

International Pediatric ORL Group (IPOG)

Hearing Loss in the Pediatric Patient Consensus Recommendations

Consensus Objectives

To make recommendations on the routine work-up for pediatric patients with hearing loss

Target Population

All infants with hearing loss identified by newborn hearing screening and older children with concerns for hearing loss

Intended Users

These consensus recommendations are targeted for:

1. Primary care providers and otolaryngologists who take care of children with hearing loss.

Recommendations and Justification

The recommendations are outlined in the following sections

- **Section 1:** Diagnostic evaluation considerations
- **Section 2:** Newborn screening and initial workup of the patient with hearing loss
- **Section 3:** Workup of child with auditory neuropathy spectrum disorder (ANSO)
- **Section 4:** Workup of child with sensorineural hearing loss
- **Section 5:** Workup of child with conductive or mixed hearing loss

Disclaimer

This report has been prepared by the members of the International Pediatric ORL Group (IPOG). Consensus recommendations are based on the collective opinion of the members of the group. Any person seeking to apply or consult the report is expected to use independent medical judgment in the context of individual patient and institutional circumstances.

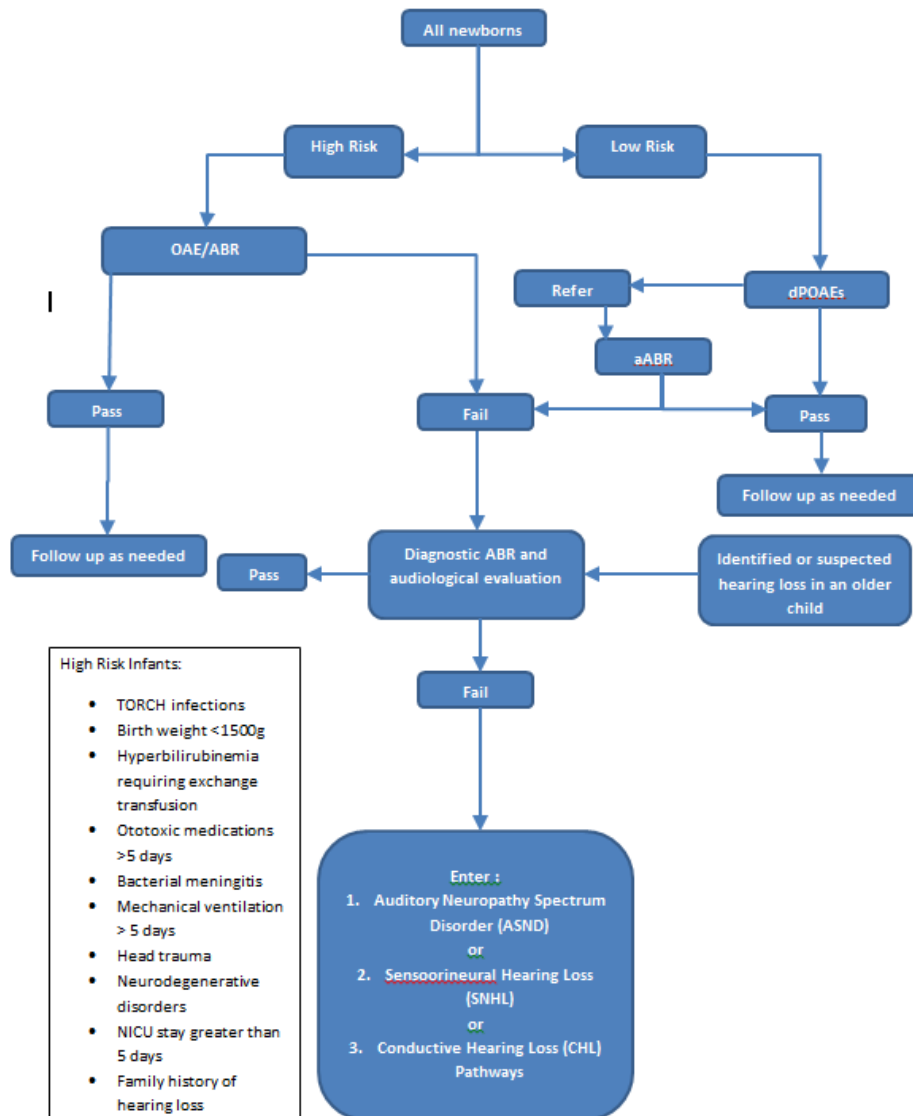
Section 1: Diagnostic evaluation considerations

