ARIZONA DEPARTMENT OF HEALTH SERVICES
BUREAU OF STATE LABORATORY SERVICES

Arizona Newborn Screening Program

GUIDELINES

DIVISION OF PUBLIC HEALTH SERVICES
AUGUST 2010 with revisions (January 2011)

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CHAPTER 1
Introduction to the Arizona Newborn Screening Program

1.1 Overview

For most types of genetic or metabolic disease, early diagnosis and treatment are critical. Although babies born with these disorders may appear to be normal at birth, with time the disorder may have a devastating or lethal effect on the infant's health and development. Early screening, detection and treatment of these disorders can, in many cases, result in normal growth and development.

Arizona has been conducting widespread newborn screening for certain disorders since 1979. A laboratory in Colorado originally performed screening tests for Arizona newborns.

In 1993, the Arizona Legislature enacted legislation requiring the Arizona Department of Health Services to develop and administer a formal Newborn Screening Program. The Arizona Newborn Screening Program was created and is now housed in the Bureau of State Laboratory Services, in the Division of Public Health Services. The Program has enabled the Department to enhance its public health role in the newborn screening process, obtain more timely results from screening, and promote better follow-up for suspected or confirmed cases.

In 2006 the Newborn Screening Program expanded its panel of screened disorders to twenty-seven disorders (27) with the addition of tandem mass spectrometry testing and added follow-up for hearing screening to follow-up done for bloodspot disorders. Cystic fibrosis screening was added in October, 2007, completing the panel of twenty-nine disorders.

The panel of twenty-nine screened disorders (including hearing loss) was recommended by the U.S Department of Health and Human Services (HHS) Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) as the original Uniform Screening Panel. In May, 2010 the Secretary added severe combined immunodeficiency (SCID) to the panel but Arizona does not yet screen for SCID.

The Advisory Committee to the Arizona Newborn Screening Program endorsed the original panel and Rules were written to allow the program to adopt this list of disorders. These disorders were selected because their detection and treatment in the newborn period is effective in preventing severe morbidity or mortality and a reliable, reasonably priced test can be used for screening. The disorders currently on the screening panel are:

Endocrine Disorders (2)
- Congenital Hypothyroidism
- Congenital Adrenal Hyperplasia

Hemoglobin Disorders (3)
- Sickle Cell Anemia
- Sickle Beta Thalassemia
- Sickle C Disease

Other Enzyme Deficiencies (2)
- Biotinidase Deficiency
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Amino Acid Disorders (6)
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- Maple Syrup Urine Disease
- Homocystinuria
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Fatty Acid Oxidation Disorders (5)
- Carnitine Uptake Defect
- Medium-chain Acyl-CoA Dehydrogenase Deficiency
- Very long-chain Acyl-CoA Dehydrogenase Deficiency
- Long-chain 3-OH Acyl-CoA Dehydrogenase Deficiency
- Trifunctional Protein Deficiency

Organic Acid Disorders (9)
- Isovaleric Acidemia
- Glutaric Acidemia Type 1
- 3-Hydroxy-3-methylglutaric Aciduria
- 3-Methylcrotonyl-CoA Carboxylase Deficiency
- Multiple Carboxylase Deficiency
- Methylmalonic Acidemia - mutase
- Methylmalonic Acidemia - cobalamin defects
- Propionic Acidemia
- Beta-Ketothiolase Deficiency

Pulmonary Disorder (1)
- Cystic Fibrosis

Disorder not detected by bloodspot screening (1)
- Hearing Loss (screening done elsewhere but results of screening reported to the program and follow-up done)

For most of these disorders, the incidence in the population is rare, but the potential for devastating consequences and the high costs of treating undiagnosed infants who do have the disorders is thought to justify mass screening. Hearing loss is the most common, occurring in approximately 2 – 4 per 1000 births.

The Arizona State Laboratory is now designated as the sole testing facility for the program. Fees charged for the testing are used to support the operation of the Newborn Screening Program.

1.11 Program Goals

The goals of the Arizona Newborn Screening Program are:

1. To identify newborns with certain, rare disorders and assist families of affected infants so that they receive appropriate and timely treatment to prevent or delay serious medical problems.

2. To identify possible hearing loss before one month of age and link families with appropriate assessment and intervention.

3. To ensure that all newborns referred for a follow-up hearing screen prior to discharge return for an outpatient screen and receive appropriate assessment by three months of age and intervention by six months of age.

1.12 Key Roles

The success of the Newborn Screening Program depends upon the coordinated efforts of many health care professionals:

- Medical Home and/or other Healthcare Professionals

Medical Home and/or other Healthcare Professionals are generally responsible for: ordering the screening tests for newborn infants in their care, informing parents about the screening tests, and collection and handling of newborn screening specimens. Practitioners, and/or their contracted laboratories, may collect and send specimens for testing. Practitioners, hospitals and laboratories work together to coordinate timely collection and rapid delivery of acceptable newborn screening specimens to the Arizona Public Health Laboratory (State Lab).

Providers are encouraged to refer to the test as Newborn Screening, not the PKU test. Although PKU was the first and for a while the only disorder screened, it is now only one of the 29 disorders on the Arizona panel.

- Screening Laboratory

  As per ARS §36-694, the sole screening laboratory is the Arizona Public Health Laboratory (State Lab). The State Lab receives all Arizona newborn screening specimens for testing of twenty-eight bloodspot disorders. Staff there review the specimens for acceptability, perform laboratory testing, keep records of the tests performed, mail results to submitters and physicians and conduct quality control studies of laboratory methods and practices.

- Arizona Department of Health Services - Newborn Screening Program

  As per ARS §36-694, the Department is responsible for the overall development, implementation, management and evaluation of the Newborn Screening Program. The Office Chief for Newborn Screening is responsible for the administration and coordination of program operations. In addition, the Chief coordinates resource networks for the specialized treatment and medical management of identified newborn screening disorders, as well as other program consultants and contractors. The Department oversees the collection of revenues from the screening tests.

  The Office Chief also administers the Follow-up component of the Newborn Screening Program. The Follow-up group has many responsibilities, including, but not limited to: reporting of positive bloodspot results to physicians and parents, tracking infants with positive results to help them get diagnostic testing and appropriate care, monitoring confirmatory testing, and reporting of confirmed cases to national newborn screening databases. They also receive reports of newborn hearing screening and track infants who do not pass hospital screening tests in order to assist families to get further testing and early intervention services when a hearing loss is confirmed.

- Contracted Specialists

  The Office of Newborn Screening works with contracted endocrinologists, metabolic geneticists, hematologists, pulmonologists (specializing in the treatment of cystic fibrosis) and audiologists. On behalf of the Office of Newborn Screening, these contracted specialists provide consultation to primary care physicians of newborns or infants with possible disorders or hearing loss. The purpose of consultation is to assure that the medical home checks on the clinical status of the baby, reviews signs and symptoms indicative of a possible disorder or hearing loss, reviews family history, recommends further testing and, if necessary, makes a referral to a specialist for diagnostic evaluation.

When newborn screening results are highly abnormal and the attempts to contact a medical home are unsuccessful, the contracted specialist may contact the family on behalf of the Newborn Screening Office in order to explain the results to the baby's parents, verify the clinical status of the baby and advise parents to schedule an appointment with a primary care physician immediately.

Contracted specialists also provide consultation services to the Newborn Screening Program. These services include case review, technical and program support, provision of technical information and recommendations regarding diagnostic testing methodologies, review of educational materials, and attendance at critical meetings where program issues are discussed.

1.2 Key Points about Newborn Screening for Discussions with Parents

1. **Every baby is at risk and you can't tell if a baby has a disorder without screening**
   Newborn screening is important for all babies because even without a family history of any of the screened disorders, every baby is at risk. Most babies with a disorder are born into a family with no other affected family members. Most affected babies look healthy at birth.

2. **Disorders are serious and can be life-threatening**
   The screened disorders are serious and can cause severe disability or even death if not detected and treated early. For some disorders, symptoms appear quickly and for others the baby appears fine for a while before symptoms appear and by that time permanent damage has been done.

   Newborn screening is not just a PKU test anymore but includes hearing screening and a panel of 28 disorders detected by bloodspot testing.

3. **How the testing will be done**
   A few large drops of blood are obtained by pricking the baby's heel. These blood drops are collected on a special filter paper kit, dried, and sent to the State Lab for analysis.

   Hearing screening is done when the baby is quiet or asleep by a trained screener using equipment that provides an automated pass or refer result – sound is introduced into the baby's ear and either an otoacoustic emission is measured or an auditory brainstem response is measured from scalp electrodes. This testing is painless and takes only a few minutes to complete.

4. **Testing must be timely**
   The first bloodspot test should be done between 24 and 36 hours of age or prior to discharge from the hospital. For some disorders, false negative results can occur with later testing. The second screen should be done at the

first outpatient visit to the medical home or other healthcare professional or between 5 and 10 days of age, whichever comes first.

The hearing test will be done before the baby is discharged from the hospital. Any rescreening should be done within two weeks and diagnostic testing should be done as soon as possible following a failed outpatient screen. Completing diagnostic testing before three months of age ensures that testing can be done without sedation or anesthesia and that early intervention and fitting of hearing aids, if appropriate, can take place before six months of age. Babies that are in the neonatal intensive care unit for more than five days should go directly to a diagnostic evaluation if they fail the inpatient screen.

5. **Results of screening tests are reported to the provider who ordered the test but parents will be contacted if there is an abnormal result**

   Parents will not be contacted by the Newborn Screening Program if their baby's results are normal. Providers who order the screening will receive mailed results and providers can request copies of tests ordered by others when they begin care for a baby. All newborn screening results should be documented in the baby's medical record.

   If results are highly abnormal, parents will get a call from their baby's doctor or from the Program with instructions for immediate diagnostic testing. For borderline results, they will usually be contacted by their doctor and will get a letter from the Program. If a specimen can't be tested (an unsatisfactory or "unsat" specimen) or if results show a hemoglobin trait, parents will receive a letter from the Program asking them to contact their baby's doctor for further information.

   Screening tests are not diagnostic and abnormal results will occur for babies that do not have a disorder (false positive results). Further testing will be needed promptly. So that parents can be notified, it is important for them to submit a current address and phone number on the specimen card. It is recommended that a second contact phone number also be included on the collection card.

   Results of the hearing screening are recorded on the back of the baby's immunization card. If the baby does not pass, a repeat test will usually be scheduled at the hospital where the baby was born. The Program does not start follow-up until the baby is a month of age and has not yet passed a hearing screening.

6. **Where parents can get further information**

   Hospitals or other health care professionals should give a copy of the brochure, "Newborn Screening, A Guide for Parents," to parents before the

first newborn screening test is ordered. Hearing screeners should also have a supply of the brochure, "Universal Newborn Hearing Screening," to share with parents at the time of their baby's hearing test.

Parents are encouraged to visit the Arizona Newborn Screening Program's website at **www.AZNewborn.com** where there is more information about the program and links to many other sites with newborn screening information.

1.3 Arizona Newborn Screening History

1.31 Bloodspot Screening History

Screening for phenylketonuria (PKU) began in Arizona in the late 60's and early 70's.

In 1979 a three-year federal grant from the Office of Maternal and Child Health was awarded to the Colorado State Laboratory to establish a regional newborn screening laboratory. The testing panel was composed of six disorders.

In 1989 a test for biotinidase deficiency was added to the panel of tests.

In 1993 legislation was passed establishing a Newborn Screening Program in Arizona funded by fees collected for screening. On November 1, 1994 the Arizona Public Health Laboratory began screening for the seven disorders on the Colorado panel and also providing follow-up for abnormal results. A second screen was encouraged but not mandated.

In 2001 legislation was passed that increased the screening fees and added congenital adrenal hyperplasia (CAH) to the newborn screening panel. Screening for CAH began in February, 2002. Second screens were now required for all babies.

In 2005 legislation again was passed that increased the screening fees and allowed the expansion of the panel to include the 29 disorders in the Uniform Screening Panel recommended by the federal Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC).

Between July and September, 2006, newborn screening for 20 disorders (fatty acid oxidation disorders, aminoacidopathies, and organic acidemias) was started using tandem mass spectrometry (MS/MS) and accompanied the testing by other methods of seven other disorders already on the panel. Follow-up for hearing loss began as well.

In October, 2007, testing for cystic fibrosis began and Arizona was now screening and doing follow-up for the complete original Uniform Screening Panel.

1.32 Hearing Screening History

Newborn Hearing Screening in Arizona began with the Never Too Young (NTY) program in 1985. NTY supported screening children with risk factors for hearing loss. By 1993 it was recognized that half of the babies born with hearing loss had no risk factors and the national move towards universal newborn hearing screening began. New technology made it faster and easier to screen all babies and new research was showing that age of identification of hearing loss was a significant factor in development of speech and language for children with hearing loss. The NTY evolved into the Arizona Early Hearing Detection and Intervention program (AzEHDI). By 2000 all birthing hospitals in Arizona were screening for hearing loss. In 2005 the current law was passed creating the follow-up program. The goals of the program within the Department of Health Services were to:

- Include parents and professionals on an advisory panel
- Educate parents, health care professionals and the public about early hearing detection and intervention
- Mandate reporting of screening and diagnostic testing
- Institute a centralized system of tracking and follow up to support parents in connecting with screening, diagnostic services and early intervention

Today 98% of all babies born in Arizona are screened for hearing loss by one month of age and 97% of babies are screened nationally. The average age of identification of hearing loss was two years of age. Now, with the goals of AzEHDI in place, the average is less than six months of age. New generations of children who are Deaf and Hard of Hearing have a greater opportunity to meet their full potential.

1.4 Statute and Rules

1.41 Arizona Statutes and Authority

The statute establishing the Newborn Screening Program, designating the State Lab as the only testing facility of the program and requiring hearing tests to be reported to the Program is found in the Appendices or in: ARS § 36-694

The statute establishing the newborn screening program fund is found in: ARS § 36-694.01

The statute regarding insurance coverage of medical foods is found in: ARS § 20-2327

1.42 Arizona Administrative Code

Rules regarding the Arizona Newborn Screening Program are in the Appendices and also available online at the Arizona Secretary of State's web site: (http://www.azsos.gov/PUBLIC_SERVICES/Title_09/9-13.htm).

For a discussion of provider responsibilities under these rules, please see Chapter 3.1 on page 33 and Chapter 5.4 on page 66.

1.5 Newborn Screening Advisory Committee

The Newborn Screening Advisory Committee is established by statute (ARS 36-694) to provide recommendations and advice to the Department of Health Services regarding tests that should be included in the Newborn Screening panel.

In 2006 the committee recommended the 29 disorders, including hearing loss, of the original core panel of the Uniform Screening Panel from the HHS Secretary's Advisory Committee on Heritable Disorders in Newborns and Children.

Any recommendation of a test to be added to the panel must be accompanied by a cost-benefit analysis.

The committee is chaired by the Director of the Department of Health Services and meets annually.

The Director appoints the members of the committee to include:

- Seven physicians representing the medical specialties of endocrinology, pediatrics, neonatology, family practice, otology and obstetrics;
- A neonatal nurse practitioner;
- An audiologist;
- A representative of an agency that provides services under part C of the Individuals with Disabilities Education Act;
- At least one parent of a child with a hearing loss or a congenital disorder;
- A representative from the insurance industry familiar with health care reimbursement issues;
- The Director of the Arizona Health Care Cost Containment System (AHCCCS) or the director's designee and
- A representative of the hospital or health care industry.

1.6 Privacy and the Health Insurance Portability and Accountability Act (HIPAA)

HIPAA became Federal law in April of 2003. Its goal is to "provide patients with access to their medical records and more control over how their personal health information is used and disclosed." (Source: DHHS, available at http://www.hhs.gov/news/facts/privacy.html)

The Arizona Department of Health Services (ADHS) is considered to be a "public health authority" as defined in the act and as such is granted access to protected health information (PHI) when necessary to prevent or control disease or where required by law (federal, tribal, state or local).

Each employee at ADHS is trained in HIPAA policies and procedures and any information obtained will be kept confidential.

As part of a HIPAA-exempt public health entity, Newborn Screening is able to lawfully receive or disclose confidential information used in the follow-up process without obtaining consent from parents.

Photocopies of the HIPAA exemption may be sent to you as explanation. Please see the Code of Federal Regulations (45 CFR 164.512) or contact the Program with any questions.

See http://www.cdc.gov/privacyrule/Guidance/PRPH.htm for further discussion of public health disclosures.

1.7 Program Fees and Payment Procedures

1.71 Testing Fees

Arizona's Newborn Screening test consists of two required screens. The first screen is billed at $30.00 each and the second screen is billed at $40.00 each. Fees support the operation of the Newborn Screening Program, including follow-up for bloodspot disorders and hearing loss.

1.72 First Screen

The first bloodspot screen is billed to and paid for by the submitter (generally a hospital, birthing center or midwife) who sends the first specimen for a baby.

1.73 Second Screen

The second bloodspot screen may be billed to and paid for by a hospital, midwife, insurance company or other responsible party (parent or guardian of a newborn).

If a baby is still a hospital inpatient at the time of collection of a second screen, the billing will be to the hospital. If a midwife collects a screen from a baby five or more days old, it will be considered to be a second screen and will be billed at the second screen rate.

Please submit the following insurance information (or a notation that the family does not have insurance) with all second screens which are not billed to a hospital or midwife:

- Policy holder's name (as shown on the insurance card)
- Policy holder's date of birth
- Policy holder's gender
- Policy holder's employer and employer's address
- Policy holder's address if different from mother's address
- Member ID#
- Group ID#
- Insurance company's name
- Insurance company's billing address
- Insurance company's phone number
- Notation of whether the insurance is primary or secondary

Please do not attach insurance information to the bloodspot card with staples. Place the insurance information with the cards or use paperclips, but these are not necessary.

If this information is not received, staff from the Program will contact physicians and facilities to obtain the information.

1.74 Recall Screen

After the second screen, a recall test for a specific disorder may be requested in response to an abnormal or borderline result. There is no fee for this test. A supplementary specimen kit should be used to collect the specimen and the Recall box above the Baby's Name field should be marked and the disorder being retested should be written on the line next to the box. Only a test for the requested disorder will be reported. Otherwise, if the specimen is not marked as a recall, it will be considered a billable specimen.

1.75 Insurance Coverage

Practitioners should encourage parents to add their newborns to their insurance as soon as possible after the birth since insurance payments cannot be made on behalf of infants until they are listed as insured.

Complete insurance information as listed above (in Chapter 1.73) should be submitted with second screens.

1.76 Payment to the Department

The Department will bill for first and second screens at least monthly. Payments are due to the Department within 30 days of the billing date.

Payments should be sent to:

Arizona Department of Health Services
Newborn Screening Program
PO Box 25046
Phoenix, AZ 85002-5046

1.8 Program Contacts

1.81 Office of Newborn Screening

Main Phone: (602) 364-1409 or (800) 548-8381 within Arizona
Fax: (602) 364-1495

Ward Jacox, Office Chief
(602) 364-1410
Ward.Jacox@azdhs.gov

Sundin Applegate, MD, NBS Medical Director
(602) 542-2950
Sundin.Applegate@azdhs.gov

Sondi Aponte, Quality Improvement Manager
(602) 364-1642
Sondi.Aponte@azdhs.gov

Brigitte Dufour, Case Management Coordinator
(602) 364-1406
Brigitte.Dufour@azdhs.gov

Wendie Jenkins, Educator
(602) 364-1407
Wendie.Jenkins@azdhs.gov

Christopher Rogers, Laboratory Manager
(602) 542-1184
Christopher.Rogers@azdhs.gov

Yvonne Carroll, Demographics Supervisor
(602) 364-1468
Yvonne.Carroll@azdhs.gov

Cheryl Gillman, Billing Manager
(602) 542-2520
Cheryl.Gillman@azdhs.gov

To obtain a submitter code (for new providers): (602) 364-1468

Billing or Insurance questions: (602) 542-0897 or (602) 542-2520

Requesting copies of lab results, fax to (602) 542-4099
or call (602) 364-3190
(Please include the baby's name, date of birth, mother's name and her date of birth with your request.)

Ordering specimen cards and envelopes:
call Lab Receiving at 542-1190
or E-mail an order form to labreceiving@azdhs.gov
or fax an order form to (602) 364-0758

Mailing address for submitting bloodspot specimens:
State Laboratory, Receiving
c/o Newborn Screening
250 N. 17th Avenue
Phoenix, AZ 85007-3231

1.82 Pediatric Specialists and Program Consultants

Endocrinology

Roger E. Johnsonbaugh, MD
Arizona Pediatric Endocrinology
(602) 274-5078

Edward I. Holland, MD
East Valley Pediatric Endocrinology
(480) 464-8600

Donnie P. Wilson, MD
Phoenix Children's Hospital, Endocrinology Dept.
(602) 546-0935

Alvin H. Perelman, MD
Southwest Pediatric Endocrinology
(480) 323-4800

Mark D. Wheeler, MD
University Physicians – Angel Wing Clinic
(520) 626-6077

Hematology

Jesse D. Cohen, MD
Pediatric Hematology-Oncology Associates
(602) 253-5993

Brenda J. Wittman, MD
University Medical Center
(520) 626-4851

Metabolic Genetics

Kirk A. Aleck, MD
R. Stephen Amato, MD, PhD
CHC Phoenix Genetics Program
St. Joseph's Hospital and Medical Center
(602) 406-3611

Melanie Colville, MS, RD, Metabolic Dietitian
(602) 406-4383

Candice Candelaria, MS, RD, Metabolic Dietitian
(602) 406-7346

Cystic Fibrosis Centers

Peggy J. Radford, MD
Pulmonology, Phoenix Children's Hospital
(602) 546-0985

Wayne J. Morgan, MD
Arizona Respiratory Center,
University of Arizona Health Sciences Center
(520) 626-7780

Hearing

Randi Winston, AuD, Audiology Consultant
The EAR Foundation of Arizona
(602) 284-1091

Lylis Olsen, MS, MPH, Audiology Consultant
Arizona EHDI Coordinator
The EAR Foundation of Arizona
(602) 690-3975

Bradley F. Golner, MD
AzEHDI Chapter Champion
(602) 971-5121

CHAPTER 2
Disorders

2.1 Arizona's Newborn Screening Panel of 29 Disorders

These are the original core disorders of the recommended Uniform Screening Panel from the US Department of Health and Human Services Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC). In May, 2010 the HHS Secretary endorsed this panel along with the addition of Severe Combined Immunodeficiency (SCID). This panel is recommended for all state newborn screening programs and is also endorsed by the American Academy of Pediatrics and the March of Dimes. Arizona does not yet screen for SCID.

Early identification and lifelong treatment of these disorders can help to prevent many of the harmful consequences of the disorders and in many cases affected babies can grow and develop normally.

<u>Endocrine Disorders</u> (2 disorders)
Endocrine disorders occur when one or more of the body's hormones cannot be produced while others are overproduced. Hormones regulate metabolism and are necessary for the normal function of the body's organs.

Congenital hypothyroidism (CH)

- A common*, endocrine condition resulting from deficient thyroid hormone secretion. It is not often an inherited condition and is most commonly caused by a failure of the thyroid gland to develop properly.
- If undetected, developmental delays, poor growth and mental retardation develop before diagnosis is made and most of this damage is not reversible with treatment. A newborn with CH may have feeding problems, lethargy, hypotonia, jaundice and constipation.
- Treated with daily oral doses of thyroid hormone. If treatment begins by two weeks of age a baby can develop normally.
- Risk of this disorder is detected by an immunoassay for elevated thyroid stimulation hormone (TSH).

Congenital adrenal hyperplasia -21-hydroxylase deficiency (CAH)

- A relatively common*, inherited group of disorders (autosomal recessive) marked by deficiency or absence of one or more enzymes essential to the production of adrenal cortex hormones. The enzyme involved most commonly is steroid 21-hydroxylase. This results in the inability to synthesize cortisol, aldosterone or both and also results in an overproduction of adrenal androgens.

- Symptoms include ambiguous genitalia in girls and in the salt-wasting form of the disorder, vomiting, dehydration, electrolyte imbalances (hyponatremia and hypokalemia), hypotension, shock and even death within 2 weeks of birth.
- Treated with hormone and electrolyte replacement with possible surgery for virilized females. Early treatment can prevent salt-wasting crises and growth and development problems.
- Risk of this disorder is detected by an immunoassay for elevated 17α-hydroxyprogesterone (17-OHP).

<u>Hemoglobin Disorders</u> (3 disorders)

Hemoglobinopathies are disorders of the red blood cells and in varying degrees affect the transport of oxygen to cells of the body.

Sickle cell anemia (Hb SS)

- A relatively common*, inherited red blood cell disorder (autosomal recessive) marked by sickling of the cells and chronic hemolytic anemia. It is caused by a mutation of both copies of the gene controlling the structure of β hemoglobin chains.
- Symptoms include intermittent severe pain, infection, damage to organs, jaundice, stroke and early death.
- Treated with prophylactic penicillin (to prevent bacterial infections), pain medications, timely vaccinations, education of parents to recognize when their child needs prompt medical care, blood transfusions and sometimes bone marrow transplants. Careful medical supervision can prevent or reduce the number of painful crises and subsequent organ damage.
- Risk of this disorder is detected by a hemoglobin result of FS by isoelectric focusing and HPLC.

Sickle beta thalassemia (Hb S/βTh)

- Rare*, inherited red blood cell disorders in which sickle genes are inherited along with genes that impair the production of hemoglobin (thalassemia). In the most serious form, normal beta hemoglobin is completely absent. This is referred to as S/β° thalassemia or S/β° thal. When some normal β chains are produced but in decreased amount it is called S/β+ thalassemia (S/β+ thal) which can cause milder symptoms, depending on the amount of normal hemoglobin produced.
- Symptoms and treatment are similar to or can be milder than those of sickle cell anemia and include splenomegaly.
- Risk of this disorder is detected by a hemoglobin result of FS or FSA by isoelectric focusing and HPLC.

Sickle C disease (Hb S/C)

- A rare*, inherited red blood cell disorder (hemolytic anemia) with hemoglobins S and C present in the absence of Hb A.
- Symptoms are variable and can be much milder than for sickle cell disease but some individuals with SC disease can have serious symptoms.
- Treatment of symptoms is similar to treatment of the same symptoms of sickle cell disease.
- Risk of this disorder is detected by a hemoglobin result of FSC by isoelectric focusing and HPLC.

<u>Other Enzyme Deficiencies</u> (2 disorders)

Biotinidase deficiency (BIO)

- A rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, biotinidase, necessary to recycle the vitamin biotin is missing or not working properly. Free biotin is needed as a cofactor for carboxylase enzymes and if it cannot be freed from its bound form by biotinidase it is not available to be used as a cofactor.
- Symptoms which usually appear in a few months of age include seizures, hypotonia, hair loss and skin rashes. Untreated, developmental delays, speech problems, hearing loss and ataxia develop.
- Treatment is daily supplementation with biotin.
- Risk of this disorder is detected by a colorimetric enzyme assay for biotinidase activity.

Galactosemia (GALT)

- Also called galactosemia type 1 or classic galactosemia.
- A rare*, inherited disorder (autosomal recessive) marked by the inability to metabolize galactose because of a deficiency of the enzyme, galactose-1-phosphate uridyl transferase (Gal-1-PUT), which is needed to convert galactose to glucose.
- Symptoms which start in the first week of life include jaundice, vomiting and feeding problems leading to failure to thrive or even death. Even with treatment (a diet without any galactose), many affected children have some development delay or mental deficit, speech or language problems, cataracts or enlarged liver.
- Risk of this disorder is detected by an enzyme assay for Gal-1-PUT activity.

Amino Acid Disorders (6 disorders)

Amino acidopathies are disorders of amino acid metabolism that occur when enzymes needed to break down specific amino acids or eliminate nitrogen from the body are deficient or absent. Toxic levels of amino acids or ammonia accumulate and cause brain damage and even death.

Phenylketonuria (PKU)

- A relatively common*, inherited disorder (autosomal recessive) that occurs when the enzyme, phenylalanine hydroxylase, necessary to break down phenylalanine, is missing or not working properly.
- Without early detection and treatment, permanent mental retardation, behavioral problems and eczema develop after a few months. Newborns with PKU seem healthy and symptoms do not appear until irreversible damage has been done.
- Treatment is a low protein diet with most protein provided in a phenylalanine-restricted formula. If treatment is started early, babies develop normally and have normal IQ.
- Risk of this disorder is detected with elevated phenylalanine by MS/MS.

Maple syrup urine disease (MSUD)

- A very rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, branched chain ketoacid dehydrogenase, necessary to break down ketoacid derivatives of leucine, isoleucine and valine, is missing or not working properly.
- Symptoms develop within the first week of life - feeding intolerance, vomiting, lethargy and progress to irreversible mental retardation, seizures, coma and death.
- Treatment is a low protein diet with most protein provided in a formula restricted in branched-chain amino acids and dietary monitoring to prevent metabolic crises.
- Risk of this disorder is detected with elevated leucine by MS/MS.

Homocystinuria (HCY)

- Also called cystathionine β-synthase deficiency or CBS deficiency.
- A very rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, cystathionine β-synthase, necessary to break down methionine, is missing or not working properly.
- Symptoms develop slowly and include developmental delay, mental retardation, skeletal abnormalities (marfanoid appearance), osteoporosis, blood clots and dislocated lens in the eye.
- Treatment is a low protein diet with most protein provided in a methionine-restricted formula. Vitamin B6 may be given along with other vitamin supplements.
- Risk of this disorder is detected with elevated methionine by MS/MS.

Citrullinemia (CIT 1)

- A rare*, inherited urea cycle disorder (autosomal recessive) that occurs when the enzyme, argininosuccinic acid synthetase, necessary to excrete the nitrogen from amino acids as urea, is missing or not working properly.
- Crisis symptoms of hyperammonemia, which can appear in the first week of life, start with loss of appetite, lethargy, hypotonia and vomiting and progress to seizures and coma. Prolonged periods of hyperammonemia can cause brain damage (intellectual disability).
- Treatment is a low protein diet and special formula, supplementary arginine and other medications along with avoiding going without eating for very long.
- Risk of this disorder is detected with elevated citrulline by MS/MS.

Argininosuccinic acidemia (ASA)

- Also called argininosuccinic aciduria or argininosuccinyl-CoA lyase deficiency.
- A rare*, inherited urea cycle disorder (autosomal recessive) that occurs when the enzyme, argininosuccinyl-CoA lyase, necessary to excrete the nitrogen from amino acids as urea, is missing or not working properly.
- Crisis symptoms of hyperammonemia, which can appear in the first week of life, start with loss of appetite, lethargy, poorly controlled breathing rate or body temperature, hypotonia and vomiting and progress to seizures and coma. Prolonged periods of hyperammonemia can cause brain damage (intellectual disability) and developmental delay.
- Treatment is a low protein diet and special formula, supplementary arginine and other medications along with avoiding going without eating for very long.
- Risk of this disorder is detected with elevated citrulline by MS/MS.

Tyrosinemia Type 1 (TYR 1)

- A very rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, fumarylacetoacetate hydrolase, necessary to break down tyrosine, is missing or not working properly.
- Symptoms develop in the first months of life but are not present immediately after birth. Untreated, a baby with tyrosinemia will develop liver disease (enlarged liver, jaundice, cirrhosis) and kidney damage leading to death. Early symptoms include lethargy, vomiting, diarrhea, irritability and failure to thrive.
- Treatment includes medication, a diet low in tyrosine and phenylalanine and possibly a liver transplant.
- Risk of this disorder is detected with elevated tyrosine by MS/MS.

Fatty Acid Oxidation Disorders (5 disorders)

Fatty acid oxidation disorders (FAODs or FODs) occur when fatty acids cannot be completely metabolized to produce energy because of defects in enzymes needed for this conversion. When the body's supply of glucose and glycogen are expended, fatty acids are broken down to supply energy during periods of fasting or increased energy demands (fever, stress). Different defects in the fatty acid oxidation pathway prevent the complete breakdown of fatty acids to produce energy and a sudden crisis occurs which can leave an affected infant or child dead or with brain damage. These conditions can yield false negative results if screening occurs after a baby is well fed.

Carnitine Uptake Defect (CUD)

- Also known as primary carnitine deficiency or carnitine transporter deficiency.
- A very rare*, inherited disorder (autosomal recessive) that is caused by a defect in the carnitine transporter that moves carnitine across the plasma membrane into cells and retains carnitine in cells. Carnitine is needed for acylcarnitine formation to transport fatty acids into mitochondria where they can be broken down.
- Symptoms which begin in infancy or early childhood include cardiomyopathy, confusion, vomiting, muscle weakness and hypoketotic hypoglycemia in acute episodes. Untreated these can lead to heart failure, encephalopathy, liver problems, coma and sudden death.
- Treatment with supplementary carnitine, a low-fat diet and avoiding fasting (frequent feedings) can prevent metabolic crises.
- Risk of this disorder is detected with lowered C0 (free carnitine) by MS/MS.

Medium-chain acyl-CoA dehydrogenase deficiency (MCAD)

- A relatively common*, inherited disorder (autosomal recessive) that occurs when the enzyme, medium chain acyl-CoA dehydrogenase, necessary to break down fatty acids of medium chain length (4 to 12 carbons long), is missing or not working properly.
- Symptoms present acutely with fasting and include hypoketotic hypoglycemia, vomiting, lethargy, seizures, metabolic acidosis, hyperammonemia, hepatomegaly and death.
- Avoiding fasting (frequent feedings) and a low-fat diet can prevent metabolic crises.
- Risk of this disorder is detected with elevated C8 by MS/MS.

Very long-chain acyl-CoA dehydrogenase deficiency (VLCAD)

- A rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, very long chain acyl-CoA dehydrogenase, necessary to break down long chain fatty acids (12 to 18 carbons long), is missing or not working properly.

- Symptoms present acutely with fasting and include hypoketotic hypoglycemia, cardiomyopathy in many cases, vomiting, lethargy, seizures, metabolic acidosis, hyperammonemia, hepatomegaly and death.
- Avoiding fasting (frequent feedings) and a low-fat diet (supplemented with MCT oil and cornstarch), can prevent metabolic crises.
- Risk of this disorder is detected with elevated C14:1 by MS/MS.

Long-chain 3-OH acyl-CoA dehydrogenase deficiency (LCHAD)

- A very rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, long chain L-3-hydroxyacyl-CoA dehydrogenase, necessary to break down certain fatty acids (between 12 and 18 carbons long), is missing or not working properly.
- Symptoms include hepatomegaly, cardiomyopathy, lethargy, hypoketotic hypoglycemia.
- Avoiding fasting (frequent feedings) and a low-fat diet (low in long chain fatty acids and supplemented with MCT oil and cornstarch), can prevent metabolic crises.
- Risk of this disorder is detected with elevated C16OH by MS/MS.

Trifunctional protein deficiency (TFP)

- Also called mitochondrial trifunctional protein deficiency.
- A very rare*, inherited disorder (autosomal recessive) that occurs when a protein containing three enzymes (long chain 3-hydroxyacyl-CoA dehydrogenase, long chain enoyl-CoA hydratase and long chain thiolase) necessary to break down long-chain fatty acids is missing or not working properly.
- Avoiding fasting (frequent feedings) and a low-fat diet (low in long chain fatty acids and supplemented with MCT oil and cornstarch), can prevent metabolic crises.
- Risk of this disorder is detected with elevated C16OH by MS/MS.

Organic Acid Disorders (9 disorders)

Organic acidemias (OAs) are a group of inherited metabolic disorders that occur when certain enzymes involved in the breakdown of amino acids and other substances are not functioning properly. Toxic acids build up in the blood and spill into the urine (metabolic acidemia). Without treatment and prevention of acute episodes, these disorders can lead to coma and death during the first days or weeks of life.

Isovaleric acidemia (IVA)

- A very rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, isovaleryl-CoA dehydrogenase, necessary to break down leucine, is missing or not working properly.
- Symptoms include metabolic ketoacidosis, poor feeding, vomiting, dehydration, lethargy, rapid shallow breathing, “sweaty feet” odor, hyperammonemia, coma and death.
- Avoiding fasting (frequent feedings) and a low-protein diet can prevent metabolic crises.
- Risk of this disorder is detected with elevated C5 by MS/MS.

Glutaric acidemia type 1 (GA-1)

- A relatively rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, glutaryl-CoA dehydrogenase, necessary to break down lysine, hydroxylysine and tryptophan, is missing or not working properly.
- Affected infant is usually macrocephalic with later signs of metabolic ketoacidosis (during periods of fasting or illness, which can progress to coma and death), failure to thrive, irritability, hypotonia, poor balance and coordination and neurological problems.
- Avoiding fasting (frequent feedings) and a low-protein diet can prevent metabolic crises.
- Risk of this disorder is detected with elevated C5DC by MS/MS.

3-Hydroxy-3-methylglutaric aciduria (HMG)

- Also known as 3-hydroxy-3-methylglutaryl-CoA lyase deficiency or HMG-CoA lyase deficiency.
- A very rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, 3-hydroxy-3-methylglutaryl-CoA lyase, necessary to break down leucine, is missing or not working properly.
- Symptoms usually appear during the first year of life and include vomiting, diarrhea, lethargy, hypotonia and metabolic acidosis (during a period of fasting or illness) which can lead to coma and death.
- Treatment includes avoidance of fasting and a low protein diet to prevent metabolic crises.
- Risk of this disorder is detected with elevated C5OH by MS/MS.

3-Methylcrotonyl-CoA carboxylase deficiency (3MCC)

- A rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, 3-methylcrotonyl-CoA carboxylase, necessary to break down leucine is missing or not working properly.
- Symptoms usually appear during the first year of life and include lethargy, vomiting, hypotonia, seizures, developmental delay and metabolic acidosis (during a period of fasting or illness) which can lead to coma and death.
- Treatment includes avoidance of fasting and a low protein diet to prevent metabolic crises.
- Risk of this disorder is detected with elevated C5OH by MS/MS.

Multiple carboxylase deficiency (MCD)

- Also known as holocarboxylase synthetase deficiency.
- A rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, holocarboxylase synthetase, necessary to attach biotin as a co-factor to certain carboxylase enzymes, is missing or not working properly.
- Symptoms can appear during the first week of life and include poor feeding, lethargy, vomiting, hypotonia, skin rash and metabolic acidosis which can lead to coma and death.
- Treatment consists of biotin supplementation.
- Risk of this disorder is detected with elevated C3 by MS/MS.

Methylmalonic acidemia - mutase deficiency (MUT)

- Also known as methylmalonic acidemia, Vitamin B_{12} unresponsive.
- A rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, methylmalonyl-CoA mutase, necessary to break down certain lipids, amino acids and cholesterol is missing or not working properly.
- Symptoms include lethargy, dehydration, vomiting, hypotonia, seizures and metabolic ketoacidosis which can lead to coma and death.
- Treatment includes avoiding fasting and a low protein diet to prevent metabolic crises.
- Risk of this disorder is detected with elevated C3 by MS/MS.

Methylmalonic acidemia - cobalamin disorders (Cbl A,B)

- Also known as methylmalonic acidemia, Vitamin B_{12} responsive.
- Rare*, inherited disorders (autosomal recessive) that occur with a defect in the synthesis of adenosylcobalamin, one of the active forms of Vitamin B_{12} which is needed as a cofactor for methylmalonyl-CoA mutase, necessary to break down certain lipids, amino acids and cholesterol.
- Symptoms include lethargy, dehydration, vomiting, hypotonia, seizures and metabolic ketoacidosis which can lead to coma and death
- Treatment includes Vitamin B_{12} injections and a low protein diet.
- Risk of this disorder is detected with elevated C3 by MS/MS.

Propionic acidemia (PROP)

- A very rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, propionyl-CoA carboxylase, necessary to break down a product of protein and fat metabolism is missing or not working properly.
- Symptoms usually appear during the first week of life and include poor feeding, lethargy, vomiting, dehydration, hypotonia and metabolic ketoacidosis which can lead to coma and death.
- Treatment includes avoidance of fasting and a protein-restricted diet to prevent metabolic crises.
- Risk of this disorder is detected with elevated C3 by MS/MS.

Beta-ketothiolase deficiency (BKT)

- A very rare*, inherited disorder (autosomal recessive) that occurs when the enzyme, β -ketothiolase, necessary to break down isoleucine, is missing or not working properly. This disorder also impairs the body's ability to process ketones, which are produced during the breakdown of fats.
- Symptoms usually appear during the first year of life and include lethargy, vomiting, seizures, and metabolic acidosis (during a period of fasting or illness) which can lead to coma and death. Long term complications include developmental delay, cardiomyopathy and hypotonia.
- Treatment includes avoidance of fasting and a low protein diet to prevent metabolic crises.
- Risk of this disorder is detected with elevated C5:1 by MS/MS.

<u>Pulmonary Disorder</u> (1 disorder)

Cystic fibrosis (CF)

- A relatively common*, inherited disorder (autosomal recessive) where a protein, cystic fibrosis transmembrane regulator (CFTR), is absent or does not work properly in the regulation of the movement of chloride ions through cell membranes. This causes the production of thick, sticky mucus that clogs lungs, pancreatic ducts and other organs.
- Symptoms include salty sweat, failure to thrive, lung and sinus infections, intestinal problems (diarrhea or constipation, pain, gas). Many babies with CF are born with meconium ileus.
- Treatment includes a higher-calorie diet, extra fluids, pancreatic enzyme supplementation, airway clearance therapy and medications (bronchodilators, mucus thinners and antibiotics).
- Risk of this disorder is detected by an immunoassay for elevated immunoreactive trypsinogen (IRT) followed by a DNA analysis of the highest 2.2% of the IRT samples for 46 mutations to the CFTR gene.

<u>Disorder Not Detected by Bloodspot Screening</u> (1 disorder)

Hearing loss (HEAR)

- A common* disorder that is both inherited and due to environmental causes.
- Defined as hearing loss that is permanent, bilateral or unilateral, sensorineural or conductive, and averaging loss of 30 decibels or more in the frequency range important for speech recognition.
- Although not required by statute, hearing screening is done in all birthing hospitals in Arizona as a standard of care. Reporting of hearing screening results to the Newborn Screening Program is required and follow-up is done for babies who do not pass a hearing screening and do not return to the hospital for further testing.
- Hearing loss is not usually diagnosed until 2 or 3 years of age without early screening programs. By that time speech and language development is delayed or impaired. Early detection and intervention helps affected infants learn speech and language.

* *common*: occurs in greater than 1 in 5,000 US births
* *relatively common*: occurs in between 1 in 5,000 and 1 in 25,000 US births
* *relatively rare*: occurs in between 1 in 25,000 and 1 in 50,000 US births
* *rare*: occurs in between 1 in 50,000 and 1 in 100,000 US births
* *very rare*: occurs in less than 1 in 100,000 US births

For further information about the disorders on the Arizona panel and other disorders:

ACT sheets, by disorder, from the American College of Medical Genetics, with suggestions for immediate action after receiving abnormal results:
http://www.acmg.net/AM/Template.cfm?Section=ACT_Sheets_and_Confirmatory_Algorithms&Template=/CM/HTMLDisplay.cfm&ContentID=5127

Disorder information from NIH, US National Library of Medicine, Genetics Home Reference:
http://www.ghr.nlm.nih.gov/search?query=%22newborn+screening%22

Disorder information from Screening, Technology, and Research in Genetics (Star-G) including fact sheets for parents:
http://www.newbornscreening.info/disorders.html

Disorder information from Genetic Alliance: http://www.geneticalliance.org

2.2 Secondary Disorders and Other Findings

Screening tests for core disorders do not exclusively test for these disorders alone. Other disorders or even carrier states may cause abnormal results and these results would be reported and followed until a diagnosis is made and a core disorder ruled out. Secondary target disorders are defined as disorders that can be detected in the differential diagnosis of a core disorder.

2.21 Other Disorders and Incidental Findings

Some of the secondary disorders that have been detected by the Arizona Newborn Screening Program along with incidental findings (carrier states) include:

- Non-classical CAH
- Hyperphenylalaninemia
- Hypermethioninemia
- Partial biotinidase deficiency
- Galactosemia carrier or Duarte variant galactosemia
- Carnitine palmitoyl transferase deficiency type II (CPT-2)
- Cystic Fibrosis carrier
- Other disease carriers
- Other hemoglobin diseases
- Hemoglobin traits

2.22 Other Hemoglobin Diseases

Other hemoglobin diseases and traits are detected because the hemoglobin screening is not specific to the three sickling disorders on the Arizona panel. These include:

Sickle/Hereditary Persistence of Fetal Hemoglobin (S-HPFH)

- A much milder form of sickle cell disease with no Hb A produced but fetal hemoglobin amounts that do not decline with time.
- Risk of this disorder is detected by a hemoglobin result of FS by isoelectric focusing and HPLC.

Hemoglobin C Disease

- An inherited red blood cell disorder (autosomal recessive) marked by target cells and chronic hemolytic anemia. It is caused by a mutation of both copies of the gene controlling the structure of β hemoglobin chains.
- It occurs most often in individuals of African descent and is characterized especially by splenomegaly, arthralgias, abdominal pain and the presence of target cells and hemoglobin C in the blood.
- Risk of this disorder is detected by a hemoglobin result of FC by isoelectric focusing and HPLC.

Alpha Thalassemia

- A hereditary hemolytic anemia marked by a decreased rate of synthesis of the α globin chains of hemoglobin.
- Four gene deletions produce alpha thalassemia major which is incompatible with life. Affected infants are stillborn or born with severe hydrops fetalis (abnormal accumulation of fluid in the entire body of the infant) and die quickly. If transfusions are given prenatally and continued throughout life, a baby can survive.
- Three gene deletions produce Hemoglobin H disease, two produce alpha thalassemia trait, and one the "silent carrier."

Hemoglobin H Disease

- An inherited red blood cell disorder (autosomal recessive) marked by chronic hemolytic anemia. It results from only one working copy of the gene controlling alpha globin synthesis (3 gene deletions or two gene deletion and one non-deletional mutation).
- It is marked by hypochromic red blood cells with inclusions that cause them to resemble golf balls (Hemoglobin H is unstable and cannot be quantified by laboratory methods).
- Risk of this disorder is detected by a hemoglobin result of FA plus Barts (in a concentration of at least 15% of total hemoglobin) by isoelectric focusing and HPLC.

Hemoglobin H – Constant Spring Disease

- A chronic inherited hemolytic anemia resulting from only one working copy of the gene for alpha globin synthesis and a structural defect called Constant Spring in one of the non-working copies of the gene. It is a more severe form of Hemoglobin H disease with variable consequences up to transfusion dependency.
- Risk of this disorder is detected by a hemoglobin result of FA + Barts (in a concentration of at least 15% of total hemoglobin) by isoelectric focusing and HPLC.

Beta Thalassemia (β thal)

- A hereditary hemolytic anemia marked by a decreased rate of synthesis of the β globin chains of hemoglobin.
- The homozygous form (Cooley's, Mediterranean or erythroblastic anemia) in which β globin is completely absent is also referred to as β° thalassemia or β° thal). The NBS hemoglobin result would be F only.
- When some beta chains are present but in decreased amount it is called β+ thalassemia or β+ thal). The NBS hemoglobin result would be FA or possibly F only). In an older individual, there would be increased amounts of Hb A2.

2.23 Hemoglobin Traits

Sickle Cell Trait

- A condition in which one sickle gene mutation is inherited along with a normal gene for β globin chain structure.
- This trait is usually a very mild condition and most people with sickle cell trait never even know that they have it. However, in rare cases a person with sickle cell trait can have problems at high altitude or when exercising hard in hot weather when not drinking enough water.
- Risk of this trait is detected by a hemoglobin result of FAS by isoelectric focusing and HPLC.

Alpha Thalassemia Trait

- A condition in which an individual inherits only two working genes for alpha globin chain synthesis. Individuals with this condition will have a mild microcytic, hypochromic anemia. This decrease in alpha globin chain production does not lead to health problems.
- Asians with alpha thalassemia trait usually, but not always, have the two non-working genes on the same chromosome (cis-type) making it possible to have a child with hydrops fetalis. African Americans with alpha thalassemia trait usually, but not always, have the two non-working genes on different chromosomes (trans-type) and therefore can only transmit one non-working gene to their offspring.
- Risk of this trait is detected by a hemoglobin result of FA plus Barts (in a concentration of at least 10% of total hemoglobin) by isoelectric focusing and HPLC.
- See the Appendices for the Arizona Hemoglobin Barts Fact Sheet for Health Care Providers.

Other Traits

- Other traits like Hemoglobin C Trait, Hemoglobin D or G trait, Hemoglobin E trait or Unidentified Traits can be identified. These do not usually have any health problems other than a mild anemia that is not responsive to iron therapy.

CHAPTER 3
Bloodspot Screening

3.1 Provider Responsibilities

Providers are responsible for timely collection of properly identified and acceptable newborn screening specimens, rapid transfer of specimens to the State Lab and follow-up on abnormal results.

1. Order a newborn screening test for each newborn and provide parents with information about newborn screening at the time the test is ordered. The brochure, "Newborn Screening, A Guide for Parents," is included in the Appendices and copies can be ordered at no charge for distribution to parents using the brochure order form in the Appendices.

   If parents refuse to have their newborn tested, the provider ordering the screen is responsible for documenting the refusal and submitting a collection kit with identifying information but no blood to the State Lab. A sample refusal form is included in the Appendices.

2. Fill out each specimen card completely. See the Appendices for sample specimen cards. Instructions for completing the demographic information on the card are in Chapter 4.4 on page 50.

3. Collect an acceptable blood sample following the instructions in Chapter 4.2 on page 48.
   Posters showing acceptable and unsatifactory specimens and specimen collection steps are available on our website, www.AZNewborn.com

4. Collect a timely specimen following instructions in Chapter 4.6 on page 57.
   - First specimens must be collected, according to whichever of the following occurs first:
     - 24 to 72 hours of age (closer to 24 hours is recommended)
     - Before a transfusion
     - Before discharge from the hospital
   - See Chapter 4.7 and 4.8 on page 59 for special considerations for sick, low birth weight or preterm infants.
   - Second specimens should be collected:
     - Between 5 to 10 days of age, or
     - At the first visit to the medical home or other health care professional (even if this visit is before 5 days of age)
     - Unless you verified that another provider collected the 2nd specimen.

- Arizona requires healthcare providers to order a second newborn screening test for babies who enter their care and are not yet one year of age unless previous test results are verified.

- Submitters of second screens must also submit payer information so that responsible parties (parents or insurance carriers) can be billed for screening.

5. Promptly send specimens to the State Lab. Submitters should send each specimen to the State Lab no later than 24 hours or the next working day after the blood sample is collected. See Chapter 4.6 on page 57 for more information about timely handling of specimens.

6. Receive a copy of the newborn screening test results and document them in the baby's medical record.

7. Follow-up on positive newborn screening results, order appropriate additional testing and refer the baby to a specialist, if needed.

8. Send copies of results from subsequent tests performed at labs other than the State Lab in response to an abnormal screening test to the Newborn Screening Program. Our fax number is 602-364-1495.

Please refer to the Arizona Administrative Code (Public Services, Title 9, Section 9-13) for the Newborn Screening rules. See the Appendices for a full copy of the rules.

3.2 Obtaining a Submitter Code

Each entity that sends specimens to the State Lab or receives test results from the State Lab must have a unique identification code. This code is assigned by the Demographics unit of the Newborn Screening Program and must appear on each newborn screening specimen collection kit submitted to the State Lab.

If you are a new provider you will need to obtain a submitter code. Please call the Demographics unit at 602-364-1468. You will need to provide complete contact information including practice name (if applicable), mailing address, telephone and fax numbers.

This information needs to be updated if anything changes since it is the information used to mail results and contact submitters when specimens are unsatisfactory or abnormal.

3.3 Ordering NBS Supplies and Education Materials

3.31 Collection Kits and Envelopes

Newborn screening collection kits and other supplies can be ordered by submitters at no charge to them.

Remember that kits expire and cannot be tested past the expiration date (found above the circles for blood on the filter paper portion of the kit). Please do not order more specimen cards than can be used promptly. Always check to be sure that the kits have not expired before using. Store kits on end, not stacked flat so that the filter paper is not compressed by the weight of kits on top of them.

Linked kits and pink envelopes are sent to hospitals only.

> The first specimen is collected on the top kit marked 1 and the bottom kit marked 2 is placed in the pink envelope and sent home with the baby. Parents should be instructed to bring this kit to their first doctor's appointment.

Supplemental kits are single kits which can be used for any screen. Above the baby's name field in the top left part of the form are check boxes to indicate what type of screen is being submitted. Please be sure to mark this area when using a supplemental kit. There are some supplemental kits circulating without the check boxes. Please note the type of specimen submitted on these forms and the disorder to be tested if it is a recall screen.

