



Protocols for Diagnostic Audiological Assessment Virginia Early Hearing Detection and Intervention Program Virginia Department of Health

These protocols are recommended for use in the assessment of infants who are referred from newborn hearing screening programs or from a primary medical care provider (PCP). The original protocols were developed in 1999 by a Task Force comprised of six audiologists, licensed in Virginia, with extensive knowledge and experience in the screening and diagnosis of newborns and young children. This is a revision of those original protocols. The judgment regarding a facility's ability to provide diagnostic audiological services to infants is equipment-driven, as there is currently no certification for pediatric audiology. Quality audiological assessment is the diagnosis of normal hearing or hearing loss specific to each ear. The assessment of hearing loss includes identification of type and severity, and whenever possible the nature of the hearing loss, as well as identification of treatment options.

An audiologist licensed by the Commonwealth of Virginia and employed by a facility designated by the Virginia Department of Health (VDH) Early Hearing Detection and Intervention (EHDI) Program as an Approved Diagnostic Audiological Facility should provide these diagnostic services.

The EHDI Program goals are to identify congenital hearing loss by three months of age and to assure enrollment in appropriate early intervention services, including amplification if appropriate, by six months of age.

General Information

Virginia law requires that hospitals give the parent information (in writing) regarding a child's hearing screening status prior to hospital discharge. Both the parent and PCP should have been told one of the following:

- The screen result is a refer (did not pass); the infant should receive follow-up hearing screening within one month.
- The infant was not screened (missed); the infant should receive a hearing screening within one month.
- The screen result is a pass; however, because one or more risk indicators for developing hearing loss later have been identified, your infant should have a hearing screening every six months until the age of three.

The Virginia EHDI Program tracks infants who 1) refer on the hearing screening prior to discharge from the hospital, 2) pass but are at-risk for progressive or delayed-onset hearing loss, 3) were not screened prior to discharge (missed), or 4) were transferred to an out-of-state hospital. VDH will send a letter to the parent of these infants and the PCP (if reported by the

hospital) to insure that they have received the appropriate recommendations at discharge. VDH will also send letters and make follow-up phone calls to the parent of any child who is diagnosed with hearing loss, to assure that they have access to the information they need to make informed decisions for their child.

The Joint Committee on Infant Hearing (JCIH) Year 2000 Position Statement identifies risk indicators that often are associated with infant and childhood hearing loss. These indicators place an infant at risk for progressive or delayed-onset sensorineural and/or conductive hearing loss. The Virginia EHDI Program tracks infants and young children who have been identified with one or more of these risk indicators. Any infant with one or more risk indicators, who has passed the birth or initial screen, should receive audiological monitoring every six months until age three years.

The indicators listed below are to be used where universal newborn hearing screening occurs.

- Parental or caregiver concern regarding hearing, speech, language and/or developmental delay.
- Family history of permanent childhood hearing loss.
- Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or Eustachian tube dysfunction.
- Postnatal infections associated with sensorineural hearing loss including bacterial meningitis.
- In-utero infections such as cytomegalovirus, herpes, rubella, syphilis and toxoplasmosis.
- Neonatal indicators-specifically Hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation and extra corporeal membrane oxygenation (ECMO).
- Syndromes associated with progressive hearing loss such as Neurofibromatosis, osteopetrosis, and Usher's syndrome.
- Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies such as Freidreich's ataxia and Charco-Marie-Tooth syndrome.
- Head Trauma.
- Recurrent or persistent otitis media with effusion for at least 3 months.

Some indicators, such as otitis media or certain syndromes, may not be determined during the course of the hospital stay. Therefore, infants and young children who have late-onset or late-identified risk indicators should be monitored for speech, language and hearing developmental milestones by the PCP during well-child visits.

Audiologists who provide audiological services have the following responsibilities as outlined in the regulations governing the program:

- Provide the screening or evaluation results to the parent and to the child's primary medical care provider.
- Offer referral to early intervention or education programs.
- Give resource information to the parent of any child found to have a hearing loss, including, but not limited to, the degrees and effects of hearing loss, communication

options, the importance of medical follow-up, and agencies and organizations that provide services to children with hearing loss and their families.

- Send the Virginia Department of Health a report including test results, diagnosis and recommendations, within two weeks of the patient visit.

The VDH Report of Follow Up form and instructions can be downloaded from the Web site, www.vahealth.org/hearing.

The Virginia EHDI Program maintains a list of Approved Diagnostic Audiological Facilities. In order to be considered as an approved facility, a practice must have both otoacoustic emissions (OAE) and auditory brainstem response (ABR) equipment located in the office, must have a Virginia-licensed audiologist on staff, and must agree to the terms and conditions listed on the form including submitting a report to VDH. A list of approved sites is placed on the VDH Web site, is communicated to all Virginia birthing hospitals, and is printed on the back of every letter VDH sends to parents and PCP.

