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MINNESOTA EARLY HEARING DETECTION AND INTERVENTION (EHDI) PROGRAM

Guidelines for Infant Audiologic Assessment

Approved, November 2008

Introduction

Newborns should have hearing screening completed before hospital discharge by either OAE or screening ABR. Re-screening if indicated should be completed no later than one month of age. For babies who have been in the NICU, screening should be completed by one month corrected age or when it is medically feasible. See MDH Guidelines for Hearing Screening in the Well Baby Nursery and MDH Guidelines for Hearing Screening in the Special Care Nursery and NICU.

If results are abnormal, the infant will be referred for diagnostic audiological evaluation which should be provided by audiologists with expertise in evaluation of infants and young children with hearing loss. The goals are to provide audiologic diagnosis with ear specific information regarding degree, configuration, and type of hearing loss as soon as possible, but no later than three months of age, and to initiate intervention as soon as possible, but no later than six months of age. For infants who have required NICU care, evaluation and referral should be accomplished using the infants' corrected age and as soon as medically possible. Complete diagnostic evaluation should not be delayed solely due to middle ear dysfunction. Infants who pass the screen but have risk indicators for hearing loss should be referred for audiological assessment. JCIH risk factors for hearing loss are provided at the end of this document.

All testing, follow-up, and tracking procedures must be consistent with current Minnesota statutory requirements.

PEDIATRIC TEST BATTERY

When evaluating infants and young children, it is necessary to cross-check results by using a battery of electrophysiologic and behavioral tests. It is also important to seek information from and listen to parents/caregivers and intervention specialists who can provide valuable information about the child's functional use of hearing and speech/language/developmental progress. The child's success is dependent upon a team approach involving parents and professionals. The following tools should be used to assess infants likely to have hearing loss.

1. CHILD'S MEDICAL and DEVELOPMENTAL HISTORY:

- a) prenatal/perinatal/postnatal history
- b) family history of childhood sensorineural hearing loss
- c) auditory responsiveness, developmental milestones
- d) cognitive, physical (including vision & hearing), social or emotional, communication & adaptive development.

2. OBJECTIVE/ELECTROPHYSIOLOGIC TESTS:

Each ear should be tested, even if the newborn hearing screening result was reported as a unilateral refer.

a) Diagnostic OAE:

OAE testing in addition to ABR testing is needed to cross-check ABR findings and to differentiate between cochlear and retrocochlear disorders.

- i. Transient Evoked Otoacoustic Emissions (TEOAEs)
 - Input stimulus level should be 78 – 82 dB SPL.
 - Pass/refer criteria specific to the test equipment is suggested.
 - Use evidence based normative data for interpretation of test results
- ii. Distortion Product Otoacoustic Emissions (DPOAEs)
 - Use L1=65/L2=55dB SPL input stimulus levels.
 - Pass/refer criteria specific to the test equipment is suggested.
 - Use evidence based normative data for interpretation of test results (see Gorga, et. al., 2005).

b) Tympanometry:

- i. 1000 Hz probe tone should be used for infants four months corrected age and younger.

- ii. Test results should be interpreted using available normative data and other clinical findings. (see Margolis, et al, 2003)
- iii. For infants over four months corrected age, the standard immittance battery becomes more reliable and valid.
- iv. Otoscopic inspection of the ear canal should be carefully timed to not awaken the infant

c) Diagnostic ABR:

If the infant has failed hospital screening and outpatient rescreen, diagnostic ABR should be scheduled. Repeated outpatient rescreening, even if there is suspected middle ear dysfunction, should not be conducted as this can delay timely diagnosis of hearing loss. Instead, **proceed to a DIAGNOSTIC ABR.** Persistent fluid does not contraindicate diagnostic ABR testing to determine hearing status. Infants in the NICU for >5days who do not pass newborn hearing screening should be referred directly to an audiologist for follow-up.

- i. Air conduction clicks should be presented through insert earphones, using a 15 to 20 msec window, and obtaining a latency/intensity function. Suggested starting point is 70 - 75 dBnHL (65dBnHL if testing is completed unседated so as not to awaken the infant), so that Waves I, III, and V can be clearly identified, then decrease to threshold. Wave III may be more prominent than wave V in the young infant which may reflect auditory immaturity rather than hearing loss and should be interpreted with caution. Wave V must be replicable at threshold, necessitating at least two runs at each intensity level. Age specific infant norms must be used for interpreting data including evaluation of absolute and interpeak wave latencies for Waves I, III, and V.
- ii. Change in stimulus polarity (i.e., condensation and rarefaction) at a high intensity is needed to evaluate neural integrity and to differentiate between cochlear and retrocochlear etiology.
- iii. Minimally, low frequency (500 Hz) and higher frequency (4000 Hz) tonebursts need to be part of the diagnostic ABR in order to provide frequency specific information necessary for amplification fitting. It is desirable to test additional frequencies if possible (1000 Hz, 2000 Hz). Be aware that in infants less than 3 months of age poor response to the 500 Hz tonebursts is a common finding due to neurological immaturity, and is not necessarily indicative of hearing loss.

