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DARS is the state agency responsible for ECI services, contracting with local ECI programs to provide services in every Texas county.

DARS Inquiries Line: 1-800-628-5115
TDD/TTY: 1-866-581-9328
www.dars.state.tx.us/ecis

Information at all stages of the EHDI process (1.3.6) should be communicated to the family in a culturally sensitive and understandable format.

(Joint Committee on Infant Hearing, 2007 Position Statement)
# Table of Contents

**Just in Time: Resources for Primary Care Providers**  
Texas Early Hearing Detection and Intervention (TEHDI)

**Acknowledgements (Inside Cover)**

**A.1 Table of Contents**

**Introduction & Background**

1.1 Introduction, Lakshmy Rajagopalan, M.D., J.D.

1.2 A Parent’s Perspective, Lisa Crawford, Texas Chapter of Hands and Voices

1.3 Facts

1.4 The Department of State Health Services

**Recommended Protocols**

2.1 American Academy of Pediatrics Guidelines for Pediatric Medical Home Providers

2.2 Texas 1-3-6 Practitioner’s Guide

2.3 Just in Time: Protocols for Primary Care Providers

**Managing Hearing Loss**

3.1 Risk Factors

3.2 Congenital Cytomegalovirus (CMV) Infection & Hearing Loss, Karen Fowler, University of Alabama

3.4 American Academy of Pediatrics EHDI Articles

3.5 Medical Care of Children With SNHL (A child in my practice has been identified with a permanent hearing loss. Now what do I do?), Susan Wiley, M.D., FAAP

3.7 Beyond Newborn Hearing Screening (Recognizing the Signs of Late Onset Hearing Loss in Infants and Young Children), Leisha Eiten, MA, CCC-A

3.9 Genetics of Early Childhood Hearing Loss-The Facts, G. Bradley Schafer, M.D.

3.11 Pediatric Auditory Neuropathy/ Dys-synchrony, Ryan McCreery, Boys Town National Research Hospital

**Early Intervention**

4.1 Physician Role in Early Intervention

4.2 Navigating the Texas Early Intervention System

4.3 Early Intervention Service System for Families with Children Who are Deaf or Hard of Hearing

4.6 Communication Opportunities for Families and Children

4.8 Speech and Language Milestones

**Resources**

5.1 Texas State Resources

5.6 Web Resources for Providers and Families

5.7 Physician Access
Introduction and Background

The goal of early hearing detection and intervention (EHDI) is to maximize linguistic competence and literacy development for children who are deaf or hard of hearing. Without appropriate opportunities to learn language, these children will fall behind their hearing peers in communication, cognition, reading, and social-emotional development.

(Joint Committee on Infant Hearing, 2007 Position Statement)
Permanent congenital hearing loss occurs in 1 to 3 per 1,000 live births. It is the most prevalent condition among all the disorders screened for in the newborn period. Hearing loss can be associated with risk factors such as craniofacial abnormalities, congenital infections with cytomegalovirus; however, about half of children with hearing impairment have no identifiable risk factors.

In the past, infants with moderate to severe hearing loss were not diagnosed until well beyond the newborn period, and diagnosis for those with mild or unilateral hearing loss may not even have occurred until school age; however, this has changed with the implementation of Early Hearing Detection and Intervention (EHDI) programs all across the country. These programs have been endorsed by multiple groups including American Academy of Pediatrics (AAP), Joint Committee on Infant Hearing (JCIH), National Institute on Deafness and other Communicative Disorders (NIDCD), and the Centers for Disease Control and Prevention (CDC).

EHDI programs have not only shown the feasibility of screening all the newborns but also the benefits of early identification and intervention. Infants who are diagnosed with hearing loss and provided with appropriate intervention before 6 months of age perform as much as 20-40 percentile points higher on school-related measures (vocabulary, intelligibility, articulation, social adjustment, and behavior) than infants who are not given this opportunity.

In Texas, 2 neonates are born every day with hearing loss. Texas established its Early Hearing Detection and Intervention program (TEHDI) in 1999, following the passage of House Bill 714. The law requires qualified birth facilities offer newborn hearing screening to all families of newborns during the birth admission. The 1-3-6 goals of TEHDI include hearing screening of all newborns before discharge from the hospital, audiologic and medical evaluation in those that do not pass the second screening before 3 months of age, appropriate intervention of the infants who are found to be hearing impaired before 6 months of age, and continued monitoring for late onset hearing loss in the pediatric population. All of this care must be provided in a culturally sensitive, family-centered medical home.

Though Texas does a stellar job screening 97–98% of all its newborns, it appears not all infants are receiving the necessary diagnostic evaluation and the appropriate intervention. Statistics from the Texas Department of State Health Services (DHS) reveal that 40–50% of infants who undergo hearing screening appear to be ‘lost to follow-up’ in the TEHDI continuum, and state records show only a fraction of infants with hearing impairment are identified and receive timely, appropriate early intervention services. In response, the Texas Deaf and Hard of Hearing Leadership Council was established in November 2006. The Council has been charged to help improve the outcomes of infants, toddlers, and families, as outlined in the 2005–2006 Comprehensive Statewide Plan for Educational Services for Students who are Deaf and Hard of Hearing.

Medical homes are central to the success of the Texas Early Hearing Detection and Intervention (TEHDI) program. As physicians taking care of children in Texas, we have both the power and the responsibility to adhere to the principles of the TEHDI program for each one of our pediatric patients, in a family-centered, culturally sensitive environment. This resource guide was created to help us fulfill these goals laid out by the TEHDI program. Let us strive to achieve the TEHDI goals for all the children in Texas, so that they have the opportunity to reach their full potentials.

All infants for whom the family has significant concerns regarding hearing or communication should be promptly referred for an audiological and speech-language assessment.

(Joint Committee on Infant Hearing, 2007 Position Statement)
A Parent’s Perspective:
Lisa Crawford, Texas Chapter of Hands and Voices

I am the parent of a daughter with a profound hearing loss. She is eleven years old now, but I vividly remember the day we were told she had a hearing loss. I was quite literally in shock and remember the helpless feeling of not knowing what to do next. My first thought was to turn to the medical professionals that surrounded me, but my husband and I soon discovered that although professionals were typically helpful, we were at the beginning of a solo journey. Now this all may sound a bit depressing, but in fact it is not. I learned that my child and my family were unique and no matter what a professional knew about hearing loss, there was always something they didn’t know about us. It was our job as parents to navigate the information, work closely with the professionals in my daughter’s life, and ultimately make the best choices for our family.

Looking back, one of the greatest gifts we received was from our daughter’s pediatrician. Newborn hearing screening was not in place when she was born so we had no idea she had a hearing loss. I started to suspect something when she was about 7 months old. The gift our doctor gave us was that he listened to me. I called him and told him my suspicion, and he immediately referred my daughter for ABR (Auditory Brainstem Response) testing. He didn’t negate my experience or tell me it was a phase—he trusted my parental instincts, and that allowed us to get her diagnosed quickly during a very critical time of language development. I will always be grateful to him for his response.

His support did not end there. When I visited him after the testing, he asked me if I had made connections with my school district and found services, etc… I informed him that I had. I believe I even surprised him a bit by telling him about a few programs he did not know about. How, may you ask, was that supportive? He didn’t pretend to know more than he did. Hearing loss is a low-incidence disability, and many doctors will never encounter it, especially in an infant. I would much rather have a doctor be willing to learn with me than one who pretends to know more than they actually do. I have heard stories from other parents who had doctors lead them in a particular direction that ultimately was not a good choice for them. My doctor did not try to lead us, but rather joined our journey for a short time and gave us the encouragement to move on to the next phase, more confident and informed than when we began.

Families should be made aware of all communication options and available hearing technologies (presented in an unbiased manner). Informed family choice and desired outcome guide the decision-making process.

(Joint Committee on Infant Hearing, 2007 Position Statement)

Lisa Crawford, Parent
Co-Founder Texas Chapter Hands and Voices
Facts

Hearing loss is one of the most common congenital anomalies occurring in approximately 12,000 children each year or 3 of every 1,000 births. This means 33 babies are born in the United States every day with permanent hearing loss.

(Universal Newborn Hearing Screening, National Center for Hearing Association and Management, Utah State University, retrieved June 30, 2008, www.infanthearing.org)

50% of infants born with hearing loss have no known risk factors.
(De Michele, Anne, Roger, Ruth; Newborn Hearing Screening, retrieved June 30, 2008, www.emedicine.com)

Without universal screening by 1 month, the average age at which hearing loss is identified is 2 to 3 years old. Earlier identification means improvements in speech and language and all developmental outcomes.
(Center for Disease Control and Prevention, retrieved November 11, 2009, www.cdc.gov/ncbddd/ehdi)

When hearing loss is not identified early, children are typically unable to develop speech, language, cognitive, and social skills with their hearing peers. This prevents them from developing the foundation skills required to be successful in school.
(Universal Newborn Hearing Screening, National Center for Hearing Association and Management, Utah State University, retrieved June 30, 2008, www.infanthearing.org)

Children with hearing loss who receive early intervention before 6 months of age are 1–2 years ahead of their later identified peers in language, cognitive, and social skills by the time they are in first grade.
(Universal Newborn Hearing Screening, National Center for Hearing Association and Management, Utah State University, retrieved June 30, 2008, www.infanthearing.org)

Infants with hearing loss can be fitted and benefit from amplification before they are 1 month of age.

Children with a unilateral or mild hearing loss that remains undetected are more likely to be held back one grade than a matched group of children with normal hearing.
(Universal Newborn Hearing Screening, National Center for Hearing Association and Management, Utah State University, retrieved June 30, 2008, www.infanthearing.org)

About 3 out of every 1,000 children in the United States are born deaf or hard of hearing. Nine out of every 10 children who are born deaf are born to parents who can hear.

Often parents think they can identify a hearing loss in their infant; however, children can compensate for a hearing loss using visual cues, such as parental expressions and reactions. Without hearing screening, a child’s hearing loss is likely to go undetected until he/she is 2 to 3 years of age.
The Department of State Health Services:

Texas Early Hearing Detection and Intervention Program

The mission of the Texas Department of State Health Services (DSHS) Texas Early Hearing Detection and Intervention (TEHDI) Program is for all babies born in Texas to receive Newborn Hearing Screening and appropriate follow-up care. It is essential infants with hearing loss be identified early and appropriate intervention services be initiated as early as possible. Without early identification and intervention, children with hearing loss may experience delays in the development of language, cognitive, and social skills that may prevent success in academic, occupational, and personal achievements.

