

- Review of prior testing, i.e., Auditory Brainstem Response (ABR) and Otoacoustic Emission (OAE) results.
- Evaluation:
 - Infectious diseases: cytomegalovirus, rubella, syphilis, toxoplasmosis.
 - Urinalysis.
 - Ophthalmologic examination.
 - Computed Tomography (CT) high resolution of temporal bones.
 - Auditory Brainstem Response for threshold (if not previously done).
 - Otoacoustic Emissions (if not previously done).
 - Medical clearance for amplification and referral.
- Referrals:
 - Genetics.
 - Audiology assessment (see Guideline IV: Recommended Protocol for Infant Audiologic Assessment).
 - Regional Colorado Hearing Resource (CO-Hear) Coordinator (Appendix 6).
 - Early intervention, if not currently being provided.

Genetic Evaluation

- History:
 - Pregnancy.
 - Family pedigree.
 - Developmental.
- Physical Examination:
 - General pediatric examination.
 - Careful dysmorphicologic examination.

- Neurologic/developmental evaluation.
- Diagnostic Tests:
 - Hearing tests on first-degree relatives (parents and siblings).
 - Ophthalmologic examination by six months of age.
 - Toxoplasmosis, herpes, rubella, or cytomegalovirus specific Immunoglobulin G and Immunoglobulin M if under six months.
 - All other laboratory tests depend upon clinical evaluation and history but may include the following:
 - Chromosomes (if dysmorphic).
 - Electrocardiograph (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.
 - Specialized genetic studies: molecular, gene testing, etc.

Ophthalmology Evaluation

Following the diagnosis of confirmed congenital hearing loss, evaluation by a pediatric ophthalmologist is indicated. Yearly re-evaluation by the ophthalmologist is also recommended.