MODIFIED PHYSIOLOGICAL HEARING SCREEN

Interim Protocol For Ontario Infants During the COVID-19 Pandemic
2020.02

MINISTRY OF CHILDREN, COMMUNITY AND SOCIAL SERVICES
Ontario Infant Hearing Program

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Scope

Introduction

As a global pandemic forced service suspensions, many newborn infants in Ontario were unable to access physiological hearing screening through the Infant Hearing Program (IHP). The hearing loss risk factor bloodspot screen (RFS) remained active during the pandemic closure period. Due to limitations and challenges with hearing screening beyond the newborn period, an interim modification is being offered so that infants who missed the ***newborn*** physiological hearing screen have access to ***early*** hearing screening in a valid and efficient way. This will support the IHP benchmarks of early hearing loss confirmation and management of infants and young children who have permanent hearing loss, without the need for large-scale efforts to provide a full assessment to young infants.

This protocol describes interim procedures for a modified physiological hearing screen for Ontario infants who are over 8 weeks corrected age and who were not offered a newborn hearing screen through the Infant Hearing Program (IHP) due to service interruptions as a result of the COVID-19 pandemic. The scope of this document includes details of the modified hearing screen as funded by the Ministry of Children, Community and Social Services (MCCSS) for the Ontario IHP. It will remain in effect as an interim service during the global pandemic and will be recalled at the discretion of the MCCSS.

Target Population

Infants who are eligible for the modified physiological hearing screen through the IHP are Ontario-residents who:

1) were not offered an IHP physiological hearing screen prior to discharge from the hospital or in the community due to COVID-19 closures;
2) are older than 8 weeks corrected age; and
3) have a negative result on the risk factor blood spot screen.

The modified physiological hearing screen will not be offered to infants who:

1) are younger than 8 weeks corrected age;
2) accessed newborn hearing screening prior to or during COVID-19 closures;
3) obtained a positive result on their bloodspot risk factor screen;
4) are identified as having a Group 3 (bypass screening) risk indicator; or
5) acquired a risk indicator later in infancy.

Routing of infants who are not eligible for the modified screen will vary based on the situation. Further direction for triaging these infants is provided in Appendix A. In some cases the infant will qualify for newborn hearing screening as described in the IHP *Protocol for Universal Newborn Hearing Screening* (2019.01) and in others the infant will bypass hearing screening (modified or universal) and go directly for an IHP audiology assessment.

Target Disorder

The IHP target disorder set includes permanent hearing loss (PHL) of $\geq$ 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 auditory brainstem response (ABR) techniques. 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.

Objectives of the Modified Hearing Screen

The main objectives of the IHP modified physiological hearing screen are to:

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

The overall modified hearing screening protocol goes beyond the audiological procedures itself and includes providing the family with the results (pass, refer, no result) and information about follow-up.

Summary of Procedures

The modified physiological hearing screen includes the following procedures and will be no more than 30 minutes in duration:

1) Risk assessment
2) Otoscopy
3) Distortion product otoacoustic emissions (DPOAE)
4) Tympanometry
5) Acoustic reflexes (for high risk infants only)

These procedures are described in detail in the Protocol section of this document.

Personnel

The modified hearing screen includes a test battery that must be interpreted by an Audiologist registered and in good standing with the College of Audiologists and Speech-Language Pathologists of Ontario (CASLPO). A non-IHP Audiologist within Ontario may conduct the modified hearing screen within the IHP provided there is an agreement to provide the services within the scope of this protocol through the IHP Lead Agency.

Supportive personnel may participate in some procedures within the modified screen under the supervision of the Audiologist responsible for the conduct of the testing and interpretation (CALSPO, 2013). Supportive personnel may include IHP Hearing Screeners, Communicative Disorders Assistants (CDA), or Speech-Language Pathologists (SLP). CDAs may conduct risk assessment, tympanometry, and/or DPOAEs only. IHP Hearing Screeners may conduct the risk assessment and DPOAEs only. IHP administrative staff may conduct the risk assessment portion of the modified screen. This may help triage infants for other procedures (e.g., bypass and go directly to IHP audiology assessment) and reduce the need to bring families into clinics for the modified screen unnecessarily. A CASLPO- registered Audiologist must interpret the results of the modified screen battery.

