IHP and shall not be deemed to provide a sufficient basis for subsequent management within the IHP. For this reason, re-screening or audiology assessment shall be conducted by an authorized IHP service provider prior to making inferences about hearing status and/or candidacy for ongoing management.

2.10 HEARING SCREENING POPULATION

Candidates for Hearing Screening include all Ontario-resident infants **two months corrected age and younger**. For hospital-based screening, the goal is to complete hearing screening prior to discharge following birth. For community-based hearing screening, infants over two months corrected age must not be screened for the following reasons:

1) The hearing screening equipment (Madsen Accurscreen) has been developed for newborn-sized ear canals. The infant's ear canals may be too large for any available eartip rendering the visit incomplete.
2) There are no adequate data on which to base an informed decision about the performance characteristics of the Madsen AccuScreen in older infants.
3) The AABR screening technology requires a sleeping infant. This is less likely to occur with older infants.
4) If the hearing screen on an older infant is incomplete due to the reasons stated, any referral for Audiological Assessment may be too late for a diagnostic ABR in natural sleep thereby delaying the EHDI process.

Infants older than two months corrected age who present for screening must be declined. The family should be counselled on signs of hearing loss and, if and when concerned, should seek medical contact. Behavioural audiometry by community audiology services may be practicable at about six months of age, with referral to the IHP then if there is clear evidence of PHL. If there is a medical record of a high risk indicator for PHL, at the discretion of the IHP Regional Coordinator the infant may be admitted into the IHP for age-appropriate Audiological Assessment. For details for clarifying entry/re-entry of children to the IHP, see Appendix D.

Occasionally, late presentation for hearing screening may arise in connection with occurrence of an acquired risk indicator later in infancy, such as a serious infection or head injury. In these situations, parents/guardians should seek medical consultation. The IHP's standard position is that questions about hearing that are associated with events requiring medical management must first be addressed outside of the IHP, again with referral into the IHP if there is clear evidence of a PHL from audiological services within the community. Community audiology assessments are most often conducted with infants six months of age and older due to the lack of expertise and equipment required for conducting ABR assessments. If the infant is younger than six months of age, they may be seen for IHP Audiological Assessment using ABR as necessary (see Appendix D).

2.11 TARGET DISORDERS

The IHP target disorder set includes PHL of ≥ 30 dB HL or more at 0.5, 1, 2, or 4 kHz in any ear, auditory neuropathy spectrum disorder (ANSD), and auditory brainstem pathway disorders that may be detectable using ABR techniques (see *IHP Auditory Brainstem Response Audiometry Protocol*). The target PHL includes conductive impairment associated with structural anomalies of the ear but does NOT include impairment attributable to minor, non-structural middle ear conditions.

2.12 SCREENING EQUIPMENT AND PARAMETERS

The equipment used within the IHP for ADPOAE and AABR screening is the Madsen AccuScreen hand-held device. The AccuScreen must have the following firmware and protocols installed:

Firmware 2012: 1.11.04160SEU

DP 5:

- The cut off is 8 dB signal-to-noise (SNR)
- Frequencies tested are 2 kHz, 3 kHz, 4 kHz, 5 kHz in descending order
- Criteria for pass is three out of four frequencies for that ear

ABR 35:

- Chirp stimulus presented at 35 dB nHL
- Criteria for pass is a suitable ABR waveform present at that stimulus

2.13 OBJECTIVE OF UNIVERSAL NEWBORN HEARING SCREENING

The main objectives of UNHS are to:

1) Determine whether the infant has a risk indicator for PHL; and
2) Determine whether further audiological assessment is required.

The overall Hearing Screening protocol goes beyond the electrophysiological procedures itself and includes providing the family with the results and information about follow-up.

2.14 IHP DESIGNATED TRAINING CENTRES (DTC)

DTCs are authorized by the MCCSS to provide IHP support, including advanced training, consultative and assessment referral services, protocol support, and clinical decision support to IHP Audiologists and Regional Trainers for various components of the IHP. DTCs also conduct standard IHP practice reviews and implement audits of services as directed by MCCSS.

The DTCs for ABRA and conditioned behavioural audiometry (CBA) are CHEO (Ottawa) and Humber River Hospital (HRH, Toronto). The National Centre for Audiology (NCA; Western University, London) is the DTC for Amplification, Screening, and Surveillance.

2.15 CONTINUOUS QUALITY IMPROVEMENT (CQI)

All high-quality EHDI programs have quality management procedures for the major program activities. This is especially important for programs that comprise a series of key stages, because shortfalls in each and every stage accumulate to rapidly result in serious deficiencies of overall program effectiveness. Programs with public funding are usually obliged to measure, control, disclose, and constantly assure and improve their overall performance. This is an issue of due diligence, transparency and accountability in the use of public resources.

The product of Hearing Screeners' work can be boiled down to a few key features that can be measured. Number of infants screened can reflect *efficiency*. The proportion of infants with successful hearing screens and reasonably low false-positive refer rates are measures of *effectiveness;* given that most hearing screening refers, especially in

infants not at risk and especially for ADPOAE screens, are false-positive. Also, consistency of these performance measures across Hearing Screeners, IHP regions and hearing screening locations, as well as consistency over time, reflect *equity* or fairness of the distribution of care quality across all infants and families.

The IHP Hearing Screening CQI program focusses on the most concrete measures of performance effectiveness. It includes recourse to expert advice, accessible materials on several aspects of 'how to do it', meetings, workshops and conference calls, and many more locally specific activities. It also includes data-driven efforts to find and address constructively situations that appear to reflect screeners' field performance challenges. Measurement actions related to the numerically biggest single volume of work type in the IHP, namely the Stage 1a ADPOAE screening in infants who are not at risk for PHL are in place and monitored on a quarterly basis within each IHP region.

IHP Lead Agencies should have documentation of hearing screening protocol adherence on file to support clinical decision support and/or standard practice review. The documentation should include results of the CQI report, AccuScreen audit results (see next section), and hearing screening notes captured in Healthy Child Development – Integrated Services for Children Information System (HCD-ISCIS). The IHP Lead Agency would send the necessary documentation to the DTC for further review and next steps.

2.15.1 ACCUSCREEN AUDITS

An AccuScreen audit must be performed by the Regional Trainer at least twice each fiscal year for every IHP Hearing Screener to monitor compliance with the current Hearing Screening protocol. The audit will include no less than 50 hearing screening results. The procedure for performing an AccuScreen audit is included in Appendix E.

2.16 INFECTION CONTROL STANDARDS

Infection control practices are typically governed by site-specific, institutional or IHP Lead Agency protocols and are outside the purview of this document. Generally accepted standards must be applied.

2.17 CLINICAL RECORDS AND DATABASE REPORTING

All hearing screening records shall be maintained in a manner satisfying site-specific and IHP Lead Agency policies and the IHP. Records detailing results of the hearing screening session and reporting of any necessary information to HCD-ISCIS are required.

2.18 PERSONAL HEALTH INFORMATION

Management of all personal health information arising from the hearing screening process shall comply with local site, IHP Lead Agency, and legislative requirements. Information communicated for approved monitoring and review procedures must be de-identified and code-referenced. All transmission of personally-identifiable information shall be consented by the appropriate family member or authorized parent/guardian.

SECTION 3: PROTOCOL FOR NEWBORN HEARING SCREENING

3.1 OVERALL IHP PROCESS FOR IDENTIFICATION OF PERMANENT HEARING LOSS (PHL)

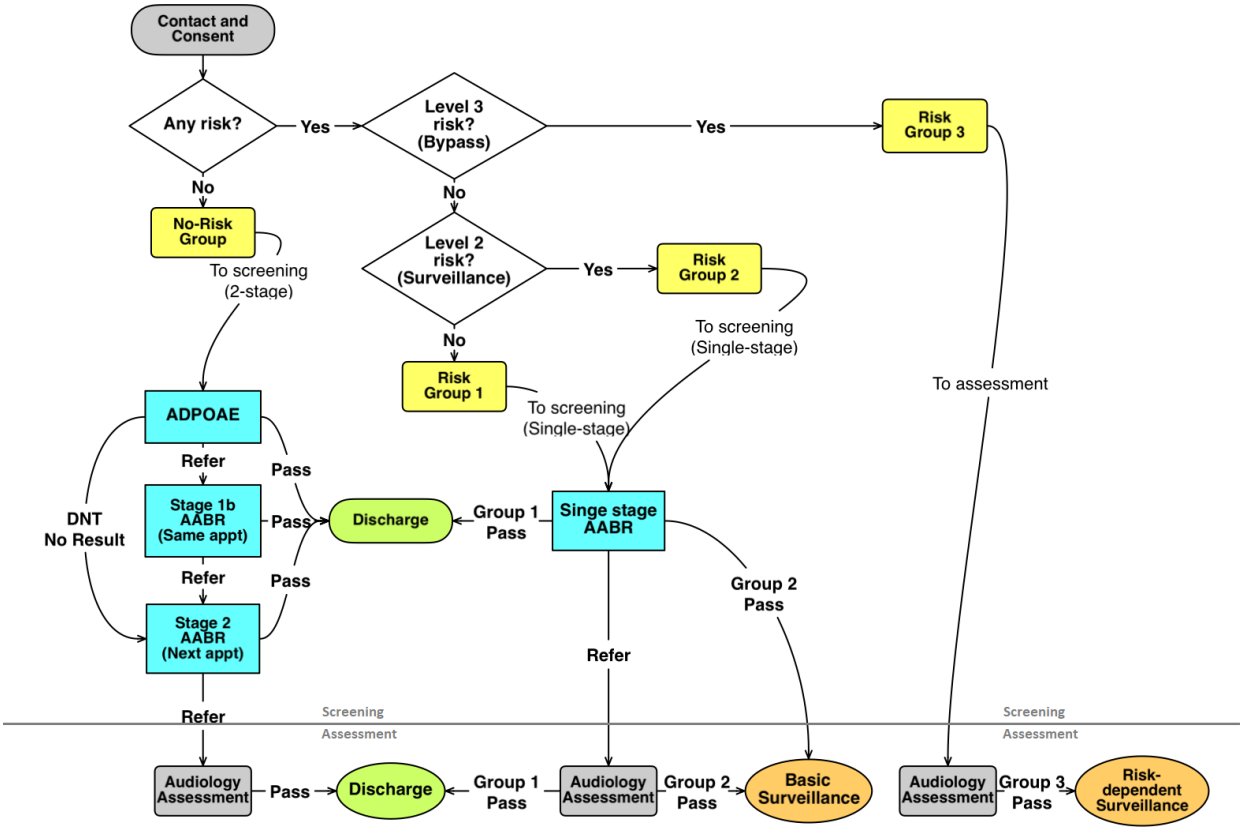

Figure 1: Flow of IHP hearing screening protocol.

3.2 HEARING SCREENING LOCATIONS

In some cases, infants in Ontario will be born in hospital and the first contact between the IHP Hearing Screener and the parent/guardian, as well as the initial hearing screening, will occur prior to discharge from the birthing facility. This process is referred to here as “Pre-Discharge” hearing screening. In other cases, the first contact will occur in a facility other than a birthing hospital and is referred to in this protocol as “Community” screening. Sometimes, the initial contact and hearing screening attempt will be Pre-Discharge but the initial hearing screening procedure will be completed in the Community.

3.3 CONTACT & CONSENT

Upon contact with a parent/guardian, the IHP Hearing Screener introduces the IHP and the hearing screen then seeks informed consent using the provided, standardized scripts. The core script has been provided in Appendix F and includes the following items:

1) Consent to share information (required to access hearing screen);
2) Consent to hearing screening; or
3) Explicit consent for the risk factor screen (only available if there is consent for hearing screening).

Any other consents or details required by local IHP Lead Agency policies (i.e., consent to share demographic information within the database with Healthy Babies Healthy Children will be added to the core script by the IHP Lead Agency and provided to the local IHP Hearing Screeners.

Note: Sharing of information includes IHP, NSO, and other providers as required for follow-up.
If consent is declined, a standardized IHP fact sheet is provided to the family describing the importance of childhood hearing.

3.3.1 EXPLICIT INFORMED CONSENT

Explicit informed consent requires the sharing of enough information about the service for the parent/legal guardian providing the consent to understand the consequences of consenting or declining the service. This can be done verbally or in writing, however, it must include an opportunity for the parent/legal guardian to clarify their understanding of the service by having any questions answered prior to making their decision.
Explicit consent requires that the parent/legal guardian making the consent decision be provided with a clear choice to consent or decline the service.

The explicit informed consent model required for the risk factor screen must meet the following criteria:

1) Provides information about the risk factor screen
    a. What it looks for;
    b. How the screen works; and
    c. Who is eligible for the screen
2) Explanation of the consent choices
    a. Risk factor screen is only available if there is consent for the hearing screen;
    b. Hearing screen is available without the risk factor screen
3) Opportunity to have questions answered about the risk factor screen prior to making a consent decision
4) Documentation of the consent choice, who provided the consent, when it was obtained and by whom

3.3.2 OBTAINING CONSENT

The consent must be obtained by someone who:

1) Has completed the MCCSS IHP Hearing Screener training;
2) Has been familiarized with what the hearing screen involves;
3) Is aware of the purpose of the hearing screen and risk factor screen;
4) Follows the script guidelines when obtaining the consent; and
5) Is comfortable responding to questions that are in scope about the hearing screen and risk factor screen.

Hospital/midwifery staff will **not** be able to complete the consent **unless** they have completed the IHP Hearing Screener training. They must be able to respond appropriately to parent questions and complete and document the explicit informed consent as required.

Wherever possible, consent should be obtained by staff associated with the IHP to allow for proper CQI. Training staff within the program helps ensure consistency in training, monitoring of service, and service quality, such as consistency in messaging.

3.3.3 PROVIDING CONSENT

Consent must be obtained from the parent/legal guardian of the infant and documented on the Hearing Screening form. In cases where the child is in the care of a child protection service, the agency becomes the legal guardian. The agency and/or caseworker must be listed as the Primary Contact. The foster family is not the legal guardian and cannot provide the necessary consent. This needs to come from the caseworker or an alternate contact at the agency.

In cases of adoption or surrogacy, issues of consent should be addressed according to local policies based on the individual circumstances.

3.4 SHARING OF INFORMATION

The sharing of information must be discussed with all families regardless of their consent decision. IHP Lead Agencies require documentation of all hearing screens for quality assurance (being part of an EHDI program) and for record-keeping purposes. No hearing screen should occur without proper documentation. Quality control is not possible if undocumented hearing screens are occurring. Therefore, those families who decline the sharing of their information are not eligible for a hearing screen.

NSO also requires documentation of the consent choice made by families for the IHP risk factor screen to know whether or not to test the blood spot sample and to generate reports for missed screens for the IHP Lead Agencies.