White envelopes are used for mailing specimens to the State Lab. Up to 4 kits can be sent in each envelope. Submitters who use a courier or FedEx will not need these envelopes.

> Hospitals can arrange for FedEx pickups paid for by the Program by calling the State Lab Receiving Dept. at 602-542-1190 or E-mail to labreceiving@azdhs.gov.

Fill out the order form in the Appendices and fax it to State Lab Receiving at (602)364-0758 or go online to www.aznewborn.com to obtain an electronic copy which can be completed and attached to an E-mail to labreceiving@azdhs.gov.

3.32 Brochures

Newborn screening parent brochures can also be ordered by submitters at no charge to them.

Fill out the brochure order form in the Appendices and fax it to the Newborn Screening Program at (602) 364-1495 or go online to www.aznewborn.com to obtain an electronic copy which can be completed and attached to an E-mail to nbs@azdhs.gov

3.4 Results and Follow-up

3.41 Results and Reference Ranges

Printed reports of newborn screening results are mailed to submitters and health care professionals listed on the specimen collection kit. Each mailer has a disclaimer reminding providers that these results are from screening tests and are not diagnostic of any condition nor can they rule out the possibility of any screened disorder.

Normal results are reported by disorder group and do not show actual values. Abnormal results are given for a disorder listed within a disorder group with a footnote requesting appropriate action. If a result in a group is not specified as abnormal, it is normal. For example, there are six amino acid disorders in that group and if the phenylalanine result is shown, it is an abnormal test for PKU. The other five disorders are not shown and they are normal. Please see the Appendices for sample mailers.

Reference ranges are listed on the back of the mailer. Further information about analytes, assays and reference ranges by disorder are shown in table form:

Arizona Newborn Screening Program: Analytes, Assays and Reference Ranges by Disorder

Disorder	Analyte	Assay	Reference Range	Abnormal Result
Endocrine Disorders				
CH	TSH	PerkinElmer AutoDelphia® time resolved fluoro-immunoassay for TSH	1st screen: < 30 µU/mL 2nd screen: < 20 µU/mL	abnormal : ≥ 60 µU/mL borderline: 1st screen: 30 - 59.9 U/mL 2nd screen: 20 - 59.9 µU/mL
CAH	17-OHP	PerkinElmer AutoDelphia® time resolved fluoro-immunoassay for 17-OHP	by birth weight: <1250 g: < 135 ng/mL 1250-1749 g: < 90 ng/mL 1750-2499 g: < 65 ng/mL ≥2500 g: < 50 ng/mL	by birth weight: <1250 g: ≥160 ng/mL 1250-1749 g: ≥ 135 ng/mL 1750-2499 g: ≥110 ng/mL ≥2500 g: ≥ 90 ng/mL borderline, by birth weight: <1250 g: 135 - 159.99 ng/mL 1250-1749 g: 90 - 134.99 ng/mL 1750-2499 g: 65 - 109.99 ng/mL ≥ 2500 g: 50 - 89.99 ng/mL
Hemoglobin Disorders				
Hb SS Hb S/β thal Hb SC	hemoglobin	Hb IEF & HPLC	Normal Hb present (FA)	F only, Fx, FAx where x is a variant Hb (S, C, D, E, G, U, Barts)
Other Enzyme Deficiencies				
BIOT	biotinidase	colorimetric enzyme assay	color change equal to control - enzyme activity present	enzyme activity absent pale - reduced enzyme activity
GALT	Gal-1-PUT	Bio-Rad fluorometric enzyme assay	≥ 1.1 U/g Hb	≤ 0.7 U/g Hb equivocal: 0.8 - 1.0 U/g Hb
Amino Acid Disorders				
PKU	phenylalanine	tandem mass spectrometry	< 3.0 mg/dL	≥ 3.0 mg/dL
MSUD	leucine	tandem mass spectrometry	1st screen: < 4.0 mg/dL 2nd screen: < 6.0 mg/dL	1st screen: ≥4.0 mg/dL 2nd screen: ≥ 6.0 mg/dL
HCY	methionine	tandem mass spectrometry	< 2.0 mg/dL	≥ 2.0 mg/dL
CIT-1 ASA	citrulline	tandem mass spectrometry	1st screen: < 1.6 mg/dL 2nd screen: < 2.6 mg/dL	1st screen: ≥ 1.6 mg/dL 2nd screen: ≥ 2.6 mg/dL
TYR-1	tyrosine	tandem mass spectrometry	< 10.0mg/dL	≥ 10.0mg/dL
Fatty Acid Oxidation Disorders				
CUD	C0	tandem mass spectrometry	> 8.00 µmol/L	≤ 8.00 µmol/L
MCAD	C8	tandem mass spectrometry	< 0.60 µmol/L	≥ 0.60 µmol/L
VLCAD	C14:1	tandem mass spectrometry	< 0.60 µmol/L	≥ 0.60 µmol/L
LCHAD TFP	C16OH	tandem mass spectrometry	< 0.60 µmol/L	≥ 0.60 µmol/L
Organic Acid Disorders				
IVA	C5	tandem mass spectrometry	1st screen: < 0.78 µmol/L 2nd screen: < 0.92 µmol/L	1st screen: ≥ 0.78 µmol/L 2nd screen: ≥ 0.92 µmol/L
GA-1	C5DC	tandem mass spectrometry	< 0.40 µmol/L	≥ 0.40 µmol/L
HMG 3MCC	C5OH	tandem mass spectrometry	1st screen: < 1.15 µmol/L 2nd screen: < 1.03 µmol/L	1st screen: ≥ 1.15 µmol/L 2nd screen: ≥ 1.03 µmol/L
MCD MUT Cbl A, B PROP	C3	tandem mass spectrometry	1st screen: < 10.0 µmol/L 2nd screen: < 7.0 µmol/L	1st screen: ≥ 10.0 µmol/L 2nd screen: ≥ 7.0 µmol/L
BKT	C5:1	tandem mass spectrometry	< 0.25 µmol/L	≥ 0.25 µmol/L
Pulmonary Disorder				
CF	IRT DNA	PerkinElmer AutoDelphia® time resolved fluoro-immunoassay for IRT/ Third Wave CFTR mutation assay	IRT in lower 97.8% of batch or < 65 ng/mL: CF not indicated by IRT or IRT in upper 2.2% of batch and no mutations found	One mutation detected Two mutations detected

3.42 Follow-up on Abnormal Results

The Follow-up Section is responsible to contact primary care providers to ensure infants will receive medical evaluation, further testing and/or proper treatment. It is the Program's goal that no infant will be lost to follow-up.

Abnormal results are communicated to the Follow-up Section by the Laboratory Section. From there, follow-up staff will take appropriate actions to locate the infant's primary care provider in a timely manner. The follow-up specialist assigned to the case will contact the primary care provider to ensure the infant will receive medical evaluation, further testing and/or proper treatment. Abnormal results are communicated to the primary care provider (and/or the designated nurse practitioner or medical assistant) by phone, by fax and by mail.

Follow-up specialists work closely with pediatric endocrinologists, metabolic geneticists, hematologists and cystic fibrosis specialists. These contracted specialists provide guidance to the Follow-up case management team until resolution is reached. The contact information for these specialists is listed in Chapter 1.81 on page 16 of this document; primary care providers are strongly encouraged to consult these specialists on specific cases.

Follow-up specialists monitor progress made and only close the case when normal results are received or it is verified that the baby is receiving appropriate care. It is important that physicians communicate results of testing performed at other labs, and the final diagnosis to the follow-up specialist. If the follow-up specialist is not informed of normal confirmatory results, unnecessary phone calls and faxes may be made to the physician or the parents.

Providers may request assistance from the Program's Follow-up staff if they are having problems locating parents or obtaining repeat tests for infants. If the infant is no longer under their care, providers should notify the follow-up specialist immediately.

If the follow-up specialist is unable to locate the primary care provider; he or she will contact the parent in order to obtain the name of the physician taking care of the baby. At that time, parents are advised to contact their primary care provider for further testing. Lab results are not communicated to the parents, only the fact that the newborn screening results are not normal and further testing is required.

When the attempts to contact the parents are unsuccessful, the follow-up staff will coordinate efforts with submitters and other agencies in order to locate parents as soon as possible.

3.43 Follow-up on Hemoglobin Traits

Letters are sent to the provider of record and the parents to notify of probable hemoglobin carrier status. There is no further follow-up beyond this notification. Parents are encouraged to talk to their primary care provider about the fact that the results indicate that the baby has a hemoglobin trait. The parent notification letter in English and Spanish directs them to contact the Office for Children with Special Health Care Needs or the Quest to Cure Foundation for more information and resources. Contact information for these organizations is in Chapter 6.3 on page 73.

A chart of hemoglobin result mnemonics with descriptions of the probable results is in the Appendices.

3.44 Follow-up on Unsatisfactory Specimens

Submitters and health care providers are notified of unsatisfactory specimens by fax and by mail. Unsatisfactory specimens are not tested by the State Laboratory. The submitter (birth hospital or provider who ordered the screen) is responsible to contact the parents in order to make arrangement for the collection of another specimen as soon as possible.

The Follow-Up staff monitors unsatisfactory specimens to ensure that a subsequent valid specimen is received. If a valid specimen is not received within two weeks, the infant's medical home or provider who ordered the screen and parents will be notified by mail that a valid specimen has not been received and will urge them to make arrangements for the collection of another specimen as soon as possible. A reminder letter will be sent if a valid specimen is not received thirty days after the date the first letter was mailed.

Follow up will be discontinued once a valid specimen indicating normal results are received, or thirty days following the issuance of the reminder letter.

3.45 Referral to Children's Rehabilitative Services (CRS)

The Children's Rehabilitative Services (CRS) Program provides medical treatment, rehabilitation and related support services as a carveout to Medicaid recipients with qualifying chronic and disabling conditions under age 21. Care is provided to more than 26,000 children and youth with special health care needs (CSHCN) through four family-centered, multi-specialty interdisciplinary clinics. CRS covers over 350 chronic and disabling health conditions including cerebral palsy, cleft lip/cleft palate, spina bifida, cystic fibrosis, sickle cell anemia, metabolic and endocrine disorders, and many congenital anomalies. Many CRS recipients are medically fragile and require complex care from multiple pediatric

physician subspecialists that are frequently in short supply. CRS manages a statewide network of specialists to provide timely access to CSHCN throughout the state. Strategies to ensure access include the use of field clinics in which specialists travel to remote areas of the state, as well as using innovative techniques such as telemedicine to minimize travel for both families and physicians.

The CRS Program is administered by the Arizona Health Care Cost Containment System through a contract with Arizona Physicians IPA.

Arizona Physicians IPA-Children's Rehabilitative Services (APIPA-CRS) does not determine eligibility based on income. To be eligible for APIPA-CRS services a child must:

- Have certain medical conditions,
- Be under age 21,
- Be an U.S. citizen or qualified alien, and
- Live in Arizona.

APIPA-CRS recipients must be members of an AHCCCS acute care health plan or an Arizona Long Term Care System (ALTCS) plan, and some recipients may have private insurance as well. APIPA-CRS will coordinate care with the recipient's acute care health plan, ALTCS or private insurance providers.

To apply for services, anyone can fill out an application including a family member, doctor, or health plan representative. Along with a completed referral/application, medical records that document the child's CRS eligible condition must be submitted. When someone other than a parent/guardian or a family member completes an application, this is called a referral.

Referrals/applications to the CRS Program are initiated by submitting a CRS Referral/Application Form. The CRS Referral/Application Form can be obtained from many sources, including physicians' offices,

and the APIPA-CRS website at:
http://www.myapipa.com/en/members/refer_child.jsp?xlang=en&xrole=members&xstate=az&xplan=uhcaz&xproduct=CRS

If you need help completing the APIPA-CRS referral/application or getting information to send in with the application packet, ask APIPA-CRS Member Services for help.

Completed applications can be mailed or faxed to:

APIPA-CRS
Attn: Eligibility and Enrollment
PO Box 33320
Phoenix, AZ 85067-3320
Fax Number: 1-866-623-1692

The APIPA-CRS Referral/Application Form shall contain the following:

a. The name, address, and phone number of the referral source;
b. The relationship of the person completing the referral/application form to the applicant;
c. The applicant's name, date of birth, social security number, gender, home address and contact information, race, citizenship, and preferred language;
d. If the applicant is a child, the name of at least one parent/guardian of the applicant;
e. If known to the referral source:
    i. The applicant's diagnosis;
    ii. The applicant's list of allergies if applicable;
    iii. The applicant's primary care physician or, if the applicant does not have a primary care physician, the name of a health care organization at which the applicant receives medical care;
    iv. Whether the applicant is enrolled in AHCCCS (Title XIX or Title XXI) or has other health insurance;
    v. Whether the applicant has ever received CRS services before;
    vi. Whether the applicant is a U.S. citizen or qualified alien; and
    vii. If a physician has not evaluated the applicant, the reason the referral source believes the applicant may be eligible for CRS.

Documentation to accompany the referral/application form for applicants who have been evaluated by a physician:

a. Documentation from a physician who has evaluated the applicant that supports the medical diagnosis;
b. Diagnostic test results that support the medical diagnosis.

Within fourteen (14) calendar days of receipt of a referral/application, APIPA-CRS will determine whether an applicant is eligible for CRS. The applicant's parent/guardian and referral source will be notified in writing of the decision.

3.5 Flow Chart of the Newborn Screening Process

See the Appendices for a summary of the follow-up processes for bloodspot and hearing shown in flow charts.

3.6 Specimen Retention

Newborn screening bloodspot specimens and attached information submitted to the Arizona Department of Health Services (ADHS) are the property of the ADHS.

Access to specimens and submitted information is restricted to ADHS employees and contractors approved by the Newborn Screening Program Manager as necessary to meet specific program needs. All applicable laws, regulations and policies safeguarding the privacy and confidentiality of medical information are followed by those with access to these specimens and attached information.

Specimens are stored at room temperature for 90 days and then autoclaved and discarded. Specimens of interest (confirmed cases) may be frozen and retained for quality assurance purposes.

During the storage period and after the newborn screening tests are complete, residual dried blood may be used for lab methodology development or validation or other quality assurance testing.

Specimens may be released to others under certain circumstances including:

1. Written request from a provider, at the request of a parent or guardian. Bloodspot specimens will not be returned directly to a newborn's parent or guardian.
2. Written request from the Office of the Medical Examiner.
3. To named persons in a court order or subpoena.

3.7 Frequently Asked Questions

What is the chance that an infant will have a disorder detected by newborn screening? The combined risk of having one of the 29 disorders on the panel is greater than 1 in 500.

What if an infant has a family history of a disorder detected by newborn screening? This family should arrange for diagnostic testing with an appropriate specialist and not wait on a newborn screening result. The possibility of this disorder should be considered by any provider of care to the baby.

What about false positive results? In any screening tests there will be some false positives. Repeat testing should show that the baby is not affected. Parents should be told that screening tests are not diagnostic and further testing is needed.

How likely is a false negative result? False negatives can occur for some disorders when specimens are collected late or early. Early collection (prior to 12 – 24 hours of age) can cause false negative results for amino acid and organic acid disorders because the specific amino acid or organic acid may not have accumulated in sufficient quantity to reach the abnormal range in the first day of life. Collection after 24 hours of age should yield a valid result for these disorders.
Late collection of a first screen (after 48 hours of age) can cause a false negative result for fatty acid oxidation disorders because by this time the baby has been fed and the fatty acid oxidation pathway is not being used as it is when glucose sources have been exhausted. Even with affected babies, a second screen can be normal after an abnormal first screen and this second normal screen does not rule out the possibility of the disorder. Further diagnostic testing is required.

Although rare, a false negative is possible for CF if the IRT is not sufficiently elevated in an affected baby and mutation testing is not done.

Timing of Specimen Collection

A baby who is only 3 days old has come for her first doctor's appointment? Isn't this too early to collect a 2nd screen? No, the second screen should be collected at this appointment. Improved testing methodology makes earlier testing results valid and reliable. False negatives for some disorders are more likely when testing is delayed. Also, many disorders on the expanded panel can cause damage very quickly if not detected early. There is no longer any advantage to delaying testing.

How long should we wait before collecting a second screen for a baby in the NICU? If a baby is still in the hospital at 5 days of age, a second screen should be collected no later than 10 days of age, or prior to discharge, whichever comes first.

A baby missed getting a 2nd screen – is 3 months too old for a second? No, the State Lab will accept specimens from babies up to a year of age. However, earlier testing is recommended as some disorders will show more false negatives when testing is delayed beyond the newborn period. Some other disorders would have already shown symptoms by 3 months of age.

Submitting Specimens for Testing

A baby's first screen was unsatisfactory for testing. Do we need to order two more screens for this baby? No, assuming that the repeat test has normal results. Further testing would only be necessary if a result came back abnormal. Be sure to check to see that a CF test was added to the repeat screen.

What about screening infants from outside Arizona? For a baby that moves to Arizona, if results of newborn screening from another state are not recorded in the medical record or cannot be verified, a newborn screen should be collected. If a baby is born in Arizona or receives health care in Arizona, a newborn screening specimen should be collected, no matter whether the family lives in another state.

What if an infant is adopted? Whose demographic information should be listed in the Mother's Information section? Note that this is an adoption and enter information about whoever has custody of the baby - the adoptive mother, if the baby is with her or the adoption caseworker or agency if they have not yet placed the baby in the adoptive home. The contact information under Mother's Information needs to show a responsible person who could be contacted about the baby if there were abnormal results.

What if an infant is in foster care? Whose demographic information should be listed in the Mother's Information section? Enter the CPS caseworker's contact information and note that this is foster care. If the caseworker's contact information is not available, enter the foster mother's name, address and phone number and note that she is the foster mother. The contact information under Mother's Information needs to show a responsible person who could be contacted about the baby if there were abnormal results. Entering "foster care" without contact information is not acceptable.

Can blood from other than a heelstick be used for newborn screening? Yes, blood obtained by venipuncture is acceptable, although not the method of choice. Indicate on the collection card that blood was obtained by venipuncture. Blood should not be drawn from an extremity or line used for infusion of IV fluids (including blood) without appropriate precautions. Umbilical cord blood is not acceptable.

Obtaining Results

What is the turnaround time for NBS reports? Once a specimen has been received at the State Lab, testing is usually finished within 3 working days. Reports are mailed the next day and should be received within about 10 days from the date of specimen collection (if it was sent promptly to the State Lab).

How do I obtain a copy of newborn screening results? Results will be mailed to the provider listed on the specimen collection kit. If you need another copy, please call the Demographics unit and provide them with identifying information about baby and mom. Results will be sent to a new medical home for the baby upon request.

The results have arrived but the infant's demographic information is incorrect. What should I do? You can call or fax the NBS Demographics unit and they will correct the information and issue an updated mailer.

It has been several weeks since I sent in a NBS sample and I have not received a report. What should I do? Call the NBS Demographics unit and request another copy of the report.

Cystic Fibrosis Testing

What kind of DNA testing is done by the Newborn Screening Program? The only DNA testing done by the Arizona Newborn Screening Program is to detect 46 mutations to the CFTR gene. Only those samples that have an elevated immunoreactive trypsinogen (IRT) level will be tested for mutations so for the great majority of babies screened, no DNA testing is done.

What does a negative test for CF mean? A negative newborn screening test for CF indicates a much reduced probability that a baby has CF but it doesn't completely rule out the possibility of disease.

The first test for CF is a test for elevated immunoreactive trypsinogen (IRT). For specimens in the lower 97.8% of the batch the result would be reported as "CF not indicated by IRT." This means that IRT was not elevated and no mutation testing was done

If the IRT was elevated (IRT in the upper 2.2% of the batch), the sample would be tested for 46 common CF mutations. If none of these 46 mutations were detected, the result would be reported as "No Mutations Detected." Since there are over 1000 mutations to the CFTR gene, not every mutation would be picked up by our testing panel and cannot be considered "normal."

Why is there no CF test on most 2nd screens? CF testing is only done once for each baby. Usually the first screen is tested and the second is not. However, if a first screen is not submitted or is unsatisfactory for testing, a CF test is added to the second screen. IRT testing is most accurate when done early since IRT falls as a baby gets older, even if the baby has CF.

It would be confusing if a first screen test was positive and a mutation was detected and then a second showed "CF not indicated by IRT" because the IRT value had fallen into the range where mutation testing was not done. In this case

the first screen results are accurate and indicate the possibility of CF or CF carrier. The second (although reporting IRT values accurately) is misleading because there is a CF mutation present.

CHAPTER 4
Specimen Collection

4.1 A message from the NBS Office Chief

Dear Health Care Professional:

Newborn Screening is one of the most important things that can happen to a newly born child. The Arizona Newborn Screening Program is working diligently to make sure every newborn is screened properly, in a timely manner, and affected children have the best outcomes possible.

Recently, data have shown that many specimens across the state are being collected improperly and are not being sent in a timely manner. These delays create extreme risk for a child who is affected with a disease. Earlier collection of properly obtained specimens, with prompt transport to the State Lab can prevent tragic outcomes for Arizona families.

The Arizona Newborn Screening program quality measures are as follows:

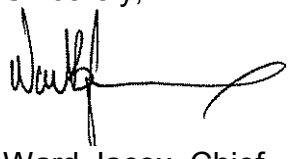
1. Submitters should have less than a 1% rejection rate for collected specimens.
2. Specimens should be collected within the recommended screening windows.
   - for 1st screens: 24 - 36 hours of age
   - for 2nd screens: 5 - 10 days of age or earlier (at the first outpatient visit to the doctor)
3. Specimens should be shipped promptly to the State Lab
   (no more than 24 hours between collection and shipment to the State Lab)

Please review the instructions for specimen collection and handling in this chapter with particular attention to Chapter 4.6. Please share these instructions with all staff that collect and handle Newborn Screening specimens and emphasize the importance and urgency of proper specimen collection and handling.

If you need further information or assistance with training, please do not hesitate to contact us directly at 602-364-1409 or visit our website, http://www.aznewborn.com.

With your help, babies can be saved from disability or even death. Your continued efforts are greatly appreciated.

Sincerely,



Ward Jacox, Chief
Office of Newborn Screening

4.2 Newborn Screening Specimen Collection Instructions

1. **Use the specimen collection kit supplied by ADHS.** Store the kits on end so that the filter paper is not compressed by the weight of kits stacked on top. To order kits and envelopes (US Mail and FedEx), call 602-542-1190 or use the order form in the Appendices or on the website, www.AZNewborn.com

2. **Check the kit's expiration date** (on the top left of the filter paper strip above the blood spot circles). Make sure the kit has not expired since expired kits will be considered unsatisfactory by the lab and will not be tested.

3. **Check the filter paper for damage** (scratched, abraded, torn or creased, detached or partially detached from the rest of the card) and discard the kit if it is not in good condition.

4. **Fill out the form completely**, printing legibly in dark ink (preferably black) and pressing hard enough so that it is possible to read the bottom copy. Record all requested information and attach insurance information to second screens. Do not use a printer or imprint card. Complete this step before collecting the blood to avoid contamination or smearing of the blood and to ensure the baby is properly identified.

5. **Gather supplies, wash hands and put on powder-free gloves** (preferred since powder can contaminate the sample). Wash hands and change gloves between babies. Observe universal precautions for handling of blood samples.

6. **Recheck the identity of the infant and mother**. Make sure that the infant being tested is the one described on the form.

7. **Position the infant with the heel lower than the heart.** Swaddling with one foot exposed can help to keep the infant comfortable. Warm the heel with a warming device or a cloth moistened with water no hotter than 42° C (107° F).

8. **Choose a collection site on the sides of the plantar surface of the heel** (the walking surface), avoiding the center curvature of the heel. See the illustration on the back of the collection kit.

9. **Wipe the collection site with 70% isopropyl alcohol or other non-iodine containing disinfectant and allow to air dry**.

10. **Use a sterile lancet to puncture the skin to a depth of no more than 2 mm**.

11. **Wipe off the first drop of blood with sterile gauze.** This drop contains tissue fluids that can dilute the sample.

12. **Allow a large drop of blood to form**. Apply gentle pressure to the heel (away from the puncture site) and release as drops of blood form. Do not squeeze or "milk" the collection site (this can cause hemolysis and addition of tissue fluids to the sample).

13. **Touch the drop of blood to a filter paper circle and allow it to fill the circle.** Do not apply multiple drops to the same circle. Apply blood to one side of the filter paper only. Collect 5 large drops of blood to fill the 5 circles on the filter paper. Fold the protective biohazard flap away from the bloodspots so that it doesn't touch the specimen until it is dry. Do not touch the filter paper with anything but blood drops.

    Do not write or put stickers on the filter paper. Cord blood is not acceptable. If blood is obtained by venipuncture, do not use tubes with EDTA.

14. **Dispose of lancets and other supplies in approved biohazard containers.**

15. **Air-dry the blood spots on a level horizontal surface**. Protect the specimen from heat, direct sunlight or contamination while drying. Do not stack until dry (and the biohazard flaps are in place).

16. **Send the specimen to the State Lab as soon as it is dry** but no later than 24 hours after collection. Specimens will not be tested if they are received more than 14 days after collection. Do not send specimens in sealed plastic containers, even when completely dry, unless desiccants are included.

    Hospitals should send a courier or FedEx envelope each weekday so that the State Lab receives specimens each week day. Hospital labs should know when the FedEx pickup is for their facility and ensure that a package is sent out daily.

    Include insurance information (see section 1.73) with second screens but do not staple paperwork to the collection card.

17. **Keep a record of specimens sent**. The kit number (or AZ number, found under the bar code and above the blood circles) can be used to track specimens. Use this record to ensure that all results have been received.

    Call the Newborn Screening Program at 602-364-3190 or fax a request for results to 602-542-4099 (include the baby's name and date of birth and either the mother's name and date of birth or the kit number in your request).

18. **If you have questions about specimen collection, please call 602-364-1409 or 800-548-8381** (within Arizona) or send an E-mail to the Newborn Screening Educator at NBS@azdhs.gov.

4.3 Sample Collection Kits

Please see the Appendices for sample collection kits with fields numbered to correspond with the instructions for entering demographic information in the section below (Chapter 4.4).

4.4 Collection Kit Demographics

INSTRUCTIONS FOR COMPLETING INFORMATION ON THE NEWBORN SCREENING SPECIMEN COLLECTION KIT

Refer to the numbered fields on the sample collection kits found in the Appendices:

When using the linked (double) kits, collect the first specimen with the top kit marked "1" and place the bottom kit marked "2" in the bright pink envelope and give it to the mother to take with her to the first doctor's appointment after discharge. Collect the second specimen with the kit marked "2".

When using the supplemental (single) kit, please mark the type of specimen in the appropriate box above the Baby's Name fields. If the specimen being collected is a repeat (after an abnormal result on a second screen), check the box marked "recall" and write in which disorder is being retested. These repeat specimens will test only the disorder(s) marked on the form.

Field #	Field Name	Instructions
1	Baby's Name (Last, First)	Enter the last name of the infant. This may not always be the same as the last name of the mother (or the father). Use the birth record or birth certificate name, if known. Enter the baby's first name, if known. If the first name is not known, enter Boy or Girl. In the case of multiple births, please enter name and birth order designation (e.g. Boy Twin A, Mary Trip B, John Quad C, etc.)
2	Date of Birth	Enter the infant's date of birth, in "mm/dd/yy" format. For example, June 13, 2009 should be entered as "06/13/09."

Field #	Field Name	Instructions
3	Time of Birth	Enter the infant's time of birth, using military time or noting am/pm. For example, if the time of birth is 7:25 am, it would be entered as "0725" or "7:25" with am circled. If the time of birth is 8:15 pm it would be entered as "2015" or "8:15" with pm circled.
4	Birth Weight	Enter the infant's birth weight in grams. If the birth weight cannot be obtained in grams, you may provide pounds and ounces, but clearly mark the form by circling "Lb/oz".
5	Date of Collection	Enter the calendar date on which the specimen was taken, in "mm/dd/yy" format.
6	Time of Collection	Enter the time at which the specimen was collected, using military time or noting am/pm.
7	Current Weight	Enter the infant's weight in grams. If the current weight cannot be obtained in grams, you may provide pounds and ounces, but clearly mark the form by circling "Lb/oz".
8	Sex	Mark the appropriate box to indicate the infant's gender.
9	Method of Collection	Mark the box that states how the specimen was collected.
10	Baby's AHCCCS #	Enter the baby's AHCCCS ID number if it is known. If the infant's AHCCCS ID # is not known, leave this space blank, but enter the mother's ID # (if she is enrolled in AHCCCS) in the Mom's AHCCCS # field under Mother's Information.
11	Medical Record #	Enter the infant's medical record number.
12	Single or Multiple Birth	Mark the appropriate box, and circle the appropriate birth order (A, B, C, D) or enter the appropriate letter for multiple births. Write which multiple (twin, trip, quad, etc.).

Field #	Field Name	Instructions
13	Race	Mark the box that identifies the infant's racial group. If unknown, mark the box that identifies the mother's racial group.
14	Hispanic	Mark the box that indicates whether the infant is of Hispanic origin.
15	Food Source	Mark the box that identifies the infant's source of nutrition (breast milk only, milk formula only, soy formula only, breast milk supplemented with milk-based formula, breast milk supplemented with soy-based formula, total parenteral nutrition (TPN) or no enteral feeding (NPO).
16	Status	Mark the box that best represents the status of the infant at the time of collection for each situation or condition listed.
17	Date Last Transfused	If the infant was transfused, enter the date of the latest transfusion. This information is needed to determine if the tests for some disorders are valid.
18	Accession Number	**FOR LAB USE ONLY. DO NOT WRITE OR PLACE STICKERS IN THIS AREA.**
19	Submitter Name/ID	Enter the name of the agency/entity submitting the specimen for testing and the unique submitter ID code for that submitter. (If you do not have a submitter ID, call the Newborn Screening Demographic Entry Section at (602) 364-1468)
20	Submitter Address	Enter the mailing address of the submitter (can be omitted if the submitter ID is entered above).

21	Physician's Name (Last, First)	Enter the physician's last name and first name. This physician or other health care professional should be the provider responsible for the management of medical services provided to the infant. Also, enter the physician's NBS ID. If the infant's medical home is not known, or if the parent has not chosen a pediatrician for the infant yet, enter the attending practitioner of record. This is the physician who will receive information about test results. (If the practitioner does not have an ID, please contact the Newborn Screening Data Entry Section at (602) 542-1187.)
22	Phone	Enter the physician's phone number, including area code.
23	Physician's Address	Enter the street address of the physician named above. This is the mailing address where a copy of the test results will be mailed.
24	City, State, Zip	Please see #23 above. Enter the city, state and zip code of the physician.
25	Mom's Name (Last, First)	Enter the mother's last and first names from the medical record. If the mother does not have physical custody of the infant, enter the name of the person who has custody. See the note below*.
26	Mom's Date of Birth	Enter the mother's birth date in "mm/dd/yy" format.
27	Maiden Name	Enter the mother's maiden name (name before her first marriage).
28	Street Address	Enter the mother's address (or mailing address). If the mother does not have physical custody of the infant, enter the address of the person who has custody. See the note below*.

29	City, State, Zip	Enter the city, state and zip code of the mother's address, even if the mother does not reside in Arizona. If the mother does not have physical custody of the infant, enter the city, state and zip code of the person who has custody. See the note below*.
30	Phone	Enter the mother's telephone number, including the area code. If the mother does not have physical custody of the infant, enter the phone number of the person who has custody. See the note below*.
31	Message Phone	Enter a number where the mother can be reached if not at her own phone. Or if the mother does not have a working phone number of her own, enter a number where the mother can be contacted (friend, relative, neighbor, etc.). This is important so the mother can be reached if there are abnormal results.
32	Mom's AHCCCS #	Enter the mother's AHCCCS # if the mother is enrolled in AHCCCS.
33	Parent Refused Testing	Mark the box if the parent refuses to have the infant screened. Parents should have been informed about the consequences of not screening their infant and their refusal should be documented in the infant's medical record. Most providers will have the parents sign a refusal form that is retained in the infant's medical record. For a sample form, see the Appendices. The completed newborn screening kit without blood should be submitted to the State Lab.

*If the infant's mother does not have physical custody of the infant, enter the name, address and phone number of the person who has custody.

In the case of a baby in foster care, enter the CPS caseworker's contact information and note that this is foster care. If the caseworker's contact information is not available, enter the foster mother's name, address and phone number and note that this is foster care.

In cases of adoption, enter the information for the adoptive mother if the baby has been placed in her care. Otherwise, enter the adoption agency caseworker information and note that this is an adoption. It is extremely important to list contact information for someone who can be reached and who will be able to assume responsibility for follow-up, if necessary.

4.5 Reasons for Specimen Rejection (UNSATS)

(With codes for Arizona unsats)

Problems with the family or baby

UPR Parent refusal
Make sure that parents are aware of the consequences of refusing the test. Document the refusal in the medical record, fill out demographic information on the collection card, check the "parent refused testing" box in the lower right corner of the card and submit the card to Arizona State Lab without blood.

UIO Infant too old (> 1 year of age)
Newborn screening reference ranges apply only to newborn infants and most accurately indicate risk of disease when specimens are collected early. Serious symptoms of these diseases appear and irreversible damage is done if treatment is not started early.

Problems with the card

UCE expired collection card
CLIA regulations state that valid results cannot be reported if the card has expired.

UNI no identifying information
Results cannot be reported if the infant cannot be identified.

UNO no blood
Tests cannot be performed without blood.

Problems with collection of the blood

UCC clotted or caked blood
This happens when a large enough drop of blood is not taken and blood begins to clot before being applied to the filter paper.
Uniform amounts of blood cannot be taken from samples with extra clotted blood in some areas of the spot.

UIS insufficient specimen
The baby didn't bleed well.
Not enough blood was submitted to complete the whole panel of tests.

UMA multiple specimen applications
Blood may have been applied to both sides of the filter paper or with overlapping drops of blood instead of one big drop in each circle. Uniform amounts of blood cannot be taken for testing.

UNS uneven saturation
With areas more saturated with blood than other areas of the spot instead of one big drop in each circle, uniform amounts of blood cannot be taken for testing.

UTS torn or scratched
This usually happens when capillary tubes are used to collect the blood and then it is spotted onto the filter paper. The rough edges of the tubes can easily abrade the filter paper if they are dragged along the surface. With areas where filter paper is missing or bunched up, uniform amounts of blood cannot be taken for testing.

UST serum or tissue fluid separation
This can happen if the first drop of blood is not wiped away, if a liquid contaminant is allowed to come in contact with the blood spots, if the area around the puncture site is squeezed excessively ("milking"), or if the specimen is dried vertically so that gravity can separate blood components before complete drying.

Problems in handling and shipping

UTO specimen too old (>14 days from collection date)
Blood components degrade with age and exposure to heat and light and cannot be accurately measured.

USD specimen detached from form
Results cannot be reported if the infant may be incorrectly identified.

USC contaminated
Filter paper has come in contact with gloved or ungloved hands or substances such as alcohol, formula, antiseptic solutions, water, hand lotion or powder, etc. before or after specimen collection. Contaminated specimens yield unreliable results.

Problem in the lab

URS results inconsistent
Repeat testing of the same sample gives results that are greatly different from one another. This could indicate that the sample was unevenly saturated or damaged in some way.

A one-page summary of these reasons for specimen rejection is found in the Appendices.

4.6 Specimen Collection and Handling

4.61 Collection and Handling of First Screens

You can collect a perfect specimen but if it wasn't collected during the recommended screening window, or if it was delayed in transport to the State Lab, a baby could suffer.

When is the best time to collect a first specimen? 24 to 36 hours of age – earlier is now better.

1. **Best screening window**
   Each disorder has a "best screening window" where the test is most likely to detect affected infants while minimizing false positive results. These windows are much earlier than those recommended in the past because the disorders detected by tandem mass spectrometry can be accurately detected by 24 hours of age and some of them can harm babies in the first few days of life (so earlier screening is needed).

2. **Arizona Rules**
   The Arizona Administrative Code R9-13-204 states that a first specimen must be collected between 24 and 72 hours of age, but collection closer to 24 hours is recommended. For all disorders except homocystinuria, ideal screening windows include the time between 24 and 48 hours of age. Waiting until 72 hours is already past the best window for many disorders.

3. **Special Considerations**
   Sick, preterm and low birth weight babies have their own complications for screening and even earlier collection is recommended to avoid interference from therapies like transfusion, parenteral nutrition or treatment with steroids and some antibiotics.

How can delays in transport to the State Lab be prevented?

1. **Prompt Mailing**
   The Arizona Administrative Code R9-13-203 A. 4 states that specimens must be sent to the State Lab within 24 hours of collection or the next business day.

2. **Hospitals using FedEx (State paid), Courier or US Mail**
   Each hospital should identify someone with the responsibility to ensure that samples move quickly from the nursery to the lab and shipping area so that

the whole process from collection to shipping takes 24 hours or less. The practice of "batching", where a facility delays sample shipment until a predetermined number of samples is collected, seriously increases the risk of irreversible harm or death for those infants born earliest in the batch. Therefore, any hospital with at least one birth a day should send a shipment of samples to the State Lab each weekday, including Friday.

3. **Other Providers**
   For those providers who don't attend at least one birth a day, the requirement is still to ship as promptly as possible. A single specimen sent alone is acceptable and shouldn't be held to be sent with others collected on a later date.

4.62 Collection and Handling of Second Screens

1. **When is the best time to collect a second specimen?** 5 to 10 days of age or at the first outpatient visit to the doctor.

   Since babies often see their primary care physicians prior to 5 days of age, second screens should be collected at that visit, even though the baby is not yet 5 days old.

   Second specimens can be collected after 10 days of age (and will be tested if collected up to a year of age). However, later-collected specimens may miss cases of certain metabolic disorders (e.g., VLCAD, MCADD, GA-1) or cystic fibrosis because false negatives are more likely in older babies. For disorders where symptoms are not apparent until irreversible damage has been done (PKU, congenital hypothyroidism) later detection would mean a poorer outcome for an affected infant.

2. **For providers who collect their own second screens**:

   Collect the specimen at the baby's first visit and send it to the State Lab as soon as it is dry. A single specimen is acceptable and shouldn't be held to be sent with others collected on a later date.

   Protect specimens from heat and sunlight. Don't put them in metal collection boxes on outside doors during the summer or drop them in an outside mailbox in the sun.

3. **For providers who send patients to an outside lab**:

   Please direct your patients to get the second screen collected as soon as possible.

We have heard that some labs are turning families away if the baby is younger than 2 weeks of age. This should not happen since earlier collection is now recommended. The Newborn Screening program will be working with the large contract labs to ensure that all of their drawing stations have the latest guidelines. Please let us know if you hear of any parents that have been turned away.

The bottom line is that early collection, with no delays in handling or shipping, gives the best chance for a disorder to be detected and treated before an infant is permanently harmed.

4.7 Special Considerations

For a baby in the NICU a first bloodspot specimen should be collected prior to transfusion, parenteral nutrition and other therapies, even if it means that the specimen will have to be collected prior to 24 hours of age.

The CLSI Guideline for preterm, low birth weight and sick newborns recommends collection of the first screen on admission to the NICU so that it can be obtained prior to any intervention. This guideline recommends a second screen between 48 and 72 hours of age and a third on day 28 or prior to discharge, whichever comes first.

Arizona rules do not have modified requirements for NICU collection. However, they do require the hospital to collect the second screen between five and 10 days of age and before discharge if a baby is still hospitalized at 5 days of age.

If a baby is transferred to another hospital prior to 48 hours of age, the receiving hospital is required to collect a first screen, unless the first screen has been collected by the sending hospital.

4.8 Factors Which Affect Test Results or Interpretation

1. Transfusions

According to the Arizona Administrative Code (R9-13-204A), the first newborn screening test should be collected prior to any transfusion, unless specified otherwise by a physician. This is necessary to avoid false negative results for many disorders.

Any transfusion of red blood cells (whole blood, packed RBCs or ECMO) can cause false negative results for galactosemia and hemoglobinopathies and this effect lasts until the donor red blood cells have been replaced (3 – 4 months after the last transfusion). Rescreening then would be necessary

unless a previous screen had been collected prior to transfusion. For both of the above disorder groups a specimen collected prior to 24 hours of age is valid and yields reliable results.

Transfusions of whole blood or ECMO (or plasma, for biotinidase deficiency screening) can cause false negative results for all screened disorders for 4 – 72 hours after the transfusion (except for galactosemia and hemoglobinopathies where the effect lasts for 3 – 4 months).

2. Parenteral Nutrition (PN)

Parenteral nutrition can cause false positives for amino acids and fatty acids. Multiple amino acid abnormal results can be an indication of excess free amino acids from the parenteral nutrition solution or liver problems (immature enzymes or illness so that liver enzymes can't handle the amino acids fast enough to prevent a rise in amino acid concentration in the blood). Medium chain fatty acids are also added to parenteral nutrition solutions and can be present in higher amounts in the blood. Prolonged PN can lead to carnitine depletion. A false positive result for IVA (elevated C5) is also possible.

A repeat screen should be collected 24 – 72 hours after PN is stopped, if a previous screen had abnormal results.

3. Maternal Conditions

A mother with hyperthyroidism treated with PTU can deliver a baby with transient hypothyroidism (elevated TSH on the newborn screen). Positive results will occur until the drug clears the newborn's system - between 7-14 days after birth. A repeat screen or other thyroid testing should be done around two weeks of age.

A mother with CAH can deliver a baby with a false positive result for 17-OHP. The newborn should be rescreened between 3 and 7 days after birth.

Transient hyperphenylalaninemia in the newborn is a result of a mother with uncontrolled PKU (high phenylalanine levels). This effect will normalize within 12 – 24 hours, unless the baby also has PKU.

A mother treated with steroids during pregnancy can deliver a baby with a false negative result for CAH since steroids can suppress fetal adrenal function. The length of the effect depends on the class of steroid and the dose and is unknown but estimated at 1 – 2 weeks after birth. A repeat screen done later than 2 weeks of age would be needed.

Maternal carnitine or Vitamin B_{12} deficiencies can cause false positive results for C0 (carnitine) and C3 (Vitamin B_{12}). The effects can last several days

depending on the nutrition provided to the newborn (for B_{12} deficiency) – the duration of the effect of carnitine deficiency is unknown).

Carnitine supplementation can cause false negative results for C0 (Carnitine uptake defect) during supplementation and for some weeks afterwards. It can also cause false positives for other acylcarnitines. This effect lasts approximately 4 days.

A mother with 3MCC can have an unaffected baby with elevated C5OH. The duration of this false positive effect is unknown.

4. **Sick or stressed infant**

   A sick or stressed newborn can have elevated 17-OHP and IRT (false positives for CAH and CF) until recovered.

   Liver disease and jaundice can cause false positives for many disorders (tyrosinemia, homocystinuria, PKU, CF, biotinidase deficiency)

5. **Preterm or low birth weight infant**

   Elevated tyrosine and 17-OHP and low biotinidase are common results for preterm or low birth weight babies (false positives for tyrosinemia, CAH and biotinidase deficiency).

   False positives for amino acids disorders are a result of immature liver enzymes.

   A false negative result for hypothyroidism caused by an immature hypothalamic/ pituitary/thyroid axis where TSH does not rise in response to low T4 levels can last for more than a month after birth. Since the newborn screen now measures only TSH, this possibility should be considered for all preterm and low birth weight babies, even though the hypothyroidism is usually transient.

6. **Steroid or antibiotic treatment**

   Dopamine therapy suppresses TSH and can cause false negative results for hypothyroidism until the drug is discontinued.

   Steroid therapy (including dexamethasone) suppresses TSH and can cause false negative results for hypothyroidism as well as false negative results for CAH. This effect can last for 1 to 2 weeks after therapy has been stopped.

Antibiotics conjugated with pivalic acid (for example, pivampicillin) can elevate C5 (false positives for IVA). This effect lasts until the drug clears the baby's system (at least 24 hours after discontinuing therapy).

7. Early collection (prior to 24 hours of age)

False positives for hypothyroidism and CAH are possible because of the normal hormone surge after birth.

False negatives for amino acidopathies and organic acid disorders are possible with early collection but specimens collected shortly after 24 hours of age are reliable.

8. Late collection

Collection of a first screen after 48 hours of age can show false negative results for fatty acid oxidation disorders. A well fed state can mask indications of a FAOD so it is important that a first screen be collected between 24 and 36 hours if at all possible.

If a baby has an abnormal result for any FAOD on a first screen and then has a normal result on the second, that second result cannot be taken to mean that the baby had a false positive on the first screen. All babies with abnormal results on first screens should have diagnostic testing done to confirm or rule out the possibility of a disorder even though their second screen is "normal." Very few false positive results for fatty acid oxidation disorders ever occur.

Late collection is also not helpful in identifying disorders that have early crises since results will not be available when symptoms start to appear. Disorders which can have serious consequences if not diagnosed early include galactosemia, MSUD, salt-wasting CAH, urea cycle disorders, organic acid disorders and some fatty acid oxidation disorders.

Please see a table in the Appendices summarizing the factors which affect screening results, including timing of the specimen collection.

4.9 Parent Refusal of Screening

It is important to convince parents of the value of newborn screening so that they get their babies screened during the best window of time to detect the disorders.

Providers are required to order newborn screening tests but parents may refuse testing. If after explaining the benefits of newborn screening and the risks involved in refusing testing, and parents still wish to refuse, they should sign a waiver in which they accept responsibility for adverse consequences.

You should check to see that the waiver you ask parents to sign has been reviewed and approved by your legal counsel (See the Appendices for a sample form available as a template for preparing a customized form for your use).

This refusal should be fully documented in the baby's medical record. The demographic information on a newborn screening kit should be fully filled out and submitted to the State Lab with no blood and the "Parent Refused Testing" box in the lower right hand corner of the form checked.

CHAPTER 5
Hearing Screening and EHDI

5.1 Hearing Screening

In Arizona approximately 98% of all infants are screened for hearing loss prior to initial hospital discharge. Both Otoacoustic Emissions (OAE) and Automated Auditory Brainstem Response (AABR) screening technologies are used, sometimes in combination. Nearly all neonatal intensive care units screen with AABR because of the increased risk of auditory neuropathy in this population.

In Arizona 3% of infants do not pass the inpatient screen. Those infants in the well baby nursery who do not pass the newborn screen should also receive one outpatient screen within the first week or two post-discharge. Because of the tenfold increased incidence of hearing loss in neonatal intensive care graduates, this population should NOT receive an outpatient screen but should go directly to a pediatric audiologist for a diagnostic evaluation.

5.2 Arizona Early Hearing Detection and Intervention (AzEHDI) Program Goals

The goals of the Arizona Early Hearing Detection and Intervention (AzEHDI) program are:

- screening all babies for hearing loss by one month of age
- completing diagnostic testing by a pediatric audiologist before three months of age for babies who fail the newborn hearing screen
- enrollment in early intervention services as soon as possible (prior to six months of age) after diagnosis of hearing loss

AzEHDI is a collaborative program with all agencies, organizations, providers and parents involved in the early hearing detection and intervention efforts in Arizona. Various aspects of the program are funded through federal grants, foundation grants, state funding and donations. The AzEHDI efforts are coordinated through the Arizona Department of Health Services, Newborn Screening Program.

5.3 A message from the NBS Program Manager

Dear Health Care Professional,

This notification is a reminder of the Arizona reporting requirements for newborn hearing screening and subsequent tests. Arizona Revised Statute §36-694, passed in 2005, mandates the reporting of all hearing screening and diagnostic results for both newborns and infants to Arizona Department of Health Services, Office of Newborn Screening.

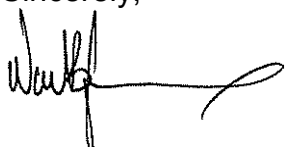
As prescribed by A.A.C. R9-13-207 (E), all screening and diagnostic hearing evaluations shall be submitted within on e week following the hearing test, **even if the results are normal**. You can report this information by:

Fax (the fastest way): (602) 364-1495
Mail:
Arizona Department of Health Services
Office of Newborn Screening
250 N 17th Ave, 1st Floor
Phoenix, AZ 85007-3242

Both screening and diagnostic reporting forms are available on the Office of Newborn Screening website, www.aznewborn.com . Please review the instructions to determine the proper reporting format. We request a copy of the supporting narrative report for any diagnostic evaluation. If you have any questions about the reporting forms or need assistance to determine if a report should be made, please contact us at 602-364-1409 or toll-free at 1-800-548-8381.

Our goal is to ensure that infants who fail their hearing screens receive prescribed treatment of intervention services in a timely manner. Your role in the Early Hearing Detection and Intervention process is fundamental to reducing the number of babies who are lost to follow-up or lost to documentation.

Sincerely,



Ward Jacox, Chief
Office of Newborn Screening

5.4 Provider Responsibilities - Mandatory Reporting

- Results of all hearing screening and subsequent tests for hearing loss performed on newborns and infants must be reported to the Newborn Screening Program, along with identifying information for the baby and the facility performing the test. Please see the Appendices for the complete reporting requirements in Rules (R9-13-207).

- Results from each hearing screening or diagnostic test performed must be sent to the Newborn Screening Program within one week of the test date.

- All tests, even if normal, should be reported. Normal results help the Newborn Screening Follow-up team know when the child no longer needs active follow-up.

5.41 Screening Forms

To report any out-patient hearing screening, please submit the Hearing Out-Patient Screening (HOPS) form or the Community Health Center Out-Patient Screening Clinic Form found in the Appendices.

- Use for reporting screening only (not diagnostic results)
- Multiple infants can be reported on the same form
- Include the mother's identifying information
- Report the results of either the Otoacoustic Emissions (OAE) screening or the Automated Auditory Brainstem Response (AABR) screening

5.42 Diagnostic Testing Forms

Use the referral forms customized by region (Phoenix, Flagstaff, Tucson and Yuma) when referring a baby for a diagnostic test. These forms are found in the Appendices.

- Refer a child who has been diagnosed with bilateral permanent hearing loss to Early Intervention Services.
- Report diagnostic test results to the state Newborn Screening Program
- Results to be reported include:
  - Unilateral hearing loss
  - Transient hearing loss
  - Hearing loss in children from birth to three years of age and
  - NORMAL results ruling out a hearing loss

5.5 Statewide Testing Locations

Lists of screening and diagnostic testing facilities with phone numbers are available in the Appendices.

A map of these screening and diagnostic testing locations in Arizona is found on our website, www.AZNewborn.com

5.6 Hearing Screening and Follow-up

The Arizona Early Hearing Detection and Intervention Program (EHDI) includes Newborn Hearing Screening Follow-up under the Newborn Screening Program. Follow-up is conducted to meet the goals of:

- Screening all infants for hearing loss by one month of age
- Completing diagnostic testing before three months of age for children who fail the newborn hearing screen
- Enrollment in Early Intervention services as soon as possible (prior to 6 months of age) after diagnosis of hearing loss

In order for the Program to know where infants are in the process of early identification and to provide appropriate follow up, it is critical that providers know when and how to report screening and diagnostic results.

Screening
If infants do not pass their newborn hearing screen in the hospital, it is important that the babies are screened again as soon as possible (no later than 1 month of age). Most can return to the birthing hospital for an outpatient rescreen within the first week or two after discharge. If it is not possible or convenient (or for insurance coverage), the provider can refer the infant to another screening location (for a list of resources, please see the Appendices or visit our website at www.AZNewborn.com).

If the infant was screened with the AABR, it is best if AABR is used for the rescreening.

NICU graduates (more than 5-day stay) or infants with risk indicators for hearing loss:

- Do not rescreen
- Refer to a pediatric audiologist for diagnostic testing
- Older infants may need sedation/anesthesia

Diagnostic testing
Refer to a pediatric audiologist if the infant fails an outpatient screen or is a NICU graduate that failed the inpatient screen. Infants may need a referral from the primary care provider.

5.61 Late Onset and Progressive Hearing Losses

Between the newborn period and school age the prevalence of significant hearing loss doubles. This increase in hearing loss is due to:

- Late onset losses
- Progressive losses
- False negative screens
- Missed newborn screens or loss to follow-up

Although some children present with hearing loss and have no risk factors, there are conditions known to cause hearing loss. The Joint Committee on Infant Hearing recommends that these children who pass their hearing screening but have risk factors receive, at minimum, a diagnostic assessment by 24 to 30 months of age. Risk indicators that are marked with an asterisk are of greater concern for delayed-onset hearing loss and should receive earlier and/or more frequent assessment.

- Caregiver concern regarding hearing, speech, language, or developmental delay.
- Family history* of permanent childhood hearing loss.
- Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO*, assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.
- In utero infections, such as CMV*, herpes, rubella, syphilis, or toxoplasmosis.
- Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
- Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
- Syndromes associated with hearing loss or progressive or late-onset hearing loss*, such as neurofibromatosis, osteopetrosis, and Usher syndrome. Other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.
- Neurodegenerative disorders*, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
- Culture-positive postnatal infections associated with sensorineural hearing loss*, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.

