INFANT HEARING PROGRAM

SCREENING TRAINING MANUAL

Rev. June 2002

SCREENING CURRICULUM (STAGE 1 AND 2)

INTRODUCTION

The Infant Hearing Screening and Communication Development Program (Infant Hearing Program) was announced in the Ontario Government's 2000 Budget. This new Program builds on the previously introduced government programs designed to give children a better start in life, including the Preschool Speech and Language Initiative and Healthy Babies/Healthy Children.

This manual has been developed to assist in the local implementation of the Infant Hearing Program across Ontario. A province wide standard will ensure that infants born deaf or hard of hearing or at risk of developing hearing loss in early childhood will be identified and that access to the necessary services and supports will be available to these babies and their families. When implemented this new program will integrate all service providers required to meet the vision and goals of the program.

CONTEXT

The importance of acquiring communication skills early in life is well understood. Research has shown that delay in language development can have significant impact on cognitive, emotional and psychosocial development and that language development is a prime indicator of future academic success. Children born with permanent hearing loss or who acquire permanent hearing loss during the early years of life are at risk for delay in language development, if the hearing loss is not identified early. Further, it is well understood that the earlier the hearing loss is identified, and supports and services for communication development are provided, the better the acquisition of language skills.

Screening of newborns to detect permanent hearing loss has been advocated for many years. In Ontario, screening of infants at high risk for hearing loss has been provided in some hospitals, but these services have reached only a small number of the 140,000 babies born in the province each year. The average age of identification of hearing loss in young children has remained about 2 ½ years of age in Ontario, while the recommended maximum age is 6 months.

VISION

Children who are born deaf or hard-of-hearing or at risk for developing hearing loss in early childhood will be identified and will receive the services and supports required for communication development, so that they will acquire the communication skills needed for performance of daily activities required for personal and social sufficiency at home and at school.

GOALS OF THE PROGRAM

The goals of this program are to provide an integrated system of services in all parts of Ontario that will:

1. identify infants born deaf or hard of hearing or at risk for developing hearing loss in early childhood and

2. provide child and family centered services to support communication development.

PRINCIPLES

1. There will be access to the services of this program across Ontario
2. Every aspect of this program will be provided based on fully informed parent/guardian choice and consent, and will comply with confidentiality requirements.
3. All services will be child and family centered taking into consideration the cultural and ethnic diversity of the people of Ontario.
4. A seamless system of services will be developed that integrates this program with other existing children's programs and services.
5. The components of this program will be developed using the principles of evidence based practice.
6. The program will be monitored and evaluated on an ongoing basis.
7. The quality of the program will be continuously improved, based on the evaluations.

THE PROGRAM

The Infant Hearing Screening and Communication Development Program (Infant Hearing Program) will have three major components.

1. Universal Hearing Screening
2. Hearing Loss Confirmation and Audiologic Assessment
3. Follow up Support and Services

Each of these three components will encompass several aspects of service delivery and supports.

All equipment and procedures used for screening and assessment have been specified by the Provincial Infant Hearing Program. Detailed protocols are being developed and will be followed for delivery of all components of the Program. These protocols will be reviewed and changed as required to reflect current evidence based practice. As new changing evidence emerges, adjustments will be made as required to the program equipment as well as protocols.

Training will be provided for individuals who will be involved in the delivery of all aspects of the program. All information on the services provided through this program will be recorded on the Integrated Services for Children Database and reported on a regular basis to the Ministry of Health and Long-Term Care. The Program will be administered through the infrastructure of the Preschool Speech and Language Initiative (PSL) using 12 Coordinating PSL Systems to coordinate local service delivery.

PROGRAM COMPONENTS

1. UNIVERSAL NEWBORN HEARING SCREENING

The hearing screening component of this program is designed, in keeping with the most current evidence, to achieve as low a false positive rate as possible, ideally less that 4%.

It is expected that the model will constantly change over time as new evidence provides information of improved techniques and methodology.

In 2000 the recommended Universal Newborn Hearing Screening protocol was a 2 stage process for all newborns. Those babies that received a "refer" result from the stage 1 screening would have a second screening. Those babies with risk factors for congenital permanent hearing loss would be screened with Automated Auditory Brainstem Response (AABR) technology for both stages while babies with no known risk factors would be screened for stage 1 with Automated Distortion Product Otoacoustic Emissions (ADPOAE) technology and AABR for stage 2.

Based on the most current evidence the recommended screening protocols have changed. Babies at risk for congenital permanent hearing loss and babies who do not have any known risk factors will now be managed differently.

As planned there will be 3 approaches to the Universal Screening:
1) HOSPITAL PREDISCHARGE WELL BABY SCREENING HOSPITAL
2) PREDISCHARGE HIGH RISK BABIES (NICU) SCREENING
3) COMMUNITY SCREENING AND HIGH RISK MONITORING

1) HOSPITAL PREDISCHARGE WELL BABY SCREENING

The mother's of all newborn's who do not have any known risk factors for congenital hearing loss will be offered a hearing screening for their baby prior to discharge from the hospital birth admission.

This predischarge screening will be provided by screening personnel hired by the Infant Hearing Program, or hospital personnel funded for this service through the Infant Hearing Program.

In keeping with the most recent evidence the "Well Baby Screening" will be a 2 stage process.

i). STAGE 1

The Stage 1 process consists of 2 parts, babies who receive a "refer" result form part 1 will have the second part of the screening.

PART 1
Part 1 of the Well Baby Screening will be done using Automated Distortion Product Otoacoustic Emissions (ADPOAE) technology. This technology has been selected mainly because it is noninvasive, very quick, straightforward and yields information about hearing sensitivity at specific frequencies of interest.

PART 2
Every baby with a "refer" result from the DPOAE, must have the second part of the Stage 1 process, which is a screening using Automated Auditory Brainstem Response (AABR). Ideally this should occur before the baby is discharged from hospital, but when this is not possible, as soon after discharge as possible.

ii). STAGE 2

Every baby with a “refer” result from the Stage 1 screening should receive another screen using the Automated Auditory Brainstem Response (AABR) technology The AABR technology is more advanced, and takes slightly longer to administer than the DPOAE, but is also relatively quick, painless and automatic. Ideally this Stage 2 screening will be arranged after 2 to 3 weeks. to allow time for the resolution of any middle ear disease that may cause a false positive result. Babies with a “refer” result from Stage 2 screening will then receive an audiology assessment for confirmation of hearing loss.

