Virginia Early Hearing Detection & Intervention Program

PROTOCOLS FOR DIAGNOSTIC AND AUDIOLOGICAL ASSESSMENT

2018

VDH VIRGINIA DEPARTMENT OF HEALTH

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

This document provides guidance and recommended procedures for how best to implement audiological services 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].

The audiological assessment protocols were first developed in 1999 and revised in 2004 and 2011. The 2018 revision is the product of the Virginia Early Hearing Detection and Intervention Program (VEHDIP) Task Force comprised of four audiological experts with extensive pediatric experience and represents best practice 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 Task Force and the VEHDIP Advisory Committee— which consists of representatives from relevant groups including, but not limited to, physicians, otolaryngologists, audiologists, speech pathologists, nurses, parents, and educators of the deaf and hard of hearing— unanimously agreed that Virginia diagnostic audiological 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. The process also requires follow-up diagnostic services, counseling, intervention programs, and parental education. This comprehensive process should involve a multidisciplinary team including, but not limited to, audiologists, physicians, educators, speech/language pathologists, nurses, parents and educators of the deaf and hard of hearing. This document, therefore, highlights key information about which audiological staff should be aware.

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

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[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

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

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

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

All infants must be given a hearing screen prior to hospital discharge (per Regulation). The hospital discharging the infant to home should screen the infant's hearing as close to discharge date as possible. Even if the infant was screened and passed at a previous facility, the discharging hospital should perform a hearing screen, as the infant's health status may have changed.

I. Audiologist and Facility Qualifications

A comprehensive audiological assessment is the diagnosis of normal hearing or hearing loss specific to each ear. The assessment of hearing loss includes identification of type, severity, and when possible, the etiology of the hearing loss as well as knowledge of the choice of treatment options.

This document describes the Joint Committee on Infant Hearing (JCIH) recommendations for infant screening, diagnostic procedures, and appropriate diagnostic equipment. Therefore, the VEHDIP maintains a roster of facilities that are equipped to provide screening and diagnostic services for children 0-36 months of age.

II. Interpretation of Hearing Screen Results

Definitions of hearing screening results categories and the recommended subsequent actions are listed below.

- **Passed**: No evidence of significant hearing impairment.
- **Missed**: Infant not born in a hospital or <u>not screened</u> at the hospital prior to discharge. Screening should be performed before **one month** of age.
- **Fail**: Infant failed the initial screen. Re-screening and/or a comprehensive diagnostic evaluation should be performed within one month of the failed initial screen.
- **Failed with risk: Infant failed the initial screen,** and has one or more risk indicators for developing progressive hearing loss**.** Re-screening and/or a comprehensive diagnostic evaluation should be performed within one month of the failed initial screen**. If the infant passes the subsequent rescreen, they should have** a diagnostic audiological assessment between 12- 24 months of age.
- **Passed with risk**: Infant passed the initial screen, but has one or more risk indicators for developing progressive hearing loss. A diagnostic audiological assessment should be completed between 12 to 24 months of age. The timing and number of hearing re-evaluations for children with risk factors should be customized and individualized depending on the relative likelihood of subsequent delayed onset hearing loss. Early and more frequent assessment may be indicated for children with Cytomegalovirus

(CMV), syndromes associated with progressive hearing loss, neuro-degenerative disorders, trauma, or culture positive postnatal infections associated with sensorineural hearing loss, for children who have received Extracorporeal Membrane Oxygenation (ECMO) or chemotherapy, and where there is a caregiver concern or family history of hearing loss (JCIH 2007[4]).

Note: For re-screening, a complete screening on both ears should be performed. (per Regulation)

III. Screening and Assessment

Persons providing audiological services to infants after hospital discharge:

- Must provide the screening or evaluation results to the parent or guardian (per Regulation), and should provide this information at the time of the visit.
- Must provide the child's Primary Care Provider (PCP) with screening results (per Regulation).
- Must 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, referral to Part C Early Intervention and/or other educational program, communication and amplification options, the importance of medical follow-up, and agencies and organizations that provide services to children with hearing loss and support for their families (per Regulation).