All personnel participating in the modified hearing screen must be familiar with this protocol. They are not required to undergo training but review of the materials developed by the MCCSS is strongly encouraged.

Instrumentation and Test Environment

DPOAEs and middle ear analysis (i.e., tympanograms, acoustic reflexes) can be reliably recorded in cooperative children in a quiet environment. A sound treated room is not required. The child must be quiet but does not need to be sleeping during the tests. The equipment used for the DPOAE portion of the modified hearing screen is at the discretion of the IHP test facility, and may include IHP screening equipment (i.e., Accuscreen) or diagnostic equipment set to meet the requirements described in the Procedures section of this protocol.

Within the IHP, the equipment used for universal newborn hearing screening is the Madsen AccuScreen hand-held device. It includes automated DPOAE and ABR protocols. For the purposes of the modified hearing screen, the Accuscreen device may be used for the DPOAE component of the test battery. This includes Firmware 2012: 1.11.04160SEU and the DP5 protocol with the criteria described in the Procedures section of this protocol.

If the Accuscreen is used, interpretation of the DPOAE screen is automated within the unit for each ear tested.

Middle ear analysis (i.e., tympanograms and acoustic reflexes) may be completed using any make or model of equipment, provided that it meets calibration requirements, and offers a 1 kHz probe tone for use with infants younger than 6 months corrected age. As per CASLPO (2008) requirements, middle ear analysis must be performed using a type 1 tympanometer. A pressure sweep range that includes -200 daPa is required to facilitate measurement of negative middle ear pressures.

Otoscopy must be conducted using a device with sufficient illuminated magnification for examination of the ear canal and tympanic membrane. Pediatric-sized speculum must be available for use with the chosen device to ensure safe and accurate otoscopic examination of small ears.

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 and follow public health protocols for COVID-19.

