EVALUATION BEFORE 3 MONTHS OF AGE

At the Second or Third Well Child Visit

☑ For those children who did not pass rescreening, check that a diagnostic auditory brainstem response test (ABR) has been scheduled or completed.

ABR Results	Action Required	Purpose/Resources
Normal	Determine if baby should be monitored for late onset of hearing loss.	See page 1-4 for more information about monitoring for late onset hearing loss.
Unknown or incomplete	Under 3-6 months: Refer for unsedated ABR or other tests by an audiologist. Over 3-6 months: Refer for sedated ABR. Over 6 months: Refer for behavioral hearing test.	Sites that provide unsedated ABRs differ as to the maximum age they will evaluate. Children must be able to sleep soundly during the test. Not all sites offer sedated ABRs. Children must be awake and alert to respond for behavioral hearing tests. (Sedated ABR testing may also be required to determine hearing status.) See page 2-2 for pediatric audiologists.
Hearing loss	Refer for EI services and amplification before age 6 months. (Hearing aids can be fit by 1 month of age.)	Refer the child for EI services by calling the Hawaii Keiki Information Services System (H-KISS) at 808-594-0066 or toll-free at 1-800-235-5477. See page 3-1 for more about EI. See page 2-2 for pediatric audiologists.

Resources for Diagnostic Audiological Evaluation

Providers on this list applied for and were awarded contracts with the Department of Health to provide pediatric audiology services. For additional audiologists in your area, please consult your phonebook.

DOH Contracted Audiologists	Appt. Phone #
Audiology Associates Hawai`i (Oahu–Honolulu, Aiea)... *Provides unsedated ABR tests at both locations and behavioral tests in Honolulu.*	486-5000
Big Island Hearing Center, LLC (Big Island–Hilo)..... *Provides behavioral tests and hearing aids.*	935-1299
Castle Medical Center Audiology Department (Oahu) ... *Provides unsedated ABR tests for infants under 4 months old.*	263-5055
Hawai`i Professional Audiology (Oahu)..... *Provides behavioral tests and hearing aids.*	597-1877
Hawai`i Professional Audiology (Kauai)..... *Provides behavioral tests and hearing aids.*	245-1530
Island Audiology (Oahu) ..... *Provides behavioral tests and hearing aids.*	375-2253
June Uyehara-Isono, Inc. (Oahu, Big Island–Hilo, Kona) *Provides behavioral tests and hearing aids.*	877-524-1432
Kapiolani Medical Center for Women and Children (Oahu) *Provides sedated and unsedated ABR tests, behavioral tests and hearing aids.*	*Behavioral tests:* 535-7000, ext. 1 *ABR tests & hearing aids:* 983-8230
Kapiolani Medical Center – Pali Momi (Oahu) ..... *Provides behavioral tests and hearing aids.*	535-7000, ext. 1
Maui Medical Group (Maui)..... *Provides non-sedated click ABR tests, behavioral tests and hearing aids.*	242-6464

Auditory Brainstem Response (ABR) test=Baby is asleep or sedated
Behavioral test=Child is awake and alert (usually at least 6-7 months old)

More about Diagnostic Audiological Evaluation

Diagnostic Testing Protocols

Unless medically contraindicated, diagnostic audiological evaluation should be completed before 3 months of age for children who do not pass newborn hearing screening. The preferred protocol in Hawai`i is to administer ear specific:

- 1000 Hz tympanometry
- comprehensive otoacoustic emissions
- click auditory brainstem response (ABR)
- at least 500 and 4000 Hz tone burst ABR testing (tone burst ABR scores corrected for age when determining hearing status)
- bone conduction testing, if air conduction results indicate hearing loss

The preferred protocol for children over 6 months of age includes ear specific behavioral assessment of hearing instead of or in addition to ABR testing. The desire for behavioral hearing test results should not delay amplification, as protocols designed for use with infants are now available.[6]

Methods Used to Assess Hearing in Infants

Tympanometry

Tympanometry, including acoustic reflexes, is used to assess middle ear function. Due to the plasticity of a newborn's ear canal, low frequency (226 Hz) tympanometry is not considered accurate until approximately 6 months of age.

High frequency (1000 Hz) tympanometry is therefore preferred for children under 6 months of age, although this procedure may not be available in every location.

Comprehensive Otoacoustic Emissions (OAE) Testing

OAE testing is used to identify outer hair cell dysfunction in the cochlea. Comprehensive, frequency specific OAE testing should be performed to help determine hearing status.

Auditory Brainstem Response (ABR) Testing

ABR testing is used to assess functioning of the cochlea, auditory nerve and auditory brainstem pathways. Comprehensive ABR tests are the accepted method of evaluating infants who are less than 6 months developmental age. Click and frequency-specific tone burst ABR tests allow an estimation of hearing thresholds for diagnostic purposes and hearing aid fitting. They may also indicate the possibility of conductive hearing loss. Tone burst ABR scores should be corrected for age when determining hearing status. Bone conduction ABR testing should be provided if air conduction results indicate hearing loss.

