

Joint Committee on Infant Hearing (JCIH) High Risk Indicators - Explanation

The Joint Committee on Infant Hearing (JCIH) High Risk Indicators list risk criteria that is often associated with infant and childhood hearing loss. It is vital to obtain complete and accurate information about the child's prenatal and birth history, results of the newborn hearing screening, and the presence of specific early childhood conditions in order to determine if factors associated with hearing loss are present. The identification of risk indicators is an essential component of a comprehensive hearing screening program for children ages birth through 2, and also for those babies who have not received Universal Newborn Hearing Screening. However, use of risk indicators alone will identify only 40-50% of infants with hearing loss.

The JCIH Year 2007 Position Statement identified eleven indicators associated with hearing loss. It is vital to obtain complete and accurate information about the child's prenatal and birth history, results of the newborn hearing screening, and the presence of specific early childhood conditions in order to determine if factors associated with hearing loss are present.

The JCIH recommends that all infants with and without risk indicators be monitored during routine medical care consistent with the AAP periodicity schedule. All infants with a risk indicator for hearing loss should be referred to an audiologist at least once by 24 to 30 months of age. Children with risk indicators that are highly associated with delayed-onset hearing loss, such as having received ECMO or having CMV infection, should have more frequent audiological assessments.

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

1. **Caregiver concern** regarding hearing, speech, language, or developmental delay.
2. **Family history** of permanent childhood hearing loss.
3. **All infants with or without risk factors requiring neonatal intensive** care for greater than 5 days, including any of the following: ECMO,* assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/lasix). In addition, **regardless of length of stay: hyperbilirubinemia requiring 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 fractures that requires hospitalization.

11. **Chemotherapy**.

Explanation of Risk Indicators

All of the risk indicators have been shown to have a potential impact on hearing in young children. A brief explanation of each indicator is provided below in order to ensure that accurate and pertinent information is obtained from each parent or caregiver.

1. Caregiver concern regarding hearing, speech, language, or developmental delay.

It is important that hearing loss be identified as early as possible to prevent speech, language and other developmental delays. Most parents are reliable reporters of their child's development. The National Institute of Health Consensus Statement on Early Identification of Hearing Loss in Infants and Young Children (1993) stated that as many as 70% of deaf and hard of hearing children are identified because of parental concern.

2. Family history of permanent childhood hearing loss.

This question is aimed at identification of hereditary (genetic) hearing loss from both maternal and paternal family members, living or deceased, when known. However, a family history of hearing loss is not necessary for the cause of a child's hearing loss to be genetic. Hearing loss that is genetic is most often sensorineural. It is important to ensure that the relative's hearing loss was not acquired (such as those resulting from meningitis, noise exposure, chemotherapy, or the aging process). Acquired hearing losses are not inherited. The type of loss which is inherited is typically present at a very young age. Semi-annual hearing screening is recommended because hereditary hearing loss may have delayed onset.

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 medication (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.

Infants admitted to NICU are at greater risk for hearing loss. For example, infants with very low birth weight are at increased risk for both sensorineural and conductive hearing loss. Additionally, jaundice is a condition which occurs when there is too much by-product from the liver in the blood, eventually resulting in high bilirubin levels. Too much bilirubin (hyperbilirubinemia) is ototoxic and may cause hearing loss. Premature infants and infants with low birth weight are at greater risk for high bilirubin

levels. Low bilirubin levels (slight jaundice) typically do not affect hearing. Also, infants who require prolonged use of mechanical ventilation are at risk for hearing loss (e.g., persistent pulmonary hypertension, conditions requiring the use of extracorporeal membrane oxygenation (ECMO)).

4. In utero infections, such as CMV, herpes, rubella, syphilis, and toxoplasmosis.

The presence of these infectious agents has been linked to hearing loss in children. The majority of infections in pregnant women involve the upper respiratory and gastrointestinal tract and are not known to cause hearing loss. However, some infectious agents, contracted by the mother during pregnancy, may cross the placental barrier and invade fetal tissue. Severe infections, especially those occurring in the first trimester, can be related to hearing loss since this is when the auditory system develops. Many infections go unrecognized due to the lack of clinical symptoms in the mother. Semi-annual hearing screening is recommended due to the potential of delayed onset sensorineural hearing loss.

