

SCREENING BEFORE 1 MONTH OF AGE

At the First Well Child Visit

- Check to see if the infant was screened for hearing loss during the inpatient stay. If the infant was not born at a hospital, it is likely that the baby has not been screened.

Screening Status	Action Required	Purpose/Resources
Passed both ears (OAE or AABR for well-babies; AABR if in a NICU for more than 5 days.)	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.
Did not pass	Assure that rescreening takes place before <u>1</u> month of age.	Contact the newborn hearing screening coordinator in the birthing hospital or the DOH Newborn Hearing Screening Program to arrange rescreening. See page 1–3 for locations.
Unknown or incomplete	Assure that screening takes place before <u>1</u> month of age.	Contact the birthing hospital's newborn nursery or the DOH Newborn Hearing Screening Program to request screening results and/or arrange screening. See page 1–3 for locations.

The likelihood of congenital hearing loss is approximately:

- 1 baby per 1000 in the well-baby population.
- 10 babies per 1000 for neonatal intensive care unit graduates.

Joint Committee on Infant Hearing Update (2006)³

SCREENING – continued

At the Second Well-Child Visit

- Check to see if outpatient screening has been completed for infants who missed or did not pass initial screening. This should be completed within 2–4 weeks of the initial screen.

Screening Status	Action Required	Purpose/Resources
Passed rescreen in both ears	Determine if baby is at high risk for late onset of hearing loss.	Both ears should be rescreened, as the baby's hearing may have changed since the previous screen. Rescreening should include AABR if the baby did not pass inpatient AABR or does not pass rescreening with OAE. See page 1–4 for more information about monitoring for late onset hearing loss.
Did not pass rescreen or needs evaluation instead of rescreen	Refer for evaluation and coordinate insurance pre-authorization.	See page 1–9 for more information about screening versus evaluation. Diagnostic ABR testing is available on Oahu and Maui for children residing anywhere in the state. Children on Kaua`i and the Big Island may first be referred to a local pediatric audiologist to confirm if diagnostic ABR is needed. See page 2–2 for pediatric audiologists.
Unknown or incomplete	Assure that screening takes place before 1 month of age.	Contact the birthing hospital's newborn nursery or the DOH Newborn Hearing Screening Program to request screening results or arrange screening. See page 1–3 for locations.

Resources for Screening

Phone numbers may change. If a more current number is needed, please contact the hospital's well baby nursery.

Castle Medical Center	808-263-5270
Hilo Medical Center	808-974-4715
Maui Memorial Medical Center (Screening by Imua Family Services)	808-244-7467
Kaiser Permanente Moanalua Medical Center (Kaiser members only)	808-432-8292
Kapiolani Medical Center for Women and Children	808-983-8230
Kona Community Hospital	808-322-4416
Molokai General Hospital (Women's Health Center)	808-553-3145
North Hawaii Community Hospital	808-881-4771
The Queen's Medical Center	808-547-4213
Tripler Army Medical Center	808-433-3197
West Kauai Medical Center/KVMH	808-338-9431
Wilcox Memorial Hospital	808-245-1433

In addition to the birthing hospitals listed above, hearing screening is available through pediatric audiologists (see page 2-2). For more information, please contact the Department of Health Newborn Hearing Screening Program at 808-594-0042.

When to Pay Extra Attention to Hearing

Infants with late onset hearing loss often have no identifiable risk factors. Therefore a more inclusive strategy of surveillance for all children is recommended. All infants should be monitored for developmental milestones, auditory skills, parental concerns about hearing, speech, and language, and middle ear effusion. All infants should be evaluated with an objective, standardized screen of global development with a validated assessment tool at 9, 18, and 24–30 months or at any time if the parent or caregiver has concern.

Healthcare providers should pay extra attention to hearing and speech–language development if a baby does not pass the newborn hearing screening test in one or both ears. However, every child's hearing should be monitored periodically throughout early childhood because:

- Mild (but still developmentally and educationally significant) hearing loss may be present even if a baby passes a screening test.
- Hearing can change over time.

Middle ear status should be carefully assessed at all well–child visits by pneumatic otoscopy and/or tympanometry. Children with persistent middle ear effusion for 3 or months should be referred for otologic evaluation.

