Introduction

Undetected hearing loss puts children at risk for significant developmental delay. Early identification and early intervention may decrease those delays. Primary care providers need to be alert for signs that children are not hearing well and make appropriate referrals for evaluation. Newborn hearing screening is performed to identify children who may have a congenital hearing loss and can benefit from early intervention. Primary care providers should obtain the newborn hearing screening results from the birth facility for all infants in their care. If the initial screen was abnormal, timely follow-up is essential to distinguish between children with false positive screening results and children with a hearing loss.

The infant's primary health care provider is responsible for:

- Assuming responsibility to ensure that the audiological assessment is conducted on infants who do not pass screening and for coordinating appropriate follow-up for children with confirmed hearing loss.
- Initiating referrals for medical specialty evaluations necessary to determine the etiology of the hearing loss and to detect and treat any associated health care problems.
- Partnering with other specialists, including the otolaryngologist, to facilitate coordinated care and ongoing relay of information between primary care, specialty care consultants, and the MDH for children with a hearing loss.
- Monitoring middle-ear status.
• Assuring that children identified with hearing loss receive appropriate referrals and that their parents are made aware of the range of medical, educational, and communication options available.

• Initiating referrals to early intervention services and monitoring developmental milestones to track the effectiveness of the intervention and the need for additional services.

• Coordinating the follow-up process with the family.

• Reviewing every infant's medical and family history for the presence of risk indicators that require monitoring for delayed-onset or progressive hearing loss and ensuring that an audiological evaluation is completed for children at increased risk of hearing loss at least once by 24 to 30 months of age, regardless of their newborn screening results. Infants with specific risk factors, such as those who received ECMO therapy and those with CMV infection have higher risk of delayed-onset or progressive hearing loss and should be monitored closely.

• Assuming responsibility for ongoing surveillance of parental concerns about language and hearing, auditory skills, and developmental milestones of all infants and children regardless of risk status, as outlined in the pediatric periodicity schedule published by the American Academy of Pediatrics (AAP).

**When the newborn hearing screen results are NORMAL**

Though infants who pass the newborn hearing screen or re-screenings can be assumed to have normal hearing, false negatives can occur. Moreover, even with normal newborn hearing screening results, there is no assurance that a child will continue to have normal hearing. Therefore, all infants should be monitored for late onset or progressive hearing loss. Parental concern about speech and language delays, at any time in a child's life, should receive prompt referral for an audiologic evaluation. Hearing testing can be performed at any age.

The Joint Committee on Infant Hearing (JCIH) 2007 Position Statement: *Principles and Guidelines for Early Hearing Detection and Intervention Programs* states that the following indicators can be associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood. Any infant with one of these risk indicators for progressive or delayed-onset hearing loss that has passed the newborn screen should, nonetheless, have at least one diagnostic audiology assessment by 24 to 30 months of age. These indicators are:

• Caregiver concern regarding hearing, speech, language, or developmental delay.

• Family history of permanent childhood hearing loss.

• Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that required exchange transfusion.
• In utero infections, such as CMV, herpes, rubella, syphilis, and toxoplasmosis.
• Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
• Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
• Syndromes associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other less frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
• Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
• Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
• Head trauma, especially basilar skull/temporal bone fracture that requires hospitalization.
• Chemotherapy.

Because some important indicators, such as family history of hearing loss, may not be ascertained at birth, the presence of any late-onset risk indicators should be determined by the primary care provider during early well-child visits. Those infants with late-onset risk factors should be carefully monitored for normal language milestones during routine medical care.

When there is a MISSED SCREEN

Primary care providers should make sure they have a report of the newborn hearing screen results for each child they see. If there is not a report on the chart, the hospital where the infant was born should be contacted. If it becomes clear that no screen was performed at birth, arrangements for screening should be made as soon as possible. Children born at home, in other non-traditional settings or in other countries may not have been screened. The provider should arrange to screen the hearing of these children as soon as possible.

• Minnesota Session Laws 2007 - Chapter 147 [144.966] EARLY HEARING DETECTION AND INTERVENTION PROGRAM requires “The hospital that discharges the newborn or infant to home is responsible for the screening;” It is important for the primary care physician to know the hospital policies and procedures for recalling and rescreening infants who miss a screen. Vigilance in checking the screening results for children who have been transferred from one hospital to another is essential.
• Screening is best performed within the first month of life; later ages require more time due to the infant’s increased alertness and may even require sedation in infants older than four months of age.

Minnesota law allows parents to decline or “opt-out” of newborn hearing screening. The hospital should document the refusal in the medical record and have the appropriate opt-out form signed by the parents and sent to the MDH Newborn Screening program and the primary provider. Parents who opt-out of newborn hearing screening in the hospital should be given information regarding the importance of early identification and the opportunity to obtain hearing screening on an outpatient basis. Providers should be vigilant for signs of language delay and other indications of hearing loss in unscreened babies.

