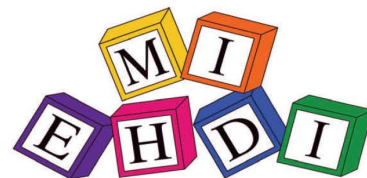


Otolaryngologic Evaluation of Infants with Hearing Loss



Michigan Early Hearing Detection
and Intervention Program

This document is a product of the Michigan Early Hearing Detection and Intervention (EHDI) Advisory Committee's desire to ensure families access to pediatric otolaryngology services that are in agreement with the current Joint Committee on Infant Hearing (JCIH) recommendations. This guideline is not intended as a sole source of guidance in evaluating children with hearing loss. It is not intended to establish a protocol for all children with this condition or to supersede clinical judgment, and may not provide the only appropriate approach to diagnosing and managing infants and children with hearing loss.

Reference: Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. The on-line version of this document can be found at <http://www.pediatrics.org/cgi/content/full/120/4/898>.

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Introduction

The Joint Committee on Infant Hearing (JCIH) endorses early detection of and intervention for infants with hearing loss. The goal of early hearing detection and intervention is to maximize linguistic competence and literacy development for children who are deaf or hard of hearing. Without appropriate opportunities to learn language, these children will fall behind their hearing peers in communication, cognition, reading, and social-emotional development. Such delays may result in lower educational and employment levels in adulthood.

The role of the Michigan Department of Health and Human Services Early Hearing Detection and Intervention (EHDI) Program is to promote the following 1-3-6 goals and to guarantee seamless transitions for infants and their families through this process.

- All newborns will be screened for hearing loss no later than **1 month of age**, preferably before hospital discharge.
- Those who do not pass screening should have a comprehensive audiological evaluation no later than **3 months of age**.
- Infants with confirmed hearing loss should receive appropriate intervention no later than **6 months** of age.

The success of EHDI programs depends on families working in partnership with professionals as a well coordinated team. The roles and responsibilities of each team member should be well defined and clearly understood. Essential team members include families, the birth hospital, the medical home (i.e., pediatricians or primary health care professionals), audiologists, otolaryngologists, speech-language pathologists, educators of children who are deaf or hard of hearing, and other early intervention professionals involved in delivering EHDI services.

Background Information on Hearing Screening

Physiologic measures must be used to screen newborns and infants for hearing loss. Such measures include Otoacoustic Emissions (OAE) and Automated Auditory Brainstem Response (A-ABR) testing. Both OAE and A-ABR technologies provide noninvasive recordings of physiologic activity underlying normal auditory function, both are easily performed in neonates and infants, and both have been successfully used for Universal Newborn Hearing Screening (UNHS). However, there are important differences between the two measures. OAE measurements are obtained from the ear canal by using a sensitive microphone within a probe assembly that records cochlear responses to acoustic stimuli. Thus, OAEs reflect the status of the peripheral auditory system extending to the cochlear outer hair cells. In contrast, A-ABR measurements are obtained from surface electrodes that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli delivered via earphone. A-ABR measurements reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway. Both OAE and A-ABR screening technologies can be used to

detect sensory (cochlear) hearing loss. As with any medical screening, hearing screening technology has limitations. The most common are listed below:

- Both technologies may be affected by transient outer or middle ear dysfunction, resulting in a “failed” screening result in the presence of normal cochlear and /or neural function.
- OAEs are generated within the cochlea, therefore OAE technology cannot detect neural (eighth nerve or auditory brainstem pathway) dysfunction. Thus, neural conduction disorders or auditory neuropathy/dyssynchrony without concomitant sensory dysfunction will not be detected by OAE testing. For this reason, A-ABR is the preferred screening method for any infant with a Neonatal Intensive Care Unit (NICU) history.
- Screening protocols are designed to detect characteristics of hearing loss most commonly identified in children. As a result, some types of hearing loss (i.e., mild or isolated frequency region losses) will be missed using standard screening protocols.
- The newborn screening assesses the auditory system at the time of screening and cannot accurately predict which infants may develop hearing loss.

Screening technologies that incorporate automated-response detection are necessary to eliminate the need for individual test interpretation, to reduce the effects of screener bias or operator error on test outcome, and to ensure test consistency across infants, test conditions, and screening personnel. When statistical probability is used to make pass/fail decisions, as is the case for OAE and A-ABR screening devices, the likelihood of obtaining a pass outcome by chance alone is increased when screening is performed repeatedly. Therefore, multiple rescreens prior to hospital discharge increase the likelihood of a false negative result.

