E. Medical Guidelines for Infants with Confirmed Hearing Loss

In 1999, the Illinois legislature passed the Hearing Screening for Newborns Act. By December 31, 2002, all hospitals delivering babies will be required to provide hearing screening to all babies born in their facility. The goals of the Illinois program are:

All infants born in Illinois will have their hearing screening at birth.
All newborns referred from the Illinois Newborn Hearing Screening Program will have diagnostic testing completed by three (3) months of age.
All infants diagnosed with significant hearing loss will receive appropriate treatment, including hearing aids and Early Intervention Program services by six (6) months of age.

Universal newborn hearing screening has quickly become the standard of care in Illinois. The Illinois chapter of the American Academy of Pediatrics and the Illinois Academy of Family Physicians endorses universal newborn hearing screening as an important strategy to identify infants with significant hearing loss. Early identification and intervention have been shown to dramatically impact the development of communication skills in young children with hearing loss. In fact, when appropriate diagnosis and treatment can be implemented prior to six months of age, babies with hearing loss can acquire speech and language much like that of babies with normal hearing.

Your involvement as a physician is critical to ensure early identification and intervention. This brochure has been developed to inform you of the most current thinking of many who have worked closely with early hearing identification and intervention programs throughout the country. This thinking has been adopted by the Illinois Universal Newborn Hearing Screening Advisory Committee as guidelines for medical follow-up in the state of Illinois. We hope this will be useful to you in the care of infants referred from universal newborn hearing screening programs. It includes information for primary care physicians, otolaryngologists, and medical geneticists, all of whom are important to the accomplishment of the goal of early intervention and intervention.

It is recommended that infants referred from a universal newborn hearing screening program and identified with a confirmed hearing loss in one or both ears should receive the following evaluations, as indicated by three months of age. It is critical to complete all diagnostic evaluations as quickly as possible so that treatment and intervention such as hearing aids, aural habilitation, and child development therapy can begin before the child is six months of age. “Wait and see” is never recommended.

Primary Care Medical Evaluation

The following should be included in the medical evaluation conducted by the baby’s primary care physician who is responsible for initiating and supervising the evaluation and referral process to otolaryngology, genetics, ophthalmology, neurology, and other professionals, as appropriate.

Infants referred from a universal newborn hearing screening program and identified with a confirmed hearing loss in one or both ears should receive the following evaluations, as indicated, by three (3) months of age, but no later than six (6) months of age.

The following should be included in the medical evaluation conducted by the baby’s primary care physician who is responsible for initiating and supervising the evaluation and referral process to otolaryngology, genetics, ophthalmology, neurology, and other professionals, as appropriate.
History

Prenatal Information

- Ototoxic medication exposure
- Any significant complications during pregnancy
- Any possibility of parental consanguinity

Perinatal information

- Indicators associated with sensorineural and/or conductive hearing loss
- In utero infection, such as cytomegalovirus, rubella, syphilis, herpes, and toxoplasmosis
- Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal
- Birth weight of less than 1,500 grams (3.3 lbs).
- Hyperbilirubinemia with risk of kernicterus
- Ototoxic medications, including but not limited to the aminoglycosides, used in multiple courses or in combination with loop diuretics
- Bacterial meningitis
- Apgar scores of 0-4 at one (1) minute or 0-6 at five (5) minutes
- Mechanical ventilation lasting five (5) days or longer
- Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss
- Intraventricular hemorrhage, seizures or inborn errors of metabolism

Family History

- Other family members with hearing loss with onset at an early age
- Family’s desire to have additional children

Physical Examination

Minor anomalies

- Unusual morphologic features occurring in less than 5% of the population with no cosmetic or functional significance

Major anomalies

- Those causing cosmetic and/or functional abnormality, e.g., cleft palate, cardiac, limb, skeletal deformities

Lab

- Urine culture for CMV prior to three (3) weeks of age, if possible

Referrals

- Audiological evaluation if appointment has not been scheduled
- Ophthalmologic examination
- Genetics evaluation
- Early Intervention
- Referral to local Child and Family Connections office if not yet done
F. Otolaryngological Medical Evaluation

The following should be included in the medical evaluation conducted by an otolaryngologist:

History

Prenatal
Perinatal
Family
Behavioral

Physical Examination

Head and neck examination
Head circumference

Review of Audiological Test Results

Evaluations

Infectious diseases: CMV, Rubella, Syphilis, and Toxoplasmosis
Urinalysis
CT with high resolution of temporal bones (if clinically indicated)