

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/did not pass 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 evaluation of an infant after a refer/did not pass result on the newborn hearing screen.
- After a definitive identification of permanent or transient hearing loss.
- After a diagnosis of congenital Cytomegalovirus (cCMV).
- Throughout childhood to monitor for delayed, late onset or progressive hearing loss.

Respect for identities statement

The Minnesota Department of Health (MDH) recognizes that everyone has their own unique identity and we respect the terms people want to use to identify themselves. People may identify as deaf, deafblind, hard of hearing, a person with

hearing loss, Deaf Plus, a person with a hearing difference, Deaf Disabled, etc.

For the purposes of this guideline, MDH uses “deaf, deafblind, and hard of hearing (DHH/DB),” in an all-inclusive manner. The term “hearing loss” may be used when talking about a medical diagnosis.



Background

Without Early Hearing Detection and Intervention (EHDI), infants who are deaf or hard of hearing are immediately at risk for experiencing delays in a variety of developmental areas, including vocabulary, articulation, intelligibility, social adjustments, and behavior (Joint Committee on Infant Hearing [JCIH], 2007; Yoshinaga-Itano et al., 1998).

The goal of an EHDI program is to promote effective communication and access to language for all children through early identification of hearing levels outside the typical range and the initiation of appropriate intervention services as soon as possible. Newborn hearing screening and subsequent 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.

Minnesota Statute 144.966¹ requires a hearing screen to be performed on all newborns prior to hospital discharge. National standards specify that screenings should be completed as soon as possible but by 1 month of age; hearing levels outside the typical range should be clinically diagnosed as soon as possible but by 3 months of age; and intervention should be initiated as soon as possible but by 6 months of age for children with hearing levels outside the typical range. With prompt referral and follow-up, Minnesota children can receive life-changing care and services even earlier than national guidelines prescribe.

Early identification and intervention can substantially reduce or even eliminate developmental delays that too often stem from a late detection of hearing levels outside the typical range. As indicated in the Joint Committee on Infant Hearing (JCIH) Year 2019 Position Statement², studies have shown that if hearing levels outside the typical range are identified before 3 months of age and intervention is initiated by 6 months of age, infants and children can be expected to achieve developmental milestones and reach their full potential. The sooner hearing loss is identified, the sooner children can receive help and begin developing critical language skills.

Many different healthcare and educational professionals and organizations play a role in the hearing screening and follow-up process. Minnesota hospitals and out-of-hospital birth providers are responsible for screening the hearing of all infants and for reporting the results to the family, the primary care provider, and 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 with audiology is scheduled if the final screen is a refer/did not pass result.

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 also play an important role in supporting timely and complete audiological assessment. Otolaryngologists often see newborns in conjunction with the infant's initial audiology outpatient rescreen, and they play an important role in emphasizing the importance of timely follow-up and definitive determination of hearing levels.



During Diagnosis After a Refer/Did Not Pass Hearing Screen Result

Definitive diagnosis of unilateral and bilateral hearing loss before 3 months of age requires establishing whether underlying permanent hearing loss is present even when middle ear dysfunction is suspected. A study of newborn hearing screenings in the presence of otitis media with effusion (OME) found 1 in 10 children had sensorineural hearing loss in addition to transient conductive hearing loss from OME (Rosenfeld et al., 2016). Infants who have not passed the newborn hearing screen **are considered at risk for hearing loss** and their parents require counseling to ensure they return for full diagnostic investigation of possible underlying mixed or sensorineural hearing loss (Rosenfeld et al., 2016). According to Clinical Practice Guidelines^{3,4} from the American Academy of Otolaryngology – Head and Neck Surgery, tympanostomy tubes may need to be considered while completing the diagnostic process, especially if a child has a risk factor for speech, language and learning problems (JCIH, 2019; Rosenfeld et al., 2016, 2022).

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 and degree of hearing loss present.

