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Pediatric Hearing Loss Guidelines and Consensus Statements – Where Do We Stand?

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Synopsis: A large number of guidelines and position statements have been published with the aim of improving outcomes for children with hearing loss. The purpose of this chapter is to review the current state of clinical practice guidelines as they relate to screening, diagnosis, and management of hearing loss in children. This summary is intended for the practicing otolaryngologist.

Key points:

- Infants should be screened for hearing loss by 1 month of age, diagnosed by 3 months, and enrolled in intervention services by 6 months (1-3-6 goal). For systems currently meeting the 1-3-6 goal, a 1-2-3 goal is recommended.
- Infants who pass the newborn hearing screening and possess or develop any of the 12 known risk factors for childhood hearing loss should be monitored regularly for indicators of congenital, delayed-onset, or progressive hearing loss.

- Once a hearing loss is diagnosed, the otolaryngologist should partner with the medical home provider and audiologist to facilitate coordinated and comprehensive care, including the referral to early intervention and fitting of amplification.

Abstract

This chapter provides a review of the current state of clinical practice guidelines regarding the screening, diagnosis, and management of children with hearing loss. An overview of the 2019 guidelines for Early Hearing Detection and Intervention (EHDI) systems is outlined. Current guidelines regarding cytomegalovirus, Zika virus, genetic screening, chronic middle ear conditions, and monitoring for ototoxicity are also discussed.

Evidence-based practice or medicine (EBP) involves the integration of scientific evidence, clinical expertise, and patient/family values to provide optimal patient care.¹ Given the current rate of technological development and scientific discovery, clinical practice guidelines (CPGs) and consensus or position statements are essential tools for healthcare professionals providing EBP.² A CPG is functionally the same as a consensus or position statement and is the preferred term for this chapter. CPGs describe how members of a given profession should adhere to a standard of practice, ensuring that patients receive consistent, high-quality care regardless of setting or provider.² The purpose of this chapter is to review the current state of CPGs as they relate to children with hearing loss.

Screening for Pediatric Hearing Loss

Many CPGs have been developed to identify children who have or are at risk for developing hearing loss. This is in response to overwhelming evidence that children with hearing loss who receive early identification and intervention achieve better academic, cognitive, communication, language, and social-emotional outcomes than those who do not.^{3,4} Given that hearing loss is one of the most prevalent developmental disabilities at birth,⁵ the first entry point for identification and intervention of childhood hearing loss is through newborn hearing screening (NBHS).

Newborn Screening via Early Hearing Detection and Intervention (EHDI) Systems

The Joint Committee on Infant Hearing (JCIH) publishes CPGs for the development and implementation of universal NBHS programs, which are now designated EHDI systems. The use of the term EHDI is preferred, as it underscores the importance of NBHS being tied to an entire system that includes the medical home, a tracking and surveillance process, a method for

monitoring system efficacy, audiologic diagnosis, and appropriate intervention services in partnership with the family.⁶

Box 12.1. Federal funding for EHDI systems currently comes from the Department of Health and Human Services.³⁸ At the state and territory level, funding for EHDI systems varies according to a variety of factors including Medicaid, Title V funding, general revenues, and procured grants. For these reasons, public and professional advocacy for EHDI at the federal, state, and territory levels is essential for the continuation and improvement of EHDI systems. As of March 2021, all 50 states and the District of Columbia have EHDI systems, with 43 states containing legislation describing minimum standards for their EHDI system.

