



— EARLY HEARING DETECTION AND INTERVENTION —

Guidelines for Otolaryngologists

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INTRODUCTION

This document is intended to promote a more standardized approach to care for infants who receive a REFER result on their newborn hearing screen. The document also aims to promote a better understanding of the rationale for early hearing detection and intervention for the general otolaryngologist. The following recommendations will help guide otolaryngologists who see infants in the following situations:

- During the process of diagnosis after a 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

BACKGROUND

The goal of an Early Hearing Detection and Intervention (EHDI) program is to promote communication from birth for all children through the early identification of hearing loss and the initiation of appropriate intervention services. Newborn hearing screening and follow-up plays

a critical role in the EHDI process by identifying newborns who are at risk for hearing loss and connecting them with diagnostic, support, and intervention services. Without EHDI, infants with hearing loss may experience delays in a variety of developmental areas, including vocabulary, articulation, intelligibility, social adjustments, and behavior.

To help ensure that every Minnesota newborn is screened for hearing loss, state law ([Minnesota Statute 144.966¹](#)) requires that a hearing screen be performed on all newborns prior to hospital discharge. National standards specify that screening should be complete as soon as possible but at no later than one month of age; hearing loss should be clinically diagnosed as soon as possible but at no later than three months of age; and intervention should be initiated as soon as possible but at no later than six months of age. With prompt referral and follow-up, Minnesota children have the opportunity to receive life-changing care and services even earlier than national guidelines prescribe.

Early identification and intervention can substantially reduce or even eliminate entirely the developmental delays that too often stem from a late diagnosis of hearing loss. As indicated in the [Joint Committee on Infant Hearing \(JCIH\) position statement²](#), studies have shown that if 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. For many children with hearing loss, early identification and intervention enables them to perform on language assessments at the same level as their hearing peers.

Many different healthcare professionals and entities play a role in the hearing screening and follow-up process. Minnesota 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). At the first well-child visit, primary care providers are expected to review newborn hearing screening results for all infants in their care and ensure that an outpatient follow-up visit is scheduled if the final screen is a REFER result. Audiologists provide the timely audiological follow-up and definitive testing that are essential in distinguishing between children with false positive screening results and children with hearing loss.

Otolaryngologists also play an important role in supporting timely and complete audiological diagnosis of infant hearing loss. Because ENT physicians often see newborns in conjunction with the infant's initial audiology outpatient rescreen, they play an important role in emphasizing the importance of timely follow-up and definitive diagnosis of hearing loss. Identification of newborn hearing loss is considered a neurodevelopmental emergency. Access to language is vital during the critical newborn period.

DURING DIAGNOSIS AFTER A REFER NEWBORN SCREENING RESULT

Definitive diagnosis of unilateral and bilateral hearing loss before three months of age requires establishing whether underlying sensorineural hearing loss is present even when middle ear dysfunction is suspected. The 2004 [Clinical Practice Guideline: Otitis Media with Effusion](#)³ (endorsed by both the American Academy of Otolaryngology-Head and Neck Surgery and the American Academy of Pediatrics) clearly indicates that prompt and complete diagnosis of children with otitis media with effusion (OME) is critical, stating: "Distinguish the child with OME who is at risk for speech, language, or learning problems and more promptly evaluate hearing... and the need for intervention." Infants who have not passed the newborn hearing screen **are considered at risk** and require full investigation of

possible underlying mixed or sensorineural hearing loss according to best practice timelines.

The rate of confirmed hearing loss (permanent and transient) after a REFER result on the initial outpatient rescreen as reported to MDH is between 30 and 50 percent in Minnesota. Recent MDH Newborn Screening Program EHDI data also indicates that 60 percent of Minnesota infants with delayed diagnosis had a history of middle ear fluid, and therefore were not referred for a complete diagnostic ABR evaluation per best practice guidelines. This data illustrates the important role otolaryngologists can play in enabling prompt diagnosis by providing timely referral for comprehensive audiological test battery, regardless of ear exam findings. Even in the presence of middle ear effusion, a complete auditory brainstem response (ABR) evaluation (consisting of air- and bone-conducted clicks and frequency-specific tone burst stimuli), supplemented by otoacoustic emissions and age-appropriate immittance measures, can determine the type of hearing loss present.

In the general population, American Academy of Pediatrics (AAP) guidelines call for the management of middle ear fluid with "watchful waiting." However, infants with REFER results on the newborn hearing screen are at risk for hearing loss, and therefore should always receive prompt referral for a definitive diagnosis. Infants should be referred to an audiologist with appropriate training and experience. A prompt referral and complete diagnosis:

- allows evidence-based decision making for the provider and family;
- avoids missing underlying sensorineural hearing loss, which can impact the course of both medical and educational intervention;
- decreases the likelihood that infants become lost to follow-up or have an incomplete diagnosis beyond six months of age; and
- reduces the number of infants who must be sedated in order to obtain results (diagnostic testing can typically be completed without sedation if performed by three months of age, adjusted for prematurity).