The Joint Committee on Infant Hearing suggests that the following indicators put an infant at risk for progressive or

delayed-onset sensorineural and/or conductive hearing loss.⁴ In Hawai`i, these risk indicators are often called “hearing loss predictors” or “indicators”. Children with risk indicators should be referred for an audiologic assessment at least once by 24–30 months of age. Children who have indicators with asterisks (*) have a higher probability of hearing loss and should have more frequent audiologic assessments. See “More About Medical Follow-up for Hearing Loss” on page 3–2.

Parental or caregiver concern* regarding hearing, speech, language, or developmental delay – Parents and caregivers are often the first to notice when their child is experiencing hearing, speech, language or developmental delays. Except for children with profound bilateral hearing loss, the chief concern first identified by parents regarding a child with hearing loss is usually “speech delay”.

Family history* of permanent childhood hearing loss (inherited) – About 60% of hearing loss present at birth or beginning in the first few months of life is due to an alteration in one or more of an estimated 400 genes that can cause deafness.⁵ If a family member had permanent hearing loss beginning in childhood, additional questions should be asked to find out if the hearing loss was acquired due to infection or trauma.

Neonatal intensive care for more than 5 days or any of the following regardless of length of stay: extra-corporeal membrane oxygenation (ECMO)*, assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin)

or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion – Conditions that require prolonged neonatal intensive care may lead to permanent hearing loss.

In utero infections, such as cytomegalovirus (CMV), herpes, rubella, syphilis, and toxoplasmosis* – Infections that can cross the placental barrier and invade fetal tissue have been linked to sensorineural hearing loss in infants. The developing auditory system is at greatest risk during the first trimester of pregnancy. An infant's hearing is most likely to be damaged if the infection occurs during this period. These infections often go unrecognized due to a lack of clinical symptoms in the mother.

- Cytomegalovirus (CMV)*: CMV is a leading cause of fetal viral infection in the United States and can cause congenital or delayed onset sensorineural hearing loss that is often progressive. It can also cause other developmental disabilities.
- Herpes: Either herpes simplex 1 or 2 may cause severe to profound sensorineural hearing loss.
- Rubella: Rubella is commonly called “German measles.” When hearing loss occurs, 50% of affected babies will have severe to profound hearing loss in both ears. Hearing loss may be progressive. In addition to hearing loss, this infection may cause heart disorders, low birth weight, developmental disability, and vision loss.

- Syphilis: Congenital syphilis may cause sensorineural hearing loss that is sudden, progressive, or fluctuating.
- Toxoplasmosis: This infection is caused by a protozoan parasite and affects approximately 1 /750 newborns in the U.S. In addition to hearing loss, toxoplasmosis may cause seizures, developmental disability, and vision loss.

Craniofacial anomalies, including those involving the pinna, ear canal, ear tags, ear pits, and temporal bone – Hearing may be affected if the head, face, neck or ears are shaped or formed in a different way than usual. Other examples of craniofacial anomalies that may be associated with hearing loss include cleft palate, shortened neck, webbed neck, abnormal head circumference, aural atresia (missing outer ear) or microtia (small outer ear), or low set ears.

Physical findings such as white forelock, associated with a syndrome known to include a sensorineural or permanent conductive hearing loss. A white forelock is a characteristic of Waardenburg's Syndrome, which may be associated with sensorineural hearing loss that can vary from profound to none.

Syndromes associated with hearing loss or progressive or late-onset hearing loss* – Conditions such as neurofibromatosis, osteopetrosis and Usher's syndrome may cause progressive hearing loss. Other frequently identified syndromes that can affect hearing include Waardenburg, Alport, Pendred, Jervell and Lange-Nielson.

Neurodegenerative disorders*, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot–Marie–Tooth syndrome, cause both neurological functioning and hearing to deteriorate over time.

Culture positive postnatal infections associated with sensorineural hearing loss*, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis – Some postnatal infections may cause hearing loss in young children. The younger the child when the infection occurs, the greater the potential impact a resulting hearing loss may have on speech and language acquisition. Bacterial meningitis is a leading cause of acquired deafness in infants. Most cases result in bilateral severe to profound sensorineural hearing loss.

Head trauma, especially basal skull/temporal bone fracture* requiring hospitalization – Trauma that causes loss of consciousness or skull fracture, with or without bleeding from the ear, may result in conductive hearing loss due to bleeding, perforation of the tympanic membrane, or disruption of the ossicular chain. Sensorineural hearing loss may occur if the temporal bone housing the inner ear is damaged.