VDH requires audiologists and re-screeners to report test results, including diagnosis and recommendations, for children ages 0-36 months into an online audiology reporting system called Virginia Infant Screening and Infant Tracking System (VISITS) within seven days of the patient's visit but no later than fourteen days (per Regulation). If a child is diagnosed with a hearing loss, the recommendation is to report the results as soon as possible in order to facilitate a prompt automatic referral to Early Intervention. However, if a baby is born out of state, born at home, or in a birthing clinic, these results should be faxed to VDH on the audiologist reporting form within seven days but no later than fourteen days of the test date.

Outlined below are descriptions and procedures regarding initial screening, re-screening, diagnostic visit, additional assessment considerations, test parameters and thresholds, confirmed hearing loss, unilateral hearing loss, and auditory neuropathy spectrum disorder (ANSD).

A. <u>Initial Screening</u>

- 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 hearing screening are:

[4] 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

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

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

- Not all infants will pass the newborn hearing screening; no more than two in-patient screenings should be attempted before discharge. Only the final in-patient screening should be reported to the VEHDIP. Excessive re-screening can cause an increase in false negatives where infants with hearing loss pass. Both ears should be tested during all screenings.

- Due to the increased incidence of ANSD in the NICU patient population, newborns who receive this level of care should have both ears screened using ABR testing prior to hospital discharge or transfer to a lower level of newborn services.

B. <u>Out-patient re-screen: all infants who failed within 1 month of hospital discharge</u>

- Infants that pass with no risk factor
  - Report in VISITS
  - PCP and Family should be provided with written results
  - Family should be provided with written information about hearing, speech, and language development.

- Infants that pass with risk for progressive or delayed onset hearing loss
  - Report in VISITS
  - PCP and Family should be provided with written results
  - Family should be provided with written information about hearing, speech, and language development
  - Parents should be counseled regarding the need for audiological follow-up

- Infants who fail OAE screening should be re-screened with an ABR whenever possible.
- Infants who fail ABR screening should not be re-screened with an OAE.
- If ABR is not available, OAE Protocol should be completed. OAE Protocol includes:
  - OAEs:
    - Considered a pass if present at 3 of 4 frequencies
    - If failed, refer for diagnostic ABR
  - Tympanometry:
    - If abnormal, refer to physician for medical intervention
  - Acoustic reflexes threshold (ART):

- Test frequencies may be consistent of ipsilateral stimuli at 500,1000, 2000 or 4000 Hz.
- Considered a pass if ipsilateral acoustic reflex thresholds are present at or below 100dB nHL.
- If failed with normal tympanometry, refer for diagnostic ABR.
- If failed with present OAE and abnormal tympanometry, return for follow up tympanometry and acoustic reflexes.

- Both ears should be tested even if only one ear failed the initial screening.
- Obtain all information from family that is requested by VISITS.
- Obtain and record specific results for each ear.
- Written audiological results should be provided to the PCP and family.
- Written information about hearing, speech, and language development should be provided to family.

C. <u>Diagnostic Visit (recommendations for audiologic 3 months assessment)</u>

Below is the recommended protocol for a comprehensive audiologic evaluation to diagnose hearing loss, or normal hearing sensitivity, in infants who have failed their newborn hearing screening.

A. Perform an otoscopic examination.
B. Evaluate hearing sensitivity utilizing ABR technology.
    a. Perform high-intensity click ABR to rule out ANSD or central pathology:
        - Present high-level click air-conducted click stimuli (70-90 dB nHL), with a condensation and rarefaction polarity, via insert earphones, to evaluate for neural integrity.
            - Increase intensity as needed until waves I, III, & V appear or no response at equipment limits is confirmed.
            - Overlap the waveforms to evaluate for ANSD.

        - If ANSD is suspected:
            - Evaluate using ABR to high-level click stimuli (70-90 dB nHL), tested with rarefaction and condensation polarity clicks in separate trials, through insert earphones.
            - A trial run with the sound-delivery tube clamped should be used to differentiate between the cochlear microphonic (CM) and stimulus artifact.
            - If the facility is unable to perform an ABR test at the same visit, refer to the PCP or VEHDIP so that the child may be referred to a diagnostic facility.