- Head trauma, especially basal skull/temporal bone fractures* that requires hospitalization.
- Chemotherapy*.

5.7 Hearing Screening Guidelines

The following guidelines are found in the Appendices:

- Newborn Screening Diagnosis and Intervention Guidelines for Medical Home Providers, 2010
- Joint Committee on Infant Hearing, AAP, Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs

The Arizona Pediatric Audiology Guidelines (last published in 2000) are currently under revision.

5.8 Family Checklists – Infant Hearing Guides

Family Checklists have been developed as a type of "roadmap" for parents and providers of children who have failed their newborn hearing screening tests. The checklists include developmental milestones, a timeline for when various assessments should be completed and the appropriate next steps.

These checklists may be given to families at the hospital and are included with letters to parents sent from the Office of Newborn Screening – Follow-up:

- A Family's Checklist – Infant Hearing (intended for well babies)
- Family Checklist for Babies at High Risk for Hearing Loss (intended for NICU graduates or at higher risk for hearing loss)

Both checklists are found in the Appendices.

5.9 Provider Resources

- The Newborn Hearing Screening Training Curriculum – Streaming Video (a competency-based training for new hearing screeners) is available from NCHAM at: http://www.infanthearing.org/nhstc_dvd/streaming.html

- NCHAM Newborn Hearing Screening Training Curriculum Scripts from the above curriculum are found in the Appendices.

- CDC Early Hearing Detection & Intervention (EHDI) Program website is found at http://www.cdc.gov/ncbddd/ehdi/default.htm

- Early Hearing Detection and Intervention – AAP Medical Home Implementation is found at http://www.medicalhomeinfo.org/how/clinical_care/hearing_screening

- National Center for Hearing Assessment and Management (NCHAM) – Utah State University website is found at http://www.infanthearing.org

- Loss & Found – What to do if your baby didn't pass the newborn hearing screening - a DVD from Hands & Voices in both English and Spanish with subtitles for the Deaf/Hard of Hearing. Call the Newborn Screening Program at (602) 364-1409 to receive a free copy to show to parents.

5.91 Family-Friendly Resources

- Universal Newborn Hearing Screening Brochure from ADHS, Office of Newborn Screening is available at no charge. A copy of it and the brochure order form are found in the Appendices.

- Frequently Asked Questions Parents May Ask (in English and Spanish) from the NCHAM curriculum can be found in the Appendices.

- A checklist "How does your child hear and talk" from the NCHAM curriculum can be found in the Appendices.

- A copy of the Hands & Voices brochure with contact information is found in the Appendices.

- The Guide By Your Side brochure provides information for parents on a program of the Arizona Chapter of Hands & Voices. GBYS is a program of trained experienced parents who can mentor new parents through the screening and diagnostic process. A copy is in the Appendices.

- The Guide By Your Side rescreen reminder card is a small pink postcard sized reminder that an infant who failed the hearing screen needs to be rescreened as an outpatient within two weeks. The pink card has information in English on one side and Spanish on the other that indicates that a Parent Guide will be contacting the family and how to opt out if they prefer not to be contacted. A copy is in the Appendices.

- The EAR Foundation website is at http://www.earfoundationaz.com

- A copy of “Hearing Aids – Resources for Parents” is found in the Appendices.

- A copy of the HEAR for Kids: Loaner Hearing Aids, Vouchers and Permanent Hearing Aid Application is found in the Appendices.

- The Hear and Now website is at http://www.hearandnow.org

- My Baby’s Hearing website from the Boys Town National Research Hospital and NIDCD is at http://www.babyhearing.org/Audiologists/index.asp

- AzEHDI is on Facebook at http://www.facebook.com/pages/Glendale-AZ/AZEHDI/162338672816

- Arizona Health Care Cost Containment System (AHCCCS): Arizona’s Medicaid agency that provides health care to Arizonans that meet income and other eligibility requirements.
  http://www.azahcccs.gov

  Children’s Rehabilitative Services (CRS) is a program administered by AHCCCS through a contract with Arizona Physicians IPA. CRS provides medical treatment, rehabilitation and related support services for children under age 21 with qualifying chronic and disabling conditions.

  http://www.myapipa.com/en/members/refer_child.jsp?xlang=en&xrole=members&xstate=az&xplan=uhcaz&xproduct=CRS

- Arizona Early Intervention Program (AzEIP): a statewide system of support and services for families of children, birth to 3, with disabilities or developmental delays.
  https://egov.azdes.gov/CMSInternet/main.aspx?menu=98&id=3026

- Arizona State Schools for the Deaf and the Blind (ASDB): provide education for children with hearing or vision loss.
  http://www.asdb.state.az.us

CHAPTER 6
Resources and Education

6.1 Brochures and Fact Sheets

Copies of the brochures and fact sheets published by the Office of Newborn Screening are found in the Appendices. These can be ordered by providers at no cost using the order form in the Appendices.

6.2 Specimen Collection Resources

Review the section of the website, www.AZNewborn.com, Bloodspot Screening – Specimen Collection Helps for color copies of posters describing proper specimen collection and common collection mistakes. Black and white versions are in the Appendices.

A one page description of common collection mistakes entitled, Specimen Rejection Quick Reference is found in the Appendices.

6.21 Bloodspot Collection Training DVD

The Clinical and Laboratory Standards Institute (CLSI) publishes a guideline for specimen collection (LA4-A5, Vol. 27 No. 20: Blood Collection on Filter Paper for Newborn Screening Programs; Approved Standard - Fifth Edition).

It is accompanied by a DVD entitled, "Making a Difference Through Newborn Screening: Blood Collection on Filter Paper." Copies of this DVD are available for loan to submitters for staff training. Check with your hospital education department to see if a copy is available.

The first 2 minutes are an introduction to the history and importance of Newborn Screening and the disorders screened. Note that Arizona does not screen for toxoplasmosis.

The instructions on proper specimen collection and handling are about 23 minutes long. This is followed by a family's story of a son born with homocystinuria (approximately 5 minutes long).

If you would like to borrow a copy, please contact the Arizona Newborn Screening Program:

Wendie Jenkins, Newborn Screening Educator
jenkinw@azdhs.gov or (602) 364-1407

6.3 Links to Additional Resources

Below are lists of links to websites for other organizations with information about newborn screening, specific disorders, support groups and other resources for providers and parents.

- **Newborn Screening and Genetics**

National Newborn Screening & Genetics Resource Center (NNSGRC): a cooperative agreement between the Maternal and Child Health Bureau (MCHB), Genetics Services Branch and the University of Texas Health Science Center at San Antonio, Department of Pediatrics that provides information and resources in the area of newborn screening and genetics.
http://genes-r-us.uthscsa.edu

American College of Medical Geneticists (ACMG): the professional organization for biochemical, clinical, cytogenetic, medical and molecular geneticists, genetic counselors and other health care professionals committed to the practice of medical genetics. ACMG provides education, resources and a voice for the medical genetics profession:
http://www.acmg.net

ACT sheets and confirmatory algorithms for professionals:
http://www.acmg.net/AM/Template.cfm?Section=ACT_Sheets_and_Confirmatory_Algorithms&Template=/CM/HTMLDisplay.cfm&ContentID=5127

Genetics Home Reference: a website offering a guide to understanding genetic conditions from the U.S. National Library of Medicine within NIH, HHS:
http://www.ghr.nlm.nih.gov/search?query=%22newborn+screening%22

March of Dimes: a non-profit organization advocating pregnancy and baby health (preventing birth defects, premature birth and infant mortality) and universal newborn screening:
http://www.marchofdimes.com/professionals/24279.asp

National Center on Birth Defects and Developmental Disabilities (CDC) – Infant topics discuss newborn screening and EHDI, sickle cell disease and thalassemia.
http://www.cdc.gov/ncbddd/index.html

Gene Tests: a publicly funded medical genetics information resource hosted at NCBI:
http://www.genetests.org

Human Genetics and Medical Research – an on-line exhibit for families from the Museum of Medical Research:
http://history.nih.gov/exhibits/genetics

HealthFinder.gov: a quick guide to healthy living provided by the US Dept. of Health & Human Services with information for parents about newborn screening:
http://www.healthfinder.gov/prevention/ViewTopic.aspx?topicID=57
(includes a toll-free number to call to get services if you don't have insurance)

- **Disorder Information**

Screening, Technology, and Research in Genetics (Star-G): a multi-state project to improve information about the financial, ethical, legal, and social issues surrounding expanded newborn screening and genetic testing. Fact sheets for parents about screened disorders are available on their website:
http://www.newbornscreening.info

Genetic Alliance: a nonprofit health advocacy organization committed to transforming health through genetics. Their network includes more than 1,000 disease-specific advocacy organizations as well as universities, private companies, government agencies and public policy organizations. Their resource repository contains information on newborn screening as well as other topics in the area of genetics:
http://www.geneticalliance.org
Resource Repository at http://resourcerepository.org

Genetics Home Reference: a website offering a guide to understanding genetic conditions from the U.S. National Library of Medicine within NIH, HHS. Newborn screening disorder information is available at:
http://www.ghr.nlm.nih.gov/search?query=%22newborn+screening%22

GeT-EQuIP (Genetic Testing Electronic Quality Information Portal): from the Division of Laboratory Systems of CDC that provides a resource to facilitate information searches about genetic diseases detected through newborn screening:
http://wwwn.cdc.gov/dls/genetics/getequip/portal.aspx

MedlinePlus: a service of the US National Library of Medicine and the National Institutes of Health that provides health information on a wide range of topics and in multiple languages:
http://www.nlm.nih.gov/medlineplus
http://medlineplus.gov
http://www.nlm.nih.gov/medlineplus/newbornscreening.html

Family Village: an on-line resource for parents of children with disabilities or chronic health conditions:
http://www.familyvillage.wisc.edu/library.htm

Medical Home Portal: a service of the University of Utah and part of a Medical Home implementation project to improve care of children with chronic conditions:
http://www.medicalhomeportal.org/newborn

PerkinElmer Genetics: a service of PerkinElmer®, provider of newborn screening testing kits, instruments and reagent:
http://www.perkinelmergenetics.com/DisordersScreened.htm

March of Dimes: a non-profit organization advocating pregnancy and baby health (preventing birth defects, premature birth and infant mortality) and universal newborn screening:
http://www.marchofdimes.com/professionals/14332_15455.asp

National Organization for Rare Disorders (NORD): maintains a database of rare diseases which can be accessed at:
http://www.rarediseases.org/search/rdblist.html
http://www.rarediseases.org

Online Mendelian Inheritance in Man® (OMIM): a database of the Johns Hopkins University housed on the NCBI website that provides information on human genes and genetic phenotypes along with links to other genetics resources:
http://www.ncbi.nlm.nih.gov/omim

- **Support Groups and Disorder Information by Disorder**

Endocrine Disorders

American Thyroid Association: a professional society of physicians and scientists who specialize in the research and treatment of thyroid diseases:
http://www.thyroid.org

The Magic Foundation (Major Aspects of Growth In Children): a non-profit organization that provides support services for the families of children with disorders that affect a child's growth (including hypothyroidism and CAH):
http://www.magicfoundation.org

Cares Foundation: Congenital Adrenal Hyperplasia Research Education and Support - a foundation providing information, advocacy and support for individuals with CAH:
www.caresfoundation.org

CAH Education and Support Network: includes message boards and support groups for people with CAH:
http://www.congenitaladrenalhyperplasia.org

National Adrenal Diseases Foundation (NADF): a non-profit organization providing support, information and education to individuals with diseases of the adrenal glands (including CAH):
http://www.nadf.us/diseases/cah.htm

Hemoglobinopathies

Sickling Disorders

Sickle Cell Information Center: housed in Atlanta, GA, the Center provides sickle cell patient and professional education, news, research updates and resources:
http://www.scinfo.org

Sickle Cell Thalassemia Patients Network: a New York-based support organization for people with sickle cell disease, thalassemia and other hemoglobin disorders:
http://sctpn.org

Information Center for Sickle Cell and Thalassemic Disorders: a website from Harvard University that provides current information on sickle cell disease, thalassemia and disorders of iron metabolism:
http://sickle.bwh.harvard.edu/index.html

Sickle Cell Disease Association of America: An advocacy organization promoting a search for a cure for sickle cell disease and an improvement in the quality of health, life and services for those affected by sickle cell disease and related conditions:
http://www.sicklecelldisease.org

American Sickle Cell Anemia Association: a United Way Agency of Cleveland, Ohio that provides services to individuals and families at risk for sickle cell disease:
http://www.ascaa.org

National Coordinating and Evaluation Center for the Sickle Cell and Newborn Screening Program: SCDAA serves as the National Coordinating and Evaluation Center (NCEC) for the projects of the Newborn Screening Sickle Cell Disease initiative of HRSA/MCHB with goals of increasing knowledge about sickle cell disease, strengthening partnerships between HRSA funded community programs and partners, including state newborn screening programs and improving the quality of follow-up activities of HRSA funded projects:
http://sicklecelldisease.net

Thalassemias

Cooley's Anemia Foundation: a support organization for people with various forms of thalassemia, including Cooley's anemia or thalassemia major:
http://www.cooleysanemia.org

Metabolic Disorders

Biotinidase Deficiency

Biotinidase Deficiency Family Support Group: a non-profit, volunteer organization that supports those affected by biotinidase deficiency by providing a forum to exchange information among affected individuals and with medical professionals:
http://www.biotinidasedeficiency.20m.com

Galactosemia

Parents of Galactosemic Children, Inc. (PGC): a national, non-profit, volunteer organization providing information, support and networking opportunities to families affected by galactosemia:
http://www.galactosemia.org

PKU

National PKU Alliance: provides research, support, education and advocacy while seeking a cure for PKU:
http://www.npkua.org

Children's PKU Network: a non-profit organization promoting public awareness, education and direct assistance to help people with PKU and other metabolic disorders reach their full potential:
http://www.pkunetwork.org

National PKU News: a non-profit organization providing up-to-date, accurate news and information to families and professionals dealing with phenylketonuria:
http://www.pkunews.org

PKU Online Community: an online resource learn about PKU, managing diet, medication and other resources as well as a forum to share in chats, blogs and other programs:
http://www.pku.com

PKU Toolkit: a guide for teens or young adults with PKU:
http://www.newenglandconsortium.org/toolkit

MSUD

The MSUD Family Support Group: provides support and personal contact for those with MSUD, distributes information and raises public awareness of MSUD, supports newborn screening and research for MSUD:
http://www.msud-support.org

Fatty Acid Oxidation Disorders

FOD Family Support Group: provides emotional support, information about living with these disorders and medical updates on new developments in screening, diagnosis, research and treatment:
http://www.fodsupport.org

United Mitochondrial Disease Foundation: promotes and funds research and education for the diagnosis, treatment and cure of mitochondrial disorders and provides support to affected individuals and families:
http://www.umdf.org

Organic Acid Disorders

Organic Acidemia Association (OAA): a source for organic acidemia metabolic disorder support and information:
http://www.oaanews.org

International Organization of Glutaric Aciduria (IOGA): an international non-profit organization engaged in patient advocacy and support for individuals with GA-1:
http://helpioga.org

Propionic Acidemia Foundation: a non-profit organization funding research and providing information and support for families and medical professionals dealing with propionic acidemia:
http://www.pafoundation.com

Isovaleric Acidemia Research: a division of the Organic Acidemia Association that provides research and information for Isovaleric Acidemia families:
http://www.ivasupport.org

Urea Cycle Disorders

National Urea Cycle Disorders Foundation: a non-profit organization providing information and support for families of children with UCDs (including citrullinemia and argininosuccinic acidemia):
http://www.nucdf.org

Cystic Fibrosis

Cystic Fibrosis Foundation: the leading organization in the US devoted to cystic fibrosis. It is a nonprofit donor-supported organization that funds and accredits CF care centers, advocates for CF research and provides families with information and resources:
http://www.cff.org
Living with Cystic Fibrosis: http://www.cff.org/LivingWithCF

Internet community for CF patients and families: http://www.cysticfibrosis.com

Hearing Loss

National Center for Hearing Assessment and Management (NCHAM), Utah State University: the national resource center for the implementation and improvement of EHDI systems and the developer of the Hi*Track data management system:
http://www.infanthearing.org

Hear and Now: from NCHAM, a resource for parents:
http://www.hearandnow.org

Hands & Voices: a support organization for families of children who are deaf or hard of hearing:
http://www.handsandvoices.org
http://www.handsandvoices.org/chapters/az.htm

Guide-By-Your-Side (GBYS) – a Hands & Voices program that provides emotional support and specialized knowledge from trained parents of children who are deaf or hard of hearing:
http://www.handsandvoices.org/services/guide.htm

Joint Commission on Infant Hearing (JCIH): composed of representatives from the American Academy of Pediatrics, the American Academy of Otolaryngology and Head and Neck Surgery, the American Speech Language Hearing Association, the American Academy of Audiology, the Council on Education of the Deaf, and Directors of Speech and Hearing Programs in state health and

welfare agencies. The committee's primary activity is publication of position statements on EHDI:
http://www.jcih.org

My Baby's Hearing: a website from the Boys Town National Research Hospital supported by the National Institute for Deafness and Communication Disorders (NIDCD):
http://www.babyhearing.org

National Institute on Deafness and Other Communication Disorders: is one of the Institutes that comprise the National Institutes of Health (NIH) and supports biomedical and behavioral research in the processes of hearing, balance, smell, taste, voice, speech and language:
http://www.nidcd.nih.gov/index.asp

American Society for Deaf Children, a parent-helping-parent network that supports and educates families of deaf and hard of hearing children and advocates for high quality programs and services.
http://www.deafchildren.org

American Speech-Language-Hearing Association – the professional, scientific, and credentialing association for members and affiliates who are speech-language pathologists, audiologists, and speech, language, and hearing scientists.
http://www.asha.org

National Center on Birth Defects and Developmental Disabilities, one of the Centers for Disease Control and Prevention (CDC) - provides information about hearing loss in children.
http://www.cdc.gov.ncbddd/hearingloss/index/html

- **Advocates for Newborn Screening**

Save Babies Through Screening Foundation, Inc. (SBTS): a national non-profit organization devoted exclusively to the advocacy of newborn screening:
http://www.savebabies.org

March of Dimes (MOD): a non-profit organization advocating pregnancy and baby health (preventing birth defects, premature birth and infant mortality) and universal newborn screening:
http://www.marchofdimes.com

American Academy of Pediatrics (AAP): the professional organization for pediatricians and pediatric medical subspecialists:
http://www.aap.org

AAP Newborn Screening Information and Fact Sheets:
http://www.aap.org/healthtopics/newbornscreening.cfm
Arizona Chapter: http://www.azaap.org/general/index.htm
AAP website for parents: http://www.HealthyChildren.org

- **Arizona Resources**

Arizona Health Care Cost Containment System (AHCCCS): Arizona's Medicaid agency that provides health care to Arizonans that meet income and other eligibility requirements:
http://www.azahcccs.gov

Children's Rehabilitative Services (CRS) is a program administered by AHCCCS through a contract with Arizona Physicians IPA. CRS provides medical treatment, rehabilitation and related support services for children under age 21 with qualifying chronic and disabling conditions.

http://www.myapipa.com/en/members/refer_child.jsp?xlang=en&xrole=members&xstate=az&xplan=uhcaz&xproduct=CRS

Arizona Early Intervention Program (AzEIP): a statewide system of support and services for families of children, birth to 3, with disabilities or developmental delays:
https://egov.azdes.gov/CMSInternet/main.aspx?menu=98&id=3026

Quest to Cure: a parent support and advocacy group based in Phoenix, AZ for children with sickle cell disease:
http://www.questtocure.org

Cystic Fibrosis Foundation – Arizona Chapter:
http://www.cff.org/Chapters/arizona

http://www.phoenixchildrens.com/emily-center/child-health-topics/cysticfibrosis.html

Phoenix Children's Hospital, Pulmonology: an accredited CF care center:
http://www.phoenixchildrens.com/medical-specialties/pulmonology.html

Arizona Commission for the Deaf and Hard of Hearing: a statewide information referral center for issues related to people with hearing loss:
http://www.acdhh.org

The EAR Foundation of Arizona: a non-profit association that provides information and resources to people who are hearing impaired:
http://www.earfoundationaz.com

HEAR for Kids™: a program of the EAR Foundation of Arizona for children 0-18 years of age that provides loaner hearing aids and permanent hearing aids if the family doesn't have insurance and meets financial criteria:
http://www.earfoundationaz.com/page_010_005.html

Application for loaner hearing aids, vouchers and permanent hearing aids:
http://aznewborn.com/pdf/hc_prov_loan_hear_aids.pdf

Hands & Voices (Arizona): http://www.azhv.org

Raising Special Kids (Arizona): a non-profit organization of families helping families of children with disabilities and special health needs (602-242-4366 or 800-237-3007):
http://www.raisingspecialkids.org

Pilot Parents of Southern Arizona: a parent training and information center for families of children with disabilities in Cochise, Gila, Graham, Greenlee, La Paz, Pima, Pinal, Santa Cruz and Yuma counties:
http://www.pilotparents.org

The Emily Center at Phoenix Children's Hospital: the largest pediatric consumer health library in the Southwest located at Phoenix Children's Hospital, 1919 E Thomas Rd, Phoenix, AZ
(602) 546-1400
http://www.phoenixchildrens.com/emily-center

American Academy of Pediatrics, Arizona Chapter:
http://www.azaap.org/general/index.htm

Note:
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6.4 Glossary

#

1-3-6
: EHDI guidelines: hearing screening before one month of age, evaluation before 3 months of age and intervention before 6 months of age

A

AABR
: automated ABR hearing test

AAC
: Arizona Administrative Code

abnormal
: a test value outside the reference range, a positive result

ABR
: auditory brainstem response hearing test

ACMG
: American College of Medical Genetics

ACT sheets
: Actions sheets developed by ACMG for use by physicians as a quick reference to NBS results and needed actions for possible disorders http://www.acmg.net/resources/policies/ACT/condition-analyte-links.htm

acylcarnitine
: a fatty acyl ester of carnitine, the transport form for a fatty acid crossing the mitochondrial membrane. Acylcarnitines are named by the number of carbons in the acyl group (the side chain attached to carnitine):
  - C0: no acyl group, free carnitine
  - C3: 3 carbon acyl group attached to carnitine - propionylcarnitine
  - C5OH: 5 carbon acyl group with hydroxyl on the 3rd carbon – 3-hydroxyisovalerylcarnitine

ADHS
: Arizona Department of Health Services

AHCCCS
: Arizona Health Care Cost Containment System

American College of Medical Genetics (ACMG)
: the professional organization for biochemical, clinical, cytogenetic, medical and molecular geneticists, genetic counselors and other health care professionals committed to the practice of medical genetics. ACMG provides education, resources and a voice for the medical genetics profession. http://www.acmg.net

ACMG 29
: 29 original target disorders for newborn screening proposed by ACMG in the report entitled, Newborn Screening: Toward a Uniform Screening Panel and System (2006) and recommended by SACHDNC as the original core disorders of the Uniform Screening Panel
http://www.acmg.net/AM/Template.cfm?Section=Practice_Guidelines&Template=/CM/HTMLDisplay.cfm&ContentID=3671

analyte
: the chemical compound being tested for (e.g. the analyte for CAH testing is 17-OHP)

Arizona Administrative Code (AAC)
: where the official rules of the state of Arizona that govern state agencies, boards and commissions are published. See Rules.

Arizona Department of Health Services (ADHS)
: the agency housing the State Laboratory and the Arizona Newborn Screening Program
http://www.azdhs.gov

Arizona Health Care Cost Containment System (AHCCCS)
: Arizona's Medicaid agency that provides health care to Arizonans that meet income and other eligibility requirements
http://www.azahcccs.gov

Arizona Revised Statutes (ARS)
: The statute establishing the Arizona Newborn Screening Program is found in ARS 36-694 (Report of blood tests; newborn screening program; committee; fees; definitions)
http://www.azleg.gov/ars/36/00694.doc

Arizona State Public Health Laboratory (State Lab)
: in the Bureau of State Laboratory Services, Division of Public Health Services, ADHS – the designated laboratory for newborn screening in Arizona
http://www.azdhs.gov/lab

ARS
: Arizona Revised Statutes

ASA
: argininosuccinic acidemia/argininosuccinic aciduria, a core disorder of the Recommended Uniform Screening panel

ASDB
: Arizona State Schools for the Deaf and the Blind

ASL
: American Sign Language

assay
: a test or analysis

atresia
: congenital absence or closure of a normal body opening or tubular structure

audiology
: the branch of science that studies hearing, balance, and related disorders. Audiologists test hearing and make recommendations about treatment options (hearing aids, cochlear implants, surgery) and appropriate medical referrals

AutoDelfia®
: An automatic immunoassay system from PerkinElmer – the method used for TSH, 17-OHP and IRT testing by the State Lab

auditory brain stem response (ABR)
: a method used for hearing screening which measures the brain's response to sound. A device near the ear makes clicking sounds while earpieces in the ear canals conduct the sound. Electrode pads placed on the forehead and behind the ear track the sound moving through the ear to the brain. Results are averaged and compared with normal hearing. This method may also be used in a more extensive, modified form as a diagnostic test.

autosomal recessive inheritance
: inheritance pattern carried on a pair of the non-sex determining chromosomes (the 22 pairs of autosomes) where matching genes for a condition must be inherited from both parents
All of the disorders on the newborn screening panel except congenital hypothyroidism and hearing loss are inherited in an autosomal recessive inheritance pattern.

AzEIP
: Arizona Early Intervention Program (acronym pronounced Ay-zip)

B

BAER
: brainstem auditory evoked response hearing test

Barts
: an abnormal, usually transitory, type of hemoglobin that is not effective in oxygen transport. It contains 4 gamma globin chains. It is often present in small amounts in newborns but larger amounts indicate alpha thalassemia trait, Hemoglobin H disease or alpha thalassemia.

BCAA
: branched chain amino acids (leucine, isoleucine, valine)

BIO
: biotinidase deficiency, a core disorder of the Recommended Uniform Screening panel

Bio-Rad
: Bio-Rad Laboratories, supplier of the kit for the galactose-1-phosphate uridyl transferase assay to screen for galactosemia. http://www.bio-rad.com

BKT
: beta-ketothiolase deficiency, a core disorder of the Recommended Uniform Screening panel

borderline
: for some tests, an equivocal or borderline range is reported. It is outside the reference range but is not frankly abnormal. The program follows these results but not as immediately or aggressively (e.g. borderline TSH or 17-OHP results)

branched chain amino acids
: leucine, isoleucine and valine

C

CAH
: congenital adrenal hyperplasia, a core disorder of the Recommended Uniform Screening panel

carrier
: a heterozygote for a recessively inherited disorder where two copies of the disease-causing gene must be present in order for the disease to be expressed. Carriers or those with traits usually have no symptoms of disease or milder symptoms under stressful conditions.

CBAVD
: congenital bilateral absence of the vas deferens, a common form of sterility in males with CF or CFTR mutations

Cbl A, B
: methylmalonic acidemia (cobalamin defects), a core disorder of the Recommended Uniform Screening panel

CDC
: Centers for Disease Control and Prevention – an agency of HHS

Centers for Disease Control and Prevention (CDC)
: the agency of HHS providing community health protection through health promotion, prevention of disease, injury and disability and preparedness for new health threats.
http://www.cdc.gov
http://www.cdc.gov/newbornscreening

CF
: cystic fibrosis, one of the 30 core disorders of the Recommended Uniform Screening panel

CFR
: Code of Federal Regulations

CFTR gene
: cystic fibrosis transmembrane conductance regulator gene

CH
: congenital hypothyroidism, a core disorder of the Recommended Uniform Screening panel

CIT-1
: citrullinemia, type I, a core disorder of the Recommended Uniform Screening panel

CLIA
: Clinical Laboratory Improvement Act of 1965 (Amendments 1988)

Clinical and Laboratory Standards Institute (CLSI)
: the developer and publisher of LA4-A5, Blood Collection on Filter Paper for Newborn Screening Programs; Approved Standard – Fifth Edition (July, 2007), I/LA27-A, Newborn Screening Follow-up; Approved Guideline (May, 2006) and I/LA31-A, Newborn Screening for Preterm, Low Birth Weight, and Sick Newborns: Approved Guideline (October, 2009) www.clsi.org

clinical diagnosis
: diagnosis based on signs, symptoms and laboratory findings

Clinical Laboratory Improvement Act of 1965 (CLIA) with amendments in 1988
: In order to ensure quality laboratory testing, the Centers for Medicare & Medicaid Services (CMS) in HHS regulates all laboratory testing (except research) performed on humans in the U.S. through the Clinical Laboratory Improvement Amendments (CLIA). In total, CLIA covers approximately 200,000 laboratory entities including the Arizona State Laboratory. http://www.cms.hhs.gov/CLIA/01_Overview.asp#TopOfPage

CLSI
: Clinical and Laboratory Standards Institute

CMV
: cytomegalovirus

collection device
: another name for collection kit, filter paper kit, dried bloodspot card or newborn screening card (sometimes called the Guthrie card in honor of its developer)

collection kit
: the numbered card with special filter paper attached for collection of drops of blood that are then dried before shipping and testing. It has areas for demographic information, accession number, date stamp, bar code, etc. Each state designs their own card but all contain the same standardized filter paper

confirmatory/diagnostic test
: a test to show whether a condition suspected because of screening test results is present or not (for screening using dried blood spots, this testing is from a specimen other than the screening specimen).

congenital
: present from birth, but not necessarily genetic

congenital bilateral absence of the vas deferens (CBAVD)
: a common form of sterility in males with CF or CFTR mutations

Cooley's anemia
: thalassemia major, ß° thalassemia

CRS
: Children's Rehabilitative Services

CUD
: carnitine uptake defect, a core disorder of the Recommended Uniform Screening panel

cutoff
: the first value outside the reference range (the point where a result is reported as abnormal or positive for the disease tested for)

Cystic fibrosis transmembrane conductance regulator gene (CFTR gene)
: Mutations to this gene can cause cystic fibrosis (over 1000 mutations to this gene have been identified). The Arizona NBS program uses a DNA kit from Third Wave Technologies to detect 46 mutations to this gene.

D

DBS
: dried blood spot

DHHS
: United States Department of Health and Human Services

diagnosis
: identification of a disease by history, lab tests and symptoms

dietary monitoring specimen
: To monitor phenylalanine levels so that diets can be adjusted for individuals with PKU, regardless of their ages, the State Lab receives specimens and reports phenylalanine levels only, at no charge

DOB
: date of birth

DOC
: date of collection (of NBS sample)

DOR
: date of receipt of NBS sample by the State Lab

DPOAE
: distortion product otoacoustic emissions

dried blood spot
: blood collected usually from a heel stick and air dried on an approved filter paper card

Duarte variant of galactosemia
: a usually benign form of galactosemia (D/G variant) where an individual inherits one gene for galactosemia (no enzyme activity) and one Duarte gene with reduced enzyme activity

E

ECMO
: extracorporeal membrane oxygenator – similar to a heart/lung bypass machine

EHDI
: Early Hearing Detection and Intervention (acronym pronounced “eddy”). The Arizona EHDI program coordinates newborn hearing screening and intervention in the state.

EI
: early intervention

ELBW
: extremely low birth weight

endocrine
: relating to or affecting those glands that secrete their products (hormones) directly into the bloodstream

enteral
: within or by way of the intestine

enzyme
: a protein produced by living organisms that catalyzes (increases the rate of) a specific chemical reaction but is not consumed in the reaction or altered upon completion of the reaction

  Enzymes are inactivated by heat and/or humidity and both enzyme assays (for biotinidase and GALT activity) in the newborn screening panel can have false positive results particularly in the summer if samples have been exposed to heat, strong sunlight and/or humidity.

extracorporeal membrane oxygenator (ECMO)
: similar to a heart/lung bypass machine, ECMO can be considered to be a continuous transfusion of red blood cells and other blood components and any NBS samples taken during ECMO are invalid.

extremely low birth weight (ELBW)
: birth weight less than 1000 g

F

false negative
: a negative (normal) result in an affected individual (one who has the disease)

false positive
: a positive (abnormal) result in an individual without the disease

fatty acid
: an organic saturated or unsaturated carboxylic acid containing a single carboxyl group and usually an even number of carbon atoms that can be combined with glycerol to form fats (glycerol plus 3 fatty acids make a triglyceride).

fatty acid oxidation
: stepwise catabolism of fatty acids in which two-carbon fragments are successively removed from the carboxyl end of the chain

fatty acid oxidation disorders (FODs)
: a group of genetic metabolic disorders in which the body is unable to oxidize (breakdown) fatty acids to make energy because an enzyme is either missing or not working correctly. When the supply of glucose, the main source of energy for the body, runs out, fat is broken down to supply energy but this can't happen in individuals with one of these disorders. Support group: FOD Family Support Group http://www.fodsupport.org

FFP
: fresh frozen plasma

filter paper
: special standardized filter paper has been designed to collect blood spots for newborn screening in the circle so that punches from the circle will each contain the same amount of blood.

filter paper kit
: another name for a specimen collection kit

first screen
: the initial specimen collected from a newborn who is less than five days of age

follow-up
: actions taken to ensure that a newborn whose screening test results are positive or unsatisfactory receives appropriate, prompt further testing and evaluation; and actions taken to ensure that the newborn screening system can evaluate the effectiveness of screening.

fresh frozen plasma
: Transfusion of this blood product can produce false negative newborn screening results for biotinidase testing. The effect is transitory and a valid repeat screen can be collected a few days to a week after transfusion.

G

GA-1
: glutaric acidemia type 1, a core disorder of the Recommended Uniform Screening panel

galactose
: a simple sugar (an isomer of glucose) which in combination with glucose forms lactose
http://www.galactosemia.org

GALT
: galactosemia, a core disorder of the Recommended Uniform Screening panel

Gal-1-PUT
: galactose-1-phosphate uridyltransferase: the enzyme whose absence or reduced activity causes galactosemia

GBYS
: Guide By Your Side Program, Hands and Voices

genotype
: the genetic material of an individual, the pair of genes present for a particular characteristic or protein

gestation
: the length of time from conception to birth. The average gestation in humans, calculated from the first day of the last normal menstrual period is 280 days (40 weeks), with a normal range of 259 days (37 weeks) to 287 days (41 weeks). Infants born prior to the 37th week are considered premature and those born after the 41st week, postmature.

gestational age
: the length of the pregnancy at birth measured from the first day of the last menstrual period, in weeks.

globin
: the protein portion of hemoglobin (consisting of 2 alpha chains and 2 beta chains in hemoglobin A)

glucose
: a simple sugar; also called D-glucose or dextrose, released by digestion of starch and other sugars, absorbed by the small intestine and circulated to the liver where excess is converted to glycogen. Within most cells, glucose is the primary energy source and is oxidized to carbon dioxide and water to produce energy.

Guide-By-Your-Side Program
: a program of Hands and Voices to provide the support of experienced families of children who are deaf, to families at the time of their child's identification of hearing loss and beyond.

Guthrie, Robert
: U.S. microbiologist (1916-1995), the "father of newborn screening"

Guthrie test
: another name for a newborn screening test or a dried bloodspot test where blood is collected on filter paper. This type of test was first developed for newborn screening by Dr. Robert Guthrie.

H

Hb S/ß thal
: S, beta-thalassemia, a core disorder of the Recommended Uniform Screening panel

Hb S/C
: S, C disease, a core disorder of the Recommended Uniform Screening panel

Hb SS
: sickle cell disease or sickle cell anemia, a core disorder of the Recommended Uniform Screening panel

HCY
: homocystinuria, a core disorder of the Recommended Uniform Screening panel

health care provider
: a physician, physician assistant, registered nurse practitioner or midwife.

Health Insurance Portability and Accountability Act of 1996 (HIPAA)
: a federal law creating national standards for recording and disclosing or keeping private protected health information (PHI).

  45 CFR 164.512 contains the exception where covered entities are not required to obtain written authorization for the disclosure of PHI for public health purposes: when the uses and disclosures are required by law or where they are disclosed to a public health authority “for the purpose of preventing or controlling disease...”

HEAR
: hearing loss, a core disorder of the Recommended Uniform Screening panel

heelstick
: a method for obtaining a blood sample from a newborn where the heel is punctured in the lateral or medial area of the plantar surface of the heel (avoiding the posterior curvature of the heel) to a depth of no more than 2.4 mm.

heme
: an iron compound that is the portion of hemoglobin that carries oxygen. There is one heme associated with each of the four globin chains of a hemoglobin molecule.

hemoglobin
: an iron-containing pigment of red blood cells that functions primarily in the transport of oxygen from the lungs to the tissues of the body, that consists of four polypeptide (globin) chains of which two are of the type designated alpha and two are of one of the types designated beta, gamma, or delta and each of which is linked to a heme molecule, that combines loosely and reversibly with oxygen in the lungs and with carbon dioxide in the tissues

hemoglobin A (Hb A)
: the hemoglobin in the red blood cells of the normal human adult that contains two alpha and two beta globin chains, each with a heme attached

hemoglobin A2 (Hb A2)
: a variant hemoglobin found in small quantities (1-2%) in normal human adults (two alpha and two delta globin chains)

hemoglobin C (Hb C)
: an abnormal hemoglobin that differs from hemoglobin A in having a lysine residue substituted for the glutamic acid residue at position 6 of each of the two β globin chains in a hemoglobin molecule

hemoglobin electrophoresis
: a test that measures the different types of hemoglobin in the blood by their movement as charged particles suspended in a liquid on gel under the influence of an applied electric field.

hemoglobin F (Hb F)
: fetal hemoglobin (containing 2 alpha and 2 gamma globin chains) is the predominant hemoglobin found in the fetus and makes up more than half of a newborn's total hemoglobin at birth. The amount of Hb F decreases after birth and a small amount of fetal hemoglobin (< 2%) can be present in adult hemoglobin.

hemoglobin H (Hb H)
: an abnormal, fast migrating hemoglobin occurring in the red blood cells when alpha globin chains are present in reduced amounts and excess beta chains come together in tetramers (4 beta chains)

hemoglobinopathy
: an inherited disorder of hemoglobin structure with characteristic clinical and laboratory abnormalities and often overt anemia.

hemoglobin S (Hb S)
: an abnormal hemoglobin occurring in the red blood cells in sickle-cell anemia and sickle-cell trait and differing from hemoglobin A in having a

valine residue substituted for the glutamic acid residue in position 6 of the two ß globin chains in the hemoglobin molecule.

hemolysis
: breakdown or destruction of red blood cells with liberation of hemoglobin

HFK
: Hear For Kids, a program of the EAR Foundation of Arizona

HHS
: U.S. Department of Health and Human Services

high performance liquid chromatography (HPLC)
: the method by which hemoglobin results are confirmed at the State Lab a form of column chromatography used to separate, identify and quantify compounds based on their polarities and interactions with the column's stationary phase (the particles packed in the column). HPLC uses different types of stationary phase, a pump to move the mobile phase (sample dissolved in a liquid) through the column and a detector that records a retention time for the analyte.

HIPAA
: Health Insurance Portability and Accountability Act of 1996

HI*TRACK
: the data management system from NCHAM used by hospitals, audiologists and the Newborn Screening Program to record and report hearing screening, diagnosis and early intervention information http://www.hitrack.org

HMG
: 3-hydroxy-3-methylglutaric aciduria, a core disorder of the recommended Uniform Screening panel

HOPS
: hearing outpatient screening - forms used for reporting outpatient hearing screening

HPLC
: high performance liquid chromatography

hydrolysis
: any reaction in which water is one of the reactants; a chemical decomposition in which a substance is split into simpler compounds by the addition of or the taking up of the elements of water

17-α-hydroxyprogesterone
an adrenal hormone that builds up in the absence of 21-hydroxylase and is used as the analyte for CAH testing

hyperplasia
abnormal increase in the number of normal cells in normal arrangement in an organ or tissue which increases its size or volume

hypo- (prefix)
deficient, low in quantity

hypochromic
in red blood cells, a decrease in the quantity of hemoglobin present so that they are abnormally pale

hypoplasia
underdevelopment of a tissue, organ or body

I

idiopathic
pertaining to conditions without clear pathogenesis, or disease without recognizable cause, as of spontaneous origin

IEF
isoelectric focusing

IEM
inborn errors of metabolism – another name for metabolic disorders

incidence
the rate at which an event occurs per unit time per person in the members of a defined population at risk; the rate of occurrence of new cases of a particular disease in a population being studied

inconclusive
a test result that cannot be determined to be either normal or abnormal because of some confounding factor. A red blood cell transfusion prior to collecting a newborn screening sample would give an inconclusive result for hemoglobin since the sample contains cells that belong to the donor and not exclusively to the baby tested.

Individuals with Disabilities Education Act (IDEA)
a law enacted in 1975 ensuring services to children with disabilities. IDEA governs how states and public agencies provide early intervention, special

education and related services to more than 6.5 million eligible infants, toddlers, children and youth with disabilities. Infants and toddlers with disabilities (birth-2) and their families receive early intervention services under IDEA Part C. Children and youth (ages 3-21) receive special education and related services under IDEA Part B.

infant
: a baby from birth until a year of age (NBS rules identify an infant as one from 29 days of age until a year of age – for the first four weeks an infant is defined as a newborn).

isoelectric focusing (IEF)
: the method used by the State Lab to identify variant hemoglobins. Results are then confirmed by HPLC. It is a technique for separating different proteins by their electrical charge differences (due to their relative content of acidic and basic side chains).

  Samples are distributed over a gel medium that has a pH gradient. An electric current is passed through the medium, creating a positive and negative end. Charged hemoglobin molecules migrate toward the oppositely charged end through the changing pH gradient until they reach a pH point that corresponds to their isoelectric points (pI) - the point where they no longer have a net charge. No further migration takes place and the proteins become focused into bands at this point.

IVA
: isovaleric acidemia, a core disorder of the recommended Uniform Screening panel

J

julian date
: consecutive numbering of the days of the year beginning with 001 for January 1st and ending with 365 for December 31st (in non-leap years)

K

ketone
: an organic compound with a carbonyl group (C=O) attached to two carbon atoms. Acetone (C_3H_6O) is a simple ketone. Ketones are formed from incomplete metabolism of fatty acids, usually from carbohydrate deficiency.

kit number
: the unique number printed on the collection kits (under the bar code, in red on the upper right hand side of the demography entry sheets and above the circles on the filter paper). This number follows the designation AZ and is sometimes known as the AZ number.

L

lab number
: unique identification number assigned by the State Lab to newborn screening bloodspot specimens (see accession number)

lactose
: a disaccharide that on hydrolysis yields glucose and galactose

lactose formula
: a milk-based formula with lactose as its carbohydrate source

LBW
: low birth weight

LCHAD
: long-chain L-3-hydroxyacyl-CoA dehydrogenase deficiency, a core disorder of the recommended Uniform Screening panel

linked kits
: double collection kits that can be taken apart at the middle perforation. The top kit is marked 1 and the bottom kit is marked 2. These kits are intended to be used by hospitals to collect a first specimen with the number 1 kit. The number 2 kit is to be placed in a bright pink envelope and sent home with the baby with instructions to take it to the first visit to the doctor (to be used to collect the second specimen). The kit numbers allow the two parts to be matched in the system.

lost to follow-up (LTF)
: a follow-up case closed when a baby whose family cannot be located for follow-up activities (further testing, referral to specialist, etc.) despite completion of follow-up activities per protocol.

low birth weight (LBW)
: birth weight of 2500 g (5.5 pounds) or less

LTF
: lost to follow-up

M

mailer
: the lab report for each newborn screening test. A copy is mailed to the submitter of the specimen and to the doctor listed who ordered the test.

Maternal and Child Health Bureau (MCHB)
: a bureau of HRSA, HHS that provides the Maternal and Child Health Block Grant to state maternal and child health programs under Title V of the Social Security Act of 1935
http://mchb.hrsa.gov

MCAD
: medium chain acyl-CoA dehydrogenase deficiency, a core disorder of the recommended Uniform Screening panel

3-MCC
: 3-methylcrotonyl-CoA carboxylase deficiency, a core disorder of the recommended Uniform Screening panel

MCD
: multiple carboxylase deficiency, a core disorder of the recommended Uniform Screening panel

MCH
: maternal and child health

MCHB
: Maternal and Child Health Bureau of HRSA, HHS

MCH Block Grant
: funding provided by MCHB to states under Title V of the Social Security Act of 1935 and its amendments
http://mchb.hrsa.gov/programs/blockgrant/overview.htm

MCT oil
: medium chain triglyceride oil

meconium
: first feces of a newborn infant (greenish black and tarry)

meconium ileus
: a blockage of the intestines with impacted meconium in a newborn – usually in infants with cystic fibrosis

metabolic disorders
: disorders affecting metabolism – usually inherited defects of enzyme production where specific enzymes are inactive and a metabolic pathway is blocked and certain substances build up to toxic levels.

metabolic formula
: a formula food designed to meet the needs of individuals with metabolic disorders (e.g. for PKU, a formula with most or all of the phenylalanine removed)

metabolic pathway
: a sequence of connected, enzyme-catalyzed reactions in cells that either builds a complex molecule (anabolic pathway) or breaks down a complex molecule into simpler compounds (catabolic pathway).

metabolism
: the sum of the processes in the buildup and destruction of living tissue, the chemical changes in living cells by which energy is provided for vital processes and activities and new material is assimilated (see anabolism and catabolism).

metabolite
: a compound that is a starting material for a metabolic process, an intermediate product of metabolism (a product of one metabolic process that is essential to another process in the same organism) or a metabolic end product that is usually excreted

microcytic
: (related to red blood cells) abnormally small red blood cells present in some anemias

microtia
: unusually small and underdeveloped external ear

MOD
: March of Dimes

MS/MS
: tandem mass spectrometry

MSUD
: maple syrup urine disease, a core disorder of the Recommended Uniform Screening panel

MUT
: methylmalonic acidemia – mutase deficiency, a core disorder of the Recommended Uniform Screening panel

mutation
: a permanent transmissible change in the genetic material (DNA)

N

National Newborn Screening and Genetics Resource Center (NNSGRC)
: a cooperative agreement between the Maternal and Child Health Bureau (MCHB), Genetics Services Branch and the University of Texas Health Science Center at San Antonio, Department of Pediatrics that provides information and resources in the area of newborn screening and genetics http://genes-r-us.uthscsa.edu

National Newborn Screening Information System (NNSIS)
: housed at NNSGRC where states report their NBS statistics and national reports are published

NBHS
: newborn hearing screening

NBS
: newborn screening

negative result
: a test value within the reference range, a normal result

neonatal
: of, relating to, or affecting the newborn and especially the human infant during the first month after birth

neonatologist
: a medical specialist (pediatrician) trained in the care of neonates that require intensive care.

neurotology
: a clinical subspecialty within otolaryngology (ENT) that focuses on the neurology and neurosurgery of the ear related to sensorineural hearing and balance disorders

newborn
: a neonate: a term applied to human infants less than a month of age

newborn screening (NBS)
: testing done within days of birth to identify infants at increased risk for specific disorders so that treatment can begin as soon as possible; when a newborn screening result is positive, further diagnostic testing is usually required to confirm the results.

  Arizona Newborn Screening Program: http://www.aznewborn.com
  State NBS contacts: http://genes-r-us.uthscsa.edu/state_contacts.pdf

Newborn Screening Advisory Committee
: A committee appointed by the Director of ADHS to provide recommendations and advice to the department regarding tests that the committee believes should be included in the Newborn Screening Program. It meets annually and includes the following members: seven physicians who represent the medical specialties of endocrinology, pediatrics, neonatology, family practice, otology and obstetrics; a neonatal nurse practitioner; an audiologist, a representative of an agency that provides services under Part C of the individuals with disabilities education act; at least one parent of a child with a hearing loss or a congenital disorder; a representative from the insurance industry; the director of AHCCCS or designee; and a representative of the hospital or health care industry. See Rules.

NICU
: neonatal intensive care unit

NIH
: National Institutes of Health – the medical research agency of HHS

NNSGRC
: National Newborn Screening and Genetics Resource Center

NNSIS
: National Newborn Screening Information System

O

OAE
: otoacoustic emissions

OCSHCN
: Office for Children with Special Health Care Needs (acronym pronounced "ocean")

Office for Children with Special Health Care Needs (OCSHCN)
in the Arizona Department of Health Services
http://www.azdhs.gov/phs/ocshcn/index.htm

17-OHP
17-α-hydroxyprogesterone, an adrenal hormone that builds up in the absence of 21-hydroxylase and is used as the analyte for CAH testing

organic acid disorders (organic acidemias)
rare inherited metabolic disorders (OAs) that occur when enzymes necessary in the breakdown of certain protein components and other substances are missing or not working properly. This results in the excretion of non-amino organic acids in urine.
Support group: Organic Acidemia Association http://www.oaanews.org

otoacoustic emissions testing (OAE)
a method used for hearing screening which measures a response from the cochlea that is generated in response to a sound. An earpiece with a microphone measures the cochlea's response to sound, listening for otoacoustic emissions. If no otoacoustic emissions are recorded, it could mean possible hearing loss. The OAE does not require the baby to respond to sound. A pass result suggests that the cochlea is working normally. A baby who doesn't pass may have a temporary hearing loss because of fluid or it may indicate a permanent hearing loss.

P

packed red blood cells (PRBC)
Transfusion of red blood cells can cause false negative newborn screening results for hemoglobinopathies and galactosemia. A valid repeat screen can be collected 3 – 4 months after the last transfusion.

parent refusal
Parents are allowed to refuse a newborn screening test for their infant. For documentation, the physician obligated to order the test will have the parents sign a form acknowledging that they understand the possible consequences of not screening their infant. The collection kit has a box to check for parent refusal. The demographics should be filled out completely, the box checked and the kit sent without blood to the State Lab.

patient number
the lab number of the screen (usually a first screen) that a subsequent screen is linked to. When a screen is unlinked its patient number is its lab

number. When a screen is linked to another screen, its patient number is the lab number of the screen it is linked to. If the patient number is different from the lab number it signifies that this screen is linked to another. If the patient number is the same as its lab number it shows that it is not linked to any other screen but other screens may be linked to it.

Perkin Elmer
: Perkin Elmer, Inc. – supplier of the AutoDelphia automatic immunoassay systems, Waters Quattro Micro tandem mass spectrometers and assay kits for TSH, 17-OHP, hemoglobin, IRT and analytes for MS/MS. http://www.perkinelmer.com

PHI
: protected health information (kept confidential under HIPAA privacy rules)

PKU
: phenylketonuria, a core disorder of the Recommended Uniform Screening panel

positive predictive value
: the percentage of true cases of disease identified in a screening test (true positives) out of the total number of positive screens- the higher the percentage, the better the test results identify those with disease. For example, a test where there were 10 abnormals reported and one true positive would have a PPV of 10% (a good test) and a test where there were 100 abnormals and one true positive would have a PPV of 1% (not as good a test).

positive result
: a test value outside the reference range, an abnormal result

practitioner
: one who has met the professional and legal requirements necessary to provide a health care service, such as a physician, nurse, dentist or physical therapist.

premature
: born at any time prior to completion of the 37th week of gestation

prevalence
: the frequency of a trait in a population, the percentage of a population that is affected with a particular disease at a given time

PROP
: propionic acidemia, a core disorder of the Recommended Uniform Screening panel

prophylactic, prophylaxis
: measures designed to preserve health and prevent the spread of disease; protective or preventive treatment

Propylthiouracil (PTU)
: a medication that suppresses thyroid hormone production and is used in the treatment of hyperthyroidism. It crosses the placenta and if a pregnant woman is treated, her baby can have transient hypothyroidism.

PTU
: propylthiouracil, a medication that suppresses thyroid hormone

Q

QNS
: quantity not sufficient

R

RBC
: red blood cell

recall screen
: a sample submitted after an abnormal repeat screen. Only 3 circles of blood are needed since only the test for the previously abnormal analyte(s) is performed (at no charge by the State Lab)

Recommended Uniform Screening Panel
: the list of 30 core disorders and 26 secondary disorders recommended by the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) for inclusion in every state newborn screening program. In May, 2010, SCID and related T-cell lymphocyte deficiencies were added to the core disorders and secondary disorders respectively. Arizona screens for the original 29 core disorders and occasionally detects a secondary disorder, but does not yet screen for SCID. http://www.hrsa.gov/heritabledisorderscommittee/uniformscreeningpanel.htm

reference range
: the range of normal values for a particular analyte or test

Rules
: Arizona Newborn Screening Rules in the Arizona Administrative Code (AAC), Title 9. Health Services, Chapter 13 Dept. of Health Services,

Article 1. Hearing Screening, Sections R9-13-101 through R9-13-110 (effective 2/18/86) and Article 2. Newborn and Infant Screening, Sections R9-13-2-1 through R9-13-205 (effective 8/31/05).
http://www.azsos.gov/PUBLIC_SERVICES/Title_09/9-13.htm

S

SACHDNC
: HHS Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (in the Maternal and Child Health Bureau of HRSA, HHS).

salt-wasting, salt-losing CAH
: see congenital adrenal hyperplasia

sample
: another name for specimen

SCID
: severe combined immunodeficiency disorder, a core disorder of the Recommended Uniform Screening panel (added to the panel in May, 2010). Arizona does not yet screen for SCID.

screening test
: testing designed to identify individuals in a given population who are at higher risk of having or developing a particular disorder, or carrying a gene for a particular disorder

second screen
: a specimen collected from a newborn after a first specimen has been collected or from an infant at least five days old and not older than one year of age, regardless of whether a first specimen was collected.

Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC)
: in the Maternal and Child Health Bureau of HRSA, HHS.
Source of the Recommended Uniform Screening Panel whose core disorders should be included in every newborn screening program. The secondary target disorders are defined as disorders that can be detected in the differential diagnosis of a core disorder. The SACHDNC maintains and periodically updates the panel. In May, 2010, a 30th disorder (SCID) was added to the panel along with a related secondary disorder (related T-cell lymphocyte deficiencies)
http://www.hrsa.gov/heritabledisorderscommittee/uniformscreeningpanel.htm

sensorineural hearing loss
: occurs when there is damage to the inner ear (cochlea) or to the nerves from the inner ear to the brain and is considered to be a permanent loss. This can be caused by diseases, birth injury, drugs and genetic syndromes or as a result of noise exposure, head trauma, aging and tumors.

severe combined immunodeficiency disorder (SCID)
: the first disorder to be added to the Uniform Screening Panel since the original panel of 29 disorders was recommended by SACHDNC. Arizona does not yet screen for SCID.
: an inherited immune system defect characterized by the absence of functional T cells and B cells.

S-HPFH
: sickle/hereditary persistence of fetal hemoglobin – a much milder form of sickle cell disease

SNHL
: sensorineural hearing loss

soy formula
: infant formula with soy protein isolate as its protein source and usually sucrose as its carbohydrate source - used when babies cannot tolerate lactose in milk-based formulas

specificity
: in a screening test – the proportion of people who are truly free of a specific disease and are so indentified by the test. A test with high specificity would have a minimum of false positives.

specimen
: a part of a thing intended to show kind and quality of the whole; for Newborn Screening, the filter paper containing dried blood spots and the identifying demographic information

specimen kit
: the strip of filter paper for collecting a blood sample attached to a form for obtaining identifying information about the infant - another name for collection kit

specimen number
: unique identification number assigned by the State Lab to each newborn screening bloodspot specimen (see accession number)

submitter
: any entity that submits newborn screening specimens to the State Lab for testing. Each submitter is assigned an identifying code by the Demographic section of the Newborn Screening program. Results are mailed to submitters and physicians ordering the tests.

supplemental kits
: single NBS collection kits, usually used for repeat tests but containing check boxes to identify what type of specimen is being submitted

T

tandem mass spectrometry (MS/MS)
: an analytical method that can determine the substances present in a sample by separating them by their weights (masses) and then measuring the amounts of the substances present. This is done by ionizing chemical compounds to generate charged molecules or molecule fragments and measuring their mass-to-charge ratios.
As a newborn screening method, MS/MS can test for amino acids and acylcarnitines and detect amino acid, fatty acid oxidation and organic acid disorders.

target cell
: a red blood cell with a darker round central area surrounded by a paler ring, which in turn is surrounded by a darker ring making it look like a bulls eye target.

TEOAE
: transient evoked otoacoustic emissions

testing fees
: Arizona NBS specimens are billed to submitters at $30.00 for a first screen and to insurance companies or responsible parties at $40.00 for a second screen. These fees are set in Rules (R9-13-208)
http://www.azsos.gov/PUBLIC_SERVICES/Title_09/9-13.htm

TFP
: trifunctional protein deficiency, a core disorder of the Recommended Uniform Screening panel

thalassemias
: a group of hereditary hemolytic anemias marked by a decreased rate of synthesis of one or more hemoglobin polypeptide (globin) chains classified according to the chain involved (α-thalassemia – absence or reduced

number of alpha chains available for combination into the globin portion of hemoglobin; ß-thalassemia – absence or reduced number of beta chains)

Third Wave
: Third Wave Technologies, Inc. now Hologic, Inc., provider of the CFTR InPlex™ reagent kits used by the Arizona Newborn Screening Program to detect 46 mutations to the CFTR gene
http://www.twt.com

Title V
: the section of the Social Security Act which funds State programs which improve health and welfare services for mothers and children. These funds are provided in the Maternal and Child Health Services Title V Block Grant.

TRECs
: T-cell receptor excision circles, a marker for SCID (absent or low amounts indicate SCID)

true positive
: a positive (abnormal) result in an individual with the disease

TYR-1
: tyrosinemia type 1, a core disorder of the Recommended Uniform Screening panel

U

Uniform Screening Panel
: see Recommended Uniform Screening Panel, SACHDNC

unilateral hearing loss
: normal hearing in one ear and hearing loss in the other ear

Universal Precautions
: an OSHA standard that protects employees who may be occupationally exposed to blood and other potential infectious materials like other body fluids.

unsat
: an unsatifactory specimen which will be accessioned as specimen type 5 (if it is a first screen or specimen type 6 if it is a second screen – if it is made unsat after accessioning, it will have unsat results listed for all tests on the panel but its specimen type will remain whatever it was originally)

unsatisfactory specimen
: a specimen which cannot be tested – no results other than the reason the specimen could not be tested will be reported

urea cycle disorder
: an inherited disorder caused by a deficiency of one of the enzymes in the urea cycle which allows ammonia to build up in the blood where it can cause irreversible brain damage, coma and death.
(of the screened disorders, citrullinemia and argininosuccinic acidemia are urea cycle disorders)

U.S. Department of Health and Human Services (HHS)
: the federal government's principal agency for protecting the health of all Americans and providing essential human services, http://www.hhs.gov

V

very low birth weight (VLBW)
: birth weight less than 1500 g

VLCAD
: very long-chain acyl-CoA dehydrogenase deficiency, a core disorder of the Recommended Uniform Screening panel

W

WAL
: within acceptable limits

WNL
: within normal limits

Arizona Revised Statutes – Newborn Screening

36-694. Report of blood tests; newborn screening program; committee; fee; definitions

A. When a birth or stillbirth is reported, the attending physician or other person required to make a report of the birth shall state on the certificate whether a blood test for syphilis was made on a specimen of blood taken from the woman who bore the child or from the umbilical cord at delivery, as required by section 36-693, and the approximate date when the specimen was taken.

B. When a birth is reported the attending physician or person who is required to make a report on the birth shall order or cause to be ordered tests for certain congenital disorders. The results of tests for these disorders must be reported to the department of health services. The department of health services shall specify in rule the disorders, the process for collecting and submitting specimens and the reporting requirements for test results.

C. When a hearing test is performed on a newborn, the initial hearing test results and any subsequent hearing test results must be reported to the department of health services as prescribed by department rules.

D. The director of the department of health services shall establish a newborn screening program within the department to ensure that the testing for congenital disorders and the reporting of hearing test results required by this section are conducted in an effective and efficient manner. The newborn screening program shall include an education program for the general public, the medical community, parents and professional groups. The director shall designate the state laboratory as the only testing facility for the program.

E. The newborn screening program shall establish and maintain a central database of newborns and infants who are tested for hearing loss and congenital disorders that includes information required in rule. Test results are confidential subject to the disclosure provisions of sections 12-2801 and 12-2802.

F. If tests conducted pursuant to this section indicate that a newborn or infant may have a hearing loss or a congenital disorder, the screening program shall provide follow-up services to encourage the child's family to access evaluation services, specialty care and early intervention services.

G. The director shall establish a committee to provide recommendations and advice to the department on at least an annual basis regarding tests that the committee believes should be included in the newborn screening program. Any recommendation by the committee that a test be added to the newborn screening program shall be accompanied by a cost-benefit analysis.

H. The committee shall include the following members who are appointed by the director and who serve without compensation or reimbursement of expenses at the pleasure of the director:

1. Seven physicians who are licensed pursuant to title 32, chapter 13 or 17 and who represent the medical specialties of endocrinology, pediatrics, neonatology, family practice, otology and obstetrics.
2. A neonatal nurse practitioner who is licensed and certified pursuant to title 32, chapter 15.
3. An audiologist who is licensed pursuant to chapter 17, article 4 of this title.
4. A representative of an agency that provides services under part C of the individuals with disabilities education act.
5. At least one parent of a child with a hearing loss or a congenital disorder.
6. A representative from the insurance industry familiar with health care reimbursement issues.
7. The director of the Arizona health care cost containment system or the director's designee.
8. A representative of the hospital or health care industry.
I. The director may establish by rule a fee that the department may collect for operation of the newborn screening program, including contracting for the testing pursuant to this section. The fee for the first specimen and hearing test shall not exceed thirty dollars. The fee for the second specimen and hearing test shall not exceed forty dollars.
J. For the purposes of this section:
1. "Infant" means a child who is twenty-nine days of age to two years of age.
2. "Newborn" means a child who is not more than twenty-eight days of age.

36-694.01. Newborn screening program fund; use; nonlapsing
A. The newborn screening program fund is established. The department of health services shall administer the fund. The fund consists of fees collected pursuant to section 36-694 and gifts and donations received by the department.
B. Subject to legislative appropriation, the department shall use fund monies to support the operation of the newborn screening program prescribed under section 36-694 and rules adopted under that section.
C. Monies in the fund are exempt from the provisions of section 35-190 relating to lapsing of appropriations.

TITLE 9. HEALTH SERVICES

CHAPTER 13. DEPARTMENT OF HEALTH SERVICES
HEALTH PROGRAMS SERVICES

Supp. 06-2

ARTICLE 1. HEARING SCREENING

Article 1 consisting of Sections R9-13-101 through R9-13-110 adopted effective February 18, 1986.

Former Article 1 consisting of Sections R9-13-111 through R9-13-117 repealed effective February 18, 1986 (Supp. 86-1).

Section

ARTICLE 2. NEWBORN AND INFANT SCREENING

Article 2, consisting of R9-13-201 through R9-13-205, recodified from R9-14-501 through R9-14-505 at 11 A.A.R. 3577, effective August 31, 2005 (Supp. 05-3).

Section

ARTICLE 3. REPEALED

Article 3 consisting of Sections R9-13-301 through R9-13-304 adopted effective July 16, 1981.

Article 3 consisting of Sections R9-13-301 through R9-13-306 repealed effective July 16, 1981.

ARTICLE 4. REPEALED

Article 4 consisting of Sections R9-13-401 through R9-13-406 repealed effective December 16, 1996 (Supp. 96-4).

Article 4 consisting of Sections R9-13-401 through R9-13-406 adopted effective July 16, 1981.

Article 4 consisting of Sections R9-13-401 through R9-13-407 repealed effective July 16, 1981.

ARTICLE 5. REPEALED

Article 5 consisting of Sections R9-13-501 through R9-13-504 adopted effective July 16, 1981.

Article 5 consisting of Sections R9-13-501 through R9-13-511 repealed effective July 16, 1981.

ARTICLE 6. REPEALED

Article 6 consisting of Sections R9-13-601 through R9-13-606 repealed effective December 16, 1996 (Supp. 96-4).

Article 6 consisting of Sections R9-13-601 through R9-13-606 adopted effective July 16, 1981.

Article 6 consisting of Sections R9-13-601 through R9-13-605 repealed effective July 16, 1981.

ARTICLE 7. REPEALED

Article 7 consisting of Sections R9-13-701 through R9-13-704 adopted effective July 16, 1981.

ARTICLE 8. REPEALED

The rules in Article 8 (R9-13-801, R9-13-802, and R9-13-806) were automatically repealed June 1, 2000. The heading for Article 8 was repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

Article 8 consisting of Sections R9-13-801 through R9-13-806 adopted effective July 16, 1981.

Section
R9-13-801. Repealed
R9-13-802. Repealed
R9-13-803. Repealed
R9-13-804. Repealed
R9-13-805. Repealed
R9-13-806. Repealed

ARTICLE 9. REPEALED

Article 9, consisting of Section R9-13-901, repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

Article 9 consisting of Section R9-13-901 adopted effective October 13, 1982.

Section
R9-13-901. Repealed
R9-13-902. Emergency expired

ARTICLE 10. REPEALED

Section
R9-13-1001. Repealed
R9-13-1002. Repealed
R9-13-1003. Repealed
R9-13-1004. Repealed

ARTICLE 11. REPEALED

Section
R9-13-1101. Repealed
R9-13-1102. Repealed
R9-13-1103. Repealed
R9-13-1104. Repealed
R9-13-1105. Repealed

ARTICLE 12. REPEALED

Section
R9-13-1201. Repealed
R9-13-1202. Emergency expired

ARTICLE 13. REPEALED

Article 13, consisting of Sections R9-13-1301 through R9-13-1303, repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

Article 13 consisting of Sections R9-13-1301 through R9-13-1303 adopted effective November 23, 1983.

Section
R9-13-1301. Repealed
R9-13-1302. Repealed
R9-13-1303. Repealed

ARTICLE 14. REPEALED

Article 14, consisting of Sections R9-13-1401 through R9-13-1415, repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

Article 14 consisting of Sections R9-13-1401 through R9-13-1415 adopted effective March 19, 1984.

Article 14 consisting of Sections R9-13-1401 through R9-13-1417 adopted as an emergency effective November 29, 1983, pursuant to A.R.S. § 41-1003, valid for only 90 days.

Section
R9-13-1401. Repealed
R9-13-1402. Repealed
R9-13-1403. Repealed
R9-13-1404. Repealed
R9-13-1405. Repealed
R9-13-1406. Repealed
R9-13-1407. Repealed
R9-13-1408. Repealed
R9-13-1409. Repealed
R9-13-1410. Repealed
R9-13-1411. Repealed
R9-13-1412. Repealed
R9-13-1413. Repealed
R9-13-1414. Repealed
R9-13-1415. Repealed
R9-13-1416. Emergency expired
R9-13-1417. Emergency expired

ARTICLE 15. RECODIFIED

Editor's Note: Article 15, consisting of R9-13-1501 through R9-3-1503 and Exhibits, was recodified to 9 A.A.C. 25.

Editor's Note: Former Article 15 was originally adopted, and subsequently amended by the addition of a new Section, under an exemption from the provisions of the Administrative Procedure Act which means that the rules were not reviewed by the Governor's Regulatory Review Council; the agency did not submit notice of proposed rulemaking to the Secretary of State for publication in the Arizona Administrative Register; the agency was not required to hold public hearings on the rules; and the Attorney General did not certify the rules.

Article 15, consisting of Sections R9-13-1501 through R9-13-1503, recodified to 9 A.A.C. 25, R9-25-801 through R9-25-803 (Supp. 98-1).

ARTICLE 1. HEARING SCREENING

R9-13-101. Definitions

In this Article, unless the context otherwise requires:

1. "Assistive listening device" has the meaning in A.R.S. § 36-1901.
2. "Audiologist" means an individual licensed under A.R.S. Title 36, Chapter 17.
3. "Audiometer" means an electronic device that generates signals used to measure hearing.
4. "Calibration" means a determination of the accuracy of an instrument by measurement of a variation from a standard.
5. "Cochlear implant" means a surgically inserted device that electrically stimulates the hearing nerve in the inner ear.
6. "dB" means decibel.
7. "dB HL" means decibel hearing level.
8. "Deaf" has the meaning in A.R.S. § 36-1941.
9. "Department" means the Arizona Department of Health Services.
10. "Documentation" means signed and dated information in written, photographic, electronic, or other permanent form.
11. "Effusion" means the escape of fluid from a blood or lymphatic vessel into tissue or a cavity.
12. "Frequency" means the number of cycles per second of a sound wave.
13. "Hard of hearing" has the meaning in A.R.S. § 36-1941.
14. "Hearing aid" has the meaning in A.R.S. § 36-1901.
15. "Hearing screening" means a test of a student's ability to hear certain frequencies at a consistent loudness performed in a school by an individual who meets the requirements in R9-13-107.
16. "Hz" means Hertz, a unit of frequency equal to one cycle per second.
17. "Immittance" means the ease of transmission of sound through the middle ear.
18. "Inner ear" means the semicircular canals, auditory nerve, and cochlea.
19. "Intensity" means the strength of a sound wave striking the eardrum resulting in the perception of loudness as expressed in decibels or decibels hearing level.
20. "Kindergarten" means the grade level immediately preceding first grade.
21. "Middle ear" means the eardrum, malleus, incus, stapes, and eustachian tube.
22. "mm H2O" means millimeters of water.
23. "Noise floor" means sounds present in the auditory canal from either the environment or bodily functions such as breathing and blood flow.
24. "Otitis media" means inflammation of the middle ear.

25. "Otoacoustic emissions" means the sounds generated from the inner ear.
26. "Outer ear" means the pinna, lobe, and auditory canal.
27. "Parent" has the meaning in A.R.S. § 15-101.
28. "Physician" means an individual licensed under A.R.S. Title 32, Chapter 13 or 17.
29. "Preschool" means the instruction preceding kindergarten provided to individuals three to five years old through a:
    a. School as defined in A.R.S. § 15-101,
    b. Accommodation school as defined in A.R.S. § 15-101,
    c. Charter school as defined in A.R.S. § 15-101, or
    d. Private school as defined in A.R.S. § 15-101.
30. "Primary care practitioner" means an individual licensed as a registered nurse practitioner under A.R.S. Title 32, Chapter 15 or a physician assistant under A.R.S. 32, Chapter 25.
31. "Pure tone" means a single frequency sound.
32. "Reproducibility" means the correlation of two responses measured simultaneously and reported by percentage.
33. "School" means:
    a. School as defined in A.R.S. § 15-101;
    b. Preschool,
    c. Kindergarten,
    d. Accommodation school as defined in A.R.S. § 15-101,
    e. Charter school as defined in A.R.S. § 15-101, or
    f. Private school as defined in A.R.S. § 15-101
34. "School administrator" means an individual or the individual's designee assigned to act on behalf of a school by the body organized for the government and the management of the school.
35. "School year" means the period between July 1 and the following June 30.
36. "Screener" means an individual qualified to perform a hearing screening in a school according to R9-13-107.
37. "Special education" has the meaning in A.R.S. § 15-761.
38. "Speech-language pathologist" means an individual licensed under A.R.S. Title 36, Chapter 17.
39. "Student" means an individual enrolled in a school.
40. "Supervision" has the meaning in A.R.S. § 36-401.
41. "Tympanogram" means a chart of the indirect measurements of the ease of movement of the parts of the middle ear as air pressure in the auditory canal changes.
42. "Tympanometer" means a device that indirectly measures the ease of movement of the parts of the middle ear as air pressure in the auditory canal changes.
43. "Tympanometry" means the indirect measurement of the ease of movement of the parts of the middle ear as air pressure in the auditory canal changes.

Historical Note

Adopted effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Amended by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-102. Hearing Screening Population

A. A school administrator shall ensure that the following students have a hearing screening each school year:
    1. A student enrolled in preschool, kindergarten, or grade 1, 2, 6, or 9;
    2. A student enrolled in grade 3, 4, or 5, unless there is written documentation that the student had a hearing screening in or after grade 2;
    3. A student enrolled in grade 7 or 8, unless there is written documentation that the student had a hearing screening in or after grade 6;
    4. A student enrolled in grade 10, 11, or 12 unless there is written documentation that the student had a hearing screening in or after grade 9;
    5. A student receiving special education; and
    6. A student who failed a second hearing screening in the prior school year.

B. A school administrator shall ensure that a student has a hearing screening at the request of the student, the student's parent, a schoolteacher, a school nurse, a school psychologist, an audiologist, a physician, a primary care practitioner, a speech language pathologist, or Department staff.

C. A hearing screening is not required if a:
    1. Student is age 16 years or over;
    2. Student's parent objects in writing to the screening as allowed under A.R.S. § 36-899.04;
    3. Written diagnosis or evaluation from an audiologist states that a student is deaf or hard of hearing; or
    4. Student has a hearing aid, an assistive listening device, or a cochlear implant.

D. In addition to meeting the requirements in subsections (A) and (B), a school administrator shall ensure that a student who meets the criteria specified in State Board of Education rule R7-2-401 has a hearing screening required under R7-2-401.

Historical Note

Former Section R9-13-112 renumbered and amended as Section R9-13-102 effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Amended by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-103. Hearing Screening Requirements

A. Before performing a hearing screening, a screener shall visually inspect a student's outer ears for:
   1. Fluid or drainage,
   2. Blood,
   3. An open sore, or
   4. A foreign object.

B. If a screener inspects a student's outer ears and finds any of the conditions in subsection (A), the screener shall not perform a hearing screening.

C. A screener shall perform a hearing screening in each ear using one of the following hearing screening methods:
   1. Four-frequency, pure tone hearing screening that screens at each of the following frequencies and intensities:
      a. 500 Hz at 25 dB HL,
      b. 1000 Hz at 20 dB HL,
      c. 2000 Hz at 20 dB HL, and
      d. 4000 Hz at 20 dB HL;
   2. Three-frequency, pure tone hearing screening with tympanometry that:
      a. Includes a tympanogram that is generated automatically or is plotted at a minimum of the following three points:
         i. +100 mm H2O,
         ii. Point of maximum immittance, and
         iii. -200 mm H2O; and
      b. Screens at each of the following frequencies at 20 dB HL:
         i. 1000 Hz,
         ii. 2000 Hz, and
         iii. 4000 Hz; or
   3. Otoacoustic emissions hearing screening using otoacoustic emissions equipment that generates a pass or no pass result:
      a. Using a minimum of three frequencies,
      b. At no less than 3 dB above the noise floor, and
      c. With reproducibility greater than 50%.

Historical Note

Adopted effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Amended by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-104. Criteria for Passing a Hearing Screening; Requirements for Performing a Second Hearing Screening

A. A student passes a hearing screening if:
   1. During a four-frequency, pure tone hearing screening, the student responds in each ear to each frequency at each intensity listed in R9-13-103(C)(1)(a) through (C)(1)(d);
   2. During a three-frequency, pure tone hearing screening with tympanometry, the student:
      a. Responds in each ear to each frequency as described in R9-13-103(C)(2)(b); and
      b. Reaches a point of maximum immittance in each ear within the range of +100mm H2O to -200mm H2O; or
   3. During an otoacoustic emissions hearing screening, the student receives a pass result in each ear according to R9-13-103(C)(3).

B. If a student does not pass a hearing screening according to subsection (A), a screener shall perform a second hearing screening on the student no earlier than 30 days and no later than 45 days from the date of the first hearing screening. The screener shall perform the second hearing screening using the same method as the first hearing screening.

Historical Note

Adopted effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Amended by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-105. Referral; Notification; Follow-up

A. If a school administrator finds that a student does not require a hearing screening under R9-13-102(C)(3) or (C)(4), the school administrator shall provide to the student's parent, within 10 days from the date the finding is made, a referral to have the student's current hearing status evaluated by an audiologist, including an electroacoustic analysis of any hearing aid or assistive listening device, unless there is documentation from an audiologist specifying a different evaluation schedule.

B. If a screener finds any of the conditions listed in R9-13-103(A) and a student does not have a hearing screening:
   1. A school administrator shall provide to the student's parent, within 10 days from the date the condition is found, a referral to have the student's outer ears evaluated by a physician or primary care practitioner; and
   2. A screener shall perform the hearing screening on the student no earlier than 30 days and no later than 45 days from the date the screener finds the condition.

C. If a student does not pass a second hearing screening or does not complete a second hearing screening within the time period required under R9-13-104(B), a school administrator shall provide to the student's parent, within 10 days from the date of

the second hearing screening or from the date the period for completing a second hearing screening ends, a referral to have the student's current hearing status evaluated by one of the following:

1. An audiologist, a physician, or a primary care practitioner if the screener used only the four-frequency, pure tone hearing screening method;
2. A physician or primary care practitioner if the student did not pass the tympanometry portion, but passed the three-frequency, pure tone portion of the hearing screening;
3. An audiologist if the student did not pass the three-frequency, pure tone portion, but passed the tympanometry portion of the hearing screening; or
4. An audiologist, a physician, or a primary care practitioner if the screener used the otoacoustic emissions hearing screening method.

D. A referral identified in subsection (C) is not required if a school-provided audiologist:

1. Assesses a student's hearing status and the condition of the middle ear at the conclusion of a hearing screening; and
2. Within 10 days from date of the assessment, provides the student's parent with a written diagnosis and recommendation for treatment, if applicable.

E. A referral required under subsections (A), (B), or (C), shall include a form requesting the following:

1. The name, address, and telephone number of the student evaluated;
2. The date of evaluation;
3. An assessment of the condition of the outer ear, if applicable;
4. An assessment of hearing status and the condition of the middle ear, if applicable;
5. A diagnosis and recommendation for treatment, if applicable;
6. The signature and title of the individual evaluating the student and completing the form; and
7. A request that the individual completing the form or the student's parent return the completed form to the school.

F. Under State Board of Education rule R7-2-401, a school administrator shall ensure that a student referred under subsections (A) or (C) is evaluated.

G. If a school receives notice of a diagnosis that a student is deaf or hard of hearing from an audiologist, the school administrator shall notify, within 10 days from the date the notice of diagnosis is received, each of the student's teachers and the person responsible for the school's special education services of the diagnosis.

Historical Note

Adopted effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Amended by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-106. Repealed

Historical Note

Adopted effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Section repealed by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-107. Screener Qualifications

A. An audiologist may perform a hearing screening.

B. An individual who is not an audiologist may perform a hearing screening only if the individual passes a hearing screener course that:

1. Includes 90 minutes of classroom instruction in the introduction to hearing covering:
   a. Development of speech and language;
   b. Anatomy and physiology of the ear;
   c. Signs and prevention of hearing loss in children; and
   d. A.R.S. Title 36, Chapter 7.2 and 9 A.A.C. 13, Article 1;
2. Includes 120 minutes of classroom instruction in hearing screening covering:
   a. Auditory development,
   b. Early identification of hearing loss,
   c. Principles of hearing screening,
   d. Selection of hearing screening methods, and
   e. Components of setting-up a hearing screening program;
3. Includes 75 minutes of classroom instruction in referral and reporting covering:
   a. Results of a hearing screening,
   b. Responses to a hearing screening outcome,
   c. Procedures for recording and tracking,
   d. Communication with parents,
   e. Role of community resources, and
   f. Reporting hearing screening results;
4. For an individual who will perform a hearing screening using three-frequency or four-frequency, pure tone hearing screening, includes 120 minutes of classroom instruction covering:
   a. Selecting and setting-up a hearing screening site,
   b. Performing a pure tone hearing screening, and

c. Identifying children who need referral and evaluation;

5. For an individual who will perform a hearing screening using tympanometry with three-frequency, pure tone hearing screening, includes 60 minutes of classroom instruction covering:
   a. The anatomy and functions of the middle ear,
   b. What tympanometry measures and identifies,
   c. Using a tympanometer,
   d. Performing a tympanometry hearing screening, and
   e. Identifying children who need referral and evaluation;
6. For an individual who will perform a hearing screening using otoacoustic emissions hearing screening, includes 60 minutes of classroom instruction covering:
   a. What otoacoustic emissions identify and measure,
   b. Using otoacoustic emissions equipment,
   c. Performing an otoacoustic emissions hearing screening, and
   d. Identifying children who need referral and evaluation;
7. Requires an individual to pass the course by scoring 80% or more on an examination that tests what the individual has learned;
8. Is taught by an individual who:
   a. Is an audiologist, or
   b. Meets the screener qualifications in subsection (B) or (C) and has performed at least 50 hearing screenings within 24 months before teaching a hearing screener course; and
9. Provides an individual who passes the course with a certificate of completion that includes:
   a. The individual's name;
   b. Whether the following were completed:
      i. Introduction to hearing,
      ii. Hearing screening,
      iii. Referral and reporting,
      iv. Pure tone hearing screening,
      v. Tympanometry hearing screening, and
      vi. Otoacoustic emissions hearing screening;
   c. An attestation that the course meets the requirements in subsection (B) or (C); and
   d. The name and signature of the individual who taught the course.

C. Every five years after completing a hearing screener course described in subsection (B), a screener who is not an audiologist shall pass a hearing screener course that:
1. Includes 195 minutes of classroom instruction covering the material required under subsections (B)(1), (B)(2), and (B)(3);
2. For an individual who will perform a hearing screening using three-frequency or four-frequency, pure tone hearing screening, includes 60 minutes of classroom instruction covering the material required under subsection (B)(4);
3. For an individual who will perform a hearing screening using tympanometry with three-frequency, pure tone hearing screening, includes 30 minutes of classroom instruction covering the material required under subsection (B)(5);
4. For an individual who will perform a hearing screening using otoacoustic emissions hearing screening, includes 30 minutes of classroom instruction covering the material required under subsection (B)(6); and
5. Meets the requirements in subsections (B)(7), (B)(8), and (B)(9).

D. Before performing a hearing screening, an individual who passes a hearing screener course described in subsection (B) or (C) shall give a copy of the certificate of completion described in subsection (B)(9) to the school.

E. An individual who does not meet the screener qualifications in subsection (A), (B), or (C) may perform a four-frequency, pure tone hearing screening, other than a second hearing screening required under R9-25-104(B), only under the supervision of an individual who meets the screener qualifications in subsection (A), (B), or (C).

Historical Note

Former Section R9-13-113 renumbered and amended as Section R9-13-107 effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Amended by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-108. Equipment Standards

A. A school administrator shall ensure that a pure tone audiometer used to perform a three-frequency or four-frequency, pure tone hearing screening is:
1. Calibrated every 12 months according to the American National Standard Specification for Audiometers, S3.6-1996, Standards Secretariat, c/o Acoustical Society of America, 120 Wall Street, 32nd Floor, New York, New York 10005-3993, January 12, 1996, incorporated by reference in R9-16-209(B)(1); and
2. Inspected within 24 hours before use to ensure that:
   a. The calibration complies with subsection (A)(1),
   b. The power source and power indicator are working,
   c. The earphone cords are securely connected and have no breaks,
   d. Each frequency and intensity required under R9-13-103(C)(1) is present,

e. A signal does not cross from one earphone to the other, and
f. Each earphone is free of noise or distortion that could interfere with a hearing screening.

B. A school administrator shall ensure that a tympanometer used to perform the tympanometry portion of a hearing screening:
1. Is calibrated every 12 months according to the American National Standard Specifications for Instruments to Measure Aural Acoustic Impedance and Admittance, S3.39-1987, Standards Secretariat, Acoustical Society of America, 335 East 45th Street, New York, New York 10017-3483, October 5, 1987, not including any later amendments or editions, incorporated by reference and on file with the Department and the Office of the Secretary of State; and
2. Is inspected within 24 hours before use to ensure that the calibration complies with subsection (B)(1).

C. A school administrator shall ensure that otoacoustic emissions equipment used to perform an otoacoustic emissions hearing screening is:
1. Calibrated every 12 months according to manufacturer's specifications; and
2. Inspected within 24 hours before use to ensure that:
a. The calibration complies with manufacturer's specifications,
b. No obstruction is in the probe microphone, and
c. The test signal is present.

Historical Note

Adopted effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Amended by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-109. Recordkeeping, Reporting Requirements

A. A school administrator shall retain, for Department review and inspection, a written record of:
1. The date and results of a student's hearing screening for no less than three complete school years beginning on the first July 1 after the student's last date of attendance at the school, and
2. All calibration dates for a piece of hearing screening equipment currently used in the school.

B. By June 30th of each year, a school administrator shall submit to the Department the following information for the school year ending that June 30th:
1. On a form available from the Department, the number of students by grade in each of the following categories:
a. Were enrolled at the time of a first hearing screening,
b. Did not have a first hearing screening under R9-13-102(C),
c. Had a first hearing screening,
d. Did not pass a first hearing screening,
e. Had a second hearing screening,
f. Did not pass a second hearing screening,
g. Were evaluated by an audiologist,
h. Were evaluated by a physician or a primary care practitioner,
i. Were first diagnosed as deaf or hard of hearing during the current school year, and
j. Were diagnosed as deaf or hard of hearing during a prior school year; and
2. The name of each individual who performed a hearing screening in the school and:
a. The individual's license number to practice audiology, or
b. Evidence that the individual successfully completed a hearing screening course described in R9-13-107(B) or (C).

Historical Note

Former Section R9-13-116 renumbered and amended as Section R9-13-109 effective February 18, 1986 (Supp. 86-1). Amended effective October 15, 1993 (Supp. 93-4). Amended by final rulemaking at 8 A.A.R. 3307, effective July 16, 2002 (Supp. 02-3).

R9-13-110. Repealed

Historical Note

Former Section R9-13-117 renumbered and amended as Section R9-13-110 effective February 18, 1986 (Supp. 86-1). Repealed effective October 15, 1993 (Supp. 93-4).

R9-13-111. Repealed

Historical Note

Effective 4-72. Amended effective November 18, 1976 (Supp. 76-5). Repealed effective February 18, 1986 (Supp. 86-1).

R9-13-112. Renumbered

Historical Note

Effective 4-72. Amended effective November 18, 1976 (Supp. 76-5). Section R9-13-112 renumbered and amended as Section R9-13-102 effective February 18, 1986 (Supp. 86-1).

R9-13-113. Renumbered

Historical Note

Effective 4-72. Amended effective November 18, 1976 (Supp. 76-5). Section R9-13-113 renumbered and amended as Section R9-13-107 effective February 18, 1986 (Supp. 86-1).

R9-13-114. Repealed

Historical Note

Effective 4-72. Amended effective November 18, 1976 (Supp. 76-5). Repealed effective February 18, 1986 (Supp. 86-1).

R9-13-115. Repealed

Historical Note

Effective 4-72. Amended effective November 18, 1976 (Supp. 76-5). Repealed effective February 18, 1986 (Supp. 86-1).

R9-13-116. Renumbered

Historical Note

Effective 4-72. Correction, Section R9-13-116 omitted in Supp. 76-5 (Supp. 77-5). Section R9-13-116 renumbered and amended as Section R9-13-109 effective February 18, 1986 (Supp. 86-1).

R9-13-117. Renumbered

Historical Note

Effective 4-72. Correction, Section R9-13-117 omitted in Supp. 76-5 (Supp. 77-5). Section R9-13-117 renumbered and amended as Section R9-13-110 effective February 18, 1986 (Supp. 86-1).

ARTICLE 2. NEWBORN AND INFANT SCREENING

R9-13-201. Definitions

In this Article, unless otherwise specified:

1. "Abnormal result" means an outcome that deviates from the range of values established by the Department for an analysis performed as part of a bloodspot test, or for a hearing test.
2. "Admitted" means the same as in A.A.C. R9-10-201.
3. "AHCCCS" means the Arizona Health Care Cost Containment System.
4. "Argininosuccinic acidemia" means a congenital disorder characterized by an inability to metabolize the amino acid argininosuccinic acid due to defective argininosuccinate lyase activity.
5. "Audiological equipment" means instruments used to measure a physiological response to determine the presence, type, or degree of hearing loss.
6. "Audiologist" means an individual licensed under A.R.S. Title 36, Chapter 17.
7. "Beta-ketothiolase deficiency" means a congenital disorder characterized by an inability to metabolize 2-methyl-acetoacetyl-CoA due to defective mitochondrial acetoacetyl-CoA thiolase activity.
8. "Biotinidase deficiency" means a congenital disorder characterized by defective biotinidase activity that causes abnormal biotin metabolism.
9. "Birth center" means a health care facility that is not a hospital and is organized for the sole purpose of delivering newborns.
10. "Blood sample" means capillary or venous blood, but not cord blood, applied to the filter paper of a specimen collection kit.
11. "Bloodspot test" means multiple laboratory analyses performed on a blood sample to detect the presence of congenital disorders listed in R9-13-202.
12. "Carnitine uptake defect" means a congenital disorder characterized by a decrease in the amount of free carnitine due to defective sodium ion-dependent carnitine transporter OCTN2 activity.
13. "Citrullinemia" means a congenital disorder characterized by an inability to convert the amino acid citrulline and aspartic acid into argininosuccinic acid due to defective argininosuccinate synthetase activity.
14. "Classic galactosemia" means a congenital disorder characterized by abnormal galactose metabolism due to defective galactose-1-phosphate uridyltranferase activity.
15. "Congenital adrenal hyperplasia" means a congenital disorder characterized by decreased cortisol production and increased androgen production due to defective 21-hydroxylase activity.
16. "Congenital disorder" means an abnormal condition present at birth, as a result of heredity or environmental factors, that impairs normal physiological functioning of a human body.
17. "Congenital hypothyroidism" means a congenital disorder characterized by deficient thyroid hormone production.
18. "Cystic fibrosis" means a congenital disorder caused by defective functioning of a transmembrane regulator protein and characterized by damage to and dysfunction of various organs, such as the lungs, pancreas, and reproductive organs.
19. "Department" means the Arizona Department of Health Services.
20. "Discharge" means the termination of inpatient services to a newborn or infant.
21. "Disorder" means a disease or medical condition that may be identified by a laboratory analysis.
22. "Document" means to establish and maintain information in written, photographic, electronic, or other permanent form.
23. "Educational materials" means printed or electronic information provided by the Department, explaining newborn and infant screening, any of the congenital disorders listed in R9-13-202, or hearing loss.

24. "Electronic" means the same as in A.R.S. § 44-7002.
25. "First specimen" means the initial specimen that is collected from a newborn who is less than five days of age and sent to the screening laboratory for testing and recording of demographic information.
26. "Glutaric acidemia type I" means a congenital disorder characterized by an accumulation of glutaric acid due to defective glutaryl-CoA dehydrogenase activity.
27. "Guardian" means an individual appointed by a court under A.R.S. Title 14, Chapter 5, Article 2.
28. "Health care facility" means a health care institution defined in A.R.S. § 36-401 where obstetrical care or newborn care is provided.
29. "Health care provider" means a physician, physician assistant, registered nurse practitioner, or midwife.
30. "Health-related services" means the same as in A.R.S. § 36-401.
31. "Hearing test" means an evaluation of both ears of a newborn or infant, using audiological equipment, for the presence, type, or degree of hearing loss.
32. "Hemoglobin S/Beta-thalassemia" means a sickle cell disease in which an individual has one sickle cell gene and one gene for beta thalassemia, another inherited hemoglobinopathy.
33. "Hemoglobin S/C disease" means a sickle cell disease in which an individual has one sickle cell gene and one gene for another inherited hemoglobinopathy called hemoglobin C.
34. "Hemoglobinopathy" means a congenital disorder characterized by abnormal production, structure, or functioning of hemoglobin.
35. "Home birth" means delivery of a newborn, outside a health care facility, when the newborn is not hospitalized within 72 hours of delivery.
36. "Homocystinuria" means a congenital disorder characterized by abnormal methionine and homocysteine metabolism due to defective cystathione-ß-synthase activity.
37. "Hospital" means the same as in A.A.C. R9-10-201.
38. "Hospital services" means the same as in A.A.C. R9-10-201.
39. "3-Hydroxy-3-methylglutaric aciduria" means a congenital disorder characterized by the accumulation of 3-hydroxy-3-methylglutaric acid due to a defective 3-hydroxy-3-methylglutaryl-CoA lyase activity.
40. "Identification code" means a unique set of numbers or letters, or a unique set of both numbers and letters, assigned by the Department to a health care facility, a health care provider, an audiologist, or another person submitting specimen collection kits to the screening laboratory or hearing test results to the Department.
41. "Infant" means the same as in A.R.S. § 36-694.
42. "Inpatient" means an individual who:
    a. Is admitted to a hospital,
    b. Receives hospital services for 24 consecutive hours, or
    c. Is admitted to a birth center.
43. "Inpatient services" means medical services, nursing services, or other health-related services provided to an inpatient in a health care facility.
44. "Isovaleric acidemia" means a congenital disorder characterized by an accumulation of isovaleric acid due to defective isovaleryl-CoA dehydrogenase activity.
45. "Long-chain 3-hydroxy acyl-CoA dehydrogenase deficiency" means a congenital disorder characterized by an inability to metabolize fatty acids that are 12 to 16 carbon atoms in length due to defective long-chain 3-hydroxy acyl-CoA dehydrogenase activity.
46. "Maple syrup urine disease" means a congenital disorder of branched chain amino acid metabolism due to defective branched chain-keto acid dehydrogenase activity.
47. "Medical services" means the same as in A.R.S. § 36-401.
48. "Medium chain acyl-CoA dehydrogenase deficiency" means a congenital disorder characterized by an inability to metabolize fatty acids that are 6 to 10 carbon atoms in length due to defective medium-chain acyl-CoA dehydrogenase activity.
49. "3-Methylcrotonyl-CoA carboxylase deficiency" means a congenital disorder characterized by an accumulation of 3-methylcrotonyl-glycine due to defective 3-methylcrotonyl-CoA carboxylase activity.
50. "Methylmalonic acidemia (Cbl A,B)" means a congenital disorder characterized by an accumulation of methylmalonic acid due to defective activity of methylmalonyl-CoA racemase or adenosylcobalamin synthetase.
51. "Methylmalonic acidemia (mutase deficiency)" means a congenital disorder characterized by an accumulation of methylmalonic acid due to defective methylmalonyl-CoA mutase activity.
52. "Midwife" means an individual licensed under A.R.S. Title 36, Chapter 6, Article 7, or certified under A.R.S. Title 32, Chapter 15.
53. "Multiple carboxylase deficiency" means a congenital disorder characterized by an inability to transport or metabolize biotin that leads to defective activity of propionyl-CoA carboxylase, beta-methylcrotonyl-CoA carboxylase, and pyruvate carboxylase.
54. "Newborn" means the same as in A.R.S. § 36-694.
55. "Newborn care" means medical services, nursing services, and health-related services provided to a newborn.
56. "Nursing services" means the same as in A.R.S. § 36-401.

57. "Obstetrical care" means medical services, nursing services, and health-related services provided to a woman throughout her pregnancy, labor, delivery, and postpartum.
58. "Organ" means a somewhat independent part of a human body, such as a salivary gland, kidney, or pancreas, which performs a specific function.
59. "Parent" means a natural, adoptive, or custodial mother or father of a newborn or infant.
60. "Person" means the state, a municipality, district, or other political subdivision, a cooperative, institution, corporation, company, firm, partnership, individual, or other legal entity.
61. "Phenylketonuria" means a congenital disorder characterized by abnormal phenylalanine metabolism due to defective phenylalanine hydroxylase activity.
62. "Physician" means an individual licensed under A.R.S. Title 32, Chapters 13, 14, 17, or 29.
63. "Physician assistant" means an individual licensed under A.R.S. Title 32, Chapter 25.
64. "Propionic acidemia" means a congenital disorder characterized by an accumulation of glycine and 3-hydroxypropionic acid due to defective propionyl-CoA carboxylase activity.
65. "Registered nurse practitioner" means the same as in A.R.S. § 32-1601.
66. "Screening laboratory" means an entity contracted with the Department under A.R.S. § 36-694(I) to perform the bloodspot test.
67. "Second specimen" means a specimen that is sent to the screening laboratory for testing and recording of demographic information, after being collected:
    a. From a newborn after a first specimen; or
    b. From an individual at least five days and not older than one year of age, regardless of whether a first specimen was collected.
68. "Sickle cell anemia" means a sickle cell disease in which an individual has two sickle cell genes.
69. "Sickle cell disease" means a hemoglobinopathy characterized by an abnormally shaped red blood cell resulting from the abnormal structure of the protein hemoglobin.
70. "Sickle cell gene" means a unit of inheritance that is involved in producing an abnormal type of the protein hemoglobin, in which the amino acid valine is substituted for the amino acid glutamic acid at a specific location in the hemoglobin.
71. "Specimen" means a blood sample obtained from and demographic information about a newborn or infant.
72. "Specimen collection kit" means a strip of filter paper for collecting a blood sample attached to a form for obtaining the information specified in R9-13-203(A)(3) about a newborn or infant.
73. "Test" means a laboratory analysis performed on body fluid, tissue, or excretion to determine the presence or absence of a disorder.
74. "Transfer" means a health care facility discharging a newborn and sending the newborn to a hospital for inpatient medical services without the intent that the patient will be returned to the sending health care facility.
75. "Transfusion" means the infusion of blood or blood products into the body of an individual.
76. "Trifunctional protein deficiency" means a congenital disorder characterized by an inability to metabolize fatty acids that are 12 to 18 carbon atoms in length due to defective mitochondrial trifunctional protein activity.
77. "Tyrosinemia type I" means a congenital disorder characterized by an accumulation of the amino acid tyrosine due to defective fumarylacetoacetate hydrolase activity.
78. "Verify" means to confirm by obtaining information through a source such as the newborn screening program, a health care provider, a health care facility, or a documented record.
79. "Very long-chain acyl-CoA dehydrogenase deficiency" means a congenital disorder characterized by an inability to metabolize fatty acids that are 14 to 18 carbon atoms in length due to defective very long-chain acyl-CoA dehydrogenase activity.
80. "Working day" means 8:00 a.m. through 5:00 p.m. Monday through Friday, excluding state holidays.

Historical Note

Amended effective October 26, 1977 (Supp. 77-5). Former Section R9-13-201 repealed, new Section R9-13-201 adopted effective July 16, 1981 (Supp. 81-4). Amended as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Amended by adding paragraphs (3), (5) and (7) and renumbering remaining paragraphs effective November 23, 1983. Amended as an emergency, by adding paragraphs (32) and (42) and renumbering remaining paragraphs, effective November 23, 1983, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency amendment expired. Permanent amendment, adding paragraphs (32) and (42) and renumbering remaining paragraphs adopted effective March 19, 1984 (Supp. 84-2). Amended as an emergency effective November 6, 1989, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 89-4). Emergency expired. Readopted as an emergency effective February 7, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-1). Re-adopted as an emergency with changes effective May 7, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-2). Readopted as an emergency with changes effective August 6, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-3). Readopted as an emergency without change effective October 31, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-4). Readopted as an emergency with changes effective January 16, 1991, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 91-1). Readopted as an emergency without change effective April 11, 1991, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 91-2). Emergency amendments permanently adopted with changes effective July 3, 1991 (Supp. 91-3). Amended effective December 16, 1996 (Supp.

96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective September 24, 1998 (Supp. 99-1). New Section recodified from R9-14-501 at 11 A.A.R. 3577, effective August 31, 2005 (Supp. 05-3). Amended by final rulemaking at 12 A.A.R. 1166, effective April 4, 2006 (Supp. 06-2).

R9-13-202. Tests for Congenital Disorders

A bloodspot test shall include laboratory analyses for the following congenital disorders:

1. Argininosuccinic acidemia,
2. Biotinidase deficiency,
3. Citrullinemia,
4. Classic galactosemia,
5. Congenital adrenal hyperplasia,
6. Congenital hypothyroidism,
7. Hemoglobin S/Beta-thalassemia,
8. Hemoglobin S/C disease,
9. Homocystinuria,
10. Maple syrup urine disease,
11. Phenylketonuria,
12. Sickle cell anemia,
13. Tyrosinemia type I,
14. 3-Methylcrotonyl-CoA carboxylase deficiency,
15. 3-Hydroxy-3-methylglutaric aciduria,
16. Beta-ketothiolase deficiency,
17. Carnitine uptake defect,
18. Glutaric acidemia type I,
19. Isovaleric acidemia,
20. Long-chain 3-hydroxy acyl-CoA dehydrogenase deficiency,
21. Medium chain acyl-CoA dehydrogenase deficiency,
22. Methylmalonic acidemia (Cbl A,B),
23. Methylmalonic acidemia (mutase deficiency),
24. Multiple carboxylase deficiency,
25. Propionic acidemia,
26. Trifunctional protein deficiency,
27. Very long-chain acyl-CoA dehydrogenase deficiency, and
28. Cystic fibrosis.

Historical Note

Amended effective October 26, 1977 (Supp. 77-5). Former Section R9-13-202 repealed, new Section R9-13-202 adopted effective July 16, 1981 (Supp. 81-4). Repealed by emergency effective November 6, 1989, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 89-4). Emergency expired. Emergency repeal readopted effective February 7, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-1). Emergency repeal readopted effective May 7, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-2). Emergency repeal readopted effective August 6, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-3). Emergency repeal readopted effective October 31, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-4). Emergency repeal readopted effective January 16, 1991, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 91-1). Emergency repeal readopted effective April 11, 1991, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 91-2). Repealed permanently effective July 3, 1991 (Supp. 91-3). New Section recodified from R9-14-502 at 11 A.A.R. 3577, effective August 31, 2005
(Supp. 05-3). Section repealed; new Section made by final rulemaking at 12 A.A.R. 1166, effective April 4, 2006 (Supp. 06-2).

R9-13-203. General Requirements for Newborn and Infant Bloodspot Tests

A. When a bloodspot test is ordered for a newborn or an infant, a health care facility's designee, a health care provider, or the health care provider's designee shall:

1. Only use a specimen collection kit supplied by the Department;
2. Collect a blood sample from the newborn or infant on a specimen collection kit;
3. Complete the following information on the specimen collection kit:
    a. The newborn's or infant's name, gender, race, ethnicity, medical record number, and if applicable, AHCCCS identification number;
    b. The newborn's or infant's type of food or food source;
    c. Whether the newborn or infant is from a single or multiple birth;
    d. If the newborn or infant is from a multiple birth, the birth order of the newborn or infant;
    e. Whether the newborn or infant has a medical condition that may affect the bloodspot test results;
    f. Whether the newborn or infant received antibiotics or a blood transfusion and, if applicable, the date of the last blood transfusion;

g. The method of blood sample collection;
h. The date and time of birth, and the newborn's or infant's weight at birth;
i. The date and time of blood sample collection, and the newborn's or infant's weight when the blood sample is collected;
j. The name and identification code of the health care facility or health care provider submitting the specimen collection kit;
k. The name, identification code, and address of the health care provider responsible for the management of medical services provided to the newborn or infant;
l. Except as provided in subsection (A)(3)(m), the mother's first and last names, date of birth, name before first marriage, mailing address, phone number, and if applicable, AHCCCS identification number; and
m. If the newborn's or infant's mother does not have physical custody of the newborn or infant, the first and last names, mailing address, and phone number of the person who has physical custody of the newborn or infant; and

4. Submit the specimen collection kit to the screening laboratory no later than 24 hours or the next working day after the blood sample is collected.

B. A health care facility or a health care provider submitting a first specimen to the screening laboratory shall pay the Department the fee in R9-13-208(A).

C. A person who submits a second specimen to the screening laboratory shall:
1. Pay the fee in R9-13-208(B) to the Department, or
2. Provide the following information to the screening laboratory for billing purposes:
a. The name, mailing address, and phone number of the newborn's or infant's parent or the individual responsible for paying, if not the parent; and
b. If the individual responsible for paying has health care insurance for the newborn or infant, information about the health care insurance, including:
i. The policyholder's name;
ii. The name and billing address of the health care insurance company;
iii. The member identification number;
iv. The group number, if applicable; and
v. The effective date of the health care insurance; or
c. That the individual responsible for paying has no health care insurance for the newborn or infant.

D. When a health care insurance company or an individual responsible for paying is identified as specified in subsection (C)(2), the health care insurance company or the individual responsible for paying shall pay the Department the fee in R9-13-208(B).

E. The screening laboratory shall perform a bloodspot test on a blood sample from a specimen collection kit if:
1. The blood sample on the specimen collection kit:
a. Contains a sufficient quantity of blood to complete the bloodspot test,
b. Is not clotted or layered,
c. Does not have serum rings,
d. Is not diluted or discolored,
e. Will elute from the filter paper,
f. Has not been applied to both sides of the filter paper, and
g. Is not contaminated;
2. The filter paper on the specimen collection kit is not contaminated, scratched, or abraded;
3. The information on the specimen collection kit is sufficient to identify:
a. The newborn or infant, and
b. The person who ordered the bloodspot test or caused the bloodspot test to be ordered; and
4. The screening laboratory receives the specimen collection kit within 14 days after the blood sample is collected.

F. When a home birth not attended by a health care provider is reported to a local registrar, a deputy local registrar, or the state registrar under A.R.S. § 36-333:
1. The local registrar, deputy local registrar, or state registrar shall notify the local health department of the county where the birth occurred; and
2. The local health department's designee shall collect a specimen from the newborn or infant according to the requirements in R9-13-204(A)(2) or R9-13-205(C).

G. A health care facility's designee, a health care provider, or the health care provider's designee shall ensure that:
1. Educational materials are provided to the parent or guardian of a newborn or infant for whom a bloodspot test is ordered, and
2. The newborn's or infant's parent or guardian is informed of the requirement for a second specimen if the second specimen has not been collected.