When there is an ABNORMAL SCREEN, either Unilateral or Bilateral

Clinicians should be familiar with the newborn hearing screening protocol at the hospitals where their patients are born so that they can best assure prompt follow-up of abnormal results.

• The hospital should send final screening results to the clinic where the family has indicated follow-up will occur.

• All newborns with abnormal screens must be rescreened or referred for an audiological evaluation. For the most reliable results, rescreening should be performed between one and two weeks of age.

• The abnormal result of the screen should be shared with the family before hospital discharge and a rescreen appointment scheduled.

• Determine the person or department responsible for the following:
  
  o Scheduling appointment for recheck.
  o Recalling no-shows; physician involvement may be required.
  o Completing all rescreening within four weeks.
  o Written notification to primary care physician and MDH.
  o Continue recall attempt.
  o Record hearing results in the child’s medical record.

• No more than two rescreens should be performed. If both screens were abnormal, the infant should be referred for comprehensive audioligic testing. All hearing screens and rescreens should be performed prior to one month of age.

• Audiological confirmation diagnosis should be obtained no later than three months of age. For infants who are premature or require intensive care, testing
should be complete by three months corrected age or as soon as medically feasible for assessment/diagnosis.

Although persistent middle ear effusion necessitates medical referral, which might delay the evaluation timeline several weeks, diagnostic audiological evaluation must not be repeatedly postponed solely due to middle ear dysfunction and should be completed before three months of age. The information from the diagnostic audiological evaluation is valuable both in determining the extent of the effect of the middle ear condition on the infant’s hearing, and identifying whether an underlying sensorineural hearing loss also exists, thereby impacting the course of both medical and educational intervention.

When the child is subsequently confirmed by an audiologist to have a hearing loss not determined to be permanent (e.g. mild-moderate conductive from middle ear fluid or with unknown etiology);

Primary care provider initiates and supervises evaluation, provides follow-up care and the option of referral to Otolaryngology (ENT), following national guidelines for otitis media with effusion: [http://aappolicy.aappublications.org/cgi/reprint/pediatrics;113/5/1412.pdf](http://aappolicy.aappublications.org/cgi/reprint/pediatrics;113/5/1412.pdf)

Summary of the CLINICAL PRACTICE GUIDELINE to manage otitis media with effusion (OME) in children aged 2 months through 12 years:

- Use pneumatic otoscopy as the primary diagnostic method and distinguish OME from acute otitis media.

- Document the laterality, duration of effusion, presence, severity of associated symptoms at each assessment of the child with OME, specific reason for referral (evaluation, surgery) and provide additional relevant information such as history of acute otitis media and developmental status of the child.

- Distinguish the child with OME who is at risk for speech, language, or learning problems from other children with OME and more promptly evaluate hearing, speech, language, and need for intervention in children at risk.

- Manage the child with OME who is not at risk with watchful waiting for 3 months from the date of effusion onset (if known) or diagnosis (if onset is unknown).

- Conduct hearing testing when OME persists for 3 months or longer or at any time that language delay, learning problems, or a significant hearing loss is suspected in a child with OME.

- Reexamine children with persistent OME who are not at risk at 3- to 6-month intervals until the effusion is no longer present, significant hearing loss is identified, or structural abnormalities of the eardrum or middle ear are suspected.
Refer to the current Minnesota Department of Health (MDH) Infant Assessment and Pediatric Amplification Guidelines at www.health.state.mn.us/newbornscreening for more information.

When the child is subsequently confirmed by an audiologist to have a permanent hearing loss;

Primary care provider initiates referrals upon diagnosis and supervises evaluation and the referral process to the following specialists; Otolaryngology (ENT), Ophthalmology, Genetics, Audiology, Speech and Language Pathology and early intervention programs. The clinician should assure that appropriate evaluations are completed by the specialists including documentation of the family and prenatal history in addition to laboratory and physical examinations with as little duplication as possible. The primary care provider is in a unique position to balance the need for timely and appropriate evaluation with the family’s need to bond with their infant and adjust to the diagnosis.

History: The following elements of the child’s history should be documented:

Prenatal

- Ototoxic medication exposure
- Any significant complications during pregnancy
- Immunization to Rubella infection
- Positive FTA-ABS or other positive syphilis confirmation
- Maternal drug use
- Multiple miscarriages

Perinatal

High Risk Factors, as listed in Appendix A of this document.