The members of the IPOG identified six frequently debated considerations in the management of pediatric hearing loss. Variation in practice among the current group members remains, and the purpose of this section is to provide a list of reasonable options based on expert opinion.

Question	
1. Which children should be offered genetic testing?	<ul style="list-style-type: none">• After an audiogram, comprehensive genetic testing has the highest diagnostic rate of any single test for bilateral sensorineural hearing loss.¹• Children with unilateral hearing loss should not be offered genetic testing.• Single gene testing is of low diagnostic yield and should not be offered as part of an initial workup.• Directed genetic testing may be considered in consultation with a geneticist if comprehensive genetic testing is negative but suspicion for a genetic cause still exists.• Comprehensive genetic testing should be offered to children with ANSD.
2. Should temporal bone imaging be performed?	<ul style="list-style-type: none">• Temporal bone imaging is of low diagnostic yield in all children.• Children who are cochlear implant candidates with profound hearing loss may benefit from CT or MRI to assess for cochlear dysplasias and cochlear nerve aplasia.• Temporal bone imaging is optional in unilateral hearing loss and in children with ANSD.
3. Should ophthalmology consult routinely be ordered?	<ul style="list-style-type: none">• Given the 2-3 fold increased risk of ocular abnormalities in children with nonsyndromic SNHL, ophthalmology evaluation is warranted.
4. Should cardiology consult be routinely requested?	<ul style="list-style-type: none">• Genetic testing may identify children who should have a cardiac workup.• Cardiology consult should not be ordered at the onset of diagnosis of hearing loss.
5. Should amplification be offered for single sided hearing loss?	<ul style="list-style-type: none">• Children with single-sided hearing loss are at a higher risk for delays in speech and language development and decreased academic performance relative to normal hearing peers.• The decision to amplify unilateral hearing loss is complex and involves factors such as the severity of the hearing loss and socioeconomic factors such as financial resources.• At minimum, a discussion regarding the benefits of amplification should be undertaken.
6. Should testing and treatment for CMV be considered?	<ul style="list-style-type: none">• Congenital CMV causes up to ¼ of all congenital sensorineural hearing loss.²• CMV saliva/urine PCR is highly specific in the first three weeks of life.• Blood spot CMV testing can be used as an adjunct diagnostic test after the first three weeks of life.• Treatment for CMV associated hearing loss currently only indicated for symptomatic CMV disease. Treatment with valganciclovir may improve hearing outcomes in children with asymptomatic congenital CMV infection but not indicated at this time.³