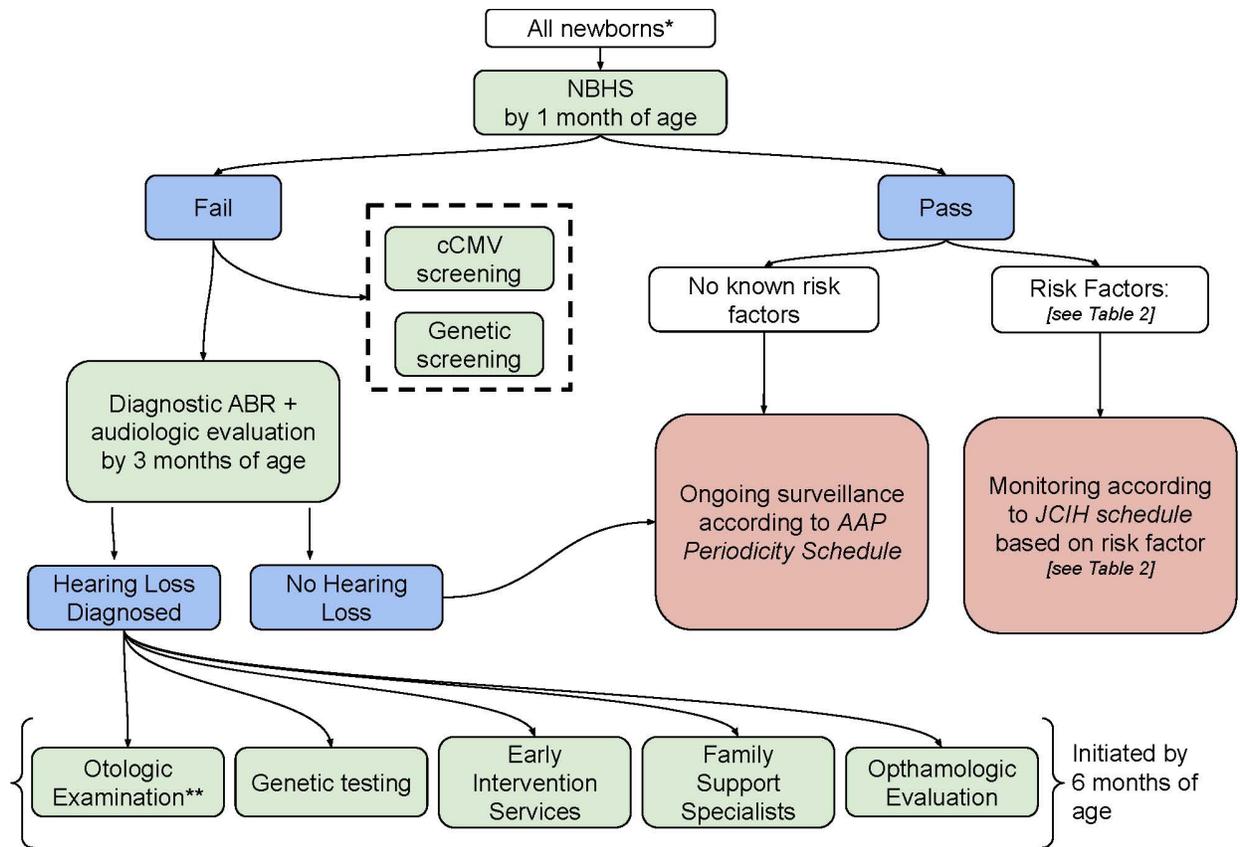
Box 12.2. Components of successful EHDI systems.

- All infants undergo NBHS by 1 month of age, prior to discharge from the birthing hospital
 - Infants who fail NBHS in either ear are scheduled for an appointment for outpatient rescreening or audiologic evaluation at the time of hospital discharge
 - Alternate contact information for a family is obtained
 - Regardless of NBHS result, the communication development of all infants and children is monitored by professionals with appropriate training and within the medical home according to the American Academy of Pediatrics Periodicity Schedule¹²
- All infants who fail the NBHS have an audiologic diagnostic assessment by 3 months of age

- For infants who are diagnosed with hearing loss, the following should occur concurrently or immediately after diagnosis:
 - Otologic evaluation
 - Referral to an appropriate interdisciplinary early intervention system through a simplified, coordinated point of entry
 - This referral is made by a professional knowledgeable about the needs and requirements of children with hearing loss
- All infants who are diagnosed with hearing loss begin early intervention services as soon as possible after audiologic diagnosis, and no later than 6 months of age
 - The early intervention approach documented in an Individualized Family Service Plan honors the family's preferences and goals for their child
 - The approach builds on the strengths, informed choices, language traditions, and cultural beliefs of the family
 - The child and family have *immediate* access to hearing aid technology through their audiologist
 - The child and family have access to cochlear implants, hearing assistive technologies, and visual alerting and informational devices
 - The family has access to:
 - Information about all resources and programs for intervention
 - Support and counseling about the child's educational and communication needs
- Individualized, family-centered care
- Informed and shared decision-making

