Virginia Early Hearing Detection & Intervention Program

PROTOCOLS FOR PRIMARY CARE PROVIDERS

2018

VDH VIRGINIA DEPARTMENT OF HEALTH

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Protocol for Primary Care Providers
Virginia Early Hearing Detection and Intervention Program
Virginia Department of Health

This document provides guidance and recommended procedures for Primary Care Providers (PCP) to implement requirements that are specified in the *Code of Virginia*, Section 32.1-64 12VAC5-80[1] and *Regulations for the Administration of the Virginia Hearing Impairment Identification and Monitoring System*[2].

These PCP protocols were first developed in 1999 and revised in 2004 and 2011. The 2018 revision represents the best practice that the Virginia Early Hearing Detection and Intervention Program (VEHDIP) Advisory Committee recommends based on the policy statement *Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs, Joint Committee on Infant Hearing*[3] and other relevant sources such as the Centers for Disease Control and Prevention (CDC), and National Center on Birth Defects and Developmental Disabilities. The VEHDIP Advisory Committee consists of representatives from relevant groups including, but not limited to, primary care physicians, otolaryngologists, audiologists, speech pathologists, nurses, parents and educators of the deaf and hard of hearing. It has been unanimously agreed that Virginia diagnostic PCP protocol standards have followed, and should continue to follow, an exceptional model of evidence-based practice and should reflect an excellence beyond minimal standards of care. This document reflects that philosophy.

It is important to recognize that newborn hearing screening is only one component of a comprehensive approach to the management of childhood hearing loss. It is recommended that the PCP inform the parents or guardian of follow-up diagnostic services, counseling, Part C Early Intervention programs, and parental education and family support organizations. This comprehensive process should be administered by a multidisciplinary team including, but not limited to, audiologists, physicians, educators of the deaf and hard of hearing, speech/language pathologists, nurses, parents and educators of deaf and hard of hearing.

VEHDIP goals are to identify congenital hearing loss by 3 months of age following the CDC *1-3-6 methodology*:

1 – All newborns will be screened for hearing loss **before 1 month** of age.

3 – All newborns who have failed their hearing screen will receive a diagnostic evaluation **before 3 months** of age.

[1] To access the *Code of Virginia* citation, go to: https://law.lis.virginia.gov/admincode/title12/agency5/chapter191/section260/

[2] To access the *Regulations for the Administration of the Virginia Hearing Impairment Identification and Monitoring System*, go to http://leg1.state.va.us/000/reg/TOC12005.HTM#C0080

[3] To access the *Executive Summary for JCIH Year 2007 Position Statement: Principles & Guidelines for Early Hearing Detection and Intervention Programs* go to http://www.asha.org/policy/PS2007-00281.htm

6 – All infants diagnosed with hearing loss will be enrolled in early intervention services **before 6 months** of age

I. Virginia Early Hearing Detection and Intervention Program Overview

The Virginia Early Hearing Detection and Intervention Program is required to collect, maintain, and evaluate hearing screening data. Program staff must provide follow-up, including communicating with the parent or guardian to assure that they have the information needed to seek timely and appropriate follow-up services. They should provide training and technical assistance to hospitals, audiologists and PCPs. Lastly, the program is required to review and evaluate the surveillance system including follow-up rates, false-positive rates, false-negative rates, referral mechanisms and effectiveness of tracking.

Best practice guidelines for the VEHDIP include:

- Monitor hospital data and rate of reporting.
- Provide epidemiological analysis of the data for planning and program management purposes.
- Provide training and technical assistance to birthing centers.

Providers performing initial hearing screening are required to perform this screen prior to hospital discharge, and should report results within seven days but no later than fourteen days after discharge to the Virginia Department of Health (VDH) and to the PCP from whom the infant will receive care after discharge. They must provide written information to the parent that includes the benefits of newborn hearing screening, the procedures used for screening, and recommendations for further follow up. A list of audiologic providers can be accessed on EHDIPALS.org or a list of pediatric audiologists can be found on the back of all the VEHDIP letters to parents and PCPs.