Those parents/guardians who choose to decline the hearing screen and not share their information should be made aware that without sharing the documentation of their choice to decline, they are likely to be contacted by their IHP Lead Agency to offer a community hearing screen as it will appear as though they were missed. By allowing the sharing of information, this additional contact can be avoided.
The information collected by IHP includes sharing with:

- The IHP Lead Agency for the region in which the infant was born as well as their home IHP Lead Agency (when follow-up is required);
- NSO for documentation of their choice regarding the risk factor screen and to exclude infants from the missed screen report; and
- Those agencies/individuals providing follow-up services for IHP (Hearing Screeners, IHP Audiologists, etc.) and/or NSO (Infectious Disease specialists, community pediatricians, physicians, etc.).

3.5 RAPPORT WITH FAMILIES

From the initial contact through to the completion of the hearing screen, the screener's behaviour, style, and tone can have a major effect on the family's cooperation, giving of informed consent, satisfaction with the screening experience, and adherence to follow-up instructions. In general, families wish to feel respected and properly informed about what will happen in the screen, what is happening as the screen proceeds, what the result was, what the result means, what should happen or what they should do next and why they should follow recommendations.

Hearing Screeners should make every effort to project calmness, empathy, knowledge, confidence, and professionalism at all times. Hearing Screeners should **never** reveal frustration or irritation, such as with a non-cooperative infant or an unsuccessful screen. When interacting with new mothers, the effects of fatigue, stress and feeling overwhelmed should not be underestimated.

3.6 ASSESS RISK FOR PERMANENT HEARING LOSS

A risk indicator is an identifiable characteristic of the child or medical procedure used with the child that is associated with increased likelihood of PHL in that child greater than the likelihood in the newborn or child population as a whole. Risk indicators are identified by the screener through document review, consulting nursing or medical staff, and talking to the family.

With respect to initial screening in the community, if the Hearing Screener's first contact with the family is in a community facility, then the family/caregiver may be the only source of risk information other than the direct physical observation of the screener, such as for an obvious malformation of the external ear. A confident and clear report of cleft palate can be accepted. The remaining risk indicators require detailed medical knowledge and if the family cannot confirm a risk indicator exists, then the infant should **not** be considered *At Risk*. When in doubt, the decision should be *No Risk* and the default is ADPOAE screening. If a risk indicator is present, screen only using AABR.

The purposes of risk assessment are to:

1) Determine whether screening should be bypassed; or
2) Decide which screening test type to use (i.e., ADPOAE or AABR); and
3) Record information that will determine whether the infant should receive later audiological testing (surveillance) and, if so, what type of surveillance.

The complete list of IHP risk indicators is provided in Appendix G. Information about modifications from the previous list are described in the next sections.

The items on the list are the **only** IHP risk indicators that exist; no other medical conditions, treatments, medications or family history items are acceptable. The only exception to this is a risk indicator specified by a physician, which is itself an IHP risk indicator.

Note that an infant is not at risk unless and until at least one of the IHP risk indicators is determined to be present. If no such determination is yet available, the infant is not at risk. There is no such thing as 'probable risk', it is either present or it is considered not to be present.

It is the Hearing Screener's responsibility to collect risk information, but it can be assisted by a nurse, except for the Family History indicator, for which the screener is responsible to ensure that the family/caregiver is questioned exactly as specified in this protocol. The role of a nurse is to assist with the indicators related to specific medical conditions or procedures.

If the family uses medical terminology that is unfamiliar to the Hearing Screener, assistance from an individual such as a nurse should be sought where feasible. If assistance is not available, the terminology used by the family does not in itself generate risk.

3.7 IHP RISK INDICATORS

The list of IHP risk indicators has been modified based on initial evidence review, expert consultation, and availability of IHP resources. In addition, the implementation of the risk factor screen has, in part, informed the hearing screening, bypass, and surveillance procedures. The summary list of IHP risk indicators and surveillance steps are found in Appendix G. In summary, the following list represents the current risk indicators for the IHP:

***Group 1*:** **AABR Screen** **No Surveillance**	***Group 2*:** **AABR Screen,** **Basic Surveillance if Pass**	***Group 3*:** **Bypass Screen, Refer to Audiology, Basic or Intensive Surveillance if Pass**
APGAR at 5 minutes ≤ 3 Birthweight ≤ 1000g Congenital Diaphragmatic Hernia Family history of parent or sibling with PHL identified by 10 years of age Hypoxic Ischemic Encephalopathy (HIE) Sarnat II or III Intraventricular Hemorrhage (IVH) Grade III or IV	Cleft palate Extracorporeal membrane oxygenation (ECMO, ECLS) Hyperbilirubinemia meeting exchange criterion, whether exchanged or not Proven TORCHES infection (toxoplasmosis, syphilis, rubella, herpes simplex virus) *except CMV* Syndrome associated with PHL *except CHARGE*	Atresia/microtia (screen of unaffected ear permitted) CHARGE Syndrome Proven Congenital Cytomegalovirus (cCMV) Proven Meningitis Genetic Screen Positive

Peri-ventricular Leukomalacia (PVL) Persistent Pulmonary Hypertension of the Newborn (PPHN) Ventilatory support with at least one of the following: • High frequency ventilation (HFJ, HFO, HFV) • Inhaled nitric oxide (iNO, NO) Other risk identified by the physician		

3.7.1 GROUP 1 RISK INDICATORS

Infants who are identified as having a Group 1 risk indicator will undergo an AABR screening procedure. If the screening outcome is an overall 'refer' result, next steps of the Screening protocol are required. If the hearing screening outcome is an overall 'pass' result, the infant does not require Audiological Surveillance and is discharged from the IHP. The purpose of the Group 1 risk indicators is to determine the type of screening technology to apply. The infant will be re-directed for audiology assessment if the risk factor screen is positive following a 'pass' result on the hearing screening.

3.7.2 GROUP 2 RISK INDICATORS

Infants who are identified as having a Group 2 risk indicator will undergo an AABR screening procedure. If the screening outcome is an overall 'refer' result, next steps of the Screening protocol are required. If the hearing screening outcome is an overall 'pass' result, the infant enters into the Basic Audiological Surveillance sequence. This single-point surveillance is targeted at 15 to 18 months of age and conducted by an IHP Audiologist (see *IHP Audiological Surveillance Protocol*).

The exceptions for Group 2 are CHARGE and CMV. CHARGE is considered a syndrome known to be associated with PHL. CMV is screened through the risk factor screen. Both are included as a separate risk indicator in Group 3 and infants identified with either of these risk indicators bypass hearing screening.

3.7.3 GROUP 3 RISK INDICATORS

Infants who are identified as having a Group 3 risk indicator will bypass hearing screening and go directly to an IHP Audiologist for an ABR assessment. Screening tests do not have the ability to identify hearing loss, that is, they have less than perfect sensitivity to PHL. For certain risk indicators, like the ones in Group 3, the probability of PHL is very high, so the possibility of false-negative screening is increased. In that case, the infant must be flagged for routing directly to Audiological Assessment. Also, if the infant has atresia or microtia, it may be difficult or impossible to obtain a satisfactory eartip insertion. In view of the likelihood of insertion failure as well as the high PHL probability, the infant must be routed directly to Audiological Assessment.

If the infant is determined to have PHL following the assessment, supports and services within the IHP will be offered. If the audiology assessment reveals normal hearing in both ears, the infant enters into surveillance, which varies in frequency and timing of appointments depending on the risk indicator (see *IHP Audiological Surveillance Protocol*).

3.7.4 GENETIC CHANGE SCREEN POSITIVE FROM RISK FACTOR SCREEN

Infants who screen positive for one of the included genetic mutations and who pass an initial audiology assessment will continue within the Intensive Surveillance sequence. This is because the likelihood of the infant developing PHL is high and they should be closely monitored.

3.8 REVIEW OF UNCHANGED RISK INDICATORS

APGAR at 5 minutes ≤ 3: This traditional, multi-component indicator largely reflects cardio-pulmonary function and has genuine, though limited, predictive relationships with long-term neurodevelopmental outcomes (Lieu, Ratnaraj & Ead, 2013). It is retained due to its accessibility and to lack of evidence that the other, more specific indicators here render it non-predictive.

Birthweight ≤ 1000g: This accessible, non-specific indicator defines the Extremely Low Birthweight (ELBW) group (World Health Organization, 2004).

Congenital Diaphragmatic Hernia (CDH): If the diaphragm does not close completely, body structures normally located below it can force their way up into the chest cavity, potentially compromising pulmonary and/or cardiac function.

Hypoxic-Ischemic Encephalopathy (HIE): Moderate (Sarnat 2) or Severe (Sarnat 3): Encephalopathy is injury to brain structures, in this case due to lack of oxygen, caused by either insufficient blood supply (ischemia) or to insufficient oxygen-delivery capacity (hypoxemia), or both. Modified Sarnat is a severity scale.

Intraventricular Hemorrhage (IVH): Grade III or IV: The ventricles generate and circulate cerebrospinal fluid. Especially in infants with low birth weight, the developing brain is vulnerable to deficient blood supply or oxygen levels. Consequent cell injury or death can cause bleeding into the ventricular lining, the ventricular fluid space itself or into nearby structures. Severity is graded I to IV, grades III and IV reflecting high risk of neurological and neurodevelopmental sequelae, as well as concurrent cochlear damage.

Periventricular Leukomalacia (PVL): This is a brain injury characterized by coagulation or necrosis of nerve fibre tracts (axons, white matter) near the lateral ventricles. It can affect the fetus or newborn; premature newborns are at greater risk for this disorder.

*Persistent Pulmonary Hypertension of the Newborn (PPHN)***:** This indicator relates to compromise of the normal post-partum circulatory transfer from the placenta to the lungs. It is a syndrome characterized by marked pulmonary hypertension and resultant hypoxemia.

Ventilatory support with at least one of the following:
*Inhaled Nitric Oxide (iNO)***:** Nitric oxide (NO) is a colorless gas with a sweet odour. It is a powerful vasodilator with a strong relaxation effect on smooth musculature in the lungs, improving oxygenation. It may be used when ventilation has insufficient effectiveness. It can decrease the need for use of major, invasive techniques such as ECMO (see below).

High-Frequency Jet Ventilation (HFV, HFJ, HFJV), High-Frequency Oscillatory Ventilation (HFV, HFO, HFOV):
These are ventilation methods that deliver very small air volumes at very high repetition rates. It is used in a variety of situations including acute respiratory failure, respiratory distress syndrome, and risk of lung injury from conventional mechanical ventilation. It may serve as a 'rescue' when the conventional methods fail to achieve the desired result.

Other risk indicator identified with confidence by a physician: Since the inception of the IHP, identification by a physician of risk that is not specified in the remainder of the IHP indicator list has been a distinct indicator in itself. It is impossible to list all conditions that incur valid risk of PHL. Even the indicators listed may be difficult or impossible to discover by any other means and for some potential indicators, the complexity and context-

specificity of disorder expression makes it impossible to derive a simple, practicable risk indicator. It is not intended that physicians adjust listed IHP indicators systematically, such as by declaring all unconfirmed meningitis cases as at risk. However, in situations such as pending but unavailable confirmatory test results, it is reasonable that if a physician judges the likelihood of risk confirmation in the individual case to be very high, then proactive declaration of risk is preferable to completely missing the risk. This is especially important given that HIV/measles/mumps have been deleted from the IHP risk indicator list (see below).

Extracorporeal Membrane Oxygenation (ECMO) or Extracorporeal Life Support (ECLS): An invasive life-support technique in which blood is routed through an external gas exchange system for oxygenation and CO2 removal. In neonates, it is used in situations of severe respiratory failure for which less drastic methods have not succeeded. The duration of ECMO is typically seven to ten days, with the intent of life support during development or recovery of improved lung function. ECMO is frequently used in cases of PPHN and CDH. It is not well-understood whether ECMO itself or its indicating conditions (or both) are the main contributors to a high rate of PHL in ECMO survivors, including late-onset PHL.

3.9 MODIFICATIONS TO CURRENT RISK INDICATORS

Based on preliminary evidence review, program data, and availability of IHP resources, some risk indicators have been modified as follows:

3.9.1 FAMILY HISTORY

Family history of parent or sibling identified with PHL by 10 years of age: The main changes for this risk indicator is that infants receive an AABR screen and are discharged from the IHP rather than entering into Surveillance. This indicator is only useful if it is identified accurately, which can be a challenge if the appropriate information is not gathered by the Hearing Screener. As such, it has a long history of false-positive identification which imposes a significant strain on audiology assessment resources. A reduction in false-positive identification is expected with the inclusion of a genetic panel within the risk factor screen. The genetic panel within the risk factor screen focuses on some common genetic changes affecting three genes associated with PHL. These account for some of the most common recessive mutations but not an exhaustive screen of all sequences. In addition, the results of the risk factor screen may not be known at the time of the hearing screen. Therefore, accurately identifying a family history must be conducted in the best way possible.

In the absence of the result of the risk factor screen, the family history risk indicator should be identified using a strictly-followed script for questioning of family members by IHP Hearing Screeners. Both questions must be asked:

1) ***Do either of the baby's parents have a hearing loss in one or both ears constantly since age 10 years or less?***

2) ***Are there any full siblings to this baby?***
   If yes to #2: Does that child/ Do any of the children have a hearing loss in one or both ears constantly since age 10 years or less?
   If yes: Was there a recommendation for use of a hearing aid(s) or cochlear implant(s), or attendance at a Provincial School for the Deaf for the child/children?

Only clear and definite affirmative responses should be accepted as placing the infant at risk on this indicator. Note that half siblings who have PHL prior to age 10 years do not put the infant being screened at risk.

Given that the IHP has been in place since 2001, any sibling with PHL diagnosed by age 10 is likely to have been involved with the IHP and may have received hearing aid(s) and possibly cochlear implant(s). Alternatively, they may have been raised with attendant sign language service provision and may have attended or may still be attending one of the Provincial Schools for the Deaf. While no question is perfect, it is better to ask very simple and direct questions, rather than some more general question about hearing loss in early childhood.

If either question yields a clear positive response, the infant is at risk on this indicator. If there is any doubt, consider the infant as not at risk. Scoring this indicator as present when it is not can cause unnecessary anxiety and concern in the family, which must be avoided as strongly as possible.

Note that if a permanent hearing loss since before age 10 is reported with confidence, it does not matter what was the cause of the loss. A definite report of constant hearing loss is sufficient. The wording of the questions above define very simple and direct questions.

An infant with a family history of PHL will be screened using AABR. If the infant passes the screening, s/he will be discharged from the IHP and not included in a surveillance sequence. If the infant has a family history that is related to an inherited syndrome associated with late-onset or progressive hearing loss, the infant could still be eligible for surveillance when the syndrome is diagnosed by a physician.

Those infants who screen positive on the risk factor screen for genetics will be re-directed to audiology for assessment regardless of any previous hearing screen outcome. Most of these children will be found to have hearing loss at birth. For those where the audiology assessment indicates that there is no hearing loss at that time, additional surveillance will be scheduled.