- Evidence-based practice
- Guarantee of family rights and privacy
- Family consent following state and federal guidelines
- Information systems that facilitate exchange of electronic health information with clinical electronic health records and population-based information systems

Figure 1. Preferred progress of early hearing detection and intervention (EHDI) activities outlined by JCIH.⁷ Green shading represents stages of assessment, with possible results shown in blue. The dashed box represents assessments not yet used in widespread clinical practice. Red shading represents patient management. *Note: NBHS = newborn hearing screening. cCMV = congenital cytomegalovirus. ABR = auditory brainstem response. AAP = American Academy of Pediatrics. JCIH = Joint Committee on Infant Hearing.*



*JCIH recognizes that this timeline may not be feasible for preterm infants who require prolonged hospitalization in the Neonatal Intensive Care Unit.
 **Sometimes the otologic examination occurs in conjunction with the diagnostic ABR + audiologic evaluation

The overall goal of EHDI is to identify infants with hearing loss as early as possible to optimize overall development through early and consistent access to language and intervention services. Components of successful EHDI systems are shown in Box 12.2 and a flowchart detailing the preferred progression of EHDI activities is provided in Figure 1. Generally, EHDI recommends that infants are screened for hearing loss by 1 month of age, receive confirmation of hearing status by 3 months of age, and begin intervention services (if indicated) by 6 months of age (1-3-6 goals). JCIH recently suggested that EHDI systems currently meeting 1-3-6 goals consider establishing 1-2-3 goals (audiologic screening by 1 month, audiologic diagnosis by 2 months, intervention initiated by 3 months). The rationale for establishing a 1-2-3 timeline includes providing infants even earlier access to language and reducing the likelihood of administering

sedation to complete audiologic testing in infancy. The 1-2-3 timeline may not be feasible for very preterm infants who could remain in the neonatal intensive care unit through the third month of life. For those infants, JCIH recommends a diagnostic audiologic evaluation prior to discharge and direct referral to audiologic follow-up and early intervention services as appropriate.

The preferred method for conducting NBHS in the well-baby nursery is either automated auditory brainstem response (AABR) or otoacoustic emissions (OAEs).⁷ For infants in the well-baby nursery who fail the initial NBHS, the second screening (e.g., rescreen) should be conducted on both ears using either method; however, it is preferred that infants who fail the initial screening conducted by AABR are rescreened using AABR.⁷ JCIH recommends initial and rescreening protocols in neonatal intensive care units be conducted solely with AABR in the hospital or by a pediatric audiologist in an outpatient setting.⁷

Table 1. Reasons children with hearing loss may not be identified by NBHS^{7,10}

EHDI Outcome	Potential reason for undiagnosed childhood hearing loss
Child passed NBHS	<p>No hearing loss present at time of NBHS (e.g., delayed onset of hearing loss)</p> <p>Hearing loss present but not detected by NBHS due to limitations of sensitivity and specificity of the NBHS protocol</p> <p>Hearing loss present but not detected by NBHS (e.g., mild hearing loss, auditory neuropathy spectrum disorder)</p> <p>Progressive or fluctuating hearing loss</p> <p>Screening equipment or personnel error</p>

Child missed NBHS	Parent refusal Not birthed in a hospital Hospital discharge or transfer prior to completion of NBHS Rescreen never completed after initial 'fail' result
Child lost to follow-up or documentation	NBHS completed in another state Miscommunication or lack of communication regarding NBHS result to family, medical and healthcare providers, hospitals, and/or state EHDI system Lack of or poor documentation of NBHS result, diagnostic assessment, or referral to early intervention services Parent or physician misunderstanding of or lack of commitment to follow-up when indicated (e.g., 'fail' result or presence of risk factor(s) listed in Table 2) Lack of skilled pediatric audiology services near the family's home Family challenges with transportation to follow-up appointments or services Infant does not have established care with a primary care provider or medical home Parent does not consent to diagnostic evaluation or early intervention services Unable to make contact with family

JCIH recognizes that NBHS protocols will not identify all children with hearing loss given the reasons listed in Table 1. Because not all childhood hearing loss is present at birth, infants who pass the NBHS and possess or develop any of the 12 known risk factors for early childhood hearing loss listed in Table 2 should be monitored as indicated.