A. Reporting

The required methodology for reporting initial newborn hearing screenings to the VDH is via the Virginia Infant Screening and Infant Tracking System (VISITS) database. If a provider is unable to enter results into VISITS, they may use the Audiological Reporting Form—including test results, diagnosis, and recommendations—and submit to the VDH within seven days but no later than fourteen days of the testing. The Audiological Reporting Form, including risk indicators and instructions, can be downloaded from the VDH website www.vahealth.org/hearing.

Best practice guidelines for PCPs:

- Review the hearing screening results and risk indicator findings from hospitals.
- Referral of infants who fail the initial screen immediately to a pediatric audiologist for a repeat hearing screening or diagnostic assessment.
- Obtain and review the diagnostic evaluation results from audiological providers.
- Ensure infants with a hearing loss diagnosis receive a referral for evaluation by an otolaryngologist, at least one examination to assess visual acuity by a pediatric ophthalmologist, and for genetic testing and counseling.

- Ensure that parents are informed and/or advised about enrollment into Part C Early Intervention, preferably by six months of age. Visit the Infant & Toddler Connection of Virginia website www.infantva.org for information on referral processes and contacts.
- Screen and refer infants diagnosed with persistent conductive hearing loss secondary to otitis media with effusion for otolaryngology and audiological assessment for confirmation of diagnosis to avoid delayed diagnosis of sensorineural hearing loss.
- Refer directly to an audiologist for rescreening NICU infants who do not pass an automated ABR. For rescreening, a complete screening of both ears is recommended even if only one ear failed the initial screening.
- All infants readmitted in the first month of life, when there are conditions associated with potential of hearing loss (such as exchange transfusions for hyperbilirubinemia or confirmed sepsis) need to have audiological testing.
- All infants and toddlers should have an objective developmental screen with a standardized tool at 9-, 18-, and 24-to-30 months of age or at any time the healthcare professional or family has concerns about speech or language skills.
- Refer infants and toddlers who do not pass the speech-language portion of a standardized developmental screen, or whose behavior/responses cause concern regarding hearing or language, for speech-language evaluation and audiology assessment.

Persons providing hearing or audiological services to infants **after** hospital discharge are required to provide the screening or evaluation results to the parent or guardian and to the child's primary care provider. If a PCP provides office-based hearing screenings, the PCP is responsible to ensure that the screening is done on the same equipment as was used for the initial hearing screening. The PCP should **only conduct one** rescreening in the office.

A variety of technologies are available to identify hearing loss in the first days of life. These techniques are physiological measures of the status of the peripheral auditory system that are highly correlated with hearing status. The two methodologies generally accepted as effective for universal newborn screening are:

1) **Auditory brainstem response (ABR) –** reflects the activity of the cochlea, auditory nerve, and auditory brainstem pathways. **For use in NICU and/or well baby nursery.**

2) **Otoacoustic emissions (OAE) –** reflects sensitivity to outer hair cell dysfunction. **For use in well baby nursery.**

Infants who fail hearing screening in one or both ears using ABR testing should not be re-screened using OAE testing. OAE is not sufficient to rule out Auditory Neuropathy. Due to the increased incidence of auditory neuropathy in the neonatal intensive care unit (NICU) patient population, newborns who receive this level of care should have both ears screened using ABR testing prior to discharge or transfer to a lower level of newborn services. Immitance testing may help distinguish conductive or mixed hearing loss from sensorineural hearing loss (SNHL).

II. Interpretation of Hearing Screen Results

Table I summarizes information on hearing screen results and associated primary healthcare provider (PHP) actions.

