

MINNESOTA EARLY HEARING DETECTION AND INTERVENTION (EHDI) PROGRAM

Guidelines for Organization and Administration of Special Care Nursery and Neonatal Intensive Care Unit (NICU) Universal Newborn Hearing Screening Programs

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Introduction

The goal of an Early Hearing Detection and Intervention (EHDI) Program is to identify and provide effective intervention for newborns and infants with permanent childhood hearing loss and those at risk for hearing loss that may affect health, communication, learning and development. To achieve this goal, Minnesota law (Minnesota Statutes 144.966) requires that a hearing screen must be performed on all newborns prior to hospital discharge, or as soon as medically feasible for infants with a prolonged stay in Special Care Nursery and the NICU. National standards hold that screening should be complete by one month of age; hearing loss should be clinically diagnosed by 3 months of age and intervention initiated by 6 months of age or as soon as medically feasible for infants with a prolonged stay in the NICU. Corrected gestational age needs to be considered for the significantly premature infant. With prompt referral and follow-up, Minnesota children have an opportunity to receive appropriate care and services even earlier.

Because of the importance of early identification of hearing loss, all screening, follow-up, and tracking procedures must be consistent with current Minnesota statutory requirements. This document provides recommended guidelines for newborn hearing screening programs in the Special Care Nursery and the NICU. Please see separate document regarding hearing screening guidelines for infants in the Well Baby Nursery. Additional resources are available from the Minnesota Department of Health (MDH) to assist hospitals and hearing screeners with specific issues of program development and management such as training, supervision, equipment options, and quality assurance issues (www.health.state.mn.us/newbornscreening).

Child and Family-Centered Communication

Minnesota statute requires hospitals to present information to parents that covers the following topics:

- Potential risks and effects of hearing loss
- Benefits of early detection and intervention
- Nature of the screening procedure
- Applicable costs of screening procedure
- Parental option to have screening performed but to request that test results be destroyed by the MDH
- Parental option to refuse screening

Best practice would also include providing the following to parents:

- Prevalence of and risk factors for permanent childhood hearing loss (Appendix A)
- Possibility of late or progressive onset of hearing loss, including otitis media
- Developmental milestones for speech, language, and hearing (Appendix B)

Personnel

Screening may be performed by trained personnel including the following:

- Audiologists, audiological technicians/assistants
- Nurses
- Nursing assistants
- Other trained medical personnel

Although licensed audiologists do not need to conduct the actual hearing screening, audiologists are uniquely qualified to develop and implement all aspects of an EHDI program. Hospital screening programs benefit from direct access to audiological consultation to address screening criteria, quality assurance, follow-up assessment and intervention services.

Training

Training qualified screeners is an ongoing process. Training typically includes three phases: initial training and demonstration of skills, ongoing quality assurance, and refresher training. Resources for training may include experienced screening program managers and local licensed audiologists.

Initial training content is based on current best practice procedures as reported in professional literature. Assuring competent screeners is a critical component for every screening program. Training components include the following:

- Competency-based demonstration of trainees' performance in the nursery environment;
- Periodic observation of each screener in the nursery environment by a skilled professional such as an audiologist and/or program manager;
- Annual staff refresher training, with individual training available as needed.

Equipment

Screening programs must use objective physiological screening methods such as Automated Auditory Brainstem Response (AABR) or otoacoustic emissions (DPOAEs or TOAEs) that do not require a behavioral response from the infant. OAE and AABR have proven to be effective screening measures. All equipment must meet technical specifications, calibration standards, and hospital safety standards. If only one test technology is to be utilized in the NICU, recent JCIH guidelines (2007) recommend that it be the AABR.

Hearing Screen Stimulus Parameters

TOAEs should be measured in response to a click at approximately 80dB SPL (78-82dB SPL). DPOAEs should be measured in response to a series of paired tones (f1 and f2), with a ratio of 1.22 at a moderate level, where L1/L2= 65/55dB SPL.

Newborn screening AABRs typically are evoked using click stimuli at 30 to 35dB nHL at a moderate rate. Non-automated ABR testing is NOT recommended for newborn hearing screening programs in hospital nurseries due to issues of potential operator error and significant time/cost effectiveness issues.

Default stimulus parameters of either OAE or AABR equipment should be reviewed by the screening program's consulting audiologist or MDH audiologists to assure they are appropriate or to adjust them to be in accordance with clinically accepted national practices.

