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PROTOCOL FOR AUDIOLOGICAL SURVEILLANCE OF CHILDREN AT RISK FOR PERMANENT HEARING LOSS

Ministry of Children, Community and Social Services
Ontario Infant Hearing Program
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SECTION 1: INTRODUCTION

This document addresses procedures for the audiological surveillance of hearing of infants and young children at risk for late onset or progressive permanent hearing loss (PHL). It is closely linked with the Ontario Infant Hearing Program (IHP) *Protocol for Universal Newborn Hearing Screening*, *Auditory Brainstem Response Assessment (ABRA) Protocol*, and the IHP protocol for *Audiometric Assessment for Children Aged 6 to 60 Months* with respect to risk indicators, hearing screening technology applied, screening bypass, and audiological assessment procedures.

Infants are most often identified as having a risk indicator for late-onset or progressive PHL at the hearing screening stage. The designation of risk at the hearing screening stage informs which hearing screening technology (automated distortion product otoacoustic emission (ADPOAE) or automated auditory brainstem response (AABR)) is applied or whether hearing screening is bypassed and audiological assessment is initiated. If the infant passes the hearing screening or the audiological assessment is within normal limits and the risk factor screen is negative, and depending on the risk indicator, the infant may be discharged from the IHP or entered into a particular surveillance sequence (Basic or Intensive). Most Surveillance sessions will consist of visual reinforcement audiometry (VRA) and some early sessions will require an ABR assessment, both of which must be conducted by an IHP Audiologist authorized in those IHP protocols. If during a Surveillance session the child is identified as having PHL, the child will be offered the necessary intervention supports provided by the IHP as guided by current protocols and guidelines (i.e., Provision of Amplification Protocol, Language Development Services Guidelines).

The scope of this document includes the details of the Surveillance procedures as funded by the Ministry of Children, Community and Social Services (MCCSS) for the IHP.

1.1 VERSION HISTORY

This version of the IHP High-Risk Surveillance Protocol (2019.01) supersedes all previous documents relating to high risk surveillance. It is substantially revised from previous documents related to Surveillance within the IHP.

VERSION DATE	DOCUMENT TITLE	PREVIOUS VERSION
2014	High Risk Surveillance Redesign: Questions and Answers	2012 Memo
2012	Memo: Redesign of IHP High Risk Surveillance	N/A
2008	N/A	Surveillance Implemented

Revisions to this version are largely due to changes to IHP Universal Newborn Hearing Screening (UNHS) 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 Surveillance protocol are largely due to the introduction of the risk factor screen.

TOPIC	DESCRIPTION	SECTION
RISK FACTOR SCREEN	With parent/guardian consent, NSO will screen the dried blood spot from the infant's heel prick for congenital cytomegalovirus (cCMV) and several	2.3

	genetic mutations associated with permanent childhood hearing loss.	
RISK INDICATORS	List of risk indicators has been updated.	3.2
GROUP 1 RISK INDICATORS	Infants with these risk indicators who pass their hearing screen will be discharged from the IHP. There will be further contact with the family if the infant screens positive on the risk factor screen.	3.2.1
GROUP 2 RISK INDICATORS	A single-point Basic Surveillance is targeted at 15 to 18 months of age for infants who have these risk indicators.	3.2.2
GROUP 3 RISK INDICATORS	Hearing screening is bypassed and surveillance will occur for infants with these risk indicators with the rate of recurrence dependent on the specific risk indicator.	3.2.3
SURVEILLANCE SEQUENCES	Due to the updated risk indicators, surveillance sequences have been modified and are determined by the infant's risk indicator.	3.6

SECTION 2: SCOPE

2.1 INFANT HEARING PROGRAM (IHP) CORE PRINCIPLES

Audiological Surveillance, using electrophysiological or behavioural measures as required, shall be provided in accordance with the IHP core principles of informed parent/guardian 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*.

2.2 WHAT IS AUDIOLOGICAL SURVEILLANCE?

Audiological Surveillance is the proactive recall for an audiological assessment of young children who have been identified within the IHP as having a risk indicator associated with late-onset or progressive PHL. Surveillance is defined here to be Basic, which involves a single recall, or Intensive, involving a varying rate of recalls depending on the risk indicator. Both sequences involve a complete audiological assessment that is developmentally appropriate for the infant (e.g., auditory brainstem response assessment (ABRA), visual reinforcement audiometry (VRA), conditioned play audiometry (CPA), standard audiometry).