Table I: Interpretation of Hearing Screen Results

Screening Result	Interpretation by PCP	PCP Follow-up Recommendations
Passed Screen	Infant passed in both ears	Monitor communication and language development.
Passed with Risk	Infant passed in both ears but is at risk for progressive or late onset hearing loss	Inform the parent of the need for a diagnostic audiological assessment between 12 to 24 months of age. Perform developmental surveillance and screening consistent with American Academy of Pediatrics recommendations and refer as needed. Early and more frequent assessments may be indicated for children with CMV syndromes and syndromes at risk for late onset of hearing loss.
Missed Screen	Infant was not screened before discharge from the hospital.	Advise the parents that screening needs to be completed immediately.
Failed Screen	Infant did not pass in one or both ears	Infants who fail Automated Auditory Brainstem Response (AABR) testing should not be rescreened by OAE testing, because they are presumed at risk for a subsequent diagnosis of auditory neuropathy/dyssynchrony. Infants should be tested again using AABR no later than one month after hospital discharge. Ensure that any infant failing their hearing screen or rescreen is referred to a pediatric audiologist as soon as possible. Refer to EHDI-PALS at http:ehdipals.org for resources.
Infant not born in a hospital	Any infant born outside of hospital facility, e.g., home birth	Refer for initial hearing screening as soon as possible after birth.
NICU Infants	Infants admitted to the NICU	All Infants admitted to NICU should be screened with AABR. If infants are admitted to the NICU > 5 days and pass the AABR then further follow up is

		needed for potential delayed onset of hearing loss. See Pass with Risk above for recommendations on follow up.

A flow chart depicting the newborn hearing screening and follow-up process is in **Attachment 1**.

III. PCP Recommended Actions for Infants with Confirmed Hearing Loss

Every infant with confirmed hearing loss needs medical evaluation to determine the etiology of hearing loss, identify related physical conditions, and make recommendations for treatment and referrals to other resources. Relevant professionals should give resource information to the parent (or guardian) of any child found to have a hearing loss, including but not limited to, the degrees and effects of hearing loss, communication options, amplification options, the importance of medical follow-up, and agencies and organizations that provide services to children with hearing loss and their families. The primary healthcare provider should thoroughly document family history and the mother's prenatal risk factors. This will support ongoing care for the child as well as monitoring other children in the family. Document:

- Prenatal conditions: ototoxic medication exposure, pregnancy complications, immunization status of mother for rubella, maternal status for syphilis, maternal drug and/or alcohol use, and history of frequent spontaneous abortions.
- Family history: hereditary childhood sensorineural hearing loss and family members with permanent hearing loss with onset before age 30 not related to trauma or medical condition.

Complete a physical examination with special attention to:

- Minor anomalies: unusual morphologic features occurring in less than 5% of the population with no cosmetic or functional significance.
- Major anomalies: dysmorphic features that cause significant cosmetic or functional abnormality, such as cleft palate, cardiac, limb, or other skeletal deformities.
- Poor growth, microcephaly, or abnormal neurological exam.

Obtain laboratory and imaging studies:

- Urine culture or oral polymerase chain reaction (PCR) swab for cytomegalovirus (CMV) before aged 2 weeks if prenatal CMV infection is suspected.
- Testing for rubella, syphilis, or toxoplasmosis consistent with history of findings and immunization status of mothers.
- EKG if cardiac condition suspected.
- Skeletal survey if growth delayed or disproportionate.
- Head CT or MRI if neurological exam abnormal. Temporal bone CT or MRI may be indicated after obtaining other laboratory or audiological testing to evaluate for inner ear abnormalities that would complicate further care recommendations (i.e. cochlear implantation).

Refer as follows:

- To otolaryngology (ENT).
- To ophthalmology/cardiology/nephrology evaluation if indicated.

- For genetic evaluation:
    - If there are significant dysmorphic features.
    - A connexin gene assay where there is a family history of deafness
    - Siblings with increased risk of having hearing loss for an audiological evaluation. Most important if moderate-to-severe-to-profound bilateral SNHL.
- To a parent to parent support organization.

