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III. Recommended Guidelines for the Medical Practitioner for Newborn Hearing Screening

When there is a NORMAL SCREEN

Infants who pass the newborn hearing screen or subsequent screenings can be assumed to have normal hearing. However all infants should be monitored for late onset or progressive hearing loss. Parental concern about speech and language delays, at any time in a child's life, should receive prompt referral for an audiological evaluation. Hearing testing can be performed at any age.

The Joint Committee for Infant Hearing's *Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs* has recommended that the following indicators be used for neonates or infants under two years. These indicators place an infant at risk for progressive or delayed-onset sensorineural hearing loss and/or conductive hearing loss. Any infant with these risk indicators for progressive or delayed-onset hearing loss that has passed the birth screen should, nonetheless, receive audiologic monitoring every six months until age three years. These indicators are:

- 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 including: hyperbilirubinemia at a serum level requiring exchange transfusion; persistent pulmonary hypertension of the newborn associated with mechanical ventilation; conditions requiring the use of extracorporeal membrane

oxygenation. Low levels of hyperbilirubinemia have been associated with auditory neuropathy/ auditory dys-synchrony.

- 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 Friedreich's ataxia and Charcot-Marie-Tooth syndrome.

- Head trauma.

- Recurrent or persistent otitis media with effusion that lasts for at least three months.

Because some important indicators, such as family history of hearing loss, may not be determined during the course of Universal Newborn Hearing Screening programs, the presence of any late-onset risk indicators should be determined in the medical home during early well-baby visits. Those infants with significant late-onset risk factors should be carefully monitored for normal communication developmental milestones during routine medical care.

When there is a MISSED SCREEN

- There is an estimated 1 in 1000 risk of undetected bilateral hearing loss.

- Typically the hospital determines who will retain initial responsibility for recall and screening. It is the responsibility of the primary care physician to know the hospital policies and procedures for recalling and rescreening infants who miss a screen.

- Screening is best performed within the first month of life, later ages require more time due to the infant's increased alertness and may even require sedation in infants older than four months of age.

- Parents who refuse should be counseled on the importance of early identification and encouraged to obtain a screen. Refusals should be documented in the medical record.

When there is an ABNORMAL SCREEN, either Unilateral or Bilateral

- All newborns with abnormal screens **must** be rescreened or referred for an audiological evaluation.

- The abnormal result of the screen is shared with the family before hospital discharge, and a rescreen appointment is scheduled.

- Determine the person or department responsible for the following:

    - Scheduling appointment for recheck.
    - Recalling no-shows; physician involvement may be required.

- Completing all rescreening within four weeks.
- Written notification to primary care physician and Colorado Department of Public Health and Environment.
- Continue recall attempt.
- Record on problem list of outpatient medical record.
- The critical window to confirm diagnosis and begin amplification is two to six months of age.

When the child is subsequently confirmed to be DEAF OR HARD OF HEARING

If the hearing loss is **Unilateral**:

- Unilateral hearing loss is a significant risk factor for later acquired hearing loss in the previously normal ear. Audiologic monitoring is recommended **every three months**, during the first three years of life, due to possible progression to bilateral hearing loss.
- Amplification may have a role in facilitating language development.
- Use of amplification and role of intervention should be explored with audiologist and Regional CO-Hear Coordinator.
- Approach to follow-up may be individualized.
- Record on problem list if rescreening or further testing is delayed.

If the hearing loss is **Bilateral**:

- Early and consistent intervention (typically including amplification) is the key to achieving normal language development. The audiologist and Regional CO-Hear Coordinator can work with the family to assess needs.
- Colorado statistics indicate that 10 percent of these children are profoundly deaf. Children with profound hearing loss have less predictable benefit from traditional amplification; consider opportunities for early intervention programs, early sign language use and nontraditional amplification techniques (e.g., cochlear implants).
- Primary care physician support is necessary:
    - Encourage timely follow-up with audiologist, CO-Hear Coordinator, and other consultants.
    - Monitor continuous use of amplification device.

    - Evaluate ongoing development of communication and language.

- Evaluations recommended in addition to audiology follow-up:

    - Comprehensive multi-disciplinary evaluation (speech, language, functional auditory skills, communication, cognition, motor skills, personal-social skills).

    - Otolaryngology or otology evaluation; required for medical clearance of amplification in children.

    - Genetics evaluation.

    - Ophthalmology exam and follow-up.

    - For more information about these recommendations, refer to Guideline VIII: Recommended Medical Evaluation for Newborns with Confirmed Hearing Loss.