Chemotherapy* – Chemotherapeutic agents such as cisplatin and carboplatin are ototoxic and may lead to late onset or progressive sensorineural hearing loss.

More about Hearing Screening

Screening does not directly measure true hearing or the brain's ability to process sounds. Screening results indicate the likelihood of a greater than 30–40 dB hearing loss at the time of screening. As mentioned in the previous section, it is important to realize that some children with mild (but still developmentally and educationally significant) hearing loss may not be identified through screening and that hearing can change over time. Healthcare providers should therefore remain alert for possible hearing loss as they provide ongoing care for infants and young children, regardless of whether or not the child passed newborn hearing screening.

Screening Protocols

Most well baby screening programs in Hawai`i perform an otoacoustic emissions (OAE) screening test first, followed by an automated auditory brainstem response (AABR) screening test if the child does not pass OAE. NICU screening programs use AABR to better detect conditions such as auditory neuropathy or use a combination of OAE and AABR. Screening is available for homebirth babies on an outpatient basis at birthing hospitals and through pediatric audiologists. Both ears are screened, since hearing loss is developmentally and educationally significant even if only one ear is affected.

Well babies who do not pass OAE or AABR during the first stage of the newborn hearing screening process are referred for rescreening before 1 month of age. Both ears are

rescreened, as the baby's hearing may have changed since the previous screen. AABR is recommended if the baby didn't pass a previous AABR screen or only had an OAE. Babies who don't pass rescreening are referred for comprehensive diagnostic audiological evaluation, including auditory brainstem response (ABR) testing, before 3 months of age.

Diagnostic audiological evaluation is recommended instead of rescreening for babies not passing inpatient screening when:

- Their head, face, neck or ears are shaped or formed in a different way than usual, including babies who have atresia, microtia, Down syndrome or cleft palate; or
- They are in a NICU for more than 5 days; or
- They have other medical conditions that require diagnostic testing to determine hearing status.

Diagnostic auditory brainstem response (ABR) testing is available on Oahu and Maui for children born anywhere in the state. If the family prefers, children not passing hospital rescreening on Kaua`i or the Big Island may be seen by a pediatric audiologist on their home island for additional testing. If the local audiologist determines that a diagnostic ABR is needed, the baby will then be referred to Oahu or Maui for evaluation before 3 months of age. Children not passing newborn hearing screening on islands where diagnostic ABR testing is available are referred immediately for a diagnostic ABR and other tests to determine if they have a hearing loss.

With prior authorization, the Hawai`i Department of Health Newborn Hearing Screening Program can help with some test and travel costs, if not covered in full by the child's insurance.

Screening Methods

Either of the following methods can usually be performed by trained personnel in about 5 to 20 minutes. Babies are often screened after 12 hours of age. Screenings before 24 hours of age are more likely to be affected by the presence of vernix/debris in the ear canal. Screenings for older children are more likely to be affected by middle ear effusion.

Auditory Brainstem Response Screening (AABR)

In AABR screening, clicking sounds are introduced through an earphone or over-the-ear coupler. Electroencephalographic (EEG) response is measured through electrodes placed on the baby's head and neck. The equipment automatically generates a "pass" or "refer" screening result based on preset criteria.

AABR screening provides information about the auditory pathway up to the brainstem, including the middle ear, the inner ear and the eighth cranial nerve. Central auditory processing and neuropathy problems, which are more likely when babies have conditions leading to prolonged NICU stays, can be detected. AABR screening is recommended for babies in the NICU, as well as for rescreening well-babies who do not pass otoacoustic emissions screening.

Otoacoustic Emissions Screening (OAE)

In OAE screening, an earphone with a built-in microphone is placed in the ear canal to introduce sounds (clicks or tones) and listen for cochlear outer hair cell response (otoacoustic emissions). If the outer hair cells of the cochlea have been damaged or if incoming sound is blocked before reaching the cochlea, otoacoustic emissions will not be produced. The equipment automatically generates a “pass” or “refer” screening result based on preset criteria.

OAE screening provides information about the auditory pathway up to and including the cochlea. Central auditory processing and neuropathy problems, which affect less than 1% of children with hearing loss, will not be detected by OAE screening. In Hawai`i, the well-baby population usually receives OAE screening. Well-babies who do not pass OAE screening are usually rescreened with AABR to obtain information about the auditory pathway up to the brainstem, including the middle ear, the inner ear and the eighth cranial nerve.