Clinical Records and Database Reporting

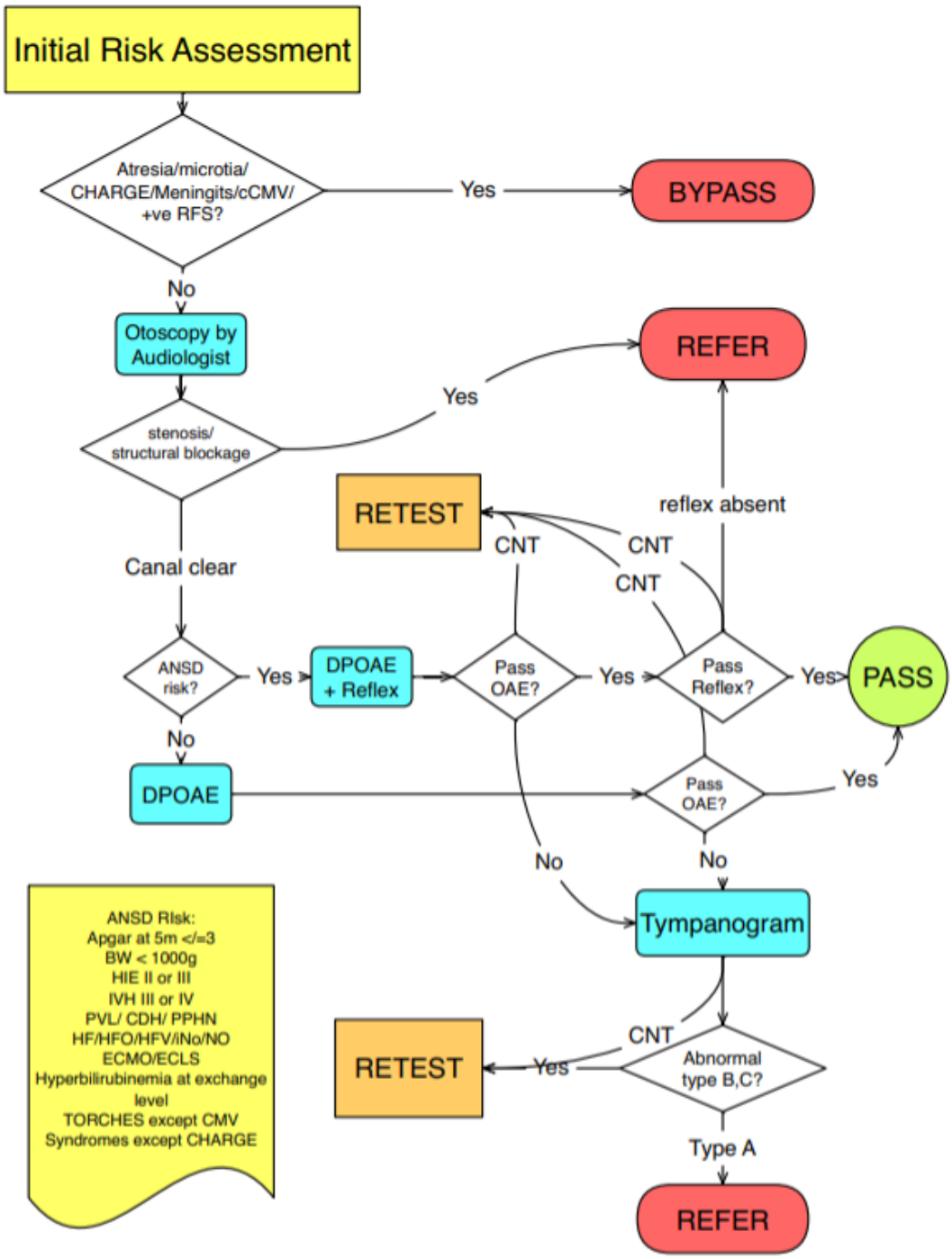
All hearing screening records shall be maintained in a manner satisfying site-specific and IHP Lead Agency policies and the IHP, including retention of the original tympanogram and OAE data. Records detailing results of the hearing screening session and reporting of any necessary information to Healthy Child Development (HCD) – Integrated System for Children Information System (ISCIS) database are required. HCD-ISCIS is a program management system for the reporting of program performance at various levels.

Personal Health Information

Management of all personal health information arising from the modified 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.

Protocol for Modified Physiological Hearing Screen

Overall Process



Risk Assessment

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. For the modified hearing screen, risk indicators are identified by screening personnel through talking to the family. This may be achieved over the phone or in person. A risk assessment conducted over the phone will support efficient triaging of infants who may bypass the modified hearing screen and be routed directly for an IHP Audiology assessment.

In this protocol, the family/caregiver may be the only source of risk information other than the direct physical observation of the child by the screening personnel, 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 provide clear and definite confirmation that a risk indicator exists, then the infant should **not** be considered *At Risk*. When in doubt, the decision should be *No Risk*.

The purposes of risk assessment are to:

1) Determine whether screening should be bypassed; or
2) Decide whether acoustic reflexes are measured; 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. 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 considered 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.

IHP Risk Indicators

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

***Group 1*:** **No Surveillance**	***Group 2*:** **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, 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

Group 1 Risk Indicators

Infants who are identified as having one or more Group 1 risk indicator, *with the exception of family history*, will have acoustic reflexes measured as part of their modified hearing screen. Acoustic reflexes are clinically relevant in the context of suspected ANSD. The likelihood of the presence of ANSD is higher

in infants with a risk indicator compared to infants with no risk. If the screening outcome is an overall 'refer' result, next steps of the protocol are required. If the modified screening outcome is an overall 'pass' result, the infant does not require Audiological Surveillance and is discharged from the IHP.