Cytomegalovirus (CMV): A virus in the herpes group and the leading cause of fetal viral infection in the U.S, the infection is most often asymptomatic in the mother. CMV can cause sensorineural hearing loss, which varies in severity, may have a delayed onset, may be unilateral, and is often progressive.

Herpes: Either systemic or simplex 1 or 2 and in the same family as the CMV virus, herpes may cause severe to profound sensorineural hearing loss.

Toxoplasmosis: Caused by a protozoan parasite, the infection is usually asymptomatic in the mother. The incidence of intrauterine toxoplasmosis averages one case per 750 deliveries in the U.S. Infection during the first trimester appears most likely to adversely affect the fetus. Central nervous system involvement, as well as mental retardation, seizures, and ocular disease are frequently seen.

Rubella (German Measles): Occurring within the first trimester of pregnancy, rubella poses a serious risk to the developing fetus. In addition to hearing loss, other anomalies which may occur include heart disorder, low birth weight, mental retardation, and vision loss. When hearing loss occurs, 50% have bilateral severe to profound loss. Hearing loss may be progressive.

Syphilis: Congenital syphilis may become apparent in the first 2 years of life, or between the ages of 8 to 20 years. Hearing loss is sensorineural and may be sudden, progressive, or fluctuating. Early onset hearing loss caused by syphilis may be reversible with early detection and prompt treatment.

5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.

Craniofacial abnormalities (e.g., cleft lip/palate, shortened neck, webbed neck, abnormal head circumference) may be indications of the presence of a hearing loss. Malformation of the ears may include atresia, low set ears, skin tags, and preauricular pits. These abnormalities may be indicative of a syndrome.

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.

There are many syndromes associated with hearing loss that include observable physical anomalies of the head, neck, and ears which frequently result in hearing loss (e.g., Down syndrome). Another example is Neurofibromatosis Type II (NF2), an inherited tumor syndrome. Some syndromes are not evident at birth.

9. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.

Type B, Hemophilus Influenza, carries the greatest incidence of hearing loss. Bacterial meningitis is the leading cause of acquired deafness in infants and childhood, ranging in incidence from 5-30%. Most occurrences result in severe, bilateral, symmetrical, sensorineural hearing loss; 30% have profound loss. The age at which meningitis occurs significantly affects rehabilitative needs; the younger the child, the greater the impact on speech and language acquisition.

10. Head trauma, especially basal skull/temporal bone fractures that requires hospitalization.

Head trauma (e.g., a skull fracture) may affect hearing due to potential damage to either the cochlea or middle ear, resulting in sensorineural or conductive hearing loss. Sensorineural loss may occur due to damage or obliteration of the temporal bone housing the inner ear. Conductive hearing loss occurs as a result of perforation of the tympanic membrane, bleeding, or disruption of the ossicular chain.

11. Chemotherapy.

"Chemotherapy" refers to a number of drugs which can be "ototoxic."

Ototoxicity is damage to the ear (oto-), specifically the cochlea or auditory nerve and sometimes the vestibular system, by a toxin. It is commonly medication-induced; ototoxic drugs include antibiotics such as the aminoglycoside gentamicin, loop diuretics such as furosemide, and platinum-based chemotherapy agents such as cisplatin. It can result in sensorineural hearing loss, dysequilibrium, or both. Either may be reversible and temporary, or irreversible and permanent.

Note: The 2007 JCIH Position Statement no longer includes in its list of risk indicators recurrent or persistent otitis media with effusion for at least 3 months. However, the JCIH recommends careful assessment of middle-ear status at all well-child visits, and children with persistent middle-ear effusion lasting 3 months or longer should be referred for otologic evaluation.

Middle ear infection is a frequently occurring illness in very young children, second only to the common cold. "Recurrent" is defined as three or more bouts of otitis media within a twelve-month period; "persistent" is defined as lasting three months or longer. PE tubes may have been inserted to address the otitis media with effusion (OME). Frequent episodes of OME may result in fluctuating conductive hearing loss, in turn influencing speech and language development. OME compounds hearing loss due to other conditions (e.g., sensorineural hearing loss) and should be monitored vigilantly.