REVIEW

Permanent Childhood Hearing Impairment and the Universal Newborn Hearing Screening Program in British Columbia

Renée M. Janssen, MSc Aud[a]

[a]Vancouver Fraser Medical Program 2013, Faculty of Medicine, University of British Columbia, Vancouver, BC

ABSTRACT

Hearing loss is a relatively common congenital disorder, affecting an estimated one to three in every 1,000 live births. Untreated hearing loss in early childhood affects the development of speech and language, and later, academic success. Prior to universal newborn hearing screening, the age of identification of permanent congenital hearing loss in infants was quite delayed, due in part to the absence of risk factors in the majority children with congenital hearing loss. This article provides a brief overview of the causes and impact of permanent childhood hearing impairment, and introduces the universal newborn hearing screening program in British Columbia (BC). The BC Early Hearing Program, announced by the provincial government in March 2005, provides newborn hearing screening, diagnostic assessments, the first set of hearing devices, and ongoing communication and family support services.

KEYWORDS: *universal newborn hearing screening, hearing screening, hearing impairment, permanent childhood hearing impairment*

DEVELOPMENT OF SPEECH AND LANGUAGE, AND THE IMPACT OF HEARING LOSS

Infants tune in to the surrounding linguistic world early in development. They are able to discriminate subtle speech sounds at birth; by six to twelve months of age, they begin to show a decline in their ability to discriminate non-native speech sounds and an enhancement in their ability to discriminate native speech sounds.[1] Thus, infants develop a bias towards their native language from a very early age.

Congenital hearing loss affects the quality and quantity of these early listening experiences. Prelingual auditory deprivation leads to language delay, the severity of which correlates with the severity of the hearing loss. Children with hearing loss have poorer language measures than their normal-hearing peers; language outcomes in children with hearing loss are much poorer than would be predicted by their non-verbal intelligence.[2-5] Language skills are critical for attaining literacy as well as other cognitive skills; as such, deaf children have notably poorer academic success (for a review, see Helfand *et al.*[6]). Additionally, children with hearing loss exhibit more behavioural problems than their normal-hearing peers.[7, 8]

Correspondence
Renée M. Janssen, renee.janssen.king@gmail.com

PERMANENT CONGENITAL HEARING IMPAIRMENT

Hearing loss is a relatively common congenital disorder. Approximately one to three in every 1,000 infants are born with permanent hearing loss; the number increases to one in every 40 for infants who require care in the Neonatal Intensive Care Unit (NICU).[9-12] The causes of congenital hearing loss in infants include:

1. anatomic abnormalities of the ear (outer, middle and/or inner);
2. syndromal genetic disorders (e.g., Down Syndrome, Waardenburg syndrome);[13]
3. non-syndromal hereditary hearing loss (i.e., hearing loss in isolation) – patterns of inheritance include autosomal dominant, autosomal recessive, mitochondrial, and X-linked[13] (see the Hereditary Hearing Loss Homepage for an overview of the genetic causes of hearing loss[14]);
4. in utero infections (toxoplasmosis, rubella, cytomegalovirus, herpes, syphilis);[15] and
5. exposure to ototoxic drugs such as alcohol, cisplatinum, and isotretinoin during pregnancy.[15]

Risk factors for hearing loss include meningitis, hyperbilirubinemia requiring exchange transfusion, low birth weight, respiratory distress, prolonged mechanical ventilation, low Apgar scores, exposure to ototoxic medications such as gentamicin and loop diuretics, and family history of childhood hearing impairment.[16]

> **Universal newborn hearing screening is the best way to ensure that all children with significant congenital hearing loss will be identified early.**

Genetic causes account for over half of children with permanent congenital hearing loss.[13] Of these, approximately 75% have non–syndromal hereditary hearing loss, i.e., they have hearing loss in isolation, with no other associated medical issues.[17] At least half of non-syndromal hereditary hearing loss is associated with a defect in the Cx26 gene, which encodes a connexin gap junction protein.[18] This genetic defect displays a recessive pattern of inheritance.

IDENTIFICATION OF PERMANENT CHILDHOOD HEARING LOSS BEFORE UNIVERSAL SCREENING

Infants with hearing loss generally develop normal early communication skills such as eye contact, smiling, gesturing, and babbling.[19] For this reason, it is difficult to identify infants who have hearing loss based on behavioural observation alone.