3.9.2 OTHERS

Atresia/Microtia: The description for this risk indicator has changed from *obvious craniofacial anomaly* to *atresia/microtia.* While there are many abnormal features of the head and neck that may be associated with PHL risk, considerable expertise is required to identify them accurately. Also, the family may not be aware of the abnormality, unless it has already been clearly identified medically as syndromic. Therefore, given that medical records or medical/nursing staff may be consulted regarding syndromes, flagging of craniofacial malformations for this risk indicator is restricted to abnormalities that are obvious to the layperson, in particular, atresia and microtia. Atresia is identified with absent, closed, or slit-like external ear canal openings. Microtia includes absent or grossly malformed ear(s) (see Figure 2). Atresia and microtia can have varying grades of severity.

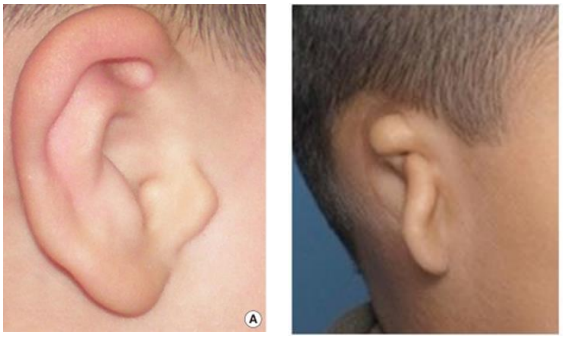

Figure 2: The picture on the left is an example of auditory ear canal atresia. The picture on the right is an example of microtia of the ear. (Source: Google Images)

CHARGE: The letters in CHARGE stand for: Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of Growth and development, and Ear abnormalities and deafness. Reports from large pediatric centres in Ontario indicate infants with CHARGE are confirmed to have PHL almost 100% of the time. As such, identification of this syndrome indicates a screening bypass and referral directly to audiology for assessment.

Proven Congenital Cytomegalovirus (cCMV): Medical diagnosis of symptomatic cCMV infection external to the risk factor screen may occur in hospital. This remains a part of the risk assessment completed by the Hearing Screener and continues to be a bypass risk indicator. As described in the Risk Factor Screen section, cCMV can also be screened with parental consent. As a result, infants who screen positive for cCMV who are not identified in hospital will also have access to earlier identification and intervention, including possible treatment. Infants who

screen positive for cCMV on the risk factor screen will bypass any further hearing screening and go directly for an audiology assessment.

Proven Meningitis: Meningitis is inflammation of the membranes (meninges) lining the brain and spinal cord. It is an IHP risk indicator regardless of the specific pathogen involved (bacterial, viral, fungal, etc.), but only if the presence of the pathogen is *proven* by a medical record or medical report. Family verbal report is not sufficient. Neonatal meningitis is usually caused by vertical transmission during labor and delivery. It occurs most frequently in the days following birth and is more common in premature infants. If meningitis is suspected, antibiotics are typically initiated immediately and may eliminate the pathogen. If the pathogen's presence has not been confirmed, the infant is not at IHP risk on this indicator.

Cleft palate: Cleft palate has a strong association with hearing loss, both permanent and temporary. Cleft lip alone (isolated cleft lip) should, however, be used as a cue for search of any medical record or medical/nursing report confirming presence of a cleft palate. Cleft lip was included in the previous list as a simple cue to possible cleft palate, but isolated cleft lip with no palatal cleft is common and not associated with PHL. Therefore, cleft lip is hereby deleted as a risk indicator component. Report of cleft palate from a family member should not prevent medical record search. In the absence of medical/nursing confirmation, family report should only be accepted as sufficient if it is clear and definite, with no doubt or uncertainty. False-positive identification of cleft palate is not a benefit for the family, because it will lead to their engagement in needless IHP surveillance.

Hyperbilirubinemia meeting exchange criterion, whether exchanged or not: Bilirubin levels that are hazardous differ substantially according to the infant's age and many other factors, including response to phototherapy. A fixed concentration criterion (e.g., ≥ 400 µmol/L) does not reflect optimally the overall risk of neurological sequelae. The modified wording reflects this clinical complexity. If an appropriate exchange criterion level is met, intensive phototherapy may be sufficiently effective to avoid the need for actual exchange transfusion. It is meeting an appropriate clinical criterion that constitutes the IHP risk indicator. Use of the term 'kernicterus' in a medical record is in itself too variable and subjective to be a useful indicator component.

Other proven TORCHES infection: TORCH is the original acronym for the group of infections: Toxoplasmosis, Rubella, CMV, Herpes Simplex Virus (HSV), and Other. Variations on the acronym have been tried and evolving epidemiology has expanded the 'Other' group. For IHP risk indicator purposes, the only qualifying 'Other' infection for this factor is Syphilis (organism: T.Pallidum), therefore TORCHES is used.

Syndrome associated with hearing loss in early childhood: About 50-60% of all hearing loss in childhood is genetic, about one fifth of which is syndromic (Toriello et al, 2004). There are hundreds of genetically-based syndromes associated with PHL in childhood. The vast majority are rare. Some of the more common, in approximate order of decreasing prevalence at birth are: Down, Pendred/Enlarged Vestibular Aqueduct (EVA), Stickler, CHARGE, Usher, Osteogenesis Imperfecta (OI), Goldenhar (OAVS), Waardenburg, Branchio-Oto-Renal (BOR)/Branchio-otic (BO), Alport, Treacher-Collins, Neurofibromatosis II (NF2), and Crouzon. CHARGE is now an indicator on its own (see above).

3.10 DELETED RISK INDICATORS

Based on preliminary evidence review, program data, and availability of IHP resources, the following risk indicators have been deleted from Ontario's list.

Gestation period ≤ 30 weeks: The 30 week criterion is too liberal. Gestational age estimates are inexact and gestational period *per se* is now considered to add negligible predictive value to that of the other indicators listed.

Cleft lip: Cleft palate has a strong association with hearing loss, both permanent and impermanent, and remains on the list (see above). Cleft lip was included in the previous list as a simple cue to possible cleft palate, but isolated cleft lip with no palatal cleft is common and not associated with PHL. Cleft lip alone (isolated cleft lip) has been

removed and should, however, be used as a cue for search of any medical record or medical/nursing report confirming presence of a cleft palate.

Congenital HIV, Measles or Mumps infection: Although these infections are reoccurring in the general population due to the increase of non-vaccinated infants, the infections are often not proven at the time of the hearing screening. As such, if an infection is proven beyond the hearing screening period (birth to two months of age), the infant should be referred to the IHP by the physician for appropriate services (see Appendix D).

Severe neonatal asphyxia/hypoxia/respiratory failure/cardiopulmonary failure: This indicator was intended to increase the ease with which IHP Screeners could identify infants with hypoxia sufficient to cause serious risk of cochlear injury. However, the terms are now recognized as too subjective and less predictive than the other indicators included within the current list that relate to specific causes, sequelae or interventions associated with severe hypoxia.

Severe neonatal sepsis: This indicator is now recognized to be neither sufficiently predictive of PHL nor specific. The association with PHL is only established if the septicemia leads to proven meningitis, which is a separate risk indicator on the current list.

Neonatal cancer treatment with cisplatin: The issue of PHL caused by either maternal cancer chemotherapy during pregnancy or, very rarely, cancer chemotherapy in the infant, is too complex and situation-specific to be addressed by a simple risk indicator. Additionally, infants undergoing this treatment are included in an ototoxic monitoring protocol which includes hearing.

3.11 HEARING SCREENING PROCEDURES AND RISK INDICATORS

Infants with no assigned IHP Risk Indicator for PHL must be screened using ADPOAE technology. About 95% of all infants are not at risk for PHL (Centers for Disease Control and Prevention, 2007). Initial likelihood of PHL is small and the number of infants large. This favours use of an initial screen (ADPOAE) to quickly filter out all infants with the greatest probability of having normal hearing at that time. The AABR screen takes more time, skill and expense, but is more accurate and will in turn eliminate many infants who are false-positive on ADPOAE. Infants who have a risk indicator for PHL must be screened using AABR technology before hospital discharge.

3.12 DEFINITION OF SUCCESSFUL AND COMPLETE SCREENS

A **successful** screen is a screen giving a Pass or Refer in any individual ear. A **complete** screen is a successful screen in both ears (see Glossary of Terms in Appendix B.) Every reasonable effort should be made to screen both ears of any given infant.

3.13 REQUIRED INFANT STATE FOR SUCCESSFUL SCREENING

Successful screening requires the infant to be asleep or resting quietly throughout the test, regardless of the screening technology used (e.g., ADPOAE, AABR). An infant who is crying or wiggling cannot be screened; **the screen must not be started** unless and until an acceptably quiet state is achieved.

Well-timed visits, manoeuvres to calm the infant if necessary or re-scheduling the screening can usually achieve a complete or at least a successful screen. The screening run may yield a pass or a refer result. A pass finishes the screening process for the passed ear. A pass in both ears finishes the screening for that infant. A refer in an ear may indicate genuine hearing loss and the need for further audiological assessment or may be false-positive (a refer when hearing is actually normal at that time). The most common causes of false-positive refers are ear canal noise due to infant movement or vocalization and/or a borderline probe fit in a situation of unacceptably high environmental noise.

3.13.1 AABR TEST CONDITIONS

The general requirement of a sleeping or at least quietly resting infant also applies to AABR screening. A sleeping infant is much preferred. The requirements for probe tip selection, correct probe positioning, and an acceptable test environment also apply.

In the AccuScreen AABR, the stimulus is a rapidly repeated, brief sound called a chirp, delivered at a low sound level. The screening device records the electrical signals (the auditory brainstem response, ABR) from the infant's hearing nerve pathways in the brain, evoked by the stimuli. These tiny ABR signals are buried in much larger electrical activity from the rest of the infant's brain as well as from the muscles of the head and neck. The computer extracts the ABR "signal" from this ongoing electrical "noise" and, if the signal meets pre-set signal to noise ratio criteria, it records a pass result. The key to extracting the ABR is the size of the electrical noise, which is quite small in a sleeping or relaxed infant but is very large if the infant is tense or moving. Excessive electrical noise may result in a non-start or a false refer.

3.14 INFANT ACTIVITY JUDGMENT, PRESSURE TO SCREEN, AND PROHIBITED 'TRIAL' HEARING SCREENING

For a hearing screen to be started, the infant must be judged to be sufficiently quiet such that the likelihood of a successful hearing screen is high. It is against this protocol to start screening if the infant's state is not acceptable in the Hearing Screener's judgment. It is also out-of-protocol to "trial" screen if an infant is in a questionable state or to use the device display as a "test" of infant state.

This judgment is not always easy and inevitably there is a grey zone of uncertainty. If an infant is clearly active on the initial contact, retrying at a more favourable time such as after feeding is the first option. Where necessary and considered to be worth trying at any given visit, an effort to settle the infant should be made. If neither of these tactics is successful, the hearing screen should not be started and the parent/guardian should be counselled firmly on the importance of attending follow-up hearing screening in the community. A community appointment, while it may be less convenient, is more likely to yield a successful hearing screen and a lower rate of false-positive refers.

3.15 EARTIP SELECTION AND PROBE INSERTION

IHP Lead Agencies must provide an appropriate range of eartip sizes and styles to Hearing Screeners at all times. Lack of basic supplies to do the job is not acceptable. It is important that eartip inventory and availability be tuned to what works well in the Hearing Screener's hands in the local infant population and local experience with the AccuScreen.

After physically inspecting the ear canal opening and superficial cleaning of fluid or debris, if needed, selection of the appropriate size and type of eartip is critical to achieving a successful test. The better the probe fit, the more stable is the insertion and the greater the immunity to environmental and physiological noise. A probe falling out by gravity or due to small movements of the infant's head clearly indicates inadequate fit, as do stimulus calibration difficulties or high noise levels in an apparently quiet infant. The eartip chosen should be the largest that can just be inserted deeply enough in the ear canal to be securely retained. A tip that is too small will go in too easily, will not exclude room noise, will probably cause a poor probe fit error or will not give a successful calibration because of air leakage past the tip preventing achievement of correct stimulus calibration levels or allowing in too much room noise. A loose tip also generates noise itself by moving against the ear canal wall. A tip that is too large will not enter the ear canal deeply enough for secure retention by the canal wall and is also likely to fall out.

It has been determined that the flanged tips (treetips) are usually the most useful. The flexible flanges can accommodate a range of canal sizes and shapes, while providing a sufficient acoustic seal. They also provide better retention because the flanges tend to expand and resist falling out or being pulled out. The eartip must be inserted correctly into the ear so as to achieve a firm, secure fit, low air leakage, and good exclusion of room noise.

This is done by gently but firmly inserting the eartip into the ear canal using a quick quarter turn screwing motion on insertion while gently pulling out, back and slightly up on the pinna (see Training Video). A good insertion will typically take no more than two seconds and is unlikely to arouse the infant, whereas a slow, tentative or misaligned insertion is far more alerting. Appropriate tip section and effective tip insertion are techniques that come with practical experience and good understanding of the acoustical and physical factors involved.

The probe body itself should **never** be held during hearing screening. The probe lead may be supported if absolutely necessary. Please use the cord clips provided to diminish the weight caused by the probe head (Figure 3). However, the question arises as to what factors render such holding/support necessary and what the effect of those factors might be on screening success and error rates. If the probe would otherwise fall out, then the fit is not adequate. If the infant is highly active, the conduct of the screening at that time is inappropriate.

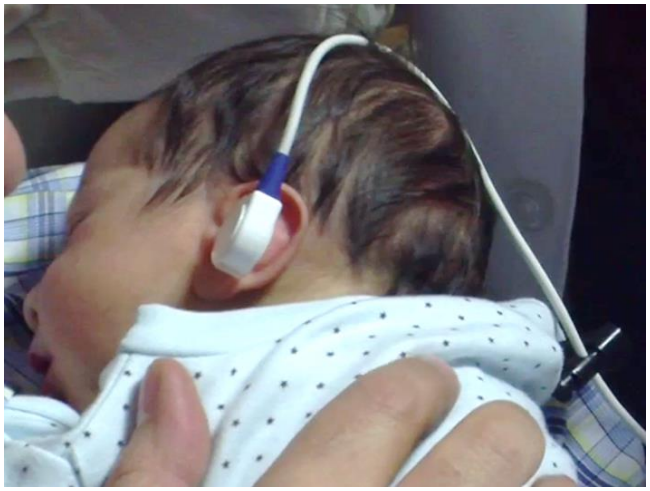

Figure 3: Example of probe in infant's ear using cord clipped to blanket.