In its broadest sense, Audiological Surveillance is a systematic process for the early detection of permanent hearing loss (PHL) that is not detected by UNHS. There are a variety of reasons why PHL may not be detected by UNHS. For instance, the child may not access UNHS through the IHP or the hearing loss was not present or detectable at the time of the hearing screen.

Failure to detect hearing loss that is not yet present is not a false-negative screen. This situation is the major focus of surveillance, as are hearing losses that are too small at the time of hearing screen and hearing losses that are frequency-specific. Because surveillance is conducted using diagnostic hearing test technology and procedures

rather than screening technology and procedures, hearing losses that are present but undetectable by hearing screening tests are detectable in those children who are selected for surveillance.

2.3 RISK FACTOR SCREEN

In 2013, the IHP undertook a collaborative project with Newborn Screening Ontario (NSO) and determined that it was feasible for NSO to detect causes of PHL through blood spot screening including selected genetic mutations and congenital cytomegalovirus (cCMV). The dried blood spot is already used to detect many other treatable diseases and disorders in newborns. The addition of some genetic mutations known to cause PHL in infants as well as cCMV, which is the leading environmental cause of PHL, will help ensure that follow-up occurs for infants who pass UNHS and have a likelihood of late-onset or progressive PHL.

The goal of implementing the risk factor screen in newborns is to improve the program's risk assessment process. It will allow for earlier and more accurate identification of infants with specific risk indicators for PHL and their subsequent assessment or surveillance monitoring.

Phase 1 was implemented in April 2018 and involved targeted cCMV screening for infants that had been referred for audiology assessment following a refer on the hearing screening and where parent/guardian explicit informed consent for the risk factor screen had been obtained.

Phase 2 was implemented in July 2019 and involves offering the risk factor screen universally. With parent/guardian explicit informed consent, NSO will screen the dried blood spot for cCMV infection and several common mutations on three genes that are known to cause PHL in infants and young children.

2.4 WHO CAN CONDUCT AUDIOLOGICAL SURVEILLANCE?

Only Audiologists registered with the College of Audiologists and Speech Language Pathologists of Ontario (CASLPO) who are trained and authorized by the IHP to conduct this protocol AND at least one of the IHP Assessment protocols (*ABRA, Audiometric Assessment for Children Aged 6 to 60 months*) may provide Audiological Surveillance services with IHP funding. The assessment protocol for which an IHP Audiologist is authorized indicates which surveillance strategy (e.g., ABR or behavioural) the Audiologist is authorized to conduct as part of this protocol. The IHP Audiologist must personally conduct the testing and interpret the results. Students may also participate with full supervision from the IHP Audiologist.

If an IHP Audiologist has been inactive in this or other IHP Assessment protocols for six months or more, the re-training review procedures in the *IHP Guidance Document* will apply.

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

2.5 PROTOCOL ADHERENCE IS A REQUIREMENT

All IHP Surveillance must be conducted in adherence to this protocol as well as the IHP Assessment protocols; such adherence is an expectation for continued authorization to provide IHP services. Sufficient documentation of protocol adherence must be kept on file by the IHP Lead Agency/Audiologist to support clinical decision support and/or standard practice reviews conducted by DTCs when necessary.

2.6 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 for IHP standard practice review or case audits (see *IHP Assessment protocols*).

2.7 PROCEDURAL CONCERNS

Prior approval by MCCSS is required in order to substantively change any element of this protocol. Program-wide changes can occur only through MCCSS directive or by a systematic process that may include survey of Audiologists' experiences or concerns, evidence review, and recommendation by a 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 IHP Lead Agency management and shall be authorized ONLY by modification of the relevant IHP protocol document (such as this document), which shall govern IHP Surveillance services throughout Ontario.

2.8 NON-IHP SERVICES

Hearing assessment services conducted by any person who is not an Audiologist 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, and because of the prevalence of progressive hearing losses and/or conductive overlays in the pediatric population, IHP Audiologists shall re-test a child prior to making inferences about hearing status and/or candidacy for ongoing management.

2.9 POPULATIONS TARGETED

Candidates for surveillance include all Ontario-resident infants who have been identified, through the IHP UNHS protocol, Risk Factor Screen, or referral into the IHP, as having at least one of the IHP risk indicators (see Appendix B). Candidates must have:

1) Passed UNHS;
2) Bypassed UNHS and been found **not** to have PHL on audiology assessment ; or
3) Referred on UNHS and been found **not** to have PHL on audiology assessment.

