V. Recommended Medical Evaluation for Newborns with Varied Hearing Levels (Hearing Loss)

This document uses the term "varied hearing levels" to encompass the wide spectrum of people with hearing differences, including those who have been categorized as deaf, hard of hearing, or those with varying degrees of hearing loss.

Pregnancy, Labor, Delivery History:

- Assess for pregnancy complications. Ask about any febrile illness, intrauterine growth restriction (IUGR), polyhydramnios/oligohydramnios, anomalies noted on ultrasound, maternal infections (e.g. toxoplasmosis, syphilis, varicella, parvovirus B19, rubella, cytomegalovirus, herpes, Zika), and maternal exposures like alcohol, prescription and non-prescription drugs and medications. Note that cytomegalovirus (CMV) is the most common infectious cause of congenital varied hearing levels and can cause progressive varied hearing levels even in children who pass the newborn hearing screen. Thus, it is important to ask whether CMV titers were obtained during pregnancy.
- Assess for neonatal risk factors known to be associated with varied hearing levels. This should include prematurity, low birth weight, severe jaundice, prolonged mechanical ventilation, birth defects, unique facial features, NICU admission, infections requiring IV antibiotics (e.g. aminoglycoside >5 days), and infections including meningitis/encephalitis, CMV, Zika, herpes, syphilis, rubella, toxoplasmosis, varicella, measles, and mumps.
- Ask about any delivery complications including hypoxia, respiratory distress, or low Apgar scores.
- Determine whether the baby passed or referred on their newborn hearing screen. Note the type of hearing screen (OAE or AABR), timing of the screen, and number of screens completed. (Note that according to the Joint Committee on Infant Hearing 2019 Position Statement, some high risk infants will need repeat screening or diagnostic follow up even if they pass the newborn hearing screen. Note that infants admitted to the NICU for 5 days or more are at increased risk for Auditory Neuropathy, and AABR/ABR is the preferred screening/testing method for these infants).
- Determine whether the baby had an abnormal result on their newborn bloodspot screen or on their critical congenital heart disease (CCHD) screen.

Family History:

- Obtain a three generation family history to assess if any relatives have had early, progressive, or delayed onset of permanent childhood varied hearing levels. (Note that family history is negative or non-contributory in most).
- Ask about stigmata of syndromes associated with varied hearing levels in relatives. This includes ear pits and other craniofacial abnormalities, pigmentary anomalies, goiter, blindness, renal disease, birth defects, and intellectual disability.
- Assess for consanguinity.

Physical Examination

- Vital signs
- General
    - Screen for major birth defects, such as cleft lip/palate, and cardiac, limb, skeletal, or genital defects
    - Screen for minor anomalies, such as transverse palmar crease
- Skin/hair (birthmarks, piebald patch, abnormal hair whorls or white forelock)
- Face/head (symmetry, shape, size, micro- or macro- encephaly)
- Eyes (reactive pupils, eye movements intact, microphthalmia, hypertelorism)
- Epicanthal Folds (absent/present)
- External ears (size, shape, position, microtia or atresia, ear pits or tags)
- External auditory canals (patency)
- Tympanic membrane (mobility, inflammation, retraction)
- Middle ear (presence/absence of fluid)
- Nose (assess choanal patency)
- Oropharynx (clefting, micro- or retro-gnathia)
- Neck: (enlarged thyroid, branchial cleft or cyst)
- Chest
- Lungs
- Heart (congenital cardiac defects, murmurs)
- Abdomen: (organomegaly present/absent)
- Back (spinal curvature, sacral dimple)
- Extremities (joint hypermobility, skeletal/limb defects, clinodactyly/syndactyly/oligodactyly/polydactyly, transverse palmar crease)
- Genitalia (genital ambiguity or anomaly, anal atresia)
- Neurological
    - Cranial Nerves: Cranial nerves II-XII
    - Muscle: (muscle mass, tone and strength)
    - Sensation

    - Reflexes: Primitive reflexes, Protective reflexes, Deep tendon reflexes, clonus present/absent
- Look for physical exam findings suggestive of syndromes associated with congenital varied hearing levels. Syndromes associated with sensorineural causes of varied hearing levels include Waardenburg syndrome, CHARGE syndrome, VACTERL syndrome, branchio-oto-renal syndrome, Pendred syndrome, and Usher syndrome. Syndromes associated with conductive causes of varied hearing levels include Down syndrome, Treacher Collins syndrome, Crouzon syndrome, Klippel-Feil syndrome, branchio-oto-renal syndrome, CHARGE syndrome, VACTERL syndrome, and Goldenhar syndrome.