Neonatal intensive care unit (NICU) graduates who do not pass the newborn hearing screen prior to discharge require a diagnostic audiological test battery as soon as medically feasible. Ideally, this evaluation should take place prior to hospital discharge. If an audiological evaluation has not occurred before the otolaryngology appointment, the infant should be scheduled as soon as possible.

As per the [2007 JCIH position statement](#)², a unilateral REFER result is as significant as bilateral REFER results and requires the same timeliness of follow-up and definitive diagnosis. Recent Minnesota data shows that unilateral and bilateral REFER results have equal risk of confirmed hearing loss after not passing the initial outpatient rescreen. A child with confirmed unilateral hearing loss is at risk for speech delay and acquired hearing loss in the opposite ear.

In order to meet the national standard that hearing loss be diagnosed at 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.

Because diagnosis can be a long and sometimes confusing process for parents, otolaryngologists should support families in scheduling appointments with a trained pediatric audiologist as needed and as soon as possible. If families do not follow through on recommended diagnostic testing or attend their clinic appointments, developing and enacting a process for rescheduling missed or cancelled appointments is strongly encouraged.



AFTER DIAGNOSIS OF PERMANENT HEARING LOSS

As with any new, life-impacting diagnosis, timely support from care professionals is critical. Ideally, ENT consultation should occur on the same day as audiological diagnosis, or within one to two weeks at the latest. Scheduling priority should be offered to families with a new diagnosis of permanent hearing loss to ensure they receive the support they need as soon as possible. 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 should participate in the assessment of options for amplification, assistive listening devices, and cochlear implantation. If amplification is chosen, the ENT physician should provide medical clearance for the fitting of amplification. Families should be fully informed about all avenues of surgical and educational intervention available to them and be encouraged to play a role in decision-making processes to help ensure that care is child- and family-centered and culturally appropriate. In partnership with the audiologist, primary care provider, and other care team members, the ENT physician should also participate in the long-term monitoring of a child's hearing.

It is important to note that unilateral hearing loss is a significant risk factor for later acquired hearing loss in the previously normal ear. Hearing loss of any degree, including mild bilateral and unilateral loss, has been shown to adversely affect speech, language, academic, and psychosocial development. For these reasons, audiological monitoring is recommended every three months during the first year of life, and every six to 12 months until age three or as clinically indicated due to possible progression to bilateral hearing loss. Studies have shown that children with unilateral hearing loss score significantly worse in oral language skills than their siblings. Additionally, as noted in the [Centers for Disease Control and Prevention \(CDC\) Unilateral Hearing Loss: Outcomes](#)⁴ document, studies have indicated that 25 to 35 percent of children with unilateral hearing loss are at risk for failing a grade in school. Appropriate amplification for children with unilateral hearing loss can make a significant difference in language development.

For all children who present with evidence of permanent hearing loss, the ENT physician should perform a complete work-up. Often, a hearing or balance disorder is an indicator of, or related to, a medically treatable condition or an underlying systemic disease. The following outlines the components of a complete work-up:

Early Hearing Detection and Intervention

1. **History:**

- 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 (toxoplasmosis, other infections, rubella, cytomegalovirus, or herpes simplex) 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

2. **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

3. **Review of diagnostic audiologic test battery:** review should consist of:

- ABR with air- and bone-conducted clicks and frequency specific tone burst stimuli
- OAE (otoacoustic emission)
- Age-appropriate immittance measures
- Behavioral testing when appropriate

4. **Laboratory studies:**

- Electrocardiogram (ECG) to check for prolonged Q-T syndrome (refer to pediatric cardiology if identified)
- Urinalysis to check kidney function (refer to pediatric nephrology as needed)
- Cytomegalovirus (CMV) testing

5. **Imaging:** no universally accepted algorithm, but consider:

- MRI – can identify presence of cochlear nerve, some anatomical abnormalities (e.g., enlarged vestibular aqueduct and some cochleovestibular anomalies), and retrocochlear abnormalities
- CT – can detect cochleovestibular anomalies and middle ear anomalies, but *cannot* detect cochlear nerve or retrocochlear abnormalities

6. **Referrals:** collaboration with and guidance to the primary care provider regarding recommended referrals to additional specialists, including:

- Genetics – A genetics referral is recommended by the JCIH, AAP, and American College of Medical Genetics (ACMG) to determine the cause of hearing loss. Genetic testing can inform prognosis as well as medical management of children with hearing loss.
- Ophthalmology – Regular ophthalmologic care is part of the management of children who are deaf or hard of hearing. Ten percent of children who are

deaf/hard of hearing will have Usher syndrome and will develop vision loss.

- Early intervention – All families of infants and young children with any degree of bilateral or unilateral permanent hearing loss should be referred for early intervention services.
- Private service options – A referral for private service options (e.g., speech pathology) should be considered as needed to supplement intervention.

