

**Pediatric Resource Guide
to Infant and Childhood
Hearing Loss**
3rd Edition



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Foreword

Foreword

Welcome to the Third Edition of the *Pediatric Resource Guide to Infant and Childhood Hearing Loss*. The Guide has become an integral part of the overall program of services provided by the Center for Early Intervention on Deafness (CEID) and we are honored to publish this latest resource for your benefit and the many children you serve.

As CEID advances toward its fourth decade of service, we continue to strive to maximize the communication potential of young children (0 to 5 years old) who are deaf, hard of hearing, or have severe speech and language delays by providing exemplary early educational services. We create diverse, inclusive, and educationally rich environments in order to empower our students and their families with the academic and social tools needed to reach their full potential.

We cannot, and do not, achieve this high calling alone. It takes an entire caring community to actualize the potential of a child who is deaf or hard of hearing. We are deeply grateful for the families, community supporters, professionals, donors, and other advocates for our work; without their steadfast support and encouragement our tasks would be unachievable.

Although the essential audience for this Guide is the Primary Pediatric Provider, we at CEID know that the Guide also has value for:

- Educators, who seek better understanding about children entrusted to their care who are deaf or hard of hearing
- Other health professionals, who can incorporate the knowledge contained in this Guide into a more holistic view of their patients
- Community leaders and public policy officials, who may be called upon to initiate or enforce relevant laws and regulations pertaining to early detection and intervention
- Parents and other family members, who seek keener comprehension, and in-depth information, and
- Advocates for stronger awareness and public policy reforms for people who are deaf or hard of hearing, advocates whose efforts are exemplary and deserving of our approbation.

I am grateful for the exceptional work of the co-authors, family contributors, and reviewers of the manuscript for their contributions to the successful publication of the Guide. I appreciate their extraordinary commitment to raise awareness with the very real, and attainable, intention of changing lives.

CEID thrives due to the generosity of others. The Guide would not be possible without the support of The Claire Gianinni Fund, the Med-EL Corporation, the Oticon Medical Corporation, the Red Oak Opportunity Foundation (ROOF), and several generous donors who wish to remain anonymous.

This sponsorship support enables CEID to provide complimentary copies of the Guide to Pediatric Residents who participate in on-site rotation training at CEID and to public health nurses and early intervention specialists in the San Francisco Bay Area.

Historically, the Guide has enjoyed the use and support from many in the United States and throughout the world. We encourage that usage with this third edition, and if we at CEID can be of any help to you and your achieving your objectives in exemplary early intervention, please contact us.

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Chapter 2

Parent Perspectives

Throughout this Guide, the reader will be provided with the latest information on childhood deafness and the recommended approaches for early screening, diagnosis, and referral – the pedestals for helping children develop meaningful and competent communication. In some ways the information provided may appear to be solely technical as it strives for currency, accuracy, and professionalism. But there is a very human dimension to childhood deafness.

Just as their hearing peers, children who are deaf and hard of hearing come into this world with different temperament styles, emotional competence, physiological rhythms, and physical strengths. It is our shared role and responsibility as key stakeholders in early intervention to help uncover their styles and strengths and help each child reach his or her potentials in all areas of development.

It is also important to remember that deafness crosses all cultural, socio-economic, linguistic, and religious communities. There is no one etiology for deafness, nor is there one best approach to helping a child develop meaningful and competent communication. The parent experiences and recommendations included in this chapter offer the Primary Pediatric Provider personal testimonials shared through the eyes and hearts of 13 families, each of whom provides you, the trusted messenger, a unique opportunity for supporting parents through early diagnosis, intervention, and educational decisions. It is our hope that you'll agree with us that while parents are a child's most important first advocates, language models, and decision makers, it is your informed and unbiased guidance that is most critical to helping families traverse the early weeks and months of their journey, helping them to overcome challenges and barriers, as well as gain confidence in their abilities to make effective decisions and learn new skills for early and meaningful communication.

Children cannot, nor should not, be defined by their deafness.