Infants with refer/did not pass results on the newborn hearing screen and middle ear fluid on exam should always receive prompt referral for definitive audiologic assessment. 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 education intervention.

- Decreases the likelihood that infants become lost to follow-up or have an incomplete diagnosis beyond 6 months of age.
- Reduces the number of infants who must be sedated to obtain results. (Around the age of four months, it can be more challenging to test a child under natural sleep. If you have questions, consult your audiology clinic).

Neonatal intensive care unit (NICU) graduates who do not pass the newborn hearing screen prior to discharge require a diagnostic audiologic 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. Ideally, this is completed prior to or concurrent with the otolaryngology appointment.

As per the 2007 and 2019 JCIH Position Statements, a unilateral refer/did not pass result is just as significant as bilateral refer/did not pass results and requires the same timeliness of follow-up and definitive diagnosis. Additionally, some children have additional risk factors for delayed onset or progressive hearing loss that could affect the opposite ear (JCIH, 2019).

In order to meet the national standard that hearing loss is diagnosed by 3 months of age, providers should expect that complete diagnosis may involve more than one clinical 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 4 to 6 weeks of age, it is more likely that any necessary subsequent testing can still be completed on time and before an age that sedation may be necessary.

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 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 canceled appointments is strongly encouraged.

After a Diagnosis of Permanent Hearing Loss

As with any new, life-impacting diagnosis, timely support from care professionals is critical. Ideally, otolaryngology consultation should occur prior to or within 30 days after a hearing loss diagnosis. Scheduling priority should be offered to families with a new diagnosis of permanent hearing levels outside the typical range to ensure they receive the support they need as soon as possible. The otolaryngologist should have expertise in childhood hearing loss and is responsible for investigating the etiology of hearing levels outside the typical range and for determining whether medical or surgical intervention may be an appropriate option. In addition, the otolaryngologist should participate in the assessment of options for amplification, assistive listening devices, and cochlear implantation. If amplification is chosen, the otolaryngologist should provide medical clearance for the fitting of amplification. Families should be fully informed about communication/language acquisition opportunities, all avenues of surgical and educational intervention available to them, and be encouraged to play a role in the decision-making process 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 otolaryngologist should also participate in the long-term monitoring of a child's hearing.

In Minnesota, about one third of the children reported to MDH have unilateral hearing loss and two thirds have bilateral hearing loss (congenital and late onset). It is important to note that almost half of children with unilateral hearing loss are at risk for declining hearing in one or both ears, usually within the first few years after initial diagnosis (Fitzpatrick et al., 2023). In Minnesota, about 1 in 10 children followed by the EHDI program initially reported with unilateral hearing loss later developed bilateral hearing loss, consistent with the rate published by Fitzpatrick et al., (2023). Children with unilateral hearing loss are at risk for delayed language development compared to children with typical hearing (Bell et al., 2022; Fitzpatrick et al., 2019, Lieu et al., 2010, 2012).



For these reasons, audiological monitoring is recommended for all children with hearing loss every three months during the first year of life, and every 6 to 12 months until age 3, or as clinically indicated due to possible progression of hearing loss. Along with monitoring, promptly refer to early intervention or their local school district for educational supports. Appropriate amplification may be considered.

For all children who present with evidence of permanent hearing loss, the otolaryngologist should perform a complete work-up. Often, hearing loss or a balance disorder is an indicator of, or related to, an underlying health condition. Clinical work-up for permanent childhood hearing loss is discussed in the literature (Hazen & Cushing, 2021; Jerry & Oghalai, 2011; Kenna, 2022; Khela & Kenna, 2020; Kimberling et al., 2010; Li et al., 2022; Liming et al., 2016; Prosser et al., 2015; Rawlinson et al., 2017; Raymond et al., 2019; Robson et al., 2023; Shave et al., 2022; Shearer, 2024; Shekdar & Bilaniuk, 2019; Sindhar & Lieu, 2021). The following outlines the components of a complete work-up:

1. History

- ✓ Prenatal history
 - Significant pregnancy complications
 - Positive fluorescent treponemal antibody absorption (FTA-ABS) test or other positive syphilis confirmation.
 - Maternal drug use
 - Multiple miscarriages
- ✓ Perinatal history – Ototoxic medication exposure, TORCH infections (Toxoplasmosis, Other infections, Rubella, Cytomegalovirus, or Herpes simplex), prematurity, NICU stay, ECMO, high frequency (HF) ventilation, or risk factors for progressive hearing loss
- ✓ Family history of childhood hearing loss, syndromes, or other disorders associated with hearing loss
- ✓ Developmental 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 audiological test battery

- ✓ 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) for bilateral profound sensorineural hearing loss*
- ✓ Urinalysis to check kidney function (refer to pediatric nephrology as needed), based on family history*
- ✓ Results of universal congenital Cytomegalovirus (CMV) screen

** If not already underway with comprehensive genetic testing/genetic referral*

5. Imaging

- ✓ No universally accepted algorithm, but consider:
 - MRI – can identify the presence of cochlear nerve, some anatomical abnormalities (e.g., enlarged vestibular aqueduct and some cochleovestibular abnormalities), and retrocochlear abnormalities.
 - CT – can detect cochleovestibular anomalies and middle ear anomalies, but cannot identify the presence of cochlear nerves 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, American Academy of Pediatrics(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 with hearing loss. 3 to 6% or more of children who are deaf/hard of hearing are estimated to have Usher syndrome and could develop vision loss (Kimberling et al., 2010, NIDCD, 2025). Connect families of children with combined vision and hearing loss to the Minnesota DeafBlind Project so that families can access specialized assistance for dual sensory loss.⁵
- Help Me Grow/early intervention – Young children 0-5 years of age are referred to Minnesota’s Part C Infant and Toddler Intervention Services/Preschool Special Education Services for an eligibility evaluation and possible services through Minnesota’s Help Me Grow intake system. Referrals can be made online at www.helpmegrowmn.org⁶ or using a toll-free phone number 1-866-693-4769.
- Private service options – A referral for private service options (e.g., speech-language pathology) should be considered as needed to supplement intervention.

- ✓ Review the Guidelines for Referral to Early Intervention, Medical Specialities, and connection to Parent-to-Parent and Family Support⁷ for details on how to make these referrals.

7. Discussion with family

- ✓ Should include surgical intervention options such as bone-anchored hearing aid (BAHA) implant, cochlear implant, or other surgical intervention – if appropriate – and a referral made as needed.
- ✓ Families have choices about language and communication for their young children. No one communication opportunity is the “right” or “best” way for all children with hearing differences or for their families as they support their child’s language learning. For unbiased information on this topic, otolaryngologists should encourage families to explore these options in Exploring Communication

Opportunities for Children with Hearing Differences: An Overview.⁸

- ✓ Discuss connections to parent-to-parent support and deaf and hard of hearing mentors and role models early on in their journey.^{9,10,11}

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 2016 Clinical Practice Guideline: Otitis Media with Effusion (Update)³ and 2022 Clinical Practice Guideline: Tympanostomy Tubes in Children (Update).⁴

The following is a summary of some key action statements for managing OME in children ages 2 months through 12 years from the 2016 Clinical Practice Guideline: Otitis Media with Effusion (Update).³

- Use pneumatic otoscopy as the primary diagnostic method and distinguish Otitis Media with Effusion (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.
- Counsel parents on the importance of follow-up to ensure that hearing is normal when OME resolves and to make sure there is no underlying sensorineural hearing loss (SNHL).
- Determine if a child with OME is “at risk for speech, language or learning problems from middle ear effusion because of baseline sensory, physical, cognitive, or behavioral factors.”
- 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 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 Assessment¹² and MDH Guidelines for Pediatric Amplification.¹³
- 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.

For infants with persistent conductive loss lasting more than 6 months, consider referral to early intervention and to audiology for amplification (temporary or long term) to ensure adequate auditory access.