3.16 ELECTRODE POSITION FOR AABR

AABR is done with electrodes placed on the high midline forehead as close as possible to the hairline and on the mastoid behind each ear. **AABR is NOT to be done using an electrode on the nape of the neck.**

Although the AccuScreen manual indicates to use an electrode on the nape of the neck, the IHP does NOT use a nape electrode site. The properties of the ABR recorded between the high forehead and the mastoid area behind each ear, both on the same side and the opposite side to the ear being stimulated are well-documented (Vander Werff et al, 2009; Casali & Santos, 2010; Ribeiro & Carvallo, 2008). Although there is substantial ABR development at the nape, there are historical data suggesting that electrical noise levels are generally higher at the nape than at the mastoid (Sininger et al, 2000; Pethe et all, 1998). Also, physical access to the nape is often less convenient and the attachment process there is consequently more likely to disturb and alert the infant.

The white electrode must be placed on the forehead in the midline, a line extending upward from the bridge of the nose and dividing the forehead into equal left and right halves. The white electrode should be placed as high as possible on the midline forehead, because the ABR gets bigger towards the top of the head, so the electrode pad should be placed as close as possible to the hairline, but not on top of hair. The red electrode should be placed on the skin of the right mastoid, as close as possible to the level of the external ear canal opening, immediately behind the pinna (outer ear). The black electrode should be placed similarly, but behind the left ear (see Figures 4 and 5).

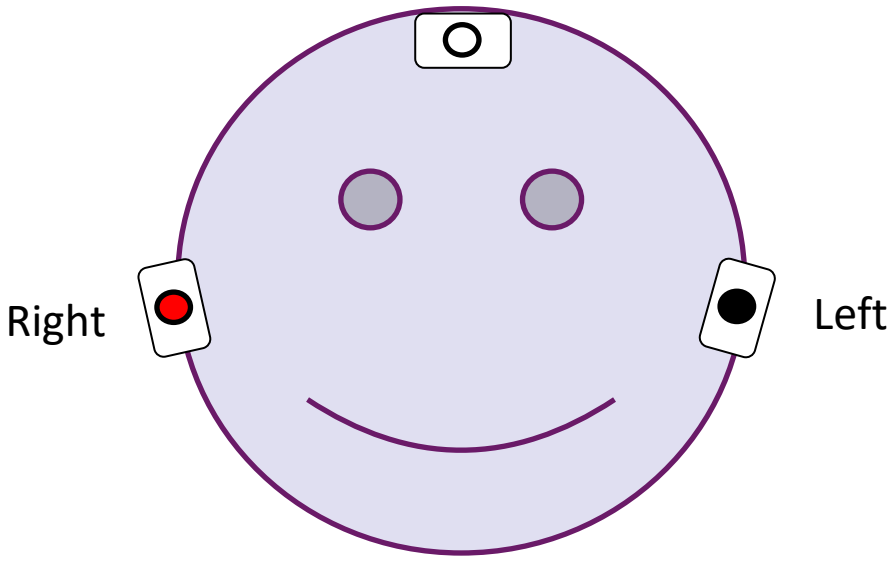

Figure 4: Schematic of AABR electrode placement. The top (white) electrode is placed on the forehead, the right (red) electrode is placed behind the right ear, and the left (black) electrode is placed behind the left ear.

Figure 5: Placement of electrodes on infant for AABR.

3.17 AABR ELECTRODE ATTACHMENT AND IMPEDANCE CHECKS

Causes of AABR non-starts include high or unbalanced electrode contact impedance, an overly active infant and inappropriate probe eartip fit. Close attention must be paid to secure electrode attachment and to the quality of the electrode-skin contact.

Impedance is a slightly more complicated version of the more familiar concept of electrical resistance. It refers to the quality of the connection between the electrode ("sensor" is a term sometimes preferred for use with families)

and the infant's skin. The better the connection is, the lower is the impedance. A good connection is less affected by any movement of the skin or the electrode leads. It also picks up less electrical interference from sources such as electric lights, cables, wall outlets and nearby electrical devices.

Extraction of the tiny ABR from electrical noise depends on the ability of the device to reject electrical noise that is similar at both the forehead and mastoid sites. This rejection ability is much reduced if the contact impedances at the forehead and mastoid are very different from each other. This is what is reflected in the 'balance' measure on the AccuScreen. The best measurement conditions have both low impedance and good balance of impedance. The electrode impedance test will reflect the forehead and right mastoid, not the forehead and nape. The absolute size of the 'forehead' impedance reflects the contact quality of both the forehead and left mastoid electrodes, while the right mastoid impedance (called "nape" on the AccuScreen) reflects the contact quality of the right and left mastoids; in both cases, the left mastoid is being used as a "ground" or "common" electrode to complete the required electrical circuit. The 'balance' impedance is actually the difference between the forehead and right mastoid impedances, because the left mastoid impedance is common to both and is subtracted out.

It is not possible to override the impedance checks in the AccuScreen. This is appropriate with respect to encouraging higher quality of electrode contact and, therefore, better screening device performance in terms of false-positive and false-negative error rates. A consequence, however, is that good electrode contact technique is essential in order to achieve a high success rate in test start-up.

Historically, practices with respect to cleaning of electrode sites for screening have been variable across the IHP. This protocol makes recommendations based on clinical experience over several decades in diagnostic ABR assessment, for which good skin contact is especially important. The main recommendation is that if any difficulty with electrode impedance is encountered, the skin at the high-impedance site should be **gently** cleaned with a prep pad such as "TENS Clean Cote". Such a pad removes skin oils and improves both electrode adhesion and contact impedance. Further improvement can be encouraged by the use of a **small** dab of a clear electrode gel such as Lectron II in the centre of the electrode pad. If gel is used, the pad should be applied to the skin with light pressure on its outer margins, not on the centre of the pad. If central pressure is used, an air pocket may be created under the pad when pressure is removed.

Screeners may choose to clean the skin and/or to use gel (or not) initially, not waiting for a possible poor impedance check. This is discretional. In newborns it is not appropriate to use abrasive pads or gel such as Nuprep to clean the skin, as the abrasive component is appropriate for use in adults and older children, but not in newborns.

3.18 ACCUSCREEN CALIBRATION TIMEOUT AND OTHER NON-START MESSAGES

On starting the screen, the AccuScreen tries to establish correct stimulus levels in the test ear (auto-calibration) and gives error messages if the stimulus and the noise levels in the test ear are not both within acceptable limits. Common causes include poor eartip fit, a wholly or partially blocked eartip, or a collapsed ear canal. The screener must address each of these issues, if and when they arise (see below). If calibration is successful, the screening data acquisition run will start automatically.

3.19 EARTIP/PROBE TIP BLOCKAGE AND INSPECTION

Complete or partial blockage of the eartip can be caused by debris or fluid in the ear canal or by direct eartip contact with the ear canal wall. The screen may indicate an error condition if the blockage is substantial, or may run and possibly give a false refer result. In either case, the probe must be removed from the ear and inspected. If fluid and/or debris are present, the eartip should be cleaned or replaced, the probe inspected and cleaned if necessary, then re-inserted correctly and the screen re-attempted.

Most newborns will have some amount of birth tissue debris and/or fluid in the external ear canal initially. Ear cleaning and suction practices in birthing units are variable. As noted earlier, fluid/debris will resolve over time by gravity, re-absorbance, and evaporation, but this may take hours or even days. The fluid/debris may entirely or

partially occlude the eartip or, if excessive, may enter the probe tip itself. Total occlusion will prevent the screening device from calibrating, whereas partial occlusion may permit starting but yield false refers. Similar effects may arise if the eartip opening contacts the ear canal wall, which may occur due to probe misalignment with the canal axis, anatomical anomaly of the canal or temporary canal collapse (see below).

If the screen runs and a refer result is obtained in a quiet infant, partial tip occlusion is a possibility. The probe must be removed from the ear and inspected for debris/fluid. If present, the eartip should be replaced, the probe tip cleaned or replaced as necessary, the probe should be correctly re-inserted into the ear and the screen repeated. Preferably, the repeat screen should be done immediately following the refer on any ear.

3.20 EAR CANAL COLLAPSE

An infant's external ear canal walls are pliable and the canal may close (collapse) partially or completely, if the ear has been against the head for a lengthy period, such as when sleeping in the caregiver's arms. Canal collapse typically occurs on one side only, usually related to a recent sleeping position.

The canal may open gradually after pressure is removed, so instruction to the mother about infant positioning and a revisit may prove successful. Ear canal opening can be encouraged by gently pulling the pinna (outer ear) outward and massaging the area just in front of the ear with a circular motion for up to about 30 seconds. Repositioning the infant and manipulation to open the canal should be tried if the infant state is favourable for an immediate attempt. If a re-visit is preferred, the caregiver should be advised to keep the ears to be screened free from physical pressure against the sides of the head.

3.21 ENVIRONMENTAL NOISE

Excessive noise in the screening environment can arouse a sleeping infant or prevent settling. As a rough guide, if one-on-one conversation in a soft voice can easily be heard at a distance of about one meter, the noise level is acceptable for screening. The effect of environmental noise on a screening run depends on the quality of the eartip fit; the better the fit, the lower the likelihood of an error message or a false-positive screen.

While the screener's control over the environment may be limited, screening in an excessively noisy environment is not appropriate and will lead to low success rates, increased test times, and high false refer rates, all of which are counter-productive and costly. Obvious improvements in environment include taking any practicable steps to avoid or at least minimize interruptions, suspending conversations in the immediate vicinity while the screening data acquisition is actually running, and turning off cell phones. For ADPOAE screening, it is acoustic noise that matters, whereas for AABR screening, direct electromagnetic interference from cell phones, computers, or equipment in the room may be an additional concern.

Screening in an environment that is permanently noisy or disruptive to infant state is completely inappropriate. If such a practice occurs and leads to high rates of screening non-starts or very high refer rates, such screening is persistently out-of-protocol. Hearing Screeners should contact their IHP Regional Trainer or their IHP Coordinator to raise this as an issue and to discuss potential solutions to the problem.

3.22 MESSAGING TO FAMILIES REGARDING AUDIOLOGICAL ASSESSMENT

When an infant has obtained an overall refer result from screening, regardless of presence/absence of risk, the likelihood of PHL being present has increased on average to at least one in 15 and frequently is higher than that average value. It is extremely important that the family promptly bring the infant to the audiological assessment appointment. Families often do not understand the difference between an assessment and a hearing screening. It is important to follow the scripts in Appendix F to explain that a screening test refer means that hearing loss is now more probable but is not certain and even if it is present, it might be a temporary hearing loss. An audiological assessment, on the other hand, determines the **true** state of the child's hearing, measures exactly what the child can or cannot hear, in each ear, finds out where the hearing problem lies and indicates what should be done about it, whether medically or by other means. The screen refer means one crucial thing: that there must be an

assessment as soon as possible. **That is the key message and the family's understanding and agreement to attend are the top priority.**

The second issue is that families often believe that they can tell whether their infant can hear. **That is absolutely not true and that fact needs to be made as clear as possible, by using the scripts provided.** An infant can be deaf in one ear but have normal hearing in the other, while sound always travels to both ears. There may be a hearing loss at high frequencies but not at low frequencies, or vice versa, so the infant may respond to some but not all sounds and families often assume that any response at all means that the hearing is fine. Hearing loss can prevent hearing of soft sounds only, but loud sounds such as a dog barking may be heard. Doors slamming cause structural vibration, which the infant may sense physically. When families try to 'test' an infant's hearing at home they often whisper in the infant's ear; the infant responds not to the sound but to the breath, or may be able to see the family member and turn accordingly. Even skilled Audiologists cannot assess the hearing of an infant under about six months of age with any certainty by observing the infant's behaviour. **Electrophysiological testing (OAEs and ABRs) is the only reliable way to test hearing in young infants**.

3.23 THE LIKELIHOOD OF PERMANENT HEARING LOSS

As the infant progresses through the screening, the probability of PHL increases after each refer result. It is important that the screener be aware of these probabilities, which may also be useful in explaining test results to some families.

The probability of a newborn chosen at random having PHL is the population *prevalence* of congenital PHL, as explained in the introduction to this protocol. It is about two per thousand (2/1000 or 0.2 %). The approximate, average changes in PHL probability at various stages are shown in Table 2 below (Meyer et al, 2009; Haghshenas et al, 2014):

Table 2: Average changes in PHL Probability at Various Screening Stages

	No Risk	At Risk
Pre-screening	1/1000	1/100 to 1/25
Stage 1 Refer	1/100 to 1/50	1/10 to 2/5
Stage 2 Refer	1/15	

In At Risk infants, the final probability of PHL after a refer varies depending on the risk indicator; it can range from below 1/10 to as high as 1/1, which is 100% certainty (such as in an infant with craniofacial anomaly risk and no external ear).

3.24 WHAT IS AN IHP AUDIOLOGICAL ASSESSMENT?

An IHP Audiological Assessment is not a hearing screening test but a detailed evaluation of the infant's hearing by an IHP Audiologist with special training. It includes several types of tests, including advanced techniques using the ABR, DPOAE and tympanometry. It is done while the infant is asleep and can take about an hour or more. It determines exactly what the infant can and cannot hear, the type and location of any disorder in the infant's auditory system and what can be done to address any hearing loss.

Audiological assessment measures the softest sounds that an infant can hear (hearing 'thresholds') for various frequencies or 'pitches' of sounds. For any infant under about six months of age, thresholds are measured with the auditory brainstem response (ABR), a brief (1/100th of a second) electrical waveform that reflects activity in the nerves of the auditory system in the base of the brain and can be measured by sensors placed on the head. It is the same response that is measured automatically by AABR screening devices such as the AccuScreen. In the assessment, the ABR thresholds are measured under manual control for various frequencies of special sounds (brief tonepips, with frequencies from 500 to 4000 Hertz, Hz, cycles per second). For example, a tuning fork at

'middle C' for a piano has a frequency of 512 Hz. The sounds are delivered both by earphones (air conduction) and by small vibrators placed behind the ear (bone conduction).

If the thresholds are higher than normal, hearing loss is present. If they are different by air- and bone conduction, there is a conductive hearing loss, whereas elevated thresholds for bone conduction reflect sensory-neural hearing loss (SNHL). SNHL is almost always permanent and is caused by a disorder affecting the tiny 'hair cells' in the cochlea, the spiral organ of hearing in the inner ear. Conductive hearing loss is a disorder of sound conduction to the inner ear, usually caused by fluid inside the middle ear (the air space behind the eardrum) or by a disorder of the tiny bones (ossicles) in the middle ear that conduct sound from the eardrum to the cochlea. Sometimes, there is a disorder of the hearing nerves or their connections to the cochlear hair cells; the most common of these (in about 7 % of all permanent hearing loss in young children) is called auditory neuropathy spectrum disorder (ANSD), which can be detected using manual otoacoustic emission (OAE) and ABR testing together.