DSHS has modeled much of the TEHDI Program after the US Department of Health and Human Services Centers for Disease Control and Prevention (CDC) for Early Hearing Detection and Intervention (EHDI). Texas is pursuing the same goals addressed nationally in the EHDI program. Each goal set forth has specific program objectives; these objectives are essential in accomplishing each goal. Each objective in the TEHDI Program has a quantitative measure (performance indicator) that is used to track progress toward realizing the program goals.

**Goal 1:** All newborns will be screened for hearing loss before 1 month of age, preferably before hospital discharge.

**Goal 2:** All infants who screen positive will have a diagnostic audiologic evaluation before 3 months of age.

**Goal 3:** All infants identified with a hearing loss will begin receiving appropriate early intervention services before 6 months of age.

**Objective:** The first three goals are referred to as the 1-3-6 Plan. The objective of these goals is to provide the earliest possible assessment, referral, and intervention for newborns.

**Goal 4:** All infants and children with late onset, progressive, or acquired hearing loss will be identified at the earliest possible time.

**Objective:** A comprehensive TEHDI tracking and surveillance system must go beyond the 1-3-6 Plan. There is a need for the TEHDI system to identify and track infants and young children who are missed or who do not have an identified hearing loss at birth. Primary health care providers (PCPs), parents, and other care providers should refer an infant for screening any time they suspect a hearing loss. Audiologists who later identify infants and children with a hearing loss should report such losses to the TEHDI tracking system.

**Goal 5:** All infants with hearing loss will have a medical home.

**Objective:** PCPs play a key role in the success of the EHDI program. It is critical infants and their families have a medical home and that services be coordinated between the medical home and the EHDI program. PCPs can help families understand the EHDI process and ensure infants referred for audiologic evaluation complete that evaluation by 3 months of age.
Recommended Protocols

Infants and children with mild to profound hearing loss who are identified in the first months of life and provided with immediate and appropriate intervention have significantly better outcomes than later identified children in vocabulary development, receptive and expressive language, syntax, speech production, and social and emotional development.

(Joint Committee on Infant Hearing, 2007 Position Statement)
Universal Newborn Hearing Screening, Diagnosis, and Intervention Guidelines for Pediatric Medical Home Providers

**Ongoing Care of All Infants**
- Provide parents with information about hearing, speech, and language milestones
- Identify and aggressively treat middle ear disease
- Provide vision screening and referral as needed
- Provide ongoing developmental surveillance and referral to appropriate resources

- Identify and refer for audiologic monitoring infants who have the following risk indicators for late-onset hearing loss:
  - Parental or caregiver concern regarding hearing, speech, language, and/or development delay
  - Family history of permanent childhood hearing loss
  - Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian tube dysfunction
  - Postnatal infections associated with sensorineural hearing loss including bacterial meningitis
  - In utero infections such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis
  - Neonatal indicators—specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation
  - Syndromes associated with progressive hearing loss such as neurofibromatosis, osteopetrosis, and Usher syndrome
  - Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth disease
  - Head trauma
  - Recurrent or persistent otitis media with effusion for at least 3 months

*OAE = Otoacoustic Emissions, ABR = Automated Auditory Brainstem Response, IDEA = Individuals with Disabilities Education Act

Notes:
(a) In screening programs that do not provide Outpatient Screening, infants will be referred directly from Initial Screening to Pediatric Audiologic Evaluation. Likewise, infants at higher risk for hearing loss, or loss to follow-up, also may be referred directly to Pediatric Audiologic Evaluation.
(b) Part C of IDEA* may provide diagnostic audiological evaluation services as part of Child Find activities.
(c) Infants who fail the screening in one or both ears should be referred for further screening or Pediatric Audiologic Evaluation.
(d) Includes infants whose parents refused initial or follow-up hearing screening.
TEHDI
Texas Early Hearing Detection and Intervention

1-3-6 MONTH PRACTITIONER’S GUIDE
TEXAS EARLY HEARING DETECTION AND INTERVENTION (TEHDI) PROCESS

BEFORE 1 MONTH SCREENING

PASS
HEARING SCREEN
at Birth Facility

PASS
2ND HEARING SCREEN
at Birth Facility before discharge

REFER
FOLLOW-UP SCREEN
as an outpatient

BEFORE 3 MONTHS CONFIRMATION DIAGNOSIS

TO AUDIOLOGIST:
DIAGNOSTIC EVALUATION
Using Texas Evaluation Protocol

HEARING LOSS CONFIRMED

REPORT RESULTS & REFERRALS - TEHDI

REFERRAL TO EARLY CHILDHOOD INTERVENTION (ECI)

BEFORE 6 MONTHS

ENROLLED IN ECI AND RECEIVING APPROPRIATE EARLY INTERVENTION, HEARING & MEDICAL SERVICES

REPORT SERVICES - TEHDI

EARLY CHILDHOOD INTERVENTION (ECI)

AUDIOMETRIC HEARING DIAGNOSTIC EVALUATION

REPORT SERVICES - TEHDI

REPORT SERVICES - TEHDI
1. Birth Screen
- Parental permission is required.
- Test is either Auditory Brainstem Response (ABR) or Transient or Distortion Product Otoacoustic Emissions (OAE).
- A second screen is done before discharge if the first is not passed.
- Written results are given to the parents and the baby’s doctor.
- Results are reported to DSHS but identifying information is removed for infants who pass; parental permission is given for identified results to be reported.
- Referral to a local audiology/hearing resource is made for outpatient re-screen when an infant does not pass the second screen.

2. Outpatient Re-Screen
- ABR or OAE tests are used.
- If the infant does not pass, referrals are made to an audiologist for diagnostic hearing testing and to Early Childhood Intervention (ECI) at 1-800-628-5115.
- Hearing services are available for children who are eligible through the Texas Medicaid Program and Children with Special Healthcare Needs (CSHCN).

3. Audiologic Evaluation
- Diagnostic ABR and, to verify cochlear involvement, OAE if not previously done.
- The Texas Pediatric Protocol for Evaluation is used; see www.dshs.state.tx.us/tehdi/assumpt.shtml.
- Results are reported to the referral source and to TEHDI.
- Referral is made to ECI upon the diagnosis of hearing loss.
- Referral to an otologist for a medical examination of the ear.
- Fitting of hearing aids by an audiologist when appropriate.
- Ongoing audiological assessment and monitoring as needed.

4. Referral to ECI
- Must be within two working days of the diagnosis of hearing loss.
- Service coordination is provided by ECI.
- Parents may refuse ECI services.
- An Individual Family Services Plan (IFSP) will be developed by ECI within 45 days of referral.
- ECI and the Local Education Agency (LEA) have shared service responsibility for children with hearing loss.

5. Deaf Education and other special education services are available from ages 3–21 when determined by the Individual Education Program (IEP).

6. For children who pass the newborn hearing screen, the Medical Home/physician continues to monitor for developing hearing loss; see http://pediatrics.aappublications.org/cgi/content/full/120/4/898 for suggested monitoring protocols.

Additional Resources:
www.callier.utdallas.edu/txc.html for Texas Connect – Educational Information; Educational Resource Center on Deafness at 1-800-332-3873.

For more information about TEHDI call 1-800-252-8023 or 512-458-7726 or visit our website at www.dshs.state.tx.us/tehdi/newbornhear.shtml
Just in Time

Protocols for Primary Care Providers

Before ONE Month—Hearing Screening

• Ensure inpatient or birth admission screening has been completed.

Utilize Provider Access at https://www.provideraccess.tehdi.com to review the following protocols:

• Review results of the initial birth admission screen.
• When the birth admission screening indicates a referral is required, ensure outpatient screening is completed by 4 weeks.
• Review risk factors for late-onset or progressive hearing loss with the parent or guardian.
• Screening technologies used for screening should be either Auditory Brainstem Response (ABR) or Transient or Distortion Product Otoacoustic Emissions (OAE).
• Schedule diagnostic audiological evaluation for all infants who did not pass the outpatient screen.
• Consider a referral to Early Childhood Intervention (ECI; IDEA Part C). ECI will accept referrals when a baby has failed the outpatient screening and provide support to families through the diagnostic process.

Before THREE Months—Diagnostic Evaluation

• Monitor results of the diagnostic evaluation performed by a pediatric audiologist.
• Tests should include a diagnostic ABR and an OAE, if not previously completed, to verify cochlear involvement.
• When a diagnosis is confirmed:
  • Schedule an ENT and ophthalmology examination.
  • Provide referrals for genetics, neurology, and developmental pediatrics.
  • Refer to Early Childhood Intervention (ECI; IDEA Part C) within 48 hours of diagnosis.
  • Provide medical clearance for hearing aids, cochlear implants, and/or therapies if chosen by the family.

Before SIX Months—Early Intervention

• Monitor completion of diagnostic processes and medical specialty referrals.
• Monitor pediatric audiology services including ear molds, hearing aids, information about cochlear implants, and other follow-up.
• Review all follow-up results with the family to help them understand the type and severity of the hearing loss and options for intervention including communication choices.
Managing Hearing Loss

All infants, regardless of the newborn hearing screening outcome, should receive ongoing monitoring for development of age-appropriate auditory behaviors and communication skills. Any infant who demonstrates delayed auditory and/or communication skills development, even if he or she passed newborn hearing screening, should receive an audiological evaluation to rule out hearing loss.

(Joint Committee on Infant Hearing, 2007 Position Statement)
Risk Factors

For Hearing Loss in Infants, Toddlers, and Children

There are conditions which may occur before birth, during infancy, or in early childhood that may affect a child’s ability to hear normally. The factors may indicate that a child is at risk for a hearing loss.

- Family history of childhood hearing loss
- Infection during pregnancy (cytomegalovirus, rubella, syphilis, herpes, and toxoplasmosis)
- Craniofacial anomalies including those affecting the pinna and ear canal
- Low birth weight (less than 3.3 lbs.)
- Ototoxic medications
- Bacterial meningitis or other infections known to cause hearing loss
- Apgar scores of 0 to 4 at one minute or 0 to 6 at five minutes
- Mechanical ventilation lasting 5 days or longer
- Symptoms of syndromes known to include hearing loss
- Head trauma associated with loss of consciousness or skull fracture
- Otitus Media with effusion (fluid) for three months or longer
- Parent or caregiver concern regarding hearing, speech, and language or any developmental delay
- If none of these conditions are present but there is concern about a child’s hearing, a referral should be made to an audiologist.

