

**VIRGINIA DEPARTMENT OF HEALTH
VIRGINIA'S NEWBORN HEARING SCREENING PROGRAM
PROTOCOLS FOR MEDICAL MANAGEMENT**

The focus of this document is to provide guidance and recommended procedures for best practice for infants and young children who are in need of follow-up from universal newborn hearing screening programs and for children who are found to have hearing loss.

The goal of Virginia's program is to achieve identification of congenital hearing loss in children by three months of age and enrollment in appropriate intervention by six months of age.

The primary care provider should direct and coordinate the evaluation and referral process within the child's medical home. The critical period to confirm diagnosis and begin intervention, including amplification if appropriate, is by age 6 months. Early and consistent intervention specific to hearing loss is key to achieving normal language development.

The goal, diagnosis of congenital hearing loss by three months of age and enrollment in appropriate intervention by six months of age, can be achieved when infants receive

- ◆ appropriate follow-up within the recommended time-frames by an audiologist with expertise in the evaluation of infants and young children, and
- ◆ referral to Part C early intervention for evaluation and service coordination as soon as a confirmed diagnosis of hearing loss is made.

What Are the Responsibilities Outlined in the Regulations Governing the Program?

Hospitals are responsible for:

- ◆ giving written information to the parent that includes screening procedures, indicators of hearing loss, recommendations for further testing and where the testing can be obtained;
- ◆ giving written information to the child's primary health care provider that includes the infant's risk status and/or screening results and recommendations for follow-up.

Persons who provide audiological services are responsible for:

- ◆ reporting children who fail to pass a hearing screening, children who were not successfully tested, and children identified with hearing loss to the Virginia Department of Health;
- ◆ giving parents information about hearing loss, including choices about learning communication, and referring them to local early intervention services.

VDH is responsible for:

- ◆ collecting and maintaining data from hospital screening and audiological follow-up;
- ◆ tracking children needing follow-up, including communicating with parents to assure that they have the information needed to seek timely and appropriate follow-up and services;
- ◆ providing training and technical assistance to hospital staff;

- ◆ monitoring hospital refer rates;
- ◆ conducting review and evaluation of the system including follow-up rates, false-positive rates, false-negative rates, referral mechanisms and effectiveness of tracking;
- ◆ providing epidemiological analysis of the data for planning and program management purposes
- ◆ communicating critical performance data to hospitals yearly;
- ◆ approving centers/providers of diagnostic audiological services for infants and young children

Interpretation of Screening Results

Passed screen: during routine and periodic assessment of health, monitor ongoing development of communication and language.

Pass with follow-up: child is at risk for progressive or late onset hearing loss (such as family history of hereditary childhood hearing loss or in utero infection-rubella, cytomegalovirus).

Missed screen: hospital responsible for providing mechanism by which screening can occur at no additional cost to the family.

Refer/fail screen: refer for diagnostic audiological assessment by a licensed audiologist at a facility approved by VDH.

Screening Follow-up Recommendations

- Refer: baby should be tested again within a month of hospital discharge. Follow-up should be done at a center designated by VDH as approved for diagnostic audiological assessment. Note: some hospital programs are giving the parent an appointment to return to the hospital for a rescreen. If the infant again fails, refer for diagnostic audiological assessment.
- Missed: the hospital is required to make arrangements for the infant to be screened; refer for hearing screening as soon as possible after birth.
- Pass with follow-up: rescreen every six months until age 3 years at a facility approved by VDH.
- Children born outside a hospital: refer for hearing screening, as soon as possible after birth, by a licensed audiologist at a facility approved by VDH.

HOSPITAL SCREENING PROTOCOL

SCREEN ALL INFANTS PRIOR TO DISCHARGE FROM HOSPITAL

Hospital identifies all infants with risk factors for progressive or delayed hearing loss

Hospital screens hearing using either Automated Auditory Brain Stem Response (AABR) or Otoacoustic Emission testing (OAE); requires $\leq 4\%$ false positive rate and 0% false negative rate. Hospital may rescreen prior to discharge using OAE or AABR or complete ABR.