2) HOSPITAL PREDISCHARGE HIGH RISK BABIES (NICU) SCREENING

Any baby with risk factors for congenital hearing loss will be screened using Automated Auditory Brainstem Response (AABR) technology. This screening is a 1 Stage process. Babies with a “refer” result from this screening will go directly for audiologic assessment .

Since about half of the target population will be found in the NICU population, it is essential that these babies receive a successful screening that yields either a “pass” or a “refer” result. If for any reason a baby was not screened prior to discharge from the birth admission or receives a “No Result” from the screening, a screening should be done as soon as possible in the community.

Any baby that, as a result of a readmission, is found to have risk factors that were not present or known at the birth admission discharge, should be screened using the Automated Auditory Brainstem Response (AABR) prior to discharge if possible.

Many babies who spend time in the NICU have risk factors for progressive or early-onset hearing loss. Therefore, even if these babies pass the Stage 1 screening procedure, they will be followed through repeat Stage 1 ADPOAE (Automated Distortion Product Otoacoustic Emissions) screenings until they are 3 years of age. Further details on this high risk monitoring are described in a later section of this document.

3) COMMUNITY SCREENING

Regularly scheduled screening clinics will be offered in community locations throughout each of the 12 regions to provide screening to babies who missed the hospital predischarge screening or did not have a successful result from Stage 1 screening either in the NICU or the well baby nursery.
These clinics will also be used to provide the Stage 2 screening for those babies who had a “refer” result from Stage 1. Stage 2 screening should be completed within 2-3 weeks, but preferably no later than 2 months of age.

This Community screening will be provided by screening personnel hired or funded through the Infant Hearing Program

HIGH RISK MONITORING

Babies at risk for progressive hearing loss will be monitored through Stage 1 rescreening until they reach 3 years of age. This screening will be conducted by appointment at the Community Screening Clinics using ADPOAE (Automated Distortion Product Otoacoustic Emissions) equipment.

The high risk monitoring screening will be carried out by screening personnel hired or funded through the Infant Hearing Program. It will be determined locally how and by whom this screening will be made available.

2. ASSESSMENT

   A. Hearing loss confirmation and Audiologic Assessment

   B. Medical Referral and Management

3. FOLLOW UP SUPPORT AND SERVICES

   A. Family Support and Access to Information
      a) Hearing Aid Dispensing
      b) Other assistive technology

   B. Communication Development

RISK ASSESSMENT

Criteria for 'Risk' present at birth
Babies with ANY of the following criteria should be screened using the ABAER equipment before discharge from Level III or Level II NICU:

- *All Retrotransfers from Level III to Level II NICU*
- *All babies born # 34 weeks gestation*
- *All babies with craniofacial anomalies (including minor anomalies such as ear tags, malformed pinnae, etc)*
- *All babies born > 34 weeks gestation with a length of stay >48 hours, but EXCLUDING those with the following conditions:*
    - *Transient tachypnea of the newborn*
    - *Transient hypoglycemia*
    - *Feeding difficulties*
    - *Babies under observation for minor conditions*

Wherever possible, testing should be performed at a corrected age of 34-35 weeks gestation and prior to discharge from NICU or Special Care Nursery. Testing should NOT be done at < 34 weeks, and NOT within 48 hours of birth.

Additional Neonatal Risk Factors for Hearing Loss

The following two risk factors place a well baby at greater risk for hearing impairment. Learning to gather this information from the parent, and learning to identify the appearance of these physical factors is covered in Appendix A - How to Conduct the Risk Assessment.
These risk factors are:

1. *There is someone in the immediate family who had a hearing problem from early childhood (parents, siblings, grandparents, aunts, uncles, first cousins ONLY)*

2. *The baby's development is different than other babies, or the baby has facial characteristics (craniofacial anomalies) that are linked to the development of hearing loss*

The following checklist identifies some of the craniofacial indicators that place the child at increased risk for hearing loss, and should be recognizable by informal inspection.

EARS
Position
*Any marked positional asymmetry, OR
*With the head vertical, the upper margin of either or both pinnae is clearly below the horizontal plane of the outer corners of the eyes. (Low-set ears are often, but not always, rotated posteriorly, with the lobe more forward than usual).

Size
*Any marked size asymmetry, OR
*The length of either or both ears (visually estimated) from the upper extremity of the pinna to the lower extremity of the lobe is about an inch or less (< 3 cm, the 3rd percentile of the normal range in the newborn).

Shape
*Any marked shape asymmetry, OR
*Obvious morphologic deformity, such as missing earlobe(s). Familial variation (e.g. bat-ears) should be taken into account.

Canal patency
*Absent or very narrow (tiny or slit-like) ear canal (meatal) opening.

Preauricular tags/pits
*Any obvious skin flap or pit on the ear just in front of the meatal opening, or on or near a line from the meatus to the corner of the mouth, IN COMBINATION with any other malformation, including but not limited to facial asymmetry, under-developed mandible or maxilla, facial droop, ocular dermoid cysts, lop ear, neck fistula.

EYES
Separation
*The distance between the inner corners of the eyes is estimated by visual inspection to be about one inch or more (2.5 cm, the 97th percentile of the normal range in the newborn). If the nasal bridge is poorly developed, guard against false impression of wide separation.

Angle

*Marked up-slant or down-slant. Slant is easier to assess with eyes closed (palpebral fissure angle). Guard against a false impression of slant, due to inner epicanthal folding. Slight upslant is not uncommon and is NOT a risk factor.

*Marked medial epicanthal folding (Trisomy 21). Familial variation should be take into account.

Iris anomalies
*Missing segment (coloboma, keyhole defect) of iris.
*Different-coloured irises (heterochromia).
*Colourless iris(es)(achromia).

MOUTH
Cleft lip and/or palate
*Expression of this anomaly may range from a thin (but definite) scar-like closure line above the upper lip through to a full cleft with secondary features such as markedly flared nares (nasal alae).

SKIN & HAIR
*Multiple, freckle-like facial spots. *White forelock or white hair patch(es).