IHP Audiological Assessment of infants who refer from screening includes ABR, DPOAE, and other tests like tympanometry (a test that measures sound reflection from the eardrum), to build a complete picture of the hearing threshold pattern and the place(s) in the auditory system where the disorder lies. A sophisticated computer system is used to measure the ABRs and the OAEs. Because the assessment is based on ABR thresholds, it is called ABR-based Audiological Assessment or ABRA. The testing is complex, requires a lot of skill and is done while the infant is asleep; it may take over an hour of sleep during a two hour appointment, depending on what is found. The infant must be brought to the test hungry and tired, to facilitate sleep. The test is done by a specially-trained audiologist, sometimes with help from a second audiologist, nurse or other person.

In some IHP regions, efforts are being made to allow 'reserved' appointments or blocks of appointments that can be booked by the hearing screener at the time of referral from the final AABR screening. Immediate booking of assessment appointments has been shown to be an effective step in reducing family non-attendance or delay in attendance. It is important to make the most effective use possible of any reserved appointments that may have been made available in specific locations.

3.25 URGENCY OF IHP AUDIOLOGICAL ASSESSMENTS

As already noted, audiological assessment appointments should occur **at the very latest** by eight weeks corrected age, provided that the infant is medically stable. Especially for infants at risk for PHL, the target is that the first audiological assessment appointment be attended by an infant age of six weeks corrected, wherever possible. The age of the infant has a strong effect on the ease with which a complete audiological assessment can be obtained quickly. Infants' sleep patterns change dramatically over the first two or three months of life. Usually, infants up to about eight weeks of age sleep a lot and it is relatively easy to induce them to sleep for long enough that the required result can be obtained reliably. At four to six weeks, most infants can be tested swaddled while laying supine in a bassinet. This allows the audiologist to test both ears much more efficiently than if the infant is in the mother's arms. Under about four weeks of age, some infants are too small to allow easy attachment of all the necessary electrodes and stimulus transducers, so the ideal age in most infants is **about four to six weeks corrected age**.

By the time an infant is older than eight weeks, in the event that more than one appointment is needed to complete an assessment, it can become very difficult to obtain the period of sleep needed to finish the testing. This is problematic, because the results at that point may be incomplete or inaccurate and it may be necessary to consider testing under sedation. This is an expensive and much more complex proposition, and limited access to sedation facilities may cause major delays in getting a complete audiological assessment. This reduces the benefit of early hearing screening and can compromise the achievement of optimal early intervention for the infant who has significant, permanent hearing loss. That is why it is so important to get to audiological assessment sufficiently early, wherever humanly possible.

SECTION 4: SCREENING OF INFANTS NOT AT RISK FOR PHL

4.1 SCREENING PROCEDURE AND EXPLANATION

Infants who are not at risk for PHL must be screened using ADPOAE. The Madsen AccuScreen using the DP 5 protocol is used for this procedure within the IHP. For all infants who are to be screened, the screening procedure itself must be explained in advance and any questions from the caregiver must be answered. It is especially important to explain the need for the infant to be asleep or at least resting quietly and that the screening can only be initiated if the screener considers the infant's state to be likely to yield a successful screen. See Appendix F for scripts.

The DPOAE itself is a very faint tonal signal generated by motion of the hair cells in the cochlea, the site within the inner ear at which mechanical vibration due to sound is converted to electrical nerve impulses. The OAE is transmitted back out through the ossicles (middle ear bones) and the tympanic membrane (eardrum), to be picked up by the miniature microphone in the OAE probe. The microphone also picks up any sound in the infant's environment that leaks past the probe eartip into the canal (hence the need for a proper ear tip fit), as well as any sound generated by the infant's vocalizations or physical movements of the head, the probe eartip or its leads. These constitute the "noise" background from which the DPOAE must be isolated and recognized by the screening device. If either or both of these noise sources is excessive, the signal to noise ratio may fail to achieve a pre-set criterion size and the result will be DPOAE detection failure and a refer outcome.

4.2 ROLE OF ADPOAE AS STAGE 1A OF A SERIAL HEARING SCREENING PROCESS

About 95% of all infants are not at known risk for PHL. The choice of ADPOAE rather than AABR is a balance among many factors, including the initial likelihood of PHL, screening efficiency, speed, accuracy, and program resources required. In infants with no risk indicators, the likelihood of PHL is small and the number of infants is large. This supports use of an initial screen that will rapidly and easily filter out many infants who have normal hearing. AABR is more time-consuming than ADPOAE and about five times more costly overall, but it has higher sensitivity to PHL (about 99% of infants refer when PHL is truly present) and a lower false-positive rate (about 2% of infants refer when PHL is truly absent) because it is less affected by contraindications such as birth fluid in the middle-ear. ADPOAE is faster and easier as well as having an acceptable sensitivity (about 90%), but it has a relatively high false-positive rate (typically 5-15%). The challenge with ADPOAE is to hold down the false-positive refer rate, preferably to below 10%. ADPOAE then acts as an effective tool that increases the probability of PHL in the infants who refer, with no more than about one in ten infants needing to have a re-screening by AABR.

Screening before hospital discharge has the advantage of relatively easy access to the mother and infant, which facilitates high screening coverage of the newborn population as well as the earliest possible start on a path to intervention if a PHL were present. While refer rates tend to be lower when screening is done after discharge from hospital, the logistical, family contact, and appointment attendance challenges with universal post-discharge hearing screening are often substantial. Generally, it is more effective overall to screen with a modest refer rate than to screen late or not at all.

4.3 TIMING OF PRE-DISCHARGE ADPOAE SCREENING: VAGINAL BIRTHS

Infants born vaginally should be screened as late as possible before hospital discharge and **not less than 15 hours after birth.** Infants who are not screened pre-discharge will be provided the opportunity to be screened in the community.

ADPOAE screening is highly susceptible to false refers (false-positive screens) in the presence of fluid or other birth debris in the external ear canal or the middle ear space directly behind the eardrum. The fluid or tissue debris can interfere with stimulus transmission to the inner ear and transmission of the OAE generated in the cochlea back out to the ear canal probe. Fluid in the external ear canal itself usually resolves rapidly in the first few hours after birth, whereas fluid in the middle ear may on occasion take a day or more to disappear.

Historical IHP data on initial ADPOAE refer rates expected from experienced, high-caseload screeners using a previous screening device (Natus/Bio-Logic AuDx) for infants delivered vaginally as a function of the infant's age, in hours, declines after 15 hours post-natal. These data indicate that prior to 15 hours of birth, the refer rate was about 15% and decreases to about 4% after about 30 hours post-natal. As such, infants should not be screened by ADPOAE within 15 hours of birth. Beyond 15 hours, infants should be screened as late as is practicable before discharge. After 20 hours is better still, with an expected refer rate of about 10%. There is no strong justification to withhold pre-discharge screening for an infant who is over 15 hours of age, provided that access to the infant in a quiet state can occur. Similarly, there is no strong justification for using a 24 hour minimum age criterion for screening an infant delivered vaginally, which is a common misconception.

Initial screens that are false-positive cause undue harm to families, as a result of needless anxiety. At some point, the ADPOAE screening false-positive rate becomes unacceptable, largely because of the AABR workload it generates and the number of families who must wait for Stage 1b in the community. Since most ADPOAE refers are false-positive, this means that refer rates must be as low as possible. There is no question that an ADPOAE refer rate over 15% is inherently unacceptable. If local hospital discharge policies result in discharge under 15 hours, the infant must be screened in the community.

4.4 TIMING OF PRE-DISCHARGE ADPOAE SCREENING: CAESARIAN BIRTHS

Infants born by Caesarian section should be screened as late as possible before hospital discharge ideally after 36 hours and **not less than 22 hours after birth.** If that timeline cannot be met, the infant must be screened in the community.

Infants delivered by C-section are much more likely than vaginal deliveries to have unresolved middle-ear fluid and resulting very high false-positive rates (Smolkin, Awadeh, Blazer, Mick, Makhoul, 2013). The later they are screened before discharge the better and they should **not** be screened with ADPOAE within 22 hours of birth. The 22 hours criterion hours allows a window of opportunity in case of a 24 hour discharge policy for C-section normal deliveries. If practicable pre-discharge, screening after a full 24 hours is preferable in C-section infants.

4.5 NUMBER OF STAGE 1A ADPOAE HEARING SCREENING ATTEMPTS

A Screening Attempt occurs when a probe is placed in the infant's ear and the screening is started on the equipment. A Successful Screen is a final screening event for that stage where a pass or refer result is obtained in any individual ear (see Glossary in Appendix B). **The maximum permissible number of hearing screening attempts for Stage 1a ADPOAE in any given ear is four.**

4.6 SWITCHING TO AABR

If the infant's state appears to be entirely satisfactory but the ADPOAE screen will not run despite several attempts, switching to AABR is an option. For example, attempt one gives a poor eartip fit, attempt two a blocked eartip/probe, and attempt three a calibration timeout. The screener has the option to switch to AABR at that point, given family permission, provided that the infant remains quiet. In most such cases, the AABR will be successful and it is advisable to stay with AABR for the other ear, if it has not yet been screened.

Similarly, if an infant has an ADPOAE refer result in one ear, it is already established that the infant will require an AABR Stage 1b. An AABR should be conducted immediately in both ears, starting with the ear that referred, if the infant remains in an appropriate state.

4.7 REPEAT HEARING SCREENING OF ALL ADPOAE REFERS: DOUBLE-REFER

ADPOAE screening must be repeated in any ear that refers if the infant state and environment are ideal.
Statistical influences within the equipment and unknown eartip/ear canal conditions may be the cause. The latter can be assessed by eartip/probe removal, inspection, and cleaning after the first refer. The repeat screen may be done immediately, following the eartip and probe inspection that must be done after the first refer. Do not re-run the ADPOAE without eartip/probe removal, inspection, and re-insertion. A pass on the repeat overrides the initial

refer. A refer on the repeat screen (double-refer) is an overall refer in that ear. **A double-refer with ADPOAE in a given ear is required to obtain an overall refer for that ear.**

There may be situations where a double-refer cannot be achieved (e.g., maximum number of attempts have been used, infant not in an appropriate state to continue). If only a single refer can be achieved, then the infant must move onto the next screening stage. The reasons for this action must be clearly documented.

4.8 MAXIMUM NUMBER OF ADPOAE ATTEMPTS AFTER A REFER

If a double-refer is obtained in a given ear on ADPOAE, **there must be no further ADPOAE screening in that ear.** The next screening event in that ear must be an AABR. Recall that the maximum permissible number of screening attempts for ADPOAE in any given ear is **four**. If a screening starts, **it must not be aborted** (see Figure 6 below).

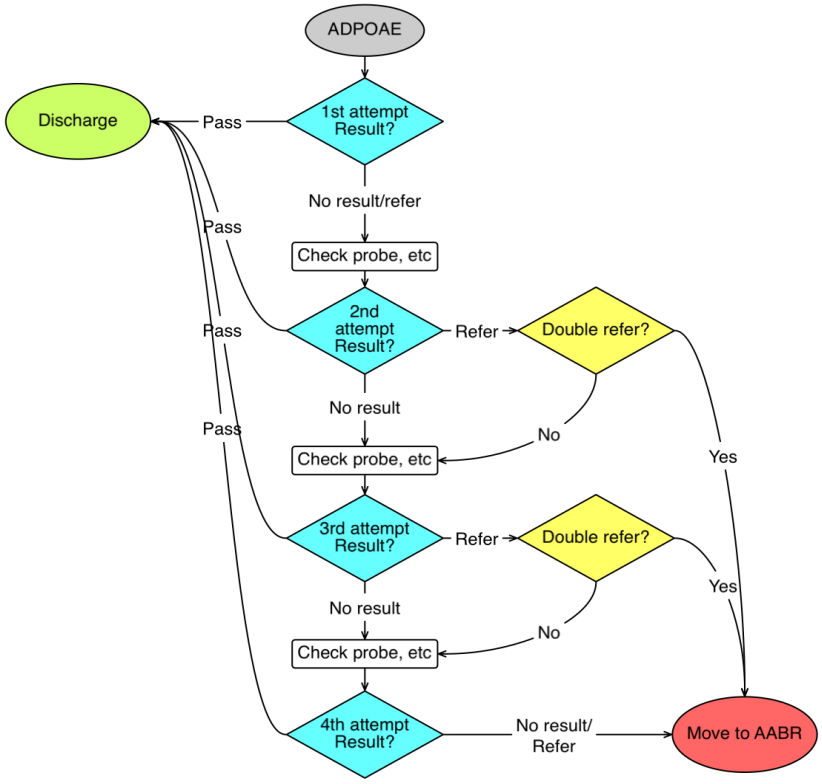

Figure 6: ADPOAE workflow

Examples:

1) Refer + refer = Refer = AABR
2) Refer + poor probe fit + cal'n timeout + cal'n timeout = Refer = AABR
3) Cal'n timeout + refer + cal'n timeout + refer = Refer = AABR
4) Cal'n timeout + cal'n timeout + too noisy + too noisy = No Result = AABR
5) Poor probe fit + refer + cal'n timeout + pass = Pass (no further screening)
6) Refer + Pass = Pass (no further screening)

Every time an infant who has a genuine PHL is screened, there is a small chance of missing the hearing loss at random and getting a false pass (false-negative screen). For ADPOAE, this probability may be as high as 2-3%, though this is very difficult to know exactly. These errors accumulate in proportion to the number of screens. If the

(totally wrong) practice were to screen 5 times, hoping for a pass, the overall probability of missing a PHL due to chance could be as high as 10-15%, which is unacceptable.

4.9 SCREENING THE OTHER EAR AFTER A FIRST-EAR DOUBLE-REFER ON ADPOAE

If the first ear screened yields a double-refer, where feasible the other ear should still be screened by ADPOAE, because a pass in one ear may reduce family anxiety prior to the AABR re-screen. This is important if the AABR is not available or feasible pre-discharge, in which case the family may wait several days or even weeks in a state of anxiety.

4.10 EXPLAINING HEARING SCREENING RESULTS TO FAMILIES: STANDARD SCRIPTS

Explaining hearing screening results to families simply and accurately is vital. Regardless of the screening result, balancing the importance of follow-up without causing undue anxiety for families is key. To ensure simple, accurate and supportive information is provided to families, standard scripts must be used by IHP Screeners and the relevant IHP Fact Sheets provided (Appendix F). This is common practice in EHDI programs worldwide.

A pass on ADPOAE in **both** ears is an overall Stage 1a Pass. The meaning of a pass result is that the infant's hearing is normal at this time. Families also should be advised that hearing loss can occur at any time later during their child's development, so it is important to always pay attention to their child's responses to sound. If the family becomes concerned about their child's ears or hearing at any time they should contact a local healthcare professional. The appropriate IHP Fact Sheet for infant's who pass IHP hearing screening must be provided.
In contrast, a refer on **either or both** ears is an overall Stage 1a Refer. It means that hearing loss is a possibility and another type of screening test, the AABR, will be conducted.