IV. Risk Indicators

Birth hospitals in Virginia are responsible for determining the risk status for hearing loss on every newborn regardless of the results of the hearing screen. Risk-status data assist with monitoring for progressive, delayed-onset, and/or conductive hearing loss. VDH recognizes the risk indicators identified by the policy statement *Year 2007 Position Statement: Principles and Guidelines for Early Detection and Intervention Programs, Joint Committee on Infant Hearing* (See **Attachment 2**).
Some indicators 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 primary healthcare provider during well-child visits.

The following diagram summarizes processes that should be followed to ensure infants and children who are at risk for hearing loss receive appropriate audiological follow-up.

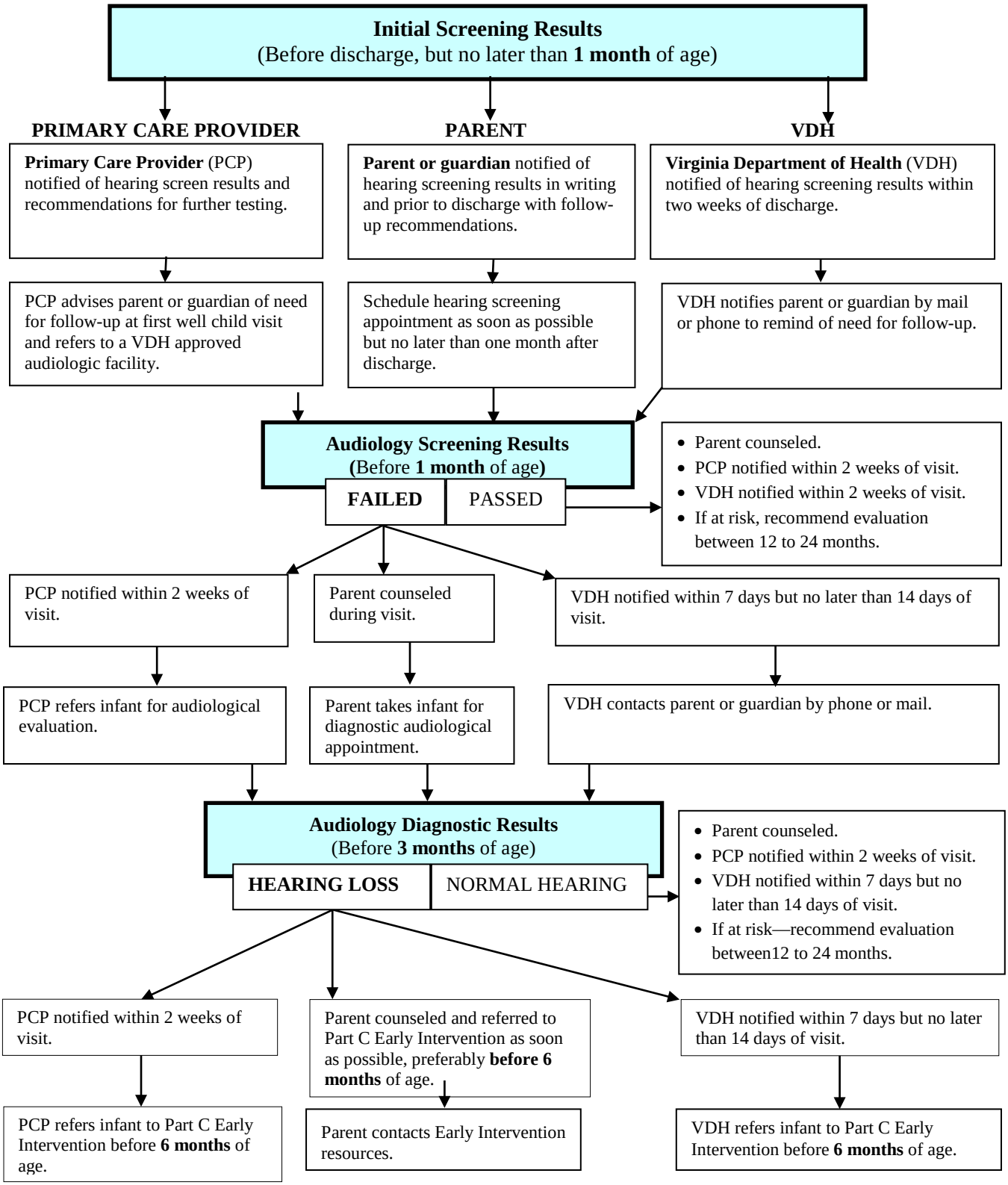
Some of these indicators are not present and/or would not be identified in the newborn period. These include parental concern and some neurodegenerative disorders or sensory motor neuropathies. These are included in the risk indicator list because parents and physicians should be informed about all indicators that can contribute to development of hearing loss beyond the newborn period. The child's physician is to report risk indicators which are identified after the newborn period to the VEHDIP in order to allow for follow-up.