OTHER MINOR ANOMALIES
Cumulatively, the large set of other anomalies such as flat facies, eye protrusion, Brushfield iris spots, under- or over-development of any part of the face or head, slight asymmetries, flat facies, facial droop, abnormal hair growth, neck fissures, large tongue, etc, can be quite common in isolation. For anomalies other than those identified and starred earlier, TWO OR MORE minor anomalies should be clearly present in order to set the risk indicator positive for hearing impairment**.**

PERFORMING THE SCREENING

(A) Stage 1 Screening with Distortion Product Otoacoustic Emissions (DPOAE)

Screening of hearing with DPOAE is usually quick, easy to do and reliable. It is completely objective. No behavioral response is needed from the baby, who ideally is tested asleep. Under good conditions, it takes less than two minutes to screen a baby, less than one minute per ear. The testing is done using a hand-held, automatic screening unit.

The test involves presenting sounds to the baby through a special probe that is gently inserted into the entrance of the ear canal. The normal inner ear actually generates tiny sounds in response to the stimulus, and these sounds are the otoacoustic emissions (OAE). A computer inside the screening unit distinguishes the OAE from ambient sound, which is always present in the baby's ear canal. Some ambient sound comes from the room where the test is done. Other sounds are generated if the baby or the probe moves during the test. The computer decides whether a real OAE is present, or just ambient sound, and then displays a 'pass' or 'refer' result.
The important ingredients of a successful screening test are a quiet room, a quiet baby and correct positioning of the probe in the ear. By a quiet room is meant a room in which it would be possible to have a conversation by whispering. There should be no

talking during the test, which will not last more than about a minute. If the environment is obviously too noisy, and cannot be made more quiet, it may be a good idea to go into another area for few minutes, if available.

The ideal subject is a sleeping baby, although it is possible to test a baby who is awake but laying still and quietly. If the baby is fussing, wriggling, fretting or crying, a successful test is unlikely. However, most neonates spend most of their time sleeping, and they can be fussing one minute and sleeping the next, so there are usually going to be plenty of opportunity to test. If the baby is not asleep or at least laying quietly, it may be necessary to wait a few minutes. Babies can be tested in their mothers' arms, if necessary but cannot usually be tested while actually feeding, because the act of sucking moves the walls of the ear canal.

If the environment is less than satisfactory but the screener has done what she or he can within reason to create a good test situation, the test can be tried on one ear, and if a pass is obtained then the other ear should be tested. It does not matter which ear is done first. If any ear produces a “refer” result the test should be tried again on that ear. If there is an obvious cause of failure the test should be re-tried after an attempt to solve the problem. There is no need to retest an ear that has passed.

The screening unit is light, portable, and easy to use and powered by a rechargeable battery. Simple operating instructions are displayed when necessary. Screening is totally automatic, and the device will check to see if the probe has been properly fitted to the ear canal; if not, a message is displayed. Test results are stored automatically, and may be printed out on a custom mini-printer, when convenient.

Parents usually want to know the results of the test right away. If there is a pass in both ears, there is usually no problem. If one or both ears give a refer result, then it is important to reassure the parent(s) that all this means is that a more detailed hearing check-up would be a good idea, preferably within a few weeks. The word 'fail' is never used in connection with a screening result, because it alarms parents unnecessarily and because it does not capture the true significance of not recording an OAE. If the test is done properly, less than about 5% of well babies will fail it. If the baby does indeed fail, what it means is that there is an increased *risk* of a hearing problem, but not that there *is* a hearing problem. The odds are that a baby who fails the screen will have normal hearing, but virtually ALL babies who DO have hearing loss will fail. Leading authorities on hearing health consider the false-positive rate for this test to be a perfectly acceptable price to pay for the ability to detect all babies with hearing problems. This test is much more reliable than any possible test based on observing the baby's behavior. Behavioral screening tests are no longer recommended.

The fact that the screening test is obviously not behavioral means that in the event of a refer result it can be emphasized that this initial screening test does not actually measure true hearing. It measures 'echoes that come out of the baby's ear, and is a good test but certainly not perfect'.

The most likely reasons for not getting a pass result are that the conditions in the ear canal were not quiet enough. The OAE are very, very faint sounds and can be hard to detect. Another possibility is that there was debris in the ear canal, blocking the OAE probe. Also, the baby might have a cold, which may cause a small hearing loss. There are many reasons not to get a pass result, so the parents can be reassured. However, it

is certainly desirable, and in the baby's best interests, that they attend for a follow-up check.

The follow-up check is a stage 2 screen using a more advanced test, the AABR (automated auditory brainstem response), that measures the baby's brainwaves (EEG). The AABR test is also quick, painless and automatic, and is best done on a sleeping baby. It is more accurate than the DPOAE test, and the equipment is MUCH more expensive. It usually takes about 15 minutes, in a quiet baby. Most babies will pass this Stage 2 screen.

(B) Stage1 - Part 2/ Stage 2 Screening with Automated Auditory Brainstem Response (AABR)

AABR screening is the recording of the EEG through noninvasive, disposable skin electrodes placed on the head and neck. Controlled acoustical stimuli sounds are delivered via earphones. A computer extracts and interprets the EEG response to sound automatically. Typically, the test takes a total of 10 minutes or less. The test is well tolerated, and is best carried out while the infant is asleep.

A key requirement for successful AABR screening is that the baby should be asleep or at least drowsy and quiet. Screening will not be successful in a baby who is crying or restless, because electrical activity from head and neck musculature will obscure the EEG response to sound. The screening equipment can inform screening personnel that test conditions are not satisfactory.

Wherever possible, the time and place of testing should be chosen to maximize the likelihood of the baby being sleepy. The act of placing the EEG electrodes may wake up the baby, so testing after a long sleep is ill advised. Wherever feasible, swaddling or other maneuvers that promote relaxation and sleep may be helpful.

Other important factors include environmental noise, activity and lighting. Sudden noise can be distracting and alerting. Steady noise at a moderate level may actually mask the infant's perception of the stimulus. Low and steady noise levels should not be a problem. While quantitative limits to the noise levels that can be tolerated are not easy to specify, the quieter the test environment the better. Testing adjacent to a source of continuous noise such as a running tap or noisy instrument is inadvisable.

