INTRODUCTION

This document is intended to promote a standardized approach to hearing screening, diagnosis and care for infants and young children who are deaf or hard of hearing. The following recommendations will help guide primary care and medical home providers who see infants and young children in the following situations:

- During the process of diagnosis after an ALERT: REFER result on the newborn hearing screen.
- After a definitive diagnosis of permanent or transient hearing loss.
- Throughout childhood to monitor for emerging hearing loss.

Without EHDI, infants with hearing loss may experience delays in many developmental areas including language, vocabulary, articulation, social interactions, and behavior.

1-3-6 Hearing Screen Model

SCREEN BEFORE 1 MONTH OF AGE
DIAGNOSE BEFORE 3 MONTHS OF AGE
INTERVENTION BEFORE 6 MONTHS OF AGE

Delay in hearing loss diagnosis can often be considered a neurodevelopmental emergency. The newborn period is a critical time for development and growth, and changes quickly.

Studies have shown that when hearing loss is identified before three months of age and intervention is initiated at no later than six months of age, children perform as much as 20 to 40 percent higher on school-related measures than children with hearing loss that was not identified early.

Many different healthcare professionals and entities play
a role in the hearing screening and follow-up process. As required by law (Minnesota Statute 144.9661), hospitals and out-of-hospital birth providers are expected to screen all infants for hearing loss and report results to the family, primary care provider, and the Minnesota Department of Health (MDH).

- Audiologists provide timely audiological follow-up and definitive testing. This is an important step to distinguish between children with false positive screening results and children with hearing loss.

- Otolaryngologists are essential in determining the cause of hearing loss and evaluating and treating ear diseases. They also provide information and participate in the assessment of candidacy for amplification, assistive devices, and surgical intervention.

**PRIMARY CARE PROVIDERS**

The primary care provider (PCP) is an essential team member who, in collaboration with the parents and other healthcare professionals, make up the infant’s “medical home.” It is the responsibility of primary care providers to monitor the general health, development, and well-being of the children in their care. A medical home provides healthcare services that are accessible, family-centered, continuous, comprehensive, coordinated, compassionate, and culturally appropriate.

The PCP oversees the EHDI process for children in his/her care by reviewing the newborn hearing screening results and ensuring that the child receives necessary audiological follow-up. Without the active assistance of primary care/medical home, the infant may become lost in this process (“lost to follow-up”) undermining the potential benefits of newborn hearing screening and increasing family stress.

If a child is identified with hearing loss, the PCP initiates referral for otolaryngology, genetics and ophthalmology evaluation to determine the cause of the hearing loss and identify related conditions. The PCP also partners with other medical specialists, including the otolaryngologist, to facilitate coordinated care for the infant and family. Because up to 40% of children with permanent hearing loss will demonstrate developmental delays or other disabilities, the PCP should closely monitor developmental milestones and initiate medical referrals related to suspected disabilities. Referrals should also be made to Part C Infant and Toddler Intervention Services through the MN Help Me Grow intake system2.

**ASSURE NEWBORN HEARING SCREEN AND RESCREEN**

Primary care providers should review hearing screening results with the infant’s parent/caregiver at the first clinic visit. If results have not arrived by the first visit, contact the birth hospital, neonatal intensive care unit (NICU), or midwife that cared for the newborn and request that the results be sent as soon as possible.

Providers are responsible for reviewing newborn hearing screening results to make sure that a hearing screen was performed.

*If the primary care provider is unable to obtain a hearing screening result for both ears or if the infant was not screened, schedule a hearing screening as soon as possible.*

**PASS—NEWBORN HEARING SCREENING RESULT**

PCPs should obtain, document, and discuss all screening results and risk factors for late onset or progressive hearing loss3.

Infants who pass their newborn hearing screen in both ears, or pass their re-screenings, are frequently assumed to have normal hearing. Providers should be aware that false negative screens may occur. Even with normal newborn hearing screening results, there is no assurance that a child will continue to have normal hearing.

- 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.
Guidelines for Primary Care and Medical Home Providers

Minnesota Department of Health

ALERT: REFER (FURTHER TESTING NEEDED) — NEWBORN HEARING SCREENING RESULT

It is the role of the PCP to coordinate care of an infant that has an ALERT: REFER result in one or both ears. Follow the steps below:

• Schedule or confirm that infant has a hearing rescreen appointment within the next two weeks.