4.11 AABR SCREENING: STAGE 1B OR STAGE 2

For any infant with any ear giving a pre-discharge ADPOAE double-refer, screening with AABR is required. Whenever feasible, the AABR should be done before discharge. If pre-discharge AABR is not available, the AABR is to be done at the most convenient site in the community. The community re-screening appointment should be made available as soon as possible and, whenever feasible, **no later than within four weeks of hospital discharge**.

All infants who double-refer from ADPOAE must receive AABR screening. The benefit of doing these screens immediately (if infant state permits), or at least before discharge, is that a majority of the refers from ADPOAE will pass AABR, it being much less affected than ADPOAE by minor, usually temporary conditions such as fluid in the middle ear. It follows that doing the AABR before discharge will eliminate quickly much family anxiety associated with false-positive ADPOAEs.

AABR screening must be done when the infant is medically stable and **never under 34 weeks gestational age.** If the infant's estimated gestation period was at least 34 weeks, pre-discharge AABR may be done at any time. However, regardless of the type of delivery (vaginal or Caesarean), the longer the interval from birth to hearing screening is, the lower is the likelihood of ear canal or eartip blockage due to fluid or tissue debris. Notwithstanding this, the AABR screen is far less sensitive to minor blockages than the ADPOAE screen. As a result, **the rules regarding the interval from birth in hours for ADPOAE screening do not apply to AABR.**

4.12 NUMBER OF STAGE 1B OR STAGE 2 AABR SCREENING ATTEMPTS

A Screening Attempt occurs when a probe is placed in the infant's ear and the screening is started on the equipment. A Successful Screen is a final screening event for that stage where a pass or refer result is obtained in any individual ear (see Glossary in Appendix B). **The maximum permissible number of screening attempts pre-discharge or per scheduled community appointment for Stage 1b or Stage 2 AABR in any given ear is <u>two</u>.**

As already stated, starting a screen as a deliberate "trial" of a questionable infant activity level is not appropriate, because of the high likelihood of either a screen non-start or a false-positive refer. The infant must be judged to be quiet enough before starting any screen. This judgment is acquired by experience. If acceptable test conditions

cannot be achieved within **two scheduled community appointments** for any given infant, **further screening will not be scheduled** and the family should be counselled on signs of hearing loss and, if and when concerned, should seek medical contact. As mentioned previously, infants older than two months corrected age are not suitable for newborn hearing screening procedures. Behavioural audiometry by community audiology services may be practicable at about six months of age, with referral to the IHP then if there is clear evidence of PHL. For details for clarifying entry/re-entry of children to the IHP, see Appendix D.

4.13 STAGE 1B AABR EAR SCREENING ORDER

The first ear tested by AABR should be an ear that referred on ADPOAE, because the infant may become active and the second ear be rendered untestable. If an infant refers on ADPOAE in one ear only, both ears still should be screened by AABR, wherever feasible. If one ear referred on ADPOAE but the other ear was not successfully screened, both ears should be screened with AABR. Information about the ADPOAE refer ear might be unavailable or incorrect. Even if ADPOAE results are correct, as should be the case for pre-discharge AABR, it is possible that an ear that passed the ADPOAE will give a refer on AABR, because the AABR checks more parts of the auditory system. The ADPOAE checks only the external ear, middle ear and part of the cochlea (the outer hair cells), whereas the AABR also checks the entire cochlea (including the inner hair cells and their connections to the auditory nerve), as well as the nerve connections in the base of the brain (the brain stem). In the event that the first ear screened by AABR gives a refer, the infant must be referred for Stage 2 AABR in the community on that basis alone, but it is still preferable to do the AABR in both ears.

4.14 REPEAT SCREENING OF ALL STAGE 1B AND STAGE 2 AABR REFERS: DOUBLE-REFER

Whenever practicable, **a Stage 1b or Stage 2 AABR screening must be repeated only once during the same visit in any ear that refers <u>if the infant state and environment are ideal</u>.** Statistical influences within the equipment and unknown eartip/ear canal/electrode conditions may be the cause. The latter can be assessed by eartip removal, inspection, and cleaning after the first refer. Do not re-run the AABR without eartip removal, inspection and re-insertion. A pass on the repeat overrides the initial refer. A refer on the repeat screen (double-refer) is an overall refer in that ear. **A double-refer with AABR in a given ear is preferred to obtain an overall refer for that ear** but is not necessary in order to move to the next stage.

The rationale for immediate repetition and no further repetition is the same as was the case for ADPOAE. Avoidance of multiple re-screening is even more critical for AABR, because the infant has already referred on both ADPOAE and the first AABR refer run, so the likelihood of a PHL being present is now quite high. To sabotage referral for the Stage 2 AABR or audiology referral by multiple re-screening is absolutely not in the best interests of the infant and the family.

4.15 INCOMPLETE AABR PASSES

If an infant passes ADPOAE in one ear, gives a refer in the other, then Stage 1b AABR gives a pass in the referred ear but is not successful (incomplete) in the ear that passed ADPOAE, the infant is to be considered an AABR Stage 1b overall pass and **need not be re-screened by Stage 2 AABR in the Community** (see Figure 8).

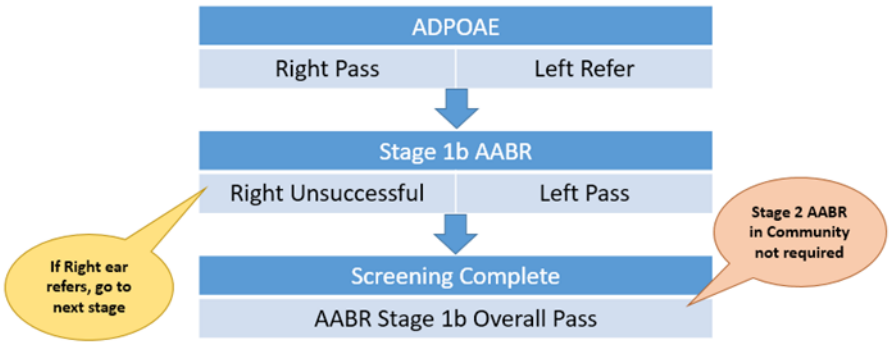

Figure 8: Handling incomplete AABR passes.

4.16 STAGE 2 AABR IN THE COMMUNITY

Infants who refer in any ear or had no result on ADPOAE and are not re-screened by AABR before discharge must be re-screened by AABR in the community (Stage 2 AABR). Community AABR should occur within four weeks of hospital discharge, wherever feasible.

The Stage 2 AABR is designed to further lower the rate of false-positive referral for detailed IHP Audiological Assessment, which is the obligatory next step if there is a further refer result. This Stage 2 AABR strategy will avoid the family having to attend for assessment if the previous screening referrals were caused by temporary hearing loss, usually due to middle ear conductive disorders. Up to about half of infants who refer from pre-discharge AABR may pass on Stage 2 AABR.

If the infant was not screened pre-discharge with AABR, if at the first community visit one ear passes AABR and one refers, at the next community visit the referred ear passes and a screen on the passed ear cannot be completed, then a revisit in the community is not necessary (see Figure 9). For this instance, the infant has an overall pass result. This is because both ears passed screening albeit different screens on different days. It is preferred to have results for both ears on the same day and only if a copy of the previous screening results is available for reference.

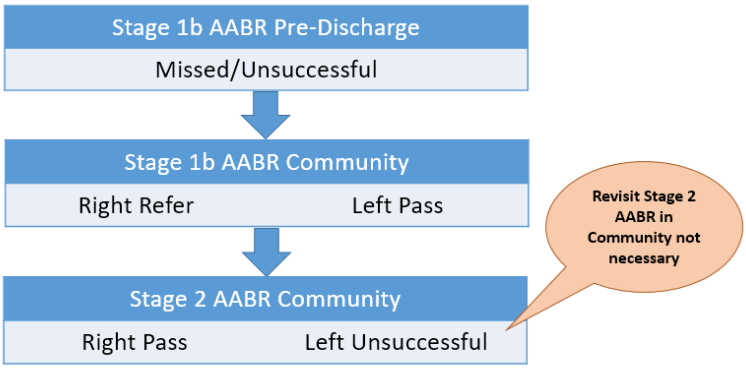

Figure 9: Handling community AABR results.

4.17 COMMUNITY AABR WHEN ALL PRE-DISCHARGE HEARING SCREENING WAS MISSED OR UNSUCCESSFUL

If an infant's first contact for any hearing screening occurs after hospital discharge, in the community, a primary concern is that if several appointments should be needed to complete the multi-stage screening protocol and it turns out that referral to Audiological Assessment is necessary, that assessment might be delayed beyond eight weeks of age. It is at the discretion of the Regional IHP Coordinator to determine whether the community

screening should be directed to AABR or whether the AABR should be preceded by an ADPOAE screen. An obvious pertinent factor is the age of the infant at the family's first attendance for community screening.

4.18 NEVER DO ADPOAE IN ANY EAR WITH REFER ON AABR

Any ear that has yielded a refer on any AABR must not be re-screened by ADPOAE at any time. **The only tests that may follow an AABR refer are a repeat AABR or a detailed Audiological Assessment.** Departure from this may result in failure to identify a child with genuine hearing loss.

4.19 REFERRAL FOR IHP ABR-BASED AUDIOLOGICAL ASSESSMENT (ABRA)

Families of infants who refer on AABR in the community should be advised and encouraged as strongly as possible to attend an 'Audiological Assessment' (see Glossary of Terms in Appendix B), at which the infant's true hearing status will be determined. Whenever feasible, these assessments must occur by six weeks corrected age and no later than eight weeks corrected age and the interval between a community AABR refer and the assessment appointment should be no greater than four weeks.

SECTION 5: SCREENING OF INFANTS AT RISK FOR PHL

5.1 INFANTS AT RISK MUST BE SCREENED ONLY WITH AABR

Except for infants who have a risk indicator for screening bypass (see Glossary of Terms), all newborns at IHP risk for PHL must be screened only by AABR. Infants at risk must not be screened by ADPOAE. The AABR screening should be done in both ears, wherever feasible. There is only one AABR screening stage in infants at risk.
Infants at risk for PHL are **not** screened with ADPOAE for three reasons:

1) The *average* probability of PHL in infants at risk is already ten times higher than in infants not at risk. It is roughly equal to the probability in an infant not at risk who has already referred on ADPOAE screening (about 1 in 100 to 1 in 50).
2) The ADPOAE will not detect auditory neuropathy spectrum disorder (ANSD), a disorder of connections between the cochlea and the auditory nerve. While some causes of ANSD are genetic in nature, other ANSD cases arise from conditions such as severe hypoxia, severe hyperbilirubinemia or congenital CMV infection. About 5-10% of all PHL in infants is associated with ANSD.
3) About half of all infants with PHL will be found in the at risk group, which is about one twentieth the size of the group who have no known risk; this concentration of PHL in a relatively small group justifies immediate use of the more complex and more accurate AABR screen.

5.2 AABR PROCEDURE

The technical aspects of the AABR procedure, causes of screening non-starts or false refers, and requirements for retesting the AABR after a refer to obtain a double-refer, having addressed any issues with the eartip/probe cable/electrode cable, electrode impedance and infant activity levels, are identical to those outlined previously for AABR in infants not at risk who refer on ADPOAE screening. **A double-refer with AABR in a given ear is preferred to obtain an overall refer for that ear** but is not necessary in order to move to the next stage.

A Screening Attempt occurs when a probe is placed in the infant's ear and the screening is started on the equipment. A Successful Screen is a final screening event for that stage where a pass or refer result is obtained in any individual ear (see Glossary in Appendix B). **The maximum permissible number of screening attempts pre-discharge or per scheduled community appointment for AABR in any given at risk infant ear is <u>two</u>.**

5.3 DO NOT SCREEN BY AABR UNDER 34 WEEKS GESTATIONAL AGE

AABR screening must be done when the infant is medically stable and **never under 34 weeks gestational age.** If the infant's estimated gestation period was at least 34 weeks, pre-discharge AABR may be done at any time. However, regardless of the type of delivery (vaginal or Caesarean), the longer the interval from birth to screening is, the lower is the likelihood of ear canal or eartip blockage due to fluid or tissue debris. Notwithstanding this, the AABR screen is far less sensitive to minor blockages than the ADPOAE screen. As a result, **the rules regarding the interval from birth in hours for ADPOAE screening do not apply to AABR.**

The reason for the requirement that the infant be at least 34 weeks gestational age to have an AABR screen is that nerve pathways and connections that generate the ABR waveform may not always have reached a sufficient stage of their development and orientation before that time. The expected ABR waveform that the AABR screening device seeks to detect may not yet be fully established, resulting in an increase in false-positive screens.

5.4 SCREEN JUST BEFORE DISCHARGE TO HOME

If an infant who has attended a neonatal intensive care unit (NICU) and is at risk for PHL is not screened in the NICU hospital but is transferred to a step-down hospital, the AABR should be done as late as possible before discharge home from the step-down hospital, whenever feasible.

5.5 COMMUNITY AABR TARGET WITHIN FOUR WEEKS

To be successful, every AABR screen requires the infant to be asleep or at least resting very quietly. If a successful AABR screen is not obtained in each ear before discharge home, the AABR must be done (or completed) in the community as soon as possible and, whenever feasible, within four weeks of hospital discharge home.

5.6 COMMUNITY AABR EAR SELECTION

If there is no successful AABR done before discharge, the choice of first ear tested in the community is discretional, often governed by the infant's most natural sleeping position. Both ears should be tested whenever practicable. If the first ear passes, the other ear **must** be screened, even if that requires a second community appointment, which must occur as soon as possible. If the first ear tested refers **and the infant state and environment are ideal**, the eartip must be inspected for debris, cleaned if necessary and the AABR repeated immediately. **As explained previously, it is essential that once a double-refer is obtained, there must be no further AABR attempts in that ear.**

A single refer result in the community should be repeated to obtain a double-refer. If the infant state does not permit a double-refer, the infant should be referred to audiology.

5.7 COMMUNITY AABR APPOINTMENT LIMITS

In a medically stable **at risk infant**, if a complete AABR screening is not obtained after a maximum of **two** community AABR screening appointments, the infant must be routed as soon as possible to IHP Audiological Assessment, regardless of the pattern of AABR screening results obtained by that point in time. This is because at risk infants have a higher likelihood of PHL. This prevents delays in the screening process which in turn delays audiology assessment by the target age of six weeks.