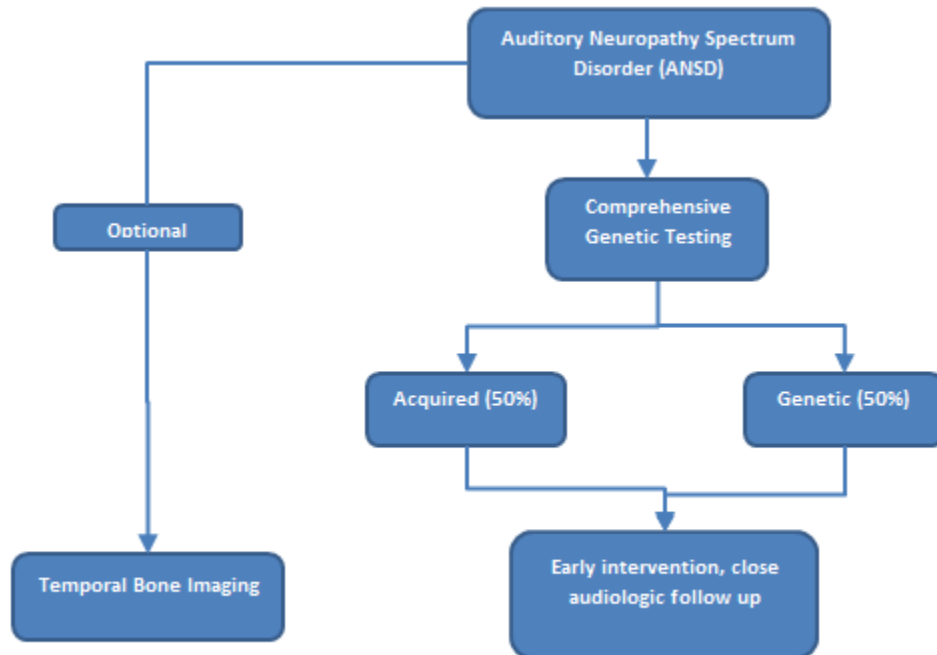
Section 2: Newborn screening and initial workup of the patient with hearing loss

The screening and initial workup algorithm is designed to guide the evaluation of all newborns with suspected hearing loss based on initial screening methodologies. The algorithm also applies to children identified later in childhood. Screening protocols recommended herein are generally derived from consensus recommendations such as those issued by the Joint Committee on Infant Hearing.⁴ Protocols may vary slightly between institutions. In general, infants meeting criteria for increased risk for congenital hearing loss should be screened with both OAEs and an automated ABR. Infants not within the high risk category can be screened with a two stage procedure that utilizes OAEs alone as the initial screening test, with an automated ABR for absent or subthreshold OAE responses. A child who fails screening protocols should undergo a confirmatory ABR.



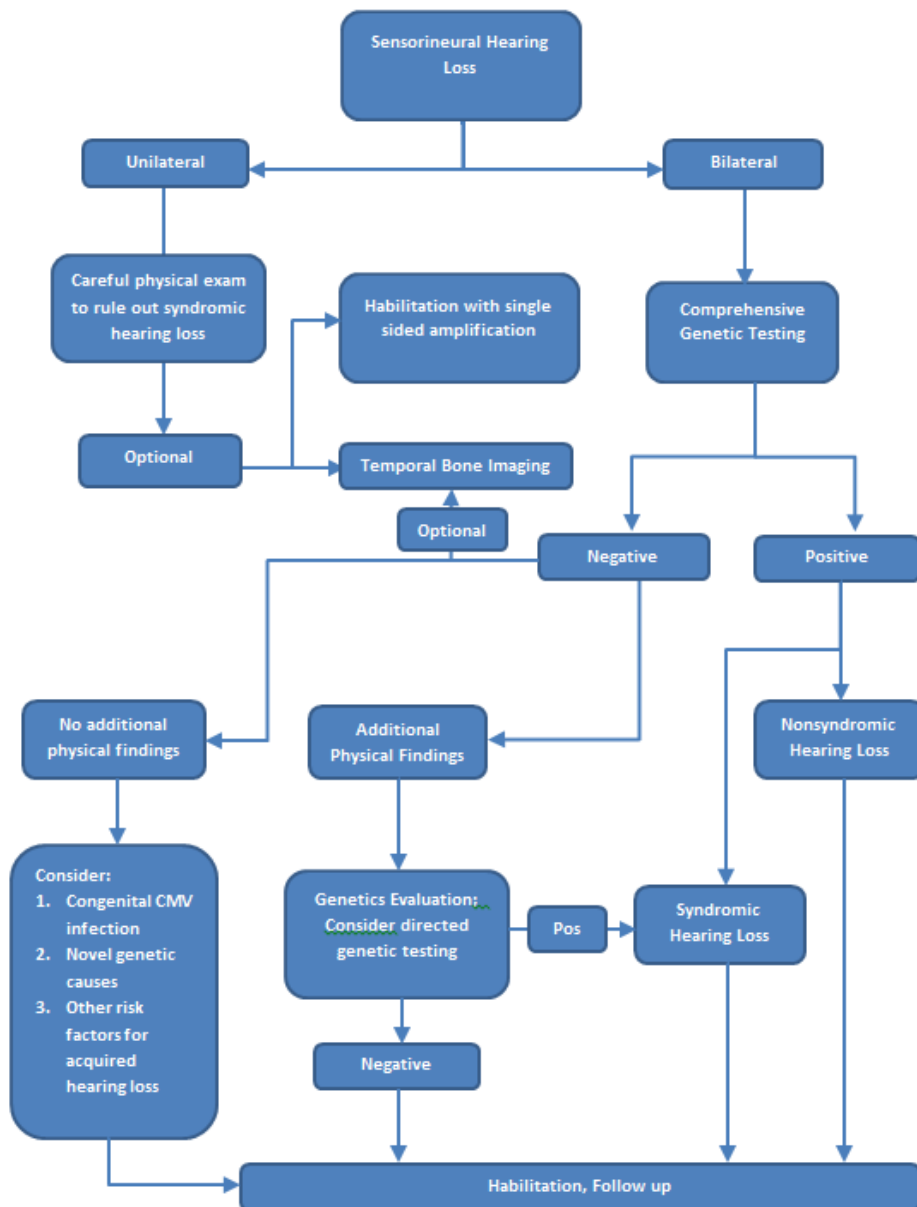
Section 3: Workup of child with auditory neuropathy spectrum disorder (ANSO)