• For practices that are able to perform a rescreen in the PCP office, use the Recommended Outpatient Rescreening Protocol in the Guidelines for the Organization and Administration of Universal Newborn Hearing Screening Programs in the Well-Baby Nursery (Refer all NICU graduates that did not pass newborn hearing screening directly to an audiologist for outpatient assessment).

• Report results back to the Minnesota Department of Health’s (MDH) EHDI Program within 48 hours.

A “wait and see” approach is never appropriate. An infant who does not pass newborn hearing screening has a potential developmental emergency.

NICU graduates who do not pass the newborn hearing screen prior to discharge require a diagnostic audiologic test battery as soon as medically possible. Ideally, this evaluation should take place prior to hospital discharge. See Guidelines for the Organization and Administration of Universal Newborn Hearing Screening Programs in the Special Care Nursery and Neonatal Intensive Care Unit (NICU).

If the infant has had an otolaryngology evaluation but did not have an audiological evaluation, the infant should be scheduled with audiology as soon as possible.

FACILITATE FOLLOW-UP EVALUATION WITH AUDIOLOGY WHEN INFANT DOES NOT PASS HEARING RESCREEN

ALERT: REFER (FURTHER TESTING NEEDED) — HEARING RESCREEN RESULT

The PCP is responsible to ensure that an audiological assessment is conducted on infants who do not pass a hearing rescreen.

Approximately 30-50% of infants reported to MDH with an ALERT: REFER result on hearing rescreen are determined to have a confirmed hearing loss (permanent or transient). The PCP should:

• Refer to an audiologist that has experience with infants for comprehensive diagnostic testing. See EHDI PALS.

• Confirm diagnostic audiology evaluation is scheduled and completed by no later than 3 months of age.

In order to meet the national standard that hearing loss be diagnosed as soon as possible but no later than three months of age, providers should expect that complete diagnosis may involve more than one clinic visit. For example, if an infant wakes up during testing, an additional visit may be necessary to complete the assessment. If the initial diagnostic visit takes place between four to six weeks of age, it is more likely that any necessary subsequent testing can still be completed on time and without sedation.

If families do not follow through on recommended diagnostic testing or attend their clinic appointments, developing and enacting a process for rescheduling missed or canceled appointments is strongly encouraged.
DEFINITIVE DIAGNOSIS OF UNILATERAL AND BILATERAL HEARING LOSS

Requires establishing whether underlying sensorineural hearing loss is present even when middle ear dysfunction (i.e. otitis media with effusion [OME]) is suspected. OME can mask a permanent underlying hearing loss and delay detection. See National Guidelines on OME. In Minnesota, 60% of infants with a late diagnosis of hearing loss had a history of middle ear fluid that delayed the final diagnosis of hearing loss.

Even in the presence of middle ear effusion, a complete diagnostic evaluation can determine the type of hearing loss present. In addition, a prompt referral and complete diagnosis from a trained pediatric audiologist reduces the number of infants who must be sedated in order to obtain results. Diagnostic testing can typically be completed without sedation if performed before three months of age, adjusted for prematurity.

Management for an infant with OME who has not passed the newborn hearing screening should include:

- Counseling of parent/caregiver regarding the importance of follow-up to ensure that the hearing is normal when OME resolves and to exclude an underlying sensorineural hearing loss.
- Follow-up hearing screen once OME resolves in order to document improvement.
- Referral to a pediatric audiologist and otolaryngologist if middle ear dysfunction does not resolve within one month.

The primary care provider may also consider coordinating care with other specialists to provide speech and language evaluation, speech and language therapy concurrent with managing OME, hearing aids or other amplification devices for hearing loss independent of OME, and tympanostomy tube insertion.

CONNECT CHILDREN IDENTIFIED WITH HEARING LOSS TO SERVICES

INFANTS IDENTIFIED WITH PERMANENT HEARING LOSS

As with any new, life-changing diagnosis, timely support from care professionals is critical. The infant’s primary care provider is responsible for monitoring the general health, development, and well-being of the infant. The PCP ensures that the child with hearing loss is receiving audiological management, habilitative, and educational services provided to children with hearing loss (i.e. Early Intervention, Speech/Language Services, visual language instruction).

The PCP also initiates referral for otolaryngology, genetics and ophthalmology evaluation to determine the cause of the hearing loss and identify related conditions.