The primary care physician should review every infant’s medical and family history for the presence of risk indicators that require monitoring for delayed-onset or progressive hearing loss and should ensure that an audiological evaluation is completed for children at risk of hearing loss at least once by 24 to 30 months of age, regardless of the newborn hearing screening results.

(Joint Committee on Infant Hearing, 2007 Position Statement)
Overview of Congenital Cytomegalovirus (CMV) Infection

Congenital CMV infection is the most common intrauterine infection in the United States today. The incidence of congenital CMV infection varies by geography and the underlying CMV seroprevalence in the maternal population. Incidence estimates of congenital CMV infection range between 0.2% and 2.2%, with an estimated 0.5% to 1.0% of all newborns in the United States infected with CMV in the prenatal period. Interestingly, the incidence of congenital CMV infection is higher in populations where the underlying CMV seroprevalence or preexisting immunity is higher in the mothers. Whether the increase in congenital CMV infection in these populations is due to certain exposure factors in the maternal delivery populations or the racial and ethnic composition of the population or a combination of these factors is not known.

Congenital CMV infection at birth may manifest with symptoms such as generalized petechiae, direct hyperbilirubinemia, hepatosplenomegaly, purpuric rash, microcephaly, seizures, focal or general neurologic deficits, retinitis, and intracranial calcifications. However, 90–95% of infants with congenital CMV infection will have no clinically apparent symptoms (asymptomatic) at birth. Most congenital CMV infections go unnoticed in the nursery since the majority of infected newborns will have no clinically apparent disease at birth.

Both symptomatic and asymptomatic infants may later develop sequelae, with more severe and frequent sequelae occurring in the symptomatic infants. Sequelae following congenital CMV infection include sensorineural hearing loss (SNHL), retinitis, mental retardation, microcephaly, seizures, and cerebral palsy. The most common sequelae following congenital CMV infection is SNHL.

Sensorineural Hearing Loss due to CMV

The association between congenital cytomegalovirus (CMV) infection and SNHL has been known for over four decades, although the mechanism by which the virus causes hearing impairment in some children and not others is still not fully understood today. Approximately 30–50% of children with clinically apparent disease (symptomatic) and 8–12% of children without clinically apparent (asymptomatic) congenital CMV infection will develop SNHL.

Hearing loss due to congenital CMV infection does not have a pathogenomonic audiometric configuration and is variable in the severity of loss. Unilateral and bilateral hearing losses may occur in children with congenital CMV infection, with loss varying from unilateral high frequency losses (4–8 khz frequencies only) to profound bilateral losses. Progression and fluctuation of hearing loss have been observed in children with congenital CMV infection. Complicating the diagnosis of SNHL in children due to congenital CMV infection is the fact that less than half of the hearing loss due to CMV infection is present at birth. Other CMV infected children may go on to develop late-onset loss during the preschool and early school years. Approximately 33 to 50% of SNHL due to congenital CMV infection is late-onset loss.

The fact that congenital CMV infection can only be confirmed in the newborn period has made it difficult to estimate the proportion of SNHL that is attributable to congenital CMV infection in childhood populations. A recent review estimates that congenital CMV infection accounts for approximately 21% of all hearing loss at birth. Since late-onset losses may occur following CMV infection, about 25% of hearing loss in children by four years of age is likely CMV-related hearing loss. These numbers suggest that CMV is the leading nongenetic cause of hearing loss in children in the United States.

Identifying Congenital CMV Infection

The diagnosis of congenital CMV infection at birth is usually by the detection of the virus in urine or saliva within the first three weeks of life. The detection of CMV in saliva or urine can be readily accomplished in newborns with congenital CMV infection because infected infants shed large amounts of virus. Traditional virus isolation in tissue culture in which viral cytopathic effect (CPE) is detected by light microscopy has been the standard. In the past
two decades, rapid viral diagnosis utilizing centrifugation-enhanced inoculation of the specimen onto the monolayer of fibroblasts followed by the detection of CMV antigens, such as shell vial assay or the detection of early antigen fluorescent foci (DEAFF), have been used with comparable sensitivity and specificity to standard viral isolation procedures. The rapid methods provide results within 24 hours, compared with the longer time frame (2–4 weeks) for tissue culture method. PCR methods to identify CMV DNA from dried blood spots, urine and saliva have been explored in recent years. Although CMV may be identified by PCR methods, the sensitivity and specificity of these methods have not been established to date.

**Children with Hearing Loss due to CMV**

Most children with congenital CMV infection do not develop hearing loss; however, it is unclear which children with congenital CMV infection will develop hearing loss, and, among those who do develop loss, whether or not the loss will continue to deteriorate. Approximately 50% of children with SNHL due to congenital CMV infection will have progressive losses. Since late-onset hearing loss along with fluctuations and progression of the loss may occur with CMV-related hearing losses, children with congenital CMV infection should have hearing tests every six months to monitor possible changes to their hearing loss. However, at times when hearing loss appears to be changing, audiological evaluations may be needed every three months to assess and document the changes in the hearing status of the child infected with CMV. In addition, parents should be encouraged to observe their child for signs that his or her hearing may be changing, i.e., not hearing the dog barking, turning up the volume of the television, not wanting to wear hearing aids when before she did not mind, etc. Finally, parents and health-care providers should select communication methods that accommodate changing hearing loss and when considering hearing aids choose those with the most flexible gain.

**References**


NOTE: This article was prepared by Karen B. Fowler, DrPH, Department of Pediatrics, University of Alabama at Birmingham. Dr. Fowler can be reached at kfowler@uab.edu.
American Academy of Pediatrics
EHDI Articles

The following articles are designed to inform the primary care provider about monitoring hearing loss in children.

A child in my practice has been identified with a permanent hearing loss. Now what do I do?
Susan Wiley, M.D., FAAP

Beyond Newborn Hearing Screening (Recognizing the Signs of Late-Onset Hearing Loss in Infants and Young Children)
Leisha Eiten, MA, CCC-A

Genetics of Early Childhood Hearing Loss—The Facts
G. Bardley Schafer, M.D.

Pediatric Auditory Neuropathy/Dys-synchrony
Ryan McCreery, Boys Town National Research Hospital
A child in my practice has been identified with a permanent hearing loss. Now what do I do?

Susan Wiley, M.D., FAAP

Johnny was screened as part of the universal newborn hearing screening program in your state and did not pass. You referred Johnny for a diagnostic audiologic evaluation, and the report back informs you that he has a moderately severe sensorineural hearing loss. His mother comes in to follow up on these results and wants your opinion on some things that the audiologist recommended.

Although this is not a daily occurrence in your office practice, you realize that providing the right guidance and care is very important for Johnny and his family. Providing psychosocial support for the family and implementing medical and therapeutic/educational interventions are key to ensuring the best outcomes possible for Johnny.

It is not just about hearing; it is also about brain development and prevention of delays. Early and prompt intervention is critical. Hearing loss is considered a developmental emergency. As such, the health-care provider must support the family and ensure that there is no delay in beginning early intervention once hearing loss has been confirmed. This might include early fitting of amplification, early use of sign language, or both. The process of completing medical referrals should never delay prompt intervention once hearing loss is confirmed.

Developmentally, the needs of children with hearing loss are often served by three types of services:

1) audioligists knowledgeable in pediatric testing and amplification,
2) early intervention providers, and
3) speech-language pathologists.

Because hearing loss affects brain development, the developmental and behavioral care of children with hearing loss is also important for primary care providers to address. All children with hearing loss should have a managing audiologist who assists in monitoring the status of their hearing and effectiveness of amplification strategies. Young children often need frequent visits to the pediatric audiologist.

Referral to Early Intervention Services as soon as a diagnosis of hearing loss is made can empower families in meeting their child’s communication and developmental needs and decrease the negative impact of hearing loss on a child’s language and cognitive development. There may be many different types of expertise among early intervention providers. It is important to have access to professionals who can provide unbiased information about communication modes, provide appropriate family support, and provide expertise in the communication mode the family chooses.

Some families access private therapies beyond those provided by Early Intervention Services. Speech-language pathologists can monitor speech and language development and provide families with support and strategies to encourage language development for their child with a hearing loss. Speech-language pathologists may have a variety of skill sets. These skill sets (i.e., sign language, auditory-oral focus, cued speech, etc.) should support a family’s choices for their child’s communication development.

Medically, the primary goals are:

1) identification of an etiology of the hearing loss to help the family anticipate their child’s needs as well as those of siblings, and
2) preventing or diminishing the impact of secondary medical concerns (i.e., vision health, additive effect of middle ear disease on hearing levels).

To attain these medical goals, there are three subspecialty referrals that are considered standard of care.

Infants for whom there is concern regarding hearing or language should be referred for a speech and language evaluation and an audiology assessment.

(Joint Committee on Infant Hearing, 2007 Position Statement)
An otolaryngologist knowledgeable in pediatric hearing loss is involved in providing clearance for hearing aid use, undertakes a number of medical tests to determine an etiology of the hearing loss, and monitors children who might be candidates for the technology of a cochlear implant.

A geneticist knowledgeable in pediatric hearing loss assists in determining genetic causes of hearing loss, including syndromic and nonsyndromic. Geneticists provide information to families about recurrence risk and risks of hearing loss to siblings and extended family members.

An ophthalmologist knowledgeable in eye conditions associated with pediatric hearing loss can help identify conditions that can affect vision in children with hearing loss.

These referrals can be helpful in the initial evaluation and work-up of a child with sensorineural hearing loss; however, monitoring of medical conditions needs to continue at subsequent health maintenance and other illness-related visits.

Particularly relevant to children with hearing loss is aggressively managing middle ear disease. Although there are clinical protocols for the management of middle ear disease in typically developing children, a child with hearing loss requires a different approach. Even small changes in hearing related to middle ear disease can substantially decrease the impact of amplification on language development and behavior. Children who use hearing aids may not be able to tolerate their amplification during an ear infection. Persistent fluid may prolong the time of ineffective amplification. Referral to the otolaryngologist for these findings is recommended.

It is also important to follow typical anticipatory care for children with hearing loss, specifically regular vision screening and immunizations. Vision health is particularly important for children who have hearing loss. Additionally, children with inner ear abnormalities are at higher risk of acquiring meningitis. Children who are being considered for a cochlear implant should receive their full primary series and booster immunizations against pneumococcal disease and Haemophilus influenzae type b.