Group 2 Risk Indicators

Infants who are identified as having a Group 2 risk indicator, *with the exception of cleft palate*, will have acoustic reflexes measured as part of their modified hearing screen. If the screening outcome is an overall 'refer' result, next steps of the 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. Both are included as separate risk indicators in Group 3 and infants identified with either of these risk indicators bypass hearing screening.

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*).

cCMV and Genetic Screen Positive from Risk Factor Screen

Infants who screen positive for cCMV or 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.

Otoscopy

Otoscopy is a recommended first step in the modified hearing screen and must be completed in both ears. Its purpose is to detect foreign bodies, canal occlusion, and any other physical condition of the ear that may invalidate or otherwise contra-indicate the modified screen or indicate referral to a physician. Within this protocol, otoscopic examination must be conducted and interpreted by an Audiologist.

Otoscopy may be more distressing to some babies than an OAE procedure, causing the baby to become active. Also, present OAEs virtually rules out any outer or middle ear dysfunction. Unless the ear looks visually abnormal, conducting OAEs as a first step is appropriate in some cases. If the result is a refer, otoscopy will be required to rule out ear canal occlusion.

Distortion Product Otoacoustic Emission (DPOAE) Testing

Purpose and Priority

DPOAEs reflect cochlear outer hair cell (OHC) function. They are measured best with an f2/f1 ratio of about 1.22 and f1/f2 levels of about 65/55 dB SPL, respectively. DPOAEs yield an approximate yes/no (DPOAE absence/presence) test for significant sensory hearing loss at each f2 value tested, with an effective binary decision criterion at about 40 dB HL.

In the context of the modified hearing screen, DPOAE measurement is mandatory in both ears.

Procedure

DPOAE testing must adhere to this protocol using a screening or diagnostic system with the following parameters:

Diagnostic DPOAE system:

- **F2/F1 ratio 1.22 (standard)**
- **L1 65dB, L2 55dB (standard)**
- **F2 at nominal frequencies of <u>either</u>: (descending order is recommended)**
  - **4kHz, 3kHz, 2kHz, 1.5kHz or**
  - **5kHz, 4kHz, 3kHz, 2kHz**
- **Stopping criteria:**
  - **Minimum DP amplitude: -5 dB**
  - **Noise floor: -17dB**
  - **Signal to noise (SNR) ratio: 8**
  - **Sample size: 1024**
  - **Minimum number of samples: 50**
- **Criteria for PASS: 3 out of 4 frequencies for that ear**

For DPOAE to be considered a PASS:

- Stimulus levels should be flat across test frequencies. Decreases in test level over test time often indicate inadequate probe fit.
- Responses should be replicable within 5 dB.
- Responses should be present: <u>≥</u>5 dB SNR and 8 dB above noise floor.

Accuscreen IHP Screening system:

- **Signal to noise (SNR) ratio: 8**
- **Frequencies tested are 2 kHz, 3 kHz, 4 kHz, 5 kHz in descending order**
- **Criteria for PASS: 3 out of 4 frequencies for that ear**

Tympanometry

Tympanometry in this protocol is mandatory if DPOAEs are absent or result in a refer, and discretional if DPOAEs are present or result in a pass in that ear. Within the protocol, tympanometry may be conducted by a CDA or Audiologist. Results must be interpreted by an Audiologist.

The primary criterion for interpreting the tympanogram in this protocol is the presence and location of a clear peak. Static compensated admittance norms (Table 1) should be used to determine whether a peak can be deemed present. Tympanometric peak pressure should be used to classify any peaks as Type A versus B in babies 6 months corrected age or older (see below). Equivalent ear canal volume should be used to crosscheck otoscopy and case history regarding middle ear pathology including perforations or tubes. If the baby has absent DPOAEs and a Type A tympanogram, the screening result is a "refer" and the next step is an IHP audiology assessment.

Table 1: Tympanometric norms.