Pass/Refer Criteria

OAE

Manufacturer's pass/fail criteria should be reviewed by the consulting audiologist or MDH audiologist and should be in accordance with clinically accepted national practices. Typical passing criteria for TOAEs include overall reproducibility greater than 50%, at least 50 low noise samples collected, stimulus stability of 75% or greater, and responses present at least 6dB above noise floor for at least three of the five test frequencies, with 4000Hz a mandatory passing frequency. Typical passing criteria for DPOAEs requires absolute response amplitude of at least -6dB and responses at least 6dB above the noise floor at three or more of the test frequency bands, with the 4000Hz region a mandatory passing frequency.

AABR

- Screening AABR pass criteria for newborns typically require repeatable Wave V evoked responses to clicks at ≤ 35 dB nHL for each ear, within specific latency parameters. Once again the consulting audiologist or MDH audiologist should review the default pass/fail criteria for the specific AABR equipment used to ensure compliance with accepted national practices.

A quality screening program benefits from incorporating new and improved evidence-based technologies and procedures as they become available.

Hearing Screening Protocol

The prevalence of hearing loss has been reported to be 10 times greater among infants in the neonatal intensive care unit than in the well baby nursery (Norton et. al., 2000). The purpose of a screening test is to identify those infants at risk for hearing loss who need further testing. A screening test is not a diagnosis. Because of the higher risk of hearing loss in the NICU population, typical referral rates in the NICU are 7 to 10%.

Hearing loss may be related to conductive involvement, sensory deficit, neural dysfunction or to some combination of the above. In addition NICU infants are at risk for late onset hearing loss (see JCIH risk factors, Appendix A). While the additional risk for neural hearing loss, specifically auditory neuropathy/dysynchrony, is not well established, it has been reported more frequently in the NICU population than among healthy newborns.

If the newborn is expected to remain in the hospital for a prolonged period, screening needs to be performed prior to three months corrected age or when medically feasible. If the infant's status changes after the initial hearing screen then a re-screen may be necessary.

Choice of the equipment used in hearing screening in the NICU needs to be made carefully. Hearing screening using otoacoustic emissions effectively identifies cochlear or conductive hearing loss but will miss hearing loss of neural origin. Therefore, infants in the NICU for more than five days and therefore presumed to be at highest risk for neural hearing loss (JCIH, 2007) must have AABR included as part of their hearing screening prior to discharge in order to be effectively screened

If nurseries have both AABR and OAE equipment available, use of a dual screening method is recommended as it may be most effective in identifying hearing loss in these infants at risk for later learning issues. Infants need to be referred for audiological assessment to include diagnostic ABR if either screening is not passed. If an infant fails both OAE and AABR screening, diagnostic ABR prior to discharge would be optimal. If scheduling is not possible, or if the infant's status or family situation does not allow for

testing prior to discharge, audiological assessment with diagnostic ABR needs to be completed on an outpatient basis as soon as medically feasible. If only one hearing screening technology is to be utilized in the NICU, recent JCIH guidelines (2007) recommend that it be the AABR.

Passing newborn hearing screening does not guarantee that hearing will remain normal nor does it eliminate the need to monitor the infant's or child's speech and language skills. Audiological re-evaluation during early childhood is recommended when parents/caregivers are concerned about hearing and/or speech/language development as well as for those infants with risk factors for emergent hearing loss.

Documentation

Minnesota Statute 144.966 requires the following documentation

- Screening results need to be recorded in the infant's medical record.
- Screening results need to be communicated to the parents of the infant so that they are well-understood.
- Screening results must be communicated to the infant's primary care physician in writing.
- Screening results must be reported to the MDH.

In order to improve follow-up for infants with failed hearing screens

- Families of infants who refer on the hearing screening should be provided with information about why their baby may not have passed the hearing screening, the importance of follow-up. In addition, a follow-up appointment for a pediatric audiological evaluation should be scheduled.
- For infants who refer, the primary care physician and MDH should be promptly notified of the follow-up audiology appointment to facilitate timely follow-up and services.

TRANSFERS

If an infant is transferred, the hospital that discharges the infant home is responsible for screening

NON-HOSPITAL BIRTHS

According to MN statute 144.966, a physician, nurse, midwife, or other health professional attending a birth outside a hospital or institution shall provide information, orally and in writing, as established by the Department of Health, to parents regarding places where the parents may have their infant's hearing screened and the importance of the screening.

RE-ADMITS

Infants re-admitted to the hospital during the first month of life who have conditions associated with potential hearing loss (e.g., hyperbilirubinemia, meningitis, sepsis) need to have the hearing screen repeated prior to discharge. Because of the high incidence of neural hearing loss associated with significantly elevated bilirubin, these infants should be referred for audiological assessment to include ABR measures.