As Johnny’s primary care provider, you play a pivotal role in monitoring health conditions that can affect his progress and in supporting and empowering his family in your role as a medical home, a coordinator of the variety of medical and educational systems and providers that promote Johnny’s overall health and well-being.

Reference
Beyond Newborn Hearing Screening
Recognizing the Signs of Late-Onset Hearing Loss in Infants and Young Children
Leisha Eiten, MA, CCC-A

Note: This article was prepared by Leisha Eiten, MA, CCC-A, Clinical Audiologist, Boys Town National Research Hospital. Supported by the National Institute on Deafness and Other Communication Disorders (NIDCD R25 DC006460-03)

With 39 out of 50 states mandating universal newborn hearing screening (UNHS), and newborn hearing screening data being collected from 40 states, it would be easy to assume that the identification of permanent childhood hearing loss is guaranteed. Yet, some childhood hearing losses have a later onset and will not be identified through newborn screening methods. This article responds to some basic questions about late-onset hearing loss in infancy and childhood.

What is the prevalence of childhood hearing loss?
Current UNHS statistics indicate an overall hearing loss prevalence rate of 1–2 per 1,000 at birth. These prevalence statistics are consistent across the U.S. and are not dependent on the particular hearing screening method being used. Statistical information about the prevalence of hearing loss in older children is difficult to find and interpret for a number of reasons. Late-onset or progressive hearing loss can be due to hereditary factors, infection, trauma, noise exposure, or teratogens. Studies also vary in how “significant hearing loss” is defined. As a result, the prevalence of late-onset hearing loss is not well defined. In general there is a trend toward increasing rates of hearing loss as children get older.

Can newborn hearing screening miss hearing loss that is present at birth?
It is possible for some children to have a mild or minimal hearing loss at birth and pass universal hearing screening. This is due, at least in part, to the underlying assumptions about newborn hearing screening. Any type of universal screening program needs to achieve a low false-alarm rate and a high “hit” rate. The goal for UNHS is that few children are referred for additional, more expensive testing who do not need it and those who are referred have a high likelihood of having hearing loss. To meet these requirements, current UNHS methods may not identify children with mild hearing losses. If no further audiological monitoring is being completed within the child’s medical home, the result could be late identification of milder degrees of hearing loss.

In some instances, mild hearing loss that is present at birth may progress to more severe hearing loss after the child goes home from the hospital. Rapidly progressive hearing loss can be associated with several congenital conditions, including Cytomegalovirus (CMV) and Large Vestibular Aqueduct (LVA) as well as some genetically inherited losses.

What are the most common causes of late-onset hearing loss?
The major categories of late-onset loss are acquired, structural, and genetic.

Acquired: Among acquired late-onset losses, congenital CMV (both symptomatic and non-symptomatic) is the most common and accounts for around 1/3 of all hearing loss in children. Hearing loss associated with CMV may be both late-onset and progressive within the first years of life. Even asymptomatic congenital CMV infection carries an increased risk of hearing loss. Hearing loss prevalence rates of 7–15% in asymptomatic cases have been reported. Congenital symptomatic CMV infection carries greater risk for hearing loss and a higher percentage of children with active CMV symptoms at birth have hearing loss identified through UNHS, with further progression reported within the first 2–3 years (Fowler, et al 1997; Barbi, et al. 2003). Other childhood illnesses may also cause hearing loss. These include viral or bacterial meningitis, mumps, and other viral infections that cause a high fever or central sequelae. Head trauma with skull fracture is one type of traumatic late-onset loss. Chemotherapeutic agents containing platinum, such as cisplatin, are among the best known ototoxic medications.

Structural: Structural causes of late-onset hearing loss may occur with a number of syndromes. Structural deformities of the cochlea, such as LVA and Mondini malformation, are congenital but not always related to a specific syndrome. Cochlear malformations affect hearing differently in different children.
Some hearing losses may occur earlier, and others may not present until later childhood. Structural malformations of the inner ear are associated with sudden and extreme progression and fluctuation of hearing.

**Genetic:** Genetic causes of late-onset hearing loss may be syndromic or non-syndromic. Full explanations of specific syndrome characteristics can be found online on OMIM, the Online Mendelian Index in Man at www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=OMIM.

**Syndromic losses include:**

- Pendred’s Syndrome, which is associated with LVA
- Branchio-Oto-Renal Syndrome (BOR), associated with Mondini deformities
- Alports Syndrome with progressive renal failure and late-occurring, progressive hearing loss
- Stickler Syndrome, a connective tissue syndrome with late-occurring vision problems and hearing loss

**Non-syndromic losses include:**

- Dominant-progressive hearing loss
- Family history of late-occurring hearing loss
- Connexin 26, which may have late-onset hearing loss in rare occurrences. A small number of studies have shown progressive hearing loss with Connexin 26.

**What are the main risk factors associated with late-onset loss?**

- Congenital CMV infection
- Meningitis or mumps infections
- Family history of late-onset hearing loss

Infants with hearing loss can be fitted and benefit from amplification before they are 1 month of age.

(De Michele, Anne, Roger, Ruth; Newborn Hearing Screening, retrieved June 30, 2008)
Genetics of Early Childhood Hearing Loss — The Facts
G. Bardley Schafer, M.D.

The past decade has seen a continued increase in the utilization of newborn screening for hearing loss. As such, the number of infants identified with hearing loss is likewise on the rise. The major impetus behind infant screening has been the improved outcome of speech and language potential in children identified early. There is, however, another great advantage in identifying hearing loss early. A significant proportion of early childhood hearing loss has a genetic etiology. The family of every child identified with a significant hearing loss should be offered clinical genetics consultation. A genetics evaluation can provide several important pieces of information to the family and the child’s health care providers. First, a specific etiology may be determined. For many families, the “why” is an important question to be answered as its own end. If an etiology is determined, then specific information can be provided to the family regarding recurrence risks for the immediate and extended family. Also, some of the causes of childhood hearing loss have associated medical conditions. Many of these associated conditions can be medically serious. Knowledge of these risk factors can lead to interventions that prevent morbidity and mortality.

Genetics of Early Childhood Hearing Loss

- 60% of congenital deafness has a primary genetic etiology.
- A complete genetics evaluation in a young child with a significant hearing loss has a high diagnostic yield. A specific etiology can be identified in close to 90% of the cases.
- Two known causes of early childhood hearing loss—congenital CMV and connexin 26 mutations—each account for about 40% of the identified children.

Of the genetic causes of congenital deafness:

- 75% are autosomal recessive.
- 20% are autosomal dominant.
- 4% are X-linked.
- 1% are mitochondrial.
- 70% of genetic deafness is nonsyndromic; 30% is syndromic.
- The empiric recurrence risk (single case without a known etiology) is 10%.
- Otherwise, recurrence is diagnosis-specific.

The genetics evaluation of a young child is complex and is best accomplished in the context of an interdisciplinary team. Important components of this team include specialists in clinical genetics, genetic counseling, otolaryngology, ophthalmology, audiology, speech pathology, and vestibular physiology. In addition, this core team should have access to additional pediatric specialists as the work-up progresses. Frequently, consultations are required from pediatric cardiology, nephrology, endocrinology, and orthopedics.

The genetics evaluation of a child with hearing loss is ideally performed in stages (tiers). The evaluations should be structured so tests obtained in higher tiers have a higher expected diagnostic yield, lower invasiveness of testing, better potential of intervention, and easier overall practicality of obtaining the tests. Experience with this approach has a high level of acceptance with third party payers and with families. A general outline of such an evaluation includes:

Stage 1: Medical genetics, audiology, otolaryngology
Stage 2: Vestibular testing, ophthalmology, CT scan of the temporal bones, serology, urinalysis, and serum creatinine
Stage 3: Selected DNA tests based on the first 2 stages, electrocardiogram (if clinical indicators), and electroretinogram (if clinical indicators)

What might be found?

1) Connexin 26 mutations Connexin 26 (GJB2) mutations are found in 40% of infants with congenital hearing loss. This is inherited as an autosomal recessive trait. The phenotype is a non-syndromic hearing loss. Patients have normal vision and vestibular function. The hearing loss is non-progressive most of the time. The hearing loss is typically mild to profound with intra- and inter-familial variability. In a few kindred, the hearing loss is progressive and asymmetric.

2) Congenital Cytomegalovirus (CMV) The phenotype of an in-utero CMV infection can range from asymptomatic to a multiple anomaly complex that includes microcephaly, intracranial calcifications, cognitive impairment, dystonia, optic atrophy, retinopathy, cataracts, and microphthalmia. Congenital CMV can present as an isolated hearing loss. Congenital
hearing loss due to CMV accounts for another 40% of early identified hearing abnormalities. While it is not genetic per se, it has significant implications for associated medical problems, progression, and recurrence.

3) Genetic syndromes A variety of genetic syndromes may be associated with hearing loss. Below is a partial list of common syndromes with hearing loss and important medical problems that can be seen with these conditions.

- Branchio-oto-renal syndrome: wide range of renal anomalies, branchial arch malformations
- Pendred syndrome: thyroid goiter due to primary iodine organification defect
- Jervell and Lange-Nielsen syndrome: syncopal attacks, long QT syndrome, sudden death
- Waardenburg syndrome: pigmentary changes, Hirschsprung disease
- Usher syndrome: retinitis pigmentosa, vestibular abnormalities
- Mitochondrial-based hearing loss: diabetes, neuromuscular disorders, lactic acidosis

Available Resources

Regional Genetic Services
Geneticists, genetic counselors, primary care physicians, families, and others are coming together to identify and address the genetic service needs through participation in the Genetics and Newborn Screening Regional Collaboratives. The regional collaborative effort is funded by the Maternal Child Health Bureau to improve the health of children and their families by promoting the translation of genetic medicine into public health and health-care services. Each region identified priority areas and activities, formed subcommittees, and initiated physician and public education efforts. Several resources are available through the regions including CME opportunities, parent education resources, research studies, small grant programs, and quick access to established networks of local genetic service providers.

