Blood Spot Screening for Hearing Loss Risk Factors

What is expanded hearing screening?
The expanded hearing screen includes two parts:

1. the hearing screen that happens in a hospital or community-based setting (performed by Ontario Infant Hearing Program hearing screeners) and
2. the hearing loss risk factor blood spot screen (performed by Newborn Screening Ontario).

The blood spot screen for hearing loss risk factors uses the same dried blood spot taken for the newborn blood spot screening program. The main risk factors are congenital cytomegalovirus (cCMV) infection and inherited predisposition to hearing loss. Children with these risk factors may be deaf at birth or may develop hearing loss early in life (before the age of 6 years).

The hearing loss risk factor blood spot screen will be performed with consent when there has been an Infant Hearing Program (IHP) recommendation that a baby see an audiologist for a hearing assessment. The hearing loss risk factor blood spot screen can detect congenital cytomegalovirus (cCMV) infections. Congenital CMV infections are the most common cause of non-genetic permanent hearing loss.

What is cytomegalovirus (CMV)?
CMV is a common virus. Most healthy people will not have any signs or symptoms and will not know they have had it. When a pregnant woman is infected there is a risk of infection of the baby. When this happens it is called congenital CMV infection.

What are the symptoms of cCMV infection in a baby?
A baby infected with CMV prenatally (before birth) may have no signs or symptoms (asymptomatic) or can be symptomatic at birth. Most babies (~85-90%) with cCMV infection will be asymptomatic. In babies who are symptomatic, hearing loss is a common symptom of cCMV infection and this is one of the reasons expanded hearing screening is offered. Some other symptoms of cCMV infection may include:

- Vision problems
- Seizures
- Jaundice
- Low platelets/ Rash from low platelets (petechial rash)
- Developmental disabilities
- Small head size
- Small size during pregnancy and at birth

What type of hearing loss can be caused by cCMV infection?
cCMV infection can cause permanent hearing loss that might be present at birth (congenital) or might develop in childhood. It can affect one or both ears. The hearing loss may affect only some sounds important for speech or all sounds important for speech. The hearing loss can be mild to profound. In some cases the hearing loss can worsen over time.
What can be done if cCMV infection is found through screening?
If cCMV infection is found through the hearing loss risk factor blood spot screen, the baby will be checked for symptoms. If symptoms are present, the baby will be followed by specialists and may be offered treatment. If they do not have symptoms at birth, will be offered ongoing appointments to check for hearing loss and problems with development during early childhood. The risk of developing a problem during early childhood is ~7-15%.

Why use the dried blood spot?
Testing for cCMV infections needs to be performed on a blood sample taken within the first 3 weeks of life. In most cases the dried blood spot is taken in the first week of life and sent to Newborn Screening Ontario (NSO) for newborn blood spot screening.

When will the hearing loss risk factor blood spot screen be performed?
The hearing loss risk factor blood spot screen will be performed with consent if there has been an IHP recommendation that the baby see an audiologist for a hearing assessment. If a baby passes their hearing screen, they will not be eligible for the hearing loss risk factor blood spot screen. Results from the hearing loss risk factor blood spot screen should be available within a week once NSO is notified to perform the screen. All results are sent to the IHP.

What happens if the baby’s results are positive (CMV detected)?
Screen positive results (CMV detected) from the hearing loss risk factor blood spot screen will be sent to the nearest Follow-Up Centre Infectious Diseases (ID) clinic and to the IHP. NSO or the ID clinic will contact families directly to talk about the screen positive results and arrange follow-up for the baby.

Can hearing screening be declined?
Yes. Hearing screening is not mandatory but is a medical recommendation. Parents/legal guardians can decline hearing screening. Parents/legal guardians can also decline expanded hearing screening if their baby is eligible for it.

If expanded hearing screening is declined, why does this get shared with NSO?
If the Infant Hearing Program has referred a baby to an Audiologist for a hearing assessment, the baby is eligible for expanded hearing screening. Parent/guardian decisions to consent or decline expanded hearing screening are documented on the hearing screening form and sent to NSO. This helps the IHP and NSO make sure that expanded hearing screening was offered to all eligible babies and monitor the rate of uptake.

For additional information about the hearing screen and for contact information, please visit: www.ontario.ca/infanthearing