GUIDELINES FOR DIAGNOSTIC AUDIOLOGIC EVALUATION

Audiologic evaluation of infants should be completed as soon as possible after a referral from the newborn hearing screening. The assessment should be completed by the time the child is three months of age to determine ear specific and frequency specific information regarding the type, degree and configuration of hearing loss. In the best interest of infants and children with hearing loss, these guidelines are strongly recommended.

Costs of hearing screening, rescreening and/or diagnostic services may be covered by third-party insurers, Medicaid, Early On, MI-Child, and/or Children's Special Health Care Services.

I. ASSESSMENT SITE REQUIREMENTS

It is recommended that infants and young children be referred to diagnostic audiologic evaluation sites having the capacity to perform the full diagnostic assessment battery as listed below. It is also recommended that audiologists, who conduct diagnostic audiologic evaluations for the pediatric population, have adequate experience in evaluating newborns and very young infants. This will help circumvent delay in diagnoses and intervention, as well as compounding anxiety for parent(s)/caregiver(s).

II. DIAGNOSTIC AUDIOLOGIC EVALUATION BATTERY

A. Obtain a complete case history containing parental/caregiver report of emerging communication and auditory behaviors.

B. Perform an otoscopic examination of the ears using great care not to scratch the ear canal.

C. Perform Behavioral Observation Audiometry (BOA) to observe the infant's responsiveness to sound. At a minimum, attempt to obtain responses for frequency modulated pure tones of 500 Hz and 3000 Hz as well as for speech. Attempt to obtain a startle response to speech.

D. Perform acoustic immittance measures, if possible, using a high frequency probe tone (tympanometry and acoustic reflex thresholds). If abnormal immittance measures are obtained, the infant should be referred to the Medical Home for diagnosis, treatment, and possible referral to an ENT for middle ear pathology prior to further testing. A one-month follow-up appointment should be scheduled to determine middle ear status and complete testing to rule out sensorineural hearing loss (Holt, Margolis, & Cavanaugh, 1991; Sprague, Wiley & Goldstein, 1985).

E. Evoked Otoacoustic Emission (OAEs), Transient Evoked or Distortion Product.

F. Diagnostic ABR (with conscious sedation if needed).

1. High intensity click stimulus to assess latency and morphology of Waves I, III, V, and interwave intervals of Waves I-III, III-V and I-V.
2. Threshold search with click stimulus.

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3. If the presence of middle ear pathology is suspected based on immittance test results, case history, or otoscopic exam, perform Wave V threshold search with bone-conducted click stimuli. If air-conducted and bone-conducted Wave V thresholds differ by 20 dB or more, the presence of conductive pathology should be suspected.
4. If possible, obtain tone burst thresholds at 500, 1000 and 4000 Hz bilaterally. (Stapells, 2000; Gorga, 1999; Stapells & Oats, 1997; Hood 1995; Gorga, Kaminski, Beauchaine, & Bergman, 1993; Ysunza & Cone-Wesson, 1987; Weber, 1982).

III. RESULTS

A. Normal/Within Normal Limits (infants with no risk indicators):

1. Discuss the test results with the parent(s)/caregiver(s). Include information on acquired, delayed onset, and progressive hearing loss and the need to monitor. Provide language and speech stimulation materials and milestone data.
2. Prepare a complete and concise report of test results, recommendations and referrals. Parent(s)/Caregiver(s) should receive a copy of the diagnostic test results at the time of diagnoses with any written recommendations.
3. Obtain a release of information for all facilities to which the infant will be referred for services or reports will be sent.
4. Report results to the Medical Home and the Michigan Department of Community Health, Early Hearing Detection and Intervention Program (FAX: (517) 335-8036).

B. Normal/Within Normal Limits ("at risk" infants and young children):

1. Retest "at risk" infants and young children at 3-month intervals for the first year and every 6 months until age 3 years. The JCIH outlines the following risk indicators for use with neonates or infants (29 days through 2 years):
   a. Parental/Caregiver concern regarding hearing, speech, language, and or developmental delay.
   b. Family history of permanent childhood hearing loss.
   c. Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian tube dysfunction.
   d. Postnatal infections associated with sensorineural hearing loss including bacterial meningitis.
   e. In-utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis.
   f. Neonatal indicators specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECMO).

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g. Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher's syndrome.
h. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome.
i. Head trauma.
j. Recurrent or persistent otitis media with effusion for at least 3 months.

2. Prepare a complete and concise report of test results, recommendations and referrals. Parent(s)/Caregiver(s) should receive a copy of the diagnostic test results at the time of diagnoses with any written recommendations.
3. Obtain a release of information for all facilities to which the infant will be referred for services or reports will be sent.
4. Report results to the Medical Home and the MDCH/EHDI Program (FAX: (517) 335-8036).

C. Hearing Loss Identified

The Joint Committee on Infant Hearing defines the targeted hearing loss for universal newborn hearing screening programs as permanent bilateral or unilateral, sensory or conductive hearing loss, averaging 30 to 40 dB or more in the frequency region important for speech recognition (approximately 500 through 4000Hz).