The Virginia EHDI Program developed the Information for Parents of Children With Hearing Loss, Virginia's Resource Guide for Parents, in collaboration with the Virginia Department of Education, Virginia Department for the Deaf and Hard of Hearing, and Virginia Early Intervention System, Department of Mental Health, Mental Retardation and Substance Abuse Services. This guide, available in English, Spanish as well as a videotape version presented in American Sign Language and open captions, was written for families of children with hearing loss. The purpose is to provide all parents with unbiased information regarding: 1) hearing loss, its effects and implications; 2) communication options; 3) amplification; 4) state agency programs and services; and, 5) local, state, and national resources. These items can be obtained by contacting the Virginia EHDI Program at 804-864-7717. The English and Spanish versions also can be downloaded from the program Web site, www.vahealth.org/hearing.

Protocols

Initial Visit (within 1 month of hospital discharge)

OAE required as the initial retest procedure. (OAE technology is sensitive to outer hair cell dysfunction)

- Obtain Evoked OAE 2000-5000Hz.
- Obtain all information requested on the VDH Report of Follow Up form; this information is crucial for tracking purposes.
- Obtain parent report of observed behavioral responses to auditory stimuli.
- Perform otoscopic examination.
- Ear specific results must be obtained for each ear.

OAE Pass or Pass with Follow-up

- If the infant passes (three of four frequencies tested), then the evaluation is complete unless the audiologist determines a need for comprehensive evaluation based on medical history.
- The family should receive information about hearing, speech and language milestones and information regarding risk indicators if present.

- For infants who pass but are at-risk for progressive or delayed-onset hearing loss, the parent should be counseled regarding the need for audiological follow-up and continued monitoring.

OAE Refer

- If the results are a refer on one or both ears, or
- OAE is normal but auditory neuropathy/desynchrony is suspected, evaluate using ABR.

If facility is unable to perform ABR at same visit, schedule diagnostic ABR at VDH-approved diagnostic facility within four weeks.

Assessment Visit (minimal requirements for audiologic assessment)

ABR (ABR technology reflects the activity of the cochlea, auditory nerve, and auditory brainstem pathways)

- Otosopic examination.
- Threshold click ABR (25dBHL-20dBHL or lower) for each ear.
- Bone conduction threshold if click stimuli are elevated (25-40 dBHL).
- Ear specific bone conduction if thresholds are elevated bilateral and/or an asymmetric loss is evident.
- Threshold tone burst to 500 Hz, as well as 3000 Hz or 4000 Hz.
- If facility has technology available to perform frequency-specific evoked potential testing, this may enhance diagnostic and (re) habilitative services for the infant.
- Repeat OAEs to again evaluate cochlear function and rule out auditory neuropathy.
- Counsel parents as to results and follow-up recommendations.

If hearing loss is confirmed

- Refer to an otolaryngologist for otologic evaluation and to obtain medical clearance for amplification.
- Discuss importance of genetic referral for testing and/or counseling. (Protocols for Medical Management, VDH 2001, recommend that children with hearing loss be referred for genetic testing and counseling.)
- Refer to Part C early intervention services regardless of type or severity of the loss.
- Refer for amplification and assistive technology; if appropriate, provide information on cochlear implant.
- Schedule follow-up audiologic assessment in three months.

Suggested Additional Assessment

- Acoustic immittance measures using high frequency (660 or 800 Hz) tone if infant is less than four months of age. A 226 Hz probe tone may be used with reliability for age four months and older.
- Acoustic Reflex thresholds at 500 Hz, 1000 Hz and 4000 Hz.
- Behavioral observation to speech stimulus, 500 and 2000 Hz (minimally). Visual reinforcement audiometry can be effectively used for infants six months or older as a

component of the evaluation. If the child will tolerate them, insertion earphones may be utilized for ear-specific information.

- Warbled pure tone or narrow band noise in sound field. Identify any minimal responses and attempt to obtain startle response.