What follows are these children's own stories, shared in their parents' own words.

- **Marcus and Kaiyu:** Brothers with hereditary hearing loss who use binaural hearing aids
- **Rafa:** A child with a progressive loss who uses one cochlear implant and one hearing aid
- **Anaya:** A child with an unknown etiology who uses bilateral cochlear implants
- **Norah:** A child with a unilateral mild loss and auditory neuropathy
- **Caitlin:** A twin with an unknown etiology–15 years ago
- **Maria:** A child who was a high-risk neonate
- **Sunnan:** A child who has Usher syndrome
- **Alondra:** A child who has Nager syndrome
- **Aaron's Story:** A child who has bilateral cochlear implants and autism spectrum disorder
- **Henry:** A child who has Cornelia de Lange syndrome
- **Josie:** A child who has Velocardiofacial syndrome
- **Scarlett:** A child who has a brain tumor
- **Jason:** A child with a family history of hearing loss and prematurity

Types of Hearing Loss

A. Conductive Hearing Loss: A failure of the sound waves to be transmitted to the inner ear. A conductive hearing loss results when conditions occur that interfere with or block sound vibrations transmitted through the outer or middle ear. Common causes are cerumen in the canal, narrow canal, middle ear effusion, ossicle damage, perforated drum, cholesteatoma. Conditions: Persistent OM with effusion; Crouzon, Treacher Collins, Alpert, CHARGE.

B. Sensorineural Hearing Loss: A failure of transduction of sound into neural activity. Causes can be both genetic and environmental, including cochlear hair cell damage, cochlear metabolic dysfunction, auditory nerve damage, agenesis, ototoxic drugs, noise, trauma or gene abnormality. Conditions: Waardenburg, Pendred, Alport, Jervell and Lange-Nielsen, Usher, and Goldenhar.

C. Mixed Hearing Loss:

A combination of conductive and SNHL. Can occur when outer ear and inner ear, or middle ear and inner ear are involved.

D. Auditory Neuropathy Spectrum Disorder (ANSO) or Auditory Dyssynchrony Hearing Loss:

Transmission of signals from inner ear to brain is impaired. Outer hair cells of cochlea initially function, but there is auditory nerve pathology or a deficit of cochlear inner hair cells or an abnormality in path from spiral ganglion fibers up through brainstem. OAE initially normal. ABR is absent or abnormal.

Discussions continue as to whether the term “auditory neuropathy” best describes this condition, which is physiologically described as auditory dyssynchrony and is thought to have a number of causes including synaptic disorders and neuropathies. Arguments against the less anatomically accurate term of “auditory neuropathy” suggest that referring to the disorder as a neural loss is more accurate, unless the etiology is known to be a neuropathy in the strictest sense. (Nachman, 2012)

Some of the known risk factors related to ANSD include neonatal hypoxia and/or anoxia requiring mechanical ventilation, neonatal hyperbilirubinemia to a level requiring exchange transfusion, low birth weight, prematurity (<28 weeks), genetic or family history of ANSD. (EHDI E-book, 2014)

Some children with ANSD have associated neurological disorders that cause problems outside of the hearing system, such as Charcot-Marie-Tooth syndrome and Friedreich’s ataxia. Children with Charcot-Marie-Tooth syndrome and Friedreich’s ataxia are at risk for late-onset ANSD and should be referred for audiology assessment at least by 24 to 30 months of age. (Guidelines, 2008)

A child with ANSD may present with any degree of hearing loss and can even present with normal responses to tonal stimuli.

Clinical audiological features of ANSD may include poor speech understanding and speech productions, despite normal pure-tone thresholds. A child may hear, but fail to understand, the words being said. Sounds may fade in and out (e.g., poor cell phone reception – “cutting in and out”).

The disorder can affect one ear (unilateral) or both ears (bilateral). It can be an unpredictable condition that can remain stable, fluctuate, and/or worsen.