1. Prepare a complete and concise report of test results, recommendations and referrals. Parent(s)/Caregiver(s) should receive a copy of the diagnostic test results at the time of diagnoses with any written recommendations and contact information.
2. Obtain a release of information for all facilities to which the infant will be referred for services or reports will be sent.
3. Report results to the Medical Home and the MDCH/EDHI Program (FAX: (517) 335-8036).
4. Counsel parent(s)/caregiver(s) on the effects of the hearing loss, communication, and the need for immediate intervention. If appropriate, earmold impressions may be taken at the diagnostic audiologic evaluation to decrease delay in amplification intervention.
5. Provide parent(s)/caregiver(s) with a copy of the *Services For Children Who Are Deaf or Hard of Hearing; A Guide to Resources for Families and Providers* (DCH-0376), which can be obtained, free of charge, by calling MDCH/EHDI Program at (517) 335-9560.
6. Refer parent(s)/caregiver(s) to educational services for children with hearing loss (Special Education Director of the Home School District or Intermediate School District). Contact information is found in the *Services For Children Who Are Deaf or Hard of Hearing; A Guide to Resources for Families and Providers* (DCH-0376). Parent(s)/caregiver(s) should be strongly urged to visit different educational programs for the hearing impaired within their intermediate school district.

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7. Refer parent(s)/caregiver to *Early On* ® Michigan County Coordinator (1-800-EARLY-ON or 1-800-327-5966) for family coordination of services.
8. Counsel parent(s)/caregiver(s) about family support through the Family Support Network (1-800-359-3722).
9. Counsel parent(s)/caregiver(s) about financial assistance through Children's Special Health Care Services (1-800-359-3722).
10. Discuss the importance of a genetic evaluation to determine etiology and other possible health related issues.
11. Schedule audiologic follow-up monitoring appointment.
12. Refer parent(s)/caregiver(s) to the Medical Home for:
    a. Medical intervention for transient conductive component (i.e., middle ear effusion) and/or referral to otolaryngologist for medical treatment with a follow-up audiologic evaluation after one month to determine status of middle ear and to rule out sensorineural hearing loss.

       AND/OR (a. and/or b.)

    b. Referral to an Otolaryngologist for medical clearance for hearing aid use if parent(s)/caregiver(s) wish to pursue this intervention option. For audiologic results indicating auditory neuropathy (ABR fail OAE pass), the appropriateness of hearing aid use may be hard to determine (Hood, 2000).
    c. Discussion of the importance of a genetic evaluation to determine etiology and other possible health related issues.
    d. Referral for follow-up audiologic evaluation to monitor hearing status based on suggested audiological follow-up schedule.
    e. Review of *Guidelines for Newborn Hearing Services,* section on Guidelines for Medical Home and Guidelines for Early Intervention, for further information to ensure appropriate hearing health care for the infants and young children identified with hearing loss (Guidelines available through the MDCH/EHDI Program at (517) 335-9560).

IV. HEARING AID OPTION

It is important for infants and children with hearing loss to have timely fitting of appropriate amplification when this intervention option is chosen. It is suggested that audiologists follow the recommendations of the Pediatric Working Group when providing amplification for infants and children with hearing loss. A systematic, quantifiable, and evidence-based approach with current technologies should be used when fitting children. The provision for audible, reliable, comfortable, and undistorted amplification for children can be determined through a four-stage process involving assessment, selection, verification, and validation (Bess, Chase, Gravel, Seewald, Stelmachowicz, Tharpe, & Hedley-Williams, 1994).

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A. Assessment:
   1. Ear specific and frequency specific information regarding the type, degree, and configuration of a hearing loss should be obtained.
   2. Once the child receives medical clearance for hearing aid use, it is important to review the physician's notes to obtain information regarding the possibility of progressive hearing loss, evidence of multiple impairments, and structural deformities.
   3. Predictive methods for obtaining loudness level of discomfort (LLD) measurements with corrections for children should be determined.
   4. Real-ear measurements should be obtained, as soon as possible
   5. Unaided sound field levels, speech detection levels and speech perception levels should be obtained as soon as possible.

B. Selection Considerations:
   1. Maximizing acoustic information for sound awareness and spoken language development is imperative, therefore binaural hearing aids should be recommended for binaural hearing loss, unless there is evidence that binaural amplification can not be tolerated.
   2. Small behind-the-ear hearing aid (s) with half moon ear hooks for ear specific and ear level amplification are preferred. Body style hearing aids may also be considered.
   3. Soft material earmolds are preferred and should be remade as the child grows.
   4. Hearing aids with the greatest flexibility in the electroacoustic parameters and signal processing should be considered. The specific method for selection used for fitting amplification for infants and young children should be based on rigorous research into the theoretical aspects of the specific method. References to selection considerations can be found in Pediatric Working Group Position Paper for Amplification for Infants and Children with Hearing Loss (Bess et al., 1994) and DSL method (Stelmachowicz, Dalzell, Peterson, Kopu, Lewis & Hoover, 1998).
   5. Safety features such as tamper resistant battery and volume control covers are recommended.
   6. Extended warranty and loss and damage coverage are recommended
   7. Parent/Caregiver kit with the contents including, but not limited to: stethoscope, dri-aid kit, battery tester, brush and pick are recommended.
   8. Retention system for keeping the hearing aid(s) secure to the head and to the body to prevent loss.
   9. Feedback management system.
   10. Directional/omni-directional microphone switching option.
   11. Remote FM microphone option and compatibility with other assistive device technology (e.g., direct audio input, telecoil/microphone-telecoil switching options).
   12. Insurance policies (Children's Special Health Care Services, Medicaid, Medicaid HMO and others third party insurers) should be contacted to

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determine specific hearing aid reimbursement coverage for children identified with hearing loss.