	If the tympanic membrane is intact:[1]			
Age group	*Equivalent ear canal volume (Vec)*	*Static compensated admittance (Ytm)*	*Tympanometric Width*[2]	*Tympanometric peak pressure (TPP)*
<6 months (1000 Hz probe)	0.2 – 0.8 cc	> 0.6 mmho (negative tail compensation)	<150 daPa	N/a
6 – 18 months	0.5 – <1.0 cc	≥0.2 mmho	<250 daPa	-75 to + 25 daPa are typical, but peak pressure as low as -199 may be consistent with transient middle ear issues and may resolve. Consider results together with other findings.
>18 months – 10 years	0.6 – 1.2 cc	≥0.3 mmho	<200 daPa	
	If otoscopy reveals a patent tube or perforation			
1 to 7 years[3]	1.0 – 5.5 cc			

[1] Hunter, 2013, BSA 2013, Rosenfeld et al., 2016

[2] For information purposes only

[3] ASHA, 2004

Babies and Infants of Corrected Age Less Than Six Months

Tympanometry must be done with a 1 kHz probe frequency for infants under six months corrected age. The test must be repeated if the trace is noisy or if it is not clearly normal. A clearly normal tympanogram need not be repeated. The key criterion is a compensated peak static admittance of ≥ 0.6 mmho, compensated from the negative tail at -400 daPa.

Infants of Corrected Age Six Months or More

For infants aged six months or more, the probe frequency must be 226 Hz. The criterion in the range 6-18 months is a compensated peak static admittance of ≥0.2 mmho, compensated from the positive tail. Above 18 months, the criterion is ≥0.3 mmho. At any age, a tympanogram that is noisy or not clearly normal must be repeated.

Acoustic Reflexes

Acoustic reflex (AR) screening is mandatory when the infant has been identified as having a Group 1 or Group 2 IHP risk indicator with the exception of family history and cleft palate, where it is discretional. AR is discretional in infants with no identified IHP risk indicator for PHL. ARs may be clinically contributory in the context of suspected ANSD.

If ipsilateral ARs are elected to be done, a 1 kHz probe must be used for infants under six months corrected age and a 226 Hz probe for infants aged six months or more. The eliciting stimulus may be a 1 kHz tone or Broad-Band Noise (BBN). BBN is the preferred stimulus because it is usually more effective than tonal stimuli for reflex elicitation, which reduces false-positive reflex absence. The goal of the AR screen is not to establish an accurate reflex threshold, but to show presence or absence of reflexes at an appropriate stimulus level. The stimulus level should start at 80 or 85 dB and be no greater than 95 dB HL. Printouts are suggested as reflex waveform anomalies do occur. Reflex presence is usually defined by a repeatable, clear, negative deflection. Biphasic and even positive deflections sometimes occur. It is the reproducibility of the elicited waveform, not its precise morphology which is the primary factor in response identification.

Maximum Number of Modified Hearing Screening Attempts

The modified hearing screen is considered complete where a PASS or REFER result is obtained in **both** ears. If the screen is incomplete during the initial visit, the child may be rebooked for ***only*** one more attempt. If this appointment results in an incomplete screen regardless of the child's risk status, please contact the IHP Lead Agency for further direction and make appropriate recommendations (e.g., monitor developmental milestones).

Messaging to Families

When an infant has obtained an overall refer result from the modified 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 within the IHP. Families often do not understand the difference between an assessment and a hearing screening. It is important to follow the scripts in Appendix D to explain that a screening test refer means that hearing loss is now more probable but is not certain. 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. **The key message and the family's understanding and agreement to attend are the top priority.**

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Appendices

Appendix A: Clarifying Access to the Modified Hearing Screen

Due to the pandemic, many IHP services paused and some Ontario-born babies did not receive newborn hearing screening or their newborn screens were incomplete. Risk factor bloodspot screening continued during the pandemic. The following questions are aimed at providing support to IHP Lead Agencies when triaging Ontario babies who missed or have incomplete newborn hearing screens.

