	reception thresholds if possible. *Otoacoustic emissions–distortion product and/or transient evoked emissions–for continued monitoring of cochlear function.
5 + Years	*Child and family case history/Parent observation report. *Otoscopic examination. *Acoustic immittance: tympanometry, physical volume, and acoustic reflexes. *Standard audiometry– to include air and bone conduction, speech reception thresholds and speech/word recognition. *Otoacoustic emissions– for continued monitoring of cochlear function.

RECOMMENDED MEDICAL PROTOCOL FOR INFANTS/CHILDREN WITH CONFIRMED HEARING LOSS

1. Primary Medical Care Provider

   A. Activities

      1. Initiates and supervises evaluation and referral process.

      2. Referral sources include ENT and/or Otology, Genetics, Audiologists and Therapists.

   B. Notification sent to parents/primary caretaker(s) and the ADPS Newborn Hearing Screening Coordinator.

   C. Important Historical Factors

      1. Exposure to ototoxic medications.

      2. Significant complications during pregnancy.

      3. Immunization to Rubella.

      4. Syphilis screening.

5. Maternal drug use.

6. History of spontaneous abortions.

D. Perinatal High-Risk Indicators

1. Family history of childhood sensorineural hearing loss.

2. Congenital infection known or suspected to be associated with sensorineural hearing loss, such as toxoplasmosis, syphilis, cytomegalovirus, bacterial meningitis, and herpes.

3. Craniofacial anomalies, including morphologic abnormalities of the pinna and ear canal.

4. Hyperbilirubinemia to degree that exchange blood transfusion needed.

5. An illness or condition requiring admission of 48 hours or greater to a NICU.

6. Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss.

7. Persistent pulmonary hypertension of the newborn associated with mechanical ventilation.

8. Conditions requiring the use of extracorporeal membrane oxygenation.

E. Post-Natal High Risk Indicator(s)

1. Family history of childhood sensorineural hearing loss.

2. Infections associated with sensorineural hearing loss including bacterial meningitis.

3. Recurrent or persistent otitis media with effusion for at least three months.

4. Head trauma.

5. Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or Eustachian tube dysfunction.

6. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome.

7. Craniofacial anomalies, including morphologic abnormalities of the pinna and ear canal.

8. Congenital infection known or suspected to be associated with sensorineural hearing loss, such as toxoplasmosis, syphilis, cytomegalovirus and herpes.

9. Parental/primary caretaker(s) concern regarding hearing, speech, language, and/or developmental delay.

2. ENT/Otology

A. History: Prenatal, Perinatal, Family and Behavioral

B. Physical Examination

1. Structure (auricle, ear canal and surrounding structures).

2. Microscopic examination (ear canal, tympanic membrane, middle ear)

C. Head and Neck Examination

1. Structural Abnormalities, e.g., microcephaly

2. Other congenital abnormalities, e.g., white forelock

D. Review prior testing

1. ABR, OAE and other test results available.

2. Possible need for additional repeat testing.

3. Tympanometry (high probe frequency).

E. Laboratory Evaluation

1. CMV, FTA(if family history of syphilis), renal, thyroid (if indicated), toxoplasmosis.

2. Urinalysis (if history of progressive hearing loss in males or gross hematuria).

F. Special testing as indicated

1. EKG (if family history of heart disease or abnormality detected on exam).

2. High resolution CT Scan of temporal bone on all babies with diagnosed sensori-neural hearing loss.

3. MRI brain and CPA (only if indicated)

G. Medical Referrals

1. Genetics referral for all diagnosed babies, including connexin-26

2. Opthamology referral for all diagnosed babies

3. Audiology referral for further diagnostic testing or amplification

H. Additional Referrals (as necessary)

1. Speech/Language evaluation

2. PT/OT evaluation

3. Social Services

I. Data Management

1. Report to primary medical care provider

2. Report to ADPH Newborn Hearing Screening Coordinator

3. Report to Alabama's Early Intervention System or other specialists as indicated by physician.