Screening Protocols in the Well-Infant Nursery

Use of either technology in the well-infant nursery will detect peripheral (conductive and sensory) hearing loss of 40 dB or greater. When A-ABR is used as the single screening technology, neural auditory disorders can also be detected. Some programs use the same technology for the initial screen and when necessary, a repeat screening at the time of discharge from the hospital. Other programs use a combination of screening technologies (OAE testing for the initial screening, followed by A-ABR for rescreening, i.e., 2-step protocol) to decrease the fail rate at discharge and the subsequent need for outpatient follow up. With this approach, infants who do not pass an OAE screening but subsequently pass an A-ABR test are considered a screening “pass.” Infants in the well-infant nursery who fail A-ABR testing should not be rescreened by OAE testing and “passed,” because such infants are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dyssynchrony.

Screening Protocols in the Neonatal Intensive Care Unit (NICU)

The 2007 JCIH position statement includes neonates at risk of having neural hearing loss (auditory neuropathy/ auditory dyssynchrony) in the target population to be identified in the NICU, because there is evidence that neural hearing loss results in adverse communication outcomes. Consequently, the JCIH recommends ABR technology as the only appropriate screening technique for use in the NICU. For infants who do not pass A- ABR testing in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, comprehensive evaluation, including diagnostic ABR testing, rather than for general outpatient rescreening.

Outpatient Rescreening for Infants Who Do Not Pass the Birth Admission Screening

Many well-infant screening protocols will incorporate an outpatient rescreening within 1 month of hospital discharge to minimize the number of infants referred for follow-up audiological and medical evaluation. The outpatient rescreening should include the testing of both ears, even if only one ear failed the inpatient screening.

As stated previously, infants in the well-infant nursery who fail A-ABR testing should not be rescreened by OAE testing and “passed,” because such infants are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dyssynchrony.

For infants who do not pass an outpatient rescreen, referral should be made directly to an audiologist for more comprehensive evaluation. A diagnostic test battery should be able to determine an underlying sensorineural component to the hearing loss, even with the presence of middle ear fluid. Repeated outpatient rescreens are discouraged, as this can delay the diagnosis of permanent sensorineural hearing loss.

Mandatory Reporting in Michigan-Public Act 31 of 2006

Effective February 23, 2006, Public Act 31 mandated that health professionals in charge of care for newborn infants (includes hospitals, health departments, audiologists, and any person administering hearing tests or screenings) must report results to the State of Michigan. To view the details of this Public Act, please visit www.michigan.gov/documents/mdch/2006-PA-0031_274659_7.pdf.

Please send all hearing results to the Early Hearing Detection and Intervention (EHDI) Program to fax number 517-763-0183. Please use the EHDI reporting form which can be obtained on www.Michigan.gov/ehdi by clicking on newborn screening. Contact the EHDI Program at 517-335-8955 if you have any questions.

Otolaryngologic Evaluation of Infants with Hearing Loss

Otolaryngologists are physicians and surgeons with special expertise in the diagnosis and management of hearing disorders. The purpose of the otolaryngologic evaluation is to determine the etiology of hearing loss, to identify associated conditions, and to provide recommendations for medical/surgical management where appropriate as well as to provide medical clearance for amplification. As such, every infant with confirmed hearing loss should be evaluated by an otolaryngologist. The otolaryngologic evaluation may include the following elements:

- Comprehensive history to identify risk-factors for childhood hearing impairment.

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. Infants who pass the neonatal screening but have a risk factor should have at least one diagnostic audiology assessment by 24 to 30 months of age per JCIH recommendations.

More frequent monitoring is recommended (customized may be considered every 6 months until age three years) for the following risk factors that are highly associated with delayed on-set hearing loss: 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 extracorporeal membrane oxygenation (ECMO) or chemotherapy; and when there is caregiver concern or a family history of childhood hearing loss.

- Complete head and neck examination.
- Careful assessment of otologic health, including the status of the auricle, external ear, tympanic membrane, and middle ear.
- Head and neck examination with attention to features which might suggest an underlying syndromic diagnosis, of which approximately 600 conditions are known to be associated with hearing loss.
- Review of prior audiologic evaluation.

For infants from birth to a developmental age of approximately 6 months, a comprehensive audiological battery should include:

- A frequency specific assessment of the ABR using air conducted click and tone bursts and bone-conducted tone bursts when indicated. When permanent hearing loss is detected, frequency specific ABR testing is needed to determine the degree and configuration of hearing loss in each ear for fitting of amplification devices, if this is a chosen option.
- Click-evoked ABR testing using both condensation and rarefaction single-

polarity stimulus, if there is an absent ABR or if there are risk indicators for neural hearing loss (auditory neuropathy/auditory dyssynchrony) such as hyperbilirubinemia or anoxia, to determine if a cochlear microphonic is present.

- Distortion product or transient evoked OAEs.
- Tympanometry using a 1000-Hz probe tone.
- Clinician observation of the infant's auditory behavior as a cross-check in conjunction with electrophysiologic measures. Behavioral observation alone is not adequate for determining whether hearing loss is present in this age group, and it is not adequate for the fitting of amplification devices.