Before the advent of universal newborn hearing screening, children were usually assessed for hearing loss based on whether they had known risk factors (such as a lengthy stay in the NICU, or a family history of hearing loss) or when there was parental concern. However, at least half of the children found to have permanent congenital hearing loss do not have risk factors.[20]

Historically, assessment of children based on the presence of risk factors led to diagnosis by approximately 12 months of age.[20] Children who had a severe or profound degree of hearing loss were typically identified earlier than those children with a lesser degree of hearing loss, as behaviours associated with a severe to profound loss are more apparent.[5] The average age of identification in children with mild to moderate degrees of hearing loss was approximately two years of age.[20]

Given that at least half of all children with permanent congenital hearing loss will have no apparent risk factor, and given the lack of early reliable behavioural indicators of hearing loss, targeted screening would miss many children with hearing loss. Universal newborn hearing screening is the best way to ensure that all children with significant congenital hearing loss will be identified early.

The U.S. Preventive Services Task Force recommends screening for hearing loss in all newborns (B recommendation[21]); with recent advances in hearing assessment techniques, this has become feasible. Universal newborn hearing screening programs are in place or under development in many countries such as the United States, the United Kingdom, Canada, Australia, and New Zealand.

THE BRITISH COLUMBIA EARLY HEARING PROGRAM (BCEHP)

The BC Early Hearing Program (BCEHP) is the first province-wide hearing screening program in British Columbia. The program was announced in March 2005 and was fully implemented across all health authorities by 2009. The program provides newborn hearing screening, follow-up diagnostic assessments, the first set of hearing aids, and ongoing communication and family support services. It is delivered by approximately 350 service providers and professionals in British Columbia's six regional health authorities.[22]

HEARING SCREENING IN THE NEWBORN

Most infants have their hearing screened before they are discharged from the hospital; for those who do not have screening completed by the time of discharge, the family is offered a follow-up appointment, usually at the local community public health audiology clinic.

Similar to universal newborn hearing screening programs in other countries, there are two screening methods in use in British Columbia:

1. Automated Otoacoustic Emissions: Otoacoustic emissions (OAEs), first characterized by Kemp in 1978, are sounds actively generated by the outer hair cells of the inner ear in response to incoming sound.[23] A probe is placed in the infant's ear, and soft clicks are presented. The probe automatically detects the presence or absence of the OAEs in response to the clicks. OAEs reflect the status of the peripheral auditory system (outer, middle, and inner ears).
2. Automated Auditory Brainstem Response: The auditory brainstem response (ABR) is generated by the synchronous firing of neurons in the auditory nerve and brainstem in response to sound.[24] Electrodes are placed on the forehead and nape of the neck, and sounds are presented to the ears via earphones. For the purpose of screening, response detection is automated. The ABR reflects the status of the peripheral auditory system and auditory neural pathway to the level of the brainstem.

Hearing screening is a two-stage process:

1. Infants who pass the first screening do not need any further testing. Sensitivity of hearing screening is estimated at 94%;[25] i.e. very few infants who have significant hearing loss will have a "pass" result.
2. Those who do not pass the first screening will have a second screening. This increases the specificity of the screening by reducing the number of false positives. The referral rate from first stage screening is approximately eight percent.[26]
3. Those who do not pass the second screening will be referred for a full diagnostic assessment with an audiologist. The referral rate to diagnostics from screening for the 2008/2009 fiscal year was two percent in BC, which is below the benchmark of four percent set by the American Academy of Pediatrics.[16,27]

