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Practical Tips for Paediatricians
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Early hearing detection and intervention in Canada

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Early detection of and intervention for hearing loss can maximize communication, reading and emotional development, resulting in optimized education and employment in later life (1). While newborn hearing screening has been endorsed by the Canadian Paediatric Society (2), Canada lags behind other developed countries (3) on addressing the major public health challenge of congenital hearing loss. With this in mind, paediatricians are key to augmenting the efficacy of congenital or early-onset hearing loss detection and intervention, particularly in cases where infants fail or do not receive the screening test.

TIP #1—WHY IS HEARING IMPORTANT?

Auditory stimulation during the first 6 months of life is critical to speech and language development (4,5), such that unevaluated hearing loss often results in a language delay (6). Studies have shown that the long-term consequences of hearing loss include impaired social and emotional development resulting in lower academic achievement and greater psychological distress (6). Early hearing detection and intervention (EHDI) programs can optimize linguistic competence and literacy development for children who experience hearing loss. The effect that hearing loss will have on a child's speech and language is directly correlated with factors such as the time delay in diagnosis and intervention for hearing loss (7). Research from other developed countries has demonstrated that infants with hearing loss who receive timely intervention may perform 20 to 40 percentile points higher on school-related measures, such as vocabulary, articulation, intelligibility, social adjustment and behaviour, than those without early intervention (8). Further, screening may also identify infants with mild and unilateral hearing impairments. Delayed intervention for hearing impairment can have irreversible negative effects on language and overall cognitive development for the child.

TIP #2—CURRENT EHDI PROGRAMS IN CANADA

Components of a comprehensive EHDI program include 1) Hearing screening of all newborns by 1 month of age, 2) Identification of babies with permanent hearing loss (PHL) via comprehensive audiologic evaluation by 3 months of age, 3) Intervention services, including support for technology and communication development, by 6 months of age, 4) Family support and monitoring, and 5) Program evaluation (9). Furthermore, all infants with or without risk factors (Box 1) should receive ongoing language, literacy, and social emotional and developmental surveillance during well-child visits from 2 months of age.

The 2019 Report Card released by the Canadian Infant Hearing Task Force gave Canada an 'insufficient' grade because

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Table 1. Canadian early hearing and detection and intervention programs

Province/Territory	Grade (sufficient)	Newborns screened (%)	All EHDI program components*
Alberta	Yes	89+	Yes
British Columbia	Yes	97+	Yes
Manitoba	No	92+	No
New Brunswick	No	98+	No
Newfoundland and Labrador	No	Unknown	No
Northwest Territories	Yes	99+	Yes
Nova Scotia	Yes	96+	Yes
Nunavut	No	Unknown	No
Ontario	Yes	94+	Yes
Prince Edward Island	No	97+	No
Quebec	No	30+	No
Saskatchewan	No	Unknown	No
Yukon	Yes	99+	Yes

Adapted from ref. (2).

EHDI Early hearing detection and intervention.

*All EHDI program components aim to include 1) universal hearing screening of all newborns, 2) identification of babies with permanent hearing loss, 3) intervention services which include support for technology and communication development, 4) family support and monitoring and 5) evaluation of the program.

all provinces and territories did not achieve all components of the EHDI program (9). Current EHDI systems fall under the umbrella of the provincial governments and exist in all provinces and territories in Canada. However, there is a large degree of variability and inconsistency across the country due to inadequate screening, missing EHDI components, unimplemented clinical protocols, or deficient database to track infants or outcomes (9). Six provinces/territories were graded as 'sufficient' with excellent screening coverage whereby more than 95% of newborns are screened and all EHDI program components are in place. Provinces, such as Manitoba, New Brunswick, and P.E.I. have excellent screening coverage but do not provide complete or consistent EHDI program components. A few provinces and territories, including Saskatchewan, Nunavut, and Newfoundland and Labrador have unknown screening coverage and incomplete EHDI components (Table 1). Of note, expanded hearing screening for congenital cytomegalovirus (cCMV) infection and some common genetic risk factors for PHL (i.e., GJB2/6, SLC26A) may be offered by some provinces (10). The incidence of congenital hearing loss in Canada is about 1 to 3/1,000 live births, and the variability in screening across provinces means that in some areas of Canada children remain unscreened (6). This leads to an average age of diagnosis of hearing loss of about 24 months of age, whereas milder hearing loss is not detected until children become school age (6). The average age of diagnosis in screened populations is about 3 months of age, which is consistent with data from other developed countries such as the USA (6,11).