Infants moving into Ontario or older siblings of infants with PHL identifed through the IHP must be assessed in the community (i.e., not through the IHP). Referral into the IHP is warranted if the child is eligible (e.g., has PHL and is younger than 6 years of age). Surveillance does not replace missed UNHS or retrospective audiology assessments for siblings of newly-identified infants with PHL through the IHP.

2.10 TARGET DISORDERS

The IHP target disorder set includes PHL of $\geq$ 30 dB HL 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 *ABRA* 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.

2.11 CONDUCTIVE HEARING LOSS

The IHP is complementary to Ontario Health Insurance Plan (OHIP)-based, physician-driven audiology services and does not replace them. Purely conductive hearing loss is not an IHP target unless obviously or presumptively structural, such as in congenital atresia or if a syndrome associated with structural, conductive anomalies is identified or suspected. For minor conductive losses, discharge from the IHP with caregiver counseling and discretional referral to a physician is the norm. For more information, see the IHP Assessment protocols.

2.12 OBJECTIVE OF AUDIOLOGICAL SURVEILLANCE

The main objectives of audiological surveillance are to:

1) Determine whether further audiological assessment is required; and
2) Potentially detect the presence of late onset or progressive PHL.

The overall surveillance goes beyond audiometry itself and includes family counseling and information about follow-up.

It is widely acknowledged that, overall, audiological surveillance is a resource-intensive method of identifying infants and young children at risk for PHL. The main reasons for this are that the majority of PHL associated with current risk indicators identifiable perinatally is congenital, the positive predictive values (PPV) of most current risk indicators for non-congenital PHL are very low, and the numbers of infants found to be at risk on some risk indicators are very large. Furthermore, the PHL case yield from any given single-point surveillance depends upon the distribution over age of the expression of the PHL (Boerst & Thorne, 2015).

Multi-point surveillance has also been applied and is resource intensive. The Joint Committee on Infant Hearing (JCIH, 2000) revised their earlier surveillance recommendations to align with the impracticability of multiple surveillance events. As such, multi-point surveillance will be applied when infants are identified as having a risk indicator known to have a relatively high PPV and short PHL expression time-frames.

A measure of this resource expenditure per case identified is the number needed to test (NNT) to find one PHL case in the group at risk on a given indicator and who did not refer on screening (Wood et al, 2013). For example, how many babies who passed newborn hearing screening and had a wide Family History risk indicator were seen for surveillance to identify one infant with PHL. In the 2.2 million babies analyzed retrospectively in the UK, the NNT for this risk indicator was 909 (Wood et al, 2013). The range median was about 500 for the other risk indicators. The corresponding median for babies who referred on screening was 4.9. This means that, overall, surveillance is at best about 1% as effective at identifying PHL as assessment is in babies who refer on screening in the UK sample.

Another issue to consider is the *harm* accrued from proactively visiting unproductive care upon families who did not seek it. Such harms include anxiety and direct costs to large numbers of families for appointment attendance that yield no benefit to them. It is about the balance of relatively small harms to the many against the relatively large benefit for the few cases identified early and for whom intervention was effective. In surveillance, an option is to limit it to those risk indicators with the lowest NNTs.

This protocol outlines the list of risk indicators used within the IHP and the surveillance sequence, if any, that must be applied. Depending on the risk indicator, the infant may not be involved in surveillance or may participate in a Basic or Intensive Surveillance sequence.

2.13 TARGET AGE RANGE

In practice, the overall target age range of children for potential surveillance within the IHP is from about three months corrected age to about five years of age. It should be noted that corrected age is to be calculated using 37 weeks as full term (World Health Organization, 2018). Corrected age shall be calculated until the child turns two years chronological age. Risk indicators associated with PHL expression only beyond five years are not included in the IHP risk indicator list and the identification of PHL expressed beyond that limit is not within the purview of the IHP. Specific age ranges for successful completion of ABR and behavioural assessment can be found in the respective IHP Assessment protocols.

It is noteworthy when considering the target age for surveillance that the lower the age, the smaller the proportion of infants with PHL already expressed. The others will likely be assessed as having normal hearing. Alternatively, the higher the age, the larger the proportion expressed, but the older the infants are at PHL identification.