AABR equipment is usually mounted on a small, movable cart that is wheeled to the test location. Having ensured that the test environment is satisfactory and that the baby is at least drowsy, the electrodes are attached usually at the high forehead and behind each ear. The skin is cleaned beforehand using an alcohol swab and the self-adhesive electrode pads applied. The stimulus is delivered to each ear in turn, via an insert earphone with a disposable eartip.

If both ears give a 'pass' result, the screening is complete. If any ear gives a 'refer' result (i.e. fails the screen), the test should be repeated on that ear. If there is an obvious reason for the failure, such as an unsettled baby, waiting a short period before re-testing may allow the baby to settle and give a screening 'pass'. Both ears must pass for the baby as a whole to pass. If on any ear there is a repeated 'refer' result, the baby has

failed the screen. If satisfactory conditions for screening (e.g. a quiet baby) cannot be achieved within reasonable time and effort, then the baby should be scheduled for re-screening.

Infection Control

Hearing screening performed in the NICU should be carried out using hospital infection control protocols.

In the community, at a minimum, the following infection control procedures should be followed:

- Wash your hands before each screening or in between screenings.
- Use anti-bacterial hand-wash if there is no sink readily available.
- Wipe down all the equipment using disinfectant spray regularly.
- The probes should be cleaned using the probe cleaners provided, whenever debris is visible or at least every 50 screens.

Never re-use the disposable supplies on another baby. (Once used, the probe tips and other supplies can be disposed of in the regular garbage collection).

DATA MANAGEMENT AND COMPLETION OF FORMS

The ABAER System

The ABAER uses a laptop computer to perform the hearing screen. Although the ABAER has its own built-in database, this will not be used for the IH Program, as all information will be entered into the provincial database (ISCIS) using data entry forms (see below). However, there are some mandatory fields in the ABAER software, which need to be populated in order to proceed with the screening:

- Child's name (where the first name is unknown, enter "Baby")
- Child's date of birth
- Child's gender

Unique identifier for the ABAER database. In this field, enter the date of birth of the child again, but in the following format [YYYY, followed by month in 3 letters, followed by day in 2 digits, followed by the first 5 or 6 letters of the last name]. For example: 2001JAN12ROBINS. The maximum number of characters is 15. Do not use the child's temporary or permanent health card number, as the IH Program is not permitted to collect this information provincially, nor is it consistently available. The unique identifier is not important for any other reason than to proceed with the screening test. A form is provided to capture all the important program information (see below).

Data for the Infant Hearing Program (IHP) is captured in the Preschool Speech and Language (PSL) version of ISCIS (Integrated Services for Children Information System). Each IH Regional Co-ordinating Agency will enter the data for babies who reside in their region.

Types of data being collected includes:

- Demographic data (name, address, date of birth, etc. for the child and for the family). Note that the health card number is not required.

- Service delivery data
- Dates of screenings/assessments/interventions
- Service status (e.g., waiting, accessing service, etc.)
- Whether the child is High Risk or not (of being born deaf or hard of hearing or of developing a hearing loss in early childhood)
- Location where service was provided
- Names of screening and other personnel
- Results of screenings/assessments

PURPOSE OF DATA COLLECTION

1. Service delivery

These data are being used to track babies and their families as they pass through the various stages of the IHP; to make sure no baby gets missed; to track wait times between service delivery points and to schedule appointments. Capturing these data is an essential part of service delivery. Parents should be encouraged to consent to sharing the above demographic and service delivery data within their regional IH System to enable services to be provided.

2. Monitoring of the Program

The Ministry of Health and Long-Term Care will receive aggregate data (no individualized or personally identifiable data) on a quarterly and annual basis from each regional IHP agency. Individualized data will only be shared at the local level.

CONSENT

The hearing screening is a quick, easy, non-invasive screening test, which does not involve the performance of any of the controlled acts under the *Regulated Health Professions Act, 1991.* However, provision of parental consent is required.
The importance of hearing screening and the procedure itself should be explained to the parent prior to doing the screen. This includes preparing the parent for both pass and refer outcomes of the screen and explaining the results in more detail afterwards. Please refer to the section “Communicating with Parents” for information on explaining the purpose and details of the hearing screening procedure to parents.

Release of Information

It is important that parents give consent to share information between specified agencies within the local/regional IHP System. As mentioned above, the IHP co-ordinating agency must have information on the babies who have received the screening (and any other services) in order to track that baby/family and make sure they receive appropriate follow-up care. The provision of this kind of consent is essential in order to provide service. It is understood that local procedures for authorization for release of information between one agency and another will be followed.

COPIES OF THE FORM AND WHERE TO SEND IT

The final version will be in triplicate – the original should be sent to your regional IH Co-ordinating Agency (not your local PSL agency); one copy should be given to the family and one copy can be sent to the primary care physician with parental consent as per local procedures. In cases where the baby has a refer result from Stage 2 and is being referred to audiology, you can ask the parents to show the audiologist the form at the time of their appointment.

APPENDIX A

How To Conduct the Risk Assessment For Permanent Childhood Hearing Impairment (PCHI)

1. **Family History,**
2. **Craniofacial Assessment**

During the initial screening appointment, screeners will perform a 'risk assessment' to determine if the baby is at risk for Permanent Childhood Hearing Impairment (PCHI) and will therefore need to have hearing monitored periodically following the initial 'pass' of the hearing screening.

In this section, terminology will be used that may now be considered inappropriate by health professionals. It is included because the general public may use these terms, or recall their use in the past, which may be informative.

Two risk indicators for PCHI can be assessed during the screening appointment. One relates to family history and the other to anatomical features of the head. The first involves a few careful questions, the other a quick inspection of the baby's appearance. The overall risk assessment should not take more than about five minutes, and typically will take less time. Whatever can be done in five minute is sufficient.

There appears to be no strong reason whether the assessment of these risk indicators should be done before or after the actual hearing-screening test. Whatever comes naturally or seems the most convenient during the individual visit may be the best way.

Risk Indicator 1. Family history of permanent childhood hearing loss.

The mother, according to research, is the first choice to be asked about any family history of PCHI. If both parents are available and seem helpful, better information may be gained by asking both parents together.