Continued Screening for Childhood Hearing Loss

Table 2. Risk factors for early childhood hearing loss: guidelines for infants who pass NBHS. Adapted from “Year 2019 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs,” by *Journal of Early Hearing Detection and Intervention*, 4(2), p. 19 (<https://digitalcommons.usu.edu/jehdi/vol4/iss2/1>). CC BY-NC.

	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 >5 days	by 9 months	As per concerns of ongoing surveillance of communication development
3	Hyperbilirubinemia with exchange transfusion regardless of length of stay	by 9 months	
4	Aminoglycoside administration >5 days or <5 days if toxic blood levels are identified or a genetic susceptibility is known	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 parent/provider concerns
7	In utero infections, such as herpes, rubella, syphilis, and toxoplasmosis	by 9 months	As per concerns of ongoing 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 positive for Zika virus:		

	<p>infant with <i>no</i> laboratory evidence and <i>no</i> clinical findings</p> <p>infant with laboratory evidence of Zika <i>and</i> clinical findings</p> <p>infant with laboratory evidence of Zika and <i>no</i> clinical findings</p>	<p>Standard</p> <p>AABR by 1 month</p> <p>AABR by 1 month</p>	<p>As per AAP Periodicity Schedule¹²</p> <p>ABR by 4-6 months or VRA by 9 months</p> <p>ABR by 4-6 months</p> <p>Monitor as per AAP Periodicity Schedule¹²</p>
8	<p>Certain birth conditions or findings:</p> <ul style="list-style-type: none"> • Craniofacial malformations including microtia/atresia, ear dysplasia, oral facial clefting, white forelock, and microphthalmia • Congenital microcephaly, congenital or acquired hydrocephalus • Temporal bone abnormalities 	By 9 months	As per concerns of ongoing surveillance of communication development
9	<p>Over 400 syndromes have been identified with atypical hearing thresholds. Visit https://hereditaryhearingloss.org/ for more information</p>	By 9 months	According to natural history of syndrome or concerns
Perinatal or Postnatal			
10	<p>Culture-positive infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis or encephalitis</p>	No later than 3 months after occurrence	Every 12 months to school age or at shorter intervals based on parent/provider concerns
11	<p>Events associated with hearing loss:</p> <ul style="list-style-type: none"> • Significant head trauma, especially basal skull/temporal bone fractures • Chemotherapy 	No later than 3 months after occurrence	According to findings and/or continued concerns
12	<p>Caregiver concern regarding hearing, speech, language, developmental delay,</p>	Immediate referral	According to findings and/or continued

	and/or developmental regression		concern
<i>Note.</i> AAP = American Academy of Pediatrics; ABR = auditory brainstem response; AABR = automated auditory brainstem response; VRA = visual reinforcement audiometry. *Infants at increased risk of delayed onset or progressive hearing loss			

The prevalence of hearing loss increases by the time children enter school.^{8,9} As of March 2021, there is no CPG or federal mandate regarding universal hearing screening after the newborn period.¹⁰ Some states mandate school-based screenings while others do not. Therefore, all infants and children, regardless of presence or absence of risk factors, should receive ongoing surveillance of communication development.^{7,11,12} It is the otolaryngologist's responsibility to evaluate all infants and children on their caseload for the risk factors listed in Table 2. For specific recommendations regarding the otolaryngologic workup of hearing loss in children, please reference Chapter 4.

Special Considerations

Cytomegalovirus. Cytomegalovirus is the leading cause of congenital infection, occurring in 0.2-2% of live births worldwide. The 2019 JCIH guidelines acknowledge that congenital cytomegalovirus (cCMV) plays a larger role in childhood hearing loss than previously thought, with 10-15% of infants with cCMV developing sensorineural hearing loss.⁷ A 2015 International cCMV Recommendations Group suggested that universal screening for cCMV in all newborns should be *considered*, but fell short of providing a recommendation for universal screening, citing the need for prospective studies and cost-effectiveness studies prior to a recommendation.¹³ In 2019, the Newborn Hearing Screening Working Group of the National Coordinating Center for the Regional Genetics Networks recommended that universal cCMV

screening, along with limited genetic testing, be integrated into the current NBHS program.¹⁴ Although the development of an appropriate and relatively inexpensive screening has been a priority research topic for nearly two decades,¹⁵ nationwide screening for cCMV does not exist as of March 2021. In line with the International Pediatric Otolaryngology Group recommendations,¹⁶ several states have implemented targeted cCMV screening - testing children who fail NBHS.¹⁷ Early findings with targeted cCMV screening show that, although sensitivity was not as high as universal cCMV screening, over two-thirds of symptomatic cCMV cases could be identified with this approach.¹⁸