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, IHP 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, Hearing Screening, and Surveillance.

2.15 IHP PROTOCOLS AND CASLPO GUIDELINES

Since surveillance includes ABR or behavioural assessment to be conducted by an Audiologist, the procedures shall be practiced in full compliance with the requirements of both CASLPO and this protocol. IHP protocols may be more specific than CASLPO guidelines. Effort is made to ensure that IHP protocols do not conflict with CASLPO guidelines. Such conflicts may arise inadvertently and if any IHP Audiologist perceives such a conflict, the Audiologist shall notify the DTC promptly and the IHP will act to resolve the issue.

2.16 INFECTION CONTROL STANDARDS

Infection control practices are typically governed by site-specific, institutional or agency protocols and are outside the purview of this document. Generally accepted standards must be applied (CASLPO, 2010).

2.17 APPROVED TEST ENVIRONMENTS

Surveillance test areas must satisfy current ANSI standards for ABR or manual puretone audiometry (see IHP Assessment protocols for details). If deviations from this cannot be avoided, they must be clearly documented. If a refer on Surveillance is suspected in a test area that does not satisfy ANSI standards, it must be confirmed at a later date in a test area that meets the standards.

2.18 CLINICAL RECORDS AND DATABASE REPORTING

All audiometric records shall be maintained in a manner satisfying both CASLPO and the IHP. The records shall be maintained in hardcopy or, if electronic medical records are used, in secure data files. For Surveillance conducted using ABR, clinical records must include the test session listing of records that details the exact order of acquisition of tracings. For Surveillance conducted using behavioural audiometry, clinical records must include details of the procedure used to condition the child for VRA, the administration of control trials, the infant's rate of correct responses on control trials, and a worksheet showing the administration of VRA including control trial administration. See IHP Assessment protocols for details. Records detailing interpretation of the Surveillance session and reporting of any necessary information to Healthy Child Development – Integrated Services for Children Information System (HCD-ISCIS) should also be available.

2.19 PERSONAL HEALTH INFORMATION

Management of all personal health information arising from the Surveillance process shall comply with local site and legislative requirements and those of CASLPO. 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 caregiver.

SECTION 3: PROTOCOL FOR AUDIOLOGICAL SURVEILLANCE

3.1 RISK ASSESSMENT WITHIN THE IHP

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 Hearing Screener through document review, consulting nursing or medical staff, and talking to the family.

With respect to initial hearing 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 Hearing Screener, such as for an obvious malformation of the external ear. A confident and clear report of cleft palate by the family 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, hearing screening is done using AABR.

The purposes of risk assessment are to:

1) Determine whether hearing screening should be bypassed; or
2) Decide which hearing screening technology 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 below and in Appendix B for quick reference. 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 Hearing Screener is responsible to ensure that the parent/guardian is questioned exactly as specified in the *IHP Hearing Screening Protocol*. The role of a nurse is to assist with the indicators related to specific medical conditions or procedures.

For infants who did not undergo a risk assessment by the Hearing Screener either in the hospital or the community, it is the IHP Audiologist's responsibility to conduct the risk assessment. It may be the case that the infant is directed to Audiology following a positive result on the dried blood spot from the risk factor screen.

3.2 CURRENT IHP RISK INDICATORS

The list of IHP risk indicators has been modified based on preliminary evidence review, expert consultation, and availability of IHP resources. In addition, the implementation of the risk factor screen has, in part, informed the screening, bypass, and surveillance procedures. The list of IHP risk indicators and surveillance steps are found in Appendix B. Table 1 represents the current risk indicators and surveillance sequences for the IHP.

Table 1: Current IHP Risk Indicators and Surveillance Sequence

***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 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	Cleft palate Extracorporeal membrane oxygenation (ECMO, ECLS) Hyperbilirubinemia meeting exchange criterion, whether exchanged or not Proven TORCHES infection (toxoplasmosis, rubella, herpes simplex virus, syphilis) *except CMV* Syndrome associated with PHL *except CHARGE*	Atresia/microtia (screen of unaffected ear permitted) CHARGE Syndrome Proven Congenital Cytomegalovirus (cCMV) including cCMV screen positive on the dried blood spot Proven Meningitis Genetic Screen Positive

3.2.1 GROUP 1 RISK INDICATORS

Infants who are identified as having a Group 1 risk indicator will undergo an AABR screening procedure. If the hearing screening outcome is an overall 'refer' result, the infant is routed for an audiology assessment. If the hearing screening outcome is an overall 'pass' result and the risk factor screen is negative, 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 hearing screening technology the Hearing Screener applies. The infant will be re-directed for audiology assessment if the risk factor screen is positive following a 'pass' result on the hearing screening.