H. For a home birth, a health care provider or the health care provider's designee shall provide educational materials to the parent or guardian of a newborn or infant for whom a bloodspot test is ordered.

Historical Note

Effective 11-74; Former Section R9-13-203 repealed, new Section R9-13-203 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R.

146, effective September 24, 1998 (Supp. 99-1). New Section recodified from R9-14-503 at 11 A.A.R. 3577, effective August 31, 2005
(Supp. 05-3). Section repealed; new Section made by final rulemaking at 12 A.A.R. 1166, effective April 4, 2006 (Supp. 06-2).

R9-13-204. First Specimen Collection

A. When a newborn is born in a hospital, the hospital's designee shall collect a first specimen from the newborn according to whichever of the following occurs first:
  1. Before a transfusion, unless specified otherwise by a physician, physician assistant, or registered nurse practitioner;
  2. When the newborn is at least 24 but not more than 72 hours old; or
  3. Before the newborn is discharged, unless the newborn:
    a. Is transferred to another hospital before the newborn is 48 hours old; or
    b. Dies before the newborn is 72 hours old.

B. If a newborn is admitted or transferred to a hospital before the newborn is 48 hours old, the receiving hospital's designee shall:
  1. Verify that the first specimen was collected before admission or transfer, or
  2. Collect a first specimen from the newborn according to the requirements in subsection (A).

C. When a newborn is born in a birth center, the birth center's designee shall collect a first specimen from the newborn according to subsections (A)(1) or (A)(2).

D. For a home birth attended by a health care provider, the health care provider or the health care provider's designee shall collect a first specimen from the newborn according the requirements in subsection (A)(2).

Historical Note

Effective 11-74; Former Section R9-13-204 repealed, new Section R9-13-204 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 6, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective September 24, 1998 (Supp. 99-1). New Section recodified from R9-14-504 at 11 A.A.R. 3577, effective August 31, 2005
(Supp. 05-3). Section repealed; new Section made by final rulemaking at 12 A.A.R. 1166, effective April 4, 2006 (Supp. 06-2).

R9-13-205. Second Specimen Collection

A. After discharge from a health care facility or after a home birth, a health care provider or the health care provider's designee shall:
  1. Collect a second specimen from a newborn or infant:
    a. When the newborn is at least 5 but not more than 10 days old; or
    b. At the time of a newborn's or infant's first visit to the health care provider; or
  2. Verify that a different health care provider has collected the second specimen from the newborn or infant.

B. If a newborn is an inpatient of a health care facility at 5 days of age, the health care facility's designee shall collect a second specimen from the newborn:
  1. When the newborn is at least 5 but not more than 10 days old; or
  2. If the newborn is discharged from the facility when the newborn is at least 5 but not more than 10 days old, before discharge.

C. For a home birth not attended by a health care provider, a local health department's designee shall collect a specimen from a newborn or infant if a second specimen has not already been collected from the newborn or infant.

D. A health care provider or the health care provider's designee shall ensure that a subsequent specimen is ordered for a newborn or child one year of age or less, according to the requirements in R9-13-203, when the health care provider or the health care provider's designee:
  1. Begins providing health care to the newborn or child, and
  2. Cannot verify the results of a bloodspot test that was conducted on a second specimen from the newborn or child.

Historical Note

Effective 11-74; Former Section R9-13-205 repealed, new Section R9-13-205 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 6, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective September 24, 1998 (Supp. 99-1). New Section recodified from R9-14-505 at 11 A.A.R. 3577, effective August 31, 2005
(Supp. 05-3). Section repealed; new Section made by final rulemaking at 12 A.A.R. 1166, effective April 4, 2006 (Supp. 06-2).

R9-13-206. Reporting Requirements for Specimens

A. The screening laboratory shall:
  1. Report in written or electronic format:
    a. The results of a bloodspot test on a specimen; and
    b. For a specimen that does not meet the requirements for testing specified in R9-13-203(E):
      i. That the bloodspot test was not performed on the specimen; and
      ii. The reason the bloodspot test was not performed; and
  2. Send the report to:

a. The health care provider identified on the specimen collection kit;
b. If applicable, the health care facility identified on the specimen collection kit; and
c. The Department.

B. The screening laboratory shall begin reporting bloodspot test results for the congenital disorders specified in:
1. R9-13-202 (1) through (13), on the effective date of these rules;
2. R9-13-202(14) through (27), no later than August 31, 2006; and
3. R9-13-202(28), no later than June 30, 2007.

C. A health care facility's designee, a health care provider, or the health care provider's designee, who orders a subsequent test on a newborn or infant in response to an abnormal result on a bloodspot test, shall send the results of the subsequent test in writing to the Department, if the subsequent test is not performed by the screening laboratory.

D. Bloodspot test results are confidential subject to the disclosure provisions of 9 A.A.C. 1, Article 3, and A.R.S. §§ 12-2801 and 12-2802.

Historical Note

Effective 11-74; Repealed effective July 16, 1981 (Supp. 81-4). Adopted as an emergency effective November 6, 1989, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 89-4). Emergency expired. Readopted as an emergency effective February 7, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-1). Emergency expired. Readopted as an emergency with changes effective May 7, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-2). Readopted as an emergency with changes effective August 6, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-3). Readopted as an emergency without change effective October 31, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-4). Readopted as an emergency without change effective January 16, 1991, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 91-1). Readopted as an emergency without change effective April 11, 1991, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 91-2). Emergency rule permanently adopted with changes effective July 3, 1991 (Supp. 91-3). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective September 24, 1998 (Supp. 99-1). New Section made by final rulemaking at 12 A.A.R. 1166, effective April 4, 2006 (Supp. 06-2).

R9-13-207. Reporting Requirements for Hearing Test Results

A. When an initial hearing test is performed on a newborn, a health care facility's designee, a health care provider, or the health care provider's designee shall provide to the Department, as specified in subsection (E), the following information:
1. The newborn's name, date of birth, gender, and medical record number;
2. Whether the newborn is from a single or multiple birth;
3. If the newborn is from a multiple birth, the birth order of the newborn;
4. The newborn's mother's first and last names;
5. The name and identification code of the health care facility or health care provider submitting the hearing test results;
6. The name and identification code of the health care facility of birth;
7. The name of the health care provider responsible for the coordination of medical services for the newborn;
8. The date of the hearing test;
9. Whether or not the hearing test was performed when the newborn was an inpatient;
10. The audiological equipment used for the hearing test and the type of hearing test performed;
11. The hearing test result for each of the newborn's ears; and
12. The name, address, and phone number of the contact person for the health care facility or health care provider.

B. In addition to the information in subsection (A), if the reported results of an initial hearing test on a newborn include an abnormal result, a health care facility's designee, a health care provider, or the health care provider's designee shall provide to the Department, as specified in subsection (E), the following information:
1. The newborn's race, ethnicity, and if applicable, AHCCCS identification number;
2. Except as provided in subsection (B)(3), the mother's date of birth, name before first marriage, mailing address, and phone number;
3. If the newborn's mother does not have physical custody of the newborn, the first and last names, mailing address, and phone number of the person who has physical custody of the newborn;
4. The name of the health care provider who will be responsible for the coordination of medical services for the newborn after the newborn is discharged from the health care facility; and
5. The name and phone number of the person to whom the newborn's mother or other person who has physical custody of the newborn was referred for a subsequent hearing test.

C. When a hearing test is performed on a newborn or an infant after an initial hearing test, the designee of the health care facility, health care provider, or other person that performs the subsequent hearing test shall provide to the Department, as specified in subsection (E), the following information:
1. The newborn's or infant's name, date of birth, and gender;
2. Whether the newborn or infant is from a single or multiple birth;
3. If the newborn or infant is from a multiple birth, the birth order of the newborn or infant;
4. The newborn's or infant's mother's first and last names and date of birth;
5. The name of the health care facility where the initial hearing test was performed, or the name and address of the health care provider who performed the initial hearing test;

6. The name of the health care facility of birth;
7. The name and identification code of the person submitting the subsequent hearing test results;
8. The date of the subsequent hearing test;
9. The audiological equipment used for the subsequent hearing test and the type of hearing test performed;
10. The result for each of the newborn's or infant's ears on the subsequent hearing test; and
11. The name, address, and phone number of the contact person for the health care facility, health care provider, or other person that performed the subsequent hearing test.

D. In addition to the information in subsection (C), if the reported results of a subsequent hearing test on a newborn or infant include an abnormal result, the person submitting the report on the subsequent hearing test shall provide to the Department, as specified in subsection (E), the following information:
1. Except as provided in subsection (D)(2), the newborn's or infant's mother's mailing address and phone number;
2. If the newborn's or infant's mother does not have physical custody of the newborn or infant, the first and last names, mailing address, and phone number of the person who has physical custody of the newborn or infant;
3. The name of the health care provider who is responsible for the coordination of medical services for the newborn or infant; and
4. If applicable, the name and phone number of the person to whom the newborn's or infant's parent was referred for further hearing tests, evaluation services, specialty care, or early intervention.

E. A health care facility's designee, health care provider, health care provider's designee, or other person required to report under subsections (A), (B), (C), or (D) shall submit, in an electronic format specified by the Department, the information specified in subsections (A), (B), (C), or (D) for hearing tests performed each week by the sixth day of the subsequent week.

Historical Note

Effective 11-74; Repealed effective July 16, 1981 (Supp. 81-4). New Section made by final rulemaking at 12 A.A.R. 1166, effective April 4, 2006 (Supp. 06-2).

R9-13-208. Fees

A. The fee for a first specimen is $30.00.

B. The fee for a second specimen is $40.00.

Historical Note

New Section made by final rulemaking at 12 A.A.R. 1166, effective April 4, 2006 (Supp. 06-2).

ARTICLE 3. REPEALED

R9-13-301. Repealed

Historical Note

Effective 11-74; Former Section R9-13-301 repealed, new Section R9-13-301 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective September 10, 1997 (Supp. 99-1).

R9-13-302. Repealed

Historical Note

Effective 11-74; Former Section R9-13-302 repealed, new Section R9-13-302 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective September 10, 1997 (Supp. 99-1).

R9-13-303. Repealed

Historical Note

Effective 11-74; Former Section R9-13-303 repealed, new Section R9-13-303 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-304. Repealed

Historical Note

Effective 11-74; Former Section R9-13-304 repealed, new Section R9-13-304 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective September 10, 1997 (Supp. 99-1).

R9-13-305. Repealed

Historical Note

Effective 11-74; Repealed effective July 16, 1981 (Supp. 81-4).

R9-13-306. Repealed

Historical Note

Effective 11-74; Repealed effective July 16, 1981 (Supp. 81-4).

ARTICLE 4. REPEALED

R9-13-401. Repealed

Historical Note

Effective 11-74; Former Section R9-13-401 repealed, new Section R9-13-401 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-402. Repealed

Historical Note

Effective 11-74; Former Section R9-13-402 repealed, new Section R9-13-402 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-403. Repealed

Historical Note

Effective 11-74; Former Section R9-13-403 repealed, new Section R9-13-403 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-404. Repealed

Historical Note

Effective 11-74; Former Section R9-13-404 repealed, new Section R9-13-404 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-405. Repealed

Historical Note

Effective 11-74; Former Section R9-13-405 repealed, new Section R9-13-405 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-406. Repealed

Historical Note

Effective 11-74; Former Section R9-13-406 repealed, new Section R9-13-406 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-407. Repealed

Historical Note

Effective 11-74; Repealed effective July 16, 1981 (Supp. 81-4).

ARTICLE 5. REPEALED

R9-13-501. Repealed

Historical Note

Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-501 repealed, new Section R9-13-501 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective March 23, 1997 (Supp. 99-1).

R9-13-502. Repealed

Historical Note

Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-502 repealed, new Section R9-13-502 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective March 23, 1997 (Supp. 99-1).

R9-13-503. Repealed

Historical Note

Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-503 repealed, new Section R9-13-503 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-504. Repealed

Historical Note

Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-504 repealed, new Section R9-13-504 adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective March 23, 1997 (Supp. 99-1).

R9-13-505. Repealed

Historical Note

Adopted effective 1977 (Supp. 77-5). Repealed effective July 16, 1981 (Supp. 81-4).

R9-13-506. Repealed

Historical Note
Adopted effective 1977 (Supp. 77-5). Repealed effective July 16, 1981 (Supp. 81-4).

R9-13-507. Repealed

Historical Note
Adopted effective 1977 (Supp. 77-5). Repealed effective July 16, 1981 (Supp. 81-4).

R9-13-508. Repealed

Historical Note
Adopted effective 1977 (Supp. 77-5). Repealed effective July 16, 1981 (Supp. 81-4).

R9-13-509. Repealed

Historical Note
Adopted effective 1977 (Supp. 77-5). Repealed effective July 16, 1981 (Supp. 81-4).

R9-13-510. Repealed

Historical Note
Adopted effective 1977 (Supp. 77-5). Repealed effective July 16, 1981 (Supp. 81-4).

R9-13-511. Repealed

Historical Note
Adopted effective 1977 (Supp. 77-5). Repealed effective July 16, 1981 (Supp. 81-4).

ARTICLE 6. REPEALED

R9-13-601. Repealed

Historical Note
Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-601 repealed, new Section R9-13-601 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-602. Repealed

Historical Note
Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-602 repealed, new Section R9-13-602 adopted effective July 16, 1981 (Supp. 81-4). Amended effective July 3, 1991 (Supp. 91-3). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-603. Repealed

Historical Note
Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-603 repealed, new Section R9-13-603 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-604. Repealed

Historical Note
Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-604 repealed, new Section R9-13-604 adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-605. Repealed

Historical Note
Adopted effective October 26, 1977 (Supp. 77-5). Former Section R9-13-605 repealed, new Section R9-13-605 adopted effective July 16, 1981 (Supp. 81-4). Amended effective July 3, 1991 (Supp. 91-3). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-606. Repealed

Historical Note
Adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

ARTICLE 7. REPEALED

R9-13-701. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective June 1, 1997 (Supp. 99-1).

R9-13-702. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective June 1, 1997 (Supp. 99-1).

R9-13-703. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-704. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective June 1, 1997 (Supp. 99-1).

ARTICLE 8. REPEALED

R9-13-801. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed June 1, 2000 (Supp. 01-1).

R9-13-802. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Amended by emergency effective November 6, 1989, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 89-4). Emergency expired, Readopted as an emergency effective February 7, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-1). Emergency expired. Readopted as an emergency with changes effective May 7, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-2). Readopted as an emergency with changes effective August 6, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-3). Readopted as an emergency without change effective October 31, 1990, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 90-4). Readopted as an emergency without change effective January 16, 1991, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 91-1). Readopted as an emergency without change effective April 11, 1991, pursuant to A.R.S. § 41-1026, valid for only 90 days (Supp. 91-2). Emergency rule permanently adopted effective July 3, 1991 (Supp. 91-3). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed June 1, 2000 (Supp. 01-1).

R9-13-803. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-804. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Repealed effective December 16, 1996 (Supp. 96-4).

R9-13-805. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Amended effective July 3, 1991 (Supp. 91-3). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed by final rulemaking at 3 A.A.R. 146, effective June 30, 1998 (Supp. 99-1).

R9-13-806. Repealed

Historical Note

Adopted effective July 16, 1981 (Supp. 81-4). Amended effective December 16, 1996 (Supp. 96-4). Section automatically repealed June 1, 2000 (Supp. 01-1).

ARTICLE 9. REPEALED

R9-13-901. Repealed

Historical Note

Adopted as an emergency effective April 6, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-2). Former Section R9-13-901 expired, new Section R9-13-901 adopted as a permanent rule effective October 13, 1982 (Supp. 82-5). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-902. Emergency expired

Historical Note

Adopted as an emergency effective April 6, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-2). Former Section R9-13-902 expired (Supp. 82-5).

ARTICLE 10. REPEALED

R9-13-1001. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 12 A.A.R. 649, effective April 8, 2006 (Supp. 06-1).

R9-13-1002. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 12 A.A.R. 649, effective April 8, 2006 (Supp. 06-1).

R9-13-1003. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 12 A.A.R. 649, effective April 8, 2006 (Supp. 06-1).

R9-13-1004. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

ARTICLE 11. REPEALED

R9-13-1101. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 12 A.A.R. 649, effective April 8, 2006 (Supp. 06-1).

R9-13-1102. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 12 A.A.R. 649, effective April 8, 2006 (Supp. 06-1).

R9-13-1103. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1104. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 12 A.A.R. 649, effective April 8, 2006 (Supp. 06-1).

R9-13-1105. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1). New Section made by final rulemaking at 8 A.A.R. 2323, effective May 9, 2002 (Supp. 02-2). Section repealed by final rulemaking at 12 A.A.R. 649, effective April 8, 2006 (Supp. 06-1).

ARTICLE 12. REPEALED

R9-13-1201. Repealed

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired. Permanent rule adopted effective March 22, 1983 (Supp. 83-2). Section repealed by final rulemaking at 12 A.A.R. 649, effective April 8, 2006 (Supp. 06-1).

R9-13-1202. Emergency expired

Historical Note

Adopted as an emergency effective September 21, 1982, pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 82-5). Emergency expired (Supp. 83-2).

ARTICLE 13. REPEALED

R9-13-1301. Repealed

Historical Note

Adopted effective November 23, 1983 (Supp. 83-6). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1302. Repealed

Historical Note

Adopted effective November 23, 1983 (Supp. 83-6). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1303. Repealed

Historical Note

Adopted effective November 23, 1983 (Supp. 83-6). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

ARTICLE 14. REPEALED

R9-13-1401. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1403 renumbered and amended as permanent rule R9-13-1401 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1402. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1404 renumbered and amended as permanent rule R9-13-1402 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1403. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1405 renumbered as permanent rule R9-13-1403 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1404. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1406 renumbered and amended as permanent rule R9-13-1404 without

change effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1405. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1407 renumbered and amended as permanent rule R90-13-1405 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1406. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1408 renumbered and amended as permanent rule R9-13-1406 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1407. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1409 renumbered and amended as permanent rule R9-13-1407 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1408. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1410 renumbered and amended as permanent rule R9-13-1408 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1409. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1411 renumber and amended as permanent rule R9-13-1409 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1410. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1412 renumbered and amended as permanent rule R9-13-1410 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1411. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1413 renumbered and amended as permanent rule R9-13-1411 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1412. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1414 renumbered and amended as permanent rule R9-13-1412 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1413. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1415 renumbered and amended as permanent rule R9-13-1413 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1414. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1416 renumbered and amended as permanent rule R9-13-1414 effective March 19, 1984 (Supp. 84-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1415. Repealed

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1417 renumbered and amended as permanent rule R9-13-1415 effective March 19, 1984 (Supp. 84-2). Correction in subsection (C)(2) to insert the word `not' which was inadvertently omitted (Supp. 94-2). Section repealed by final rulemaking at 7 A.A.R. 1082, effective February 13, 2001 (Supp. 01-1).

R9-13-1416. Emergency expired

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1416 renumbered and amended as permanent rule R9-13-1414 effective March 19, 1984 (Supp. 84-2).

R9-13-1417. Emergency expired

Historical Note

Adopted as an emergency effective November 29, 1983 pursuant to A.R.S. § 41-1003, valid for only 90 days (Supp. 83-6). Emergency expired. Former Section R9-13-1417 renumbered and amended as permanent rule R9-13-1414 effective March 19, 1984 (Supp. 84-2).

Editor's Note: Article 15 was recodified to 9 A.A.C. 25, Article 8 (Supp. 98-1).

Editor's Note: Former Article 15 contained Sections and Exhibits which were adopted under an exemption from the provisions of the Administrative Procedure Act (A.R.S. Title 41, Chapter 6) pursuant to A.R.S. § 36-2205(C). Exemption from A.R.S. Title 41, Chapter 6 means that the Department of Health Services did not submit these rules to the Governor's Regulatory Review Council for review; the Department did not submit notice of proposed rulemaking to the Secretary of State for publication in the Arizona Administrative Register; the Department was not required to hold public hearings on these rules; and the Attorney General did not certify these rules.

ARTICLE 15. RECODIFIED

R9-13-1501. Recodified

Historical Note

Adopted effective July 11, 1994; received by the Office of the Secretary of State August 4, 1994, under an exemption from the provisions of the Administrative Procedure Act pursuant to A.R.S. § 36-2005(C) (Supp. 94-3). Former Section R9-13-1501 recodified to A.A.C. R9-25-801 (Supp. 98-1).

R9-13-1502. Recodified

Historical Note

Adopted effective October 12, 1994; received by the Office of the Secretary of State October 24, 1994, under an exemption from the provisions of the Administrative Procedure Act pursuant to A.R.S. § 36-2205(C) (Supp. 94-4). Former Section R9-13-1502 recodified to A.A.C. R9-25-802 (Supp. 98-1).

Exhibit 1. Recodified

Historical Note

Adopted effective July 11, 1994; received by the Office of the Secretary of State August 4, 1994, under an exemption from the provisions of the Administrative Procedure Act pursuant to A.R.S. § 36-2005(C) (Supp. 94-3). Former R9-13-1502, Exhibit 1 recodified to A.A.C. R9-25-802, Exhibit 1 (Supp. 98-1).

Exhibit 2. Recodified

Historical Note

Adopted effective July 11, 1994; received by the Office of the Secretary of State August 4, 1994, under an exemption from the provisions of the Administrative Procedure Act pursuant to A.R.S. § 36-2005(C) (Supp. 94-3). Former R9-13-1502, Exhibit 2 recodified to A.A.C. R9-25-802, Exhibit 2 (Supp. 98-1).

Exhibit 3. Recodified

Historical Note

Adopted effective July 11, 1994; received by the Office of the Secretary of State August 4, 1994, under an exemption from the provisions of the Administrative Procedure Act pursuant to A.R.S. § 36-2005(C) (Supp. 94-3). Former R9-13-1502, Exhibit 3 recodified to A.A.C. R9-25-802, Exhibit 3 (Supp. 98-1).

Exhibit 4. Recodified

Historical Note

Adopted effective July 11, 1994; received by the Office of the Secretary of State August 4, 1994, under an exemption from the provisions of the Administrative Procedure Act pursuant to A.R.S. § 36-2005(C) (Supp. 94-3). Former R9-13-1502, Exhibit 4 recodified to A.A.C. R9-25-802, Exhibit 4 (Supp. 98-1).

R9-13-1503. Recodified

Historical Note

Adopted effective November 27, 1995, under an exemption from the provisions of the Administrative Procedure Act pursuant to A.R.S. § 36-2205(C) (Supp. 95-4). Former Section R9-13-1503 recodified to A.A.C. R9-25-803 (Supp. 98-1).

Exhibit 1. Recodified

Historical Note

Adopted effective November 27, 1995, under an exemption from the provisions of the Administrative Procedure Act pursuant to A.R.S. § 36-2205(C) (Supp. 95-4). Former R9-13-1503, Exhibit 1 recodified to A.A.C. R9-25-803, Exhibit 1 (Supp. 98-1).

NEWBORN SCREENING - PARENT REFUSAL FORM

Name of Infant	Hospital of Birth
Date of Birth	Hospital Street Address
Medical Record Number	City/State/Zip

I, _____, have received current information about the Arizona Department
Parent's Name
of Health Services' Newborn Screening Program. I understand there are many rare, inherited disorders for which Arizona newborns are screened.

I have been informed and understand that these tests are offered by State Law for all infants born in Arizona.

I have been informed and understand that, if untreated, these conditions may cause permanent damage to my child, including serious mental retardation, growth failure and, in some cases, death.

I have discussed the testing requirements with_____. I have had the
Healthcare Provider
testing requirements explained to me, and I understand all the risks involved if the screening tests are not given to my child.

I have been informed and understand the nature of the screening tests and how these tests are given.

I object to these tests, and I do not want _____ tested for the conditions at
Child's Name
this time. I understand that I may request the Newborn Screening from my physician at a future date, but no later than my child is one year of age.

My decision was freely made without undue influence or encouragement by any person.

Printed Name	Relationship to Child
Signature	Date
Printed Name of Witness	Witness Title/Address
Witness Signature	Date

Original: Infant's Medical Record
Copies: Newborn Screening Follow-up Program
Parent, Healthcare Provider

Newborn Screening
1st

PRINT ALL INFORMATION ENTERED

Accession Number: 18

SPECIMEN Date / Time Stamp

Baby's Name

Last: 1 First:

Date of Birth 2	Time of Birth 3 a.m. p.m.	Birth Weight 4 Grams Lb/oz	Sex
___/___/___	___	___	M 8 F
Date of Collection 5	Time of Collection 6 a.m. p.m.	Current Weight 7 Grams Lb/oz	
___/___/___	___	___	

Method of Collection: 9 ☐ Heelstick (preferred) ☐ Venipuncture

Baby's AHCCCS # 10

Medical Record # 11

12 ☐ Single Birth ☐ Multiple Birth (circle one) A B C D

Race 13	Food Source 15	Status 16
1 White	1 Breast Only	Sick/Premature Y N
2 African Amer.	2 Milk Formula Only	Cleft Lip/Palate Y N
3 Asian	3 Soy Formula Only	Neural Tube Def. Y N
4 Amer. Indian	4 Breast & Milk Formula	Transfused Y N
5 Other	5 Breast & Soy Formula	Date Last Transfused 17
Hispanic Y N 14	6 TPN 7 NPO	

AZ 091029353 SN

Submitter / Physician Information AZ091029353 SN

Submitter Name/ID: 19

Submitter Address: 20

Physician's Name (Last, First): 21

Phone: (22)

Physician's Address: 23

City, State, Zip: 24

Mother's Information

Mom's Name Last: 25 First:

Mom's Date of Birth: 26 / / Maiden Name: 27

Street Address: 28

City, State, Zip: 29

Phone: (30)

Message Phone: (31)

Mom's AHCCCS #: 32 ☐ Parent Refused Testing 33

EXP DATE 2011-12 Alsion 226 8040201 / Lot 0901072 AZ091029353 SN

2011-12

Newborn Screening
2nd

PRINT ALL INFORMATION ENTERED

Accession Number:

SPECIMEN Date / Time Stamp

Baby's Name

Last: First:

Date of Birth	Time of Birth a.m. p.m.	Birth Weight Grams Lb/oz	Sex
___/___/___	___	___	M F
Date of Collection	Time of Collection a.m. p.m.	Current Weight Grams Lb/oz	
___/___/___	___	___	

Method of Collection: ☐ Heelstick (preferred) ☐ Venipuncture

Baby's AHCCCS #

Medical Record #

☐ Single Birth ☐ Multiple Birth (circle one) A B C D

Race	Food Source	Status
1 White	1 Breast Only	Sick/Premature Y N
2 African Amer.	2 Milk Formula Only	Cleft Lip/Palate Y N
3 Asian	3 Soy Formula Only	Neural Tube Def. Y N
4 Amer. Indian	4 Breast & Milk Formula	Transfused Y N
5 Other	5 Breast & Soy Formula	Date Last Transfused
Hispanic Y N	6 TPN 7 NPO	

AZ 092029353 SN

Submitter / Physician Information AZ092029353 SN

Submitter Name/ID:

Submitter Address:

Physician's Name (Last, First):

Phone: ( )

Physician's Address:

City, State, Zip:

Mother's Information

Mom's Name Last: First:

Mom's Date of Birth: / / Maiden Name:

Street Address:

City, State, Zip:

Phone: ( )

Message Phone: ( )

Mom's AHCCCS #: ☐ Parent Refused Testing

EXP DATE 2011-12 Alsion 226 8040201 / Lot 0901072 AZ092029353 SN

Newborn Screening

A Guide for Parents

NBS-PB (01/10)

What is Newborn Screening?

Babies born in Arizona have a few drops of blood taken from their heels to test for certain medical disorders. They also have their hearing tested.

Why are babies tested?

- The blood tests could save your baby's life.
- Most babies are healthy when they are born. A few babies look healthy but have a rare health problem.
- If the problem is found early, we can help prevent serious results like mental retardation or death.
- Finding hearing loss will help your child learn speech and language.

When are babies tested?

- The hospital will do the first blood spot test and the hearing test.
- Your baby's doctor will order the second blood spot test at your first visit.

For what disorders are babies tested?

- The 29 core disorders including hearing loss on the Recommended Uniform Screening Panel from the U.S. Department of Health and Human Services (HHS)
- This panel is also endorsed by the American Academy of Pediatrics and the March of Dimes.

How will I know the results?

Blood spot results:

- Ask you baby's doctor for the results of both the first and second tests.
- Give the hospital and the baby's doctor your correct address and phone number.

Hearing results:

- The hospital will write the results on the back of the immunization record. Bring the blue shot card to your baby's doctor.
- A second test may need to be done.

What if my baby's results are abnormal?

- You will get a call from your baby's doctor asking you to get more testing done immediately.
- An abnormal second blood spot or hearing test may not mean your baby has a problem. Your baby may be referred to a specialist for diagnostic testing and treatment. If needed, the Arizona program will help you get special services for your baby.

Where can I get more information?

- Ask your baby's doctor
- Call the Newborn Screening Program at 602-364-1409 (1-800-548-8381 outside the Phoenix metropolitan area)
- Visit our website at: www.aznewborn.com

Arizona Department of Health Services

Arizona Department of Health Services
Bureau of State Laboratory Services
Newborn Screening Program
250 N. 17th Ave., 1st Floor
Phoenix, Arizona 85007-3231
Phone: 602-364-1409
Outside the Phoenix metropolitan area:
1-800-548-8381
Deaf and Hard of Hearing call 711
for AZ Relay Service

Análisis para niños recién nacidos

Una Guía para Padres

NBS-PB (01/10)

¿Qué es la evaluación de los recién nacidos?

A los bebés que nacen en Arizona se les toma unas gotas de sangre de sus talones para evaluar si padecen de ciertos trastornos. También se les evalúa la audición.

¿Por qué se evalúa a los bebés?

- Las pruebas de sangre pudieran salvar la vida de su bebé.
- La mayoría de los bebés son saludables al nacer. Unos cuantos bebés lucen saludables pero tienen problemas de salud pocos comunes.
- Si se detecta el problema a tiempo, podemos ayudar a prevenir resultados graves, tales como el retraso mental o la muerte.
- La detección de la pérdida de la audición puede ayudar a su niño/a a aprender el lenguaje y a hablar.

¿Cuándo se evalúa a los bebés?

- El hospital hará la primera prueba de sangre y la prueba de oído.
- El/la doctor(a) de su bebé mandará a que le hagan la segunda prueba de muestreo de sangre en la primera cita.

¿Qué trastornos se evalúan en los bebés?

- Los 29 trastornos fundamentales, incluso la pérdida de audición, que enumera la Lista Uniforme de Diagnósticos Recomendados de la Secretaría de Salud y Servicios Humanos de los Estados Unidos (HHS)
- La Academia Americana de Pediatría y la Fundación March of Dimes recomiendan la misma lista.

¿Cómo me enteraré de los resultados?

Prueba de muestreo de sangre:

- Pídale al o a la doctor(a) de su bebé que le informe los resultados de la primera y la segunda prueba.
- Dele el domicilio y el teléfono correctos al hospital y al/a la doctor(a) de su niño/a.

Resultado de la prueba de audición:

- En el hospital apuntarán el resultado de la prueba al dorso de la tarjeta de vacunas. Lleve la tarjeta azul de vacunas al/a la doctor(a) de su bebé.
- Tal vez haya que hacer otra prueba.

¿Qué sucede si la prueba de mi bebé no resulta normal?

- El/la doctor(a) de su bebé se comunicará con usted para pedirle que le hagan más pruebas de inmediato.
- Que el segundo muestreo de sangre o de la audición no resulte normal no tiene que significar que su bebé tenga un problema. Pudieran enviar a su bebé a un(a) especialista para pruebas diagnósticas y tratamiento. Si hiciera falta, el programa de Arizona le ayudará a conseguir servicios especiales para su bebé.

¿Dónde puedo obtener mayor información?

- Pregúntele al/a la doctor(a) de su bebé
- Llame al Programa de Análisis Para Recién Nacidos al 602-364-1409 (1-800-548-8381 fuera del área metropolitana de Phoenix)
- Visite nuestra página en Internet: www.aznewborn.com

Arizona Department of Health Services

Departamento de Servicios de Salud de Arizona
Negociado de Servicios Estatales de Laboratorios
Programa de Análisis Para Recién Nacidos
250 N. 17th Ave., Piso 1°
Phoenix, Arizona 85007-3231
Teléfono: 602-364-1409
Fuera del área metropolitana de Phoenix:
1-800-548-8381
Los sordos o con dificultades para oír llamen
al AZ Relay Service al 711

ADHS - Newborn Screening Kit
Order Form

Please do not write in shaded areas

Order Date: _____

Ship Date: _____

Submitter ID: _____

Contact & Ph # _____

Ship To:

Submitter Name: _____

Attn: _____

Address _____

City _____,AZ Zip: _____

Special _____

Instructions _____

Linked Kits	Qty
Linked Kits	
WHITE Envelopes	
PINK Envelopes	

Starting Kit #	Ending Kit #

Supplementals	Qty
Supplementals	
White Envelopes	

Starting Kit #	Ending Kit #

Order Taken By: _____

Order Pulled By: _____

Verified and Shipped By: _____

To Place Order, Please Fax Form To: 602-542-0760, Or Call: 602-542-1190 ... Thank You!

Newborn Screening Program Order Form

Please Print and Fill Out the Following Information Completely.
Please See the Unit Quantities and Maximums and Enter Your Requirements Accordingly.
Incomplete Information Will Cause a Delay in Processing Your Order.

Date of Request	Requestor's Area Code & Telephone Number	Requestor's Area Code and Fax Number
Organization or Agency		
Number & Street Address		Room No./Floor
City	State	Zip Code
Ship to the Attention of (Name)	Department	

Date Order Received @ NBS	Date Order Sent	By

These Items will be shipped directly from Standard Register.

Item Number	Brochure or Item Name	No per Unit Maximums	Unit	Units Ordered	Units Shipped
NBS-PB	Newborn Screening: A Guide for Parents **(English/Spanish)**	100/Pkg *Max: 10 Pkgs.*	Pkg.		
NBS-EHDI01	Universal Newborn Hearing Screening **(English/Spanish)** for Hospitals	25/Pkgs. *Max:* 10 Pkgs.	Pkg.		
NBS-NHS	Newborn Hearing Screening Labels **(for back of Lifetime Immunization Record)**	250/Roll *Max:* 10 Rolls	Roll		

These Items will be Shipped from Newborn Screening.

	Your Baby's Hearing **(English) for Doctors/Clinics**	100/Pkg *Max: 10 Pkgs.*	Pkg.		
	Your Baby's Hearing **(Spanish) for Doctors/Clinics**	100/Pkg *Max: 10 Pkgs.*	Pkg.		
	Arizona Pediatric Audiology Guidelines	*Max:* 2	Ea.		
	Arizona Hospitals' Universal Newborn Hearing Screening Guidelines	*Max:* 2	Ea.		

You May Fax or Mail Your Order to the Newborn Screening Program:

FAX YOUR ORDER TO:	MAIL YOUR ORDER TO:
(602) 364-1495	***Arizona Department of Health Services*** ***Attn: Newborn Screening Program*** ***250 N. 17th Ave., 1st Floor*** ***Phoenix, AZ 85007-3231***

If You Have Any Questions, Please Call (602) 364-1409 or 1-800-548-8381 (outside Phoenix area)

Please Allow Two (2) Weeks for Your Order to be Processed and Shipped

ARIZONA DEPARTMENT OF HEALTH SERVICES
BUREAU OF STATE LABORATORY SERVICES
250 North 17th Avenue Phoenix, Arizona 85007
State Lab 602-364-3190 http://www.AZNewborn.com

ARIZONA NEWBORN SCREENING REPORT

Date	: 05/13/2010	Specimen Type*	: First Screen
Infant's Name	:	Lab Number	: 2010131
Date of Birth	: 05/05/2010 @ 17:37	Patient Number	: 2010131
Date of Collection	: 05/08/2010 @ 05:10	Medical Record	:
Date Received	: 05/11/2010	Sex	: Female
Mother's Name	:	Race	: White
Address	:	Birth Weight	:
City/St/Zip	:	Transfused	: No
Phone	:	Date Transfused+	:
Physician	:	Food Source	: Breast & Lactose
Submitter	:	Kit Number	:

SCREENING RESULTS

365

DISORDERS	RESULTS
Endocrine Disorders	Normal
Hemoglobinopathies	Normal
Biotinidase Deficiency	Normal
Galactosemia	Normal
Amino Acid Disorders	Normal
Fatty Acid Oxidation Disorders	Normal
Organic Acid Disorders	Normal
Cystic Fibrosis	CF not indicated by IRT

*****Effective 9/28/09: Please note that birthweight ranges and cut-offs for CAH have been changed. Please refer to our website, www.aznewborn.com, for more information.**

*A second screen is required for all babies born in Arizona. If this specimen is the FIRST SCREEN, please collect an additional specimen at the first visit to a healthcare provider after discharge from the hospital or no later than five to ten days of age.
+ Unless transfusion is marked, the assumption is that the infant has not been transfused.

The purpose of the Arizona Department of Health Services Newborn Screening Program is to identify infants at increased risk for a variety of disorders. Since this is a screening test, the possibility of a false positive or negative result must be considered. The test may need to be repeated and diagnosis confirmed or ruled out by additional specialized studies. A negative screen does not rule out the possibility of a disorder. Health care providers should remain watchful for any signs or symptoms of these disorders with their patients.

ARIZONA DEPARTMENT OF HEALTH SERVICES
BUREAU OF STATE LABORATORY SERVICES
250 North 17th Avenue Phoenix, Arizona 85007
State Lab 602-364-3190 http://www.AZNewborn.com

ARIZONA NEWBORN SCREENING REPORT

Date	: 06/03/2010	Specimen Type*	: Second Screen
Infant's Name	:	Lab Number	: 2010152
Date of Birth	: 05/21/2010 @ 01:06	Patient Number	: 2010145
Date of Collection	: 05/26/2010 @ 14:50	Medical Record	:
Date Received	: 06/01/2010	Sex	: Female
Mother's Name	:	Race	: White
Address	:	Birth Weight	:
City/St/Zip	:	Transfused	: No
Phone	:	Date Transfused+	:
Physician	:	Food Source	:
Submitter	:	Kit Number	:

SCREENING RESULTS

15

DISORDER GROUP	RESULT VALUE	RESULT
Endocrine Disorders	Normal	Normal
Hemoglobinopathies	FA	Normal
Biotinidase Deficiency	Normal	Normal
Galactosemia	Normal	Normal
Amino Acid Disorders	Normal	Normal
Fatty Acid Oxidation Disorders	Normal	Normal
Organic Acid Disorders	**C5OH 1.13 µmol/L**	**Abnormal - See Comments (HMG or 3MCC)**
Cystic Fibrosis (CF)	Refer To First Screen Results.	

Comments

(3-Hydroxy-3-methylglutaric aciiduria - HMG or 3-Methylcrotonyl coA carboxylase deficiency - 3MCC) - Please consult the Phoenix metabolic genetics specialists at (602) 406-3611.

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+ Unless transfusion is marked, the assumption is that the infant has not been transfused.

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ARIZONA DEPARTMENT OF HEALTH SERVICES
BUREAU OF STATE LABORATORY SERVICES
250 North 17th Avenue Phoenix, Arizona 85007
State Lab 602-364-3190 http://www.AZNewborn.com

ARIZONA NEWBORN SCREENING REPORT

Date	: 07/14/2010	Specimen Type*	: Second Screen
Infant's Name	:	Lab Number	: 2010193
Date of Birth	: 06/18/2010 @ 23:15	Patient Number	: 2010175
Date of Collection	: 07/07/2010 @ 10:30	Medical Record	:
Date Received	: 07/12/2010	Sex	: Male
Mother's Name	:	Race	: NP*
Address	:	Birth Weight	:
City/St/Zip	:	Transfused	: No
Phone	:	Date Transfused+	:
Physician	:	Food Source	: Lactose Formula
Submitter	:	Kit Number	:

*NP = Not Provided

SCREENING RESULTS

DISORDER GROUP	RESULT VALUE	RESULT
Endocrine Disorders	Normal	Normal
Hemoglobinopathies	**FAS**	**See Comments (Hemoglobin)**
Biotinidase Deficiency	Normal	Normal
Galactosemia	Normal	Normal
Amino Acid Disorders	Normal	Normal
Fatty Acid Oxidation Disorders	Normal	Normal
Organic Acid Disorders	Normal	Normal
Cystic Fibrosis (CF)	Refer To First Screen Results	

Comments

(Hemoglobin) - Possible Sickle Cell Trait

*****Effective 9/28/09: Please note that birthweight ranges and cut-offs for CAH have been changed. Please refer to our website, www.aznewborn.com, for more information.**

*A second screen is required for all babies born in Arizona. If this specimen is the FIRST SCREEN, please collect an additional specimen at the first visit to a healthcare provider after discharge from the hospital or no later than five to ten days of age.
+ Unless transfusion is marked, the assumption is that the infant has not been transfused.

The purpose of the Arizona Department of Health Services Newborn Screening Program is to identify infants at increased risk for a variety of disorders. Since this is a screening test, the possibility of a false positive or negative result must be considered. The test may need to be repeated and diagnosis confirmed or ruled out by additional specialized studies. A negative screen does not rule out the possibility of a disorder. Health care providers should remain watchful for any signs or symptoms of these disorders with their patients.

ARIZONA DEPARTMENT OF HEALTH SERVICES
BUREAU OF STATE LABORATORY SERVICES
250 North 17th Avenue Phoenix, Arizona 85007
State Lab 602-364-3190 http://www.AZNewborn.com

(Updated)

ARIZONA NEWBORN SCREENING REPORT

Date	: 07/21/2010	Specimen Type*	: First Unsat
Infant's Name	:	Lab Number	: 2010172
Date of Birth	: 06/14/2010 @ 09:46	Patient Number	: 2010172
Date of Collection	: 06/15/2010 @ 12:00	Medical Record	:
Date Received	: 06/21/2010	Sex	: Male
Mother's Name	:	Race	: Asian
Address	:	Birth Weight	:
City/St/Zip	:	Transfused	: NP*
Phone	:	Date Transfused+	:
Physician	:	Kit Number	:
Submitter	:		

*NP = Not Provided

SCREENING RESULTS

Specimen Unsatisfactory For Testing

Unsatisfactory specimen due to multiple specimen applications. Please send another newborn screening sample as soon as possible.

Resubmit Another Newborn Screening Specimen Promptly

***Effective 9/28/09: Please note that birthweight ranges and cut-offs for CAH have been changed. Please refer to our website, www.aznewborn.com, for more information.

*A second screen is required for all babies born in Arizona. If this specimen is the FIRST SCREEN, please collect an additional specimen between five and ten days of age or at the first provider visit after discharge from the hospital

+ Unless transfusion is marked, the assumption is that the infant has not been transfused

The purpose of the Arizona Department of Health Services Newborn Screening Program is to identify infants at increased risk for a variety of disorders. Since this is a screening test, the possibility of a false positive or negative result must be considered. The test may need to be repeated and diagnosis confirmed or ruled out by additional specialized studies.

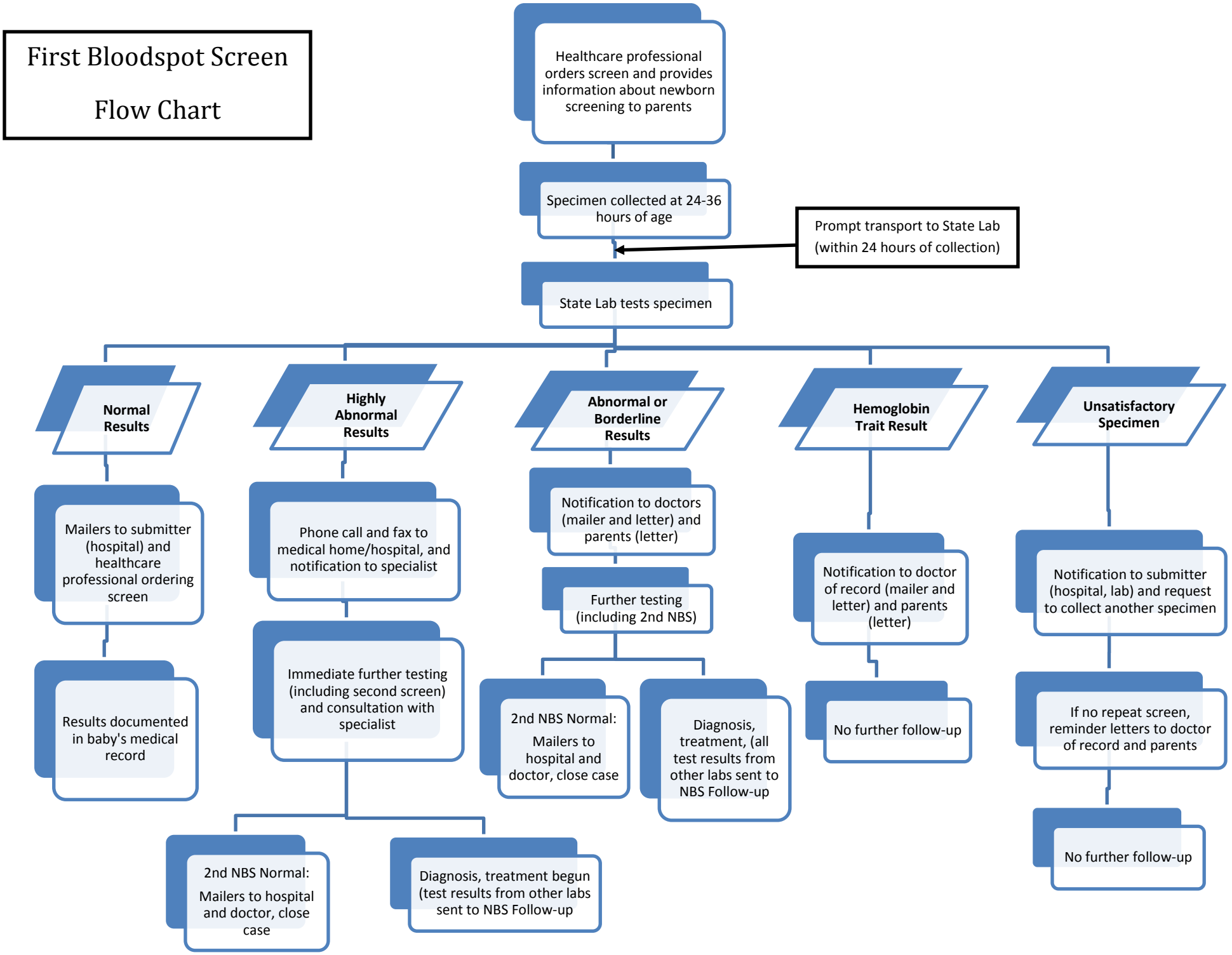
HEMOGLOBIN RESULTS

Hemoglobin results are reported in order of predominance with the highest percentage listed first:

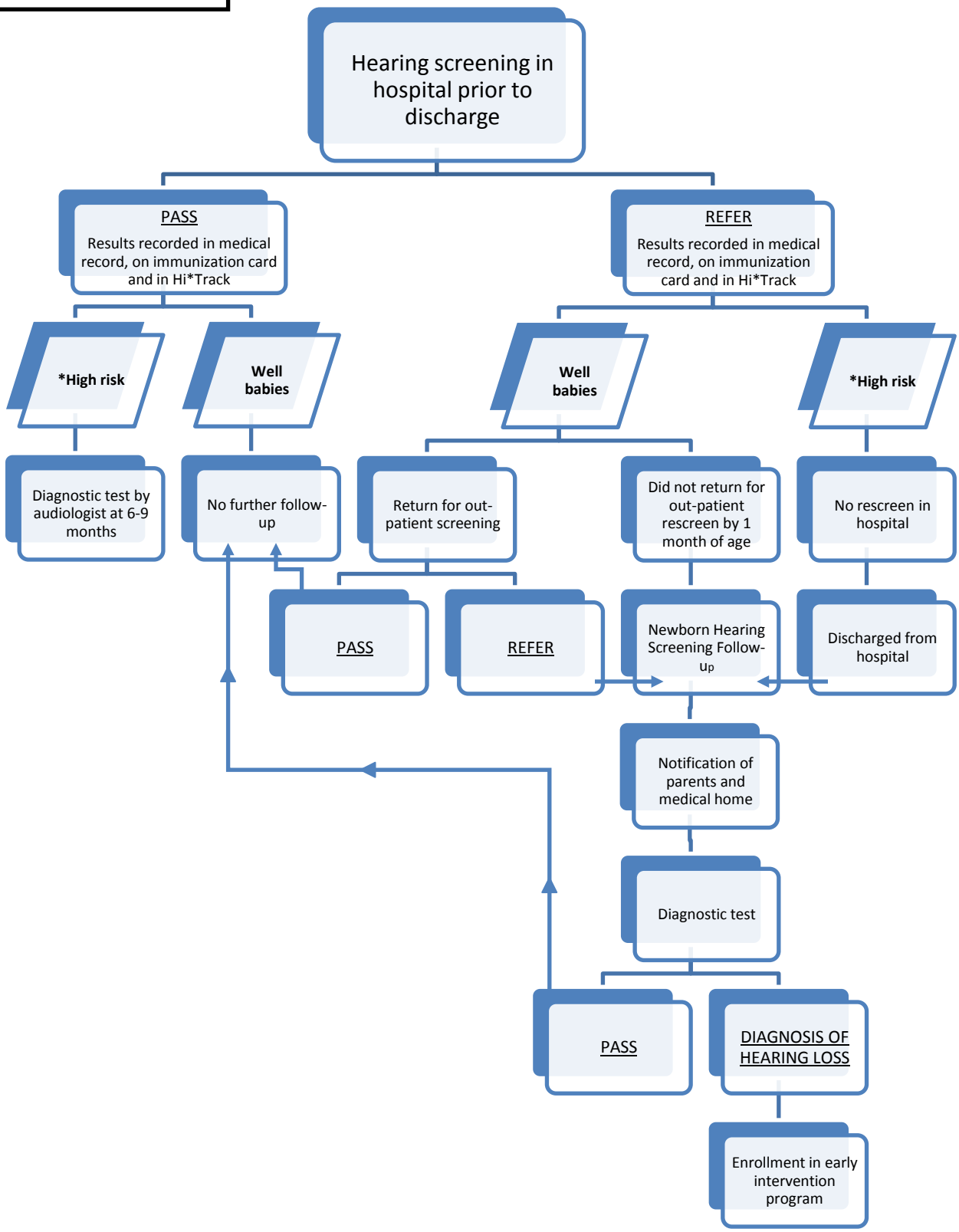
F = fetal hemoglobin (2 alpha globin chains, 2 gamma globin chains)
A = adult hemoglobin (2 alpha globin chains, 2 beta globin chains)
S, C, D, G, E, O = hemoglobin, each with a different mutation in the beta globin chains
U = unidentified hemoglobin variant
Barts = non-oxygen carrying, transient hemoglobin variant (4 gamma globin chains)

Mnemonic	Description
FA	Normal newborn infant hemoglobin pattern with fetal hemoglobin (F) predominant plus measurable A hemoglobin
AF	Normal older infant result where fetal hemoglobin is declining and A hemoglobin is greater than 50%. It could also indicate a possible transfusion in a newborn.
PAH	Predominantly adult hemoglobin (would be reported as AA in older individuals) in an infant less than 60 days old. This indicates a probable prior transfusion.
AA	Normal child or adult hemoglobin with A hemoglobin present and no other variant hemoglobin in measurable amounts
FS, SS	Probable sickle cell anemia - could also be S/ß° thalassemia or S/hereditary persistence of fetal hemoglobin (Hb S/HPFH)
FSU	Possible sickle cell anemia or sickle ß thalassemia
FSA, SA	Possible S/ß+ thalassemia or Sickle cell trait
FSA1	Sickle cell trait or possible S/ß+ thalassemia
FAS, AFS, ASF, AS	Probable sickle cell trait
FSC, SC	Probable Sickle C disease
FC	Probable Hemoglobin C disease (homozygous) or Hemoglobin C with thalassemia
FAC, AFC, ACF	Probable Hemoglobin C trait
F only	Possible ß thalassemia in full term infant or a premature infant (not yet producing measurable A hemoglobin)
FE, EE	Possible Hemoglobin E disease (homozygous) or Hemoglobin E with thalassemia
FSE	Possible Sickle E disease
FAE, AFE, AEF, AE	Probable Hemoglobin E trait
FD	Possible Hemoglobin D disease (homozygous) or Hemoglobin D with thalassemia
FAD, AFD, AD	Probable Hemoglobin D trait
FAD/G	Possible Hemoglobin D or G trait
FAG	Probable Hemoglobin G trait
FAO	Probable Hemoglobin O trait
FU	Possible unidentified hemoglobin disease
FUA	Possible ß+ thalassemia trait or Thalassemia Intermedia
FAU, AFU	Possible unknown (slow migrating) hemoglobin variant
FA fast, AFFAST	Possible unknown (fast migrating) hemoglobin variant
Bart 10	At least 10% Barts along with F and A hemoglobin - Possible alpha thalassemia trait. Barts may be present in normal newborns and is even more likely to appear in premature infants
Bart 15	15% or more Barts along with F and A hemoglobin - Possible alpha thalassemia trait or Hemoglobin H disease

First Bloodspot Screen Flow Chart



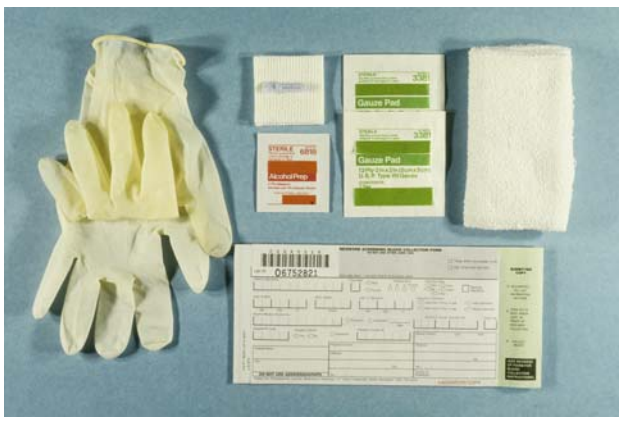
Hearing Screen Flow Chart



Newborn Screening

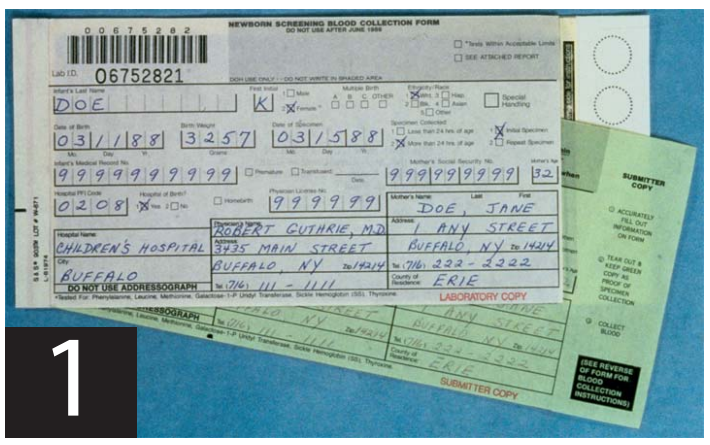
Sample Collection and Handling Procedure

ID Biological SYSTEMS

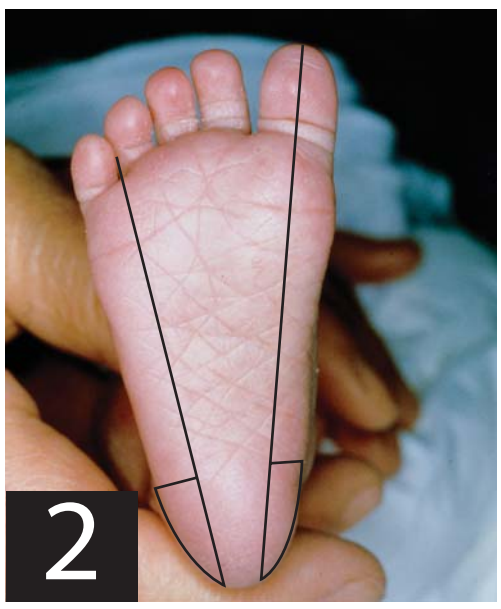


Before collecting the sample, the following materials must be gathered:

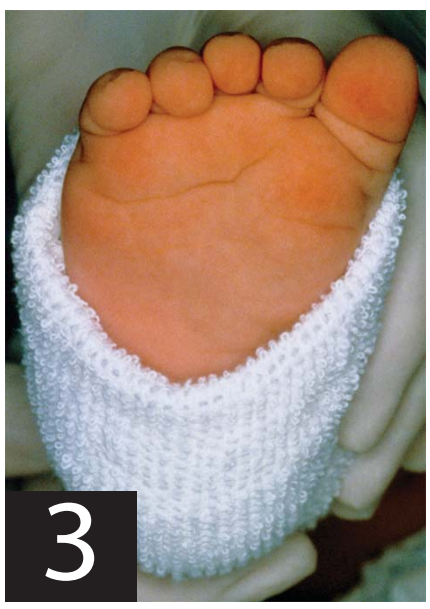
- Sterile lancet with tip less than 2.4mm long
- Sterile 70% alcohol pads
- Sterile gauze pads
- Warm moist towel or compress
- Fully completed, in-date filter paper blood collection form
- Sterile gloves



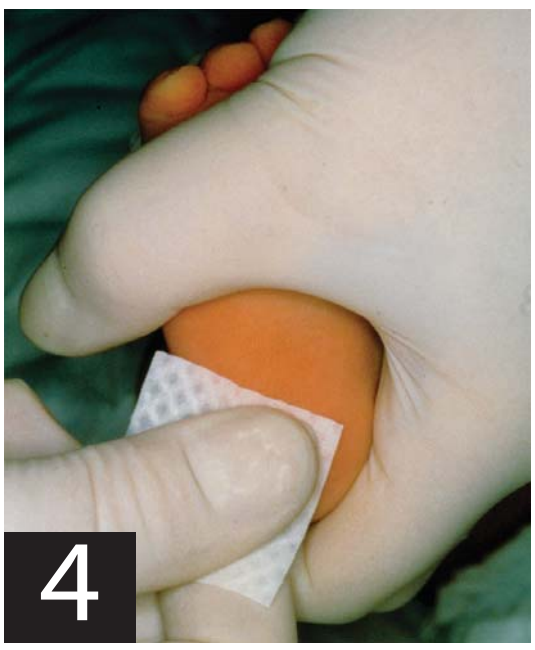
All fields on the form must be completed. Take steps to ensure that the filter paper is not touched or contaminated in any way during this process. If a submitter's copy is present, remove it and keep it for your records.