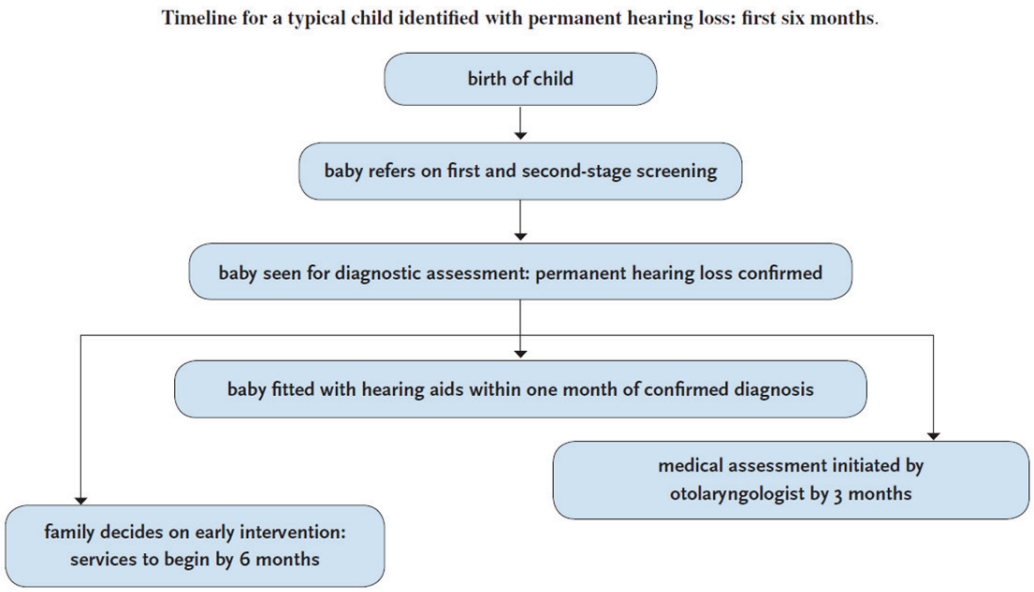

Figure 1. Timeline of the first six months of life for a typical child diagnosed with permanent congenital hearing loss through the BCEHP.

DIAGNOSIS AND MANAGEMENT OF CONGENITAL PERMANENT HEARING LOSS

Infants who do not pass screening are seen for a full diagnostic assessment with an audiologist at one to two months of age. Infants at this age are too young for reliable behavioural testing: the gold-standard method of infant hearing assessment is by diagnostic tone-evoked ABR in conjunction with other supportive tests.[16] The diagnostic tone ABR is very similar to the screening ABR, but does not employ automatic response detection, providing a much larger degree of diagnostic detail about hearing status. It is performed by an audiologist with training in infant assessment techniques. An infant is fit with electrodes and headphones, and the elicited ABR waveforms are interpreted by an audiologist. The infant must be asleep, as any muscle movement or tension creates myogenic artefacts that interfere with waveform interpretation. This test takes approximately one hour of sleep time (often less if hearing is normal).[28] When hearing loss is identified, a second appointment is often required to obtain more information.[28] In the case of normal hearing it is sufficient to determine that hearing sensitivity is within the normal range; however, when there is hearing loss present, more ABR waveforms must be obtained to accurately characterize the hearing loss for optimal habilitation with amplification devices such as hearing aids.

Once the hearing loss has been confirmed and quantified, the child requires a medical evaluation by an otolaryngologist. The purpose of the medical evaluation is to investigate the etiology of the hearing loss, to evaluate for other complications or concomitant disorders, and to prevent/manage future deterioration of the hearing loss. Investigation of etiology involves a thorough history and physical examination, imaging of the ear, and targeted genetic testing.[29] At this time, the only genetic screening available in British Columbia for non-syndromic hereditary hearing loss is for the Cx26 gene, as discussed above. A full review of the BCEHP guidelines for the medical management of children newly diagnosed with permanent hearing loss is available on the BCEHP website.[22]

Additionally, the child will receive a fitting with hearing aid(s) (if appropriate), and the family will enrol in an early intervention program to aid in the development of speech, listening, and language skills. The first set of hearing aids, including batteries and earmolds, are provided by the BCEHP at no cost to the family. For more information about Early Intervention and the Hearing Aid Program, please see the BCEHP website.[22]

Figure 1 shows the timeline of events from birth through the first six months of a child identified with permanent congenital hearing loss through the BCEHP. The BCEHP protocol, which requires screening by one month, diagnosis of hearing loss by three months, and early intervention (including hearing devices) by six months of age, is in line with the Joint Committee on Infant Hearing recommendations.[16]

THE ROLE OF THE FAMILY PHYSICIAN/ PAEDIATRICIAN

Identification of permanent hearing loss in a newborn is often a time of intense emotions for the family.[30] While children who are identified with hearing loss, fitted with hearing aids, and enrolled in an early intervention program by age six months have a good prognosis for speech and language development, it is still a difficult time of adjustment for the family, and family-centred support is important during this period.[31,32] Good interdisciplinary communication between the audiologist and the primary care physician is critical to ensure families receive consistent information.