3.2.2 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, the infant is routed for an audiology assessment. If the screening outcome is an overall 'pass' result, the infant enters into the Basic Audiological Surveillance sequence.

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.

3.2.3 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. 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 including appropriate audiologic assessment follow-up which is not to be confused with surveillance. Children with PHL do not by definition require surveillance. If the audiology assessment reveals normal hearing in both ears, the infant enters into surveillance, which varies in frequency of appointments depending on the risk indicator.

3.2.4 GENETIC MUTATION 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.

3.3 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 for example. 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 7-10 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.

3.4 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:

3.4.1 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 selected common genetic mutations affecting three genes associated with significant and early PHL. These account for some of the most common recessive mutations but is not an exhaustive screen of either all mutations within the three genes, or all genes associated with pediatric PHL. In addition, the results of the risk factor screen may not be known at the time of the hearing screen. Therefore, it is imperative for the Hearing Screener to accurately identify a family history of hearing loss.

In the absence of the result of the risk factor screen, the family history risk indicator should be identified by the Hearing Screener using a strictly-followed script for questioning of family members by IHP Hearing Screeners (see IHP Screening Protocol). 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 at risk.

An infant with a family history of PHL will be screened using AABR. If the infant passes the hearing 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, Intensive Surveillance will be scheduled.

3.4.2 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). Atresia and microtia can have varying grades of severity.

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. Infants with CHARGE have a very high incidence of PHL (~90%; e.g., Arndt et al, 2010; Blake et al, 1998; Holcomb et al, 2013). As well, these children usually have a very complex audiological profile, with the possibility of sensorineural, both temporary and/or permanent conductive, and ANSD components all present in the same child. 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. Many infants with cCMV will be asymptomatic (85-90%) with a 10% chance of developing PHL in childhood (Fowler et al, 2017). Ten to 15% of infants will have signs or symptoms of cCMV identified at birth such as rash, jaundice, or growth problems. Infants with symptomatic cCMV have a 30% chance of developing PHL (Fowler et al, 2017), which supports the need for different surveillance sequence timing for the two groups (Lanzieri et al, 2018).

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. Between 25 and 50 % of survivors will manifest some type of morbidity in the first five years post-illness. Hearing loss is reported in about 25 % of cases (Bedford et al, 2001). Wellman et al (2003) recommended ABRA in all cases at four to six weeks post-recovery.

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.

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).

3.5 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 seen in the community for appropriate services.

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.

3.6 SURVEILLANCE SEQUENCES AND RISK INDICATORS

Infants with no assigned IHP Risk Indicator for PHL will be screened using ADPOAE technology. About 95% of all infants are not at risk for PHL. Initial likelihood of PHL is small and the number of infants large. This favours use of an initial hearing screen using ADPOAE technology to quickly filter out all babies with the greatest probability of having normal hearing at that time. The AABR screen takes more time, skill and expense, but is more resilient to minor middle and outer ear dysfunction and will in turn eliminate many babies who are false-positive on ADPOAE.

Infants who have a risk indicator from Group 1 will be screened using AABR technology. If the infant passes the hearing screen or is determined to have normal hearing at the audiology assessment, s/he will not enter into surveillance. S/he will be discharged from the IHP if they are also determined to have a negative result on the risk factor screen. Infants who are assigned an IHP risk indicator from Group 2 or 3 will enter into either Basic or Intensive Surveillance, which are described in detail in the following sections (see Table 2). Infants with a Group 3 risk indicator will bypass screening and be routed directly for audiology assessment. If the infant is determined to have normal hearing at the initial assessment, s/he is entered into either a Basic or Intensive Surveillance sequence depending on the risk indicator. All surveillance appointments include a developmentally appropriate audiology assessment. A quick guide for the clinical application of the surveillance sequences for each IHP risk indicator can be found in Appendix C.

Table 2: Summary of risk indicator groups and surveillance sequences.

