Hearing Loss Facts

Prevalence and identification:
- Approximately 3 in 1,000 babies are born with permanent hearing loss, making it the most common birth defects in America (Ross et al., 2008).
- Testing only the “at-risk” children results in the identification of only 50% of children with hearing loss.

Average age of identification:
- Reduced, close to 2 months of age with the implementation of newborn hearing screening programs.
- Without EHDI (Early Hearing Detection & Intervention) programs, the average age of identification for HL is age 2.5–3.0 years.

GOAL OF EARLY HEARING DETECTION AND INTERVENTION

Hearing loss can affect a child’s ability to develop speech, language, and social skills. Early detection of hearing loss enables us to provide interventions that can maximize the child’s developmental potential.
- Hearing screening before age 1 month.
- Diagnostic audiologic evaluation before age 3 months for infants who do not pass the screening.
- Enrollment of infants identified with HL in early intervention services before age 6 months.

CALIFORNIA NEWBORN HEARING SCREENING PROGRAM

- An automated Auditory Brainstem Response and/or automated Otoacoustic Emissions (OAEs) is used for inpatients and outpatient hearing screenings on infants.

If a child REFERS (or fails an) inpatient hearing screening then that child will be referred for an outpatient screening. If the child refers/fails again on outpatient newborn hearing hearing screening then that infant will be referred for a DIAGNOSTIC Auditory Brainstem Response Evaluation (ABR) to:
- confirm the presence or absence of hearing loss.
- determine the TYPE and DEGREE of hearing loss.
- obtain frequency and ear specific information.
- rule out auditory neuropathy.

DIAGNOSTIC AUDITORY BRAINSTEM RESPONSE (ABR)

- Auditory Brainstem Response (ABR): averaging of brain wave responses to sound.
  - Natural sleep ABR — Children less than 5 months of age are tested in a natural sleep state. It is necessary for the child to sleep approximately 2 hours for the assessment.
  - Sedated ABR — Children over 5 months of age are sedated in conjunction with nursing personnel for ABR testing. Performed at beside for in-patients in NICU, in the surgery center, and in the Main OR for children at risk.
AUDIOLOGIC PROTOCOLS

To rule out a hearing loss with older kids who come to us (6 months and up), the audiologist will use several cross-check measures:

- **Behavioral Audiologic Measurements**—Behavioral observational audiometry, Visual Reinforcement audiometry, Play audiometry
- **Objective Measures**—OAes, tympanometry, evaluation of middle ear muscle reflexes, and ABR for children unable to perform behavioral audiologic assessments

IDENTIFYING A HEARING LOSS

The goal of the objective and behavioral measures when a hearing loss is present is to identify:

- **Type**—conductive, sensorineural, mixed, neural (auditory neuropathy spectrum disorder)
- **Degree**—mild, moderate, severe, profound
- **Configuration**—flat, sloping, cookie bite, high-frequency, low frequency, etc.
- **Appropriate Counseling and Management**

AUDITORY NEUROPATHY SPECTRUM DISORDER

- Patient displays auditory characteristics consistent with normal outer hair cell function and abnormal neural function at the level of the VIIIth (vestibulo-cochlear) nerve.
- Prevalence of 1 in 10 of children with hearing loss, relatively uncommon.
- These characteristics are observed on clinical audiological tests as present otoacoustic emissions, in the presence of an absent or severely abnormal ABR, and absent middle ear muscle reflexes.
- These patients are distinguished from patients with space-occupying lesions, such as VIIIth nerve tumors, or multiple sclerosis, in that radiological evaluation yields normal results and even the most peripheral responses from the VIIIth nerve are absent.

DEGREE’S OF HEARING LOSS

- **Profound (over 90 dB)**
  - No response to speech
  - Must use visual cues to communicate
  - May respond to vibration from sound
- **Severe (71dB-90dB)**
  - Speech heard only when loud and at close range
  - Difficulty hearing and understanding always
  - May show awareness, not recognition to sound (clap)
- **Moderate (41-55dB)**
  - May do well close up and face to face but problems hearing across a room
  - Difficulty discriminating consonants
- **Mild (26-40dB)**
  - Trouble understanding (Hears sound, but not all info)
  - More trouble at distance or in noisy environment
- **Moderately severe (56-70dB)**
  - Misses nearly all speech signal necessary to develop normal speech and language
  - May respond to loud environmental sounds

ONCE A CHILD HAS A DIAGNOSED HEARING LOSS

- Once identified, a referral to an ENT will be made for a work-up and to obtain medical clearance for hearing aids if appropriate.
- A baby as young as 3 weeks of age CAN be fit with hearing aids.
- A child’s hearing loss should continue to be monitored and appropriate referrals made for early intervention services.
- Child is routinely followed within our department every 3-6 months for routine audiological evaluations, and hearing aid checks.
- Child may be referred to the cochlear implant team depending on degree and configuration of hearing loss.
Locations and Services

UCSF Benioff Children's Hospital Oakland
744 52nd St., Oakland CA 94609
Phone: (510) 428-3344

Oakland Services:
• Comprehensive newborn hearing screening
• Sedated / non-sedate ABR assessments
• Behavioral audiologic assessments
• Cochlear Implant evaluation and implantation, follow-up mapping and management
• Therapy, auditory training and aural habilitation/rehabilitation
• Cortical auditory evoked potential assessments
• Family counseling and support following diagnosis

UCSF Benioff Children's Hospital Oakland
Walnut Creek Campus
2401 Shadelands Dr., Walnut Creek, CA 94598
Phone: (925) 979-3440

Walnut Creek Services:
• Comprehensive newborn hearing screening
• Sedated / non-sedate ABR assessments
• Central Auditory Processing Assessments
• Behavioral audiologic assessments
• Therapy, auditory training and aural habilitation/rehabilitation
• Cortical auditory evoked potential assessments
• Family counseling and support following diagnosis

UCSF Benioff Children's Hospital Oakland
Specialty Care Brentwood
1181 Central Blvd., Suite B, Brentwood, CA 94513
Phone: (888) 530-3034

Brentwood Services:
• Comprehensive newborn hearing screening
• Sedated / non-sedate ABR assessments
• Behavioral audiologic assessments
• Cortical auditory evoked potential assessments
• Family counseling and support following diagnosis