It is important to be aware that not all permanent childhood hearing loss is present at birth; some children will develop hearing loss later in childhood. Within the BCEHP, children who pass screening at birth but have risk factors for progressive hearing loss* are monitored until three years of age. However, as discussed, not all children with hearing loss have identifiable risk

*Risk factor criteria for BCEHP monitoring[27] are: craniofacial anomalies (not ear pits or tags); close relative (parent, sibling, uncle, aunt, cousin, grandparent) who had permanent hearing loss before 12 years of age; syndrome associated with late onset/progressive hearing loss; birthweight less than 1200 grams; five-minute Apgar score less than or equal to 3; hypoxic-ischemic encephalopathy (HIE) moderate/severe (Sarnat II or III); congenital diaphragmatic hernia (CDH); extra-corporeal membrane oxygenation (ECMO), or inhaled Nitrous Oxide (iNO) or High-Frequency Oscillatory (HFO) or Jet (HFJ) ventilation; intra-ventricular hemorrhage (IVH), Grade III or IV; peri-ventricular leukomalacia (PVL); hyperbilirubinemia > 400 µmol or meeting any standard criteria for exchange; perinatal (in the baby) TORCHS infection (toxoplasmosis, rubella, cyto-megalovirus (CMV), herpes, syphilis); meningitis, irrespective of the pathogen; and accidental overdose of gentamicin or other aminoglycosides, five-fold or greater.

factors. If there is a speech-language delay, or parental concern about hearing, then the child should be referred for a hearing assessment at the nearest public health audiology clinic regardless of screening results or the absence of risk factors.

The American Academy of Pediatrics[16] outlines the following roles for the pediatrician/primary care physician:

1. Monitor the general health, development, and well-being of the infant, including developmental milestones;
2. Ensure that families follow through with hearing screening and diagnostic audiology appointments;
3. Initiate medical referrals for medical specialty evaluations for the determination of etiology of the hearing loss;[#]
4. Monitor middle ear status for temporary outer/middle ear pathology such as otitis media, as this can impede screening and diagnostic testing, or add to the degree of loss in the case of permanent hearing loss;
5. Review the infant's medical and family history for the presence of risk factors that require monitoring for progressive or delayed-onset hearing loss;
6. Attend to parental concerns regarding speech and language development, hearing, and other developmental milestones.

The BCEHP is quite new, and permanent childhood hearing impairment, while not rare, is not a common diagnosis; for these reasons, primary care physicians may not be aware of the BCEHP, and may not know how to communicate with families who have just had their child diagnosed with hearing loss. The BCEHP website has information for physicians about hearing screening, diagnostic assessment of hearing loss, early intervention programs, and the BCEHP guidelines for medical management of children newly diagnosed with hearing loss.[22]