        - If ANSD is suspected due to a ringing Cochlear Microphonic (CM), clamp the insert earphone tube, or detach the insert

earphone tube from the transducer, and record an additional rarefaction and condensation polarity at the same intensity. The waveforms recorded should be essentially flat in order to rule out stimulus artifact from the CM.

- If the click ABR suggests ANSD or other retro-cochlear hearing loss, frequency specific ABR may be performed. If ANSD is diagnosed follow up ABR testing should be performed in the future to monitor for neural maturation.

b. Frequency-Specific Toneburst Threshold ABR:
   - Utilizing air-conducted toneburst stimuli, at minimum, two frequencies should be evaluated in each ear, to investigate hearing sensitivity: one high-frequency and one low-frequency, and may include:
     - 500, 1000, 2000, 3000, 4000, 8000 Hz
   - If hearing loss is confirmed, bone-conducted toneburst stimuli should be utilized to determine the nature of the hearing loss: conductive vs. sensorineural vs. mixed, at the same frequencies where hearing loss was identified.
   - If frequency specific thresholds are normal, follow protocols for monitoring of risk indicators for the child.

C. Perform acoustic immittance in each ear.
   a. Tympanometry:
      - High-frequency tympanometry preferably 1000 Hz for infants under 4 months of age; do not use 226 Hz for infants under 4 months
      - Low-frequency tympanometry (226 Hz) for infants over 4 months of age
   b. Acoustic Reflex Threshold (screening or diagnostic) (ipsilateral or contralateral):
      - Utilize appropriate probe tone depending on age (high-frequency (1000 Hz) vs. low-frequency (226 Hz))
      - Test frequencies: 500, 1000, 2000, 4000 Hz
      - Denote if reflexes were screened or if threshold search was performed

D. Perform initial or repeat Otoacoustic Emissions (OAE) testing.
   a. Transient Evoked Otoacoustic Emissions (TEOAEs)
   b. Distortion Product Otoacoustic Emissions (DPOAEs)
   c. A TEOAE or DPOAE test is passed when 3 out of 4 frequencies yield present responses

- Counsel parents/guardians on the results and make appropriate follow-up recommendations

D. Additional Assessment Considerations

If the appointment is limited due to the sleep state of the patient and hearing loss cannot be confidently ruled out:

- Schedule the infant for a repeat audiologic evaluation as soon as possible. After two unsuccessful natural sleep ABR tests have been attempted, a physician recommended sedated ABR should be considered.

E. Test Parameters/Thresholds

If you do not have research based or manufacturer recommended protocols, these parameters may be used as a guideline.

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 ≥6dB than measured noise floor levels

Auditory Brainstem Response (ABR) (*Adapted from Stapells & Oats, 1997***)**

- General ABR parameters:
    - 2 Channel Recording:
        - Vertex - left mastoid or left ear lobe
        - Vertex - right mastoid or right ear lobe
        - Low-forehead- ground (use high forehead for newborn < 6 months due to present fontanel)
    - One Channel Recordings:
        - Vertex to ipsilateral mastoid or ear lobe
        - Opposite mastoid or earlobe acts as ground
        - Slope: 6 or 12 dB per octave, analog
    - EEG Filter:
        - High Pass: 20-30 Hz