Review Hearing Assessment

- Results of infant hearing screening
- Results of audiologic testing to date
    - Auditory Brainstem Response (ABR)
    - Otoacoustic Emissions (OAE)
    - Auditory Steady State Response (ASSR)
    - Tympanometry
- Plan for periodic hearing rescreening to at least age 5

The Role of the Medical Home:

The importance of the primary care physician and medical home in ensuring timely follow up and intervention cannot be overstated. The medical home plays a critical role in ensuring that infants have completed hearing screening by 1 month of age, have had an audiologic diagnosis by 3 months of age, and have received necessary interventions by 6 months of age, including enrollment into early intervention, placement of hearing devices when appropriate and aligned with family desires, and consultation for cochlear implantation if indicated. (Justification: Some families may choose not to use assistive listening devices if they are already fluent or pursuing visually accessible language (ASL or cued speech)). An outpatient hearing screen for infants who have not passed the newborn hearing screen prior to hospital discharge or who were born at home should be scheduled within the first 2-3 weeks of life. Referral to an audiologist by 3 months of life for infants who have not passed the newborn hearing screen should be facilitated by the medical home, if not already done at the birth hospital. Any infant diagnosed with a varied hearing level should be referred to the Colorado Regional Hearing Resource Coordinator, or CO-Hear (early intervention Part C) by the medical home, if not already done by the audiologist.

Referrals and Other Testing

- Consider CMV testing by PCR on saliva or urine, within 3 weeks of birth, for all children who refer on the newborn hearing screen or who have other risk factors for congenital CMV infection.
- Audiologic evaluation and amplification should not be delayed while awaiting referral for specialist evaluations. Every infant confirmed as deaf or hard of hearing, with or without middle ear dysfunction, should be referred by the medical home for specialty evaluations including:
    - ENT/Otology
        - Medical clearance for amplification
        - Discuss inner ear imaging options (e.g. MRI or CT scan)
        - Discuss cochlear implantation (Note that the CDC recommends pneumococcal conjugate vaccine (PCV13) for all people who have or are candidates for cochlear implants, and pneumococcal polysaccharide vaccine (PPSV23) for people 2 years and older who have or are candidates for cochlear implants).
    - Genetic Counseling
        - Review with a geneticist or genetic counselor the multiple etiologies of varied hearing levels.
        - Discuss genetic testing options (e.g. connexin panel, chromosomal microarray), and help families decide what testing is appropriate.
    - Ophthalmology
- Consider EKG if physical exam raises suspicion of syndromes associated with long QT interval (e.g. Jervell and Lange-Nielsen syndromes).
- Consider renal ultrasound if physical exam raises suspicion of syndromes associated with renal disorders (e.g. branchio-oto-renal syndrome).
- Inform the family of local and national parent support groups (see www.cohandsandvoices.org).

Ongoing Primary Care in a Medical Home:

In addition to speech and language delays, approximately 40% of children confirmed as deaf or hard of hearing or with varied hearing levels will demonstrate additional delays, including autism and learning differences, or will have genetic syndromes, or blindness. It is important that a primary care physician in a medical home provide:

- Periodic developmental and speech & language screening, and early referral for evaluation and therapies if any concerns

- Assess parental impression of hearing and speech at every visit, and educate the family about expected auditory, speech, and language development.
- Recognize cultural and linguistic diversity and provide accommodations as needed.
- Assess hearing device use to determine if re-evaluation by the patient's audiologist or ENT/otologist is indicated.
- Provide ongoing assessment for persistent or recurrent otitis media, with a low threshold for referral to ENT.
- Assess the need for screening siblings, depending on suspicion of a familial diagnosis or results of any genetic testing.

For additional information please refer to:

1. American Academy of Pediatrics, Early Hearing Detection and Intervention Guidelines for Medical Home Providers (see https://tinyurl.com/y5zzowco).

2. The Joint Committee on Infant Hearing. (2019). Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Journal of Early Hearing Detection and Intervention 2019*; *4*(2): 1-44. JCIH 2019 Position Statement