The regions are:
1) New England Regional Genetics Group (CT, MA, ME, NH, RI, VT) www.nergg.org
2) New York/Mid-Atlantic Region (District of Columbia, DE, MD, NJ, NY, PA, VA, WV) wadsworth.org/newborn/nymac
3) Southeastern Regional Genetics Group (AL, FL, GA, LA, MS, NC, Puerto Rico, SC, TN, Virgin Islands) www.sergginc.org
4) Region IV Genetics Collaborative (IL, IN, KY, MI, MN, OH, WI) region4genetics.org
5) Heartland Genetics and Newborn Screening Collaborative (AR, IO, KS, MO, ND, NE, OK, SD) heartland.ohsc.edu
6) Mountain States Genetics Regional Collaborative Center (AZ, CO, MT, NM, TX, UT, WY) www.mostgene.org
7) Western States Genetic Services Collaborative (AK, CA, Guam, HI, ID, NV, OR, WA) www.westernstatesgenetics.org

“Ask the Geneticist” A project of the Southeastern Regional Genetics Group. The mission of “Ask the Geneticist” is to answer questions about genetic concepts, and the etiology, treatment, research, testing, and predisposition to genetic disorders. Questions that meet these criteria are answered and posted to the site. Previously answered questions are archived on the website. “Ask the Geneticist” is a collaborative effort of the Department of Human Genetics at Emory University and the Department of Genetics at the University of Alabama at Birmingham and can be found at http://www.askthegen.org.

National Library of Medicine’s Genetics Home Reference
Physicians can help patients and families access up-to-date, reliable, consumer-friendly information about a genetic condition and the basics of genetic science by referring them to Genetics Home Reference, a free, patient-friendly website of the National Institutes of Health (NIH) at http://ghr.nlm.nih.gov. Genetics Home Reference includes over 500 topics on genetic conditions and related genes including congenital hearing loss and genetic conditions that may lead to hearing loss. The site features a richly illustrated tutorial that explains the basics of genetics, from the cellular level on up, and a glossary of genetics terms. The site is regularly updated by scientific staff and reviewed by external experts.

NOTE:
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Pediatric Auditory Neuropathy/Dys-synchrony
Ryan McCreery, Boys Town National Research Hospital

Universal newborn hearing screening (UNHS) is currently the standard of care in 42 states. In addition to lowering the average age of identification of permanent childhood hearing loss, advances in audiological assessment have resulted in greater specification of what portion of the auditory system might be affected in cases of permanent hearing loss. The ability to evaluate the function of the cochlea separately from the auditory nerve now exists. Children with auditory neuropathy or dys-synchrony have evidence of normal cochlear function, but show impairment in the function of the auditory nerve. Auditory neuropathy/dys-synchrony presents significant challenges related to identification and management of hearing loss during childhood.

What is auditory neuropathy/dys-synchrony (AN/AD)?
AN/AD is a type of sensorineural hearing loss that can be congenital or acquired. Unlike other types of sensorineural hearing loss where both Otoacoustic Emissions (OAE) and Auditory Brainstem Response (ABR) tests are likely to be abnormal, AN/AD is characterized by normal OAE results and significantly abnormal ABR responses, even when measured with very loud sounds. The combination of normal OAE responses and severely impaired ABR responses is thought to reflect normal outer hair cell (OHC) function in the cochlea and abnormal auditory nerve function. The site of lesion for AN/AD is often unknown, but possibilities include cochlear inner hair cells, cochlear spiral ganglia, and the auditory nerve. Audiograms of children with AN/AD vary from hearing in the normal range with complaints of difficulty hearing in background noise to profound hearing loss.

Who is at risk for AN/AD and how many children are affected?
Multiple risk factors for AN/AD have been described in the research literature: Hyperbilirubinemia (> 20 mg/dL total serum bilirubin, Shapiro, 2005), anoxia/hypoxia, family history of childhood hearing loss, prenatal/neonatal infections, and congenital neurological problems. The incidence of permanent childhood hearing loss is estimated to be between 2-3 of every 1,000 babies born in the United States. Children with AN/AD represent a subset of the overall number of children with hearing loss. Recent data suggest that the incidence of AN/AD in the general population may be 1-2 per 10,000 births (Foerst, et al. 2006). However, with OAEs being widely used as the primary screening method, and the fact that infants with AN/AD will pass OAEs, the exact incidence could be higher.

Does UNHS detect AN/AD?
If AN/AD is present at birth, detection depends on the type of screening protocol utilized. As previously discussed, children with AN/AD often have normal OAE results with a significantly abnormal ABR response. If newborn hearing screening consists only of OAE testing or if OAE is the first stage in the screening process, most children with AN/AD will not be identified. Hearing screening programs that utilize ABR will appropriately refer infants with AN/AD. Unfortunately, the cost of providing ABR screening to every infant born in the U.S. would increase the costs of UNHS substantially. The 2007 Joint Committee on Infant Hearing Position Statement recently recommended that infants who spend more than 5 days in the NICU should receive an ABR screening to improve identification of AN/AD. Therefore, many newborn hearing screening programs have elected to perform ABR screening on all babies with risk factors for AN/AD. Physicians should familiarize themselves with the type of screening tests that are utilized in local hospitals. Infants with risk factors for AN/AD or a history of residence in the NICU should be referred for ABR evaluation if one was not completed as part of the hearing screening.

What types of assessments and intervention should be provided?
Just as in all cases of childhood hearing loss, referral for early intervention services is critical as soon as a diagnosis is confirmed. Pediatric audiologists can provide diagnostic confirmation of hearing sensitivity and ongoing monitoring. Management of referrals to specialists is best coordinated through the medical home. Evaluations by an otolaryngologist, neurologist, geneticist, and ophthalmologist help to identify any additional medical conditions. Childhood AN/AD can result from a wide variety of etiologies. Because of this variability, children with AN/AD also have a wide range of responsiveness to sound. Some have behavioral hearing thresholds in the normal range, while others may have limited or no responsiveness to sound. Unfortunately, results from diagnostic OAE and ABR results will not predict a child’s behavioral auditory responsiveness, despite the importance of these measures.
in the diagnosis of AN/AD. Given the spectrum of possible behavioral outcomes, intervention strategies should be customized based on the needs of each child. No single type of intervention is likely to be effective for every child with AN/AD. Behavioral audiological evaluation should be completed as soon as it is developmentally appropriate, typically around 6 months of age. Behavioral audiometric results may not be obtainable until 12 months or later in children with concomitant developmental delays. Because hearing may fluctuate in children with AN/AD, follow-up audiological monitoring should ideally occur every three months until the child is three years old and every six months between three and five years.

Hearing aids
For children with AN/AD who have behavioral audiometric responses to sound in the mild to severe hearing loss range, a trial with appropriately fit hearing aids is an essential part of the early intervention process. Hearing aids provide audibility for speech sounds and improve access to language. Even with hearing aids, some children with AN/AD will not show improved sound awareness or progress in speech and language development. Research has indicated that approximately 50% of children with AN/AD will show improvement in speech recognition scores with hearing aids, while the remaining 50% show limited benefit (Rance, 2005). Clinical tools to predict which children with AN/AD will be successful with hearing aids are not currently available. Therefore, a trial with amplification is recommended as soon as reliable behavioral audiometric data can be obtained. Since the child may or may not derive benefit from hearing aids, loaner hearing aids or an extended trial period through the hearing aid manufacturer should be provided whenever possible. Parents should be advised that their child's responsiveness with hearing aids may or may not improve. Monitoring of speech and language development as well as auditory skills should be undertaken to help assess the impact of amplification.

Cochlear implants
For children with AN/AD who do not show improved responsiveness with hearing aids, research has suggested cochlear implants (CI) can improve auditory awareness and speech and language development for some children. Several studies have compared children with AN/AD who received cochlear implants to children with other types of sensorineural hearing loss who use hearing aids and have not found significant differences between the two groups. However, in some cases of AN/AD where the auditory nerve is either small or absent, cochlear implantation has not been as effective, and the results are much more variable (Bradley, et al. 2008). Based on these findings, cochlear implantation should be considered with caution in cases where hearing aids do not improve auditory responsiveness or result in improvement of speech and language skills. Parents should participate in extensive discussions with the ENT surgeon and audiologist to evaluate the risks and benefits of cochlear implantation for their child.

Other considerations
Children with AN/AD are a unique and heterogeneous group. While some can develop auditory and oral skills necessary for spoken language development, others may require sign language or other visual cues to support their progress, even in cases where hearing aid and cochlear implant use has been implemented. Supporting the child's development based on individual skills and abilities is critical. Because nearly all children with AN/AD demonstrate significant difficulty listening in background noise, additional hearing assistance technology may be appropriate in school or in specific listening situations.


NOTE: This article was prepared by Ryan McCreery from Boys Town National Research Hospital, Omaha.
Early Intervention

Children [with hearing loss] enrolled in early intervention within the first year of life have been shown to have language development within the normal range of development at 5 years of age.

(Joint Committee on Infant Hearing, 2007 Position Statement)
Physician Role in Early Intervention

According to the statement from the 2007 Joint Commission on Infant Hearing (JCIH), the goal of Early Hearing Detection and Intervention (EHDI) is to maximize linguistic competence and literacy development for children who are deaf or hard of hearing. Without appropriate opportunities to learn language, these children will fall behind their hearing peers in communication, cognition, reading, and social-emotional development. Such delays may result in lower educational and employment levels in adulthood.

The role of the primary care physician in early hearing detection and intervention cannot be underestimated. Below are some steps you should take:

• Ensure that all children in your practice have been screened prior to hospital discharge or prior to one month of age.

• Refer all babies who do not pass screening for a diagnostic audiology evaluation.

• Review risk factors and discuss hearing monitoring with parents.

• Refer babies to Early Intervention as soon as a hearing loss is suspected.

Early Intervention

Early intervention is provided to families of children, ages birth to age 3, who have disabilities or developmental delays. Services are designed to support families as they help their infant or toddler to reach his or her potential.

Early intervention in Texas is provided by Early Childhood Intervention (ECI), a division of the Texas Department of Assistive and Rehabilitative Services. For children who have a hearing loss, services are provided by a qualified Teacher of the Deaf through an agreement with the Texas Education Agency, Regional Day School Programs for the Deaf, and local school districts.

Who to Refer to ECI:

• Refer an infant who did not pass his/her follow-up screening and whose family may need help in finding or getting to an audiologist. An ECI service coordinator can assist a family with accessing diagnostic testing needed to rule out or confirm a hearing loss.

• Refer every infant who has a confirmed hearing loss. An ECI team, including a qualified Teacher of the Deaf, will perform an evaluation to determine what services will most benefit the child and family.

• Refer an infant who has a confirmed hearing loss and is receiving private services. ECI can assist the family with community resources and transition into a school program when the child turns three.

• Refer an infant whose family needs help finding a hearing aid.

How do I refer a child for ECI services?