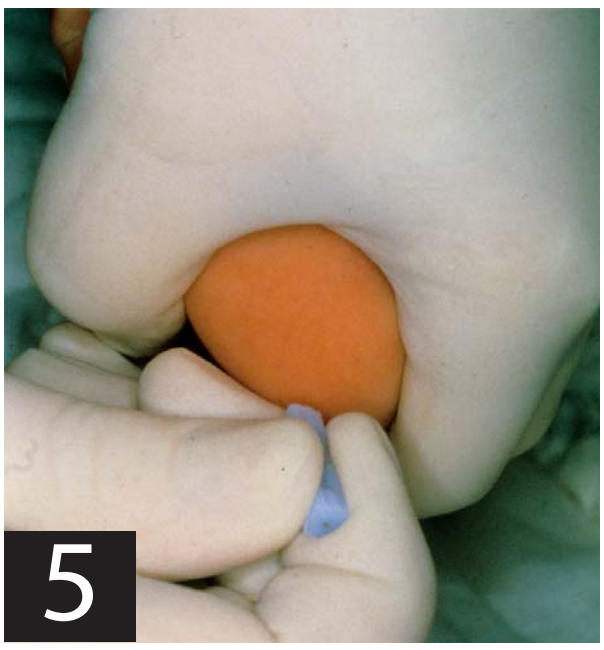
Areas on the side of the heel containing hatch marks are acceptable puncture sites.



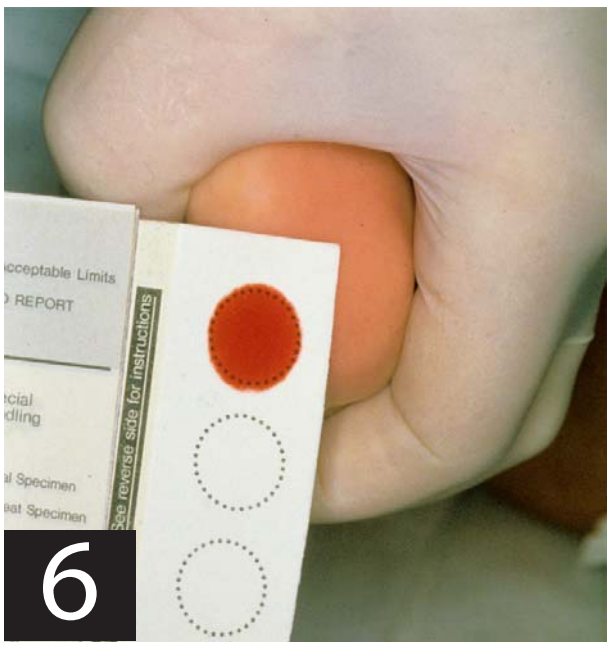
Warm puncture site for 3-5 minutes with a warm, moist towel no hotter than 42˚C.



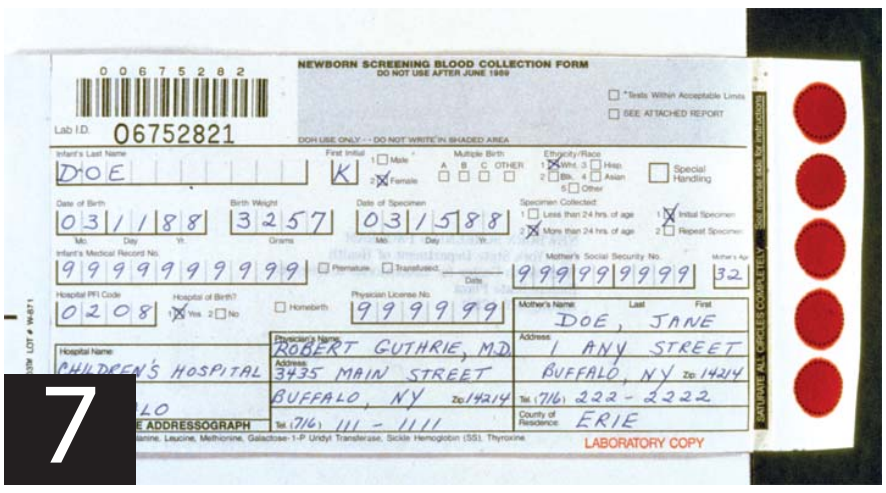
Cleanse the puncture site with a sterile alcohol pad and allow to air dry.



Puncture skin and wipe away the first drop of blood. Allow a second large drop to form



Lightly touch the filter paper to the second drop of blood. Allow the blood to soak through and fill the preprinted circle. DO NOT APPLY MORE THAN ONE DROP TO A CIRCLE. Fill all remaining circles in this manner. If blood flow is diminished, repeat steps 3-6 with sterile equipment.



Allow the speciment to dry in a horizontal position for at least three hours, away from direct contact with surfaces, heat, and sunlight.

Send the specimen to the appropriate location no more than 24 hours after collection.

ID Biological SYSTEMS

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USA: 17 P & N Drive • Greenville, SC 29611 USA • Tel. (864) 299-8787 • Fax (864) 299-8797 • id-biological.com • bdavin@id-biological.com • ggaillard@id-bioloigcal.com
UK: 15 Tanners Hill • Abbots Langley • Hertfordshire WD5 0LT UK • Tel. + 44 (0) 7525 844431 • Fax + 44 (0) 1923 682120 • id-biological.com • jdinan@id-biological.com

Blood Spot Check

ID Biological SYSTEMS

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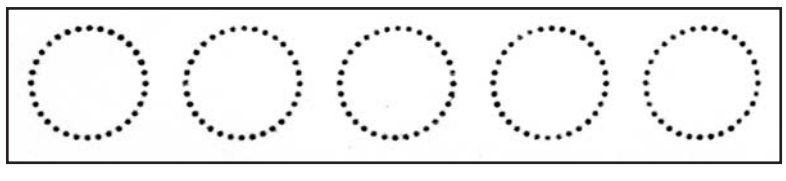
Acceptable Specimen

- No foreign substances have contaminated the filter paper.
- The blood fills all printed circles and is applied evenly on one side of the filter paper only, free of layering and clots.
- The specimen dried in a horizontal position for at least three hours, away from direct contact with surfaces, heat, and sunlight.
- The speciemen is sent to the appropriate location no more than 24 hours after collection.

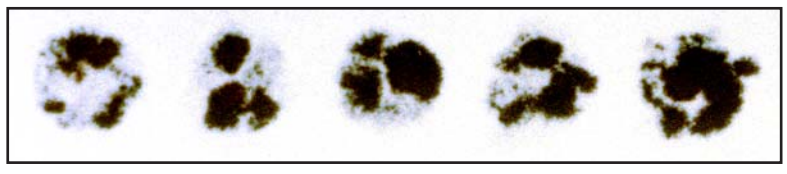
Unacceptable Specimens — Possible Causes

No blood



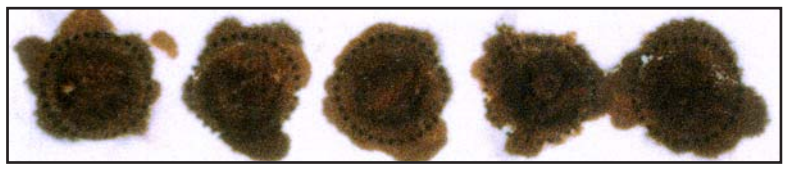
- Failure to obtain any blood

Quantity of blood insufficient



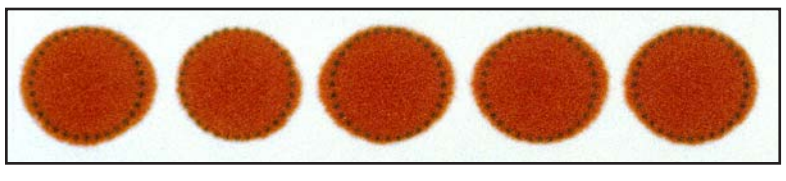
- Filter paper circle incompletely filled or not saturated
- Blood applied to filter paper with needle or capillary tube
- Contamination of surface of filter paper circle

Scratched or abraded blood spots



- Blood applied improperly with capillary tube or by other means

Wet or discolored blood spots

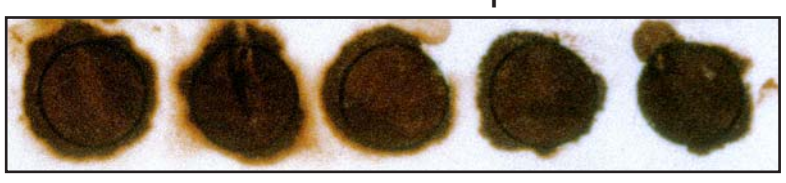


- Specimen not properly dried before mailing

Supersaturated blood spots

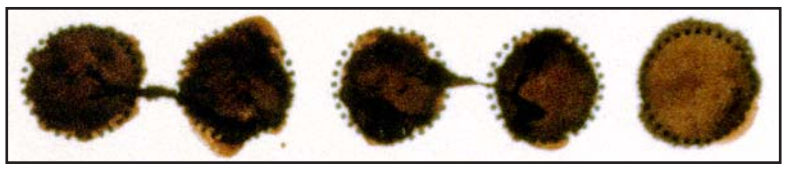
- Excess blood applied to the filter paper, usually with capillary tube or needle
- Blood applied to both sides of the filter paper

Diluted blood spots



- Puncture site squeezed or "milked" to expel blood
- Exposure of blood spots to direct heat
- Contamination of filter paper before or after blood collection by gloved or ungloved hands or by substances such as alcohol, feeding or antiseptic solutions, hand lotion or powder

Clotted or layered blood spots



- Touching the same filter paper circle to a blood drop several times
- Filling the circle from both sides of the filter paper

Serum rings evident in blood spots

- Alcohol not allowed to dry completely before skin puncture is made
- Allowing filter paper to come in contact with alcohol, water, hand lotion, etc.
- Squeezing the area around the puncture site excessively
- Drying the specimen improperly
- Applying blood to filter paper with a capillary tube

ID Biological SYSTEMS

USA: 17 P & N Drive • Greenville, SC 29611 USA • Tel. (864) 299-8787 • Fax (864) 299-8797 • id-biological.com • bdavin@id-biological.com • ggaillard@id-bioloigcal.com
UK: 15 Tanners Hill • Abbots Langley • Hertfordshire WD5 0LT UK • Tel. + 44 (0) 7525 844431 • Fax + 44 (0) 1923 682120 • id-biological.com • jdinan@id-biological.com

REASONS FOR SPECIMEN REJECTION (UNSATS) - (with codes for Arizona unsats)

Code	Reason	Comments
Problems with the family or baby		
UPR	Parent refusal	Document the refusal in the medical record and submit the card with no blood (check the "parent refused testing" box)
UIO	Infant too old (>1 year of age)	Reference ranges apply only to newborn infants and cannot accurately indicate risk of disease when collected from older children
Problems with the card		
UCE	Expired collection card	Valid results cannot be reported if the card has expired per CLIA. Check the date printed above the circles for blood.
UNI	No identifying information	Results cannot be reported if the infant cannot be identified.
UNO	No blood on the card	Tests cannot be performed without blood.
Problems with collection of the blood		
UCC	Clotted or caked blood	Uniform amounts of blood cannot be taken from samples with extra clotted blood in some areas of the spot.
UIS	Insufficient specimen	Not enough blood was submitted to complete the whole panel of tests
UMA	Multiple specimen applications	Blood may have been applied to both sides of the filter paper or with overlapping drops of blood instead of one big drop in each circle.
UNS	Uneven saturation	With some areas more saturated with blood than other areas of the spot instead of one big drop in each circle, uniform amounts of blood cannot be taken for testing.
UTS	Torn or scratched	With areas where filter paper is missing or bunched up, uniform amounts of blood cannot be taken for testing. This usually happens when capillary tubes are used to collect the blood. The rough edges of the tubes can abrade the filter paper if they are dragged along the surface.
UST	Serum or tissue fluid separation	This can happen if the first drop of blood is not wiped away, if a liquid contaminant is allowed to come in contact with the blood spots, if the area around the puncture site is squeezed excessively ("milking"), or if the specimen is dried vertically so that gravity can separate blood components before complete drying.
Problems in handling and shipping		
UTO	Specimen too old (>14 days from collection date)	Blood components degrade with age and exposure to heat and light and cannot be accurately measured.
USD	Specimen detached from form	Results cannot be reported if the infant may be incorrectly identified.
USC	Contaminated	Filter paper has come in contact with gloved or ungloved hands or substances such as alcohol, formula, antiseptic solutions, water, hand lotion or powder, etc. before or after specimen collection. Contaminated specimens yield unreliable results.
Problem in the lab		
URS	Results inconsistent	Repeat testing of the same sample gives results that are greatly different from one another. This could indicate that the sample was unevenly saturated or damaged in some way.

Timing of NBS Specimen Collection and Factors that Influence Results

Disorder	Appearance of symptoms &/or need for treatment	Risk of acute crisis?	Best Screening Window	Factors causing false positive result	Factors causing false negative result	Duration of Effect	Comments
CH	first year of life, treatment by 2 weeks of age prevents mental retardation, developmental delays	no	12 - 72 hr and 2 - 6 weeks	TSH surge in first 12-24 hours of life		until 12 - 24 hours of age	
				topical iodine on baby or breastfeeding mother		2 - 6 weeks after topical iodine discontinued	
				maternal hyperthyroidism treated with PTU		until drug clears – typically 7 - 14 days	transient hypothyroidism
				acute illness		until recovered	
				iodine deficiency		until supplemented	
					delayed rise of TSH in affected infants, particularly if preterm (immature hypothalamic-pituitary-thyroid axis)	up to 6 weeks of age	
					dopamine therapy (suppresses TSH)	until drug therapy is stopped	
					steroid treatment (suppresses TSH & T4)	unknown – depends on class of steroid and dose; estimate of 1 - 2 weeks	
CAH	first week of life	yes	12 - 48 hr and 2 - 4 weeks	preterm birth or LBW		until stable	
				sick or stressed infant		until recovered	
				mother with CAH and elevated 17-OHP		unknown – estimate of 3 - 7 days	
				early collection (<24 hr of age)		until 24 hours of age	
					maternal steroid treatment	unknown – depends on class of steroid and dose; estimate of 1 - 2 weeks	suppresses fetal adrenal function
					steroid (dexamethasone) treatment in infant	unknown – depends on dose; estimate of 1 - 2 weeks	
Hb diseases	first months of life, prophylactic penicillin by 6 weeks of age prevents infection and early complications	no	birth - 72 hours	variants found with uncertain clinical significance			not affected by maternal conditions or treatments
					red blood cell transfusion, ECMO	3 - 4 months after the last RBC transfusion	

Disorder	Appearance of symptoms &/or need for treatment	Risk of acute crisis?	Best Screening Window	Factors causing false positive result	Factors causing false negative result	Duration of Effect	Comments
BIO	1 week – 10 years of age (most show symptoms between 3 – 6 months of age)	no	birth - 72 hr	heat with humidity damage to specimen		until another specimen (protected from heat and promptly delivered to the lab) is tested	not affected by maternal conditions or treatments
				prematurity		until 40 weeks gestational age	
				liver disease, jaundice		until resolved	
					transfusion of plasma or other blood products	transient effect: 1 – 3 days after transfusion	
GAL	first week of life	yes	birth - 48 hours	heat damage to specimen, age of specimen (received by lab more than 4 – 5 days after collection)		until another specimen (protected from heat and promptly delivered to the lab) is tested	not affected by maternal conditions or treatments
					red blood cell transfusion	3 - 4 months after the last RBC transfusion	
PKU	6 - 8 months of age (irreversible brain damage happens if treatment is not started in first weeks of life)	no	24 - 48 hours	PN		4 - 24 hours after discontinuing PN	
				liver dysfunction or immaturity		a few weeks or until resolved	
				maternal PKU or hyperphe uncontrolled by diet or medication		12 - 24 hours unless infant has PKU	transient hyperphe
					early collection (<24 hours of age) or collection only a few hours after transfusion or discontinuation of ECMO	1 – 3 days after transfusion	
MSUD	first two weeks of life	yes	24 - 48 hours	PN		4 - 24 hours after discontinuing PN	
				liver dysfunction or immaturity		a few weeks or until resolved	
					early collection (<24 hours of age) or collection only a few hours after transfusion or discontinuation of ECMO	1 – 3 days after transfusion	
HCY		no	3 - 7 days	PN		4 - 24 hours after discontinuing PN	
				liver dysfunction or immaturity		a few weeks or until resolved	
					early collection, pyridoxine responsive cases are not identified by NBS		

Disorder	Appearance of symptoms &/or need for treatment	Risk of acute crisis?	Best Screening Window	Factors causing false positive result	Factors causing false negative result	Duration of Effect	Comments
CIT & ASA	first two weeks of life	yes	24 - 48 hours	PN		4 - 24 hours after discontinuing PN	
				liver dysfunction or immaturity		a few weeks or until resolved	
					early collection or collection only a few hours after transfusion or discontinuation of ECMO	1 – 3 days after transfusion	
TYR 1	3 – 4 months of age (liver is damaged by that time)	no	more than 1 week of age	liver dysfunction or immaturity		a few weeks or until resolved	
FAO disorders	first few days to months or years (more easily detected during acute illnesses or during times of increased energy need)	yes	birth - 48 hours	carnitine supplementation, MCT oil		for duration of supplementation and some weeks later	
					well-fed state (later collection)	could be until fasting or ill	
					transfusion or ECMO	1 – 3 days after transfusion	
				fatty liver of pregnancy or HELLP syndrome* can cause elevated even chain acylcarnitines		unknown	
CUD		yes	birth - 48 hours	maternal carnitine deficiency		unknown	
					carnitine supplementation	for duration of supplementation and some weeks later	
MCD, MMAs, PA		yes	24 - 48 hours	maternal Vitamin B_{12} deficiency		a number of days depending on nutrition provided	
Organic acid disorders	first two weeks of life	yes	24 - 48 hours	PN		4 - 24 hours after discontinuing PN	
IVA	first two weeks of life	yes	24 - 48 hours	pivalic acid antibiotic therapy		24 hours after discontinuing therapy	
3MCC		yes	24 - 48 hours	asymptomatic mother with 3MCC, unaffected infant		unknown	

Disorder	Appearance of symptoms &/or need for treatment	Risk of acute crisis?	Best Screening Window	Factors causing false positive result	Factors causing false negative result	Duration of Effect	Comments
CF	first months of life – early treatment prevents early, progressive damage	no	24 hr - 7 days	hypoxic organ damage, neonatal stress (low Apgar), respiratory distress, hypoglycemia, acute illness, preterm birth, trisomies, early collection (<12 hours of age)		until recovered, until 40 weeks gestational age (will elevate IRT but won't be reported unless a mutation is found)	not affected by maternal conditions or treatments
				elevated IRT and a single mutation detected		a sweat test is needed to rule out CF and confirm carrier status	not always false positive - CF has been diagnosed with only one mutation detected by the screening panel
					in-range IRT and no DNA testing done	without elevated IRT, no mutation testing will be done	
					later collection (IRT declines with age, even in affected infants)	without elevated IRT, no mutation testing will be done	
					the mutation panel used – mutations vary among ethnic groups	only mutations on the screening panel will be detected	

ECMO: extra corporeal membrane oxygenation
PTU: propylthiouracil, an antithyroid medication
PN: parenteral nutrition
HELLP: Hemolysis, Elevated Liver enzymes, Low Platelets. Mothers carrying fetuses with FAO disorders have a 16% chance of developing HELLP syndrome compared with the general population risk of 0.88%.

Arizona Early Hearing Detection and Intervention
Arizona Department of Health Services
Hearing Out-Patient Screening Form

Screener or Contact Name: _____
Screener or Contact Phone:_____
Date Submitted:_____
Facility Name: _____

FAX TO 602-364-1495 within one week of screening

- **Submit for all infants screened up to two years of age.**
- **Do not submit if previous testing shows normal hearing and child is being screened for otitis media.**
- **Submit diagnostic report form if diagnostic testing was completed**

Patient Last Name:	Date of Birth:	Gender: ☐ Male ☐ Female
Patient First Name:	Birth Order (if multiple births): ☐ A ☐ B ☐ C ☐ D	
Mother's Full Name:	Birth Facility:	
Mother's Date of Birth:	Date of Screen:	
Primary Care Physician:	Right: ☐ Pass ☐ Fail	Left ☐ Pass ☐ Fail
Comments:	☐ OAE ☐ ABR ☐ Behavioral	
Patient Last Name:	Date of Birth:	Gender: ☐ Male ☐ Female
Patient First Name:	Birth Order (if multiple births): ☐ A ☐ B ☐ C ☐ D	
Mother's Full Name:	Birth Facility:	
Mother's Date of Birth:	Date of Screen:	
Primary Care Physician:	Right: ☐ Pass ☐ Fail	Left ☐ Pass ☐ Fail
Comments:	☐ OAE ☐ ABR ☐ Behavioral	
Patient Last Name:	Date of Birth:	Gender: ☐ Male ☐ Female
Patient First Name:	Birth Order (if multiple births): ☐ A ☐ B ☐ C ☐ D	
Mother's Full Name:	Birth Facility:	
Mother's Date of Birth:	Date of Screen:	
Primary Care Physician:	Right: ☐ Pass ☐ Fail	Left ☐ Pass ☐ Fail
Comments:	☐ OAE ☐ ABR ☐ Behavioral	

Fax to: 602-364-1495 Questions call 602-364-1409
Always start a new log sheet after faxing to ADHS

Within one week of screening, submit results to the Arizona Department of Health Services.
Fax to **602-364-1495** or mail to: ***Newborn Screening Program***
150 North 18th Avenue Suite 320
Phoenix, Arizona 85007-3242

If you have questions or need help finding resources for further testing call 800-548-8381

Hearing Screening Record

First Name_____ **Last Name**_____ **Gender**_____

Birth Hospital_____ **Birth Order** _____ **Date of Birth (DOB)**_____

Mother's First Name_____ **Last Name (prior to first marriage)** _____

Mother's DOB_____ **Clinic Site**_____

☐ **Periodic Screen**

Child's Age **Days** ☐ 2 - 4
Months ☐ 1 ☐ 2 ☐ 4 ☐ 6 ☐ 9 ☐ 12 ☐ 15 ☐ 18 ☐ 24 ☐ 36

Is this the first time the OAE hearing screening has been performed on this child in your clinic/practice?
☐ No ☐ Yes--if Yes:

Was child screened for hearing loss at birth? ☐ Unknown ☐ Not Screened ☐ Passed ☐ Referred

Any neonatal risk factors or complications? ☐ Unknown ☐ No ☐Yes: _____

Any family history of early hearing loss? ☐ No ☐Yes: _____

- **Is the parent/caregiver concerned about the child's:** **Hearing?** ☐ No ☐Yes: _____
  Speech? ☐ No ☐Yes: _____
- **Has the child experienced:** **Head trauma?** ☐ No ☐Yes: _____
  Recurrent ear infections? ☐ No ☐Yes: _____
- **Does the child have Pressure Equalization (PE) tubes?** ☐ No ☐Yes: _____

☐ **Pre-Referral Rescreen** (Conduct rescreen; refer for Audiological Assessment unless OAE pass is obtained.)

☐ **Other** _____

Screener: _____ **Date**: ___/___/___

Recommendations :

Target Date ___/___/___

Dx Code _____

Referred to :_____

LEFT

OAE 1 → **Middle Ear Evaluation** (Tympanometry/Pneumatic Otoscopy) → **OAE 2**

Pass ☐ → **Pass** ☐ → **Pass** ☐

***Can't test** ☐ → **Can't test** ☐ → ☐ Schedule Pre-Referral Rescreen (w/in 2 weeks) or ☐ Refer for **Audiological Assessment**

Refer ☐ → **Refer** ☐ (☐ If wax removal, ☐ If OM or other) → **Refer** ☐ → ☐ Rescreen 4 weeks after medical clearance or ☐ Refer for **Audiological Assessment** as needed; also if OM remains unresolved after 3 months

RIGHT

OAE 1 → **Middle Ear Evaluation** (Tympanometry/Pneumatic Otoscopy) → **OAE 2**

Pass ☐ → **Pass** ☐ → **Pass** ☐

***Can't test** ☐ → **Can't test** ☐ → ☐ Schedule Pre-Referral Rescreen (w/in 2 weeks) or ☐ Refer for **Audiological Assessment**

Refer ☐ → **Refer** ☐ (☐ If wax removal, ☐ If OM or other) → **Refer** ☐ → ☐ Rescreen 4 weeks after medical clearance or ☐ Refer for **Audiological Assessment** as needed; also if OM remains unresolved after 3 months

***If can't test, indicate why**: ☐child uncooperative ☐internal noise ☐ external noise ☐ear wax ☐drainage ☐equipment problem

REFERRAL AND TRACKING FORM
ARIZONA EARLY HEARING DETECTION AND INTERVENTION &
ARIZONA STATE SCHOOLS FOR THE DEAF AND THE BLIND
PHOENIX

PLEASE FAX WITH ASSESSMENT RESULTS WITHIN 48 HOURS TO:

ASDB fax: 602-544-1704 phone: 602-771-5200
AzEHDI fax: 602-364-1495 phone: 602-364-1409

NAME OF CHILD: **CHILD BIRTH DATE:**

DATE REFERRED: **BIRTH HOSPITAL:** ☐ MALE ☐ FEMALE

MOTHER'S FULL NAME: **MOTHER BIRTHDATE:**

ADDRESS WITH CITY & ZIP:

PRIMARY PERSON TO CONTACT: **HOME PHONE:**

CELL PHONE: **WORK PHONE:** **HOME LANGUAGE :**

TO WHAT AGENCY OR SPECIALIST HAVE YOU REFERRED THIS CHILD?

DDD REFERRAL MADE: YES ☐ NO ☐ ALREADY ENROLLED ☐
CRS REFERRAL MADE: YES ☐ NO ☐ ALREADY ENROLLED ☐
ENT REFERRAL MADE: YES ☐ NO ☐ ENT PROVIDER NAME: _____

OTHER AGENCY: _____

OTHER SPECIALTY: _____

AUDIOLOGIST NAME: _____ **DATE OF EVALUATION:** _____

☐ **Fax to both if…**
- o Under 3 years of age
- o Bilateral hearing loss
- o Sensorineural or Permanent Conductive
- o Auditory Neuropathy

☐ **Fax to AzEHDI only if...**
- o Over 3 years of age or
- o Unilateral
- o Ruled out Hearing Loss in a Child under 3 (normal hearing results)

TESTING THAT DETERMINED HEARING LOSS (MARK ALL THAT APPLY)

ABR:
☐ CLICKS
☐ TONE BURSTS/PIPS
☐ BONE CONDUCTION
☐ ASSR

BEHAVIORAL:
☐ VRA
☐ BOA
☐ PLAY
☐ CONVENTIONAL

HEARING LOSS:
☐ CONFIRMED ☐ Preliminary NEXT APPT: _____

DEGREE:	RIGHT	LEFT	**TYPE:**	RIGHT	LEFT
	☐	☐ NORMAL		☐	☐ CONDUCTIVE
	☐	☐ MILD		☐	☐ PERMANENT CONDUCTIVE
	☐	☐ MODERATE		☐	☐ MIXED
	☐	☐ SEVERE		☐	☐ SENSORINEURAL
	☐	☐ PROFOUND		☐	☐ NEUROPATHY

AMPLIFICATION: RIGHT ☐ LEFT ☐ **ANTICIPATED FITTING DATE:** _____

OTHER DISABILITIES/CONCERNS: _____

_____

_____

_____

REFERRAL AND TRACKING FORM
ARIZONA EARLY HEARING DETECTION AND INTERVENTION &
ARIZONA STATE SCHOOLS FOR THE DEAF AND THE BLIND
FLAGSTAFF

PLEASE FAX WITH ASSESSMENT RESULTS WITHIN 48 HOURS TO:

ASDB fax: 928-773-9229 phone: 928-774-0655
AzEHDI fax: 602-364-1495 phone: 602-364-1409

NAME OF CHILD: **CHILD BIRTH DATE:**

DATE REFERRED: **BIRTH HOSPITAL:** ☐ MALE ☐ FEMALE

MOTHER'S FULL NAME: **MOTHER BIRTHDATE:**

ADDRESS WITH CITY & ZIP:

PRIMARY PERSON TO CONTACT: **HOME PHONE:**

CELL PHONE: **WORK PHONE:** **HOME LANGUAGE :**

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CRS REFERRAL MADE: YES ☐ NO ☐ ALREADY ENROLLED ☐
ENT REFERRAL MADE: YES ☐ NO ☐ ENT PROVIDER NAME: _____

OTHER AGENCY: _____

OTHER SPECIALTY: _____

AUDIOLOGIST NAME: _____ **DATE OF EVALUATION:** _____

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BEHAVIORAL:
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☐ BOA
☐ PLAY
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HEARING LOSS:
☐ CONFIRMED ☐ Preliminary NEXT APPT: _____

DEGREE:	RIGHT	LEFT	**TYPE:**	RIGHT	LEFT
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	☐	☐ SEVERE		☐	☐ SENSORINEURAL
	☐	☐ PROFOUND		☐	☐ NEUROPATHY

AMPLIFICATION: RIGHT ☐ LEFT ☐ **ANTICIPATED FITTING DATE:** _____

OTHER DISABILITIES/CONCERNS: _____

_____

_____

_____

REFERRAL AND TRACKING FORM
ARIZONA EARLY HEARING DETECTION AND INTERVENTION &
ARIZONA STATE SCHOOLS FOR THE DEAF AND THE BLIND
TUCSON

PLEASE FAX WITH ASSESSMENT RESULTS WITHIN 48 HOURS TO:

ASDB fax: 520-770-3010 phone: 520-770-3703
AzEHDI fax: 602-364-1495 phone: 602-364-1409

NAME OF CHILD: **CHILD BIRTH DATE:**

DATE REFERRED: **BIRTH HOSPITAL:** ☐ MALE ☐ FEMALE

MOTHER'S FULL NAME: **MOTHER BIRTHDATE:**

ADDRESS WITH CITY & ZIP:

PRIMARY PERSON TO CONTACT: **HOME PHONE:**

CELL PHONE: **WORK PHONE:** **HOME LANGUAGE :**

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CRS REFERRAL MADE: YES ☐ NO ☐ ALREADY ENROLLED ☐
ENT REFERRAL MADE: YES ☐ NO ☐ ENT PROVIDER NAME: _____

OTHER AGENCY: _____

OTHER SPECIALTY: _____

AUDIOLOGIST NAME: _____ **DATE OF EVALUATION:** _____

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BEHAVIORAL:
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HEARING LOSS:
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	☐	☐ MODERATE		☐	☐ MIXED
	☐	☐ SEVERE		☐	☐ SENSORINEURAL
	☐	☐ PROFOUND		☐	☐ NEUROPATHY

AMPLIFICATION: RIGHT ☐ LEFT ☐ **ANTICIPATED FITTING DATE:** _____

OTHER DISABILITIES/CONCERNS: _____

_____

_____

_____

REFERRAL AND TRACKING FORM
ARIZONA EARLY HEARING DETECTION AND INTERVENTION &
ARIZONA STATE SCHOOLS FOR THE DEAF AND THE BLIND
YUMA

PLEASE FAX WITH ASSESSMENT RESULTS WITHIN 48 HOURS TO:

ASDB fax: 928-783-4941 phone: 928-783-4003
AzEHDI fax: 602-364-1495 phone: 602-364-1409

NAME OF CHILD: **CHILD BIRTH DATE:**

DATE REFERRED: **BIRTH HOSPITAL:** ☐ MALE ☐ FEMALE

MOTHER'S FULL NAME: **MOTHER BIRTHDATE:**

ADDRESS WITH CITY & ZIP:

PRIMARY PERSON TO CONTACT: **HOME PHONE:**

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ENT REFERRAL MADE: YES ☐ NO ☐ ENT PROVIDER NAME: _____

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	☐	☐ MODERATE		☐	☐ MIXED
	☐	☐ SEVERE		☐	☐ SENSORINEURAL
	☐	☐ PROFOUND		☐	☐ NEUROPATHY

AMPLIFICATION: RIGHT ☐ LEFT ☐ **ANTICIPATED FITTING DATE:** _____

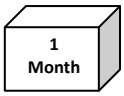
OTHER DISABILITIES/CONCERNS: _____

_____

_____

_____

Health Care Providers - Screen All Babies by One Month
Resources for Outpatient Hearing Screening



If a baby did not pass the newborn hearing screening at birth, it is important that the baby is screened again as soon as possible, *no later than 1 month of age*. If the baby was in the NICU for more than 5 days or has a risk indicator for hearing loss, the baby should not receive an outpatient screen but should go directly to a pediatric audiologist for testing. The family may need a referral from the primary care provider in order to schedule an appointment.

NOTE: Not all hospitals can screen infants over 1 month of age.

Phoenix (Metro Area) - Central, North and Scottsdale

OAE/A-ABR		HOSPITALS		OAE/AABR		OTHER PROVIDERS	
	√	Banner Good Samaritan	602-839-2178	√		Affiliated Audiology	602-254-6041
√	√	Maricopa Medical Audiology Clinic	602-344-5185	√	√	Arizona Balance & Hearing	602-265-9000
	√	John C. Lincoln North Mountain	602-277-4161 x44 *		√	Neonatology Associates (NAL)	602-277-4161 x44 *
	√	Paradise Valley Hospital		√		Mountain Park Clinic – Baseline	602-243-7277
	√	Phoenix Baptist Hospital		√		North Valley ENT	602-688-6500
	√	St. Joseph's Hospital		√	√	Premiere ENT Surgeons	602-678-5001
√	√	Phoenix Children's Audiology	602-546-0905	√		Valley ENT	480-614-0499
	√	Scottsdale Healthcare (North)	480-323-3638				
	√	Scottsdale Healthcare Osborn	480-882-4171				

Phoenix Area – West Valley

OAE/AABR		HOSPITALS		OAE/AABR		OTHER PROVIDERS	
	√	Arrowhead Hospital	602-277-4161 x44 *	√		Adelante Healthcare	623-544-5108
	√	Banner Thunderbird Medical		√		Valley ENT	623-566-4718
	√	Maryvale Hospital		√		Mayo Clinic – Arrowhead	623-561-5282
	√	West Valley Hospital		√	√	West Valley ENT	602-843-4844
	√	Banner Estrella Medical Ctr.	623-327-5873	√	√	Metro Hearing Services	623-866-0147
	√	Banner Del E. Webb	602-344-1015		√	Neonatology Associates (NAL)	602-277-4161 x44 *

Phoenix Area – East Valley

OAE/AABR		HOSPITALS		OAE/AABR		OTHER PROVIDERS	
	√	Banner Baywood Medical Ctr.	602-277-4161 x44 *	√		Advance Hearing Group	480-218-1328
	√	Banner Gateway Medical Ctr.		√	√	ASU Speech and Hearing	480-965-2373
	√	Chandler Regional Hospital		√	√	Arizona Ear and Hearing	480-292-7100
	√	Mercy Gilbert Medical Ctr		√		Arizona Hearing and Balance	480-558-5306
	√	Mountain Vista Medical		√		Cigna Healthcare Hearing Ctr.	480-464-6870
√	√	Cardon Children's/Banner Desert	480-412-4099	√	√	Desert Sounds	480-497-3285
√	√	Phoenix Children's Audiology	602-546-0905	√		ENT Specialists of Arizona	480-894-5550 X502
√	√	Tempe St. Luke's	480-784-5557	√	√	Good Sound Audiology	480-497-0780
				√		Harper Hearing Solutions	480-838-1212
				√		Hearing Solutions of Arizona	480-833-4330
				√	√	Premier ENT Surgeons	602-678-5001

Tucson Area & Southern Arizona

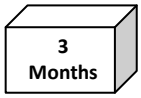
OAE/AABR		HOSPITALS		OAE/AABR		OTHER PROVIDERS	
√	√	Carondelet St. Joseph's	520-873-3761	√		Adobe Hearing Center	520-322-8211
√	√	Carondelet St. Mary's	520-872-4846	√		Carlson Ear, Nose & Throat	520-795-8777
	√	Northwest Medical Center	520-877-4021	√		Grunewald Blitz (U of A)	520-621-7070
√	√	Tucson Medical Center	520-324-2075	√	√	Marana Health Center	520-682-4560
√		The Children's Clinic	520-324-3600	√		Oro Valley & Tanque Verde Aud	520-751-3901
	√	Casa Grande Regional	520-381-6475	√		Tucson ENT	520-777-0495
√	√	Carondelet Holy Cross (Nogales)	520-287-8048	√		Birth and Women's Health Center	520-795-9912
	√	Cobre Valley Community (Safford)	928-402-1260	√		Chricahua Health (Douglas)	520-364-3285
	√	Mt. Graham (Safford)	928-348-4185	√	√	Audiology Inc. (Safford)	928-428-1613
√	√	Sierra Vista Hospital	520-417-3184	√		Southern AZ Hearing & Balance	520-459-0688
√	√	Yuma Regional Medical Ctr	928-336-3893	√	√	Cochise ENT Assoc (Sierra Vista)	520-458-4919
				√		Sells Special Services Program	520-383-8665

Flagstaff Area & Northern Arizona

OAE/AABR		HOSPITALS		OAE/AABR		OTHER PROVIDERS	
√	√	Flagstaff Medical Ctr. Audiology	928-214-3728	√		North Country CHC (Flagstaff)	928-213-6154
√	√	Verde Valley (Cottonwood)	928-639-6521	√		Northern Arizona University	928-523-8110
	√	Little Colorado (Winslow)	928-289-4691	√	√	Audiology Inc (Show Low)	928-537-3456
√	√	Tuba City IHS	928-283-2629	√		ASDB Eastern Highland Reg. Coop	928-524-6770
	√	Payson Regional	928-472-1226	√		Fort Defiance	928-729-8915
√	√	Banner Page Hospital	928-645-0777	√	√	Chinle IHS Audiology	928-674-7223
	√	Summit Healthcare	928-537-6362		√	White River IHS (Whiteriver)	928-338-4911 x3728
	√	Havasu Regional	928-453-0660	√	√	White Mtn Hearing Svc (Lakeside)	928-537-7373
	√	Kingman Regional	928-757-0690	√	√	AZ Coast ENT (Lake Havasu)	928-854-5368
	√	WARMC (Bullhead City)	928-763-0676	√		Valley View (Fort Mohave)	928-788-7095
√	√	Yavapai Regional	928-771-5200	√		Tri State Audiology (Bullhead City)	928-758-3337
				√		Prescott ENT	928-778-9190

AABR – Automated Auditory Brainstem Response
OAE – Otoacoustic Emissions

Health Care Providers – Diagnostic Evaluation by Three Months
Resources for Infant Diagnostic Hearing Testing



If a baby has failed the outpatient screen, or has been in the NICU for more than five days, the baby should be referred by their primary care provider to a pediatric audiologist for a diagnostic evaluation. Under three to six months of age the testing is usually completed without sedation/anesthesia. After an infant is older than six months of age, sedation/anesthesia may be required to complete the test.

SITES MARKED WITH * ARE ABLE TO PERFORM TESTING WITH SEDATION/ANESTHESIA IF NEEDED

Phoenix Area (Metro Area, Central, North & Scottsdale)	
*Phoenix Children's Hospital (Phoenix & East Valley locations)	602-546-0905
*St. Joseph's Hospital Children's Rehabilitative Services	602-406-6420
Arizona Balance & Hearing Associates	602-265-9000
Maricopa Medical Audiology Clinic	602-344-5185
Phoenix Indian Medical Center	602-263-1514
Premier ENT Surgeons	602-678-5001
Phoenix Area – West Valley	
Valley ENT (Glendale)	623-566-4718
Phoenix Area – East Valley	
*Arizona Hearing and Balance Center (Chandler)	480-558-5306
*Cardon's Children's/Banner Desert Audiology (Mesa)	480-412-4099
Arizona State University Speech and Hearing Clinic (Tempe)	480-965-2373
Arizona Ear & Hearing (Queen Creek)	480-292-7100
Desert Sounds Audiology (Mesa)	480-497-3285
Good Sound Audiology (Mesa)	480-497-0780
Tucson Area & Southern Arizona	
*Carondelet St. Joseph's Hospital Hearing Services	520-873-3761
*Tucson Medical Center Audiology Department	520-324-2075
Carondelet St. Mary's Audiology Department	520-872-4846
Tucson ENT (East & Northwest Tucson)	520-777-0495
University of Arizona Grunewald-Blitz Center	520-621-7070
Audiology Inc. (Safford)	928-428-1613
Flagstaff Area & Northern Arizona	
*Chinle Indian Health Services Audiology Department	928-674-7223
*Flagstaff Medical Center Audiology Department	928-214-3728
White Mountain Hearing Services (Lakeside)	928-537-7373
Audiology Inc. (Show Low)	928-537-3456

NOTE: Sites have self identified as providing services and are not endorsed or recommended.

If you have questions or concerns about the hearing screening or would like more information, you can call the Arizona Department of Health Services Newborn Screening Program at 602-364-1409 or visit online at www.aznewborn.com.

Si usted tiene algunas preguntas o preocupaciones sobre la prueba auditiva o si quisiera más información, puede llamar al Departamento de Servicios de Salud de Arizona al Programa de Análisis para el Niño Recién Nacido al número 602-364-1409. También puede visitarnos en el Internet a www.aznewborn.com.

1 Month

Screen All Babies by One Month
Resources for Parents for Outpatient Hearing Screening

Your baby did not pass the newborn hearing screening. It is important that your baby is screened again as soon as possible (before 1 month of age). Contact one of the places listed below to schedule an appointment. If you need a referral for insurance, talk to your baby's doctor as soon as possible.

Su bebé no pasó la prueba de audición del recién nacido. Es importante que haga otra prueba auditiva para su bebé lo más pronto posible. Debe hacer esta prueba antes que su bebé cumpla 1 mes de edad. Llame a uno de los lugares en la lista abajo para hacer una cita. Si usted necesita una referencia de su seguro médico, hable con el doctor de su bebé lo más pronto posible.

NOTE: Not all hospitals can screen infants over 1 month of age.

Phoenix (Metro Area) - Central, North and Scottsdale			
HOSPITALS		*OTHER PROVIDERS*	
Banner Good Samaritan	602-839-2178	Affiliated Audiology	602-254-6041
Maricopa Medical Audiology Clinic	602-344-5185	Arizona Balance & Hearing	602-265-9000
John C. Lincoln North Mountain Paradise Valley Hospital Phoenix Baptist Hospital St. Joseph's Hospital	602-277-4161 x44 *	Mayo Clinic – Scottsdale	480-301-8484
		Neonatology Associates (NAL)	602-277-4161 x44 *
		Mountain Park Clinic – Baseline	602-243-7277
Phoenix Children's Audiology	602-546-0905	North Valley ENT	602-688-6500
Scottsdale Healthcare (North)	480-323-3638	Premiere ENT Surgeons	602-678-5001
Scottsdale Healthcare Osborn	480-882-4171	Valley ENT	480-614-0499
Phoenix Area – West Valley			
HOSPITALS		*OTHER PROVIDERS*	
Arrowhead Hospital Banner Thunderbird Medical Maryvale Hospital West Valley Hospital	602-277-4161 x44 *	Adelante Healthcare	623-544-5108
		Valley ENT	623-566-4718
		Mayo Clinic – Arrowhead	623-561-5282
		West Valley ENT	602-843-4844
Banner Estrella Medical Ctr.	623-327-5873	Metro Hearing Services	623-866-0147
Banner Del E. Webb	602-344-1015	Neonatology Associates (NAL)	602-277-4161 x44 *
Phoenix Area – East Valley			
HOSPITALS		*OTHER PROVIDERS*	
Banner Baywood Medical Ctr. Banner Gateway Medical Ctr. Chandler Regional Hospital Mercy Gilbert Medical Ctr Mountain Vista Medical	602-277-4161 x44 *	Advance Hearing Group	480-218-1328
		ASU Speech and Hearing	480-965-2373
		Arizona Ear and Hearing	480-292-7100
		Arizona Hearing and Balance	480-558-5306
		Cigna Healthcare Hearing Ctr.	480-464-6870
Cardon's Children's/Banner Desert	480-412-4099	Desert Sounds	480-497-3285
Phoenix Children's Audiology	602-546-0905	ENT Specialists of Arizona	480-894-5550 X502
Tempe St. Luke's	480-784-5557	Good Sound Audiology	480-497-0780
		Harper Hearing Solutions	480-838-1212
		Hearing Solutions of Arizona	480-833-4330
		Premier ENT Surgeons	602-678-5001
Tucson Area & Southern Arizona			
HOSPITALS		*OTHER PROVIDERS*	
Carondelet St. Joseph's	520-873-3761	Adobe Hearing Center	520-322-8211
Carondelet St. Mary's	520-872-4846	Carlson Ear, Nose & Throat	520-795-8777
Northwest Medical Center	520-877-4021	Grunewald Blitz (U of A)	520-621-7070
Tucson Medical Center	520-324-2075	Marana Health Center	520-682-4560
The Children's Clinic	520-324-3600	Oro Valley & Tanque Verde Aud	520-751-3901
Casa Grande Regional	520-381-6475	Tucson ENT	520-777-0495
Carondelet Holy Cross (Nogales)	520-287-8048	Birth and Women's Health Center	520-795-9912
Cobre Valley Community (Safford)	928-402-1260	Chricahua Health (Douglas)	520-364-3285
Mt. Graham (Safford)	928-348-4185	Audiology Inc. (Safford)	928-428-1613
Sierra Vista Hospital	520-417-3184	Southern AZ Hearing & Balance	520-459-0688
Yuma Regional Medical Ctr.	928-336-3893	Cochise ENT Assoc. (Sierra Vista)	520-458-4919
		Sells Special Services Program	520-383-8665
Flagstaff Area & Northern Arizona			
HOSPITALS		*OTHER PROVIDERS*	
Flagstaff Medical Ctr. Audiology	928-214-3728	North Country CHC (Flagstaff)	928-213-6154
Verde Valley (Cottonwood)	928-639-6521	Northern Arizona University	928-523-8110
Little Colorado (Winslow)	928-289-4691	Audiology Inc (Show Low)	928-537-3456
Tuba City IHS	928-283-2629	ASDB Eastern Highland Regional Coop	928-524-6770
Payson Regional	928-472-1226	Fort Defiance	928-729-8915
Page Hospital	928-645-0777	Chinle Indian Health Services Audiology	928-674-7223
Summit Healthcare	928-537-6362	White River IHS (Whiteriver)	928-338-4911 x3728
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Kingman Regional	928-757-0690	AZ Coast ENT (Lake Havasu)	928-854-5368
WARMC (Bullhead City)	928-763-0676	Valley View (Fort Mohave)	928-788-7095
Yavapai Regional	928-771-5200	Tri State Audiology (Bullhead City)	928-758-3337
		Prescott ENT	928-778-9190

**previsión del español disponible en 602-277-4161x 43*

3 Months

Diagnostic Evaluation by Three Months
Resources Parents for Infant Diagnostic Hearing Testing

If your baby does not pass a second screen, further testing will be needed to see if your baby has a hearing loss.