Before questioning, the ground is prepared as follows:

"I'd like to talk for a minute about whether there's any history of childhood hearing problems in your family or the family of your baby's father. The reason is that if there IS a family history of certain kinds of hearing problems in childhood, it might be a good idea for us to check from time to time and make sure that <baby's name>'s hearing is fine. Even if we DO find a family history, the chance of <baby's name> having a problem is very, very small. So, is it OK with you to talk about family history?"

The task is to get at the required information and to assess its reliability. The first question asked is

"Do you know if any of <baby's name>'s close relatives have had hearing problems or deafness in early childhood?" By 'close relatives', what is meant is any of the child's brothers or sisters, mother and father, aunts and uncles, maternal or paternal first cousins and grandparents. NO ONE ELSE MATTERS.

'Hearing problems' means any perception or memory of the affected family member having difficulty hearing for a period of years, or having a hearing aid as a child, or being considered or referred to as 'deaf' or 'hearing-impaired' or 'hard-of-hearing' or as having 'problems with his/her ears'.

'Early childhood' means that the problem probably started at age less than about ten years. There is no hard and fast limit, but it does not mean over 15 years.

A possible cause of confusion is that ear infections and other disorders affecting the middle ear (the space behind the eardrum) are common and often cause hearing loss but DO NOT lead to positive risk, because such hearing problems are rarely permanent.

Only permanent hearing loss indicates risk, so if there is any indication of an actual or possible hearing problem, the screener's task is to try and sort out whether the problem was likely to have been due to PCHI or to ear infections.

A strong clue about whether a hearing loss is or was permanent is the affected person having used a hearing aid. However, lack of knowledge about any hearing aid does not rule out PCHI. For example, many years ago some children with PCHI never got hearing aids. New devices such as cochlear implants are definite indicators of PCHI. Usually, if the word 'deaf' is or was used in reference to the affected child, it suggests PCHI, as does any indication of use of sign language or having attended a special school for the deaf.

Reports about hearing problems in more than one close family member increase the likelihood that there is positive risk of permanent hearing loss of genetic origin.

Clues that suggest that a 'hearing problem' or 'ear problem' may have been due to ear infections include recollection of pain, ear discharge, treatment with antibiotics and/or ear surgery to put in 'tubes' or 'grommets'. There may have been a 'no swimming' order. There may have been many episodes, because ear infections often recur in young children.

Remember that even children who have PCHI get ear infections. The question is whether any hearing problem reported is likely to have been EXCLUSIVELY due to ear infections.

To maximize the net benefit from screening, it is important to minimize parental anxiety throughout the processes of testing and risk indicator ascertainment. The screener should always remember that information is being volunteered by the parent(s), not extracted from them. They are not obliged to respond. They should be encouraged gently to do so, if willing, but they should on no account be pressed, especially if there is any indication of reluctance, anxiety or frustration. When in doubt, the parent should be reassured that the matter is not crucial and the screener should move on to the next activity.

Note that the baby may be adopted, or the father may not be known. In the case of adoption, the parents may know little about family history. If the father is unknown, the information will clearly be incomplete even if mom's history is clear and negative.

If there is a clear indication of PCHI, or if on balance it seems very likely that there is or was PCHI, the family history indicator is marked YES. If there is fairly clear recollection and no memory of any problem, AND the information base is complete (both maternal and paternal information known), the indicator is marked NO. In every other case, the indicator is marked UNKNOWN. It can be quite difficult to assess the reliability of the information, especially for the first few assessments. It may be helpful for the screener to think back about his or her own family, and imagine having said what the mother just said. Does it add up to being fairly sure that there was PCHI, or just raising a vague possibility?
There is much clinical experience indicating that for individual screeners, the speed, reliability and ease of determining the quality of the information will increase rapidly over time.

Risk Indicator 2. Craniofacial anomalies or stigmata

Certain unusual features of the face or head are associated with increased risk of PCHI. An example would be low-set ears, skin tags near the ear, eyes of a different color, a white forelock, etc. These kinds of anomalies are quite often associated with PCHI, as a result of genetic or environmental factors affecting common structures during early fetal development. Some of the anomalies may occur as part of a recognized syndrome (e.g., Down, Goldenhar, Pierre-Robin, Treacher-Collins, Turner, Waardenberg, etc).

The screener should carry out a brief and cursory inspection of the baby's head and neck, as the opportunity arises naturally. A detailed, deliberate inspection is not advisable, because anatomical characteristics may be a delicate issue with some parents. A brief check for obvious anomalies is all that is needed. This physical inspection is limited to what is immediately observable in the head and neck area. It will not be necessary for the screener to develop a detailed knowledge of craniofacial anomalies.
If any of the key characteristics is clearly apparent, the risk indicator is positive. Otherwise, it is considered unknown, because the physical examination is not exhaustive.

An important area in relation to this risk indicator is that of parental awareness, anxiety and sensitivity. A given anomaly may have many possible etiologies. Parents may or may not be aware of the anomaly or even of a syndrome, given that some syndromes can vary in their level of expression in the individual, and also that professional awareness of the significance of isolated anomalies may be limited. It is possible to code the risk indicator so that discussion of the issue need not arise inadvertently, but there are several concerns with masking the finding of an anomaly. It is conceivable, for example that the anomaly was not noticed previously and is important clinically. Also, the anomaly indicator may be the only thing that triggers a need for surveillance re-screening of hearing. Ideally the parents would be prepared for the re-screen, and at the time of scheduling the appointment, the parents would want to know why the test is necessary. Disclosure of a risk indicator at that time may be more problematic than doing it in an up-front manner.

One approach to this is to be quite overt in the logging of the indicator, and to mention the anomaly in a matter-of-fact way such as to minimize anxiety. The precise method will depend on the situation and be dictated by the judgement of the screener. In the case of an isolated anomaly such as moderately low-set ears, for example, the approach might

be: “It looks to me as if <baby's name>'s ears might be a little bit lower down than usual. That's one of the things on my list so I'll make a note of it. What it means is that it would be a good idea for us to check from time to time…” Then, the various parental reactions and counseling options should be dealt with as considered appropriate by the screener.