        - Low Pass: 1500-3000 Hz
    - Number of Trials:
        - 2000 per average
        - At least 2 repetitions
- High Intensity click ABR
    - Time window: 0-12 ms
    - Stimulus: air conduction clicks
    - Rate: no greater than 39.1 clicks/second
        - Use a slower click rate 21.7 clicks/sec or 7.7 clicks/sec if waves I, III, and V are unclear (always confirm click rate is compatible with your equipment settings).
    - Polarity: collect one rarefaction and one condensation run and overlap to evaluate for ANSD
    - Gain: 100 mV
- Frequency-Specific Threshold ABR
    - Time window: 0-25 ms
    - Stimulus: air conduction tonebursts
    - Rate of no greater than 39.1 tone pips/second
    - Polarity: Alternating for 1000, 2000, 4000 or 8000 Hz stimuli, single polarity for 500 Hz
    - Gain: 100 mV
    - Stimuli 2-1-2 linear:
        - 500 Hz: 4 ms 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
    - Ipsilateral Noise: Band-reject (notched noise, with 1-octave-wied notch centered on the tone frequency)
    - Contralateral Noise: White noise 30 dB below tone masking level (when possible)
- Bone-Conducted ABR
    - Time window: 0-25 ms
    - Stimulus: bone conduction tonebursts
    - Rate of no greater than 39.1 tone pips/second
    - Polarity: Alternating for 1000, 2000, 4000 or 8000 Hz stimuli, single polarity for 500 Hz
    - Gain: 100 mV
    - Utilize contralateral masking when indicated for ear specific bone-conduction results
        - If unable to mask the contralateral ear, evaluate ipsilateral vs. contralateral recording for a wave I at a higher intensity level.

- If there is an ipsilateral wave I, 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.

F. <u>If hearing loss is confirmed</u>:

- Report the results into VISITS immediately.
- Notify the child's PCP.
- Refer to an otolaryngologist for otologic evaluation and to obtain medical clearance for amplification and/or assistive technology if appropriate.
- If appropriate, provide information on cochlear implant.
- If appropriate, provide information for American Sign Language.
- Prepare families of children with hearing loss that they be referred for:
    - Genetic testing and counseling
    - Ophthalmology evaluation
    - Part C Early Intervention services, regardless of type or severity of the loss
    - parent to parent support network

- Explain results to Family
    - Provide parent or guardian with information about services needed for specific type and severity of hearing loss.
    - Inform the family that Virginia EHDI will be in contact with them to provide additional resources.

G. <u>Unilateral Hearing Loss:</u>

Unilateral hearing loss refers to a hearing loss in one ear—whether mild or profound, congenital or acquired—that warrants a definitive diagnosis. Parents or guardians 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. A child with unilateral hearing loss:

- May exhibit delays in speech and language development.
- Will have difficulty localizing sounds; therefore, crossing the street or riding a bike are safety concerns.
- May exhibit behavior and/or social problems.
- May have difficulty following directions, be distractible, and may appear inattentive; may show signs of fatigue as the school day progresses.
- May lag substantially behind in math, language, cognitive and social functioning as compared to their peers, and may be at risk for failing a grade in school.
- May benefit from assistive listening devices in the classroom.

- May benefit from amplification.

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 Part C Early Intervention services for developmental evaluation and support is automated based on the reporting of a child's hearing diagnosis. Visit www.infantva.org for information on referral processes and contacts.
- Noise protection counseling to protect the "good ear."
- Prompt medical management of middle ear disorders.
- Counsel to see ENT if hearing loss suddenly changes (within 48 hours of change).

H. Auditory Neuropathy Spectrum Disorder (ANSD):

According to the Joint Committee on Infant Hearing, 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 ANSD. This is a hearing impairment characterized by a unique constellation of behavioral and physiologic auditory test results. Behaviorally, children with ANSD may exhibit mild to profound hearing loss, with typically poor speech perception. Physiologic measures of auditory function may include the finding of normal otoacoustic emissions, an intact CM and atypical or absent auditory brainstem response to high intensity click stimuli suggesting neural conduction dysfunction. CMs will show a characteristic reversal in polarity with reversal in polarity of the stimulating click; ABR will show a constant polarity regardless of polarity of the click. CMs generally remain present in individuals with ANSD despite loss of OAEs.

ABR recordings might appear as (1) a "flat" ABR with no evidence of any peaks, (2) presence of early peaks (waves up to III) with absence of later waves, or (3) some poorly synchronized but evident later peaks (wave V) that appear only to stimuli at elevated levels.