For subsequent testing of infants and toddlers at developmental ages of 6 to 36 months, the confirmatory audiological test battery should include:

- Behavioral audiometry (either visual reinforcement or conditioned-play audiometry, depending on the child's developmental level), including pure-tone audiometry across the frequency range for each ear and speech detection and recognition measures.
- OAE testing.
- Acoustic immittance measures (tympanometry and acoustic reflex thresholds).
- ABR testing if responses to behavioral audiometry are not reliable or if ABR testing has not been performed in the past.

- **Consideration of Additional Testing**

If after a careful history and physical examination, the etiology of the hearing loss is not apparent, consideration should be given to additional diagnostic testing.

Congenital Cytomegalovirus (CMV).

- Estimated to account for between 20-30% of congenital hearing loss (depending on prevalence of CMV infection).
- If hearing loss is confirmed within the first 4 weeks of life, testing for CMV should be considered.
- Outside of this time frame, the prevalence of postnatal infection makes interpretation of positive results difficult.

Temporal Bone Imaging.

- Temporal bone imaging identifies diagnostic information in approximately 30% of children with hearing loss, and may provide prognostic information or be used to assess potential candidacy for surgical interventions.
- Traditionally, this imaging is performed with high-resolution computed tomography. Magnetic resonance imaging has been introduced more recently and may provide complementary information.

Other Testing.

- Historically, an extensive battery of laboratory testing was frequently performed. However, this battery is expensive and has low yield in identifying the etiology of hearing loss, and is therefore not routinely recommended.
- Additional testing should be based on the judgment of the treating physician on the basis of their evaluation.

• **Referral for Genetic Evaluation**

A referral to the Medical Geneticist can provide families with information on the etiology of the hearing loss, prognosis for progression, associated disorders (e.g., renal, vision, cardiac), and the likelihood of recurrence in future offspring. This information may influence the parents' decision-making regarding intervention options for their child. The medical geneticist is responsible for the following:

- Interpreting family history data.
- Clinically evaluating and diagnosing inherited disorders, if present.
- Performing and assessing genetic tests.
- Providing genetic counseling to the family.

Historically, at least 50% of congenital hearing loss has a genetic etiology. This proportion is likely to increase as the prevalence of acquired causes including infectious agents and ototoxic medications decreases. Consideration should be given to testing for mutations in GJB-2 (Connexin 26), which are responsible for approximately 50% of autosomal recessive non-syndromic hearing loss.

The brochure, "Hearing Loss, Genetics and Your Child", has contact information on local genetic services and is available at no charge by contacting the EHDI Program at 517-335-8955.

Other Medical Specialists

JCIH recommends that infants with confirmed hearing loss should have an evaluation by an ophthalmologist to document visual acuity and rule out concomitant or late-onset vision disorders such as Usher syndrome. Indicated referrals to other medical subspecialists, including developmental pediatricians, neurologists, cardiologists, and nephrologists, should be facilitated and coordinated by the primary health care professional.

- The otolaryngologist should conduct longitudinal monitoring to detect and manage coexisting otologic pathology (such as otitis media with effusion) which might impact on management of the hearing loss.

Next Steps

- Once hearing loss is confirmed, medical clearance for hearing aids and referral to *Early On*[®], (1-800 EARLY ON), Michigan's Birth to Three Program, should be initiated as soon as possible. Amplification and early intervention should not be delayed pending the outcome of the diagnostic process. Hearing aids (if a chosen option) should be fit within one month of diagnosis. Hearing aid fittings should be provided by a licensed audiologist with pediatric experience and the equipment necessary to provide adequate verification measures. Families should be provided with a copy of the medical clearance form.
- Closely monitor unilateral hearing loss. Monitoring may be considered as often as every 6 months. Audiological management will be necessary to monitor speech and language development and to assess the child's auditory needs relative to amplification or other assistive listening technology.
- Cochlear implantation is a consideration for infants with bilateral profound hearing loss.
- The family should be given information regarding the Guide By Your Side Program. This is a free service offered to connect parents of newly identified infants to other parents of children with hearing loss. Contact the EHDI Program at 517-335-8955 for more information.

Attachment

The Joint Committee on Infant Hearing, Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs (www.jcih.org) recommended the eleven risk indicators listed below that are associated with either congenital or delayed-onset hearing loss. All infants with a risk indicator for hearing loss should be referred for an audiological assessment at least once by 24 to 30 months of age. Children with risk indicators that are highly associated with delayed-onset hearing loss, such as having received ECMO or having CMV infection, should have more frequent audiological assessments.

Risk Indicators Associated with congenital, delayed-onset, or progressive hearing loss in childhood are listed below. Risk indicators that are marked with an asterisk* 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. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO*, assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires 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*.

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