Approaches to questions from parents about the significance of various anomalies will vary. The two most likely issues are on the one hand the syndromic possibilities and on the other the significance for hearing. If appropriate, the former might be best handled by suggesting that the finding be mentioned to the family doctor or pediatrician at the next visit. The hearing facet can be dealt with in the usual way with risk indicators, by minimizing the likelihood of anything being wrong but at the same time recommending that the child attend the check-up appointment when the call comes.

APPENDIX B

COMMUNICATING WITH PARENTS
SAMPLE QUESTIONS:

Before the screening:

1. Why are you screening my baby's hearing?
2. What is a DPOAE screening test? How does it work?
3. What is an AABR screening test? How does it work?
4. What does it mean to "Pass" the screening?
5. What does a "Refer" result mean?
6. What can cause a "Refer" result?

After the screening:

7. If baby "Passes" the screening, what is the next step?
8. If baby "Passes" the screening, and there are risk factors, what happens next?
9. If the result is "No Result" what does it mean?
10. If there is a "Refer" result, what is the Stage 2 screen?
11. If the result is "Refer" from Stage 2, the next step is an Audiology Assessment.

SUGGESTED ANSWERS:

1. Why are you screening my baby's hearing?

The importance of acquiring communication skills early in life is well understood. Research has shown that delay in language development can have significant impact on cognitive, emotional and psychosocial development and that language development is a prime indicator of future academic success. Children born with permanent hearing loss or who acquire permanent hearing loss during the early years of life are at risk for delay in language development, if the hearing loss is not identified early. Further, it is well understood that the earlier the hearing loss is identified, and supports and services for communication development are provided, the better the acquisition of language skills.

Serious hearing loss is found in only six of every 1000 babies born in Canada. Some babies may develop hearing loss later in life from repeated ear infections, meningitis, head injury or other medical conditions. Sometimes babies lose their hearing and the reason in never known.

For babies in intensive care for a number of days after birth, 1 in 50 babies will have significant hearing loss. There is also a higher risk of late onset or progressive hearing loss for babies who graduate from NICU care.

2. What is a DPOAE test? How does it work?

Stage 1 screening is performed using a technology called Distortion Product Otoacoustic Emissions (DPOAE). Screening of hearing with DPOAE is usually quick, easy to do and reliable. It is completely objective. No behavioral response is needed from the baby, who ideally is tested asleep. Under good conditions, it takes less than two minutes to screen a baby, less than one minute per ear. The testing is done using a hand-held, automatic screening unit called the AudX. The test involves presenting sounds to the baby through a special probe that is gently inserted into the entrance of the ear canal. The normal inner ear actually generates tiny sounds in response to the stimulus, and these sounds are the otoacoustic emissions (OAE). A computer inside the screening unit distinguishes the OAE from ambient sound, which is always present in the baby's ear canal. Some ambient sound comes from the room where the test is done. Other sounds are generated if the baby or the probe moves during the test. The computer decides whether a real OAE is present, or just ambient sound, and then displays a 'pass' or 'refer' result.

3. What is an AABR screening test? How does it work?

The Auditory Brainstem Response is the response of a part of the brain (the brainstem) to sound. The ABR is a type of EEG which can be measured very early after birth. Screening of hearing with Automated ABR equipment is usually quick, easy to do and reliable. It is completely objective. No behavioral response is needed from the baby, who ideally is tested asleep. Under good conditions, it takes less than ten minutes to screen a baby. AABR screening is performed using a machine called an ABAER which consists of an automatic screening unit installed on a cart or on a table and a portable computer. Disposable skin electrodes are placed on the head and neck and soft sounds are presented to the baby through earphones. The ABAER computer extracts and interprets the EEG response to sound automatically. Typically, the test takes a total of 10 minutes or less.

4. What does it mean to “Pass” the screening?

‘Pass’ means that the computer detected a response to sound for all test frequencies at normal levels in both ears.

5. What does a “Refer” result mean?

‘Refer’ means that for any one of the test conditions (e.g. one frequency, one ear), a response could not be detected by the computer

6. What can cause a “Refer” result?

The most likely reason for not getting a pass result is that the baby’s activity level was too high or the ambient room noise was too loud. The ABR is a subtle response and can be hard to detect. Another possibility is that there was wax or debris in the ear canal,

blocking the probe. Or the baby might have a cold, which may cause a small hearing loss. There are many reasons not to get a pass result, so the parents can be reassured. However, it is certainly desirable, and in the baby's best interests, that they attend for a follow-up check.

7. If baby “Passes” the screening, what is the next step?

The results of the hearing screening indicate that your baby is able to hear normally in both ears at this time. The hearing screening is the first step. But you should continue to pay attention to how your baby responds to sounds and speech, and to how your baby’s language and speech develop. If at any time you become concerned about your baby’s hearing, contact the infant hearing centre closest to you.

8. If baby “Passes” the screening, and there are risk factors, what happens next?

The results of the hearing screening indicate that your baby is able to hear normally in both ears at this time. Even if a baby passes the hearing screening, there may be factors that cause the baby to be at greater risk for developing hearing problems. Some of these factors are:

- The baby had to stay in the hospital for intensive care for a few days before coming home
- There is someone in the immediate family who had a hearing problem from early childhood
- The baby’s development is different than other babies, or has facial characteristics that are linked to the development of hearing loss.

Because your baby has one of these factors, your baby will be re-screened periodically until reaching the age of 3 years.

9. If the result is “No Result” what does it mean?

“No Result” means that the screening test could not be completed for some equipment related reason. If the baby was too active and the noise levels are too high, the computer will not be able to recognize a response. There could be a problem with the probe, or probe fit, or with the computer software, although that would be rare. Whatever the reason, a “No Result” means that the screening will have to be repeated in order to determine whether the true result for that baby is “Pass” or “Refer”.