5.8 PROMPT AUDIOLOGICAL ASSESSMENT IS ESSENTIAL IN INFANTS AT RISK

All infants at risk who double-refer by AABR in **one or both ears** require a prompt audiological assessment. The probability of PHL in such infants is about 1 in 5 on average but may be as high as certainty (1 in 1), depending on the specific risk indicator(s) present. **The provided scripts and handouts (IHP fact sheets) must be used by the Hearing Screener to inform the family of the necessity of attending the assessment appointment as soon as it is made available. The audiology assessment target is six weeks corrected age and an age of four to six weeks is preferable, wherever feasible medically.**

All of the previously explained rationale, concerns about family understanding and key messages apply more strongly than ever to these families. If pre-established appointments are available to the individual screener, as they are increasingly in certain IHP sites, they should be utilised wherever possible.
Infants who refer in only one ear by AABR are already established as requiring prompt audiological assessment. Trying to obtain a successful AABR in both ears is important, but if the second ear cannot be successfully screened, the at risk infant should be referred to Audiology and not be rebooked in the community.

5.9 EARLIER ASSESSMENT IN VERY PREMATURE INFANTS

If an infant at risk is screened at 34 weeks gestational age or soon thereafter and the overall result is refer, the usual target of six weeks corrected age for audiological assessment might involve a delay of several weeks from the AABR refer to the assessment date. Note that for infants whose perinatal hospital stay extends beyond 44 weeks gestational age, audiology assessment is targeted within four weeks of hospital discharge to home. A four week target minimum allows some time for transient external or middle-ear conditions to resolve, increasing the accuracy and efficiency of the ABR assessment.

SECTION 6: APPENDICES

APPENDIX A: REFERENCES

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APPENDIX B: GLOSSARY OF TERMS

GENERAL

Acquired: Developed or originating after birth.

Auditory Neuropathy Spectrum Disorder (ANSD): A hearing condition whereby the transmission of sound from the inner ear (cochlea) to the auditory brain is impaired. It is a component of a conventional loss of hearing sensitivity which results in impairments in speech perception, especially in noisy situations. Individuals with ANSD vary widely in their performance and outcomes.

Bilateral: Both sides, in this case, both ears.

Chronological age: The age of an individual as measured in number of days, months, and/or years from birth to a given date. It is sometimes referred to as actual age.

Cochlea: A portion of the inner ear that is the spiral organ of hearing.

Congenital: Present at birth or associated with the birth process.

Corrected age: The chronological age of the infant adjusted for a 37-week gestation period. Infants are considered premature if they are born before 37 weeks of pregnancy are completed (World Health Organization, 2015 [fact sheet 363]). Therefore, corrected age is to be calculated using **37 weeks** as full term. For example, an infant born 6 weeks ago at an estimated 34 completed weeks of gestation has a corrected age of: 6 – (37 – 34) = 6 - 3 = 3 weeks. Except where otherwise stated, any age specified in this document is a corrected age.

Critical period: The time within the first two to three years of life when neurological development involved in acquiring, understanding, and expressing language are rapidly developing.

Gestational age: Describes how far along a pregnancy is, measured in weeks, from the first day of the woman's last menstrual cycle to the current date.

Permanent hearing loss: Hearing loss that is expected to last six months or more and will not recover spontaneously.

Pre-discharge: Before discharge from the birth hospital.

Risk indicator: A condition or event associated with increased likelihood of developing a particular condition. In the case of permanent hearing loss risk indicators, they may be present at birth, develop later, or be acquired.

HEARING SCREENING

Complete screen: A Successful hearing screen in **both ears**. See below for definition of a Successful screen.

Double-refer: A repeat ADPOAE/AABR screen on any ear that refers on initial ADPOAE/AABR screening.

False positive: A screening or test result that incorrectly indicates a given condition exists. For newborn hearing screening, this occurs when a refer result is obtained on an infant who has normal hearing.

False negative: A screening or test result that incorrectly indicates a given condition does not exist. For newborn hearing screening, this occurs when a pass result is obtained on an infant who has hearing loss.

Screening attempt: Occurs when a probe is placed in the infant's ear and the screening is started on the equipment.

No result screen: Maximum number of attempts reached without a pass or refer result.

Screening bypass: A screening protocol component whereby newborns identified as having certain risk indicators for permanent hearing loss are routed directly for audiological assessment without first undergoing a hearing screening in the hospital or community.

Screening non-start: Occurs when the infant has been prepped for screening and auto-calibration is not satisfied. That is, stimuli and noise levels within the screening equipment are not within acceptable limits for screening to begin. The start indicator is pressed and the auto-calibration cannot be completed resulting in an error message. The capture of data to indicate screening is occurring does not begin. This counts as a screening attempt.

Successful screen: A final screening event for that stage where a pass or refer result is obtained in any individual ear.

Trial Screening: Starting the hearing screen when the infant's state and/or environment are not acceptable. Inappropriately using the screening itself as a tool to determine when screening is appropriate.

True negative: A screening or test result that correctly indicates a given condition does not exist. For newborn hearing screening, this occurs when a pass result is obtained on an infant who has normal hearing.

True positive: A screening or test result that correctly indicates a given condition exists. For newborn hearing screening, this occurs when a refer result is obtained on an infant who has hearing loss.

TECHNOLOGY

Audiological assessment: A detailed evaluation of hearing performed by an Audiologist or other trained hearing healthcare provider. Using a comprehensive test battery approach, the assessment indicates the degree, type, and configuration of hearing in each ear. Within Ontario's IHP, Audiologists must be authorized through training and monitoring to conduct infant hearing assessments.

Audiological surveillance: A component of an EHDI program that involves the proactive recall for audiological assessment of young children who have been identified within the IHP as having a risk indicator associated with non-congenital (late-onset/delayed-onset) permanent hearing loss (ncPHL). Surveillance is defined in the IHP to be Basic, involving one recall and assessment only, or Intensive, involving more than one recall and assessment.

Auditory Brainstem Response (ABR): A response recorded from the lower level of the brain (brainstem) to rapid auditory stimuli. The responses are elicited by soft, rapid clicking sounds and measured from electrodes strategically-placed on the individual's head.

Automated Auditory Brainstem Response (AABR): A type of screening technology used to screen auditory brainstem function, usually in newborns. The technology is set up to deliver a soft, rapid, clicking stimuli at a fixed level to elicit an ABR response for the determination of hearing. Based on criteria embedded into the screening unit, the system will provide a "pass" or "refer" result.

Automated Distortion Product Otoacoustic Emissions (ADPOAE): A type of screening technology used to screen cochlear hair cell function, usually in newborns. The technology is set up to deliver stimuli at a specific set of frequencies at specific levels to measure OAEs for the determination of healthy cochlear function. Based on criteria embedded into the screening unit, the system will provide a "pass" or "refer" result.

Cochlear Implant: A device used to provide access to sound for individuals who have little or no hearing. An electrode array is surgically placed into the cochlea to provide electric stimulation collected from the sound processor seated on the individual's ear. A magnet is also surgically placed on the skull just under the skin behind the ear to allow for the internal electrode array to connect to the externally worn processor.

Otoacoustic Emissions (OAE): Responses from the hair (sensory) cells within the cochlea. They are elicited by placing a probe in the ear canal and delivering a combination of soft stimuli that generate a response from healthy cochlear hair cells. The responses, or OAEs, are recorded by the same probe that delivered the stimulus.
Tympanometry: Used as part of an audiological assessment test battery to describe middle ear function. Specifically, it measures the movement (or not) of the individual's eardrum.

KEY ROLES WITHIN THE IHP

Audiologist: Practitioners who assess, treat, and educate about the prevention and treatment of hearing loss and balance problems. They are university-trained (minimum Master's level) health care professionals who perform a variety of clinical tests to determine the nature and degree of an individual's hearing and/or balance status and manage the various conditions with (re)habilitation, including various of technology (e.g., hearing aids, cochlear implants). They have the expertise and equipment to work with infants, children, and adults. In Ontario, Audiologists must be registered with the College of Audiologists and Speech-Language Pathologists of Ontario (CASLPO) in order to practice and receive additional training to provide services within the IHP.

Early Hearing Detection & Intervention (EHDI) Program: Healthcare programs that aim to: 1) universally screen all newborns, regardless of the presence of risk indicators for early hearing loss; 2) identify infants with permanent hearing loss using appropriate diagnostic techniques; 3) provide intervention services which include support for technology (e.g., hearing devices) and communication development (i.e., spoken and/or signed languages) based on informed and engaged parental choice; 4) provide family support; and 5) monitor and evaluate the impact of the interventions (Hyde, 2016b; Bagatto & Moodie, 2016).

Hearing Screener: An individual trained by a Regional Trainer to complete hearing screening on newborns in full accordance of this protocol.

Ministry of Children, Community & Social Services (MCCSS): The Ontario ministry that is responsible for funding and oversight of Ontario's EHDI program (IHP).

Universal Newborn Hearing Screening (UNHS): A component of EHDI programs whereby population screening of newborn hearing occurs.

Regional Trainer (RT): Each of the 12 geographic regions of the IHP in Ontario will have one or more RTs to provide consistent training and monitoring of the Hearing Screeners in their region. As well, they will be active Hearing Screeners. They will act as a resource to the Hearing Screener. The MCCSS has established criteria for the qualifications to be an RT (Appendix C).

APPENDIX C: REGIONAL TRAINER CRITERIA

To be considered for the role of Regional Trainer (RT) for the IHP, the following criteria must be met:

1) Have screened at least 250 infants using ADPOAE with 100 consecutive infants having a 12% refer rate;
2) Maintain a 12% or less refer rate for ADPOAE;
3) Screen at least 250 infants per year (with the required refer rate) and no less than 50 infants in a three month period;
4) Thorough understanding of the IHP Hearing Screening Protocol including communicating results to parents using required scripts;
5) Thorough knowledge of troubleshooting techniques; and
6) Working knowledge of AABR screening technology.

RTs will be identified by IHP Regional Coordinators based on the above criteria. RTs are responsible for training personnel new to hearing screening within the IHP, as well as retraining current Hearing Screeners as part of the Continuous Quality Improvement (CQI) initiative.

APPENDIX D: CLARIFYING ENTRY/RE-ENTRY TO THE IHP

The following questions should be asked of the person requesting an IHP hearing assessment for a child. Based on the response, the appropriate action is indicated. This procedure should be followed regardless of who is calling (i.e., parent, physician). Should uncertainty arise about how to proceed, the IHP Coordinator must be contacted.

Item	Question	Response	Action	Notes
1	How old is the child? (corrected age)	≤ 2 months	Community screen	Exit triage
		2.1 months to 6 years	Question #2	
		≥ 6 years	Audiology Assessment in Community	Not eligible for IHP services
2	Did the child have his/her hearing screened at birth?	Yes	Question #3	
		No / Don't know	Question #4	
3	If screened, what was the result?	**Pass**	Question #4	
		Refer with no further testing	IHP Reactivation and Audiology Assessment	Obtain relevant information and provide to IHP Lead Agency[a]
		No Result / Don't know	Question #4	
4	Did the child require any special care following delivery? (e.g., neonatal ICU)	**Yes**	Ask caller to specify, then discuss with IHP Coordinator	Obtain relevant information and provide to IHP Lead Agency[a]
		No	Question #5	
5	Has the child been proven to have meningitis, head injury or extreme jaundice since the screening?	**Yes**	IHP Reactivation and Audiology Assessment	Obtain relevant information and provide to IHP Lead Agency[a]
		No	Question #6	
6	Does the child have frequent ear infections?	**Yes**	Assessment in Community[b]	Not eligible for IHP services
		No	Question #7	
7	Are you concerned that the child has a hearing loss?	**Yes**	Question #8	
		No: 2.1 to 6 months	Monitor developmental milestones	Provide appropriate resources.
		No: ≥ 6 months	Audiology Assessment in Community[b]	Encourage caller to contact IHP if results abnormal.
8	Has the child had his/her hearing assessed in the community?	**Yes**	Obtain community assessment results.	Review with IHP Audiologist and Coordinator prior to considering reactivation.
		No / Incomplete	Question #9	
9	Are you concerned that the child has a speech and/or language delay?	**Yes**	Refer to PSL and Audiology Assessment in Community if necessary	Obtain relevant information and provide to PSL Lead Agency[a]
		No / Don't know	Audiology Assessment in Community[b]	Not eligible for IHP services

Notes:

a	Relevant information for the IHP Lead Agency includes:	Child's Full Name Date of Birth Gestational Age	Parent/caregiver Full Name Address Phone number
b	If child is < 6 months of age, appropriate audiological assessment (e.g., ABR) is not typically feasible in community-based clinics. Behavioural audiometry by community audiology services may be practicable at about six months of age, with referral to the IHP then if there is clear evidence of PHL.		

APPENDIX E: PERFORMING A MADSEN ACCUSCREEN AUDIT

An AccuScreen audit must be performed at least twice each fiscal year for every IHP Hearing Screener. As part of the CQI process, this activity serves to monitor compliance with the current IHP Hearing Screening Protocol. For an audit to occur, there must be no less than 50 screening results within the AccuScreen unit. The AccuScreen holds a maximum of 250 screens. Audits must be performed by an IHP Regional Trainer whenever possible. During an audit, the last two months of screening data shall be reviewed, if available.

SETUP

1. With the AccuScreen in hand, record the serial number and date of the audit.
2. Log on with the screener password "ihp".
3. From the home page select "Test View".
4. Record the first and last log numbers to be audited on that AccuScreen and calculate the total number of screens being audited.

REVIEW

5. Select the first screen in the list and open it. A list of all screening attempts for that log number will appear.
6. Count the number of attempts (both successful and unsuccessful).

RECORD RESULTS

7. Document the date and the number of screens that exceed the protocol attempt limit.
8. Document the date and the number of ADPOAE screens that did not meet the double-refer requirement (double-refer was not obtained despite attempt limit not being reached)

RETURN AND CONTINUE

9. Press the back arrow to return to the test view page.
10. Select the next sequential log number and continue to review and document.

SUMMARIZE

11. Complete a summary for each audited unit/screener and include the following information:
    a. Date of audit
    b. AccuScreen serial number
    c. Screener identification number (PN)
    d. Number of screens reviewed
    e. Number of screens that met protocol
    f. Number of screens that exceeded maximum protocols attempts
    g. Number of attempts over maximum protocol attempts
    h. Number of screens where ADPOAE double-refer requirement was not met

EXAMPLE ACCUSCREEN AUDIT

L AABR	P	9:22:37 05-21-2015
R AABR	P	9:20:09 05-21-2015
R ADPOAE	X	9:17:54 05-21-2015
R ADPOAE	?	9:16:46 05-21-2015
R ADPOAE	?	9:16:32 05-21-2015
R ADPOAE	?	9:16:15 05-21-2015
R ADPOAE	X	9:14:48 05-21-2015
R ADPOAE	?	9:14:21 05-21-2015
L ADPOAE	?	9:12:17 05-21-2015
L ADPOAE	?	9:11:43 05-21-2015

*Note: P = pass, X = Refer, ? = No result

Data are presented in reverse chronological order. In this example, the left ear had two ADPOAE attempts followed by a single AABR in the left ear which resulted in a “pass” result. The sequence of events in the left ear follows protocol. The right ear indicates six ADPOAE attempts which exceeds the maximum number of four attempts for a single ear and therefore does not meet protocol. Results that exceed the number of attempts are recorded. Therefore, the results for this audit would be recorded as: May21, 2 over

APPENDIX F: SCRIPTS USED BY IHP HEARING SCREENERS

HEARING SCREEN CORE CONSENT SCRIPT

The following are scripts to be used by IHP Hearing Screeners to obtain explicit informed consent from families. This is common practice in many EHDI programs in order to standardize the way information is provided throughout the program. The scripts aim to support the delivery of high quality, accurate information to families so they can provide informed consent for the screening.

