BC Early Hearing Program

A service of BC Children's Hospital and the Provincial Health Services Authority

Medical Management of Infants and Young Children with Sensorineural Hearing Loss

Developed April 12, 2007

PURPOSE OF GUIDELINES:

The purpose of these guidelines is to assist physicians in the management of infants and young children newly diagnosed with sensorineural hearing loss through the B.C. Early Hearing Program (BCEHP). These guidelines are anticipated to be informational only, and are not intended or designed to substitute reasonable exercise of independent clinical judgment by physicians and medical providers. Guidelines directed towards infants or children newly diagnosed with sensorineural hearing loss (SNHL) are presented. Structural conductive impairments can arise from maldevelopment of the external or middle ears. The medical management of these children with the resultant conductive hearing loss (CHL) is beyond the scope of these guidelines.

DEFINITION OF SENSORINEURAL HEARING LOSS:

For the purpose of the Early Hearing Program, a significant SNHL has been defined as thresholds greater than 40 dB in the better hearing ear (i.e. bilateral SNHL). The prevalence of hearing impairment diagnosed may triple if the severity threshold is changed from 40 dB to 20 dB, and if unilateral hearing loss is included. However, current hearing screening technology limits the detection of hearing loss that is less severe than 40 dB. The appropriate evaluation of children diagnosed with unilateral SNHL requires further research and is not addressed by these guidelines. However, there may be a significant risk of later acquired hearing loss in the previously normal contralateral ear in children with unilateral SNHL. Regular audiologic monitoring during the first 5 years of life to detect possible progression to bilateral hearing loss should be instituted until further research clarifies the frequency with which this may occur.

CHARACTERISTICS OF MEDICAL PROVIDERS:

Children who have been diagnosed with sensorineural hearing loss require adequate initial medical assessment as well as knowledgeable follow-up by their pediatrician/family practitioner and an otolaryngologist. Physicians who provide these services must exhibit the following characteristics:

- *Have comprehensive and current knowledge of the etiology of sensorineural hearing loss in infants.*

- *Have the appropriate level of competence in determining the evidence-based investigations required and in explaining the medical investigations to parents, including why they are needed and what the results mean for their child.*

- *Be able to provide family and child oriented care in a timely and accessible manner*

- *Be able to coordinate care with other health care providers.*

Physicians that care for children with sensorineural hearing loss must ensure they possess these characteristics and manage children with SNHL on a frequent enough basis to be thoroughly familiar with the tasks involved. If they do not, they are obligated to ensure the appropriate referrals are in place to physicians who do possess them.

CHARACTERISTICS OF MEDICAL GUIDELINES:

In general, the goals of the medical management of children with SNHL are to:

- *Diagnose the etiology of SNHL*

  The BCEHP medical management guidelines reflect an evidence-based approach to determining the etiology of bilateral SNHL in children while limiting the risk to the child and providing services in the least invasive and cost efficient manner possible. Medical investigations should be completed as soon as possible, preferably within three months depending on parental readiness and the health of the child.

- *Evaluate for other complications and/or concomitant disorders*

  A thorough history and physical exam by a clinician familiar with the etiologies of SNHL in infants and children is essential and may aid in the diagnosis of hearing loss. Familiarity with conditions and syndromes associated with SNHL in children is essential.

- *Predict future progression of the hearing loss if possible*

  After the initial completed workup, children with bilateral SNHL should have a medical review in 3 years, barring other clinical concerns, by an otolaryngologist with expertise in childhood hearing loss. Medical management is an ongoing process as some conditions take years to manifest.

- *Prevent and/or manage further deterioration of hearing*

  A certain proportion of children will have progression of their SNHL. Those who have a precipitous progression require urgent review and those with an otherwise slow undetected progressive course found on follow-up audiologic examination warrant less urgent review. Children with SNHL may have additional conductive hearing loss and require close monitoring by audiologists and their pediatrician/family practitioner and/or otolaryngologist to optimize management of the child's hearing loss.

- *Provide information for family members*

  Parents or caregivers are to be provided adequate information in oral and/or written format so they can understand the advantages and disadvantages of performing investigations. Management options need to be discussed highlighting the implications for the possible outcomes and prognosis of their child's hearing loss.

ALGORITHM FOR MEDICAL EVALUATION OF INFANTS AND CHILDREN WITH SNHL

Bilateral SNHL

Evaluation of a child newly diagnosed with bilateral SNHL should include a stepwise approach to determine the etiology of the hearing loss. Once a cause for the hearing loss is determined at any given stage, further investigation is usually not warranted.