10. If there is a “Refer” result, what is the Stage 2 screen?

Parents usually want to know the results of the test right away. If there is a pass in both ears, there is usually no problem. If one or both ears give a refer result, then it is important to reassure the parent(s) that all this means is that a more detailed hearing check-up would be a good idea, preferably within a few weeks. The word 'fail' is never used in connection with a screening result, because it alarms parents unnecessarily and because it does not capture the true significance of not recording an OAE. If the test is done properly, less than about 5% of well babies will fail it. If the baby does indeed fail, what it means is that there is an increased *risk* of a hearing problem, but not that there *is* a hearing problem. The odds are that a baby who fails the screen will have normal hearing, but virtually ALL babies who DO have hearing loss will fail. Leading authorities

on hearing health consider the false-positive rate for this test to be a perfectly acceptable price to pay for the ability to detect all babies with hearing problems. This test is much more reliable than any possible test based on observing the baby's behavior. Behavioral screening tests are no longer recommended.

The fact that the screening test is obviously not behavioral means that in the event of a refer result it can be emphasized that this initial screening test does not actually measure true hearing. It measures 'echoes that come out of the baby's ear, and is a good test but certainly not perfect'.

The follow-up check is a stage 2 screen using a more advanced test, the AABR (automated auditory brainstem response), that measures the baby's brainwaves (EEG). The AABR test is also quick, painless and automatic, and is best done on a sleeping baby. It is more accurate than the DPOAE test, and the equipment is MUCH more expensive. It usually takes about 15 minutes, in a quiet baby. Most babies will pass this Stage 2 screen.

11. If the result is ”Refer” from Stage 2, the next step is an Audiology Assessment.

If a baby doesn’t pass the screen, the parents should be informed in a low-key manner that there will be a follow-up contact for an audiology assessment within a few weeks, to be done at a time convenient to them and provided the general health of the baby is satisfactory. The parents should be reassured that the pre-discharge test does NOT mean that the baby is deaf. There are many possible causes of screening failure, such as fluid in the middle ear that will resolve naturally in some cases or with medical management. Most infants who fail the pre-discharge screen will turn out to have normal hearing. However, the 'refer' screening result does indicate increased risk and it should be indicated that a hearing check-up would be a good idea after the acute-care stage is passed.

The Next Step:
An audiology assessment is another step in checking your baby’s hearing. It includes a number of tests to get a complete picture of how your baby hears. The assessment will be completed by an audiologist, a health care professional who specializes in hearing.

How is the Assessment Done?
While your baby is sleeping, special equipment is used to play soft sounds through earphones specially made for testing babies’ hearing. Your baby’s responses to sound are electronically recorded through the earpieces or through little wires attached by sticky pads on the head. Because more tests are done, an audiology assessment takes longer than the screening. It is possible you may be at the office for an hour, and may have to return for a second visit.

APPENDIX C

INFANT HEARING PROGRAM CO-ORDINATING SYSTEMS

1. Essex Preschool Speech and Language (PSL) System (Includes Windsor-Essex and Kent-Chatham PSL Systems)
   Windsor Regional Hospital
   1453 Prince Road
   Windsor, Ontario, N9C 3Z4
   Tel: (519) 252-0636 (general number)
   Infant Hearing Program: 519-254-5577 Ext. 52595
   Co-ordinator: Rose Grant Rennie: rgrennie@wrh.on.ca
   Tel: (519) 257-5100 x76555

2. Thames Valley PSL System (Includes Middlesex, London, Oxford, Elgin-St. Thomas (=Thames Valley PSL), Sarnia-Lambton, Huron-Perth and Grey Bruce Owen Sound PSL Systems)
   C/O Middlesex-London Health Unit
   50 King Street
   London, Ontario, N6A 5L7
   Tel: (519) 663-5317 ext. 2224 (general number)
   Co-ordinator: Debbie Shugar: dshugar@mlhu.on.ca
   Tel: (519) 663-5317 x2430
   Web Page: www.tyketalk.com

3. Hamilton-Wentworth PSL System (Includes Hamilton-Wentworth, Niagara, Brant and Haldimand-Norfolk PSL Systems)
   Hamilton Health Sciences Corporation
   C/O Affiliated Services for Children and Youth
   1171 Upper James Street, Unit 8
   Hamilton, Ontario, L9C 3B2
   Co-ordinator: Sue Honeyman: honeyman@binatech.on.ca
   Tel: (905) 381-2828 x235

4. Peel PSL System (Includes Peel, Halton, Waterloo, and Wellington-Dufferin PSL Systems)
   Erinoak Serving Children with Physical Disabilities
   2277 South Millway
   Mississauga, ON, L5M 2M5
   (905) 820-7111 (general number)
   Project Manager: Bridget Fewtrell
   Tel: (905) 820-7111

5. Toronto PSL System (Includes the City of Toronto only)

Toronto Public Health Unit
225 Duncan Mill Road, #201
Toronto, Ontario, M3B 3K9
Tel: (416) 338-8255 (general number) Fax: (416) 338-8511
Co-ordinator: Martha Cole: Tel: (416) 338-8626
Web Page: www.tpsls.on.ca

6. Simcoe County PSL System (Includes Simcoe County and Muskoka-Parry Sound PSL Systems)
   Royal Victoria Hospital of Barrie
   201 Georgian Drive
   Barrie, Ontario, L4M 6M2
   Tel: 705-739-5696 or 1-800-675-1979 (PSL/Infant Hearing/Children's Rehabilitation Services Intake)
   Co-ordinator: Mary Riggin Springstead: rigginm@rvh.on.ca
   Tel: (705) 728-9090 x4736

7. York Region PSL System (Includes York Region, Durham, Haliburton, Kawartha, Pine-Ridge PSL System)
   Markham Stouffville Hospital
   381 Church Street
   Markham, Ontario, L3P 7P3
   Tel: 1-888-703-5437 (general number)
   Co-ordinator: Sara Koke; skoke@msh.on.ca Tel: (905) 472-7373 x6070
   Web Page: www.Beyond-Words.org

8. Early Expressions Speech and Language Preschool Services (Includes Kingston, Frontenac, Lennox and Addington, Leeds, Grenville and Lanark and Hastings Prince Edward PSL Systems)
   Kingston, Frontenac, Lennox & Addington Health Unit
   221 Portsmouth Avenue
   Kingston, Ontario, K7M 1V5
   Tel: (613) 546-3854 or 1-800-267-7875
   Co-ordinator: Paula Varette Cerre
   Tel: (613) 544-3400 x 3628