Behavioral Assessment

Behavioral hearing tests should be provided to validate physiological results. Behavioral test results become more accurate as a baby approaches 6 months of age. Bone conduction behavioral testing should be provided if air conduction results indicate hearing loss.

Department of Health Programs

Upon request, the Newborn Hearing Screening Program assists families in scheduling appointments for infants who need diagnostic evaluation to rule out permanent hearing loss. Children under three years of age who are not eligible for Part C early intervention can participate. Health insurance is billed first, if the child has coverage. The Department of Health is billed last. Families are not billed.

The Department of Health is Lead Agency for Part C early intervention (EI) in Hawai`i. Part C Care Coordinators assist families of EI enrolled children in scheduling evaluations, hearing aid related services (not including purchase of hearing aids), and auditory rehabilitation. Families are encouraged to allow health insurance to be billed first, if the child has coverage. The Department of Health is billed last. Families are not billed. EI has a lending library which includes FM systems.

For income eligible families, the Children with Special Health Needs Program can assist families of children diagnosed with hearing loss in obtaining ongoing evaluations, hearing aids, and hearing aid related services. Health insurance is billed first, if the child has coverage. The Department of Health is billed last. Families are not billed.

The Department of Health also has loaner hearing aids available for short term loan to children enrolled in EI and/or the Children with Special Health Needs Program. For more information, call 808-733-9067.

More about Hearing Loss

Types of Hearing Loss

Hearing loss can be present from birth or acquired after birth. About 60% of congenital hearing loss is associated with genetic causes.[5] For many children, the associated clinical findings regarding the etiology (cause) of their hearing loss is important in patient care. See Appendix D for additional information from the Hawai`i Genetics Program.

Hearing can change over time and the incidence of hearing loss increases with age. According to statistics compiled by the National Institute on Deafness and Other Communication Disorders (NIDCD), hearing loss affects 17/1000 children under age 18 and 314/1000 adults older than age 65. Nearly 28 million Americans have hearing loss in at least one ear.[7]

Conductive hearing loss occurs when sound cannot pass through the outer or middle ear into the inner ear. It can be caused by reversible conditions such as impacted wax, a punctured eardrum, fluid in the middle ear, or a middle ear infection. Conductive hearing loss can often be corrected through medical treatment or surgery. Without treatment, conductive hearing loss may become permanent.

Permanent conductive hearing loss occurs when sound cannot pass through the outer or middle ear into the inner ear due to conditions that cannot be corrected by medical

treatment or surgery or due to conditions that are expected to be of life-long or long-term duration. It can be caused by acquired conditions such as middle ear scarring from ear infections or injury and by congenital conditions such as aural atresia (no ear) or microtia (small ear).

Sensorineural hearing loss occurs when the auditory nerve or hair cells in the inner ear are damaged. Causes may include noise, aging, infection, head trauma, toxic medications or an inherited condition. Researchers estimate that 30-40% of children with confirmed hearing loss also demonstrate developmental delays or other disabilities.[4] The most commonly found additional disability is visual impairment. Mild cognitive disabilities are the next most common. Specific learning disabilities are third.

Mixed hearing loss occurs when both conductive and sensorineural hearing losses are present.

Auditory neuropathy affects a small number of children with hearing loss. Although many appear to be NICU graduates, the exact prevalence of auditory neuropathy is unknown. This disorder is identifiable through combined use of OAE and ABR tests. This disorder is characterized by normal OAE results and absent or abnormal ABR results, suggesting functioning outer hair cells with abnormal neural conduction. Some children with auditory neuropathy benefit from amplification.

Unilateral vs. Bilateral Hearing Loss

Children with hearing loss in one ear should receive the same medical assessment to determine etiology as children with bilateral hearing loss, as well as audiological follow-up to monitor for progressive hearing loss and early intervention to support language acquisition. Some will develop bilateral hearing loss by the time they enter school. Many will experience early language delays and may have difficulty understanding speech depending upon the amount of background noise or their distance from the person speaking.

Degrees of Hearing Loss

Children without amplification who have mild hearing loss may be unable to hear as much as 50% of spoken language. Those with moderate hearing loss may be unable to hear as much as 90% of spoken language. Thus, any degree of hearing loss should be considered developmentally and educationally significant. The Hawai`i Department of Health utilizes the scale from the Centers for Disease Control and Prevention annual Early Hearing Detection and Intervention data report when describing degree of hearing loss:

Degree	Responds at
Mild Hearing Loss:	21-40 dB
Moderate Hearing Loss:	41-70 dB
Severe Hearing Loss:	71-90 dB
Profound Hearing Loss:	91 or greater dB