Infants who may be deaf or hard of hearing
Special considerations for Otolaryngology

Suggestions for practice of special interest to Otolaryngologists
Drawn from the report of the Joint Committee on Infant Hearing[1]

PRESENTING PATIENTS

Infants and children through 3 years of age with possible hearing loss initially indicated by:

- Parent concern
- Did not pass newborn hearing screening
- One or more risk factors for hearing loss, regardless of screening result (See Risk Indicators section)
- Audiologic or other test results suggesting peripheral hearing loss in one or both ears

No child is too young to test. Hearing status should be confirmed by audiologic evaluation no later than 3 months of age.

Early intervention services[2] should begin no later than 6 months.

Delay in obtaining intervention services can compromise language development and reduce later communication and school performance.

If audiologic evaluation[3] has not been completed, refer immediately to pediatric audiologist.

"Don't delay—hearing can be evaluated at any age".

—Craig A. Buchman, MD, FACS, University of North Carolina School of Medicine

KEY PRACTICE POINTS

- ENT clearance for hearing aid fitting should not be delayed while other medical exams are on-going or while the etiology for the hearing loss is being explored.
- Clearance by the ENT should be interpreted as meaning "no medical contraindication."
- Hearing aid fitting should take place within 1 month of diagnosis, if the child's parents/caregivers choose to do so.
- Hearing aid fitting should not be delayed when there is transient effusion.
- Hearing aids can and should be fitted for chronic middle-ear effusion associated with cleft palate and Down Syndrome.
- At all times, observe and respond to parents' stated and unstated questions and emotions, working through stages of denial, grief, anger, etc.
- Once any degree of hearing loss is diagnosed (including unilateral or bilateral hearing loss due to aural atresia and "transient" hearing loss associated with cleft palate), referral to Early Intervention must be made within 7 working days of confirmation of hearing loss.
- Ongoing care for all hearing losses ("transient" and permanent) by an audiologist is essential, in addition to medical care.

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RECOMMENDATIONS FOR CLINIC VISIT

HISTORY	• Review all medical records and case history information for risk factors associated with congenital hearing loss and/or late-onset or progressive hearing loss (See Risk Indicators). • Review the audiological record to assess need for further audiological testing. • Elicit whether there is parental concerns about the child's hearing.
PHYSICAL EXAM	• Pay special attention to malformed features, and presence of ear pits or brachial cysts.
LABORATORY TESTS	• If hearing loss etiology remains undetermined, consider referring family to a genetics specialist to determine possible genetic etiology. • To assess possible auditory neuropathy or when medical causation is unknown, may consider CT/MRI of the temporal bones.
REFERRALS	• All infants with confirmed hearing loss should be referred to appropriate specialists including an ophthalmologist JCIH 2007[1]. • Every infant with confirmed hearing loss should be evaluated by an otolaryngologist who has knowledge of pediatric hearing loss and have at least 1 examination to assess visual acuity by an ophthalmologist who is experienced in evaluating infants. • Verify/ Refer to Early Intervention within 7 days of confirmation of hearing loss. http://ectacenter.org/ • Ask parents if referral to parent support has been done.

RISK INDICATORS

Risk indicators associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood

Risk indicators that are marked with a "§" are of greater concern for delayed-onset hearing loss.

1. Caregiver concern§ regarding hearing, speech, language, or developmental delay.
2. Family history§ of permanent childhood hearing loss.
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO,§ assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.
4. In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,§ such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
8. Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
9. Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.
11. Chemotherapy.§

[1] **Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs:**
Joint Committee on Infant Hearing. Pediatrics Vol. 120 No. 4 October 1, 2007 pp. 898-921 (doi: 10.1542/peds.2007-2333)
http://pediatrics.aappublications.org/content/120/4/898.full?ijkey=oj9BAleq21OlA&keytype=ref&siteid=aapjournals

[2] **Early Intervention Services:** Services that are provided to young children (Birth to three) who have or at risk for disabilities or special needs. Services are family-based, ranging from speech and occupational therapy to general intervention and instruction.

[3] **A complete audiologic evaluation for infants consists of (may vary depending on age, circumstances, diagnosis):**

- Acoustic immittance (tympanometry and acoustic reflex thresholds {high-frequency probe tone for infants under 6 months of age})
- Otoacoustic emissions (Distortion Product or Transient)
- Auditory Brainstem Response (Air- and bone-conduction) for specific frequencies (in particular, 500 Hz and 2000 Hz, others as time permits). ABR protocols to assess possible Auditory Neuropathy when indicated.
- Auditory Steady-State Response as a secondary measure (after tone-burst ABR).
- Behavioral Audiometry at age- and developmentally-appropriate stages.