Because 30% to 40% of children with confirmed hearing loss will have developmental delays or other disabilities, the PCP should closely monitor developmental milestones and initiate referrals related to suspected disabilities such as neurology, genetics or developmental pediatrics.

Middle-ear status should be monitored, because the presence of middle-ear effusion can further compromise hearing. Referrals to otolaryngology for early management of middle ear effusion is recommended.

The PCP 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.

**PCP EVALUATION OF A CHILD WITH PERMANENT HEARING LOSS**

**HISTORY**

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

- Prenatal history
- Ototoxic medication exposure
- Significant pregnancy complications
- Positive fluorescent treponemal antibody absorption (FTA-ABS) test or other positive syphilis confirmation
- Maternal drug use
- Multiple miscarriages
- Perinatal history – TORCH infections or risk factors for progressive hearing loss
- Family history of childhood hearing loss, syndromes, or other disorders associated with hearing loss
- Behavioral history
- Review of child’s motor milestones (may point toward vestibular dysfunction related to hearing loss)
- Review of prior audiologic testing

**PHYSICAL EXAM**

- Craniofacial abnormalities such as microcephaly, mandibular or midface anomalies
- Shape and location of pinna, presence of pre-auricular pits or sinuses, external ear canal stenosis, presence of middle ear fluid
- Growth trajectory
- Neurologic exam including cranial nerves

- Basic balance evaluation
- Evidence for genetic syndromes associated with hearing loss

**DEVELOPMENTAL/SOCIAL EMOTIONAL ASSESSMENT**

- Delays in speech and language, general development, social/emotional development

**LABORATORY STUDIES**

- Electrocardiogram (ECG) to check for prolonged Q-T syndrome or SA node dysfunction (refer to pediatric cardiology if identified), particularly in children with profound hearing loss.
- Cytomegalovirus (CMV) PCR testing through urine or saliva, performed prior to 3 weeks of age.
Early Hearing Detection and Intervention

REFERRALS FOR A CHILD WITH HEARING LOSS

AUDIOLOGY

EHDI GOAL: Amplification fit within 1 month of diagnosis (if chosen)

Once a child is identified with hearing loss, the audiologist will evaluate the child for candidacy for amplification and other sensory devices and assistive technology. Audiologists provide timely fitting and monitoring of amplification devices and also provide ongoing evaluation and management of the hearing loss. In addition, the audiologist will work in collaboration with other professionals, such as otolaryngologists, educational audiologists, teachers of the deaf and hard of hearing, speech-language pathologists, and parent support organizations, to provide comprehensive parent education about communication opportunities.

FOR THE CHILD WITH 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.

FOR THE CHILD WITH BILATERAL HEARING LOSS

• For families pursuing listening and spoken language and use of residual hearing, early and consistent intervention (typically including hearing aids) is the key to achieving language development.

• Children who gain limited benefit from amplification should be advised about additional opportunities such as cochlear implantation and visual language instruction.

OTOLARYNGOLOGY

The PCP should ensure that children who are deaf or hard of hearing have an otolaryngology evaluation within four months of diagnosis to determine etiology, and identify appropriate medical and surgical management. See Guidelines for Otolaryngologists.

GENETICS

The PCP should ensure that children who are deaf or hard of hearing have a genetics evaluation within one year of diagnosis even if there is a negative family history of hearing loss. This evaluation provides families with important information on etiology of hearing loss, prognosis for progression, and risk of associated disorders (e.g., renal, vision, cardiac). Genetic information may influence parents’ and physician’s decision-making regarding intervention options for their child.

OPHTHALMOLOGY

The PCP should ensure that children who are deaf or hard of hearing have an ophthalmologic evaluation within six months of diagnosis to document visual acuity and rule out concomitant or late-onset vision disorders such as Usher syndrome. This information may influence parents’ decision-making regarding intervention options for their child.

EARLY INTERVENTION

Young children with permanent hearing loss are at risk for delays in many developmental areas including language and communication, social interaction and behavior, as well as literacy and later academic achievement. It is critical that children with permanent hearing loss and their families receive appropriate support services as soon as possible.

The population of children who are deaf and hard of
hearing has diverse and unique needs and requires individualization in order to help each child reach his/her full potential. Access to highly qualified service providers who have specialized knowledge and skills in early childhood development, supporting families, and the communication opportunities chosen by each family is critical. To provide the highest quality of intervention, more than one provider may be required.