Item	Question	Response	Action	Notes
1	How old is the child? (corrected age)	≤ 8 weeks	IHP Universal Newborn Hearing Screen	Arrange IHP Universal Newborn Hearing Screen
		> 8 weeks 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	Check database
3	If screened, what was the result?	**Pass**	Check database	Describe next steps
		Refer with no further testing	IHP Audiology Assessment	Obtain relevant information and provide to IHP Lead Agency[a]
		No Result / Don't know	Question #4	Check database
4	Has the child been proven to have atresia/microtia, CHARGE syndrome, meningitis, cCMV, or screen positive for genetic riskon the bloodspot?	**Yes**	Bypass Modified Screen for IHP Audiology Assessment	Obtain relevant information and provide to IHP Lead Agency[a]
		No	IHP Modified Screen	Arrange IHP Modified Screen

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

Appendix B: Description of IHP Risk Indicators

Group 1

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).

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.

Family history of parent or sibling with PHL identified by 10 years of age: 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. 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 on either question 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.

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.

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 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 recently been deleted from the IHP risk indicator list.

Group 2

Cleft palate: Cleft palate has a strong association with hearing loss, both permanent and temporary. Cleft lip alone (isolated cleft lip) can be used as a cue for investigation of the presence of a cleft palate.

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.

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. 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.

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 PHL except CHARGE: 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.

Group 3

Atresia/Microtia: 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 1). Atresia and microtia can have varying grades of severity.

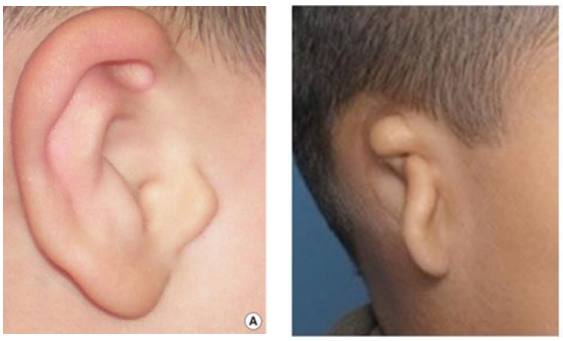


Figure 1: The picture on the left is an example of auditory ear canal atresia. The picture on the right is an example of microtia of the ear. (Source: Google Images)

CHARGE Syndrome: 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.