C. Verification:

After the hearing aid has been selected and preset, the frequency-gain and output characteristics should be verified through probe-tube measurements (Stelmachowicz & Seewald, 1991; Moodie, Seewald & Sinclair, 1994). Aided sound field measurements for children over 6 months of age can also provide verification information but limitations should be considered (e.g., cooperation from the child, limited frequency resolution, poor test-retest, and limited gain and output level information from nonlinear hearing aids.

D. Validation:

The purpose of validation is to demonstrate benefits/limitations of a child's aided listening abilities for perceiving the speech of others, the child's own speech, and sound awareness (Bess, et al, 1994). This information is usually obtained over time in various settings and from various people. A validation tool designed for the pediatric population is the Infant-Toddler: Meaningful Auditory Integration Scale (Zimmerman-Phillips, Osberger & Robbins, 1997).

E. Amplification Management and Maintenance

Families should be counseled regarding the need for audiologic follow-up to monitor the function, use, and appropriateness of amplification. Families should be counseled regarding the need to perform daily listening checks and the need for audiological reevaluation of a child's amplification. Periodic audiological re-evaluation should include a recheck within 1-2 weeks after the initial fitting, and at 3 month intervals for children age 0-3 years; every 6 months for ages 4-6 years; and every 6-12 months for school age children. The frequency of follow-up may need to be increased if fluctuation/progression of the hearing loss is noted and/or if progress is questioned. Ongoing communication between the clinical audiologist and the members of the early intervention team is critical.

F. Other Considerations:

1. Assistive Technology

   Families should be counseled regarding the benefits and limitations of assistive technology use in the home. Amplification options include but are not limited to, FM systems and alerting devices.

2. Amplification Orientation

   Families should be counseled regarding hearing aid care, safety issues, troubleshooting techniques, incorporating use of hearing aids into the family routine, and other important topics related to hearing aid care (Elfenbein, 2000).

3. Amplification Follow-up

   Families should be counseled regarding the need for amplification follow-up. A child's external ears change rapidly therefore earmolds will need to be remade to prevent acoustic leakage from occurring. When earmolds are remade, Real Ear to Coupler Difference measurements should be reevaluated to account for acoustic changes in the earmolds and for developmental changes in the child's external ear. During earmold

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appointments, hearing aids should be evaluated through listening checks and electroacoustic checks in order to ensure proper function.

V. SUGGESTED AUDIOLOGIC FOLLOW-UP AND MEDICAL MANAGEMENT

Families should be counseled regarding the need for audiologic and medical follow-up. An immediate audiologic evaluation should be scheduled when there is concern related to change in hearing or hearing aid function.

A. Bilateral sensorineural hearing loss and permanent conductive hearing loss:
   1. Age 0-3: Every 3 months, after hearing loss is confirmed.
   2. Age 4-6: Every 6 months, if intervention progress is satisfactory.
   3. Age 6 years or older: Every 6-12 months if progress is satisfactory.

B. Transient conductive hearing loss (e.g., otitis media with effusion), unilateral or bilateral:
   Should be monitored after medical treatment (completion of antibiotic treatment, PE tubes, etc.), and/or at least on a 3-4 month basis until resolved and normal hearing is confirmed.

C. Unilateral hearing loss (sensorineural or permanent conductive):
   Infants with unilateral hearing loss should be monitored every 3 months during the first year, then on a 6-months basis after the first year, to rule out changes in the normal hearing ear.

VI. PERSONNEL REQUIREMENTS

At a minimum, an audiologist should complete the diagnostic audiologic evaluation. Any audiologist accepting infants and young children for initial diagnostic audiologic evaluation should have the ability to complete the procedures identified above. The following is preferred:

A. Certification
   A qualified audiologist holds a Certificate of Clinical Competence from the American Speech-Language-Hearing Association (ASHA) or holds board certification by the American Board of Audiology or is a clinical fellow in audiology under the direct supervision of a certified audiologist.

B. Experience in assessment of newborns and young infants
   1. Knowledge and experience in administering and interpreting ABRs and OAEs for the pediatric population.
   2. Affiliation with a medical facility in which sedation can be administered and monitored safely is needed.

C. Experience in recommending, fitting, and dispensing pediatric amplification
   1. Experience in making earmold impressions for the pediatric population.
   2. Ability to provide hearing aids on a trial basis.
   3. Ability to provide loaner hearing aids.
   4. Access to repair of hearing aids for infants and young children in a timely manner.

D. Knowledge of auditory development and auditory habilitation options

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