For toneburst measurement, use a 20 to 30 msec window and a protocol proven to effectively limit spectral splatter. Again, Wave V needs to be replicable at threshold, and since toneburst responses are often less replicable than clicks, three or four repetitions may be necessary to obtain clear replication.

- iv. If results are abnormal, bone conducted responses should be part of the test battery. An abnormal bone conduction result in an infant under four months of age should be interpreted with caution because of the nature of the young infant's skull.

d) **Sedation:**

- i. In order for the ABR to be accurate, it is essential that the baby be sleeping so that there is no bodily or eye movement causing electrical activity that could obscure the auditory evoked potentials.
- ii. Infants under 3 months of age often will fall asleep allowing for testing to be completed without sedation. Evidence suggests that this is best practice.
- iii. For older babies, as well as those who do not routinely sleep well during the day, sedation as prescribed by the managing sedation physician is indicated.
- iv. **SEDATION SHOULD ONLY BE ADMINISTERED ON SITE AT THE TESTING FACILITY.** Each facility needs to have a comprehensive Sedation Policy, which outlines the steps required to ensure patient safety. Adequate sedation is needed for completion of the full diagnostic protocol which is essential for accurate assessment, and for planning the child's educational future.
- v. If any component cannot be completed unседated, sedation is justified to provide the most reliable route for obtaining the information needed to initiate intervention in a timely manner.
- vi. Since not all facilities are equipped to safely provide sedation, it may be feasible to attempt unседated diagnostic evaluations, and to refer those requiring sedation to other centers.

e) **Emerging Technologies:**

Emerging technologies can be used to add information but should not substitute diagnostic ABR.

- i. **Auditory Steady State Response (ASSR)** is an emerging technology that may be incorporated into the test battery when adequate methodologies and normative data are available. ASSR is useful as a supplement to ABR data, particularly in cases of severe to profound hearing loss, but is NOT at this time recommended as a substitute for ABR testing.
- ii. **Middle Ear Reflectance** is an additional emerging technology and may be incorporated into the test battery when methodologies and normative data are available.

- iii. **Cortical Evoked Auditory Potentials (CAEP)** recording is emerging as a useful technology in assessment of infants with Auditory Neuropathy/Dys-Synchrony (AN/AD) in whom the ABR is abnormal, thus limiting threshold estimation. While the CAEP data is not used for threshold determination, presence or absence of CAEP to conversational speech may assist in case management (Pearce et al., 2007.) In addition, cortical response recording may be valuable in assessing cortical reorganization and benefit from amplification and/or cochlear implantation. (Sharma, 2007)

3. BEHAVIORAL ASSESSMENT:

Essential to cross-check electrophysiologic data and provide measure of functional use of hearing

- a) Behavioral testing, using visual reinforcement audiometry (VRA), should be included as part of the audiological evaluation if the child is over 4 months of age, taking into account that at less than 6 months developmental age there will be many babies unable to reliably condition to this task. It is important to note that children with global developmental delay and certain syndromes may also be less able to condition to this task.
- b) Use of insert earphones is ideal in order to obtain ear specific data; if insert testing is not successful, loudspeaker assessment in combination with physiological individual ear data can be used to proceed with the diagnostic process.
- c) Use of behavioral observation audiometry (BOA) is helpful in providing information about a young infant's auditory awareness and presence or lack of startle response, but cannot be used to determine thresholds information or the presence / absence of significant hearing loss.
- d) If hearing loss is diagnosed, ongoing behavioral audiological assessment and follow-up is required as the baby matures to obtain additional information and to monitor hearing levels. Refer to Minnesota Department of Health (MDH) Guidelines for Pediatric Amplification Follow-up section www.health.state.mn.us/newbornscreening.

FOLLOW-UP AND CASE MANAGEMENT

If results indicate abnormal hearing, the family should be counseled regarding the results and implications of the hearing loss and written literature should be provided.

1. If the audiological assessment results are abnormal, and it appears that **conductive** involvement is likely, the primary care provider should be contacted so that a plan of treatment/ referral to an otolaryngologist with expertise in examination of infants and young children can be developed for the infant.

Although persistent middle ear effusion might delay the evaluation timeline several weeks, **diagnostic audiological evaluation must not be repeatedly postponed solely due to middle ear dysfunction and still 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 the existence of an underlying sensorineural hearing loss, thereby impacting the course of both medical and educational intervention.

If medical intervention is recommended, hearing should be re-evaluated after treatment is completed. If results continue to be abnormal, discussion of treatment options to include the use of hearing aids (if elected) should begin (see MDH Guidelines for Pediatric Amplification). Treatment should not be repeatedly postponed due to persistent middle ear dysfunction.