Zika virus. The Zika virus is a mosquito-borne infection that can be passed from a pregnant woman to a developing fetus. Congenital Zika syndrome (CZS) causes birth defects such as microcephaly, decreased brain tissue, and vision and hearing impairment. Approximately 7% of infants with CZS have sensorineural hearing loss.¹⁹ Although the Zika virus is not an epidemic in the United States, CZS is still a concern in Middle and South American countries where the Zika virus remains a threat. For the first time, JCIH has included the Zika virus in its list of risk factors for hearing loss. JCIH and CDC recommendations for testing the hearing of infants with Zika virus are shown in Box 12.3. While the primary concern for Zika virus infections is CZS, children infected with Zika beyond the neonatal period are at risk for Guillain-Barre syndrome and transient sensorineural hearing loss.

Box 12.3. JCIH and CDC share recommendations for audiologic testing of infants with Zika virus.

- Infants with laboratory-confirmed Zika - regardless of symptoms - should receive an automated ABR by 1 month even if they passed a newborn hearing screening with OAE.

- Infants with laboratory-confirmed Zika with clinical symptoms should also receive a diagnostic ABR by 4-6 months or testing using Visual Response Audiometry by 9 months of age.
- Infants with laboratory-confirmed Zika with no clinical symptoms should receive a diagnostic ABR by 4-6 months.
- From thereafter, the AAP 2017 schedule of testing for infants with risk factors¹² should be followed for all infants with Zika virus.

Diagnosing Pediatric Hearing Loss

Audiologic diagnosis of hearing loss should be completed no later than 2-3 months of age by an audiologist with the specific skills, knowledge, and access to equipment required for infant and early childhood diagnostic evaluations. Pediatric audiologists can be found through a roster maintained by the American Board of Audiology²⁰ or through the EHDI's Pediatric Audiology Links to Service.²¹

Table 3. Key aspects of audiologic assessment for infants and young children.

Auditory brainstem response	Gold standard test for infants and children who cannot complete behavioral audiologic assessment. Provides information necessary to diagnose the type, degree, and configuration of hearing loss.
Tympanometry or wideband reflectance	Measures of middle ear function.
Acoustic reflexes	Test of middle ear function and integrity of auditory brainstem pathways.
Otoacoustic emissions	Assessment of integrity of the outer hair cells of the cochlea. Critical for the differential diagnosis of auditory neuropathy

	spectrum disorder and sensorineural hearing loss.
Behavioral assessment of hearing	Gold standard test for infants and children who can provide reliable and valid responses.

Testing included in an audiologic diagnostic appointment is listed in Table 3. It is important to note that the auditory brainstem response is not a test of hearing, but a measure of an electrophysiologic response to auditory stimuli. To confirm a child’s hearing (perception), a behavioral evaluation should be conducted as soon as the child is developmentally capable of providing reliable responses (approximately 6 months of age).

The Role of the Medical Home

Because the rate of childhood hearing loss increases from 1.2/1000 in newborns to 3/1000 in early school age,²¹ all children, regardless of NBHS result, should receive surveillance of speech and language milestones and auditory development in the medical home beginning at two months of age.¹² Ongoing screening allows for children with delayed-onset or progressive hearing loss or those who might have been missed by NBHS (e.g., mild hearing loss), to receive timely intervention. Once diagnosed, the medical home provider should refer the child with hearing loss to an otolaryngologist, clinical geneticist, genetic counselor, audiologist, speech and language specialist, early hearing intervention provider, and family support specialist. This team of professionals should collaborate with the family in informed decision making for their child.²² This care is ideally found at a multidisciplinary care center; however, in rural areas, families might need to seek services from individual providers. Finally, because the otolaryngologist’s evaluation includes a comprehensive history to identify risk factors for and findings associated with congenital or delayed-onset childhood hearing loss (see Ch 5), the otolaryngologist should

partner with the medical home provider and audiologist to facilitate coordinated and comprehensive care.