Follow-up and Case Management

EHDI is part of a continuum of care that progresses from screening to assessment to amplification (if elected) to educational intervention. Assessment referrals need to be made to audiologists with expertise in pediatric physiological and behavioral assessment and management. See the Minnesota Department of Health (MDH) website for Audiology Regional Centers that provide pediatric diagnostic assessments and habilitation services in Minnesota (www.health.state.mn.us/newbornscreening).

The National recommended time line for hearing screening and follow-up includes: complete screening no later than one month corrected age or as soon as medically feasible for screening, assessment no later than three months corrected age or as soon as medically feasible for assessment/diagnosis, and amplification (if elected) and intervention no later than six months corrected age or as soon as medically feasible for intervention. With prompt referral and follow-up, Minnesota children will receive appropriate care and services even earlier.

Middle Ear Effusion

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 3 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 exists, thereby impacting the course of both medical and educational intervention.

Positive Risk Factor (JCIH 2007 clarification document)

The timing and number of hearing re-evaluations for children with risk factors (Appendix A) should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. Infants who pass the neonatal screening but have a risk factor should have at least 1 diagnostic audiology assessment by 24 to 30 months of age. Early and more frequent assessment may be indicated for children with cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture-positive postnatal infections associated with sensorineural hearing loss; for children

who have received ECMO or chemotherapy; and when there is caregiver concern or a family history of hearing loss.

Without an adequate follow-up plan, even the best EHDI program is ineffective. Refer to the current Minnesota Department of Health (MDH) Infant [Assessment](#) and Pediatric [Amplification](#) Guidelines.

QUALITY ASSURANCE

Components of a quality assurance program include data management, screener performance, site performance, outcome measures, and follow-up compliance. The goal of quality assurance is information management and accountability to the following stakeholders:

- Family and the infant
- Screeners
- EHDI manager
- Clinical and educational audiologist
- Primary Care Provider (Medical Home)
- Hospital
- State of Minnesota
- Otolaryngologists
- Parents
- Medical and Educational Specialists
- Advocates

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

Appendix B

Hearing & Speech Milestones

Birth to 3 months	<ul style="list-style-type: none">• startles to loud sounds• quiets or smiles when spoken to• seems to recognize your voice and quiets if crying• increases or decreases sucking behavior in response to sound• makes pleasure sounds (cooing, gooing)• cries differently for different needs• smiles when sees you
3 to 6 months	<ul style="list-style-type: none">• looks or turns toward a new sound• responds to changes in tone of voice• vocalizes excitement and displeasure• enjoys rattles and other toys that make sounds• babbling sounds more speech-like (such as ooh, aah, and ba-ba)
6 to 12 months	<ul style="list-style-type: none">• enjoys games like peek-a-boo and pat-a-cake• responds to his/her own name, telephone ringing, someone's voice, even when not loud• knows words for common things (cup, shoe) and sayings ("bye-bye")• imitates different speech sounds• starts to respond to requests such as "come here"• looks at things or pictures when someone talks about them• has one or two words (bye-bye, dada, mama). although they may not be clear

Appendix C

HEARING SCREENING RESULT AND FOLLOW-UP PROCESS

(a) Pass Both Ears, No Risk Indicators

- ✓ Passes screen in both ears (AABR or both OAE and AABR).
- ✓ Does not have any risk indicator for hearing loss (see risk indicators in Appendix A).
- ✓ Parents receive information (oral and written, culturally appropriate) regarding developmental milestones for auditory, speech, and language skills. In addition, parents should be encouraged to return for audiological evaluation if their child is not meeting these milestones, or if they have concerns about their child's hearing at any point in the future.

(b) Pass Both Ears, Positive Risk

- ✓ Passes screen in both ears (AABR or both OAE/AABR). However, infant has one or more risk factors for hearing loss (see risk indicators in Appendix A).
- ✓ Infant is referred and scheduled for repeat hearing screen at three months of age.
- ✓ Parents receive information (oral and written) regarding developmental milestones for auditory, speech, and language skills. In addition, parents should be encouraged to return for audiological evaluation if their child is not meeting these milestones, or if they have concerns about their child's hearing at any point in the future.
- ✓ If the three month hearing re-screen is normal, a complete audiological evaluation should occur at six months developmental age, using visual reinforcement audiometry (VRA).
- ✓ If follow-up testing is abnormal at three or six months, diagnostic ABR is performed as soon as feasible.

(c) Does Not Pass in One or Both Ears, or Incomplete Test

- ✓ Does not pass AABR hearing screen in one or both ears or does not pass OAE or AABR (when dual technology is utilized) in one or both ears.
- ✓ Testing cannot be completed in one or both ears.
- ✓ Infant is referred and scheduled for diagnostic ABR as soon as medically feasible. When possible, diagnostic ABR should be completed while an inpatient or scheduled and completed prior to three months of age.

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