Medical Evaluation when an infant or young child has been identified with hearing loss

- Obtain birth history, family history other significant medical history.

- Perform complete physical exam.

- Refer to otolaryngology to determine etiology of hearing loss and provide medical clearance for hearing aids if chosen by family. Examination to identify factors associated with hearing loss including possible structural or anatomic abnormalities, dysmorphic or syndromic features known to be associated with hearing loss, presence of fluid in the middle ear. Other tests may be ordered such as high-resolution CT scan of the temporal bones to identify conditions such as Mondini malformation or large vestibular aqueduct. Progression of hearing loss may be prevented with appropriate intervention. Cytomegalovirus (CMV) titer testing has become important with administration of gancyclovir in the first month of life to lessen the progression of the hearing loss.

- Other tests may be ordered to investigate association with a syndrome such as: renal functioning (Branchio-oto-renal syndrome), cardiac evaluation (Jervell-Lang-Neilsen, CHARGE), endocrine function (Pendreds), and autoimmune evaluation.

- Refer to Ophthalmology to assure that the visual stimuli to the brain are in no way compromised, and to assess for any associated eye abnormalities or genetic syndromes with both visual and hearing loss.

- Refer to genetics evaluation to determine etiology of hearing loss in order to anticipate whether the child has or is at risk for other medical conditions. Other important benefits are to identify other family members, particularly young siblings who should be evaluated for possible hearing loss or associated medical conditions and provide recurrence information for family planning. Parents (caregivers) who are deaf may view hearing loss as a difference that is a normal variation in humans, not a disorder. Since it is a common misconception that the only purpose of a genetic evaluation is to reduce the recurrence of a condition in the family, parents may be reluctant to accept a genetic referral. The medical home should counsel families regarding the role of genetics in the medical management and intervention strategies needed.

  Genetic factors are thought to cause between 50% to 60% of congenital hearing loss in children. In about 70% of cases the hearing loss occurs as an isolated trait and is called nonsyndromic. The remaining 30% is classified as syndromic. There are at least 75 genetic types of nonsyndromic hearing loss and over 400 genetic types of syndromic hearing loss.

A very common “nonsyndromic” hearing loss is caused by one gene known as Connexin 26. Connexin 26 is the cause in about 1/3 of all children with nonsyndromic hearing loss.

Some common genetic syndromes in which hearing loss is one of the known characteristics is Down syndrome, Usher syndrome, Treacher Collins syndrome, Fetal alcohol syndrome, Crouzon syndrome and Alport syndrome.

- Monitor timely follow up with an audiologist. An immediate diagnostic audiologic evaluation should be scheduled when there is concern related to change in hearing. Bilateral sensorineural and permanent conductive hearing loss should be monitored every 3 months for children age 0-3 years. Unilateral hearing loss should be monitored every 3 months during the first year and then on a 6-month basis after the first year to rule out changes in the normal ear. Many unilateral hearing losses progress to bilateral.

- Monitor middle ear status to ensure the presence of middle ear fluid does not further compromise hearing. Transient conductive hearing loss or a mixed loss (sensorineural with conductive component) should be monitored after medical treatment, completion of antibiotic or PE tubes at least on a 3-4 month basis.

- Monitor consistent use of amplification if appropriate for the family. Hearing aid use is an important option for many children. Hearing aid(s) should be fit by one month after confirmation of the hearing loss by a pediatric audiologist who specializes in fitting hearing aids on infants and young children.

- Refer to early intervention services. Upon confirmation of the hearing loss a referral to Early On (1-800-327-5966) must take place within 2 working days. Intervention services should be in place as soon as possible or with 45 days of identification of the hearing loss.