9. Ottawa PSL System (Includes Ottawa, Renfrew County and District, and Eastern Ontario PSL Systems)
   Pinecrest-Queensway Health and Community Services
   1365 Richmond Road, 2nd Floor,
   Ottawa, Ontario, K2B 6R7
   Tel: (613) 724-4179 (parent information line)
   Co-ordinator: Suzanne Larocque: firstwords@pinecrest-queensway.com
   Tel: (613) 820-492

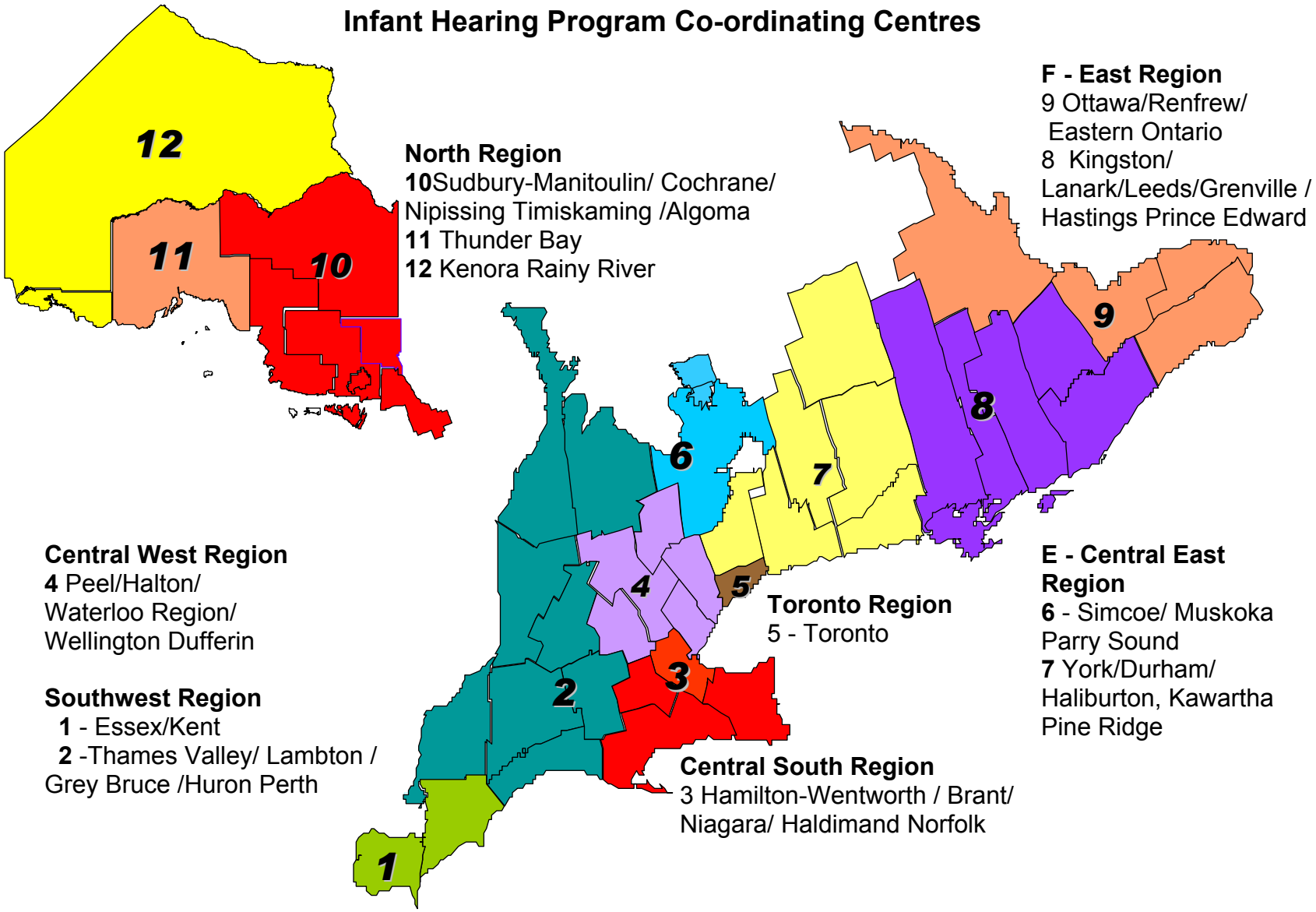
10. Manitoulin Sudbury PSL System (Includes Manitoulin-Sudbury, Algoma, Cochrane and Nipissing-Timiskaming PSL Systems)

Northeast Mental Health Centre
2041 Long Lake Road
Sudbury, Ontario, P3E 4M8
Tel: 1-877-522-6655 (general number)
Co-ordinator: Frank Demarco: fdemarco@networknorth.on.ca Tel: (705) 522-6655 x32

11. Thunder Bay PSL System (Includes Thunder Bay PSL only)
    Thunder Bay District Health Unit
    999 Balmoral Street
    Thunder Bay, Ontario, P7B 6E7
    Tel: 1-888-294-6630 (general number)
    Co-ordinator: Bill Campbell (807) 625-5949
    Web Page: www.tbdhu.com

12. Kenora Rainy River PSL System (Includes Kenora Rainy River only)
    Northwestern Health Unit
    396 Scott Street
    Fort Frances, Ontario, P9A 1G9
    Co-ordinator: Debby Cousineau: Tel: (807) 274-0709
    dcousineau@nwhu.on.ca
    Toll–free: 1-877-553-7122
    Web Page: www.northwords.com

Infant Hearing Program Co-ordinating Centres



North Region
10Sudbury-Manitoulin/ Cochrane/ Nipissing Timiskaming /Algoma
11 Thunder Bay
12 Kenora Rainy River

F - East Region
9 Ottawa/Renfrew/ Eastern Ontario
8 Kingston/ Lanark/Leeds/Grenville / Hastings Prince Edward

Central West Region
4 Peel/Halton/ Waterloo Region/ Wellington Dufferin

Southwest Region
1 - Essex/Kent
2 -Thames Valley/ Lambton / Grey Bruce /Huron Perth

Toronto Region
5 - Toronto

E - Central East Region
6 - Simcoe/ Muskoka Parry Sound
7 York/Durham/ Haliburton, Kawartha Pine Ridge

Central South Region
3 Hamilton-Wentworth / Brant/ Niagara/ Haldimand Norfolk