V. Contacts

For more information or further assistance, families are encouraged to contact:
Virginia Department of Health
Office of Family Health Services
Virginia Early Hearing Detection and Intervention Program
109 Governor Street, 9th Floor
Richmond, VA 23219
Phone: Toll Free 1-866-493-1090 TTY 7-1-1
Fax: 804-864-7771
Website: newbornhearingtestva.com

VI. Attachments

Attachment I: Virginia Early Hearing Detection and Intervention Program Process Flow Chart



Attachment 2: Risk Indicators for Progressive or Delayed-Onset Hearing Loss

(For Use with Neonates and Infants Through 2 Years of Age)

Family history of permanent childhood hearing loss		
• Mother of child	• Grandmother of child	• 1st cousin of child
• Father of child	• Grandfather of child	• More than one relative of the same parent
• Sister of child	• Aunt of child	
• Brother of child	• Uncle of child	
Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or Eustachian tube dysfunction		
• Branchio-oto-renal (BOR)	• Stickler	• Trisomy 21 – Down syndrome
• Noonan	• Williams	• Trisomy 18 – Edwards syndrome
• CHARGE association	• Zellweger	• Trisomy 13 – Patau syndrome
• Pierre Robin	• Goldenhar (oculo-auriculo-vertebral or OAV)	• Trisomy 9 – Mosaic syndrome
• Rubenstein-Taybi	• Trisomy 8 – Warkany syndrome	
Postnatal infections associated with sensorineural hearing loss		
• Confirmed bacterial meningitis	• Confirmed viral meningitis	
In utero infections		
• Cytomegalovirus	• Rubella	• Toxoplasmosis
• Herpes	• Syphilis	• Zika
Neonatal indicators		
• Intensive care greater than (>) 5 days	• Exposure to ototoxic medications: at risk aminoglycoside exposure	• Hyperbilirubinemia requiring exchange transfusion
• Extracorporeal membrane oxygenation (ECMO)	• Mechanical ventilation	
Syndromes associated with progressive hearing loss		
• Neurofibromatosis	• Jervell & Lange-Nielson	• Usher
• Osteopetrosis	• Waardenburg	
• Alport	• Pendred	
Neurodegenerative disorders, such as		
• Hunter syndrome	• Charcot-Marie-Tooth syndrome	• Friedreich's ataxia
Head trauma requiring hospitalization		
• Basal skull/temporal bone fracture	Other – specify if chosen	
Parental or caregiver concern regarding hearing, speech, language, and/or developmental delay		
Craniofacial Anomalies		
• Pinna	• Atresia	• Choanal atresia
• Cleft palate	• Microtia	• Temporal bone anomalies
Chemotherapy		

Based on *Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs, Joint Committee on Infant Hearing.*

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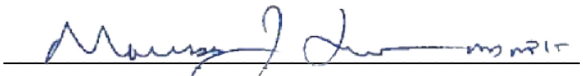
Approved by:



Marissa Levine, MD, MPH
State Health Commissioner

Date

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