Special Considerations

Genetic screening. It is estimated that up to 60% of congenital and early-onset hearing loss is caused by genetic factors.^{23,24} In conjunction with the American College of Medical Genetics and Genomics, JCIH recommends that all infants and children with confirmed hearing loss be offered a genetics evaluation and counseling.^{7,25} Results of genetic testing can be valuable for families of children with hearing loss, providing answers to two common questions: (1) “what caused my child’s hearing loss?”, and (2) “will their hearing loss change over time?” In addition to information about the etiology and prognosis for progression, genetic evaluation can uncover disorders associated with the hearing loss (e.g., renal, vision, cardiac) and explain the likelihood of recurrence of hearing loss in future offspring.

Intervention and Management of Pediatric Hearing Loss

Together with the AAP and CDC, JCIH recommends that, at the time of hearing loss diagnosis, children should be referred for medical and otologic evaluations to the state EHDI program and to the state Part C early intervention program.^{11,12,26,27} The purposes of the medical and otologic evaluations are to identify any conditions related to the hearing loss, provide medical/surgical recommendations and treatments, refer the child for ancillary services, and engage the family in informed decision making for their child.²²

Box 12.4. For families who have chosen listening and spoken language as the communication goal for their child, medical clearance to obtain hearing aids is included as part of these medical recommendations. Specifically, the Food and Drug Administration requires a written statement

from a licensed physician declaring that a medical evaluation has determined the patient to be a candidate for hearing aids.³⁹ As of 2020, federal law allows a fully informed adult to sign a waiver declining the medical evaluation - this option is not permitted for children. Importantly, medical clearance for hearing aid fitting should not be delayed until other medical examinations are completed or a diagnosis of etiology made.

Once medical clearance is obtained, children with hearing loss should be fitted with hearing aids by an audiologist with expertise, skills, and knowledge in pediatric audiology.²⁸ JCIH recommends fitting of amplification as soon as possible following the diagnosis of hearing loss. Children with sensorineural hearing loss will be fitted with traditional air conduction hearing aids, which might be contraindicated for some children (e.g., draining ears, microtia). In these cases, it is important that infants and young children with conductive hearing loss be fitted with bone conduction hearing devices on a softband until they are considered implant candidates at age five years. Once fitted, the child will receive regular surveillance of hearing status and ongoing validation of amplification fitting. Note that a child's referral to early intervention should not be deferred until hearing aid fitting; referral should occur within 48 hours of hearing loss diagnosis.²⁹

For children who fail to make expected progress with appropriately fitted amplification, cochlear implantation evaluation should be offered, especially if parental goals for their child include improved hearing and understanding and the use of spoken language. The cochlear implant evaluation process should be conducted by a team consisting of audiologists, otolaryngologists, medical home providers, early intervention specialists, and the family. Cochlear implantation is currently approved by the U.S. Food and Drug Administration for

children 9 months of age and older, and clinical trials to further reduce age are underway. Age at implant and audiologic candidacy for cochlear implantation is expanding (see Ch 10). The American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) now considers cochlear implantation (unilateral and bilateral) an appropriate treatment for children, including infants between 6 and 12 months of age with severe to profound hearing loss having failed a trial with appropriately fitted hearing aids. Children aged 12 months and older with more residual hearing (pure tone average between 65 and 85 dB) might be candidates if aided auditory skill development and speech and language progress indicate persistent or widening gap in age-appropriate skills.³⁰

Special Considerations

Box 12.5. Risk factors for developmental difficulties associated with childhood OME as specified by the AAO-HNS' CPG for OME.

1. Permanent hearing loss independent of OME
2. Suspected or confirmed speech and language delay or disorder
3. Autism spectrum disorder
4. Syndromes or craniofacial disorders that include cognitive, speech, or language delays
5. Blindness or uncorrectable visual impairment
6. Cleft palate
7. Developmental delay

Chronic middle ear conditions. Otitis media with effusion (OME) typically resolves within 3 months, but 30-40% of children have repeated episodes of OME, with 5-10% of

episodes lasting more than one year.^{31,32} This middle ear fluid can be associated with conductive hearing loss, placing some children at risk for developmental difficulties. The AAO-HNS CPG for OME³³ identifies seven risk factors for developmental difficulties in children with OME (see Box 12.5).

Recommendations for management of OME in children without permanent hearing loss vary widely. Below are recent guidelines pertaining to OME management.

