SoundBeginnings

KANSAS MEDICAL ASSESSMENT GUIDELINES FOR INFANTS WITH CONFIRMED HEARING LOSS

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ACKNOWLEDGMENTS

Sound Beginnings expresses extreme gratitude to the members of the Assessment and Amplification Task Force for sharing their expertise, time and energy to develop the Kansas Medical Assessment Guidelines for Infants with Confirmed Hearing Loss. Their commitment demonstrates their strong belief in the importance of providing the highest quality of assessment services for infants and their families. They recognize that the early months in the life of an infant "are not a rehearsal, but the real show."

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Guidelines will be reviewed every two years and updated as needed.

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I. INTRODUCTION

Health and Human Services' Healthy People 2010 includes the following goal for infants: *to confirm hearing loss by three months of age with appropriate intervention no later than six months of age*. Once a hearing loss is confirmed, the infant should be referred for a medical evaluation. The purposes of the medical evaluation are "to determine the etiology of the hearing loss, to identify any related physical conditions, and to provide recommendations for other medical treatment as well as referral for other services." (Joint Committee on Infant Hearing *Year 2000 Position Statement).* In addition to the infant's pediatrician/primary care physician and the Ear, Nose and Throat (ENT) physician, referral may be needed to other medical specialists (*e.g*., cardiologist, developmental pediatrician, medical geneticist, nephrologist, neurologist, ophthalmologist). Surveys indicate that 30-40% of children with hearing loss have other disabilities which may affect communication and related development. (Joint Committee on Infant Hearing *Year 2000 Position Statement).*

The purpose of the Kansas Medical Assessment Guidelines for Infants with Confirmed Hearing Loss is to identify the essential components of the medical evaluations after hearing loss has been confirmed. Test selection is determined by the medical practitioner in partnership with the infant's family, and is based on the child/family history and other presenting factors.

II. PEDIATRICIAN/PRIMARY CARE PHYSICIAN

Pediatricians and other primary care physicians work with families and other physicians by establishing a medical home for the child. They advocate for the child's health care, and they initiate and coordinate the evaluation and the referral processes to other medical specialists and to other services and intervention therapies as needed by the child and family.

Essential components of the medical evaluation include:

- History
  - Prenatal
    - Ototoxic medication exposure
    - Any significant complications during pregnancy
    - Immunization to rubella
    - Syphilis screening
    - Maternal drug use
    - Frequent spontaneous abortions

- Perinatal
  - •Risk indicators for hearing loss (Appendix A)

- Family
  - •Other family members with hearing loss with onset at age less than 30 years.

- Review of hearing assessment results
  - Test results should identify degree and type of hearing loss at a minimum.

  - See Kansas Guidelines for Infant Audiologic Assessment[1] for recommended tests.

- Physical
  - 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 (*i.e.*, cleft palate, cardiac, limb or skeletal deformities)

  - Poor growth and/or microcephaly

  - Abnormal neurologic examination

  - Abnormal ear examination: tympanic membrane abnormality; middle ear status (middle ear effusion can further limit hearing)

- Lab
  - Cytomegalovirus (CMV) is the most common cause of viral induced hearing loss. The hearing loss is sensorineural; it may be unilateral or bilateral; it may be progressive or delayed-onset.

    - •Urine culture for CMV: test must be prior to 3 weeks of age to determine congenital CMV

    - •CMV specific IgG and IgM antigens if under 6 months of age. Titers to detect CMV must be drawn early, before 6 months of age, to be accurate.

- Referrals
  - ENT (Chapter III)

  - Geneticist (Chapter IV)

  - Early intervention services (Chapter V)

[1]These guidelines can be obtained from SoundBeginnings KDHE. Call 1-800-332-6262

- Communication
  - Communication is essential in an effective medical home approach to providing coordinated health care services. The pediatrician/primary care physician is one of several team members working with the infant with a confirmed hearing loss and his/her family. The sharing of information among the family, audiologist, ENT, other medical specialists, and early intervention service providers (*e.g.*, speech-language pathologist, teacher of the deaf/hard of hearing, early childhood special educator) is efficient, cost effective, and most importantly, results in better outcomes for the child and family.

III. EAR, NOSE, AND THROAT (ENT) PHYSICIAN

The ENT physician treats ear diseases, determines the etiology of the hearing loss, determines whether medical and/or surgical intervention may be appropriate, and determines the presence of related syndromes. In addition, this specialist provides information and participates in the assessment for candidacy for amplification, assistive devices and cochlear implantation, and should be involved in long-term monitoring and follow-up as part of the team within the infant's medical home.

Essential components of the medical evaluation include:

- History
  - Prenatal
  - Perinatal
  - Family
  - Behavioral
- Physical
  - Ear, head and neck examination
  - Other systems affected by syndromes (*e.g.*, skin, eye, thyroid)
- Review of prior hearing assessment results
  - The ENT physician should be in contact with the infant's audiologist in order to not duplicate audiologic tests.
  - See Kansas Guidelines for Infant Audiologic Assessment[2] for recommended tests.
- Diagnostic Tests
  - The ENT physician should be in contact with the infant's primary care physician in order to not duplicate diagnostic tests.
  - Lab Tests

[2]These guidelines can be obtained from SoundBeginnings KDHE. Call 1-800-332-6262

•Urinalysis: positive protein or heme may suggest Alport syndrome.

•TORCH titers: to evaluate for congenital infectious etiologies of hearing loss.

•Urine culture for cytomegalovirus (CMV): test must be prior to 3 weeks of age to determine congenital CMV.