Prior to discharge, hospital gives written information to all parents that includes:

1. Purpose and benefit of newborn hearing screening
2. Indicators of hearing loss
3. Procedure performed
4. Hearing screening results (pass or fail)
5. Recommendation for follow-up hearing test if indicated
6. Where follow-up testing can be obtained (VDH-approved audiological facility, see Appendix 2)

For infants who are missed, hospital informs parent of need for hearing screening and provides a mechanism by which screening can occur at no additional cost to the family.

Hospital completes VDH report and submits within one week of discharge via electronic database

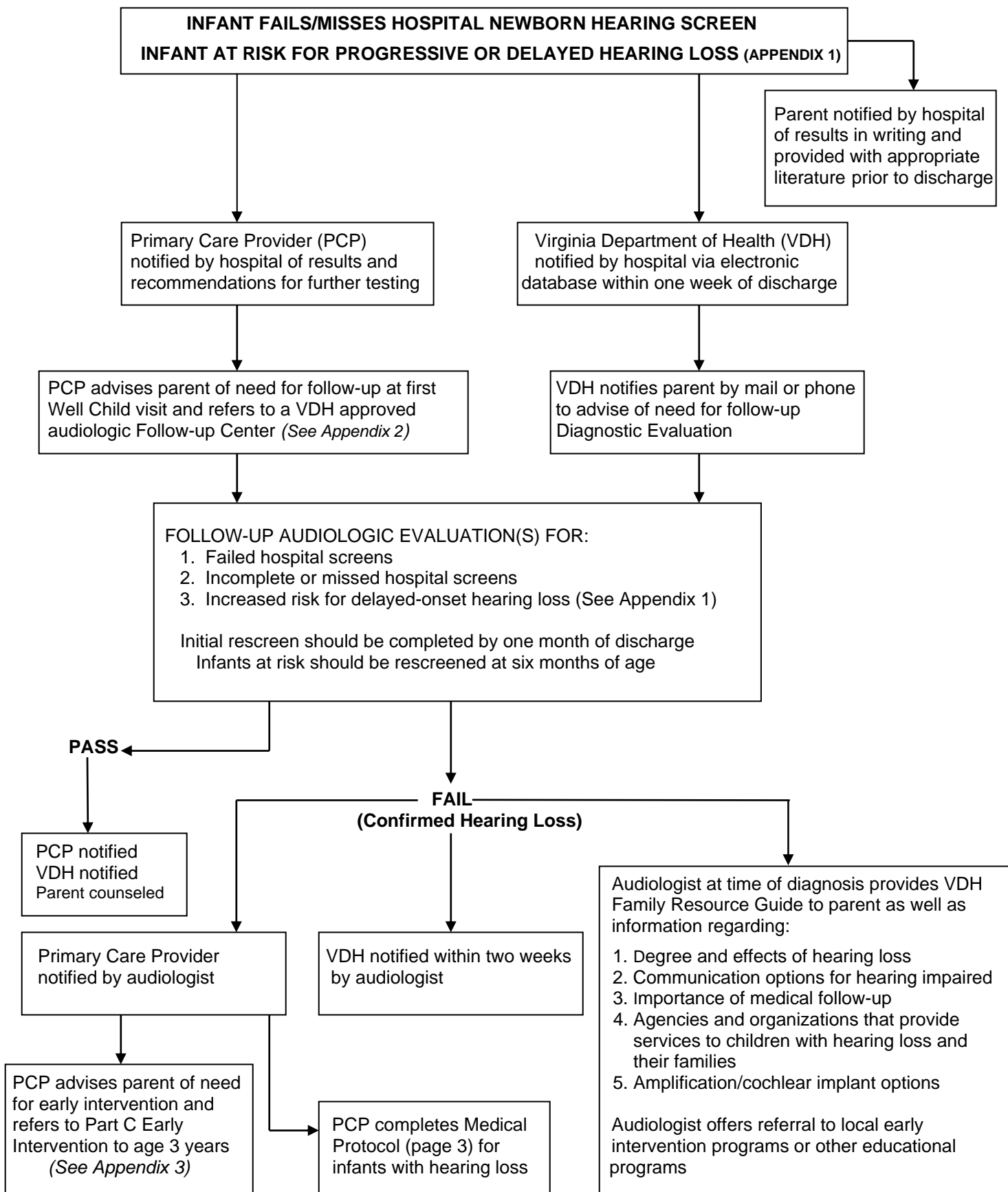
Hospital provides written information to the Primary Care Provider that includes:

1. Procedure performed and limitations of newborn hearing screening
2. Results (passed, failed or missed)
3. Recommendation for further follow-up testing if indicated
4. Where the follow-up testing can be obtained (VDH-approved audiological facility, see Appendix 2)

For infants at increased risk for hearing loss (Appendix 1), the written information should also include:

1. Identification of the risk indicators
2. Recommendation to retest hearing at 6 month intervals up to a minimum of three years of age

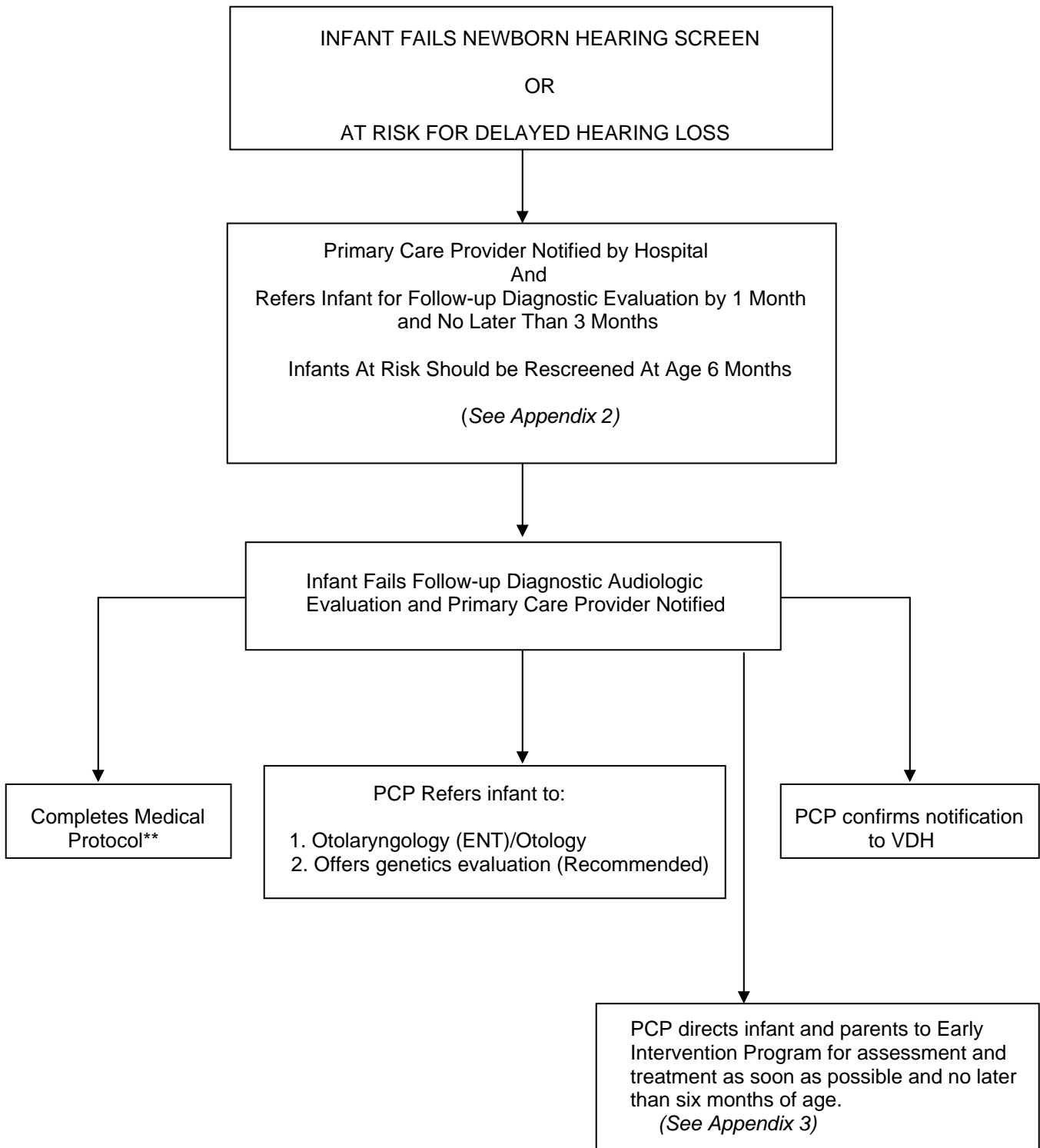
MANAGEMENT FOR INFANT WHO REFERS FROM HOSPITAL HEARING SCREEN



MEDICAL PROTOCOL FOR INFANTS WITH CONFIRMED HEARING LOSS

1. Document history of:
 - a. 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
 - b. Perinatal high risk factors: family history of hereditary childhood sensorineural hearing loss, in-utero infections, craniofacial or external ear anomalies, birthweight less than 1500 grams, hyperbilirubinemia >25 mg/dl, bacterial meningitis, Apgar scores of 0-4 at one minute or 0-6 at five minutes, administration of ototoxic drugs, mechanical ventilation lasting 5 days or longer, stigmata of syndromes known to have sensorineural hearing loss, neurofibromatosis Type II, and persistent pulmonary hypertension
 - c. Family History: family members with permanent hearing loss with onset before age 30 years not related to trauma or medical condition; Plan for other children (need for genetic counseling)
2. Complete a physical examination, with special attention to:
 - a. Minor anomalies: unusual morphologic features occurring in less than 5% of the population with no cosmetic or functional significance
 - b. Major anomalies: dysmorphic features that cause significant cosmetic or functional abnormality, such as cleft palate, cardiac, limb or other skeletal deformities
 - c. Poor growth, microcephaly, or abnormal neurological exam
3. Laboratory
 - a. Obtain urine culture for cytomegalic inclusion virus before age 3 weeks if possible
 - b. Consistent with history of findings, consider testing for rubella, syphilis, or toxoplasmosis
 - c. Chromosomes if significant dysmorphic features
 - d. EKG if cardiac condition suspected
 - e. Skeletal survey if growth delayed or disproportionate
 - f. Head CT or MRI if neurological exam abnormal