ECI programs accept referrals for children with hearing loss from providers through direct telephone contact or fax. ECI programs provide services to families within defined geographical boundaries based upon the family’s residence.

You can refer a child by contacting any local ECI program in your community; the staff there will ensure the family will be contacted by the ECI program that serves their residential area.

OR

You can refer a child to a specific ECI program within the family’s residential area.

There are several methods to identify a family’s local ECI program.

• Call the DARS Inquiries Line at 1-800-628-5115.

• Send an e-mail to: DARSinquiries@dars.state.tx.us with the child’s zip code information. You will receive an email back with the contact information for the ECI program that serves the family’s residential area.

• Utilize the ECI website search page to find the child’s local ECI program at: www.dars.state.tx.us/ecis.
Navigating the Texas Early Intervention System

**Newborn Hearing Screening**
- All newborns screened for hearing loss
- Outpatient screening completed by or before 1 month of age

**Follow-up Hearing Screening**
- Diagnostic testing completed by audiolgist by or before 3 months of age
- Hearing Loss Confirmed

**Referral to Early Intervention by or before 6 months of age**
- Your family is referred to your local Early Childhood Intervention Program (ECI) within two days of identification of your child’s hearing loss.
- Your family is referred to your Local Education Agency (LEA)/Regional Day School Program for the Deaf (RDSPD)/Texas School for the Deaf (TSD) within 5 days of referral to ECI.

**Assessment and development of Individual Family Service Plan (IFSP) within 45 days of referral to ECI**
- Service Coordination/additional early intervention services provided by ECI
- Deaf Education Early Intervention Services provided by LEA/RDSPD/TSD

**ECI and local education agency’s Deaf Education Early Intervention Services (school district, RDSPD, TSD) are available, when determined by an IFSP, until your child is three years old. ECI and LEA will coordinate transition services before your child’s third birthday.**

**Audiological Monitoring**
- Hearing Aid Evaluation & Fitting
- Continued Monitoring
- Hearing Services and Hearing Aids are provided to eligible children through Medicaid and Children with Special Healthcare Needs

**Medical Home Monitoring**
- Pediatrician / Primary Care / ENT provider to coordinate medical care

**Important Note**
- Children whose hearing loss is identified later than infancy or who develop hearing loss after they are born are referred for early intervention as soon as the hearing loss is suspected or confirmed. It is important that children, especially those with certain high risk factors, be monitored for late-onset hearing loss.

Services for students who are deaf or hard of hearing and/or other special education services are available through your LEA/RDSPD/TSD from ages 3–21 when determined by an Individual Education Plan (IEP).

This page is designed to provide a visual overview of Texas’ comprehensive system of early intervention services for families of infants and toddlers with hearing loss. Texas Connect Topic Card #10 has more information on early intervention services.
Early Intervention Service System
for Texas Families with Children Who are Deaf or Hard of Hearing (Birth to Age 3)

Early intervention describes a wide range of services available to children, ages birth to three, who have disabilities or developmental delays. A federal law, the Individual with Disabilities Education Act (IDEA) Part C, mandates early intervention services and establishes the guidelines for eligibility and service delivery. Early intervention services may include home visits, family training, counseling, special instruction, and therapy. Early intervention services are designed to help families help their infant or toddler reach his or her potential.

Texas provides a comprehensive system of services for families of infants and toddlers who have a hearing loss. Two state agencies, the Department of Assistive and Rehabilitative Services Early Childhood Intervention (DARS-ECI) and the Texas Education Agency (TEA) have outlined a plan to ensure a full range of services is available throughout all areas of Texas.

DARS-ECI is the state’s lead agency for early childhood intervention (ECI) services and provides oversight to the local ECI programs that assist families. Local ECI programs work in partnership with local education agencies (school districts), Regional Day School Programs for the Deaf (RDSPDs), and the Texas School for the Deaf (TSD) so that specialized services, specific to children with hearing loss, are made available.

ECI coordinates a team of professionals, including a Teacher of the Deaf, to work with deaf and hard of hearing children and their families. Additional team members often include an audiologist, a speech and language pathologist, and other disciplines as appropriate. Together with the family, the team develops an Individual Family Service Plan (IFSP) designed to help babies maximize their developmental potential. The team also supports families as they explore questions and concerns regarding their child’s hearing, communication, and language development.

Transition Services
The IFSP team works with families before their child turns three to begin the transition to services with their local education agency or other service provider. Services for students who are deaf or hard of hearing are available through the local education agency, Regional Day School Programs for the Deaf, or Texas School for the Deaf from ages 3–22.

For information on Early Childhood Intervention Programs
http://www.dars.state.tx.us/ecis/index.shtml
DARS Inquiries Line 1-800-628-5115 | TDD/TTY 1-866-581-9328

For information on Deaf Education Early Intervention Programs
Region 10 Education Service Center
ESC State Leadership Office, Deaf and Hard of Hearing Services: Birth-5
972-348-1594 | http://www2.ednet10.net/SpecialEducation/DHHState0_5.html

Deaf Education Programs and Services

Regional Day School Program for the Deaf (RDSPD)
In order to provide services statewide, Texas operates a system of Regional Day School Programs for the Deaf (RDSPDs), serving students birth–22. Through a shared service agreement with local education agencies, RDSPDs provide early intervention services to families of children with diagnosed hearing loss—from birth to 36 months—in partnership with Early Childhood Intervention (ECI). Each specialized teacher, commonly known as a parent advisor, is a member of a child and family’s IFSP team and assists in helping families navigate through their appointments as they work towards a diagnosis and follow-up care. Drawing on their specialized training, the parent advisor supports families as they explore questions regarding their
child’s communication development by answering questions and providing information. In addition, the parent advisor helps families to access resources, including introduction to other families of children who are deaf or hard of hearing, individuals who are deaf or hard of hearing, and family support groups.

Contact the local Education Service Center (ESC) for more information about RDSPD programs and for services for students who are deaf or hard of hearing in your area.

**Education Service Center Directory:** [http://www.tea.state.tx.us/ESC](http://www.tea.state.tx.us/ESC)

**Education Service Center Support:** 512-463-9371

**RDSPDs for the Deaf Directory:** [http://ritter.tea.state.tx.us/deaf/rdspddirectory0809.pdf](http://ritter.tea.state.tx.us/deaf/rdspddirectory0809.pdf)

**Texas Education Agency, Division for IDEA Coordination:** [http://www.tea.state.tx.us/deaf](http://www.tea.state.tx.us/deaf)

**Local Education Agency (LEA)**

Local education agencies (school districts) may provide deaf education services to deaf or hard of hearing children. See below for more information about how these services are coordinated within a specific school district:

**School district locator:** [http://deleon.tea.state.ts.us/sdl/Forms/#](http://deleon.tea.state.ts.us/sdl/Forms/#)

**Education Service Centers’ Special Education Directors:**
[http://ritter.tea.state.tx.us/special.ed/escinfo/contact.html](http://ritter.tea.state.tx.us/special.ed/escinfo/contact.html)

**Texas School for the Deaf (TSD)**

Texas School for the Deaf is a public day and residential school for deaf and hard of hearing students who are preschool through the 12th grade or to age 22 if transitional services are needed. TSD is a day school program for children below the age of five. For students age 5 or above, Austin-area children may attend classes during the day and return home at night while students outside the Austin area may reside in the dorms during the week and return home each weekend.

**Birth to Age Three Services:**

Texas School for the Deaf also works jointly with local ECI programs to provide deaf education early intervention services to Austin-area children from birth to age three. These services include home visits/consultations related to maximizing developmental potential of children with hearing loss as well as early childhood classroom time for infants and toddlers. Parents whose children transition from birth to age three services with ECI may keep Texas School for the Deaf as their educational choice or choose placement in another local educational, Regional Day School Program for the Deaf, or private program.

Texas School for the Deaf can provide resources, support, and services for Texas families of children who are deaf or hard of hearing through the Educational Resource Center on Deafness (ERCOD). See “Statewide Outreach and Resources” on the following page.
Statewide Outreach and Resources

Region 10 Education Service Center (ESC-10)
ESC-10 provides leadership, staff development, consultation, and technical assistance via the statewide ESC Deaf and Hard of Hearing (ESC DHH) network. ESC-10 assists Regional Day School Programs for the Deaf and local school districts with the development and implementation of comprehensive early intervention services to families of infants and toddlers with hearing loss and instructional programs for preschool age students with hearing loss.

ESC-10 works to promote practices and protocols that support a comprehensive, seamless statewide system of coordinated services between newborn hearing screening programs, audiology and otolaryngology services, medical home services, local ECI programs, RDSPDs, and local school districts.

Region 10 Education Service Center
ESC State Leadership Office for Deaf & Hard of Hearing Services: Birth-5 | 972-348-1594
http://www2.ednet10.net/SpecialEducation/DHHState0_5.html

Educational Resource Center on Deafness (ERCOD)
ERCOD provides resource information and outreach programs to families across Texas with children who are deaf and hard of hearing and the professionals who serve them. ERCOD also provides assistance to parents to help them navigate the multiple private and public services and resources that are available in Texas.

1-800-DEAFTSD or 512-462-5329 (V/T) | Email ercod@tsd.state.tx.us | www.tsd.state.tx.us/outreach

Texas Department of State Health Services (DSHS)
The Texas Department of State Health Services, Newborn Hearing Screening provides information and resources to families and professionals related to newborn hearing screening protocols and follow-up care.

For more information about the Department of State Health Services, visit http://dshs.state.tx.us/audio
Did You Know?

ECI and Deaf Education Programs Collaborate to Provide Support and Services for Children with Hearing Loss

As a physician, families of children with a newly diagnosed or suspected hearing loss will look to you for guidance. Navigating the health care delivery system can be overwhelming for parents who are facing the possibility of a having a child with a special need.

Early Childhood Intervention (ECI) and deaf education programs recognize the importance of the link between the child's physician and other supports and services. We want to work with you to ensure families are connected to the supports and services they need. Here are some items we want you to know!

**Did you know...**
In Texas, ECI is the agency responsible for referring children with hearing loss to their local education agency. ECI coordinates a statewide system of services for families with children, birth to 36 months, who have developmental delays and disabilities.

**Did you know...**
Deaf education programs can be a resource to you by providing your office important information on newborn hearing screening, the importance of a diagnosis, the implication of early childhood hearing loss, and resources families can access in support of a diagnosis and care.