After Diagnosis of Congenital CMV

In 2023, Minnesota became the first state in the nation to screen every newborn for cCMV. A standardized approach to hearing and balance-related follow-up care for these children was developed to ensure consistency in care and help providers minimize possible loss to follow-up or parent confusion about ongoing care. For infants with a medical diagnosis of cCMV that is confirmed with urine testing, the suggested clinical protocol for audiology follow-up care is detailed in the EHDl Guidelines for Audiologists – Section 4: Audiology Guidelines for Infants with Congenital Cytomegalovirus.¹⁴ Otolaryngologists working with these children can help support this timeline to aid in early detection of developing hearing loss associated with cCMV.

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.¹⁵ At any time in a child's life, parental concern about speech and language delays or risk factors¹⁶ associated with hearing loss should prompt timely referral for an audiologic evaluation. Hearing testing can be performed at any age.

The following is a summary of some key action statements for evaluating the need for and managing tympanostomy tubes in children ages 6 months to 12 years from the 2022 Clinical Practice Guideline: Tympanostomy Tubes in Children (Update):⁴

- 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 evaluation.
- 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, blindness, cleft palate, developmental delay, intellectual disability, learning disorder, or attention-deficit hyperactivity disorder – may be offered tympanostomy tubes.

Selected Links

¹ Minnesota Statute 144.966 <https://www.revisor.mn.gov/statutes/cite/144.966>

² Joint Committee on Infant Hearing (JCIH) Year 2019 Position Statement <https://doi.org/10.15142/fptk-b748>

³ Clinical Practice Guideline: Otitis Media with Effusion (Update) <https://aao-hnsfjournals.onlinelibrary.wiley.com/doi/10.1177/0194599815623467>

⁴ Clinical Practice Guideline: Tympanostomy Tubes in Children (Update) <https://aao-hnsfjournals.onlinelibrary.wiley.com/doi/full/10.1177/01945998211065662>

⁵ Minnesota DeafBlind Project <https://www.dbproject.mn.org>

⁶ Minnesota Help Me Grow intake system www.helpmegrowmn.org

⁷ EHDI Guidelines for Audiologists - Section 2: Guidelines for Referral to Early Intervention, Medical Specialities and Connection to Parent-to-Parent and Family Support www.health.state.mn.us/docs/people/childreneyouth/improveehdi/guidereferrei.pdf

⁸ Exploring Communication Opportunities for Children with Hearing Differences: An Overview <https://www.mnlowincidenceprojects.org/Projects/ehdi/ehdiCommunicationOpp.html>

⁹ Minnesota Hands and Voices www.lssmn.org/mnhandsandvoices/

¹⁰ Deaf Mentor Family Services <https://www.lssmn.org/services/families/deaf-hard-of-hearing/mentor-services>

¹¹ MN Hands & Voices Deaf and Hard of Hearing Guide Program <https://www.lssmn.org/mnhandsandvoices/about-us/deaf-and-hard-hearing-guide-program>

¹² Guidelines for Infant Audiologic Assessment health.state.mn.us/docs/people/childreneyouth/improveehdi/guideehdiaudiol.pdf

¹³ Guidelines for Pediatric Amplification <https://www.health.state.mn.us/docs/people/childreneyouth/improveehdi/guideamplification.pdf>

¹⁴ EHDI Guidelines for Audiologists – Section 4: Audiology Guidelines for Infants with Congenital Cytomegalovirus <https://www.health.state.mn.us/docs/people/childreneyouth/improveehdi/audiogdlnccmv.pdf>

¹⁵ Guidelines for Hearing Screening After the Newborn Period to Kindergarten Age <https://www.health.state.mn.us/docs/people/childreneyouth/improveehdi/guideafternb.pdf>

¹⁶ Risk Factors for Early Childhood Hearing Loss <https://www.health.state.mn.us/docs/improveehdi/riskindicators.pdf>

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