Risk Indicator	Action	Surveillance Classification	Surveillance Sequence
Group 1	AABR Screen Discharge if Pass	None	N/A
Group 2	AABR Screen Surveillance if Pass	Basic Sequence	15 to 18 months corrected age
Group 3	Bypass Hearing Screen Refer for Audiology Assessment If hearing within normal limits, enter Surveillance.	*Basic Sequence:* Atresia/Microtia CHARGE Syndrome	15 to 18 months corrected age
		Intensive Sequence: Proven Meningitis Proven cCMV: Asymptomatic* Proven cCMV: Symptomatic* Genetic Screen Positive	*Proven Meningitis* 3 assessments at 3 month intervals *Proven cCMV: Asymptomatic* 10 to 12 months, 15 to 18 months, 3 years, 5 years *Proven cCMV: Symptomatic* 3 assessments at 3 month intervals, 15 to 18 months, 3 years, 5 years *Genetic Screen Positive*: 3 assessments at 3 month intervals, 15 to 18 months, 3 years

*Note: Audiologists should categorize infants with proven cCMV as asymptomatic unless otherwise notified.

3.6.1 BASIC SURVEILLANCE

Basic Surveillance is likely to be optimal if done between one and two years of age. This corresponds to the JCIH (2007) recommendations, among others (e.g., Sutton et al, 2012; Vos et al, 2015). It reflects the limited interest shown by families in attending assessments after too-lengthy periods of apparently normal hearing in their infants. Wood et al (2013) reported that only 55% of all families offered such appointments attended them. The resources expended in tracking and contacting all such families to no avail are not to be underestimated. It also takes account of the window of practicality for VRA in the target population.

Infants identified as having a risk indicator from Group 2 and atresia/microtia and CHARGE syndrome from Group 3 and who pass their hearing screen or audiological assessment will enter into Basic Surveillance. This single-point activity is targeted to occur between 15 to 18 months corrected age. Earlier than about 15 months limits the amount of PHL likely to be expressed. Later than about 18 months of age limits the likelihood of successful and efficient VRA, while play audiometry is still unlikely to be widely effective.

The Basic Surveillance appointment will consist of a complete audiological assessment as described in the *IHP Audiometric Assessment for Children Aged 6 to 60 months* protocol. Briefly, in each ear, VRA shall be conducted via

air conduction at 500, 2000, and 4000 Hz. Case history, otoscopy, immittance, and DPOAEs shall also be obtained and the results interpreted accordingly to satisfy the cross-check principle (Norrix, 2015).

Priority is given to obtaining VRA results with recommended cross-check from DPOAEs. If the result on Basic Surveillance is a pass in both ears, then the child shall be discharged from the IHP. No further surveillance appointments are required. If the child is identified as having PHL during the surveillance appointment, next step IHP supports and services shall be offered and no further surveillance is required.

3.6.2 INTENSIVE SURVEILLANCE

Infants identified as having a risk indicator from Group 3 (*except* atresia/microtia and CHARGE syndrome) and who pass their audiological assessment will enter into Intensive Surveillance. This surveillance sequence is multi-point and varies depending on the risk indicator. Regardless of the number of surveillance appointments, all infants with the relevant risk indicator from Group 3 will undergo a complete audiology assessment that is developmentally appropriate. That is, for infants under six months corrected age, an ABR assessment must be conducted and conditioned behavioural assessment must be conducted in infants six months and older, as developmentally appropriate. For either strategy, the current IHP Assessment Protocols apply (e.g., *ABR Assessment* and *Audiometric Assessment for Children aged 6 to 60 months*).

The risk-dependent Intensive Surveillance sequences are as follows:

1) *Proven Meningitis:* 3 assessments at 3 month intervals from the time of the first normal assessment
2) *Proven cCMV with no symptoms (asymptomatic)**: 10 to 12 months, 15 to 18 months, 3 years, and 5 years
3) *Proven cCMV with symptom(s) (symptomatic)**: 3 assessments at 3 month intervals from the time of the first normal assessment, then 15 to 18 months, again at 3 years and 5 years
4) *Genetic screen positive*: 3 assessments at 3 month intervals from the time of the first normal assessment, then 15 to 18 months and again at 3 years

* Audiologists should categorize infants with proven cCMV through the risk factor screen as asymptomatic unless otherwise notified.