**Did you know...**
ECI and deaf education programs work together to provide early intervention services to children birth to 36 months with hearing loss. We coordinate our services to help babies develop speech, language and social skills and support families as they begin to explore questions regarding their child's communication and development.

**Did you know...**
It is appropriate for you to refer a family to ECI anytime a child has failed a hearing examination. Early referral and identification are critical to speech and language development. Through service coordination, ECI will help families navigate through their appointments as they work toward a diagnosis.

**Did you know...**
At our first contact with the family, ECI will begin to assemble a team of professionals to work with a child who has a hearing loss. The team may consist of an audiologist, a speech and language pathologist, and a teacher of the deaf and hard of hearing, and other appropriate disciplines.

**Did you know...**
As the child’s primary care provider, you are a part of the team along with the child’s parents. Together, the team decides which services and supports will be needed for the child.

**Did you know...**
An Individualized Family Service Plan (IFSP) is developed, and with parental consent, ECI will provide you with regular communication about the IFSP and the child's progress.

**Did you know...**
Even when families choose to receive services from other agencies or providers, service coordination is still available through ECI.

With your assistance, we want to ensure that every family is linked to community, state, and national resources, including parent support groups. It’s very important that families have access to these resources as they prepare to handle a newly diagnosed or suspected hearing loss diagnosis for their child.
Hearing loss is the most common congenital condition in the United States. Everyday 33 infants are born with some degree of hearing loss.

The following are local resources to assist you and your practice in supporting families who are concerned about their infant or toddler’s hearing status, or have confirmed that their infant or toddler has a diagnosed hearing loss of any degree.

Local Early Childhood Intervention (ECI) Program: Local Regional Day School Program for the Deaf (RDSPD)/Local School District Deaf Education Early Intervention Services:

Parent Support Organizations/Groups

Hands & Voices – Texas Chapter
www.txhandsandvoices.org
Austin Telephone: 512-470-4065

Texas Parent to Parent
Laura Warren, Executive Director
www.txp2p.org
Toll Free: 866-896-6001
Austin Telephone: 512-458-8600

For further information about ECI or to make a referral, please contact your local ECI Program or call the DARS Inquiries Line at 1-800-628-5115 or visit www.dars.state.tx.us/ecis.
Speech and Language Milestones

Speech and language in infants and toddlers occurs best in an environment that is filled with varied sights, sounds, and opportunities to listen and experience the speech and language of others. Recent research suggests there are critical periods for speech and language development. The most intensive period of speech and language development is during the first three years of life when the brain is developing. Early exposure to language is critical at the very beginning of a child’s life.

Milestones are identifiable skills that follow a natural progression and serve as a guide to highlight typical development. While children vary in their development, the following milestones can help doctors and other health professionals identify when a child may need additional support. Whenever there is a concern about a child’s speech and language development, it is appropriate to refer a child to their primary care provider for follow-up recommendations.

Language milestones for a child who is deaf or hard of hearing and who is learning to communicate using American Sign Language are listed in bold.

Birth to Three Months

- Startles to loud sounds, **looks around with alertness**
- Quiets or smiles when spoken to, **looks attentively at a person’s face**
- Seems to recognize familiar voices and quiets when crying, **is attracted to any human movement**
- Increases or decreases sucking behavior in response to sound
- Makes pleasure sounds (cooing)
- Cries differently for different needs
- Smiles when he/she sees Mom/Dad, **responds to smiling by smiling back**

Four to Six Months

- Moves eyes in direction of sounds, **likes to be held facing out to view that action**
- Responds to changes in tone of voice, especially Mom/Dad’s, **laughs when sees fingers approaching to tickle**
- Notices toys that make sounds, **is attracted to moving and colored objects**
- Pays attention to music, **turns eyes to flashing light**
- Babbling sounds more speech-like with many different sounds, including “p,” “b,” and “m”

- Vocalizes excitement and displeasure, **smiles and makes eye contact and laughs**
- Makes gurgling sounds when left alone and when playing with you, **plays with hands and fingers and enjoys hand plays**

Seven Months to One Year

- Enjoys games like peekaboo and pat-a-cake
- Turns and looks in direction of sounds, **turns toward vibrations, phone, door bell, music**
- Listens when spoken or **signed to**
- Shows understanding of words by appropriate behavior and gesture
- Recognizes words for common item like “cup,” “shoe,” or “juice”
- Begins to respond to verbal requests such as “come here” or “want more”
- Uses speech/signs or non-crying sounds to get and keep attention
- Imitates different speech sounds, **begins hand babbling with varied patterns**
- Says one or two words/signs (“bye-bye,” “dada,” “mama”)
- Uses single word sentences in speech or **signs (simple hand shapes, mine, more, milk, mommy)**
- **Points to self and things**

One to Two Years

- Points to a few body parts when asked via speech or sign, **points to self and things**
- Follows simple commands and understands simple questions ("Roll the ball," "Kiss the baby," "Where’s your shoe?")
- Listens to simple stories, songs and rhymes, **listens to ASL stories**
- Points to pictures in a book when named, **points and can sign some letters of the alphabet**
- Puts two words together in speech or **sign**

Early intervention programs need to provide families with access to skilled and experienced early intervention professional to facilitate communication and language development in the communication option chosen by the family.

(Joint Committee on Infant Hearing, 2007 Position Statement)
• Uses many different consonant sounds at the beginning of words
• **Uses expressive vocabulary** of 50+ words in sign but understands many more

**Two to Three Years**

• Understands differences in meaning in speech or **sign** (“stop-go,” “in-out,” “big-little”)
• Follows two-command requests (“Get the book and put it on the table.”)
• Uses two to three word “sentences” (speech or **sign**) to talk about and ask for things
• Ask questions beginning with what, where and when (speech or **sign**)
• Uses expressive vocabulary of 200+ words (speech or **sign**)
Texas State Resources

Educational Resources and Programs

Education Service Centers (ESCs)
Statewide leadership in addressing identified areas of need in special education services is provided through eleven functions and five projects directed by various ESCs. The primary responsibility of each ESC is to provide leadership, training, technical assistance, and the disseminate information throughout the state. The ESCs coordinating these statewide leadership functions and projects are responsible for the implementation of many of the state’s continuous improvement activities.

Though both functions and projects provide statewide leadership, ESC function leads establish and coordinate a 20-region network. This ensures ongoing communication among ESCs about state-level needs assessment processes and planning, as well as implementing and evaluating statewide activities. Project leadership focuses on a specific activity.

The following ESCs have statewide leadership responsibilities, in partnership with the Texas Education Agency, for students who are deaf or hard of hearing:

ESC Region 10
Contact: Alicia M. Favila, M.Ed.; Senior Consultant/State Coordinator;
ESC State Leadership Office for the Deaf and Hard of Hearing Services: Birth-5
Voice/TTY: 972-348-1594

ESC Region 11
Contact: Penny Morrison; Coordinator;
ESC State Leadership Office for the Deaf and Hard of Hearing Services: Communication Access
Voice/TTY: 817-740-7580

ESC Region 20
Contact: John Bond; Coordinator III;
ESC State Leadership Office for the Deaf and Hard of Hearing Services: Access to General Curriculum
Voice/TTY: 210-370-5418
Texas Education Agency (TEA)

Texas School for the Blind and Visually Impaired (TSBVI)
Address: 1100 W. 45th St. Austin, Texas 78756
Voice: 512-454-8631   TTY: 512-206-9451

TSBVI is a partner with independent school districts in Texas. Provides instructional and related services to students who are blind, deaf-blind, or visually impaired, including those with additional disabilities.

Texas School for the Deaf (TSD)
TSD is a public (tuition-free) day and residential school for deaf and hard of hearing students from all across Texas who are preschool through 12th grade or through age 21 if continued transitional services are needed. Additionally, the school works jointly with local Early Childhood Intervention (ECI) programs to provide services to Austin-area children from birth to age three. These services include home visits with consultation related to maximizing the developmental potential of children with hearing loss as well as early childhood classroom time for infants and toddlers. Parents whose children transition from birth to three services under IDEA Part C may keep TSD as their educational choice under IDEA Part B or choose placement in another program. Also, through the Educational Resource Center on Deafness (ERCOD), TSD can provide resources, support, and services to families of children 0–3 who do not live in the Austin area. See section on ERCOD for more information.

Educational Resource Center on Deafness (ERCOD)
Address: 1102 South Congress Avenue, Austin, Texas 78704
Voice/TTY: 1-800-DEAF-TSD or 512-462-5329

ERCOD serves families, school districts, and communities throughout Texas through outreach programs. Information/referral workshops, summer programs, distance learning, and interpreter training are some of the services provided for deaf and hard of hearing children, their families, and professional service providers.
State Agencies

Texas Department of Assistive and Rehabilitative Services (DARS)
Early Childhood Intervention
Address: 4900 North Lamar Blvd.
Austin, Texas 78751
Voice: 1-800-628-5115
TTY: 1-866-581-9328

Serves children, birth to 36 months, with disabilities or delays. Assists families in helping their children reach their potential through developmental services. Programs are located throughout the state. For children who have a hearing loss, ECI works in partnership with deaf education parent infant services provided through regional or local school programs or Texas School for the Deaf.

Texas Department of State Health Services (DSHS)
Texas Early Hearing Detection and Intervention (TEHDI Program)
Address: 1100 West 49th Street
Austin, Texas 78756
Voice: 1-800-252-8023

Texas Early Hearing Detection and Intervention (TEHDI) Program is dedicated to ensuring all babies born in Texas will receive Newborn Hearing Screening and appropriate follow-up care. It is essential infants with hearing loss be identified early and appropriate intervention services be initiated as early as possible. Without early identification and intervention, children with hearing loss may experience delays in the development of language, cognitive, and social skills that may prevent success in academic, occupational, and personal achievements.

Texas Department of Aging and Disability (DADS)
Address: 701 West 51st Street
Austin, Texas 78757
Voice: 1-877-438-5658

The agency offers a waiver called the Deaf Blind with Multiple Disabilities (DBMD) program. The DBMD Program provides services and supports for people with deafblindness and one or more other disabilities as an alternative to living in an ICF/MR. Recipients may reside in their own or family home or in small group homes. Services include adaptive aids and medical supplies, dental services, assisted living, behavioral support services, case management, chore services, minor home modifications, residential habilitation, intervener, nursing services, occupational therapy, physical therapy, orientation and mobility, respite, speech, hearing and language therapy, supported employment, employment assistance, dietary services, financial management services for the consumer directed services option, and transition assistance services. The CDS option is available for residential habilitation, respite, and intervener services.