Test Criteria

(Click) Transient evoked Otoacoustic emissions (TEOAE) Stimulus- air conduction click
Intensity - 80 ± 3 dB SPL

Pass Criteria

- Frequencies 2000 Hz through 5000Hz
- three of four frequencies having reproducibility minimally: 70% @ 2400, 3200, 4000 and 5000 Hz

Distortion Product Otoacoustic Emissions (DPOAE) Stimulus - pure tone complex
Intensity - maximum levels <70 dB SPL

Pass Criteria

- $F2 = 2000, 3000, 4000$ and 5000 Hz
- Three of four frequencies have a distortion product ($2F1-F2$) amplitude ≥ 6 dB than measured noise floor levels

Auditory Brainstem Response (ABR)

Pass Criteria

- Air conduction click stimulus for both ears
- Replicable wave V response thresholds less than or equal to 25-20dBHL

ABR Bone Conduction Testing

Stimulus and recording parameters

- Alternating Click, 21.1/sec
- Window 15 msec
- Low Filter 30
- High Filter 1500
- Gain 100,000
- Earlobe/mastoid non-inverting electrode

Bone Conduction 2-Channel Recording (Ipsilateral/Contralateral)

Setup

- Right-Ref I
- Left-Ref II
- High Forehead - Active jump between Ch I & Ch II
- Low Forehead - Ground

Bone Conduction stimulus: Ear-specific two-channel recording

- At higher intensity levels, evaluate Ipsilateral vs. Contralateral recording.

- If there is a Wave I on the Ipsilateral in conjunction with the absence of a Wave I on the contralateral ear, then it can be assumed that the response is being generated from the stimulated ear.
- Record Wave V responses down to threshold.

Recording Parameters Guidelines for Tone Burst ABR

Air Conduction

- EEG Channels:
 - Vertex-left mastoid (Cz-M1)
 - Vertex-right mastoid (Cz-M2)
- EEG filter:
 - High Pass: 20-30 Hz
 - Low Pass: 1500-3000 Hz
- One-channel recordings (Cz-ipsilateral mastoid)
 - (Slope: 6 or 12 dB/octave, analog)
- Window: 25 msec
- Polarity: Alternating onset
- Rate: 39.1/s
- Ipsilateral Noise: Band-reject (notched noise, with 1-octave-wide notch centered on the tone frequency)
- Contralateral Noise: White noise 30dB below tone masking level (when possible)
- Stimuli 2-1-2 linear
 - 500 Hz: 4ms rise/fall, 2 ms plateau
 - 1000 Hz: 2 ms rise/fall, 1 ms plateau
 - 2000 Hz: 1 ms rise/fall, 0.5 ms plateau
 - 4000 Hz: 0.5 rise/fall, 0.25 ms plateau

(Adapted from Stapells and Oats, 1997)

Unilateral Hearing Loss

Unilateral hearing loss refers to a hearing loss in one ear, whether mild or profound, congenital or acquired, that warrants a definitive diagnosis. Parents have the right to know what their child's hearing level is in each ear. Depending on the etiology of the unilateral loss, there may be associated medical conditions that require further medical attention and management.

Children with a unilateral hearing loss are at risk for progressive and/or bilateral hearing loss. Unilateral hearing loss has significant implications for a child's development, socialization, and success in school including:

- May exhibit delays in speech and language development.
- Will have difficulty localizing sounds; therefore, crossing the street or riding a bike create safety concerns.
- May exhibit behavior and/or social problems.
- May have difficulty with following directions.
- Is at risk for failing a grade in school.

- May lag substantially behind in math, language, cognitive and social functioning as compared to their peers.
- Is distractible and may appear inattentive.
- May show signs of fatigue as the school day progresses.
- May benefit from assistive listening devices in the classroom.

Children with a unilateral hearing loss should receive:

- Routine audiologic evaluations at six-month intervals to monitor for progressive or delayed-onset hearing loss.
- Referral to early intervention services for developmental evaluation.
- Noise protection counseling to protect the "good ear".
- Prompt medical management of middle ear disorders.

Auditory Neuropathy/Desynchrony

According to JCIH a small population of infants, particularly those cared for in the NICU, may be at increased risk for neural conduction and/or auditory brainstem dysfunction, including auditory neuropathy. Auditory neuropathy is a recently identified disorder, characterized by a unique constellation of behavioral and physiologic auditory test results. Behaviorally, children with auditory neuropathy have been reported to exhibit mild to profound hearing loss and poor speech perception. Physiologic measures of auditory function demonstrate the finding of normal otoacoustic emissions (suggesting normal outer hair cell function) and atypical or absent auditory brainstem response (suggesting neural conduction dysfunction). Reports suggest that those at increased risk for auditory neuropathy are infants with a compromised neonatal course, infants with hyperbilirubinemia and children with a family history of childhood hearing loss. Currently, neither the prevalence of auditory neuropathy in newborns nor the natural history of the disorder is known. Audiologic and medical monitoring of infants at risk for auditory neuropathy is recommended, every six months until the age of three. Infants with these disorders can be detected only by the use of **OAE** and auditory **ABR** technology **in combination**.