The diagnosis of auditory neuropathy spectrum disorder (ANSO) is given when otoacoustic emissions and/or cochlear microphonics are present in the setting of absent or abnormal ABR. Given that 50% of ANSO has a genetic basis, comprehensive genetic testing should be offered to children in whom ANSO is identified. Identification of a specific genetic cause may determine candidacy for habilitation options, including cochlear implantation. Temporal bone imaging is optional in the setting of ANSO and may reveal abnormalities in up to 2/3 of patients.⁵



Section 4: Workup of child with sensorineural hearing loss

Genetic testing has a limited role in the workup of children with a unilateral sensorineural hearing loss unless syndromic hearing loss (i.e. Branchio-oto-renal syndrome, Waardenburg syndrome) is suspected based on physical exam and/or family history. Temporal bone imaging should be optional and discussed with parents at the time of diagnosis. Comprehensive genetic testing should guide subsequent workup in children with bilateral sensorineural hearing loss. The diagnostic rate is approximately 40-65%.¹ The physical examination and presence of other risk factors will dictate additional workup needed in the child with negative genetic testing. Asymptomatic congenital CMV is a significant contributor to congenital hearing loss and may present as mild to profound, unilateral or bilateral and stable or progressive. Vestibular dysfunction is also possible. Testing for congenital CMV is not currently standardized but should be considered in children with identified hearing loss. Treatment is controversial but has been shown to have some benefit.



Section 5: Workup of child with conductive or mixed hearing loss

The management of an effusion in infants is dependent on the age at which it is identified. In a child who is older than 3 months, a rapid course of action is indicated to ensure that any appropriate rehabilitation occurs prior to 6 months. For a child under the age of 3 months with purely conductive hearing loss, the effusion may be observed for a period of 3 months for spontaneous resolution. If the hearing loss is mixed in nature, then a shorter observation period of 4-6 weeks is appropriate to ensure that the sensorineural component is appropriately worked up and diagnosed prior to the age of 6 months.



References

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4. American Academy of Pediatrics, Joint Committee on Infant Hearing. Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics.* 2007;120(4):898–921
5. Roche JP, Huang BY, Castillo M, Bassim MK, Adunka OF, Buchman CA. Imaging characteristics of children with auditory neuropathy spectrum disorder. *Otol Neurotol.* 2010 Jul;31(5):760-8