PART C INFANT AND TODDLER INTERVENTION SERVICES, (BIRTH THROUGH TWO YEARS OF AGE) AND EARLY CHILDHOOD SPECIAL EDUCATION (THREE YEARS OF AGE TO KINDERGARTEN ENTRANCE)

The Federal Individuals with Disabilities Education Act (IDEA) Part C Guidelines require medical professionals to refer all infants and toddlers (birth to three years of age) who have developmental concerns to their state’s Part C Infant and Toddler Intervention Program as soon as possible, but no later than 7 days after a developmental concern or delay is noted.

All young children who have hearing loss should be referred to Minnesota’s Part C Infant and Toddler Intervention Services / Preschool Special Education Services for an eligibility evaluation and possible services as soon as the hearing loss is identified.

Young children 0-5 years of age are referred through MN Help Me Grow intake system. Referrals can be made online or using a toll-free phone number.

Help Me Grow: 1-866-693-4769
www.helpmegrowmn.org

School-aged children who have been identified with hearing loss may be referred for possible educational support services through connection with their local school district.

In Minnesota, nearly all young children who have hearing loss are eligible to receive Part C Early Intervention and educational support services through their local school district. These services often include individualized home-based early intervention services to support the family’s needs and the child’s development in all areas, support to the child in a variety of early learning environments, consultation, information resources, and service coordination. Services are provided free of charge regardless of a family’s income or immigration status.

Early intervention team members may include educators of the deaf, speech-language pathologists, and educational audiologists. The PCP should provide ongoing support and guidance to the family and the child’s early intervention team.

SPEECH AND LANGUAGE THERAPY (PRIVATE/CLINIC BASED)

Speech-language pathologists provide both evaluation and intervention services to young children who are deaf and hard of hearing (including aural habilitation, language, speech, and cognitive-communication development beginning in infancy).

Referral to a speech-language pathologist should be considered for all children with hearing loss particularly those children whose families have chosen for them to learn to communicate through listening and spoken language.

CONNECT FAMILIES TO ADDITIONAL SUPPORTS

- Encourage active involvement in parent to parent support, advocacy and information networks such as Minnesota Hands & Voices. See Minnesota Hands & Voices Secure online referral.
- For families who choose visual communication, trained Deaf adult mentors provide instruction in American Sign Language, early visual communication methods, and Deaf Culture. See Deaf Mentor Family Program at Lifetrack.
- Children who are deaf or hard of hearing often benefit from a positive relationship with a trusted adult who is also deaf or hard of hearing. Role Models can help children who are deaf or hard of hearing practice
important, age-appropriate self-advocacy skills and help develop their sense of identity. See Deaf and Hard of Hearing Role Model Program.\(^{14}\)

**RISK FACTORS FOR HEARING LOSS BEYOND THE NEWBORN PERIOD**

The PCP should review every infant's medical and family history for the presence of risk indicators that require monitoring for delayed-onset or progressive hearing loss. They should ensure that an audiological evaluation is completed for children at risk of hearing loss at least once by 24 months of age, regardless of newborn screening results.

The Joint Committee on Infant Hearing (JCIH) 2007 Position Statement Principles and Guidelines for Early Hearing Detection and Intervention Programs\(^ {15}\) states that the following indicators can be associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood:

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

- Family history of permanent childhood hearing loss.

- NICU care for greater than five days, including any of the following: ECMO, assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide/Lasix). In addition, hyperbilirubinemia requiring exchange transfusion regardless of length of stay.

- 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 frequently identified syndromes include 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 meningitis.

- Head trauma, especially basal skull/temporal bone fracture that requires hospitalization.

- Chemotherapy.

Infants with any of these specific risk factors are at increased risk of delayed-onset or progressive hearing loss and should be monitored closely at regular intervals.

**Details:** Risk Indicators Associated with Permanent Congenital, Delayed-onset, or Progressive Hearing Loss in Childhood\(^ {3}\)

In addition, the PCP is responsible for ongoing surveillance of parent concerns about language and hearing, auditory skills, and developmental milestones of all infants and children regardless of risk status. Parental concern about speech and language delays, at any time in a child’s life, should receive prompt referral for an audiologic evaluation.

**REFERENCES**


Selected Links

1. Minnesota Statute 144.966: https://www.revisor.mn.gov/statutes/?id=144.966


15. Joint Committee on Infant Hearing (JCIH) Position Statement - http://pediatrics.aappublications.org/content/120/4/898