If normal hearing in both ears is confirmed during an Intensive Surveillance appointment within the sequence or a clinical decision is made that PHL is unlikely, the child shall be booked for the next surveillance appointment. If normal hearing in both ears is confirmed at the final appointment of the sequence, the child shall be discharged from the IHP. No further surveillance appointments are required. If the infant is identified as having PHL during any Intensive Surveillance appointment, next step IHP supports and services shall be offered and no further surveillance is required. For infants with proven meningitis who are identified as having PHL, the IHP Audiologist's prompt referral to a cochlear implant program is recommended due to the possibility of ossification of the cochlea. Infants with incomplete or inconclusive results during the Intensive Surveillance appointment shall be rebooked for the next appointment within the sequence.

3.7 NUMBER OF SURVEILLANCE ATTEMPTS

Every effort should be made to complete the surveillance apointment in as few visits as possible. This should include parent counselling on how to prepare the child for the next visit and activities that can be done at home to help train the child to be better prepared to condition for the testing at the next visit (e.g., working on sound awareness). Review the relevant sections of the *IHP Assessment* protocols for helpful strategies for ABR and behavioural audiometry.

Infants who have incomplete or inconclusive results during a surveillance appointment should be rebooked no more than two times for a total of three attempts. These appointments should be closely scheduled and occur within a three month time period if part of the Basic Surveillance sequence. For Intensive Surveillance, the child should be moved to the next scheduled surveillance visit.

If a complete assessment has not been achieved after the third attempt, the Audiologist must be prepared to make a clinical decision based on any information that was obtained. This may include ear-specific or soundfield responses, tympanometry, DPOAEs, acoustic reflexes, progress in listening skills, and parent reports on responsiveness to sound, which may include the LittlEARS auditory questionnaire (see *IHP Provision of Amplification Protocol*). If there is no information to indicate that PHL has developed, the child should either be moved to the next scheduled surveillance visit if the risk indicator requires Intensive Surveillance, or be discharged from the IHP with appropriate counselling to the family regarding monitoring the child's responsiveness to sound, and clear documentation of the rational for the decision. If the family has future concerns about their child's hearing, they should arrange for a hearing assessment in the community (i.e., not through IHP).

If there is concern that a PHL may be present based on the information that was obtained, consideration of a natural sleep ABR or sedated ABR referral may be warranted. If the audiologist is unable to come to a clinical decision, consultation with the DTC for clinical decision support must occur.

APPENDICES

APPENDIX A: REFERENCES

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APPENDIX B: 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 Meningitis	**Yes**	No	**Yes**
Proven Congenital Cytomegalovirus: Asymptomatic	**Yes**	No	**Yes**
Proven Congenital Cytomegalovirus: Symptomatic	**Yes**	No	**Yes**
Genetic Screen Positive	**Yes**	No	**Yes**

APPENDIX C: QUICK GUIDE FOR CLINICAL APPLICATION OF SURVEILLANCE SEQUENCES

Risk Indicator	3 ax at 3 month intervals	10 – 12 months	15 – 18 months	3 years	5 years
Group 1					
Apgar ≤ 3 at 5 minutes	---	---	---	---	---
Birthweight ≤ 1000 g	---	---	---	---	---
Congenital Diaphragmatic Hernia	---	---	---	---	---
Family history <10 yrs of age parent or sibling	---	---	---	---	---
Hypoxic Ischemic Encephalopathy (Sarnat II or III)	---	---	---	---	---
Intraventricular Hemorrhage (Grade III or IV)	---	---	---	---	---
Peri-ventricular Leukomalacia	---	---	---	---	---
Persistent Pulmonary Hypertension of the Newborn	---	---	---	---	---
Ventilatory support: iNO/NO, HFJ/HFO/HFV	---	---	---	---	---
Other risk identified by physician	---	---	---	---	---
Group 2					
Cleft Palate	---	---	✓	---	---
ECMO	---	---	✓	---	---
Hyperbilirubinemia (exchange levels)	---	---	✓	---	---
Proven TORCHES infection	---	---	✓	---	---
Syndrome associated with childhood PHL (not CHARGE)	---	---	✓	---	---
Group 3					
Atresia/Microtia	---	---	✓	---	---
CHARGE Syndrome	---	---	✓	---	---
Proven Meningitis	✓	---	---	---	---
Proven cCMV: Asymptomatic	---	✓	✓	✓	✓
Proven cCMV: Symptomatic	✓	---	✓	✓	✓
Genetic Screen Positive	✓	---	✓	✓	---