A. General History:

A thorough history should be predicated on the age of the child at time of diagnosis, but may include details of the following:

- maternal health in prenatal period, including substance exposure
- details of peri- and postnatal period including admission to a Neonatal Intensive Care Unit (NICU)
- attainment of developmental milestones
- ototoxic medication exposure
- head injury
- ear disease
- meningitis
- congenital viral illness (i.e. toxoplasmosis, rubella, cytomegalovirus, herpes simplex virus)
- immunization status
- family history of hearing loss, and
- risk factors for Auditory Neuropathy Spectrum Disorder (ANSD).

The issue of Auditory Neuropathy Spectrum Disorder warrants specific discussion as our understanding of this condition continues to evolve. There appear to be multiple risk factors for ANSD, the most common of which are:

a. Anoxia or hypoxia at birth
b. Hyperbilirubinemia requiring exchange transfusion during the newborn period
c. Infectious disease (such as meningitis or mumps)
d. Immune disorders (e.g. Guillain-Barré Syndrome)
e. Neurological and Genetic disorders

B. *Physical examination to detect stigmata associated with syndromic hearing loss*.

A full examination of the child is essential, assessing for the presence of conditions or malformations that may be associated with SNHL and aid in the determination of the etiology, i.e. preauricular pits, branchial sinuses.

C. Audiograms for siblings of child with hearing loss.

Audiometric evaluation of siblings of children with SNHL may be warranted to determine normal hearing is present. This evaluation may also assist with determination of a possible genetic cause for the hearing loss.

D. Audiological assessment for Auditory Neuropathy Spectrum Disorder if risk factors present.

Assessment for ANSD may include:

a. Ear specific tone pip ABR, both air and bone conduction.
b. Ear specific click ABR, depending on results of tone pip ABR
c. 95 dB nHL records are assessed for the present of cochlear microphonics and stimulus artifacts.
d. Diagnostic Distortion Product or Transient Otoacoustic Emission
e. Otoscopy and tympanometry (both ears)

This testing should be performed by an audiologist familiar with assessment of young children and familiar with the condition of ANSD.

E. Targeted genetic testing

Connexin 26 (Cx26) is currently the only genetic mutation generally available for screening purposes in British Columbia. Evaluation for other genetic mutations will likely become more widely available in the future. A genetics consultation should be considered for all families with a child with SNHL.

F. Ophthalmologic and other specialist assessment

All children with SNHL should have ophthalmologic assessment by an Ophthalmologist familiar with associated eye syndromes to identify concurrent visual abnormalities (e.g. strabismus, myopia, amblyopia), to maximize visual acuity and to identify ocular findings associated with congenital syndromes coupled with SNHL. Additional referrals are warranted only if indicated by findings on history and/or physical examination.

G. Electrocardiogram (ECG)

An ECG is indicated for any children with:

- SNHL greater than 70 dB (severe)
- a history of syncopal episodes or arrhythmia
- a family history of childhood death

If no cause identified after above:

H. High-resolution CT scan of temporal bones

A CT scan is not always needed to evaluate a child with SNHL i.e. a child identified with homozygous mutations for Cx26 is unlikely to have malformations identified on a CT scan.

A CT scan should be considered only in consultation with an Otolaryngologist skilled in the management of children with SNHL. The CT scan should be performed in a tertiary care centre experienced in radiologic evaluation of children such that the appropriate and adequate study is performed and children are not exposed unnecessarily to radiation.

Note: There are currently no recommendations surrounding routine serologic screening for infectious etiologies at this time (CMV, Rubella, Herpes and Toxoplasmosis). This is an area requiring further research. However, children identified as having congenital CMV infection must be followed with hearing screening regularly for a minimum period of five years to identify in a timely manner the possible development of delayed SNHL.

References:

1. Colorado Infant Hearing Advisory Committee: Guidelines for Infant Hearing Screening, Audiologic Assessment and Early Intervention. Submitted to the Colorado Board of Health. By the Division of Prevention of Public Health and Environment. December 14, 2000. Revision Aug 4, 2004.
2. Medical Management of Infants with Significant Congenital Hearing Loss Identified through the National Newborn Hearing Screening Programme – Best Practice Guidelines. 10/20/2004
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4. Sinninger, Yvonne and Starr, Arnold. 2001. Auditory Neuropathy/Auditory Dyssynchrony: A New Perspective on Hearing Disorders.
5. Canadian Working Group on Childhood Hearing (CWGCH) Resource Document, http://www.phac-aspc.gc.ca/publicat/eh-dp/index.html, accessed February 11, 2007.
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