2. If the audiological assessment results are abnormal, and the hearing loss appears **sensorineural**, the primary care provider should be contacted and a referral to a otolaryngologist should be made. Audiological results and treatment options should be discussed with the family, and referral to early intervention should be made with parental consent. Once medical clearance is obtained, use of amplification (if elected) can be initiated (see MDH Amplification Guidelines.) If assessment is based on ABR data, results should be clearly marked as to whether they have been converted to estimated hearing levels. Infants with confirmed hearing loss should have at least one exam by an ophthalmologist experienced in evaluating infants (JCIH, 2007). A genetics consultation should be offered to the families of infants with confirmed hearing loss (JCIH, 2007).
3. If unilateral hearing loss is identified, case management should be similar to that listed above for a bilateral hearing loss.
4. If the audiological assessment results are abnormal, and **auditory neuropathy/dysynchrony** is suspected, ie, otoacoustic emissions (OAEs) and/or a cochlear microphonic(CM) are present and ABR indicates abnormal brainstem function, the primary care provider should be contacted to discuss results and recommendations.

Once medical/neurological evaluation has ruled out retrocochlear lesion, results and implications should be discussed with the parents. Information regarding typical auditory, speech, and language developmental milestones should be provided with behavioral assessment of the infant using visual reinforcement audiometry at six months developmental age. If parents express concerns sooner, age appropriate audiological re-evaluation should be conducted.

As the impact of auditory neuropathy/dysynchrony on auditory, language, and speech skills can vary tremendously from one individual to another, behavioral audiological follow-up is a critical piece in determining the child's needs for intervention. If age appropriate auditory skills are not developing, a referral to early intervention should be made. If auditory skills are not developing normally

and visual reinforcement audiometry suggests a hearing loss, a trial period with amplification using loaner hearing instruments should be initiated. If adequate progress is not seen with consistent amplification, referral for cochlear implant evaluation would be an appropriate consideration.

On rare occasions, evidence of improvement in ABR thresholds has been noted and may be due to neurological maturation. Patients born prematurely or with histories of hyperbilirubinemia should be carefully monitored for change in audiological findings.

5. Infants with any degree of bilateral or unilateral permanent hearing loss (sensorineural or conductive) should be referred to determine eligibility for early intervention services. Referral to Help Me Grow (Infant and Toddler Early Intervention/Part C) should be made within two business days of hearing loss diagnosis with signed consent. In cases of persistent or chronic conductive hearing loss (including loss due to effusion) not receiving or responding to medical treatment, referral to early intervention should be offered to the family.

Minnesota's "Help Me Grow" (Infant and Toddler Early intervention or Part C) is designed for children birth to three years of age who have delays in their development or a diagnosed condition that has a high probability of resulting in developmental delay. Services are designed to meet the unique developmental needs of each child and their family. Families work together with local service providers to access services and resources to help their child grow and develop. Accessing these services as early as possible will ensure best developmental outcomes for the child. These services are offered at no cost to the family and are provided under federal law.

Please refer to the Minnesota Department of Health Guidelines for Audiologist Referral to Early Intervention and Family-to-Family Support for more information regarding these referrals.

6. The diagnosing audiologist should also facilitate family connection to parent-to-parent support, such as Minnesota Hands & Voices, with parental consent. Please refer to the Minnesota Department of Health Guidelines for Audiologist Referral to Early Intervention and Family-to-Family Support.
7. If the audiological assessment results are normal, and the infant has a positive risk factor for progressive or late-onset hearing loss (Appendix A), the primary care provider should be made aware of the need for on-going follow-up and the results and implications should be discussed with the parents.

The JCIH 2007 statement specifically recommends, "The timing and number of hearing re-evaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. National recommendations indicate 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.”

Early and more frequent assessment can be interpreted as every 6 months, or more, depending on the clinical findings and concerns. Children with conditions known to be associated with middle ear dysfunction, such as Down syndrome and cleft palate, should have hearing monitored at 6 month intervals in the first few years of life or until language is well established.

DOCUMENTATION

1. Audiologic assessment results need to be clearly communicated to the parents of the infant verbally and in writing.
2. Audiologic assessment results and/or fail appointments should be communicated to the infant’s primary care physician in writing.
3. Audiologic assessment results and/or failed appointments must be reported to the MDH as required by MN Statute 144.966. Fax all reports within one week of assessment.

FACILITY AND PROFESSIONAL CONSIDERATIONS

1. If a particular facility does not have the equipment or staff with appropriate training or experience to conduct the above described evaluation, the infant should be referred elsewhere for the initial audiological evaluation. The diagnosing pediatric audiologist can then work with the local audiologist on the amplification fitting and follow-up so that the family will be able to receive much of the ongoing audiological care near their home. Please refer to the Minnesota Department of Health Guidelines for Pediatric Amplification .
2. Every effort should be made to offer audiologic care that is child and family centered so that care may be provided in an order and manner that is tailored to an individual family.

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*

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