Introduction:
The hearing screen tells us if your baby needs to have his/her hearing checked. If your baby isn't hearing well, it can make it harder to learn and talk so the earlier you find out the sooner we can help your baby. The hearing screen is not mandatory, but it is strongly recommended.

Information Sharing and Privacy:
First, I would like to let you know that the information I collect about you and your baby, what you decide about the screen, and any results are stored by the Infant Hearing Program. The information will be shared with Newborn Screening Ontario and those who are providing any follow-up. Your information is protected by privacy legislation, so it's always kept secure. Only the program's overall statistics are made public. If you need more information you can call [Lead Agency] at [phone number] or visit our website.
Can I confirm/collect some information?

*PLEASE NOTE: Screening **cannot proceed** if there is not consent for the sharing of information as all screens must be documented.*
Confirm baby's identity, demographics, primary contact information, ability to provide consent.

Explain the Hearing Screen and Discuss Consent
For the **hearing screen**, I'll put a soft tip in your baby's ear and possibly some stickers on your baby's head. I'll use these to record either the ear or the brain's response to some sounds. The test is very reliable, takes only a few minutes and won't hurt your baby. There's no fee for the test and I can give you the results right away.
Is it okay for me to do the hearing screen or do you have questions?

1) **Decline: Review decline decision, confirm documentation and review IHP Fact Sheet**
   Ok, I've recorded your choice to not have the hearing screen. I'll share this with the Infant Hearing Program and Newborn Screening Ontario, so it doesn't look like we missed you. That way no one should try to contact you again to book an appointment for the hearing screen. Remember your baby might have trouble learning and talking if there is a hearing loss so make sure you are watching for any problems and, if you notice anything, have it checked out right away. If you change your mind and decide you would like to have a hearing screen, please contact us here [provide Fact Sheet]. Hearing screens are only available up to two months of age.

2) **Consent – Review risks and discuss Risk Factor Screen consent**
   There are some things that could put your baby at risk for hearing loss either now or in the future. *Review any known risk factors.* Were there any (other) complications during the pregnancy or birth? Has the baby had any health issues since the birth? Have either of the baby's parents or any full siblings to this baby had a hearing loss in one or both ears constantly since before the age of 10 years? (*If yes:* Was there a recommendation for a hearing aid or cochlear implant or did they attend a Provincial School for the Deaf?)

   We can also test for some common risk factors for childhood hearing loss to see if your baby needs any extra monitoring. This test uses a sample that the hospital or midwife usually collects for the newborn (heel prick) screen and results are usually ready in one to two weeks. Most babies have negative results, meaning their chance of having one of these risks is lower. If your baby has a positive result, there is a

higher chance your baby could have childhood hearing loss, and you'll be contacted so that follow-up with an audiologist and a doctor can be set up.

The test will look for an infection called cytomegalovirus (or CMV) which can cause hearing loss. Most babies with this infection won't have any symptoms at birth but we double-check their hearing and then monitor them during their first few years if they have a positive result.

It will also look for some common genetic risk factors. Babies who inherit the same risk factor from both parents will have a positive result. They usually don't have anyone with hearing loss in their family. These babies have a high chance of hearing loss in early childhood.

This test may also pick up babies who inherit a risk factor from only one parent but you won't be contacted with this kind of result since it does not cause hearing loss.

Is it okay for me to arrange for this test or do you have questions?

a. Decline – Review decline decision, complete hearing screen and review IHP Fact Sheet

Ok, I've recorded your choice to not have the risk factor screen.
Complete the hearing screen and discuss the outcome and any next steps with the family. Mark the screen results and consent choice on the appropriate Fact Sheet based on the outcome of the screen.
Remember your baby might have trouble learning and talking if a hearing loss develops so make sure you are watching for any problems and, if you notice anything, have it checked out right away. If you change your mind and decide you would like to have the risk factor screen, please contact us here [provide Fact Sheet].

b. Consent – Complete hearing screen and review IHP Fact Sheet

Complete the hearing screen and discuss the outcome and any next steps with the family. Mark the screen results and consent choice on the appropriate Fact Sheet based on the outcome of the screen and provide to the family.

EXPLAINING SCREENING RESULTS TO FAMILIES

The following are scripts to be used by IHP Hearing Screeners to explain screening results to families. This is common practice in many EHDI programs in order to standardize the way information is provided throughout the program. The scripts aim to support the delivery high quality, accurate information to families as well as facilitate necessary follow-up and regulate family anxiety.

Families who Decline Screening:
If you don't mind, I'd like to leave you with something to read that explains the benefits and the importance of screening your baby's hearing. If you should change your mind and decide you do want the screening, you can let me or your baby's attending physician or nurse know and we can discuss it more at any time.

Family History Risk Assessment:

1) *Do either of the baby's parents have a hearing loss in one or both ears constantly since age 10 years or less?*

   Interpretation notes: If the family member does not have the required hearing loss and/or does not know the answer for the other parent, the infant is to be considered not at risk for this indicator. If there is hearing loss but whether it was present before 10 years is uncertain, the infant is to be considered not at risk for this indicator.

2) *Are there any full siblings to this baby?*
   If yes to #2: Does that child/ Do any of the children have a hearing loss in one or both ears constantly since age 10 years or less?

If yes: Was there a recommendation for use of a hearing aid(s) or cochlear implant(s), or attendance at a Provincial School for the Deaf for the child/children?

Interpretation notes: If there were previous partners, half-brothers or half-sisters do not count and the infant is considered not at risk for this indicator. If either question yields a clear positive response, the infant is at risk on this indicator. If there is any doubt, consider the infant as not at risk.

Hearing Screen Next Steps

Discharge (pending risk factor screen result, if consent)

The hearing screen is done and the result is a 'Pass' for both ears. Please share the results with your baby's doctor.

If there is consent for the risk factor screen: If a risk factor is found, you will be contacted by Newborn Screening Ontario to review the results and follow-up will be arranged.

It is always important to monitor your baby's development, especially their speech and language because your baby's hearing can change at any time. Have your baby's hearing tested by an audiologist if you have any concerns.

*Provide **Your baby's hearing screen** IHP Fact Sheet indicating the outcome of the hearing screen and the consent choice for the risk factor screen.*

Moving from ADPOAE screen to AABR screen

The first part of the hearing screen is done and your baby did not get a 'Pass' result. I don't know why your baby didn't pass, the unit doesn't give me that information but the 'Refer' means I need move on, like a referral to the next test, so I'm going to put the stickers on your baby's head so I can do that test.

Community Screen due to "No Result"

We've attempted to screen your baby's hearing but we weren't able to complete the screen. Some babies need to be screened more than once in order to get an accurate result. Your baby should be seen at one of our community clinics to re-attempt the hearing screen.

Review local process for scheduling the follow-up appointment.

It is important that your baby be sleeping for the follow-up appointment. If you bring your baby in a car seat, we will try to leave the baby in it for the screening.

If there is consent for the risk factor screen: If a risk factor is found, you will be contacted by Newborn Screening Ontario to review the results and follow-up will be arranged.

*Provide and review **Your baby's hearing screen** IHP Fact Sheet indicating the outcome of the hearing screen and the consent choice for the risk factor screen.*

Community Screen due to "Did Not Test"

Although we attempt to provide newborn hearing screening to all babies born in the hospital, we are unable to complete the screening on your baby. It is important that your baby be screened as soon as possible.

Review local process for scheduling the follow-up appointment.

It is important that your baby be sleeping for the follow-up appointment. If you bring your baby in a car seat, we will try to leave the baby in it for the screening.

If there is consent for the risk factor screen: If a risk factor is found, you will be contacted by Newborn Screening Ontario to review the results and follow-up will be arranged.

*Provide and review **Can your baby hear?** IHP Fact Sheet indicating the consent choice for the risk factor screen, if the consent was discussed.*

Community Screen due to "Refer"
Your baby did not get a "Pass" result. The "Refer" result means more testing is needed and your baby will need to have another hearing screen. It is very important that you follow-up with the next test. Your baby should be seen at one of our community clinics for a follow-up hearing screen.

Review local process for scheduling the follow-up appointment.
It is important that your baby be sleeping for the follow-up appointment. If you bring your baby in a car seat, we will try to leave the baby in it for the screening.

If there is consent for the risk factor screen: If a risk factor is found, you will be contacted by Newborn Screening Ontario to review the results and follow-up will be arranged.

*Provide and review **Your baby's hearing screen** IHP Fact Sheet indicating the outcome of the hearing screen and the consent choice for the risk factor screen.*

Audiology High Risk Surveillance
The hearing screen is done and the result is a "Pass" for both ears. Please share the results with your baby's doctor.

Because of your baby's history, there is a chance your baby may develop a hearing loss later on and this could affect his/her speech and language development. The Infant Hearing Program will arrange to check on your child's hearing when he/she is around 15 to 18 months old. If you have any concerns that there has been a change in hearing before your follow-up appointment, please contact us so we can arrange to see you sooner.

If there is consent for the risk factor screen: If a risk factor is found, you will be contacted by Newborn Screening Ontario to review the results and follow-up will be arranged.

*Provide **Your baby's hearing screen** IHP Fact Sheet indicating the outcome of the hearing screen and the consent choice for the risk factor screen.*

Audiology Assessment due to Screen Bypass (Group 3 Risk Indicator)
Because of your baby's history, he/she is at greater risk for hearing loss and this could affect his/her speech and language development, so the program will arrange for detailed testing with an Audiologist who can tell you how well your baby hears. Please let your baby's doctor know about the follow-up that has been recommended by the Infant Hearing Program.

If there is consent for the risk factor screen: If a risk factor is found, you will be contacted by Newborn Screening Ontario to review the results and additional follow-up will be arranged. The screen result will also be forwarded to the audiologist who should have it by your appointment.

*Provide **Your baby needs a hearing assessment** IHP Fact Sheet indicating the outcome of the hearing screen and the consent choice for the risk factor screen.*

Please make sure you attend the appointment. Only the Audiologist can determine for certain whether or not your baby has a hearing loss. If a hearing loss is confirmed, there will be services available to help support you and your child.

The appointment will be booked for approximately two hours. You will be asked to arrive with your baby awake, tired, and ready to eat. Some stickers will be placed on your baby, similar to the ones we used today, then you will be given a chance to feed your baby and get him/her to sleep.

After the appointment, the Audiologist will discuss the results with you. It is best not to come with other children, if possible. You may want to bring another adult with you for support.

Audiology Assessment due to High Risk Refer
The hearing screen is done and your baby did not pass. There can be simple reasons for this, and you'll need to go for more detailed testing with an Audiologist to find out how well your baby hears.

Because of your baby's history, he/she is at greater risk for hearing loss so it is very important for you to attend this follow-up appointment. Please let your baby's doctor know about these results and the follow-up that has been recommended by the Infant Hearing Program.

If there is consent for the risk factor screen: If a risk factor is found, you will be contacted by Newborn Screening Ontario to review the results and additional follow-up will be arranged. The screen result will also be forwarded to the audiologist who should have it by your appointment.

Provide Your baby needs a hearing assessment Fact Sheet indicating the outcome of the hearing screen and the consent choice for the risk factor screen.

Please make sure you attend the appointment. Only the Audiologist can determine for certain whether or not your baby has a hearing loss. If a hearing loss is confirmed, there will be services available to help support you and your child.

The appointment will be booked for approximately two hours. You will be asked to arrive with your baby awake and possibly ready to eat. Some stickers will be placed on your baby, similar to the ones we used today, then you will be given a chance to feed your baby and get him/her to sleep.

After the appointment, the Audiologist will discuss the results with you. It is best not to come with other children, if possible. You may want to bring another adult with you for support.

Audiology Assessment Due to No Risk Refer
The hearing screen is done and your baby did not pass. There can be simple reasons for this, and you'll need to go for further testing with an Audiologist to find out how well your baby hears. Hearing loss could affect his/her speech and language development so it is very important that you attend this follow-up appointment. Please let your baby's doctor know about these results and the follow-up that has been recommended.

If there is consent for the risk factor screen: If a risk factor is found, you will be contacted by Newborn Screening Ontario to review the results and additional follow-up will be arranged. The screen result will also be forwarded to the audiologist who should have it by your appointment.

*Provide **Your baby needs a hearing assessment** IHP Fact Sheet indicating the outcome of the hearing screen and the consent choice for the risk factor screen.*

Please make sure you attend the appointment. Only the Audiologist can determine for certain whether or not your baby has a hearing loss. If a hearing loss is confirmed, there will be services available to help support you and your child.

The appointment will be booked for approximately two hours. You will be asked to arrive with your baby awake and possibly ready to eat. Some stickers will be placed on your baby, similar to the ones we used today, then you will be given a chance to feed your baby and get him/her to sleep.

After the appointment, the Audiologist will discuss the results with you. It is best not to come with other children, if possible. You may want to bring another adult with you for support.

APPENDIX G: IHP RISK INDICATORS FOR PERMANENT HEARING LOSS

Risk Indicator	Screen Bypass	Basic Surveillance	Intensive Surveillance
Group 1			
Apgar ≤ 3 at 5 minutes	No	No	No
Birthweight ≤ 1000 g	No	No	No
Congenital Diaphragmatic Hernia	No	No	No
Family history <10 yrs of age parent or sibling	No	No	No
Hypoxic Ischemic Encephalopathy (Sarnat II or III)	No	No	No
Intraventricular Hemorrhage (Grade III or IV)	No	No	No
Peri-ventricular Leukomalacia	No	No	No
Persistent Pulmonary Hypertension of the Newborn	No	No	No
Ventilatory support: iNO/NO, HFJ/HFO/HFV	No	No	No
Other risk identified by physician	No	No	No
Group 2			
Cleft Palate	No	**Yes**	No
Extracorporeal Membrane Oxygenation (ECMO)	No	**Yes**	No
Hyperbilirubinemia (exchange levels)	No	**Yes**	No
Other proven TORCHES infection	No	**Yes**	No
Syndrome associated with childhood PHL (not CHARGE)	No	**Yes**	No
Group 3			
Atresia/Microtia	**Yes**	**Yes**	No
CHARGE Syndrome	**Yes**	**Yes**	No
Proven Congenital Cytomegalovirus infection	**Yes**	No	**Yes**
Proven Meningitis	**Yes**	No	**Yes**
Genetic Screen Positive	**Yes**	No	**Yes**