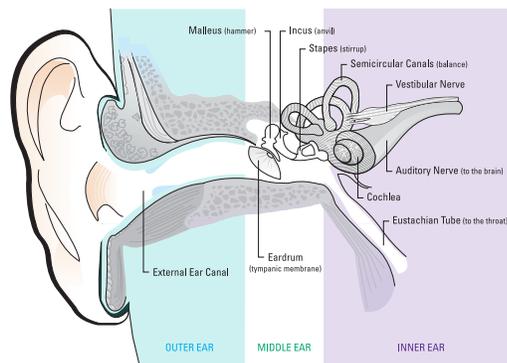
Patients with ANSD can be distinguished from patients with

other hearing disorders by four typically presenting physiologic responses:

- Present otoacoustic emissions, reflecting normal function of the outer hair cells in the cochlea, which can disappear over time
- Absent or very abnormal diagnostic ABR
- Present cochlear microphonics
- Absent acoustic middle ear muscle reflexes

Special care should be made to include imaging (MRI) and referral to a neurologist in the case of a unilateral ANSD. A recent case documented a child who later developed a tumor (a juvenile pilocytic astrocytoma) which would present with the same clinical presentation as ANSD (normal OAEs and absent ABR). (Nachman, 2012)

A child’s understanding or perception of speech cannot be determined by the level of residual hearing. (Rance et al, 1999)



A Step-by-Step Approach to Early Intervention

After the initial screenings using AABR and OAE are completed and a “refer” for comprehensive diagnostic testing is determined, support from the Primary Pediatric Provider and Medical Home can help the family to take the following steps.

1. Confirmation of Hearing Loss

(Prior to three months of age)

It is essential that a specially trained and experienced Pediatric Audiologist administer a variety of tests to confirm the degree and type of hearing loss. Most parents are unfamiliar with and often anxious about the tests, including the various evaluation components and interpretation of the findings. It is helpful to provide written information to the parents and primary caretakers that describes the comprehensive test battery, including:

- a. Diagnostic Auditory Brainstem Response (frequency-specific and bone conduction)
- b. Diagnostic Otoacoustic Emissions
- c. Acoustic Immittance (to evaluate status of middle ear)
- d. Behavioral Measures (children six months and older)

2. Referral to Early Intervention Programs and Services

(Can be simultaneous with the diagnostic process)

With the support of the Early Hearing Detection and Intervention Program (EHDI), all 50 states, the District of Columbia, and territories have created and implemented their respective newborn hearing screening programs and associated services for infants and their families eligible for their programs. Part C of the IDEA outlines such services. (www.ed.gov/about/offices/list/osers/osep/index.html).

The first step in referring a newborn and his or her family for direct services and family support is usually to the Department of Education, i.e., the Special Education Local Planning Agency (SELPA), the County Office of Education (COE), the Local Education Agency (LEA), or a state coordination center within the Department of Health. Some states link their programs with a local university or contracted non-profit agency. For more information about your state’s specific contacts and point of entry please refer to www.infantheating.org.

Qualified specialists from the designated state agency will work to develop an Individualized Family Service Plan (IFSP) for the child (and his or her family), who is found eligible for services based on each state’s unique eligibility criteria. (Some states determine eligibility when the newborn does not pass the hospital screen, while other states consider the infant eligible when the diagnosis is confirmed.) An IFSP is a written plan, designed around the unique needs of the child and family that identifies the strengths of the child and his or her family, and outlines the major outcomes expected to be achieved by the family’s involvement in the Early Intervention (EI) process. EI services may include:

- Assistive Technology Devices
- Audiology Services
- Family Training, Counseling, and Home Visits
- Health Services
- Medical Services (for diagnostic and evaluation purposes only)
- Nursing Services
- Nutrition Services
- Occupational Therapy Services
- Opportunities to interact with members of the Deaf community
- Physical Therapy Services
- Psychological Services
- Service Coordination
- Social Work Services
- Special Instruction
- Speech-Language Pathology Services
- Transportation and Related Costs
- Vision Services
- Other States-Specific Services