Family

Other family members with hearing loss with onset before age 30
Family history of syndromes or genetic disorders

Systematic Physical Examination, including the head, neck and extremities:

- Minor anomalies: Unusual morphologic features occurring in less than 5% of the population with no cosmetic or functional significance (e.g. clinodactyly, skin tags, transverse palmar crease)
- Major anomalies: Those causing cosmetic and/or functional abnormality (e.g. cleft palate, cardiac, limb, or skeletal deformities)
• Poor growth and/or microcephaly
• Abnormal neurologic examination
• Abnormal ear exam (external ear anomalies, stenotic canal, middle ear fluid)

**Laboratory Studies:**
Infectious Diseases: rule out *in utero* infections with agents including CMV, Rubella, Syphilis, Toxoplasmosis
CMV urine culture: CMV Titer, done at 0 to 3 weeks of age

Upon diagnosis of a permanent hearing loss:
• All infants should have an Otolaryngology evaluation
• All infants should have a Genetics evaluation even if there appears to be a clear non-genetic cause of hearing loss.
• All infants should have an Ophthalmologic evaluation.

**Otolaryngology (ENT) Evaluation of Children with a Confirmed Hearing Loss:**

The ENT physician should have expertise in childhood hearing loss and is responsible for investigating the etiology of hearing loss and for determining whether medical or surgical intervention may be an appropriate option. In addition, the ENT physician provides information and participates in the assessment of the options for amplification, assistive listening devices, and cochlear implantation. The ENT physician should participate in the long-term monitoring of the hearing in partnership with the primary care team.

**History:**

Prenatal, Perinatal, Family and Behavioral
Review of prior audiomeric testing

**Physical Exam:**

Complete head and neck examination

**Diagnostic testing:**

Review of audiometric tests
ABR for threshold (if not previously done)
Otoacoustic emissions (if not previously done)
Behavioral audiogram, when appropriate
Radiologic Studies:

CT or MRI with detailed images of the temporal bone

Other tests:
Depending on previous testing and consultations the following tests will be considered: ECG (prolonged Q-T syndrome) for children with sensorineural hearing loss, urinalysis, and CMV testing by urinalysis and blood sample, if not already performed. Additional testing may be indicated depending on the specific clinical situation.

Ophthalmologic Examination of Children with a Permanent Confirmed Hearing Loss:
Children with hearing loss often have vision problems. The role of the ophthalmologist is to assess for the presence of syndromic visual loss associated with hearing loss, such as Usher’s syndrome. Evaluation for more common types of visual impairment including refractive error is essential for children who will likely be strong visual learners.

History:
Prenatal, Perinatal, Family, and Behavioral

Physical exam:
Complete ophthalmologic exam

Follow-up:
Routine annual ophthalmology evaluation for all children with hearing loss.
Special consideration should be given to children with the following diagnoses:
- Symptomatic congenital CMV
- Down syndrome (at 6 months and then annually)
- Very low birth weight (until deemed unwarranted by Ophthalmologist)
- Usher Syndrome

Genetic Evaluation and Counseling for Children with a Confirmed Hearing Loss:
Half of all hearing loss in children is genetic. The purpose of the evaluation performed by a clinical geneticist is to determine the cause of hearing loss, identify other medical issues that are associated with hearing loss and develop long term medical management
plans based on associated conditions that will impact the health and well-being of the child with hearing loss.

**History:**

- Pregnancy
- Family pedigree
- Medical History
- Developmental

**Physical:**

- Full physical examination
- Careful dysmorphologic examination
- Neurologic/developmental evaluation

**Genetic Diagnostic Tests:**

Genetic testing will be positive in nearly half of all children with hearing loss. Genetic testing for Connexin26 (GJB2) and Connexin 30 (GJB6) or Pendrin (SLC26A4), the most common single gene causes of hearing loss, should be considered based on physical examination, assessment of the type, severity, progression, and onset of hearing loss, and temporal bone anatomy based on CT or MRI studies.

There are over 400 syndromes associated with hearing loss. Syndromes such as Branchio-oto-renal syndrome, Stickler Syndrome, Waardenberg Syndrome, Usher Syndrome and others should be considered based on physical findings, radiologic evaluations and severity, progression, type and onset of hearing loss.

All other laboratory tests depend upon clinical evaluation and history, but may include the following:

- Chromosomal analysis
- Skeletal survey if short stature or disproportional growth.
- Evaluation of other systems: renal, cardiac, skin
- CT or MRI of the brain if neurologically abnormal
- Specialized genetic studies: such as comparative genomic hybridization or specific gene testing if indicated.