7. **Discussion with family:** should include surgical intervention options such as bone-anchored hearing aid (BAHA) implant, cochlear implant, or surgical repair—if appropriate—and a referral made as needed

the date of effusion onset (if known) or diagnosis (if onset is unknown).

- Conduct hearing testing when OME persists for three months or longer or at any time that language delay, learning problems, or a significant hearing loss is suspected in a child with OME.
- Re-examine children with persistent OME who are not at risk at three- to six-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 MDH [Guidelines for Infant Audiologic Assessment](#)⁶ and MDH [Guidelines for Pediatric Amplification](#)⁷.

AFTER DIAGNOSIS OF TRANSIENT HEARING LOSS

Once a definitive diagnosis of transient hearing loss (mild to moderate conductive from middle ear fluid or unknown pathology) is complete, otolaryngologists should follow both the Academy of Otolaryngology's 2004 [Clinical Practice Guideline: Otitis Media with Effusion](#)³ and 2013 [Clinical Practice Guideline: Tympanostomy Tubes in Children](#)⁵.

The following is a summary of the guidelines for managing OME in children ages two months through 12 years from the 2004 [Clinical Practice Guideline: Otitis Media with Effusion](#)³:

- Use pneumatic otoscopy as the primary diagnostic method and distinguish OME from acute otitis media.
- Document the laterality, duration, and presence of effusion, as well as the severity of associated symptoms at each assessment of the child with OME. Additionally, document the specific reason for referral (e.g., evaluation, surgery) and provide additional relevant information, such as history of acute otitis media and the 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 of children at risk.
- Manage and carefully monitor the child with OME who is not at risk for speech, language, or learning problems with watchful waiting for three months from

The following is a summary of facts and guidelines for evaluating the need for and managing tympanostomy tubes in children ages six months to 12 years from the 2013 [Clinical Practice Guideline: Tympanostomy Tubes in Children](#)⁵:

- Many children with a fluid build-up (otitis media with effusion) in the middle ear improve on their own, especially when the fluid is present for less than three months.
- Children with OME that lasts for more than three months should have an age-appropriate hearing test.
- If OME and hearing difficulty are present for more than three months in both ears, hearing and quality of life can be improved with tympanostomy tubes.
- Tubes should not be used in children with recurrent or frequent ear infections (acute otitis media) who do not have middle ear effusion.
- Children who are at risk for developmental difficulties when OME is present in one or both ears and is unlikely to resolve quickly—such as children with permanent hearing loss, speech/language delays or disorders, autism-spectrum disorder, Down syndrome, craniofacial disorders, cleft palate, and/or developmental delay—may be offered tympanostomy tubes.
- When ear infections occur in children with tubes, the authors recommend prescribing topical antibiotic ear drops rather than systemic oral antibiotics. Topical antibiotic ear drops are more effective and present fewer side effects.

MONITORING FOR EMERGING CHILDHOOD HEARING LOSS

The incidence of childhood hearing loss doubles by the time children are of school age. Therefore, all infants should be monitored for late-onset or progressive hearing loss per accepted national recommendations and as recommended by the MDH [Guidelines for Hearing Screening After the Newborn Period to Kindergarten Age](#)⁸. At any time in a child's life, parental concern about speech and language delays or risk factors (refer to [Appendix A](#)⁹) associated with hearing loss should prompt timely referral for an audiologic evaluation. Hearing testing can be performed at any age.

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SELECTED LINKS

¹ Minnesota Statute 144.966

<https://www.revisor.mn.gov/statutes/?id=144.966>

² Joint Committee on Infant Hearing (JCIH) position statement

<http://www.jcih.org/posstatemts.htm>

³ Clinical Practice Guideline: Otitis Media with Effusion

http://oto.sagepub.com/content/130/5_suppl/S95.full.pdf+html

⁴ CDC Unilateral Hearing Loss: Outcomes

www.cdc.gov/ncbddd/hearingloss/documents/unilateral/DRoss%20Pkg06-Unilateral-Outcomes%20TABLE-cleared.pdf

⁵ Clinical Practice Guideline: Tympanostomy Tubes in Children

http://oto.sagepub.com/content/149/1_suppl/S1.full.pdf+html

⁶ Guidelines for Infant Audiologic Assessment

<http://www.improveehdi.org/wi/library/files/MDH%20Audiologic%20Assessment%20Guidelines%281%29.pdf>

⁷ Guidelines for Pediatric Amplification

<http://www.improveehdi.org/wi/library/files/MDH%20Pediatric%20Amplification%20Guidelines.pdf>

⁸ Guidelines for Hearing Screening After the Newborn Period to Kindergarten Age

<http://www.improveehdi.org/mn/library/files/afternewbornperiodguidelines.pdf>

⁹ Appendix A: Risk Factors Associated with Permanent Congenital, Delayed-onset, or Progressive Hearing Loss in Childhood

<http://www.improveehdi.org/wi/library/files/Risk%20Indicators%20from%20JCIH%202007%20Position%20Statement.pdf>