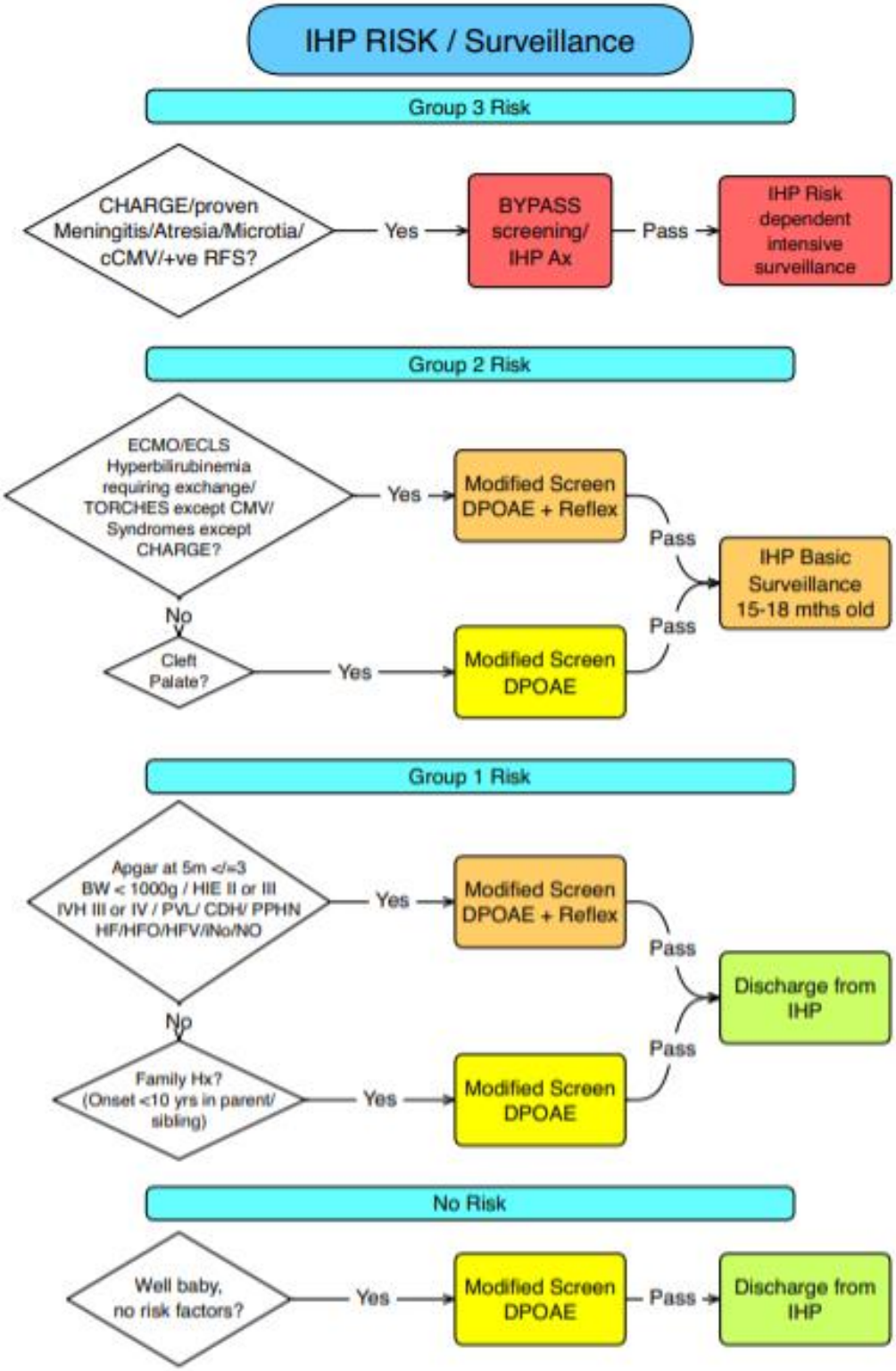
Proven Congenital Cytomegalovirus (cCMV): Medical diagnosis of symptomatic cCMV infection external to the bloodspot risk factor screen may occur in hospital. This remains a part of the risk assessment and continues to be a bypass risk indicator. Infants who screen positive for cCMV who are not identified 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.

Genetic Screen Positive: Those infants who screen positive on the bloodspot risk factor screen for genetics will bypass screening and be re-directed to audiology for assessment. 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.

Appendix C: IHP Risk Indicators and Surveillance



Appendix D: Scripts Used for IHP Modified Hearing Screen

Explaining Screening Results to Families

Pass and Infant At Risk with Group 2 Risk Indicator

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 the IHP so arrangements to see you sooner can be made.

Pass and Infant Not At Risk or Group 1 Risk Indicator

The hearing screen is done and the result is a "Pass" for both ears. Please share the results with your baby's doctor. Since your baby does not have any risk indicators for developing hearing loss later, no further testing is needed now.

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.

Refer and Infant At Risk with Group 1 or 2 Risk Indicator

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 IHP 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 share the results with your baby's doctor and the follow-up that has been recommended by the Infant Hearing Program.

Refer and Infant Not At Risk

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 IHP Audiologist to find out how well your baby hears.

Hearing loss could affect his/her speech and language development, so it is very important for you to attend this follow-up appointment. Please share the results with your baby's doctor and the follow-up that has been recommended by the Infant Hearing Program.

No Result, Regardless of Risk

I have attempted to screen your baby's hearing but was not able to complete the screen. Some babies need to be screened more than once in order to get an accurate result. We will arrange another appointment to re-attempt the hearing screen.