Genetic counseling (performed by a M.S. Genetic Counselor) should be provided in conjunction with the genetic evaluation (performed by a clinical genetics physician) to obtain a careful three generation family history, explain the benefits, risks and limitations of genetic testing to families, and explain the significance of testing results for the child and for the family.
Early Intervention (EI) Referrals from Primary Care Providers for Children with a Confirmed Hearing Loss:

All children with identified hearing loss should have a referral made to early intervention. All families of infants in Minnesota with any degree of bilateral or unilateral hearing loss should be referred to determine eligibility for Help Me Grow early intervention services. The primary care provider should assure that the infant has been referred to the local agency providing Part C intervention services, and provide ongoing support and guidance to the family and the child’s early intervention team. Members of the early intervention team may include educators of the deaf, speech-language pathologists, and educational audiologists in addition to other teachers and therapists. Early intervention options can be both home-based and center-based and are provided without charge to the family. Additional referrals may include private intervention services.

Parental concern about speech and language delays, and/or behavior at any time in a child’s life, should receive prompt referral for evaluation by Special Education in addition to audiologic testing.

Audiology Referrals from Primary Care Providers for Children with a Confirmed Unilateral Hearing Loss:

- Unilateral hearing loss is a significant risk factor for later acquired hearing loss in the previously normal ear. Audiological monitoring is recommended every three months during the first year of life, and every six to twelve months until age three or as clinically indicated due to possible progression to bilateral hearing loss.

- Amplification may have a role in facilitating language development.

- Use of amplification and role of intervention should be explored with the audiologist and otolaryngologist.

- Approach to follow-up should be individualized.

Audiology Referrals from Primary Care Providers for Children with a Confirmed Bilateral Hearing Loss:

- The audiologist will work with the family to assess the child and family’s needs and communication goals. In addition, the audiologist will work in collaboration with other professionals, such as otolaryngologists, educational audiologists, teachers of the deaf and hard of hearing, and parent support organizations, to provide comprehensive parent education about options.

- For families pursuing oral communication and use of residual hearing, early and consistent intervention (typically including conventional hearing aids) is the key to achieving typical oral language development. Children who gain limited
benefit from conventional amplification should be advised about additional opportunities such as cochlear implantation.

- For families pursuing a visual form of communication without the use of residual hearing, early and consistent intervention, and exposure to visual language is key. The audiologist will refer to professionals skilled in this area. Networking with older children and adults using sign language or cueing will increase the family’s skill with visual language.

- For families pursuing a combination of communication modalities, discussion with the family and coordination with the early intervention team to monitor effectiveness of the mode of communication and language milestones by the audiologist can benefit the child’s development.

**Primary care provider involvement is essential for children with hearing loss:**

- Make sure that routine health maintenance is not ignored because of all the time and attention paid to issues surrounding the hearing loss.

- All children with sensorineural hearing loss should have a routine 12 lead ECG performed to evaluate for prolonged QT duration. Approximately 1% of children with hearing loss will have this finding and be at risk for serious cardiac rhythm disturbance and sudden death. If prolonged QT is identified, the patient should be referred to a pediatric cardiologist for formal evaluation and treatment. Genetics evaluation and genetic counseling for the patient are required. ECG evaluation of family members from both sides is required.

- Facilitate timely follow-up with audiologist, and other consultants.

- Encourage active involvement with family support groups such as Minnesota Hands and Voices.

- Monitor continuous use of amplification device (if chosen) and maintain good contact with the family’s audiologist.

- Evaluate ongoing development of communication and language and refer to speech-language pathologists, other language specialists, or early intervention team should concerns arise.

For more detailed protocols on newborn hearing screening, diagnosis, amplification and early intervention please go to MDH website: www.health.state.mn.us/newbornscreening

*These guidelines are adopted and revised from the Colorado and North Carolina Guidelines*
Appendix A

RISK INDICATORS ASSOCIATED WITH PERMANENT CONGENITAL, DELAYED-ONSET, OR PROGRESSIVE HEARING LOSS IN CHILDHOOD

Risk indicators that are marked with a "§" are of greater concern for delayed-onset hearing loss.

1. Caregiver concern§ regarding hearing, speech, language, or developmental delay.
2. Family history§ of permanent childhood hearing loss.
3. All infants with or without risk factors requiring neonatal intensive care for greater than 5 days, including any of the following: ECMO,§ assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix). In addition, regardless of length of stay: hyperbilirubinemia requiring exchange transfusion.
4. In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,§ such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
8. Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
9. Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.
11. Chemotherapy.§

* Risk factor references available in JCIH 2007 Position Statement
References


CDC EHDI National Goals and Objectives, Final Version by the EHDI Data Committee, July 13, 2006.


mothers and their newborn infants with asymptomatic congenital cytomegalovirus infections. *Journal of Infectious Diseases*, 167, 72-77.


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