•CMV specific IgG and IgM antigens if child is under 6 months of age. CMV is the most common cause of viral induced hearing loss. Hearing loss is sensorineural; it may be unilateral or bilateral; it may be progressive or delayed-onset. Titers to detect CMV must be drawn early, before 6 months of age, to be accurate.

•Syphilis: FTA-absorption test is needed because usual RPR is not sensitive enough. Test is important because this condition can be treated medically.

•Other laboratory tests depending on clinical evaluation and history.

- EKG

- Imaging studies (CT or MRI) of the temporal bones may be helpful to determine cochlear abnormalities. The scan is not necessary in infancy but can be performed at a later date. Indications for early imaging are fluctuating hearing loss or consideration of cochlear implant.

- Referrals
  - Ophthalmology referral (to monitor for accompanying vision disorders; to rule out disorders such as Usher's syndrome)
  - Other systems affected by syndromes (*e.g.*, heart; kidney)
  - Geneticist (Chapter IV)
  - Early intervention services (Chapter V)

- Communication
  - Communication is essential to providing coordinated health care services. The ENT is one of several team members working with the infant with a confirmed hearing loss and his/her family. The sharing of information among the family, primary care physician, audiologist, other medical specialists, and early intervention service providers (*e.g.*, speech-language pathologist, teacher of the deaf/hard of hearing, early childhood special educator) is efficient, cost effective, and most importantly, results in better outcomes for the child and family.

IV. GENETICIST

Whether or not the etiology of the hearing loss is determined based on physical and laboratory tests, the infant and family may be referred for a genetic evaluation. The medical geneticist collects and interprets family history data, diagnoses inherited diseases, performs and interprets genetic tests, and provides genetic counseling.

Essential components of the evaluation include:

- History
  - Pregnancy
  - Family pedigree
  - Developmental
- Physical (or review of prior test results)
  - General Pediatric examination
  - Dysmorphologic examination
  - Neurologic/developmental evaluation
- Diagnostic Tests (or review of prior test results)
  - Review of the infant's hearing assessment results
  - Review of hearing test results on first degree relatives (parents and siblings)
  - Ophthalmologic examination by 6 months of age
  - Laboratory tests
    - TORCH titers or CMV specific IgG and IgM if under 6 months
    - Chromosomes if dysmorphic
    - EKG
  - Skeletal survey if there is short stature or disproportional growth
  - Evaluation of other systems: renal, cardiac, skin
  - CT or MRI of brain if neurologically abnormal
  - Other specialized genetic studies such as DNA testing (*e.g.*, Connexin 26)

- Communication
  - Communication is essential to providing coordinated health care services. The geneticist is one of several team members working with the infant with a confirmed hearing loss and his/her family. The sharing of information among the family, primary care physician, audiologist, ENT, other medical specialists, and early intervention service providers (*e.g.*, speech-language pathologist, teacher of the deaf/hard of hearing, early childhood special educator) is efficient, cost effective, and most importantly, results in better outcomes for the child and family.

V. TRANSITION TO EARLY INTERVENTION SERVICES

The outcomes of a successful early hearing detection and intervention (EHDI) program are that a) all infants with hearing loss are identified as soon as possible, preferably within three months of age; and b) infants with confirmed hearing loss begin receiving early intervention services, as appropriate for the child and family, as soon as possible and preferably by six months of age.

The infant's primary care physician, within the medical home, should be knowledgeable about community-based programs that provide early intervention services for the infant and family. Follow-up care for an infant with a confirmed hearing loss should include collaboration of all medical specialists with other early intervention services to assure that the individualized needs of the child and family are being met. The physician is encouraged to participate in the planning of these services, such as being an active team member in the development and ongoing review of the child's Individualized Family Service Plan (IFSP).

SoundBeginnings A Kansas Resource Guide for Families with Infants and Toddlers who are Deaf/Hard of Hearing[3] has been developed for families whose infant has a confirmed hearing loss. Additional information can be obtained by contacting:

The Make a Difference Information Network (1-800-332-6262 V/TTY)
SoundBeginnings at KDHE (1-800-332-6262 V/TTY)
Infant-Toddler Services at KDHE (1-800-332-6262 V/TTY)
The Kansas Commission for the Deaf and Hard of Hearing (1-800-432-0698 V/TTY)
The community's local Infant-Toddler Early Intervention Network
The local Health Department

[3]These guidelines can be obtained from SoundBeginnings KDHE. Call 1-800-332-6262

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WEB SITES

www.kumc.edu/gec/prof/kugenes.html

www.geneclinics.org

APPENDIX A
RISK INDICATORS FOR HEARING LOSS

From the Joint Committee on Infant Hearing *Year 2000 Position Statement*

Risk Indicators: Birth through age 28 days where universal newborn hearing screening is not mandated

This information should be used by all primary health care providers (e.g., physicians; local health department personnel) to assess risk status for hearing loss during the well-baby visit.

- an illness or condition requiring admission of 48 hours or greater to a NICU
- stigmata or other findings associated with a syndrome known to include a sensorineural and or conductive hearing loss
- family history of permanent childhood sensorineural hearing loss
- craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal
- in-utero infection such as cytomegalovirus (CMV), herpes, toxoplasmosis, or rubella

Risk Indicators: For use with infants (29 days through 2 years) when the newborn hearing screening test was passed

Passing the newborn hearing screening does not mean that the child will not develop or acquire a hearing loss. The presence of any of these risk indicators for progressive or delayed-onset sensorineural hearing loss and/or conductive hearing loss denotes the need to provide audiologic monitoring every 6 months until age 3 years.

- 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 (CMV), herpes, rubella, syphilis, and toxoplasmosis
- neonatal indicators - specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation (ECMO)
- 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 for at least 3 months

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