PEP PROMOTING EHDI PRACTICES

# CHECKLIST
# RISK FACTORS

**Table 1**

***Risk Factors for Early Childhood Hearing Loss: Guidelines for Infants who Pass the Newborn Hearing Screen***

| | Risk Factor Classification | Recommended Diagnostic Follow-up | Monitoring Frequency |
|---|---|---|---|
| | Perinatal | | |
| 1 | Family history* of early, progressive, or delayed onset permanent childhood hearing loss | by 9 months | Based on etiology of family hearing loss and caregiver concern |
| 2 | Neonatal intensive care of more than 5 days | by 9 months | As per concerns of on-going surveillance of hearing skills and speech milestones |
| 3 | Hyperbilirubinemia with exchange transfusion regardless of length of stay | by 9 months | |
| 4 | Aminoglycoside administration for more than 5 days** | by 9 months | |
| 5 | Asphyxia or Hypoxic Ischemic Encephalopathy | by 9 months | |
| 6 | Extracorporeal membrane oxygenation (ECMO)* | No later than 3 months after occurrence | Every 12 months to school age or at shorter intervals based on concerns of parent or provider |
| 7 | In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis | by 9 months | As per concerns of on-going surveillance |
| | In utero infection with cytomegalovirus (CMV)* | No later than 3 months after occurrence | Every 12 months to age 3 or at shorter intervals based on parent/provider concerns |
| | Mother + Zika and infant with no laboratory evidence & no clinical findings | standard | As per AAP (2017) Periodicity schedule |
| | Mother + Zika and infant with laboratory evidence of Zika + clinical findings<br>Mother + Zika and infant with laboratory evidence of Zika - clinical findings | AABR by 1 month<br><br>AABR by 1 month | ABR by 4-6 months or VRA by 9 months<br><br>ABR by 4-6 months<br><br>Monitor as per AAP (2017) Periodicity schedule (Adebanjo et al., 2017) |
| 8 | Certain birth conditions or findings:<br>• Craniofacial malformations including microtia/atresia, ear dysplasia, oral facial clefting, white forelock, and microphthalmia<br>• Congenital microcephaly, congenital or acquired hydrocephalus<br>• Temporal bone abnormalities | by 9 months | As per concerns of on-going surveillance of hearing skills and speech milestones |
| 9 | Over 400 syndromes have been identified with atypical hearing thresholds***. For more information, visit the Hereditary Hearing Loss website (Van Camp & Smith, 2016) | by 9 months | According to natural history of syndrome or concerns |
| | Perinatal or Postnatal | | |
| 10 | Culture-positive infections associated with sensorineural hearing loss***, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis or encephalitis | No later than 3 months after occurrence | Every 12 months to school age or at shorter intervals based on concerns of parent or provider |
| 11 | Events associated with hearing loss:<br>• Significant head trauma especially basal skull/temporal bone fractures<br>• Chemotherapy | No later than 3 months after occurrence | According to findings and or continued concerns |
| 12 | Caregiver concern**** regarding hearing, speech, language, developmental delay and or developmental regression | Immediate referral | According to findings and or continued concerns |

*Note.* AAP = American Academy of Pediatrics; ABR = auditory brainstem response; AABR = automated auditory brainstem response.

* Infants at increased risk of delayed onset or progressive hearing los

**Infants with toxic levels or with a known genetic susceptibility remain at risk

***Syndromes (Van Camp & Smith, 2016)

****Parental/caregiver concern should always prompt further evaluation.

JCIH JOINT COMMITTEE on INFANT HEARING

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# PROMOTING EHDI PRACTICES

## Risk Factors for Early Childhood Hearing Loss

The JCIH 2019 position statement includes risk factors that are important to consider for ongoing monitoring of late onset or progressive hearing loss for those that pass the newborn hearing screening. Parents, medical providers and audiologists can benefit from understanding these risk factors as the prevalence of children confirmed as deaf or hard of hearing by school age doubles compared to the neonatal period.

When risk factors are present, comprehensive audiologic evaluation should occur. The schedule for ongoing re-evaluation is based on both the specific risk factors and the observations by the family of their child's auditory and speech/language development.

When a baby is readmitted, within the first month of life, the baby may need to be rescreened. Rescreening hearing should be completed any time there are conditions associated with elevated hearing levels. Automated ABR rescreening should be performed prior to discharge from that readmission even when the baby passed the initial newborn hearing screening. Risk factors are divided into predominantly perinatal and postnatal.

## Perinatal

History of family members being deaf or hard of hearing with onset in childhood. Monitoring continues to be based on both the etiology and the level of family concern. Diagnostic evaluation recommended by 9 months of age or earlier if parent or caregiver concern is expressed.

Infants who require care in the NICU or special care nursery for more than five days is used as an indicator of illness severity.

Hyperbilirubinemia, is impacted by factors including illness severity, birth weight, rate of rise of bilirubin, clinical findings, postnatal age of the infant, and gestational age. Close follow up is recommended for those requiring exchange transfusion regardless of length of stay in the NICU.

Aminoglycoside administration of more than five days (or less than five days if toxic blood levels are identified), or if there is a family history of a mitochondrial genetic mutation associated with sensitivity for sensorineural hearing loss.

Perinatal asphyxia, also termed hypoxic ischemic encephalopathy, is noted because of the illness severity and increase in permanently elevated hearing thresholds.

Extracorporeal membrane oxygenation (ECMO) is specifically noted because of the increased risk of delayed-onset hearing loss.

In-utero infections pose a risk and require follow up by 9 months of age. cCMV is a leading cause of congenital infection and a leading cause of non-genetic unilateral or bilateral early, progressive, and delayed onset sensorineural hearing loss. The recommendation for audiologic assessment for infants with cCMV is no later than 3 months of age. Those infants born to mothers with possible Zika virus exposure during pregnancy or with findings consistent with congenital Zika syndrome should receive a standard newborn hearing screen at birth or by one month of age using the automated ABR (not OAE).

All craniofacial conditions and physical conditions associated with hearing loss are included as risk factors.

More than 400 syndromes and genetic disorders associated with atypical hearing thresholds are now included as risk factors.

## Perinatal or Postnatal Risk Factors

- Perinatal and postnatal confirmed bacterial and/or viral meningitis or encephalitis.
- Predominantly post-natal events of chemotherapy, significant head trauma and particularly injury to the mastoid.
- Family concern regarding development, hearing, speech, or language should result in immediate referral.

This QR code will take you to the JCIH website

This QR code will take you to the EHDI Pals website

This QR code will take you to AAP EHDI resources.

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