4. Obtain hearing tests on first-degree relatives (parents and siblings) if family history positive
5. Ophthalmology/cardiology/nephrology evaluation if indicated
6. Developmental evaluation if indicated
7. Refer for genetics evaluation and counseling for both syndromic and non-syndromic forms of hearing loss
8. Refer to Otolaryngology (ENT)/Otology

PRIMARY CARE PROVIDER MEDICAL PROTOCOL



Appendix 1

Factors that increase risk for progressive or delayed hearing loss include:

- (a) Family history of permanent childhood hearing loss
- (b) In-utero infections including Cytomegalovirus, Herpes, Toxoplasmosis or Rubella
- (c) Craniofacial or external ear anomalies
- (d) Postnatal infections associated with sensorineural hearing loss including bacterial meningitis
- (e) Stigmata of syndromes known to have sensorineural or conductive hearing loss
- (f) Neurofibromatosis Type II
- (g) Persistent pulmonary hypertension associated with mechanical ventilation, hyperbilirubinemia requiring exchange transfusion, or conditions requiring extracorporeal membrane oxygenation (ECMO)
- (h) Neurodegenerative disorders including Hunter Syndrome, Friedreich's ataxia and Charcot-Marie-Tooth Syndrome
- (i) Head trauma
- (j) Recurrent or persistent otitis media with effusion for at least 3 months
- (k) Syndromes associated with progressive hearing loss including Neurofibromatosis, Osteopetrosis, Usher's Syndrome, Goldenhar Syndrome, Branchio-Oto-Renal Syndrome, CHARGE Association, Pendred Syndrome, Pierre Robin Syndrome, Trisomy 21 (Down) Syndrome, Waardenburg Syndrome, choanal atresia, Stickler Syndrome and Rubinstein-Taybi Syndrome
- (l) Parental or caregiver concerns

Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs, Joint Committee on Infant Hearing, June 2000

APPENDIX 2

Virginia Department of Health
Virginia Early Hearing Detection and Intervention Program
formerly known as
Newborn Hearing Screening Program

Approved Sites for Audiological Assessment

The current list is available on the Program Web site at:

www.vahealth.org/hearing/providers.asp

or, call the Virginia Department of Health at

804-864-7718

Toll Free: 1-866-493-1090

APPENDIX 3

Part C Early Intervention Central Points of Entry

The central directory is available on the Web site at:

www.infantva.org

or

via Information and Referral at:

1-800-234-1448