Common EHDI challenges include lack of follow-up of children who do not pass initial screening for confirmation of hearing loss or those who do not receive appropriate early intervention services. System-wide problems include the reported inability of current EHDI programs to provide culturally appropriate information and services, adequate data tracking systems, and expert resources to provide follow-up for infants referred from the hearing screen (1).

TIP #3—WHAT IS THE PAEDIATRICIAN'S ROLE?

Paediatricians are responsible for monitoring the general health, development and well-being of infants. They must have knowledge of provincial/territorial EHDI programs to ensure that infants who do not pass screening receive appropriate care. Furthermore, they should provide the developmental surveillance, as more than a third of infants with PHL will demonstrate developmental delays or other disabilities.

The patient-centred medical home with a team-based health care delivery model, with both the paediatrician and audiologist, can care for infants with proven or suspected PHL by:

- Confirming completion and reviewing the result of the newborn hearing screen,
- If newborn hearing screen is abnormal, reinforcing the need for early intervention, reviewing the result of the diagnostic evaluation, and advocating for early hearing amplification or family counselling if needed,

Box 1. Risk factors associated with permanent congenital, delayed onset or progressive hearing loss in childhood

1. Parental concern† for hearing, speech, language or developmental delay
2. Family history*,† of permanent childhood hearing loss
3. Neonatal indicators* (prolonged NICU stay > 5 days, hyperbilirubinemia requiring exchange transfusion, persistent pulmonary hypertension needing mechanical ventilation and conditions requiring use of ECMO†)
4. In utero infections* (CMV†, herpes, rubella, syphilis, and toxoplasmosis)
5. Craniofacial anomalies* (pinna or ear canal abnormalities)
6. Stigmata of syndrome known to have sensorineural* or conductive hearing loss or eustachian tube dysfunction (such as a white forelock)
7. Syndromes associated with progressive hearing loss† (neurofibromatosis, osteopetrosis, Usher syndrome, other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson)
8. Neurodegenerative disorders (Hunter syndrome, sensory motor neuropathies such as Friedreich ataxia and Charcot-Marie-Tooth disease)
9. Postnatal infection associated with sensorineural hearing loss† (bacterial and viral meningitis)
10. Head trauma (especially basal skull/temporal bone fracture that require hospitalization)
11. Chemotherapy†

Adapted from ref. (1).
ECMO Extracorporeal membrane oxygenation; NICU Neonatal intensive care unit.
*Risk factors for newborn sensorineural hearing loss. †Greater concern for delayed onset hearing loss. CMV, cytomegalovirus.

- Informing parents on hearing, speech and language milestones. In addition, informing parents on local multidisciplinary teams, that may include speech and language therapy, psychology, and social work,
- Ensuring referrals for genetic counselling and otolaryngology assessment,
- Diagnosing and aggressively treating middle ear disease,
- Referring for vision screening as appropriate,
- Specifically assessing balance and motor development, as nearly 50% of children with hearing loss may have associated vestibular impairment (12),
- Providing ongoing developmental surveillance and referral to resources,
- Identifying and referring for audiologic monitoring those infants at risk for late-onset hearing loss (1),
- Re-referring children to the infant hearing program when there are concerns, and
- Resourcing family and grief support following the diagnosis of infant's hearing loss.

Regardless of the newborn screening outcome, paediatricians should identify infants with medical or family risk indicators (Box 1) to monitor for delayed-onset or progressive hearing loss that will require audiological evaluation in early childhood. These include those infants who present with the following risk factors: screened positive for cCMV, had hyperbilirubinemia requiring exchange transfusion, received extracorporeal membrane oxygenation therapy in the neonatal period, or whose parents have any concern about language, hearing or developmental milestone attainment, among others. Furthermore, children with cochleovestibular anomalies and/or cochlear implants may be at increased risk of acquired bacterial meningitis from an otogenic source; this risk can be decreased by administration of both routine and specific pneumococcal immunization. Finally, paediatricians have to advocate at provincial and national levels for comprehensive EHDI systems.

CONCLUSION

Canada has received an 'insufficient' grade for addressing the major public health issue of congenital deafness (9). More newborns are being screened for hearing loss than previously. However, EHDI programs are not delivered consistently across provinces and territories due to inadequate screening, missing EHDI components, unimplemented clinical protocols, or deficient database to track infants or outcomes. Until the international gold standard of universal newborn hearing screening is realized in Canada, paediatricians may play an integral role in building and supporting EHDI systems that optimize communication, social and academic outcomes for children with PHL.

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