Organizations

HEAR ME Foundation
Address:
4814 Woodstream Village Dr.,
Kingwood, Texas 77345
Voice: 281-359-6725

A non-profit organization that offers support and empowerment through public awareness and educational program assistance to families who have chosen an oral communication option. Provides a weekend camp in June for the entire family. Offers opportunities for children to have a mentor.
Texas Association of the Deaf (TAD)  
Address: P.O. Box 1982  
Manchaca, Texas 78652  
www.deaftexas.org/wp  

A non-profit, membership based organization. Provides information and education, including surveys and studies, on various issues affecting the lives of those who are deaf or hard of hearing at all levels of the community.

Texas Association of Parents and Educators of the Deaf (TAPED)  
Address: 1307 Memorial Dr.  
Bryan, Texas 77802  
Connie Ferguson, Brazos Valley Regional Day School for the Deaf  
Voice: 979-209-2890  
www.taped.org/  

TAPED is an information and networking resource for parents and educators. Offers a website for educators and parents and is a voice for the field of deaf education.

Texas Chapter of Alexander Graham Bell Association for the Deaf and Hard of Hearing  
Address: 103 Tuleta Drive  
San Antonio, Texas 78212  
Voice: 210-495-0398  

Texas AG Bell provides both services and support for those with a hearing loss through a biannual conference, newsletter for its members, directory of services, and an opportunity for scholarships and awards.

Texas Chapter of the Hearing Loss Association of America  
Address: 1402 Saint Mary’s Lane  
Houston, Texas 77079  
Voice: 281-497-2670  

This organization promotes an improved quality of life for people who are hard of hearing through education, advocacy, and self-help.

Texas Hands and Voices  
Email: TXHandsandVoices@gmail.com  
www.txhandsandvoices.org  

Texas Hands and Voices is a state chapter of a nationwide non-profit organization (Hands and Voices). This is a parent driven organization that supports families and their children who are deaf or hard of hearing with a focus on being unbiased toward communication modes and methods.

Texas Parent to Parent (TxP2P)  
Address: 3710 Cedar Street  
Austin, Texas 78705  
Voice: 512-458-8600  
Toll Free: 866-896-6001  

TxP2P is a non-profit statewide program providing support, resources, and information to parents/caregivers of children of all ages and disabilities, as well as professionals who work with them. Services include matching parents to trained parent volunteers who have children with similar disabilities or issues. Also offers quarterly newsletter, listservs, annual parent conference, and educational opportunities across the state. Services in Spanish and English.

Texas Speech Language Hearing Association (TSHA)  
Address: 918 Congress Avenue  
Austin, Texas 78701  
Voice: 512-494-1127  
Toll Free: 1-888-729-8742  

Professional and scientific association comprised of speech-language pathologists, audiologists, associates, and students. Provides materials, literature, and workshops.

Services Activities

Advocacy Incorporated/Disability Rights Texas  
Address: 7800 Shoal Creek Blvd.  
Austin, Texas 78757-1024  
Voice/TTY: 512-454-4816  

Advocacy Inc. is a non-profit corporation that advocates for the legal rights of Texans of all ages with disabilities. It provides information for parents on federal and state law to help parents work as equal partners with the school in planning their children’s educational program.

Callier Center For Communication Disorders, The University of Texas at Dallas  
Address: 1966 Inwood Rd.  
Dallas, Texas 75235  
Voice: 214-905-3000  
TDD: 214-905-3012  

Callier Center is a non-profit educational, clinical, and research center for individuals with communication disorders of all types. Clinical services specific to hearing loss include complete audiological testing, amplification services, aural rehabilitation classes, and cochlear implant evaluation. Educational programs include specialized services for children with hearing loss, ages 2-5, within an early childhood preschool.

Partners Resource Network Inc.  
Address: 1090 Longfellow Drive  
Beaumont, Texas 77706  
Voice/TTY: 1-800-866-4762  
(Texas parents only)  

Non-profit that operates statewide network of federally funded Parent Training and Information Centers (PTIs), which provide training and technical assistance.
to families who have children with all types of disabilities, including the rights of students to a free appropriate public education.

**Camps**

Camp SIGN, DARS-Deaf and Hard of Hearing Services  
Address: P.O. Box 12306  
Austin, Texas 78711  
Voice: 512-407-3250  
TTY: 512-407-3251  
Ages: 8–17 years old

Camp SIGN is a communication barrier-free environment for students who are deaf or hard of hearing. The goal is to have all students from around the state, regardless of their communication mode, participate in the program and to create an environment of acceptance and encouragement.

Cochlear Implants Summer Listening Camp  
Address: 1966 Inwood Rd.  
Dallas, Texas 75235  
Voice: 214-905-3139  
Ages: 3–11 years old

Provides a one-week day camp for children with cochlear implants. Each camper is paired with a graduate student buddy. Camp activities focus on listening and spoken language activities.

Texas School for the Deaf (TSD)  
Address: 1102 South Congress Ave.  
Austin, Texas 78704  
Voice/TTY: 512-462-5329  
Ages: Infant through high school

TSD is a public (tuition-free) day and residential school for deaf and hard of hearing students from all across Texas who are preschool through 12th grade or through age 21 if continued transitional services are needed. Additionally, the school works jointly with local Early Childhood Intervention (ECI) programs to provide services to Austin-area children from birth to age three. These services include home visits with consultation related to maximizing the developmental potential of children with hearing loss as well as early childhood classroom time for infants and toddlers. Parents whose children transition from birth to three services under IDEA Part C may keep TSD as their educational choice under IDEA Part B or choose placement in another program. Also, through the Educational Resource Center on Deafness (ERCOD), TSD can provide resources, support, and services to families of children 0–3 who do not live in the Austin area. See section on ERCOD for more information.

**Funding Sources**

Supplemental Security Income (SSI)  
Toll Free: 1-800-772-1213  
www.ssa.gov

Provides Medicaid coverage and monthly cash assistance to eligible individuals. Eligibility is based on citizenship, financial status, and disability.

Caring for Children Foundation  
Toll Free: 1-800-258-5437  
E-mail: Resha_Wafer@bcbstx.com  
www.carevan.org

Provides access to preventative care for children ages 6–18 whose families are not eligible for Medicaid and cannot afford private insurance.

CHIP and Children’s Medicaid  
Toll Free: 1-877-KIDS-NOW  
TTY: 1-800-735-2988  
www.chipmedicaid.org

Texas families with uninsured children may be eligible for health insurance through Children’s Medicaid and CHIP (Children’s Health Insurance Program). Both programs provide a wide range of benefits, including regular check-ups and dental care to keep kids healthy.
Web Resources for Providers and Families

Alexander Graham Bell Association for the Deaf
www.agbell.org

American Academy of Audiology
www.audiology.org

American Academy of Pediatrics
www.aap.org

American Sign Language
www.signmedia.com

American Society for Deaf Children
www.deafchildren.org

American Speech-Language-Hearing Association
www.asha.org

Auditory-Verbal International, Inc.
www.auditoryverbal.org

Baby Hearing
www.babyhearing.org

Better Hearing Institute
www.betterhearing.org

Boys Town National Research Hospital
www.boystownhospital.org

Callier Center for Communication Disorders, The University of Texas at Dallas
www.callier.utdallas.edu

Centers for Disease Control and Prevention, Early Hearing Detection and Intervention
www.cdc.gov/ncbddd/ehdi

Cochlear Implants: Navigating a Forest of Information
clerccenter.gallaudet.edu/kidsworldDeafNet/e-docs/CI/index.html

Deaf.com
www.deaf.com

Deaf Linx
www.deaflinx.com

Deaf.com
www.deaf.com

Family Village
www.familyvillage.wisc.edu/lib_deaf.htm

Family Voices
www.familyvoices.org

Hands & Voices
www.handsandvoices.org

Hand Speak
www.handspeak.com

Kidsource
www.kidsource.com/nfpa/social.html

John Tracy Clinic
www.johntracyclinic.org

Joint Committee on Infant Hearing
www.jcih.org

League for the Hard of Hearing
www.lhh.org

Marion Downs National Center for Infant Hearing
www.colorado.edu/slhs/mdnc

National Association for the Deaf
www.nad.org

National Center for Hearing Assessment and Management
www.infanthearing.org

National Cued Speech Association
www.cuedspeech.org

Laurent Clerc National Deaf Education Center National Institutes of Health, National Institute on Deafness and Other Communication Disorders
clerccenter.gallaudet.edu/InfoToGo/

Oral Deaf Education
www.oraldeafed.org

Parenting Deaf and Hard of Hearing Children
deafness.about.com/od/growingupdeafhoh/Education_and_Parenting.htm

Raising Deaf Kids
www.raisingdeafkids.org

Telecommunications for the Deaf, Inc.
www.tdi-online.org
Provider Access

The Texas Early Hearing Detection and Intervention (TEHDI) program is pleased to offer Provider Access to you as one of the many services for our providers. As the oversight agency for the TEHDI program, the Texas Department of State Health Services provides this tool to you at no cost. The site is your gateway to comprehensive hearing health records for each of your patients and can assist you in coordinating care to those babies needing further intervention.

Once you receive your user name and password, you will be instructed to complete the Provider Registration page and will be directed to the Medical Home Physician Workflow. Below is the depiction of the Workflow you will see once accessing your account.

Medical Home Physician Workflow

Physician Workflow

The first screen you see after logging in to your account.

Patients I Have Not Reviewed

Displays a list of patients whose records have not yet been reviewed by you, the assigned primary care provider.
Patients Who Need Follow-Up

Displays those patients whose birth screening status is unilateral refer, bilateral refer, missed, moved out of state, or lost contact.

Find a Patient Not On My List

Allows you to search for a patient who is referred to you.
Manage My Account

Allows you to review and edit your username, password, security questions and office email address.

- The Department of State Health Services values your input and hopes you will advise us about what you need to better care for your patients.

- For all questions, comments and concerns, feel free to contact Mary Gwyn Allen at 512-458-7111 ext. 7726 or by email at marygwyn.allen@dshs.state.tx.us.

- For technical questions, contact TEHD1 Customer Support (Help Desk) at 1-866-427-5768 Opt. 2 or by email at ozhelp@oz-systems.com.

- For assistance online, go to https://www.provideraccess.tehdi.com and select Help.
Early Hearing Detection and Intervention (EHDI)
for Infants and Young Children

Just in Time: Resources for Primary Care Providers