REFERENCES

1. Polka L., Werker J. Developmental changes in perception of nonnative vowel contrasts. J Exp Psychol [Hum Percept]. 1994;20(2):421-435.
2. Moeller MP. Early intervention and language development in children who are deaf and hard of hearing. Pediatrics. 2000;106:E43.
3. Davis JM, Elfenbein JL, Schum R, Bentler R. Effects of mild and moderate hearing impairments on language, educational, and psychosocial behavior of children. J Speech Hear Disord. 1986 Feb;51:53-62.
4. Elfenbein JL, Hardin-Jones MA, Davis JM. Oral communication skills of children who are hard of hearing. J Speech Hear Res. 1994;37:216-26.
5. Wake M, Poulakis Z, Hughes EK, Carey–Sargeant C, Rickards FW. Hearing impairment: A population study of age at diagnosis, severity, and language outcomes at 7-8 years. Arch Dis Child. 2005;90(3):238-44.
6. Helfand M, Thompson D, Davis R, McPhillips H, Homer C, Lieu T. Newborn hearing screening. Systematic evidence review number 5. Rockville (MD): Agency for Healthcare Research and Quality; 2001. AHRQ Publication No. 02-S001. Contract No.: 290-97-0018to the Oregon Health & Science University Evidence-based Practice Center, Portland, Oregon.
7. Davis A, Hind S. The impact of hearing impairment: a global health problem. Int J Pediatr Otorhinolaryngol. 1999;49(suppl 1):S51-S54.
8. Vostanis P, Hayes M, Feu MD, Warren J. Detection of behavioural and emotional problems in deaf children and adolescents: comparison of two rating scales. Child Care Health Dev. 1997; 23(3):233-46.
9. Davis A, Wood S. The epidemiology of childhood hearing impairment: factors relevant to planning of services. Br J Audiol. 1992 Apr:26,77-90.
10. Prieve BA, Stevens F. The New York State newborn hearing screening demonstrations project: introduction and overview. Ear Hear. 2000;21:85-91.
11. Wessex Universal Hearing Screening Trial Group. Controlled trial of universal neonatal screening for early identification of permanent childhood hearing impairment. Lancet. 1998;f52:1957-64.
12. Hille E, van Straaten H, Verkerk P. Prevalence and independent risk factors for hearing loss in NICU infants. Acta Paediatr. 2007;96:1155-8.
13. Toriello H, Reardon W, Gorlin R, editors. Hereditary Hearing Loss and its Syndromes. 2nd ed. New York: Oxford University Press; 2004.
14. Van Camp G, Smith RJH. Hereditary hearing loss [Internet]. [cited 2010 Feb 26]; Available from: http://webhost.ua.ac.be/hhh/.
15. Gifford KA, Homes MG, Bernstein HH. Hearing loss in children. Pediatr Rev. 2009;30(6):207-15.
16. Joint Committee on Infant Hearing. Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. Pediatrics 2007;120:898-921.
17. Morton NE. Genetic epidemiology of hearing impairment. Ann N Y Acad Sci. 1991;630:16-31.
18. Denoyelle F, Lina-Granade G, Plauchu H, Bruzzone R, Chaib H, Levi-Acobas F, *et al.* Connexin26 gene linked to a dominant deafness. Nature. 1998;393:319-20.
19. Oller DK, Eilers RE. The role of audition in infant babbling. Child Dev. 1988;59(2):441-9.
20. Harrison M, Roush J. Age of suspicion, identification, and intervention for infants and young children with hearing loss: a national study. Ear Hear. 1996;17(1):55-62.
21. U.S. Preventive Services Task Force. Universal screening for hearing loss in newborns: US Preventive Services Task Force recommendation statement. Pediatrics. 2008;122:143-8.
22. B.C. Early Hearing Program [homepage on the Internet]. [updated 2009 Sept 29; cited 2010 Feb 26]; Available from: http://www.phsa.ca/AgenciesAndServices/Services/BCEarlyHearing/default.htm.
23. Kemp DT. Stimulated acoustic emissions from within the human auditory system. J Acoust Soc Am. 1978;64:1386-91.
24. Sininger YS. The use of auditory brainstem response in screening for hearing loss and audiometric threshold prediction. In: Burkard RF, Don M, Eggermont JJ, editors. Auditory Evoked Potentials. Basic Principles and Clinical Application. Philadelphia: Lippincott, Williams & Wilkins; 2007. pp. 254-74.
25. Kennedy CR, McCann DC. 7 to 9 year outcome of the Wessex Controlled Trial of Universal Neonatal Hearing Screening (UNHS). Arch Dis Child. 2003;88:S1, A3.
26. Watkin PM. Neonatal screening – methods and outcome. Audioll Med. 2003;1:165-74.
27. B.C. Early Program Information Update [Internet]. Fall 2009 [cited 2010 April 4]; Available from: http://www.phsa.ca/AgenciesAndServices/Services/BCEarlyHearing/ForPhysicians/default.htm.
28. Janssen RM, Stapells DR. The British Columbia's Children's Hospital tone–evoked ABR protocol: how long do infants sleep, and how much information can be obtained in one appointment? Ear Hear. 2010 May [Epub ahead of print];31(5).
29. Morzaria S, Westerberg BD, Kozak FK. Evidence–based algorithm for the evaluation of a child with bilateral sensorineural hearing loss. J Otolaryngol. 2005;34(5):297-303.
30. Wood Jackson C, Randi J, Traub RJ, Turnbull AP. Parents' experiences with childhood deafness: implications for family–centered services. Communication Disorders Quarterly. 2008;29:82-98.
31. Kennedy CR, McCann DC, Campbell MJ, Law CM, Mullee M, Petrou S, *et al.* Language ability after early detection of permanent childhood hearing impairment. N Engl J Med. 2006;354:2131-41.
32. Meadow–Orlans KP, Mertens DM, Sass-Lehrer MA, Scott-Olson K. Support services for parents and their children who are deaf or hard of hearing: a national survey. Am Ann Deaf. 1997 Oct ;142(4):278-88.

[#]The BCEHP has an expedited medical referral process whereby designated otolaryngologists will accept referrals and ensure prompt medical evaluation of the child newly diagnosed with hearing loss. Please see the BCEHP website www.phsa.ca/earlyhearing for more information.