- The AAO-HNS recommends that a child with persistent OME and no known risk factors for permanent hearing loss be managed with watchful waiting for three months from the date of effusion onset (if known) or from date of diagnosis if onset is unknown. If OME persists for ≥ 3 months or if OME of any duration is present in an at-risk child, a full audiologic evaluation should be recommended. AAO-HNS also recommends that children with chronic OME be evaluated in 3-6 month intervals until OME is resolved, significant hearing loss is identified, or structural abnormalities are suspected. Note that the definition of “significant hearing loss” was intentionally not defined in this guideline.
- JCIH takes a more conservative approach by recommending that a child with OME persisting for ≥ 6 months should be referred for early intervention services to ensure adequate auditory access.²⁹
- Perhaps the most conservative approach to chronic OME is taken by the International Pediatric Otolaryngology Group, which recommends that children with persistent OME within the first few months of life receive myringotomy and tubes with a follow/up ABR if OME is still present at 6 months of age.¹⁶

Guidelines for management of OME were developed for children with normal hearing sensitivity. More aggressive management of OME in children with permanent hearing loss

should be considered. Quickly resolving OME in children with permanent hearing loss is essential for the accurate initial diagnosis of degree of loss and to optimize benefits from amplification. Furthermore, because some infants and children do not tolerate hearing devices when OME is present, speedy resolution of OME can aid in consistent use of amplification.

Box 12.6. Audiologic surveillance should begin prior to the end of oncologic treatment. When treatment begins at <6 years of age, audiologic surveillance should continue, at minimum, annually. For children who begin treatment at 6-12 years of age, audiologic surveillance should occur every other year. When treatment begins at >12 years of age, audiologic surveillance should occur every 5 years.³⁶ Beyond these recommendations, there is no consensus regarding how long ototoxicity-induced hearing loss monitoring should continue past treatment.

Ototoxicity-induced hearing loss. Roughly half of pediatric survivors of cancer will develop hearing loss as a result of ototoxicity from treatment with platinum-base compounds and/or radiation of the head or brain.^{34,35} Because ototoxicity-induced hearing loss often affects the high frequencies and presents with tinnitus, audiologic surveillance is recommended for pediatric survivors of cancer treated with head or brain radiotherapy or with cisplatin, with or without high-dose carboplatin (see Box 12.6).³⁶ This recommendation holds true regardless of co-treatment with otoprotective agents.^{36,37} Ideally, a baseline audiologic evaluation will establish hearing thresholds prior to the initiation of oncologic treatment. Otolaryngologists should communicate directly with oncology regarding treatment and with audiology regarding hearing surveillance. Refer to Chapter 1 for additional information about ototoxicity and otoprotection.

Summary

The implementation of CPGs that guide screening, diagnosis, and management of hearing loss in children has resulted in improved outcomes for children with hearing loss. However, not all infants and children with hearing loss will receive timely diagnosis and intervention. As a result, it is critical that the otolaryngologist evaluate all infants and children for risk factors for hearing loss, regardless of NBHS result, and make prompt referrals when indicated. Although not yet covered in CPGs, otolaryngologists should also consider the impact of cerumen, noise exposure, and potentially COVID-19 infection on children's hearing status.

Clinical Care Points

1. *Newborn Hearing Screening Testing Method.*

For infants in the well-baby nursery who fail the initial NBHS, it is preferred that the second screening be conducted on both ears using AABR. Neonatal intensive care unit initial and rescreening protocols should be conducted solely with AABR.

2. *Risk Factor Monitoring*

Risk factor assessment is important when evaluating children who passed NBHS. Those at risk for postnatal onset of HL (Table 2) should be referred for audiologic evaluation and monitoring. Of special consideration are children with positive neonatal viral cultures or microcephaly and other findings concerning cCMV or Zika.

3. *Otitis Media Management*

Guidelines for management of otitis media were developed for children with normal hearing sensitivity who are not at risk for language delay. Children with permanent

hearing loss could benefit from timelier placement of myringotomy tubes than indicated in these guidelines.

4. *Cochlear Implantation Evaluation*

Referral of infants with bilateral severe profound hearing loss, and of young children with moderate to severe loss not progressing despite amplification would benefit from candidacy evaluation by a pediatric cochlear implant program.

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