Reports suggest that those at increased risk for ANSD are infants with a compromised neonatal course, infants with hyperbilirubinemia, and children with a family history of childhood hearing loss. Currently, 1 in 10 cases of sensorineural hearing loss cases are ANSD. Audiologic and medical monitoring of infants that pass the newborn hearing screen at risk for ANSD is recommended between 12 to 24 months of age. (NHS 2008[5])

I. Risk Indicators

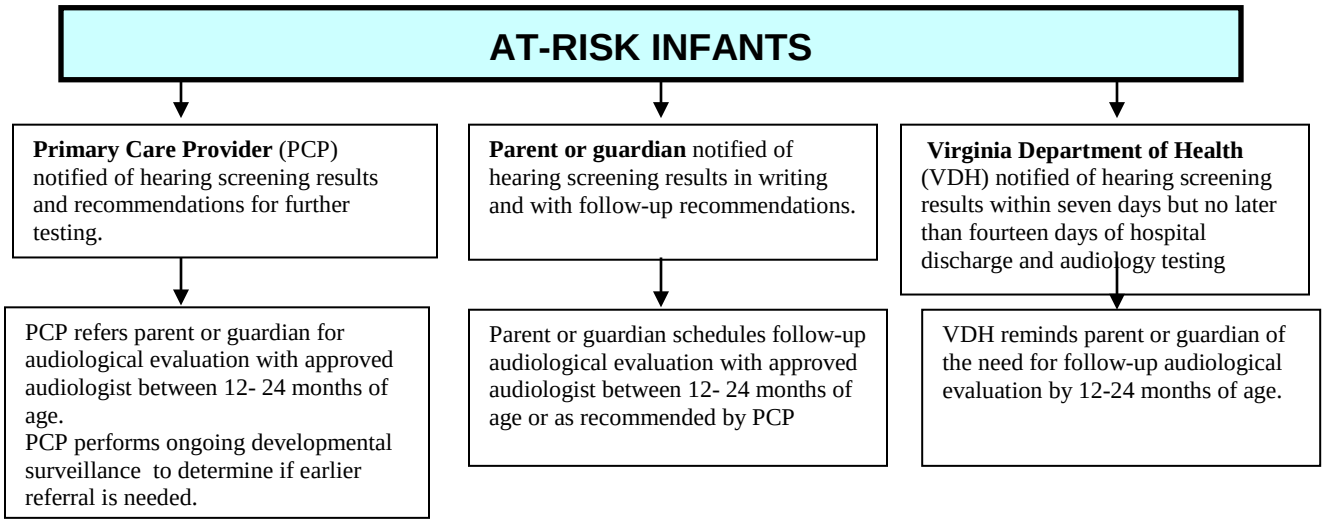
The *Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs, Joint Committee on Infant Hearing* identifies risk indicators that often are associated with infant and childhood hearing loss (see Table I).

[5] Adapted from *Guidelines for Identification and Management of Infants and Young Children with Auditory Neuropathy Spectrum Disorder*, NHS 2008

These indicators place an infant at risk for progressive or delayed-onset sensorineural and/or conductive hearing loss.

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 PCP 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



VDH recommends that a medical professional obtain the risk information from the infant's and mother's charts; family history of permanent childhood hearing loss should be identified by a direct question to the parent(s). The parent should not simply be given the whole list of indicators to check off, as they may not understand the meaning of all indicators. The medical professional should review the risk information with the parent.

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.

Infants who pass but have an identified risk indicator for progressive or delayed-onset hearing loss (**pass with risk**) should have a complete diagnostic evaluation between 12-24 months of age.

J. 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, Virginia 23219
Phone: Toll Free 1-866-493-1090 TTY 7-1-1
Fax: 804-864-7771
Website: newbornhearingtestva.com

If a VISITS user is locked out of VISITS, please call the help desk at (804) 864-7200 option 2 between 8:00 am – 5:00 pm or use the forgot password link at the login page to reset your password.

For technical and reporting issues, such as inability to locate a child, please contact VEHDIP Staff.

Table I. 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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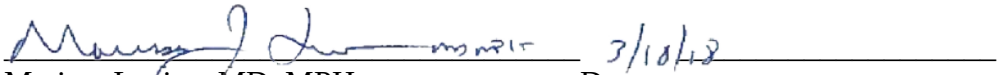
Approved by:



Marissa Levine, MD, MPH Date
State Health Commissioner

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