Si su bebé no pasa la segunda prueba auditiva, será necesario hacer pruebas adicionales para determinar si su bebé tiene una pérdida auditiva.

SITES MARKED WITH * ARE ABLE TO PERFORM TESTING WITH SEDATION IF NEEDED

Phoenix Area (Metro Area, Central, North & Scottsdale)	
*Phoenix Children's Hospital (Phoenix & East Valley locations)	602-546-0905
*St. Joseph's Hospital Children's Rehabilitative Services	602-406-6420
Arizona Balance & Hearing Associates	602-265-9000
Maricopa Medical Audiology Clinic	602-344-5185
Phoenix Indian Medical Center	602-263-1514
Premiere ENT Surgeons	602-678-5001
Phoenix Area – West Valley	
Valley ENT (Glendale)	623-566-4718
Phoenix Area – East Valley	
*Arizona Hearing and Balance Center (Chandler)	480-558-5306
*Cardon's Children's/Banner Desert Audiology (Mesa)	480-412-4099
Arizona State University Speech and Hearing Clinic (Tempe)	480-965-2373
Arizona Ear & Hearing (Queen Creek)	480-292-7100
Desert Sounds Audiology (Mesa)	480-497-3285
Good Sound Audiology (Mesa)	480-497-0780
Tucson Area & Southern Arizona	
*Carondelet St. Joseph's Hospital Hearing Services	520-873-3761
*Tucson Medical Center Audiology Department	520-324-2075
Carondelet St. Mary's Audiology Department	520-872-4846
Tucson ENT (East & Northwest Tucson)	520-777-0495
University of Arizona Grunewald-Blitz Center	520-621-7070
Audiology Inc. (Safford)	928-428-1613
Flagstaff Area & Northern Arizona	
*Chinle Indian Health Services Audiology Department	928-674-7223
*Flagstaff Medical Center Audiology Department	928-214-3728
White Mountain Hearing Services (Lakeside)	928-537-7373
Audiology Inc. (Show Low)	928-537-3456

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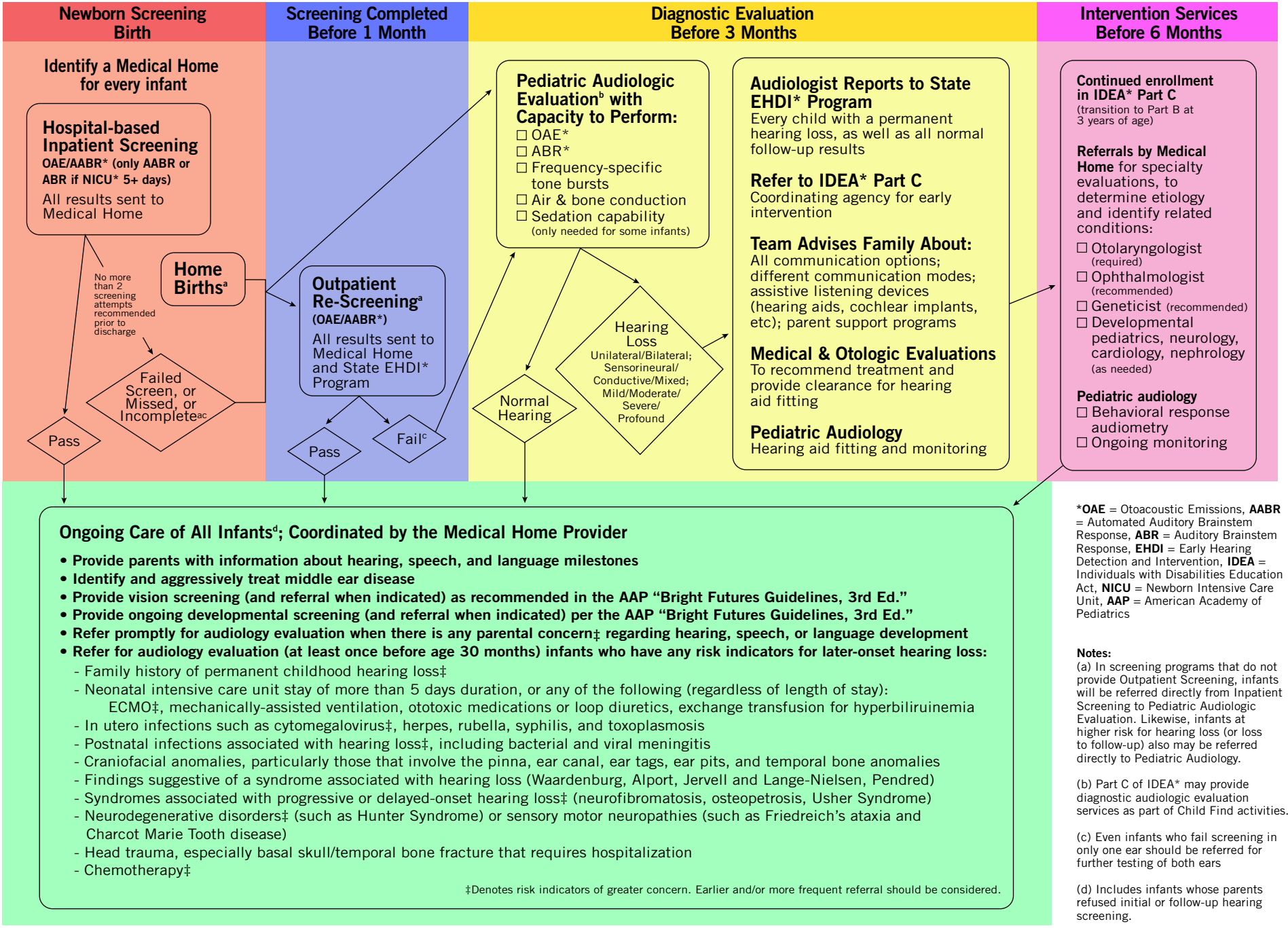
Si usted tiene algunas preguntas o preocupaciones sobre la prueba auditiva o si quisiera más información, puede llamar al Departamento de Servicios de Salud de Arizona al Programa de Análisis para el Niño Recién Nacido al número 602-364-1409. También puede visitarnos en el Internet a www.aznewborn.com.

Early Hearing Detection and Intervention (EHDI) Guidelines for Pediatric Medical Home Providers



***OAE** = Otoacoustic Emissions, **AABR** = Automated Auditory Brainstem Response, **ABR** = Auditory Brainstem Response, **EHDI** = Early Hearing Detection and Intervention, **IDEA** = Individuals with Disabilities Education Act, **NICU** = Newborn Intensive Care Unit, **AAP** = American Academy of Pediatrics

Notes:

(a) In screening programs that do not provide Outpatient Screening, infants will be referred directly from Inpatient 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 Audiology.

(b) Part C of IDEA* may provide diagnostic audiologic evaluation services as part of Child Find activities.

(c) Even infants who fail screening in only one ear should be referred for further testing of both ears

(d) Includes infants whose parents refused initial or follow-up hearing screening.

February 2010 - American Academy of Pediatrics Task Force for Improving Newborn Hearing Screening, Diagnosis and Intervention (www.medicalhomeinfo.org)

Appropriate Referrals

1. Audiologist knowledgeable in pediatric screening and amplification

Name:
Telephone number:
Fax:
Date of referral:

2. Otolaryngologist knowledgeable in pediatric hearing loss

Name:
Telephone number:
Fax:
Date of referral:

3. Local early intervention service coordinator

Name:
Telephone number:
Fax:
Date of referral:

4. Family support resources, financial resources

Name:
Telephone number:
Fax:
Date of referral:

5. Speech/language therapist and/or aural rehabilitation therapist knowledgeable in pediatric hearing loss

Name:
Telephone number:
Fax:
Date of referral:

6. Sign language classes if parents choose manual approach

Name:
Telephone number:
Fax:
Date of referral:

7. Ophthalmologist knowledgeable in co-morbid conditions in children with hearing loss

Name:
Telephone number:
Fax:
Date of referral:

8. Clinical geneticist knowledgeable in hearing loss

Name:
Telephone number:
Fax:
Date of referral:

9. Equipment vendor(s)

Name:
Telephone number:
Fax:
Date of referral:

10. State EHDI Coordinator
http://www.infanthearing.org/status/cnhs.html

Name:
Telephone number:
Fax:
Date of referral:

11. AAP Chapter Champion
www.medicalhomeinfo.org/screening/hearing.html

Name:
Telephone number:
Fax:
Date of referral:

12. Family physician(s)

Name:
Telephone number:
Fax:
Date of referral:

National Resources

Alexander Graham Bell Association for the Deaf and Hard of Hearing (AG Bell)
202/337-5220
www.agbell.org

American Academy of Audiology (AAA)
800/AAA-2336
www.audiology.org

American Academy of Pediatrics
847/434-4000
www.aap.org

American Society for Deaf Children
717/703-0073
www.deafchildren.org

American Speech-Language- Hearing Association (ASHA)
800/498-2071
www.asha.org

Boys Town Center for Childhood Deafness
www.babyhearing.org

Centers for Disease Control and Prevention
www.cdc.gov/ncbddd/ehdi

Families for Hands and Voices
217/357-3647
www.handsandvoices.org

Laurent Clerc National Deaf Education Center and Clearing-house at Gallaudet University
clerccenter.gallaudet.edu/InfoToGo

National Association of the Deaf (NAD)
301/587-1788
www.nad.org

National Center on Hearing Assessment and Management (NCHAM)
435/797-3584
www.infanthearing.org

National Institute on Deafness and Other Communication Disorders (NIDCD)
800/241-1044
www.nidcd.nih.gov

Oberkotter Foundation
www.oraldeafed.org

The recommendations in this document do not indicate an exclusive course of treatment or serve as a standard of medical care. Variations, taking into account individual circumstances, may be appropriate.

This project is funded by an educational grant from the Maternal and Child Health Bureau, Health Resources and Services Administration, US Department of Health and Human Services.

American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN™

NCHAM National Center for Hearing Assessment and Management Utah State University™

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OFFICIAL JOURNAL OF THE AMERICAN ACADEMY OF PEDIATRICS

Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs

Joint Committee on Infant Hearing

Pediatrics 2007;120;898-921
DOI: 10.1542/peds.2007-2333

American Academy of Pediatrics
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POLICY STATEMENT

Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs

Joint Committee on Infant Hearing

THE POSITION STATEMENT

The Joint Committee on Infant Hearing (JCIH) endorses early detection of and intervention for infants with hearing loss. 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.[1] To maximize the outcome for infants who are deaf or hard of hearing, the hearing of all infants should be screened at no later than 1 month of age. Those who do not pass screening should have a comprehensive audiological evaluation at no later than 3 months of age. Infants with confirmed hearing loss should receive appropriate intervention at no later than 6 months of age from health care and education professionals with expertise in hearing loss and deafness in infants and young children. Regardless of previous hearing-screening outcomes, all infants with or without risk factors should receive ongoing surveillance of communicative development beginning at 2 months of age during well-child visits in the medical home.[2] EHDI systems should guarantee seamless transitions for infants and their families through this process.

2007 JCIH POSITION STATEMENT UPDATES

The following are highlights of updates made since the 2000 JCIH statement[3]:

1. Definition of targeted hearing loss
   - The definition has been expanded from congenital permanent bilateral, unilateral sensory, or permanent conductive hearing loss to include neural hearing loss (eg, "auditory neuropathy/dyssynchrony") in infants admitted to the NICU.
2. Hearing-screening and -rescreening protocols
   - Separate protocols are recommended for NICU and well-infant nurseries. NICU infants admitted for more than 5 days are to have auditory brainstem response (ABR) included as part of their screening so that neural hearing loss will not be missed.
   - For infants who do not pass automated ABR testing in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, comprehensive evaluation including ABR.
   - For rescreening, a complete screening on both ears is recommended, even if only 1 ear failed the initial screening.
   - For readmissions in the first month of life for all infants (NICU or well infant), when there are conditions associated with potential hearing loss (eg, hyper-

www.pediatrics.org/cgi/doi/10.1542/peds.2007-2333

doi:10.1542/peds.2007-2333

All policy statements from the American Academy of Pediatrics automatically expire 5 years after publication unless reaffirmed, revised, or retired at or before that time.

Key Word
hearing screening

Abbreviations
JCIH—Joint Committee on Infant Hearing
EHDI—early hearing detection and intervention
ABR—auditory brainstem response
CMV—cytomegalovirus
ECMO—extracorporeal membrane oxygenation
AAP—American Academy of Pediatrics
MCHB—Maternal and Child Health Bureau
HRSA—Health Resources and Services Administration
NIDCD—National Institute on Deafness and Other Communication Disorders
CDC—Centers for Disease Control and Prevention
UNHS—universal newborn hearing screening
OAE—otoacoustic emission
IFSP—individualized family service plan
OME—otitis media with effusion
FM—frequency modulation
DSHPSHWA—Directors of Speech and Hearing Programs in State Health and Welfare Agencies
GPRA—Government Performance and Results Act
OMB—Office of Management and Budgets

PEDIATRICS (ISSN Numbers: Print, 0031-4005; Online, 1098-4275).

bilirubinemia that requires exchange transfusion or culture-positive sepsis), a repeat hearing screening is recommended before discharge.

3. Diagnostic audiology evaluation
   - Audiologists with skills and expertise in evaluating newborn and young infants with hearing loss should provide audiology diagnostic and auditory habilitation services (selection and fitting of amplification device).
   - At least 1 ABR test is recommended as part of a complete audiology diagnostic evaluation for children younger than 3 years for confirmation of permanent hearing loss.
   - The timing and number of hearing reevaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss. Infants who pass the neonatal screening but have a risk factor should have at least 1 diagnostic audiology assessment by 24 to 30 months of age. Early and more frequent assessment may be indicated for children with cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture-positive postnatal infections associated with sensorineural hearing loss; for children who have received extracorporeal membrane oxygenation (ECMO) or chemotherapy; and when there is caregiver concern or a family history of hearing loss.
   - For families who elect amplification, infants in whom permanent hearing loss is diagnosed should be fitted with an amplification device within 1 month of diagnosis.
4. Medical evaluation
   - For infants with confirmed hearing loss, a genetics consultation should be offered to their families.
   - Every infant with confirmed hearing loss should be evaluated by an otolaryngologist who has knowledge of pediatric hearing loss and have at least 1 examination to assess visual acuity by an ophthalmologist who is experienced in evaluating infants.
   - The risk factors for congenital and acquired hearing loss have been combined in a single list rather than grouped by time of onset.
5. Early intervention
   - All families of infants with any degree of bilateral or unilateral permanent hearing loss should be considered eligible for early intervention services.
   - There should be recognized central referral points of entry that ensure specialty services for infants with confirmed hearing loss.
   - Early intervention services for infants with confirmed hearing loss should be provided by professionals who have expertise in hearing loss, including educators of the deaf, speech-language pathologists, and audiologists.
   - In response to a previous emphasis on "natural environments," the JCIH recommends that both home-based and center-based intervention options be offered.
6. Surveillance and screening in the medical home
   - For all infants, regular surveillance of developmental milestones, auditory skills, parental concerns, and middle-ear status should be performed in the medical home, consistent with the American Academy of Pediatrics (AAP) pediatric periodicity schedule. All infants should have an objective standardized screening of global development with a validated assessment tool at 9, 18, and 24 to 30 months of age or at any time if the health care professional or family has concern.
   - Infants who do not pass the speech-language portion of a medical home global screening or for whom there is a concern regarding hearing or language should be referred for speech-language evaluation and audiology assessment.
7. Communication
   - The birth hospital, in collaboration with the state EHDI coordinator, should ensure that the hearing-screening results are conveyed to the parents and the medical home.
   - Parents should be provided with appropriate follow-up and resource information, and hospitals should ensure that each infant is linked to a medical home.
   - Information at all stages of the EHDI process is to be communicated to the family in a culturally sensitive and understandable format.
   - Individual hearing-screening information and audiology diagnostic and habilitation information should be promptly transmitted to the medical home and the state EHDI coordinator.
   - 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.
8. Information infrastructure
   - States should implement data-management and -tracking systems as part of an integrated child health information system to monitor the quality of EHDI services and provide recommendations for improving systems of care.

- An effective link between health and education professionals is needed to ensure successful transition and to determine outcomes of children with hearing loss for planning and establishing public health policy.

BACKGROUND

It has long been recognized that unidentified hearing loss at birth can adversely affect speech and language development as well as academic achievement and social-emotional development. Historically, moderate-to-severe hearing loss in young children was not detected until well beyond the newborn period, and it was not unusual for diagnosis of milder hearing loss and unilateral hearing loss to be delayed until children reached school age.

In the late 1980s, Dr C. Everett Koop, then US Surgeon General, on learning of new technology, encouraged detection of hearing loss to be included in the *Healthy People 2000*[4] goals for the nation. In 1988, the Maternal and Child Health Bureau (MCHB), a division of the US Health Resources and Services Administration (HRSA), funded pilot projects in Rhode Island, Utah, and Hawaii to test the feasibility of a universal statewide screening program to screen newborn infants for hearing loss before hospital discharge. The National Institutes of Health, through the National Institute on Deafness and Other Communication Disorders (NIDCD), issued in 1993 a consensus statement on early identification of hearing impairment in infants and young children.[5] In the statement the authors concluded that all infants admitted to the NICU should be screened for hearing loss before hospital discharge and that universal screening should be implemented for all infants within the first 3 months of life.[4] In its 1994 position statement, the JCIH endorsed the goal of universal detection of infants with hearing loss and encouraged continuing research and development to improve methods for identification of and intervention for hearing loss.[6,7] The AAP released a statement that recommended newborn hearing screening and intervention in 1999.[8] In 2000, citing advances in screening technology, the JCIH endorsed the universal screening of all infants through an integrated, interdisciplinary system of EHDI.[3] The *Healthy People 2010* goals included an objective to "increase the proportion of newborns who are screened for hearing loss by one month, have audiological evaluation by 3 months, and are enrolled in appropriate intervention services by 6 months."[9]

The ensuing years have seen remarkable expansion in newborn hearing screening. At the time of the National Institutes of Health consensus statement, only 11 hospitals in the United States were screening more than 90% of their newborn infants. In 2000, through the support of Representative Jim Walsh (R-NY), Congress authorized the HRSA to develop newborn hearing screening and follow-up services, the Centers for Disease Control and Prevention (CDC) to develop data and tracking systems, and the NIDCD to support research in EHDI. By 2005, every state had implemented a newborn hearing-screening program, and approximately 95% of newborn infants in the United States were screened for hearing loss before hospital discharge. Congress recommended cooperation and collaboration among several federal agencies and advocacy organizations to facilitate and support the development of state EHDI systems.

EHDI programs throughout the United States have demonstrated not only the feasibility of universal newborn hearing screening (UNHS) but also the benefits of early identification and intervention. There is a growing body of literature indicating that when identification and intervention occur at no later than 6 months of age for newborn infants who are deaf or hard of hearing, the infants perform as much as 20 to 40 percentile points higher on school-related measures (vocabulary, articulation, intelligibility, social adjustment, and behavior).[10–13] Still, many important challenges remain. Despite the fact that approximately 95% of newborn infants have their hearing screened in the United States, almost half of newborn infants who do not pass the initial screening do not have appropriate follow-up to either confirm the presence of a hearing loss and/or initiate appropriate early intervention services (see www.infanthearing.org, www.cdc.gov/ncbddd/ehdi, and www.nidcd.nih.gov/health).

State EHDI coordinators report system-wide problems including failure to communicate information to families in a culturally sensitive and understandable format at all stages of the EHDI process, lack of integrated state data-management and -tracking systems, and a shortage of facilities and personnel with the experience and expertise needed to provide follow-up for infants who are referred from newborn screening programs.[14] Available data indicate that a significant number of children who need further assessment do not receive appropriate follow-up evaluations. However, the outlook is improving as EHDI programs focus on the importance of strengthening follow-up and intervention.

PRINCIPLES

All children with hearing loss should have access to resources necessary to reach their maximum potential. The following principles provide the foundation for effective EHDI systems and have been updated and expanded since the 2000 JCIH position statement.

1. All infants should have access to hearing screening using a physiologic measure at no later than 1 month of age.
2. All infants who do not pass the initial hearing screening and the subsequent rescreening should have appropriate audiological and medical evaluations to

confirm the presence of hearing loss at no later than 3 months of age.

3. All infants with confirmed permanent hearing loss should receive early intervention services as soon as possible after diagnosis but at no later than 6 months of age. A simplified, single point of entry into an intervention system that is appropriate for children with hearing loss is optimal.
4. The EHDI system should be family centered with infant and family rights and privacy guaranteed through informed choice, shared decision-making, and parental consent in accordance with state and federal guidelines. Families should have access to information about all intervention and treatment options and counseling regarding hearing loss.
5. The child and family should have immediate access to high-quality technology including hearing aids, cochlear implants, and other assistive devices when appropriate.
6. All infants and children should be monitored for hearing loss in the medical home.[15] Continued assessment of communication development should be provided by appropriate professionals to all children with or without risk indicators for hearing loss.
7. Appropriate interdisciplinary intervention programs for infants with hearing loss and their families should be provided by professionals who are knowledgeable about childhood hearing loss. Intervention programs should recognize and build on strengths, informed choices, traditions, and cultural beliefs of the families.
8. Information systems should be designed and implemented to interface with electronic health charts and should be used to measure outcomes and report the effectiveness of EHDI services at the patient, practice, community, state, and federal levels.

GUIDELINES FOR EHDI PROGRAMS

The 2007 guidelines were developed to update the 2000 JCIH position statement principles and to support the goals of universal access to hearing screening, evaluation, and intervention for newborn and young infants embodied in *Healthy People 2010*.[9] The guidelines provide current information on the development and implementation of successful EHDI systems.

Hearing screening should identify infants with specifically defined hearing loss on the basis of investigations of long-term, developmental consequences of hearing loss in infants, currently available physiologic screening techniques, and availability of effective intervention in concert with established principles of health screening.[15–18] Studies have demonstrated that current screening technologies are effective in identifying hearing loss of moderate and greater degree.[19] In addition, studies of children with permanent hearing loss indicate that moderate or greater degrees of hearing loss can have significant effects on language, speech, academic, and social-emotional development.[20] High-risk target populations also include infants in the NICU, because research data have indicated that this population is at highest risk of having neural hearing loss.[21–23]

The JCIH, however, is committed to the goal of identifying all degrees and types of hearing loss in childhood and recognizes the developmental consequences of even mild degrees of permanent hearing loss. Recent evidence, however, has suggested that current hearing-screening technologies fail to identify some infants with mild forms of hearing loss.[24,25] In addition, depending on the screening technology selected, infants with hearing loss related to neural conduction disorders or "auditory neuropathy/auditory dyssynchrony" may not be detected through a UNHS program. Although the JCIH recognizes that these disorders may result in delayed communication,[26–28] currently recommended screening algorithms (ie, use of otoacoustic emission [OAE] testing alone) preclude universal screening for these disorders. Because these disorders typically occur in children who require NICU care,[21] the JCIH recommends screening this group with the technology capable of detecting auditory neuropathy/dyssynchrony: automated ABR measurement.

All infants, regardless of 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.

Roles and Responsibilities

The success of EHDI programs depends on families working in partnership with professionals as a well-coordinated team. The roles and responsibilities of each team member should be well defined and clearly understood. Essential team members are the birth hospital, families, pediatricians or primary health care professionals (ie, the medical home), audiologists, otolaryngologists, speech-language pathologists, educators of children who are deaf or hard of hearing, and other early intervention professionals involved in delivering EHDI services.[29,30] Additional services including genetics, ophthalmology, developmental pediatrics, service coordination, supportive family education, and counseling should be available.[31]

The birth hospital is a key member of the team. The birth hospital, in collaboration with the state EHDI coordinator, should ensure that parents and primary health care professionals receive and understand the hearing-screening results, that parents are provided with appropriate follow-up and resource information, and

that each infant is linked to a medical home.[2] The hospital ensures that hearing-screening information is transmitted promptly to the medical home and appropriate data are submitted to the state EHDI coordinator.

The most important role for the family of an infant who is deaf or hard of hearing is to love, nurture, and communicate with the infant. From this foundation, families usually develop an urgent desire to understand and meet the special needs of their infant. Families gain knowledge, insight, and experience by accessing resources and through participation in scheduled early intervention appointments including audiological, medical, habilitative, and educational sessions. This experience can be enhanced when families choose to become involved with parental support groups, people who are deaf or hard of hearing, and/or their children's deaf or hard-of-hearing peers. Informed family choices and desired outcomes guide all decisions for these children. A vital function of the family's role is ensuring direct access to communication in the home and the daily provision of language-learning opportunities. Over time, the child benefits from the family's modeling of partnerships with professionals and advocating for their rights in all settings. The transfer of responsibilities from families to the child develops gradually and increases as the child matures, growing in independence and self-advocacy.

Pediatricians, family physicians, and other allied health care professionals, working in partnership with parents and other professionals such as audiologists, therapists, and educators, constitute the infant's medical home.[2] A medical home is defined as an approach to providing health care services with which care is accessible, family centered, continuous, comprehensive, coordinated, compassionate, and culturally competent. The primary health care professional acts in partnership with parents in a medical home to identify and access appropriate audiology, intervention, and consultative services that are needed to develop a global plan of appropriate and necessary health and habilitative care for infants identified with hearing loss and infants with risk factors for hearing loss. All children undergo surveillance for auditory skills and language milestones. The infant's pediatrician, family physician, or other primary health care professional is in a position to advocate for the child and family.[2,16]

An audiologist is a person who, by virtue of academic degree, clinical training, and license to practice, is qualified to provide services related to the prevention of hearing loss and the audiological diagnosis, identification, assessment, and nonmedical and nonsurgical treatment of persons with impairment of auditory and vestibular function, and to the prevention of impairments associated with them. Audiologists serve in a number of roles. They provide newborn hearing-screening program development, management, quality assessment, service coordination and referral for audiological diagnosis, and audiological treatment and management. For the follow-up component, audiologists provide comprehensive audiological diagnostic assessment to confirm the existence of the hearing loss, ensure that parents understand the significance of the hearing loss, evaluate the infant for candidacy for amplification and other sensory devices and assistive technology, and ensure prompt referral to early intervention programs. For the treatment and management component, audiologists provide timely fitting and monitoring of amplification devices.[32] Other audiologists may provide diagnostic and auditory treatment and management services in the educational setting and provide a bridge between the child/family and the audiologist in the clinic setting as well as other service providers. Audiologists also provide services as teachers, consultants, researchers, and administrators.

Otolaryngologists are physicians whose specialty includes determining the etiology of hearing loss; identifying related risk indicators for hearing loss, including syndromes that involve the head and neck; and evaluating and treating ear diseases. An otolaryngologist with knowledge of childhood hearing loss can determine if medical and/or surgical intervention may be appropriate. When medical and/or surgical intervention is provided, the otolaryngologist is involved in the long-term monitoring and follow-up with the infant's medical home. The otolaryngologist provides information and participates in the assessment of candidacy for amplification, assistive devices, and surgical intervention, including reconstruction, bone-anchored hearing aids, and cochlear implantation.

Early intervention professionals are trained in a variety of academic disciplines such as speech-language pathology, audiology, education of children who are deaf or hard of hearing, service coordination, or early childhood special education. All individuals who provide services to infants with hearing loss should have specialized training and expertise in the development of audition, speech, and language. Speech-language pathologists provide both evaluation and intervention services for language, speech, and cognitive-communication development. Educators of children who are deaf or hard of hearing integrate the development of communicative competence within a variety of social, linguistic, and cognitive/academic contexts. Audiologists may provide diagnostic and habilitative services within the individualized family service plan (IFSP) or school-based individualized education plan. To provide the highest quality of intervention, more than 1 provider may be required.

The care coordinator is an integral member of the EHDI team and facilitates the family's transition from screening to evaluation to early intervention.[33] This person must be a professional (eg, social worker, teacher, nurse) who is knowledgeable about hearing loss. The care coordinator incorporates the family's preferences for outcomes into an IFSP as required by federal legisla-

tion. The care coordinator supports the family members in their choice of the infant's communicative development. Through the IFSP review, the infant's progress in language, motor, cognitive, and social-emotional development is monitored. The care coordinator assists the family in advocating for the infant's unique developmental needs.

The deaf and hard-of-hearing community includes members with direct experience with signed language, spoken language, hearing-aid and cochlear implant use, and other communication strategies and technologies. Optimally, adults who are deaf or hard-of-hearing should play an integral part in the EHDI program. Both adults and children in the deaf and hard-of-hearing community can enrich the family's experience by serving as mentors and role models. Such mentors have experience in negotiating their way in a hearing world, raising infants or children who are deaf or hard of hearing, and providing families with a full range of information about communication options, assistive technology, and resources that are available in the community.

A successful EHDI program requires collaboration between a variety of public and private institutions and agencies that assume responsibility for specific components (eg, screening, evaluation, intervention). Roles and responsibilities may differ from state to state. Each state has defined a lead coordinating agency with oversight responsibility. The lead coordinating agency in each state should be responsible for identifying the public and private funding sources available to develop, implement, and coordinate EHDI systems.

Hearing Screening

Multidisciplinary teams of professionals, including audiologists, physicians, and nursing personnel, are needed to establish the UNHS component of EHDI programs. All team members work together to ensure that screening programs are of high quality and are successful. An audiologist should be involved in each component of the hearing-screening program, particularly at the level of statewide implementation and, whenever possible, at the individual hospital level. Hospitals and agencies should also designate a physician to oversee the medical aspects of the EHDI program.

Each team of professionals responsible for the hospital-based UNHS program should review the hospital infrastructure in relationship to the screening program. Hospital-based programs should consider screening technology (ie, OAE or automated ABR testing); validity of the specific screening device; screening protocols, including the timing of screening relative to nursery discharge; availability of qualified screening personnel; suitability of the acoustical and electrical environments; follow-up referral criteria; referral pathways for follow-up; information management; and quality control and improvement. Reporting and communication protocols must be well defined and include the content of reports to physicians and parents, documentation of results in medical charts, and methods for reporting to state registries and national data sets.

Physiologic measures must be used to screen newborns and infants for hearing loss. Such measures include OAE and automated ABR testing. Both OAE and automated ABR technologies provide noninvasive recordings of physiologic activity underlying normal auditory function, both are easily performed in neonates and infants, and both have been successfully used for UNHS.[19,34–37] However, there are important differences between the 2 measures. OAE measurements are obtained from the ear canal by using a sensitive microphone within a probe assembly that records cochlear responses to acoustic stimuli. Thus, OAEs reflect the status of the peripheral auditory system extending to the cochlear outer hair cells. In contrast, ABR measurements are obtained from surface electrodes that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli delivered via an earphone. Automated ABR measurements reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway.

Both OAE and ABR screening technologies can be used to detect sensory (cochlear) hearing loss[19]; however, both technologies may be affected by outer or middle-ear dysfunction. Consequently, transient conditions of the outer and middle ear may result in a "failed" screening-test result in the presence of normal cochlear and/or neural function.[38] Moreover, because OAEs are generated within the cochlea, OAE technology cannot be used to detect neural (eighth nerve or auditory brainstem pathway) dysfunction. Thus, neural conduction disorders or auditory neuropathy/dyssynchrony without concomitant sensory dysfunction will not be detected by OAE testing.

Some infants who pass newborn hearing screening will later demonstrate permanent hearing loss.[25] Although this loss may reflect delayed-onset hearing loss, both ABR and OAE screening technologies will miss some hearing loss (eg, mild or isolated frequency region losses).

Interpretive criteria for pass/fail outcomes should reflect clear scientific rationale and should be evidence based.[39,40] Screening technologies that incorporate automated-response detection are necessary to eliminate the need for individual test interpretation, to reduce the effects of screener bias or operator error on test outcome, and to ensure test consistency across infants, test conditions, and screening personnel.[41–45] When statistical probability is used to make pass/fail decisions, as is the case for OAE and automated ABR screening devices, the likelihood of obtaining a pass outcome by chance alone is increased when screening is performed repeatedly.[46–48]

This principle must be incorporated into the policies of rescreening.

There are no national standards for the calibration of OAE or ABR instrumentation. Compounding this problem, there is a lack of uniform performance standards. Manufacturers of hearing-screening devices do not always provide sufficient supporting evidence to validate the specific pass/fail criteria and/or automated algorithms used in their instruments.[49] In the absence of national standards, audiologists must obtain normative data for the instruments and protocols they use.

The JCIH recognizes that there are important issues differentiating screening performed in the well-infant nursery from that performed in the NICU. Although the goals in each nursery are the same, numerous methodologic and technological issues must be considered in program design and pass/fail criteria.

Screening Protocols in the Well-Infant Nursery

Many inpatient well-infant screening protocols provide 1 hearing screening and, when necessary, a repeat screening no later than at the time of discharge from the hospital, using the same technology both times. Use of either technology in the well-infant nursery will detect peripheral (conductive and sensory) hearing loss of 40 dB or greater.[19] When automated ABR is used as the single screening technology, neural auditory disorders can also be detected.[50] Some programs use a combination of screening technologies (OAE testing for the initial screening followed by automated ABR for rescreening [ie, 2-step protocol[5]]) to decrease the fail rate at discharge and the subsequent need for outpatient follow-up.[34,35,37,51–53] With this approach, infants who do not pass an OAE screening but subsequently pass an automated ABR test are considered a screening "pass." Infants in the well-infant nursery who fail automated ABR testing should not be rescreened by OAE testing and "passed," because such infants are presumed to be at risk of having a subsequent diagnosis of auditory neuropathy/dyssynchrony.

Screening Protocols in the NICU

An NICU is defined as a facility in which a neonatologist provides primary care for the infant. Newborn units are divided into 3 categories:

- Level I: basic care, well-infant nurseries
- Level II: specialty care by a neonatologist for infants at moderate risk of serious complications
- Level III: a unit that provides both specialty and subspecialty care including the provision of life support (mechanical ventilation)

A total of 120 level-II NICUs and 760 level-III NICUs have been identified in the United States by survey, and infants who have spent time in the NICU represent 10% to 15% of the newborn population.[54]

The 2007 JCIH position statement includes neonates at risk of having neural hearing loss (auditory neuropathy/auditory dyssynchrony) in the target population to be identified in the NICU,[55–57] because there is evidence that neural hearing loss results in adverse communication outcomes.[22,50] Consequently, the JCIH recommends ABR technology as the only appropriate screening technique for use in the NICU. For infants who do not pass automated ABR testing in the NICU, referral should be made directly to an audiologist for rescreening and, when indicated, comprehensive evaluation, including diagnostic ABR testing, rather than for general outpatient rescreening.

Conveying Test Results

Screening results should be conveyed immediately to families so that they understand the outcome and the importance of follow-up when indicated. To facilitate this process for families, primary health care professionals should work with EHDI team members to ensure that:

- communications with parents are confidential and presented in a caring and sensitive manner, preferably face-to-face;
- educational materials are developed and disseminated to families that provide accurate information at an appropriate reading level and in a language they are able to comprehend; and
- parents are informed in a culturally sensitive and understandable manner that their infant did not pass screening and informed about the importance of prompt follow-up; before discharge, an appointment should be made for follow-up testing.

To facilitate this process for primary care physicians, EHDI systems should ensure that medical professionals receive:

- the results of the screening test (pass, did not pass, or missed) as documented in the hospital medical chart; and
- communication directly from a representative of the hospital screening program regarding each infant in its care who did not pass or was missed and recommendations for follow-up.

Outpatient Rescreening for Infants Who Do Not Pass the Birth Admission Screening

Many well-infant screening protocols will incorporate an outpatient rescreening within 1 month of hospital discharge to minimize the number of infants referred for follow-up audiological and medical evaluation. The out-

patient rescreening should include the testing of both ears, even if only 1 ear failed the inpatient screening.

Outpatient screening at no later than 1 month of age should also be available to infants who were discharged before receiving the birth admission screening or who were born outside a hospital or birthing center. State EHDI coordinators should be aware of some of the following situations under which infants may be lost to the UNHS system:

- Home births and other out-of-hospital births: states should develop a mechanism to systematically offer newborn hearing screening for all out-of-hospital births.
- Across-state-border births: states should develop written collaborative agreements among neighboring states for sharing hearing-screening results and follow-up information.
- Hospital-missed screenings: when infants are discharged before the hearing screening is performed, a mechanism should be in place for the hospital to contact the family and arrange for an outpatient hearing screening.
- Transfers to in-state or out-of-state hospitals: discharge and transfer forms should contain the information of whether a hearing screening was performed and the results of any screening. The recipient hospital should complete a hearing screening if one was not previously performed or if there is a change in medical status or a prolonged hospitalization.
- Readmissions: for readmissions in the first month of life when there are conditions associated with potential hearing loss (eg, hyperbilirubinemia that requires exchange transfusion or culture-positive sepsis), an ABR screening should be performed before discharge.

Additional mechanisms for states to share hearing-screening results and other medical information include (l) incorporating the hearing-screening results in a statewide child health information system and (2) providing combined metabolic screening and hearing-screening results to the primary care physician.

Confirmation of Hearing Loss in Infants Referred From UNHS

Infants who meet the defined criteria for referral should receive follow-up audiological and medical evaluations with fitting of amplification devices, as appropriate, at no later than 3 months of age. Once hearing loss is confirmed, coordination of services should be expedited by the infant's medical home and Part C coordinating agencies for early intervention services, as authorized by the Individuals With Disabilities Education Act, following the EHDI algorithm developed by the AAP (Appendix 1).

Audiological Evaluation

Comprehensive audiological evaluation of newborn and young infants who fail newborn hearing screening should be performed by audiologists experienced in pediatric hearing assessment. The initial audiological test battery to confirm a hearing loss in infants must include physiologic measures and, when developmentally appropriate, behavioral methods. Confirmation of an infant's hearing status requires a test battery of audiological test procedures to assess the integrity of the auditory system in each ear, to estimate hearing sensitivity across the speech frequency range, to determine the type of hearing loss, to establish a baseline for further monitoring, and to provide information needed to initiate amplification-device fitting. A comprehensive assessment should be performed on both ears even if only 1 ear failed the screening test.

Evaluation: Birth to 6 Months of Age

For infants from birth to a developmental age of approximately 6 months, the test battery should include a child and family history, an evaluation of risk factors for congenital hearing loss, and a parental report of the infant's responses to sound. The audiological assessment should include:

- Child and family history.
- A frequency-specific assessment of the ABR using air-conducted tone bursts and bone-conducted tone bursts when indicated. When permanent hearing loss is detected, frequency-specific ABR testing is needed to determine the degree and configuration of hearing loss in each ear for fitting of amplification devices.
- Click-evoked ABR testing using both condensation and rarefaction single-polarity stimulus, if there are risk indicators for neural hearing loss (auditory neuropathy/auditory dyssynchrony) such as hyperbilirubinemia or anoxia, to determine if a cochlear microphonic is present.[28] Furthermore, because some infants with neural hearing loss have no risk indicators, any infant who demonstrates "no response" on ABR elicited by tone-burst stimuli must be evaluated by a click-evoked ABR.[55]
- Distortion product or transient evoked OAEs.
- Tympanometry using a 1000-Hz probe tone.
- Clinician observation of the infant's auditory behavior as a cross-check in conjunction with electrophysiologic measures. Behavioral observation alone is not adequate for determining whether hearing loss is present in this age group, and it is not adequate for the fitting of amplification devices.

Evaluation: 6 to 36 Months of Age

For subsequent testing of infants and toddlers at developmental ages of 6 to 36 months, the confirmatory audiological test battery includes:

- Child and family history.
- Parental report of auditory and visual behaviors and communication milestones.
- Behavioral audiometry (either visual reinforcement or conditioned-play audiometry, depending on the child's developmental level), including pure-tone audiometry across the frequency range for each ear and speech-detection and -recognition measures.
- OAE testing.
- Acoustic immittance measures (tympanometry and acoustic reflex thresholds).
- ABR testing if responses to behavioral audiometry are not reliable or if ABR testing has not been performed in the past.

Other Audiological Test Procedures

At this time, there is insufficient evidence for use of the auditory steady-state response as the sole measure of auditory status in newborn and infant populations.[58] Auditory steady-state response is a new evoked-potential test that can accurately measure auditory sensitivity beyond the limits of other test methods. It can determine frequency-specific thresholds from 250 Hz to 8 kHz. Clinical research is being performed to investigate its potential use in the standard pediatric diagnostic test battery. Similarly, there are insufficient data for routine use of acoustic middle-ear muscle reflexes in the initial diagnostic assessment of infants younger than 4 months.[59] Both tests could be used to supplement the battery or could be included at older ages. Emerging technologies, such as broad-band reflectance, may be used to supplement conventional measures of middle-ear status (tympanometry and acoustic reflexes) as the technology becomes more widely available.[59]

Medical Evaluation

Every infant with confirmed hearing loss and/or middle-ear dysfunction should be referred for otologic and other medical evaluation. The purpose of these evaluations is to determine the etiology of hearing loss, to identify related physical conditions, and to provide recommendations for medical/surgical treatment as well as referral for other services. Essential components of the medical evaluation include clinical history, family history of childhood-onset permanent hearing loss, identification of syndromes associated with early- or late-onset permanent hearing loss, a physical examination, and indicated radiologic and laboratory studies (including genetic testing). Portions of the medical evaluation, such as urine culture for CMV, a leading cause of hearing loss, might even begin in the birth hospital, particularly for infants who spend time in the NICU.[60–62]

Pediatrician/Primary Care Physician

The infant's pediatrician or other primary health care professional is responsible for monitoring the general health, development, and well-being of the infant. In addition, the primary care physician must assume responsibility to ensure that the audiological assessment is conducted on infants who do not pass screening and must initiate referrals for medical specialty evaluations necessary to determine the etiology of the hearing loss. Middle-ear status should be monitored, because the presence of middle-ear effusion can further compromise hearing. The primary care physician must partner with other specialists, including the otolaryngologist, to facilitate coordinated care for the infant and family. Because 30% to 40% of children with confirmed hearing loss will demonstrate developmental delays or other disabilities, the primary care physician should closely monitor developmental milestones and initiate referrals related to suspected disabilities.[63] The medical home algorithm for management of infants with either suspected or proven permanent hearing loss is provided in Appendix 1.[15]

The pediatrician or 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 their newborn screening results.[25] Infants with specific risk factors, such as those who received ECMO therapy and those with CMV infection, are at increased risk of delayed-onset or progressive hearing loss[64–67] and should be monitored closely. In addition, the primary care physician is responsible for ongoing surveillance of parent concerns about language and hearing, auditory skills, and developmental milestones of all infants and children regardless of risk status, as outlined in the pediatric periodicity schedule published by the AAP.[16]

Children with cochlear implants may be at increased risk of acquiring bacterial meningitis compared with children in the general US population.[68] The CDC recommends that all children with, and all potential recipients of, cochlear implants follow specific recommendations for pneumococcal immunization that apply to cochlear implant users and that they receive age-appropriate *Haemophilus influenzae* type b vaccines. Recommendations for the timing and type of pneumococcal vaccine vary with age and immunization history and should be discussed with a health care professional.[69]

Otolaryngologist

Otolaryngologists are physicians and surgeons who diagnose, treat, and manage a wide range of diseases of the head and neck and specialize in treating hearing and vestibular disorders. They perform a full medical diagnostic evaluation of the head and neck, ears, and related structures, including a comprehensive history and physical examination, leading to a medical diagnosis and appropriate medical and surgical management. Often, a hearing or balance disorder is an indicator of, or related to, a medically treatable condition or an underlying systemic disease. Otolaryngologists work closely with other dedicated professionals, including physicians, audiologists, speech-language pathologists, educators, and others, in caring for patients with hearing, balance, voice, speech, developmental, and related disorders.

The otolaryngologist's evaluation includes a comprehensive history to identify the presence of risk factors for early-onset childhood permanent hearing loss, such as family history of hearing loss, having been admitted to the NICU for more than 5 days, and having received ECMO (see Appendix 2).[70,71]

A complete head and neck examination for craniofacial anomalies should document defects of the auricles, patency of the external ear canals, and status of the eardrum and middle-ear structures. Atypical findings on eye examination, including irises of 2 different colors or abnormal positioning of the eyes, may signal a syndrome that includes hearing loss. Congenital permanent conductive hearing loss may be associated with craniofacial anomalies that are seen in disorders such as Crouzon disease, Klippel-Feil syndrome, and Goldenhar syndrome.[72] The assessment of infants with these congenital anomalies should be coordinated with a clinical geneticist.

In large population studies, at least 50% of congenital hearing loss has been designated as hereditary, and nearly 600 syndromes and 125 genes associated with hearing loss have already been identified.[72,73] The evaluation, therefore, should include a review of family history of specific genetic disorders or syndromes, including genetic testing for gene mutations such as *GJB2* (connexin-26), and syndromes commonly associated with early-onset childhood sensorineural hearing loss[72,74–76] (Appendix 2). As the widespread use of newly developed conjugate vaccines decreases the prevalence of infectious etiologies such as measles, mumps, rubella, *H influenzae* type b, and childhood meningitis, the percentage of each successive cohort of early-onset hearing loss attributable to genetic etiologies can be expected to increase, prompting recommendations for early genetic evaluations. Approximately 30% to 40% of children with hearing loss have associated disabilities, which can be of importance in patient management. The decision to obtain genetic testing depends on informed family choice in conjunction with standard confidentiality guidelines.[77]

In the absence of a genetic or established medical cause, a computed tomography scan of the temporal bones may be performed to identify cochlear abnormalities, such as Mondini deformity with an enlarged vestibular aqueduct, which have been associated with progressive hearing loss. Temporal bone imaging studies may also be used to assess potential candidacy for surgical intervention, including reconstruction, bone-anchored hearing aid, and cochlear implantation. Recent data have shown that some children with electrophysiologic evidence suggesting auditory neuropathy/dyssynchrony may have an absent or abnormal cochlear nerve that may be detected with MRI.[78]

Historically, an extensive battery of laboratory and radiographic studies was routinely recommended for newborn infants and children with newly diagnosed sensorineural hearing loss. However, emerging technologies for the diagnosis of genetic and infectious disorders have simplified the search for a definitive diagnosis, which obviates the need for costly diagnostic evaluations in some instances.[70,71,79]

If, after an initial evaluation, the etiology remains uncertain, an expanded multidisciplinary evaluation protocol including electrocardiography, urinalysis, testing for CMV, and further radiographic studies is indicated. The etiology of neonatal hearing loss, however, may remain uncertain in as many as 30% to 40% of children. Once hearing loss is confirmed, medical clearance for hearing aids and initiation of early intervention should not be delayed while this diagnostic evaluation is in process. Careful longitudinal monitoring to detect and promptly treat coexisting middle-ear effusions is an essential component of ongoing otologic management of these children.

Other Medical Specialists

The medical geneticist is responsible for the interpretation of family history data, the clinical evaluation and diagnosis of inherited disorders, the performance and assessment of genetic tests, and the provision of genetic counseling. Geneticists or genetic counselors are qualified to interpret the significance and limitations of new tests and to convey the current status of knowledge during genetic counseling. All families of children with confirmed hearing loss should be offered, and may benefit from, a genetics evaluation and counseling. This evaluation can provide families with information on etiology of hearing loss, prognosis for progression, associated disorders (eg, renal, vision, cardiac), and likelihood of recurrence in future offspring. This information may influence parents' decision-making regarding intervention options for their child.

Every infant with a confirmed hearing loss should have an evaluation by an ophthalmologist to document

visual acuity and rule out concomitant or late-onset vision disorders such as Usher syndrome.[1,80] Indicated referrals to other medical subspecialists, including developmental pediatricians, neurologists, cardiologists, and nephrologists, should be facilitated and coordinated by the primary health care professional.

Early Intervention

Before newborn hearing screening was instituted universally, children with severe-to-profound hearing loss, on average, completed the 12th grade with a 3rd- to 4th-grade reading level and language levels of a 9- to 10-year-old hearing child.[81] In contrast, infants and children with mild-to-profound hearing loss who are identified in the first 6 months of life and provided with immediate and appropriate intervention have significantly better outcomes than later-identified infants and children in vocabulary development,[82,83] receptive and expressive language,[12,84] syntax,[85] speech production,[13,86–88] and social-emotional development.[89] Children enrolled in early intervention within the first year of life have also been shown to have language development within the normal range of development at 5 years of age.[31,90]

Therefore, according to federal guidelines, once any degree of hearing loss is diagnosed in a child, a referral should be initiated to an early intervention program within 2 days of confirmation of hearing loss (CFR 303.321d). The initiation of early intervention services should begin as soon as possible after diagnosis of hearing loss but at no later than 6 months of age. Even when the hearing status is not determined to be the primary disability, the family and child should have access to intervention with a provider who is knowledgeable about hearing loss.[91]

UNHS programs have been instituted throughout the United States for the purpose of preventing the significant and negative effects of hearing loss on the cognitive, language, speech, auditory, social-emotional, and academic development of infants and children. To achieve this goal, hearing loss must be identified as quickly as possible after birth, and appropriate early intervention must be available to all families and infants with permanent hearing loss. Some programs have demonstrated that most children with hearing loss and no additional disabilities can achieve and maintain language development within the typical range of children who have normal hearing.[12,13,85,90] Because these studies were descriptive and not causal studies, the efficacy of specific components of intervention cannot be separated from the total provision of comprehensive services. Thus, the family-centered philosophy, the intensity of services, the experience and training of the provider, the method of communication, the curricula, the counseling procedures, the parent support and advocacy, and the deaf and hard-of-hearing support and advocacy are all variables with unknown effects on the overall outcomes of any individual child. The key component of providing quality services is the expertise of the provider specific to hearing loss. These services may be provided in the home, a center, or a combination of the 2 locations.

The term "intervention services" is used to describe any type of habilitative, rehabilitative, or educational program provided to children with hearing loss. In some cases of mild hearing losses, amplification technology may be the only service provided. Some parents choose only developmental assessment or occasional consultation, such as parents with infants who have unilateral hearing losses. Children with high-frequency losses and normal hearing in the low frequencies may only be seen by a speech-language pathologist, and those with significant bilateral sensorineural hearing losses might be seen by an educator of the deaf and receive additional services.

Principles of Early Intervention

To ensure informed decision-making, parents of infants with newly diagnosed hearing loss should be offered opportunities to interact with other families who have infants or children with hearing loss as well as adults and children who are deaf or hard of hearing. In addition, parents should also be offered access to professional, educational, and consumer organizations and provided with general information on child development, language development, and hearing loss. A number of principles and guidelines have been developed that offer a framework for quality early intervention service delivery systems for children who are deaf or hard of hearing and their families.[92] Foundational characteristics of developing and implementing early intervention programs include a family-centered approach, culturally responsive practices, collaborative professional-family relationships and strong family involvement, developmentally appropriate practice, interdisciplinary assessment, and community-based provision of services.

Designated Point of Entry

States should develop a single point of entry into intervention specific for hearing impairment to ensure that, regardless of geographic location, all families who have infants or children with hearing loss receive information about a full range of options regarding amplification and technology, communication and intervention, and accessing appropriate counseling services. This state system, if separate from the state's Part C system, should integrate and partner with the state's Part C program. Parental consent must be obtained according to state and federal requirements to share the IFSP information with providers and transmit data to the state EHDI coordinator.

Regular Developmental Assessment

To ensure accountability, individual, community, and state health and educational programs should assume the responsibility for coordinated, ongoing measurement and improvement of EHDI process outcomes. Early intervention programs must assess the language, cognitive skills, auditory skills, speech, vocabulary, and social-emotional development of all children with hearing loss at 6-month intervals during the first 3 years of life by using assessment tools that have been standardized on children with normal hearing and norm-referenced assessment tools that are appropriate to measure progress in verbal and visual language.

The primary purpose of regular developmental monitoring is to provide valuable information to parents about the rate of their child's development as well as programmatic feedback concerning curriculum decisions. Families also become knowledgeable about expectations and milestones of typical development of hearing children. Studies have shown that valid and reliable documentation of developmental progress is possible through parent questionnaires, analysis of videotaped conversational interactions, and clinically administered assessments.* Documentation of developmental progress should be provided on a regular basis to parents and, with parental release of information, to the medical home and audiologist. Although criterion-referenced checklists may provide valuable information for establishing intervention strategies and goals, these assessment tools alone are not sufficient for parents and intervention professionals to determine if a child's developmental progress is comparable with his or her hearing peers.

