

## *Audiological Diagnostic Protocol*

This protocol is intended as a guide for audiologists who are performing diagnostic evaluations on infants. It is assumed that these children have not passed an initial and repeat hearing screening. (\* Denotes optional procedure, but highly recommended for more complete information.)

1. Case history/parent observation report.
2. Otoscopic inspection.
3. ABR assessment:
  - a. Obtain a threshold search to a click ABR in 10 dB steps.
  - b. If no neural response is identified, compare responses obtained to rarefaction and condensation clicks presented at 80 to 90 dB nHL using a fast click rate (>30 second). If a response (i.e., cochlear microphonic) is observed, an auditory neuropathy may be present.
  - c. \* Obtain a threshold response to a 500 Hz and 3000 Hz tone pip ABR.
  - d. \* Obtain a bone conduction click ABR.
4. \* Obtain acoustic immittance measures (tympanograms, physical volume, reflexes) using a high frequency probe tone.
5. Obtain an evoked OAE (either DP or Transient) to verify cochlear function.
6. Perform Behavioral Observation Audiometry (BOA) to a speech stimulus and/or a 500 and 2000 Hz tone or noise, by air conduction and bone conduction. Identify any minimal responses and attempt to obtain startle responses. Perform Visual Reinforcement Audiometry (VRA) with infants five months of age and older.
7. Discuss the results and follow-up recommendations with the parents; discuss all options (amplification, intervention, communication choices) with parents. Give parents a copy of the "Georgia Resource Guide for Families of Children with Hearing Loss or Deafness."
8. Prepare written report, interpreting the test results and describing the diagnostic profile.

9. If hearing loss is confirmed, recommend referral to an ENT for evaluation and to obtain an otologic diagnosis and clearance for amplification if indicated (with parental permission).
10. Disseminate written report and other information to the infant's primary care physician, Children 1<sup>st</sup>, and other care providers/agencies, as requested by the parents.

The CPT codes per procedure are listed below for reimbursement purposes.

- Otitic evaluation
  - New patient - CPT code 99201
  - Established patient - CPT code 99212
- Auditory Brainstem Response (ABR)
  - CPT code 92585
- Acoustic immittance measurements
  - Tympanometry - CPT code 92567
  - Acoustic reflexes - CPT code 92568
- Otoacoustic emissions (OAE)
  - Limited - CPT code 92587
  - Comprehensive or diagnostic - CPT code 92588
- Behavioral observation audiometry / Visual response audiometry
  - CPT code 92579

### ***Medical Diagnostic Protocol***

The following is a recommended protocol for medical evaluation once an infant has been identified with hearing loss. This is just to serve as a guide. Many medical practitioners may perform other or additional procedures and referrals.

#### **Identify Historical Factors**

##### Prenatal

- Ototoxic medication exposure
- Complications during pregnancy
- Immunization to Rubella
- Syphilis screening

- Maternal drug use
- Frequent spontaneous abortions

#### Perinatal

- Craniofacial abnormalities
- Birth weight < 1500 grams
- Hyperbilirubinemia
- In-utero infection such as TORCH (Toxoplasmosis, Rubella, Cytomegalovirus, Herpes)
- Low apgar score

#### Family

- Family history of childhood sensorineural hearing loss

### **Physical Examinations**

- Ear specific micro exam
- Morphologic features
- Poor growth and / or microcephaly
- Abnormal neurologic examination

### **Laboratory Evaluations Considered**

- Urine culture for CMV at < 3 weeks
- Specific Syphilis Test
- Renal
- Thyroid
- Infectious disease - CMV, Toxoplasmosis, Rubella, Syphilis
- EKG
- Ophthalmology
- CT high resolution of temporal bone
- MRI brain and CPAs
- Audiological evaluation (see "Audiological Diagnostic Protocol", pg. 27)
- Neurological
- Developmental

### **Referrals**

- Genetics
- Audiology
- Children 1<sup>st</sup> (entry point for public health programs, such as Babies Can't Wait, Children's Medical Services)
- District UNHSI Coordinator
- Georgia PINES (Georgia Parent Infant Network for Educational

- Services)
- Speech-Language Pathology
  - ENT
  - Family Counseling/Education
  - Social Services Agencies

### ***Risk Indicators for Hearing Loss***

Since 1972, the Joint Committee on Infant Hearing (JCIH) has identified specific risk indicators that are often associated with infant and childhood hearing loss. Recently, the JCIH issued their Year 2000 Position Statement and Guidelines (see <http://www.audiology.com/positions/jcih-early.php#iie2> for full text) and further delineated hearing risk indicators. Risk indicators can be divided into two categories: those present during the neonatal period and those that may develop as a result of certain medical conditions or essential medical interventions in the treatment of an ill child.

Based on data from an eight year study of permanent childhood hearing impairment in Great Britain and the recent NIH multicenter study, the JCIH risk indicators have been modified for use in neonates (birth through 28 days) where universal newborn hearing screening is not yet available. These indicators include:

- a. An illness or condition requiring admission of 48 hours or greater to a NICU.
- b. Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss.
- c. Family history of permanent childhood sensorineural hearing loss.
- d. Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
- e. In-utero infection such as cytomegalovirus, herpes, toxoplasmosis, or rubella.

The Joint Committee on Infant Hearing (JCIH) recommends using risk indicators with neonates and infants (29 days through 2 years). The following indicators put an infant at risk for progressive or delayed-onset sensorineural and/or conductive hearing loss. These infants should receive audiologic monitoring every six months until the child's third birthday. The indicators are:

- a. Parental or 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).
- g. Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher's syndrome.
- h. Neuro-degenerative 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 three months.

There is a possibility that some of the important indicators, such as family history of hearing loss, may not be known at the time of screening in a universal newborn hearing screening program. The identification of these indicators should occur during early well-baby visits by the primary care physician/provider. Infants who are identified with these risk indicators need to be monitored closely for normal communication developmental milestones during routine medical care. The parents of infants with these risk factors should be given the brochure "Have You Heard?" which emphasizes the importance of periodic monitoring.