

III. Primary Care Physician/Medical Home Responsibilities in UNHS Program

A. PASS SCREENING RESULTS

Certain infants who pass the newborn hearing screen or subsequent screenings should be monitored for delayed-onset (a hearing loss that is not present at birth but develops shortly after) or progressive hearing loss (hearing loss that worsens over time).

In October 2007, the federal Joint Committee on Infant Hearing issued a policy statement in Pediatrics (Vol. 120 No. 4, pp. 898-921) that updated its earlier 2002 recommendations concerning principles and guidelines for early hearing detection and intervention programs. An updated listing of the 11 risk indicators that place an infant at risk for progressive or delayed-onset sensorineural hearing loss and/or conductive hearing loss was included in that document. **Any infant with these risk indicators for delayed onset or progressive hearing loss who has passed the hearing screen at birth, should, nonetheless, receive audiologic monitoring every six months until three years of age and at appropriate intervals thereafter.** The risk factors are:

- Caregiver concern regarding hearing, speech, language, and/or developmental delay.
- Family history of permanent childhood hearing loss.
- Neonatal intensive care of more than 5 days, or any of the following regardless of length of stay: extracorporeal membrane oxygenation (ECMO), assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide/lasix), and hyperbilirubinemia that requires exchange transfusion.
- In utero infections such as cytomegalovirus, herpes, rubella, syphilis and toxoplasmosis.
- Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
- Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss
- Syndromes associated with hearing loss or progressive or late-onset hearing loss such as neurofibromatosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred and Jervell and Lange-Nielson.
- Neurodegenerative disorders such as Hunter syndrome, or sensory motor neuropathies such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
- Culture-positive postnatal infections associated with sensorineural hearing loss including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
- Head trauma, especially basal skull/temporal bone fracture that requires hospitalization
- Chemotherapy

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 delayed-onset risk indicators should be determined by the primary care physician/medical home during early well-baby visits. Rack cards and posters to alert families to the risk factors are available from the state program at no charge for doctor's offices (see order form in Appendix K).

B. REFER SCREENING RESULTS

Montana hospitals are required to: (1) notify the baby's primary care provider if the newborn hearing screening was completed with a "Refer" result and, (2) to recommend a pediatric audiological assessment before the baby is three months of age. When receiving such notice, the primary care physician/medical home should:

1. Work with the family to ensure that the infant has timely audiological follow-up to determine whether there is a confirmed hearing loss – this should be performed before the baby is three months old. Early intervention should begin before the baby is six months old at the latest to prevent language development deficiencies that may never be recouped.
2. Issue referrals for an otolaryngologist and audiological assessment as needed
3. Be knowledgeable about community resources that can help the family obtain outpatient rescreening or the audiologic assessment. The state newborn hearing screening and assessment monitoring program will provide the primary care professional with a list of those audiologists who have the capacity to perform the pediatric audiological assessment (Appendix G) in accordance with the state's recommended Infant Audiological Assessment Guidelines (see section IV of these Guidelines). The state program will also provide a current listing of the audiologists who contract with the state's Hearing Conservation Program (see Appendix H) and who can provide newborn hearing screening at not cost to the parents under two conditions:
 - a. the baby needs an outpatient rescreening after receiving an initial "Refer" result with inpatient newborn hearing screening in the birth hospital; OR
 - b. the baby was NOT born in a hospital and needs an initial newborn hearing screen and/or repeat screening.

C. MISSED SCREEN OR RESCREEN

The primary care professional should work with the local UNHS program at the hospital/birthing facility to: (1) Communicate to the family the importance of the hearing screening; (2) provide educational material to the family regarding speech and hearing milestones; and (3) recall families that have missed screening or rescreening appointments. The state program has parent educational materials to assist in the education (Appendix D).

D. CONFIRMED HEARING LOSS

The primary care physician/medical home works with the parents and other healthcare professionals in the coordination of care for infants identified with a hearing loss. Those professionals should include, but are not limited to the following:

- Otolaryngologists (ENT specialists)
- Geneticists
- Audiologists
- Speech Language Pathologists

Early Intervention Specialists (Part C personnel) Educators of the Deaf/Hard of Hearing