Opportunities for Interaction With Other Parents of Children With Hearing Loss

Intervention professionals should seek to involve parents at every level of the EHDI process and develop true and meaningful partnerships with parents. To reflect the value of the contributions that selected parents make to development and program components, these parents should be paid as contributing staff members. Parent representatives should be included in all advisory board activities. In many states, parents have been integral and often have taken leadership roles in the development of policy, resource material, communication mechanisms, mentoring and advocacy opportunities, dissemination of information, and interaction with the deaf community and other individuals who are deaf or hard of hearing. Parents, often in partnership with people who are deaf and hard of hearing, have also participated in the training of professionals. They should be participants in the regular assessment of program services to ensure ongoing improvement and quality assurance.

Opportunities for Interaction With Individuals Who Are Deaf or Hard of Hearing

Intervention programs should include opportunities for involvement of individuals who are deaf or hard of hearing in all aspects of EHDI programs. Because intervention programs serve children with mild-to-profound, unilateral or bilateral, permanent conductive, and sensory or neural hearing disorders, role models who are deaf or hard of hearing can be significant assets to an intervention program. These individuals can serve on state EHDI advisory boards and be trained as mentors for families and children with hearing loss who choose to seek their support. Almost all families choose at some time during their early childhood programs to seek out both adults and child peers with hearing loss. Programs should ensure that these opportunities are available and can be delivered to families through a variety of communications means, such as Web sites, e-mail, newsletters, videos, retreats, picnics and other social events, and educational forums for parents.

Provision of Communication Options

Research studies thus far of early-identified infants with hearing loss have not found significant differences in the developmental outcomes by method of communication when measured at 3 years of age.† Therefore, a range of options should be offered to families in a nonbiased manner. In addition, there have been reports of children with successful outcomes for each of the different methods of communication. The choice is a dynamic process on a continuum, differs according to the individual needs of each family, and can be adjusted as necessary on the basis of a child's rate of progress in developing communication skills. Programs need to provide families with access to skilled and experienced early intervention professionals to facilitate communication and language development in the communication option chosen by the family.

Skills of the Early Intervention Professional

All studies with successful outcomes reported for early-identified children who are deaf or hard of hearing have intervention provided by specialists who are trained in parent-infant intervention services.[12,90,97] Early intervention programs should develop mechanisms to ensure that early intervention professionals have special skills necessary for providing families with the highest quality of service specific to children with hearing loss. Professionals with a background in deaf education, audiology, and speech-language pathology will typically have the skills needed for providing intervention services. Professionals should be highly qualified in their respective fields and should be skilled communicators who are knowledgeable and sensitive to the importance of en-

*Refs 10–13, 51, 85, 87–90, and 93–96.

†Refs 10–13, 85, 87, 88, 90, 93, and 96.

hancing families' strengths and supporting their priorities. When early intervention professionals have knowledge of the principles of adult learning, it increases their success with parents and other professionals.

Quality of Intervention Services

Children with confirmed hearing loss and their families have the right to prompt access to quality intervention services. For newborn infants with confirmed hearing loss, enrollment into intervention services should begin as soon after hearing-loss confirmation as possible and no later than 6 months of age. Successful early intervention programs (1) are family centered, (2) provide families with unbiased information on all options regarding approaches to communication, (3) monitor development at 6-month intervals with norm-referenced instruments, (4) include individuals who are deaf or hard of hearing, (5) provide services in a natural environment in the home or in the center, (6) offer high-quality service regardless of where the family lives, (7) obtain informed consent, (8) are sensitive to cultural and language differences and provide accommodations as needed, and (9) conduct annual surveys of parent satisfaction.

Intervention for Special Populations of Infants and Young Children

Developmental monitoring should also occur at regular 6-month intervals for special populations of children with hearing loss, including those with minimal and mild bilateral hearing loss,[98] unilateral hearing loss,[99,100] and neural hearing loss,[22] because these children are at risk of having speech and language delay. Research findings indicate that approximately one third of children with permanent unilateral loss experience significant language and academic delays.[99–101]

Audiological Habilitation

Most infants and children with bilateral hearing loss and many with unilateral hearing loss benefit from some form of personal amplification device.[32] If the family chooses personal amplification for its infant, hearing-aid selection and fitting should occur within 1 month of initial confirmation of hearing loss even when additional audiological assessment is ongoing. Audiological habilitation services should be provided by an audiologist who is experienced with these procedures. Delay between confirmation of the hearing loss and fitting of an amplification device should be minimized.[51,102]

Hearing-aid fitting proceeds optimally when the results of physiologic audiological assessment including diagnostic ABR, OAE, and tympanometry and medical examination are in accord. For infants who are below a developmental age of 6 months, hearing-aid selection will be based on physiologic measures alone. Behavioral threshold assessment with visual reinforcement audiometry should be obtained as soon as possible to cross-check and augment physiologic findings (see www.audiology.org).

The goal of amplification-device fitting is to provide the infant with maximum access to all of the acoustic features of speech within an intensity range that is safe and comfortable. That is, amplified speech should be comfortably above the infant's sensory threshold but below the level of discomfort across the speech frequency range for both ears. To accomplish this in infants, amplification-device selection, fitting, and verification should be based on a prescriptive procedure that incorporates individual real-ear measures that account for each infant's ear-canal acoustics and hearing loss.[32] Validation of the benefits of amplification, particularly for speech perception, should be examined in the clinical setting as well as in the child's typical listening environments. Complementary or alternative technology, such as frequency modulation (FM) systems or cochlear implants, may be recommended as the primary and/or secondary listening device depending on the degree of the infant's hearing loss, the goals of auditory habilitation, the infant's acoustic environments, and the family's informed choices.[3] Monitoring of amplification, as well as the long-term validation of the appropriateness of the individual habilitation program, requires ongoing audiological assessment along with electroacoustic, real-ear, and functional checks of the hearing instruments. As the hearing loss becomes more specifically defined through audiological assessments and as the child's ear-canal acoustics change with growth, refinement of the individual prescriptive hearing-aid gain and output targets is necessary. Monitoring also includes periodic validation of communication, social-emotional, and cognitive development and, later, academic performance to ensure that progress is commensurate with the child's abilities. It is possible that infants and young children with measurable residual "hearing" (auditory responses) and well-fit amplification devices may fail to develop auditory skills necessary for successful spoken communication. Ongoing validation of the amplification device is accomplished through interdisciplinary evaluation and collaboration with the early intervention team and family.

Cochlear implantation should be given careful consideration for any child who seems to receive limited benefit from a trial with appropriately fitted hearing aids. According to US Food and Drug Administration guidelines, infants with profound bilateral hearing loss are candidates for cochlear implantation at 12 months of age and children with bilateral severe hearing loss are eligible at 24 months of age. The presence of developmental conditions (eg, developmental delay, autism) in addition to hearing loss should not, as a rule, preclude the consideration of cochlear implantation for an infant or child who is deaf. Benefits from hearing aids and cochlear implants in children with neural hearing loss

have also been documented. The benefit of acoustic amplification for children with neural hearing loss is variable.[28,103] Thus, a trial fitting is indicated for infants with neural hearing loss until the usefulness of the fitting can be determined. Neural hearing loss is a heterogeneous condition; the decision to continue or discontinue use of hearing aids should be made on the basis of the benefit derived from amplification. Use of cochlear implants in neural hearing loss is growing, and positive outcomes have been reported for many children.[28]

Infants and young children with unilateral hearing loss should also be assessed for appropriateness of hearing-aid fitting. Depending on the degree of residual hearing in unilateral loss, a hearing aid may or may not be indicated. Use of "contralateral routing of signals" amplification for unilateral hearing loss in children is not recommended.[104] Research is currently underway to determine how to best manage unilateral hearing loss in infants and young children.

The effect of otitis media with effusion (OME) is greater for infants with sensorineural hearing loss than for those with normal cochlear function.[73] Sensory or permanent conductive hearing loss is compounded by additional transient conductive hearing loss associated with OME. OME further reduces access to auditory cues necessary for the development of spoken English. OME also negatively affects the prescriptive targets of the hearing-aid fitting, decreasing auditory awareness and requiring adjustment of the amplification characteristics. Prompt referral to either the primary care physician or an otolaryngologist for treatment of persistent OME is indicated in infants with sensorineural hearing loss.[105] Definitive resolution of OME should never delay the fitting of an amplification device.[73,106]

Medical and Surgical Intervention

Medical intervention is the process by which a physician provides medical diagnosis and direction for medical and/or surgical treatment options for hearing loss and/or related medical disorder(s) associated with hearing loss. Treatment varies from the removal of cerumen and the treatment of OME to long-term plans for reconstructive surgery and assessment of candidacy for cochlear implants. If necessary, surgical treatment of malformation of the outer and middle ears, including bone-anchored hearing aids, should be considered in the intervention plan for infants with permanent conductive or mixed hearing loss when they reach an appropriate age.

Communication Assessment and Intervention

Language is acquired with greater ease during certain sensitive periods of infant and toddler development.[107–109] The process of language acquisition includes learning the precursors of language, such as the rules that pertain to selective attention and turn taking.[20,110,111] Cognitive, social, and emotional development are influenced by the acquisition of language. Development in these areas is synergistic. A complete language evaluation should be performed at regular intervals for infants and toddlers with hearing loss. The evaluation should include an assessment of oral, manual, and/or visual mechanisms as well as cognitive abilities.

A primary focus of language intervention is to support families in fostering the communication abilities of their infants and toddlers who are deaf or hard of hearing.[20] Spoken- and/or sign-language development should be commensurate with the child's age and cognitive abilities and should include acquisition of phonologic (for spoken language), visual/spatial/motor (for signed language), morphologic, semantic, syntactic, and pragmatic skills, depending on the family's preferred mode of communication.

Early intervention professionals should follow family-centered principles to assist in developing communicative competence of infants and toddlers who are deaf or hard of hearing.[112–114] Families should be provided with information specific to language development and access to peer and language models as well as family-involved activities that facilitate language development of children with normal hearing and children who are hard of hearing or deaf.[115,116] Depending on family choices, families should be offered access to children and adults with hearing loss who are appropriate and competent language models. Information on spoken language and signed language, such as American Sign Language[117] and cued speech, should be provided.

Continued Surveillance, Screening, and Referral of Infants and Toddlers

Appendix 2 presents 11 risk indicators that are associated with either congenital or delayed-onset hearing loss. A single list of risk indicators is presented in the current JCIH statement, because there is significant overlap among those indicators associated with congenital/neonatal hearing loss and those associated with delayed-onset/acquired or progressive hearing loss. Heightened surveillance of all infants with risk indicators, therefore, is recommended. There is a significant change in the definition of risk-indicator 3, which has been modified from NICU stay more than 48 hours to NICU stay more than 5 days. Consistent with 2000 JCIH position statement,[3] the 2007 position statement recommends use of risk indicators for hearing loss for 3 purposes. Historically, the first use of risk indicators is for the identification of infants who should receive audiological evaluation but who live in geographic locations (eg, developing nations, remote areas) where universal hearing screening is not yet available.‡ This use has become less common as a result of the expansion of

‡Refs 3, 19, 21, 24, 25, 64, and 118–124.

UNHS. The second purpose of risk-indicator identification is to help identify infants who pass the neonatal screening but are at risk of developing delayed-onset hearing loss and, therefore, should receive ongoing medical, speech and language, and audiological surveillance. Third, the risk indicators are used to identify infants who may have passed neonatal screening but have mild forms of permanent hearing loss.[25]

Because some important indicators, such as family history of hearing loss, may not be determined during the course of UNHS,[14,72] the presence of all risk indicators for acquired hearing loss should be determined in the medical home during early well-infant visits. Risk indicators that are marked with a section symbol in Appendix 2 are of greater concern for delayed-onset hearing loss. Early and more frequent assessment may be indicated for children with CMV infection,[118,125,126] syndromes associated with progressive hearing loss,[72] neurodegenerative disorders,[72] trauma,[127–129] or culture-positive postnatal infections associated with sensorineural hearing loss[130,131]; for children who have received ECMO[64] or chemotherapy[132]; and when there is caregiver concern or a family history of hearing loss.[16]

For all infants with and without risk indicators for hearing loss, developmental milestones, hearing skills, and parent concerns about hearing, speech, and language skills should be monitored during routine medical care consistent with the AAP periodicity schedule.

The JCIH has determined that the previously recommended approach to follow-up of infants with risk indicators for hearing loss only addressed children with identifiable risk indicators and failed to consider the possibility of delayed-onset hearing loss in children without identifiable risk indicators. In addition, concerns were raised about feasibility and cost associated with the 2000 JCIH recommendation for audiological monitoring of all infants with risk indicators at 6-month intervals. Because approximately 400 000 infants are cared for annually in NICUs in the United States, and the 2000 JCIH recommendation included audiology assessments at 6-month intervals from 6 months to 36 months of age for all infants admitted to an NICU for more than 48 hours, an unreasonable burden was placed on both providers of audiology services and families. In addition, there was no provision for identification of delayed-onset hearing loss in infants without an identifiable risk indicator. Data from 2005 for 12 388 infants discharged from NICUs in the National Perinatal Information Network indicated that 52% of infants were discharged within the first 5 days of life, and these infants were significantly less likely to have an identified risk indicator for hearing loss other than NICU stay. Therefore, the 2007 JCIH recommends an alternative, more inclusive strategy of surveillance of all children within the medical home based on the pediatric periodicity schedule. This protocol will permit the detection of children with either missed neonatal or delayed-onset hearing loss irrespective of the presence or absence of a high-risk indicator.

The JCIH recognizes that an optimal surveillance and screening program within the medical home would include the following:

- At each visit, consistent with the AAP periodicity schedule, infants should be monitored for auditory skills, middle-ear status, and developmental milestones (surveillance). Concerns elicited during surveillance should be followed by administration of a validated global screening tool.[133] A validated global screening tool is administered to all infants at 9, 18, and 24 to 30 months or, if there is physician or parental concern about hearing or language, sooner.[133]
- If an infant does not pass the speech-language portion of the global screening in the medical home or if there is physician or caregiver concern about hearing or spoken-language development, the child should be referred immediately for further evaluation by an audiologist and a speech-language pathologist for a speech and language evaluation with validated tools.[133]
- Once hearing loss is diagnosed in an infant, siblings who are at increased risk of having hearing loss should be referred for audiological evaluation.[14,75,134,135]
- All infants with a risk indicator for hearing loss (Appendix 2), regardless of surveillance findings, should be referred for an audiological assessment at least once by 24 to 30 months of age. Children with risk indicators that are highly associated with delayed-onset hearing loss, such as having received ECMO or having CMV infection, should have more frequent audiological assessments.
- All infants for whom the family has significant concerns regarding hearing or communication should be promptly referred for an audiological and speech-language assessment.
- A careful assessment of middle-ear status (using pneumatic otoscopy and/or tympanometry) should be completed at all well-child visits, and children with persistent middle-ear effusion that last for 3 months or longer should be referred for otologic evaluation.[136]

Protecting the Rights of Infants and Families

Each agency or institution involved in the EHDI process shares responsibility for protecting infant and family rights in all aspects of UNHS, including access to information including potential benefits and risks in the family's native language, input into decision-making, and confidentiality.[77] Families should receive information about childhood hearing loss in easily understood language. Families have the right to accept or decline hearing screening or any follow-up care for their newborn

infant within the statutory regulations, just as they have for any other screening or evaluation procedures or intervention.

EHDI data merit the same level of confidentiality and security afforded all other health care and education information in practice and law. The infant's family has the right to confidentiality of the screening and follow-up assessments and the acceptance or rejection of suggested intervention(s). In compliance with federal and state laws, mechanisms should be established that ensure parental release and approval of all communications regarding the infant's test results, including those to the infant's medical home and early intervention–coordinating agency and programs. The Health Insurance Portability and Accountability Act (Pub L No. 104-191 [1996]) regulations permit the sharing of health information among health care professionals.

Information Infrastructure

In its 2000 position statement,[3] the JCIH recommended development of uniform state registries and national information databases that incorporate standardized methodology, reporting, and system evaluation. EHDI information systems are to provide for the ongoing and systematic collection, analysis, and interpretation of data in the process of measuring and reporting associated program services (eg, screening, evaluation, diagnosis, and/or intervention). These systems are used to guide activities, planning, implementation, and evaluation of programs and to formulate research hypotheses.

EHDI information systems are generally authorized by legislators and implemented by public health officials. These systems vary from a simple system that collects data from a single source to electronic systems that receive data from many sources in multiple formats. The number and variety of systems will likely increase with advances in electronic data interchange and integration of data, which will also heighten the importance of patient privacy, data confidentiality, and system security. The appropriate agencies and/or officials should be consulted for any projects regarding public health surveillance.[69]

Federal and state agencies are collaborating in the standardization of data definitions to ensure the value of data sets and to prevent misleading or unreliable information. Information management is used to improve services to infants and their families; to assess the quantity and timeliness of screening, evaluation, and enrollment into intervention; and to facilitate collection of demographic data on neonatal and infant hearing loss.

The JCIH endorses the concept of a limited national database to permit documentation of the demographics of neonatal hearing loss, including prevalence and etiology across the United States. The information obtained from the information-management system should assist both the primary health care professional and the state health agency in measuring quality indicators associated with program services (eg, screening, diagnosis, and intervention). The information system should provide measurement tools to determine the degree to which each process is stable and sustainable and conforms to program benchmarks. Timely and accurate monitoring of relevant quality measures is essential.

Since 1999, the CDC and the Directors of Speech and Hearing Programs in State Health and Welfare Agencies (DSHPSHWA) have collected annual aggregate EHDI program data needed to address the national EHDI goals. In 1999, a total of 22 states provided data for the DSHPSHWA survey. Participation had increased to 48 states, 1 territory, and the District of Columbia in 2003. However, many programs have been unable to respond to all the questions on the survey because of lack of a statewide comprehensive data-management and reporting system.

The Government Performance and Results Act (GPRA) of 1993 (Pub L No. 103-62) requires that federal programs establish measurable goals approved by the US Office of Management and Budget (OMB) that can be reported as part of the budgetary process, thus linking future funding decisions with performance. The HRSA has modified its reporting requirements for all grant programs. The GPRA measures that must be reported to the OMB by the MCHB annually for the EHDI program are:

- the number of infants screened for hearing loss before discharge from the hospital;
- the number of infants with confirmed hearing loss at no later than 3 months of age;
- the number of infants enrolled in a program of early intervention at no later than 6 months of age;
- the number of infants with confirmed or suspected hearing loss referred to an ongoing source of comprehensive health care (ie, medical home); and
- the number of children with nonsyndromic hearing loss who have developmentally appropriate language and communication skills at school entry.

One GPRA measure that must be reported to the OMB by the CDC annually for the EHDI program is the percentage of newborn infants with a positive screening result for hearing loss who are subsequently lost to follow-up.

EHDI programs have made tremendous gains in their ability to collect, analyze, and interpret data in the process of measuring and reporting associated program services. However, only a limited number of EHDI programs are currently able to accurately report the number of infants screened, evaluated, and enrolled in intervention, the age of time-related objectives (eg, screening by 1 month of age), and the severity or laterality of hearing loss. This is complicated by the lack of data standards and

by privacy issues within the regulations of the Family Educational Rights and Privacy Act of 1974 (Pub L No. 93-380).

Given the current lack of standardized and readily accessible sources of data, the CDC EHDI program, in collaboration with the DSHPSHWA, developed a revised survey to obtain annual EHDI data from states and territories in a consistent manner to assess progress toward meeting the national EHDI goals and the *Healthy People 2010* objectives. In October 2006, the OMB, which is responsible for reviewing all government surveys, approved the new EHDI hearing screening and follow-up survey. To facilitate this effort, the CDC EHDI Data Committee is establishing the minimum data elements and definitions needed for information systems to be used to assess progress toward the national EHDI goals.

The JCIH encourages the CDC and HRSA to continue their efforts to identify barriers and explore possible solutions with EHDI programs to ensure that children in each state who seek hearing-related services in states other than where they reside receive all recommended screening and follow-up services. EHDI systems should also be designed to promote the sharing of data regarding early hearing loss through integration and/or linkage with other child health information systems. The CDC currently provides funds to integrate the EHDI system with other state/territorial screening, tracking, and surveillance programs that identify children with special health care needs. Grantees of the MCHB are encouraged to link hearing-screening data with such child health data sets as electronic birth certificates, vital statistics, birth defects registries, metabolic or newborn dried "blood-spot" screenings, immunization registries, and others.

To promote the best use of public health resources, EHDI information systems should be evaluated periodically, and such evaluations should include recommendations for improving quality, efficiency, and usefulness. The appropriate evaluation of public health surveillance systems becomes paramount as these systems adapt to revise case definitions, address new health-related events, adopt new information technology, ensure data confidentiality, and assess system security.[69]

Currently, federal sources of systems support include Title V block grants to states for maternal and child health care services, Title XIX (Medicaid) federal and state funds for eligible children, and competitive US Department of Education personnel preparation and research grants. The NIDCD provides grants for research related to early identification and intervention for children who are deaf or hard of hearing.[137]

Universities should assume responsibility for special-track, interdisciplinary, professional education programs for early intervention for infants and children with hearing loss. Universities should also provide training in family systems, the grieving process, cultural diversity, auditory skill development, and deaf culture. There is a critical need for in-service and preservice training of professionals related to EHDI programs, which is particularly acute for audiologists and early interventionists with expertise in hearing loss. This training will require increased and sustained funding for personnel preparation.

Benchmarks and Quality Indicators

The JCIH supports the concept of regular measurements of performance and recommends routine monitoring of these measures for interprogram comparison and continuous quality improvement. Performance benchmarks represent a consensus of expert opinion in the field of newborn hearing screening and intervention. The benchmarks are the minimal requirements that should be attained by high-quality EHDI programs. Frequent measures of quality permit prompt recognition and correction of any unstable component of the EHDI process.[138]

Quality Indicators for Screening

- Percentage of all newborn infants who complete screening by 1 month of age; the recommended benchmark is more than 95% (age correction for preterm infants is acceptable).
- Percentage of all newborn infants who fail initial screening and fail any subsequent rescreening before comprehensive audiological evaluation; the recommended benchmark is less than 4%.

Quality Indicators for Confirmation of Hearing Loss

- Of infants who fail initial screening and any subsequent rescreening, the percentage who complete a comprehensive audiological evaluation by 3 months of age; the recommended benchmark is 90%.
- For families who elect amplification, the percentage of infants with confirmed bilateral hearing loss who receive amplification devices within 1 month of confirmation of hearing loss; the recommended benchmark is 95%.

Quality Indicators for Early Intervention

- For infants with confirmed hearing loss who qualify for Part C services, the percentage for whom parents have signed an IFSP by no later than 6 months of age; the recommended benchmark is 90%.
- For children with acquired or late-identified hearing loss, the percentage for whom parents have signed an IFSP within 45 days of the diagnosis; the recommended benchmark is 95%.
- The percentage of infants with confirmed hearing loss who receive the first developmental assessment with

standardized assessment protocols (not criterion reference checklists) for language, speech, and nonverbal cognitive development by no later than 12 months of age; the recommended benchmark is 90%.

CURRENT CHALLENGES, OPPORTUNITIES, AND FUTURE DIRECTIONS

Despite the tremendous progress made since 2000, there are challenges to the success of the EHDI system.

Challenges

All of the following listed challenges are considered important for the future development of successful EHDI systems:

- Too many children are lost between the failed screening and the rescreening and between the failed rescreening and the diagnostic evaluation.
- There is a shortage of professionals with skills and expertise in both pediatrics and hearing loss, including audiologists, deaf educators, speech-language pathologists, early intervention professionals, and physicians.
- There is often a lack of timely referral for diagnosis of, and intervention for, suspected hearing loss in children.
- Consistent and stable state and federal funding is needed for program sustainability.
- When compared with services provided for adults, pediatric services in all specialties are poorly reimbursed.
- Access to uniform Part C services is inadequate among states and within states.
- There is a lack of integrated state data-management and -tracking systems.
- Demographics and cultural diversity are changing rapidly.
- Funding for hearing aids, loaner programs, cochlear implants, and FM systems is needed.
- There is a lack of specialized services for children with multiple disabilities and hearing loss.
- Children may not qualify for services (state Part C guidelines) before demonstrating language delays (prevention model versus deficit model).
- Children may not qualify for assistive technology (prevention model versus deficit model).
- There is a lack of in-service education for key professionals.
- There are regulatory barriers to sharing information among providers and among states.
- No national standards exist for the calibration of OAE or ABR instrumentation, and there is a lack of uniform performance standards.

Opportunities for System Development and Research

- Establish programs to ensure the development of communication for infants and children with all degrees and types of hearing loss, allowing them access to all educational, social, and vocational opportunities throughout their life span.
- Develop improved, rapid, reliable screening technology designed to differentiate specific types of hearing loss.
- Develop and validate screening technologies for identifying minimal hearing loss.
- Develop state data-management systems with the capacity for the accurate determination of the prevalence for delayed-onset or progressive hearing loss.
- Develop state data-tracking systems to follow infants with suspected and confirmed hearing loss through individual state EHDI programs.
- Track the certification credentials of the service providers for children with confirmed hearing loss who are receiving Part C early intervention services and early childhood special education.
- Track genetic, environmental, and pharmacologic factors that contribute to hearing loss, thus allowing for tailored prevention and intervention strategies.
- Continue to refine electrophysiologic diagnostic techniques, algorithms, and equipment to enable frequency-specific threshold assessment for use with very young infants.
- Continue to refine techniques to improve the selection and fitting of appropriate amplification devices in infants and young children.
- Conduct translational research pertaining to young children with hearing loss, in particular, genetic, diagnostic, and outcomes studies.
- Initiate prospective population-based studies to determine the prevalence and natural history of auditory neural conduction disorders.
- Conduct efficacy studies to determine appropriate early intervention strategies for infants and children with all degrees and types of hearing loss.
- Conduct additional studies on the efficacy of intervention for infants and children who receive cochlear implants at younger than 2 years.
- Conduct additional studies on the efficacy of hearing-aid use in infants and children younger than 2 years.

- Conduct additional studies of the auditory development of children who have appropriate amplification devices in early life.
- Expand programs within health, social service, and education agencies associated with early intervention and Head Start programs to accommodate the needs of the increasing numbers of early-identified children.
- Adapt education systems to capitalize on the abilities of children with hearing loss who have benefited from early identification and intervention.
- Develop genetic and medical procedures that will determine more rapidly the etiology of hearing loss.
- Ensure transition from Part C (early intervention) to Part B (education) services in ways that encourage family participation and ensure minimal disruption of child and family services.
- Study the effects of parents' participation in all aspects of early intervention.
- Test the utility of a limited national data set and develop nationally accepted indicators of EHDI system performance.
- Encourage the identification and development of centers of expertise in which specialized care is provided in collaboration with local service providers.
- Obtain the perspectives of individuals who are deaf or hard of hearing in developing policies regarding medical and genetic testing and counseling for families who carry genes associated with hearing loss.[139]

CONCLUSIONS

Since the 2000 JCIH statement, tremendous and rapid progress has been made in the development of EHDI systems as a major public health initiative. The percentage of infants screened annually in the United States has increased from 38% to 95%. The collaboration at all levels of professional organizations, federal and state government, hospitals, medical homes, and families has contributed to this remarkable success. New research initiatives to develop more sophisticated screening and diagnostic technology, improved digital hearing-aid and FM technologies, speech-processing strategies in cochlear implants, and early intervention strategies continue. Major technological breakthroughs have been made in facilitating the definitive diagnosis of both genetic and nongenetic etiologies of hearing loss. In addition, outcomes studies to assess the long-term outcomes of special populations, including infants and children with mild and unilateral hearing loss, neural hearing loss, and severe or profound hearing loss managed with cochlear implants, have been providing information on the individual and societal impact and the factors that contribute to an optimized outcome. It is apparent, however, that there are still serious challenges to be overcome and system barriers to be conquered to achieve optimal EHDI systems in all states in the next 5 years. Follow-up rates remain poor in many states, and funding for amplification in children is inadequate. Funding to support outcome studies is necessary to guide intervention and to determine factors other than hearing loss that affect child development. The ultimate goal, to optimize communication, social, academic, and vocational outcomes for each child with permanent hearing loss, must remain paramount.

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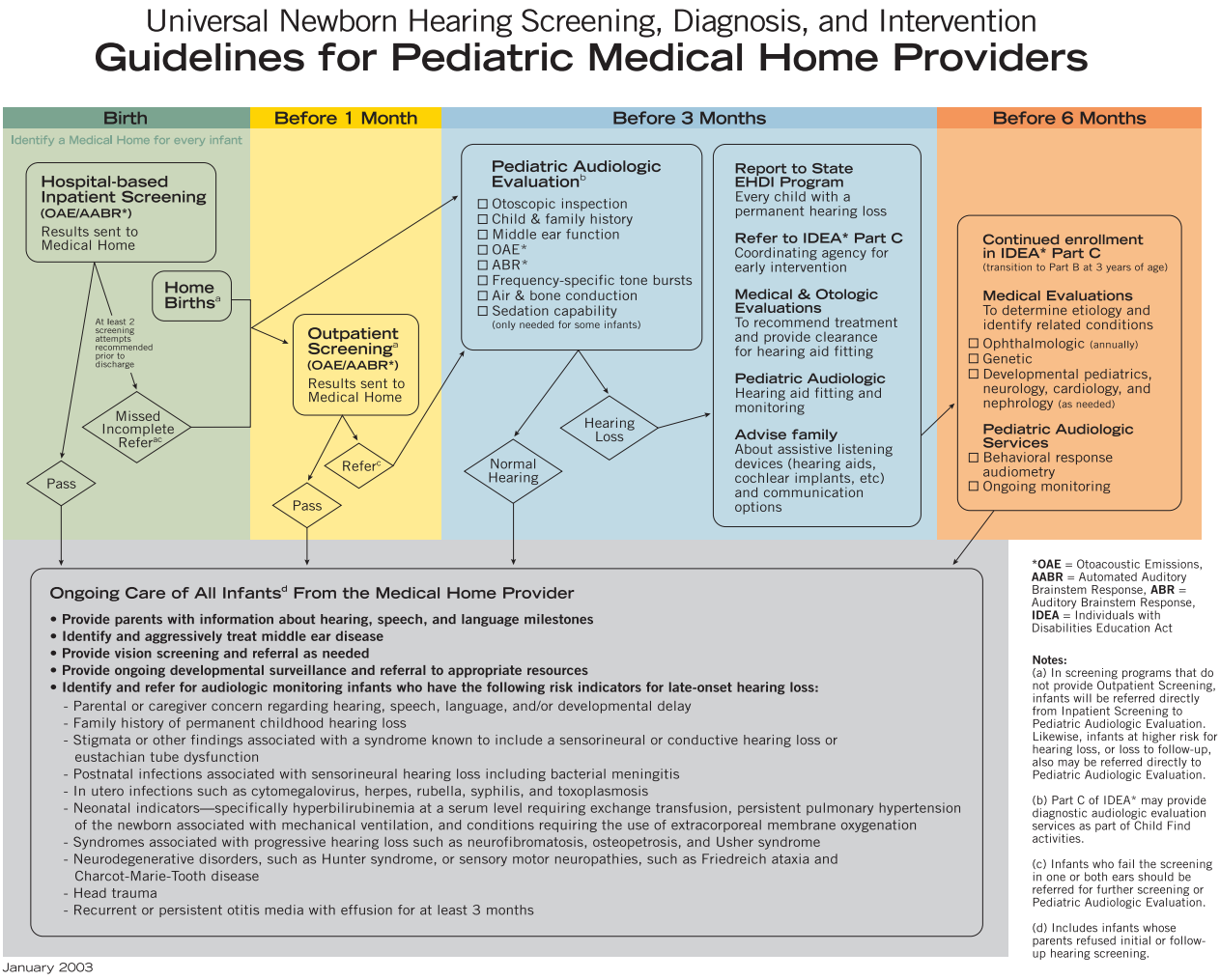
APPENDIX 1 **Algorithm for Hearing Screening. Available at: http://www.medicalhomeinfo.org/screening/Screen%20Materials/Algorithm.pdf**



APPENDIX 2: RISK INDICATORS ASSOCIATED WITH PERMANENT CONGENITAL, DELAYED-ONSET, OR PROGRESSIVE HEARING LOSS IN CHILDHOOD

Risk indicators that are marked with a "§" are of greater concern for delayed-onset hearing loss.

1. Caregiver concern§ regarding hearing, speech, language, or developmental delay.[62]
2. Family history§ of permanent childhood hearing loss.[24,140]
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: ECMO,§ assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion.[64,131]
4. In utero infections, such as CMV,§ herpes, rubella, syphilis, and toxoplasmosis.[64–67,125,126]
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.[24]
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.[24]
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,§ such as neurofibromatosis, osteopetrosis, and Usher syndrome[131]; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson.[72]
8. Neurodegenerative disorders,§ such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.[131]
9. Culture-positive postnatal infections associated with sensorineural hearing loss,§ including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.[130,131,141]
10. Head trauma, especially basal skull/temporal bone fracture§ that requires hospitalization.[127–129]
11. Chemotherapy.§[132]

Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs

Joint Committee on Infant Hearing

Pediatrics 2007;120;898-921
DOI: 10.1542/peds.2007-2333

Child's Name: _____

Child's Date of Birth: _____/_____/_____

A Family's Checklist – Infant Hearing

Normal Milestones

Many babies meet normal milestones even if they have a hearing loss in one or both ears. You should only use these milestones to monitor your baby's hearing and development if your baby has PASSED the hearing screening or been evaluated by a Pediatric Audiologist…

Months of Age

2	Quiets when hearing a familiar voice. Makes vowel sounds like ahh, ohh
4	Looks for sounds with his eyes. Uses sounds such as squeals, whimpers, chuckles
6	Turns head toward sound. Babbles ba-ba, ma-ma, da-da
9	Imitates speech sounds of others. Understands no-no or bye-bye. Turns head toward soft sounds.
12	Correctly uses ma-ma or da-da.

Before 1 Month

☐ **Hospital Inpatient Screen**

Place: _____

Screen Date: ____/____/____

Results:

Left Ear		Right Ear
☐	**Pass**	☐
☐	**Refer**	☐

☐ **Repeat Screen**

Place: _____

Screen Date: ____/____/____

Results:

Left Ear		Right Ear
☐	**Pass**	☐
☐	**Refer**	☐

If your baby does not pass the screening in one or both ears, talk to your doctor about seeing a Pediatric Audiologist as soon as possible.

If your baby passes, testing is done. Watch for normal milestones.

Be sure your doctor gets the results.

www.AZNewborn.com

Before 3 Months

☐ **Evaluation by Pediatric Audiologist. Be sure your doctor gets the results.**

Place: _____

Date: ____/____/____

Results:

Right Ear		Left Ear
☐	**Normal**	☐
☐	**Hearing Loss**	☐

If your baby has a HEARING LOSS, the next steps are:

☐ **Evaluation by an ENT (Ear, Nose, and Throat) doctor**

Place: _____

Date: ____/____/____

☐ **Hearing aid fitting (if appropriate) of loaner or permanent hearing aids by a Pediatric Audiologist.**

☐ **Contact Hands & Voices for family support: by phone at 866-685-1050 or www.AZHV.org**

Before 6 Months

☐ **Enroll in Early Intervention program, if hearing loss in both ears**

Program: _____

Date: ____/____/____

☐ **Learn about communication Options**

☐ **Learn about cochlear implants, if applicable**

☐ **Regular visits to a Pediatric Audiologist**

Evaluations:

☐ **Ophthalmologist (eye doctor)**

Place: _____

Date: ___/____/____

☐ **Genetic Specialist**

Place: _____

Date: ___/____/____

You may need a referral from your doctor to see these specialists.

TAKE THIS TO YOUR BABY'S DOCTOR

Lista de Comprobación Familiar – Audición Infantil

Nombre del Niño: _____

Fecha de Nacimiento: _____/_____/_____

Etapas Normales

Muchos bebés alcanzan las etapas normales aunque sean sordos de uno o ambos oídos. Sólo debe usar estas etapas para vigilar el desarrollo de la audición de su niño si este ha PASADO la prueba de audición o ha sido revisado por un Audiólogo Pediátrico…

Meses de Edad

2 Se calla cuando oye una voz conocida. Hace sonidos de vocales como a-a-ah, o-o-o

4 Busca los sonidos con sus ojos. Usa sonidos como chillidos, quejidos, risitas

6 Voltea hacia el sonido. Balbuce ba-ba, ma-ma, da-da

9 Imita a los demás. Entiende no-no o bye-bye. Voltea la cabeza hacia los sonidos suaves.

12 Usa correctamente ma-ma o da-da

Antes de 1 Mes

☐ **Prueba en el Hospital**

Lugar: _____

Fecha de la Prueba: ___/___/___

Resultados:

Oído Izq.		Oído Derecho
☐	Pasó	☐
☐	Recomendación	☐

☐ **Repetición de la Prueba**

Lugar: _____

Fecha de la Prueba: ___/___/___

Resultados:

Oído Izq.		Oído Derecho
☐	Pasó	☐
☐	Recomendación	☐

Si su bebé no pasa la evaluación en uno o en ambos oídos, hable con su doctor para que lo mande con un Audiólogo Pediátrico lo más pronto posible.

Si su bebé pasa, se hace la prueba y usted debe esperar etapas normales.

Asegúrese que su doctor reciba los resultados.

www.AZNewborn.com

Antes de 3 Meses

☐ **Evaluación del Audiólogo Pediátrico. Asegúrese que su doctor reciba el resultado.**

Lugar: _____

Fecha: _____/_____/_____

Resultados:

Oído Derecho		Oído Izq.
☐	Normal	☐
☐	Sordera	☐

Si su bebé tiene SORDERA, los pasos a seguir son:

☐ **Prueba de un Otorrinolaringólogo (Oído, Nariz y Garganta)**

Lugar: _____

Fecha: _____/_____/_____

☐ **Medida del audífono (si es apropiado) prestado o audífonos permanentes por un Audiólogo Pediátrico.**

☐ **Contacte a Hands & Voices para apoyo familiar; por teléfono al 866-685-1050 o en www.AZHV.org**

Antes de 6 Meses

☐ **Regístrelo en el programa de Intervención Temprana, si es sordo de los dos oídos**

Programa: _____

Fecha: ____/_____/_____

☐ **Aprenda sobre opciones de comunicación**

☐ **Aprenda sobre implantes cocleares, si es aplicable**

☐ **Visitas regulares a un Audiólogo Pediátrico**

Evaluaciones:

☐ **Oftalmólogo (oculista)**

Lugar: _____

Fecha: ___/____/____

☐ **Especialista en Genética**

Lugar: _____

Fecha: ___/____/____

Quizá necesite recomendación de su doctor para ver a estos especialistas

LLEVE ESTO AL DOCTOR DE SU BEBÉ

Family Checklist for Babies at High Risk for Hearing Loss

Infants who are in the NICU for 5 days or more may be at a higher risk for hearing loss. Other risk factors are listed at the bottom of the page.

Child's Name: _____

Child's Date of Birth: _____/_____/_____

HOSPITAL

Screen Before 1 Month of Age

While in the hospital, your baby should have had an ABR hearing screen. It is important that you talk to your baby's doctor right away about the information below.

Place: _____

Screen Date: ____/____/_____

Results:

Right Ear		Left Ear
☐	Fail/Refer	☐
☐	Pass	☐

If Your Baby:

Referred / Failed:

See a pediatric audiologist for diagnostic testing right away.**

Passed:

There is still a chance of developing hearing loss. See a pediatric audiologist for further testing at 9 months of age (corrected age).**

****See list of phone numbers attached**

AUDIOLOGIST

Diagnose Before 3 Months of Age

It is important that your baby have diagnostic hearing testing with a qualified pediatric audiologist that can perform the following 3 tests:

- √ **Auditory Brainstem Response (ABR) - Frequency Specific with Bone Conduction**
- √ **Otoacoustic Emissions (OAEs)**
- √ **Tympanometry - Age Appropriate**

Place: _____

Date: ____/____/____ **Corrected Age:** _____

Results:

Right Ear:	Left Ear:
☐ Within Normal Limits	☐ Within Normal Limits
☐ Hearing Loss	☐ Hearing Loss

If your baby has a hearing loss the next steps are:

- ☐ **See an Ear Nose and Throat (ENT) Doctor**
- ☐ **Contact the Arizona Early Intervention Parent Outreach Program**
- ☐ **Contact Hands and Voices at www.AZHV.org or 1-866-685-1050**

EARLY INTERVENTION

Enroll in the Arizona Early Intervention Program Before 6 Months of Age

Program: _____

Date: ____/____/_____

- ☐ **Learn about communication options**
- ☐ **Learn about hearing aids and/or cochlear implants, if recommended**
- ☐ **Regular visits to a Pediatric Audiologist**

Evaluations:
(May need order from your doctor and insurance approval first)

Ophthalmologist (eye doctor)

Place: _____

Date: ____/____/_____

Genetic Specialist

Place: _____

Date: ____/____/_____

☐ **Other Medical Specialists, as needed**

Babies with the following "Risk Indicators for Late Onset and Progressive Hearing Loss" should have hearing checked regularly

- *ECMO
- *Hyperbilirubinemia requiring exchange transfusion
- *Family history of permanent childhood hearing loss
- *Exposure to ototoxic medication
- *Assisted Ventilation
- *Neurodegenerative disorders
- *CMV
- *Caregiver Concern
- *Syndromes associated with progressive or late-onset hearing loss
- *Culture positive postnatal infections associated with sensorineural hearing loss

TAKE THIS TO YOUR NEXT APPOINTMENT WITH YOUR BABY'S DOCTOR

Rev 1/10

Lista de cotejo para familias de bebés con gran peligro de pérdida auditiva

Los infantes que permanezcan en cuidados intensivos por 5 días o más pudieran peligrar más de padecer merma o pérdida auditiva . Se relatan otros factores de peligro al calce de la página.

Nombre de niño/a: _____

Fecha de nacimiento: ____/____/_____

HOSPITAL

Diagnóstico antes de 1 mes de edad

A su niño/a le debieron hacer el diagnóstico de potenciales provocados auditivos del tallo cerebral (PPATC, o *ABR* en inglés) en el hospital. Es importante que hable de inmediato con su doctor(a) sobre la información siguiente.

Lugar: _____

Fecha de diagnóstico: ____/____/_____

Resultado:

Oído Derecho	Oído Izquierdo
☐ Falló/Atiéndase	☐
☐ Aprobó	☐

Si su bebé:

Falló/ Atiéndase:

Atiéndase de inmediato con un(a) audiólogo/a pediátrico/a para un estudio diagnóstico.**

Aprobó:

Todavía es posible que padezca merma o pérdida auditiva. Atiéndase con un(a) audiólogo/a pediátrico/a para hacerle más estudios a los 9 meses de edad (edad corregida).**

****Vea la lista adjunta de teléfonos**

AUDIÓLOGO/A

Diagnostique antes de los 3 meses de edad

Resulta importa que su bebé se haga un diagnóstico de la audición con un(a) audiólogo/a pediátrico/a calificado/a que pueda efectuar las 3 pruebas siguientes:

- √ **Potenciales provocados auditivos del tallo cerebral (PPATC, o *ABR* en inglés) – de frecuencia precisa con osteoconducción**
- √ **Emisiones otoacústicas (EMOA, u *OAEs* en inglés)**
- √ **Timpanometría – apropiadas para la edad**

Lugar: _____

Fecha: ____/____/____ **Edad corregida:** _____

Resultado:

Oído Derecho:	Oído Izquierdo:
☐ Dentro de lo normal	☐ Dentro de lo normal
☐ Merma o pérdida auditiva	☐ Merma o pérdida auditiva

Si su bebé padeciera merma o pérdida auditiva, las medidas siguientes serían:

- ☐ **Atenderse con un(a) doctor(a) de oídos, nariz y garganta (*ENT*, en inglés)**
- ☐ **Comuníquese con el programa de Acercamiento a los Padres Para la Intervención Temprana en Arizona (*Arizona Early Intervention Parent Outreach Program)***
- ☐ **Comuníquese con *Hands and Voices* por www.AZHV.org o al 1-866-685-1050**

INTERVENCIÓN TEMPRANA

Inscríbase en el Programa de Intervención Temprana de Arizona antes de los 6 meses de edad

Programa: _____

Fecha: ____/____/_____

- ☐ **Entérese de las opciones para comunicarse**
- ☐ **Entérese de los dispositivos auditivos y/o de los implantes de cóclea, si los recomendaran**
- ☐ **Consulte con regularidad a su audiólogo/a pediátrico**

Evaluaciones:
(Pudiera necesitar antes una orden médica y la aprobación del seguro)

Oftalmólogo/a (oculista)

Lugar: _____

Fecha: ____/____/_____

Especialista en genética

Lugar: _____

Fecha: ____/____/_____

☐ **Otros especialistas médicos que haga falta**

Los bebés que padezcan los siguientes "indicadores de peligro de merma auditiva tardía o paulatina" deben examinarse la audición con regularidad

- ***oxigenación con membrana extracorpórea (ECMO)**
- ***hiperbilirubinemia que requiera exsanguinotransfusión**
- ***antecedentes familiares de pérdida auditiva permanente en la niñez**
- ***exponerse a medicamentos ototóxicos**
- ***respirar con ventilador**
- ***trastornos neurodegenerativos**
- ***citomegalovirus**
- ***preocupación de quien cuide a su cría**
- ***síndromes asociados a la merma auditiva paulatina o de comienzo tardío**
- ***infecciones posnatales de resultado positivo en el cultivo que se asocien a la pérdida auditiva neurosensorial**

LLEVE ESTO A LA PRÓXIMA CITA CON EL O LA DOCTOR(A) DE SU BEBÉ

Rev 1/10

Newborn Hearing Screening Training Curriculum Scripts

Informing Parents of the Screen:

Hi! Congratulations on the birth of your baby. You have received information that we provide hearing screening to all babies born. We are going to screen your baby now.

Informing Parents of the Screen (Spanish):

¡Hola! Felicitaciones por el nacimiento de su bebé. Usted recibió información sobre el tamizaje auditivo que le hacemos a todos los recién nacidos. Ahora vamos a hacerle el tamizaje a su bebé.

Passing:

Congratulations on the birth of your baby. We just completed the hearing screen; the results are a pass. Here is a brochure that talks about development of speech and language. It is always important to monitor the progress of your baby's development, especially their speech and language because your baby's hearing can change any time. If you are ever worried that your baby can't hear, talk to your baby's doctor right away and ask for a referral to an audiologist that is skilled at testing infants and young children.

Passing (Spanish): Pasó:

Felicitaciones por el nacimiento de su bebé. Acabamos de finalizar el tamizaje auditiva de su bebé y él/ella la pasó. Este es un folleto que trata sobre el desarrollo del habla y del lenguaje. Es importante observar el desarrollo de su bebé especialmente de su habla y lenguaje ya que la audición de su bebé puede cambiar en cualquier momento. Si usted está preocupado de que su bebé no pueda oír, hable con el médico pediatra inmediatamente y pídale que lo envíe a donde un audiólogo especializado en hacer pruebas a bebés y niños pequeños.

Not Passing:

Congratulations on the birth of your baby. We just finished screening your baby's hearing. Your baby did not pass the screen today. This does not necessarily mean that your baby has a permanent hearing loss, but without additional testing we can't be sure. The screening results will be provided to your baby's doctor. Please be sure you make or keep (depending on your hospital's protocol) the appointment for further hearing testing.

Not Passing (Spanish): No Pasó:

Felicitaciones por el nacimiento de su bebé. Los resultados del tamizaje auditivo que le hicimos hoy a su bebé indican que él/ella no lo pasó. Esto no necesariamente significa que su bebé tenga una pérdida auditiva permanente, pero sin hacer pruebas adicionales no podemos estar seguros. Los resultados del tamizaje le serán enviados al médico de su bebé. Asegúrese de hacer una cita para hacer más exámenes auditivos o acudir a esta (dependiendo del protocolo de su hospital).

Inconclusive:

Although we attempt to provide newborn hearing screening to all babies born at our hospital, we were unable to complete the screening on your baby. It is important that your baby be screened as soon as possible. Let's schedule a time for the screening to be completed within the next 2 weeks.

Inconclusive (Spanish): No Concluyente:

Aunque tratamos de hacerle un tamizaje auditivo a todos los recién nacidos en nuestro hospital, no pudimos completar el tamizaje de su bebé. Es importante hacerlo lo más pronto posible. Hagamos una cita para terminar de hacerle la prueba durante las dos semanas entrantes.

Not Passing Outpatient Rescreen:

Your baby did not pass the second screen. The screening does not tell us whether your baby has a hearing loss; it just tells us that further testing should be done as soon as possible. The next step is to get a diagnostic ABR as soon as possible. This should be discussed immediately with your baby's doctor who may need to help you with obtaining a referral to a pediatric audiologist.

Not Passing Outpatient (Spanish): No Pasó El Segundo Tamizaje Auditivo:

Su bebé no pasó el segundo tamizaje auditivo. Esto no significa que su bebé tiene una pérdida auditiva; solamente nos indica que se deben hacer más pruebas lo más pronto posible. El siguiente paso es realizar una prueba de potenciales evocados auditivos del tronco cerebral (conocida por sus siglas en inglés ABR). Hable de manera inmediata con el médico de su bebé quien puede ayudarle a conseguir una cita con un audiólogo pediatra.

High Risk: Passing:

Congratulations on the birth of your baby. We screened your baby's hearing and the results are a pass. However, because your baby has had some medical problems at birth, there is a chance that your baby can develop a hearing loss after you leave the hospital. Your baby's hearing is critical for normal speech and language development, so it is important that you speak to your baby's doctor who can help you in knowing when your baby should have further tests with a pediatric audiologist and can also help you to monitor for normal speech and language development.

High Risk: Passing (Spanish)

Felicitaciones por el nacimiento de su bebé. Su bebé pasó el tamizaje auditivo que le realizamos. Sin embargo y debido a que tuvo algunas complicaciones médicas durante su nacimiento , existe la posibilidad de que desarrolle una pérdida auditiva después de que sea dado de alta del hospital. La audición es importante para el desarrollo normal del habla y lenguaje de su bebé. Es importante que hable con el médico de su bebé quien le puede indicar cuando debe ser visto por un audiólogo pediatra y también le puede ayudar a hacer el seguimiento del desarrollo del habla y lenguaje de su bebé.

High Risk: Not Passing

Congratulations on the birth of your baby. Your baby received a hearing screen and the results show that your baby *did not pass*. There can be simple reasons for this, but without further testing with a pediatric audiologist, I cannot tell you what your baby hears. Because your baby has had some medical problems at birth, your baby is at a greater risk for a hearing loss. Please discuss these results with your baby's doctor and ask them to help you schedule diagnostic tests with a pediatric audiologist as soon as possible. Finding out about hearing issues as early as possible will help to ensure that your baby has the best chance to develop normal speech and language.

High Risk: Not Passing (Spanish)

Felicitaciones por el nacimiento de su bebé. Su bebé no pasó el tamizaje auditivo que le realizamos. Las razones no son necesariamente complicadas pero sin tener resultados de exámenes realizados por un audiólogo pediatra no puedo informarle cual es la capacidad auditiva de su bebé. Debido a que su bebé tuvo algunas complicaciones médicas durante su nacimiento tiene una mayor posibilidad de desarrollar una pérdida auditiva. Es importante que discuta estos resultados con el médico de su bebé, él le puede ayudar a hacer una cita con un audiólogo pediatra lo más pronto posible. El diagnóstico de problemas auditivos lo más temprano posible ayudará a que su bebé tenga una mejor oportunidad para desarrollar un habla y lenguaje normal.

NCHAM
National Center for Hearing Assessment and Management
Utah State University™
2800 Old Main Hill
Logan, UT 84321-9981

Universal Newborn Hearing Screening

Arizona EHDI
Early Hearing Detection and Intervention Program

It's Never Too Early to Test Your Baby's Hearing

NBS-EHDI01 (8/09)

The most common problem in newborns is...

Hearing loss. The first three years are the most important for learning speech and language. Babies who can't hear well may have problems learning to talk. If hearing loss is identified early, your child will have the best chance to learn to speak and do well in school.

About Hearing Screening...

Hearing screenings are performed in the hospital just hours after your baby is born. They can be completed in as little as 10-15 minutes if the baby is quiet or asleep. A newborn's hearing can be tested on special equipment using one or both quick methods:

Otoacoustic Emissions (OAE)
uses a soft ear probe to measure the ear's "echo".

Auditory Brainstem Response (ABR)
uses sensors that measure your baby's brainwaves to determine if sounds are heard normally.

If your baby PASSES the screening...

It means your baby's hearing tested within the normal range during the screening. However, hearing loss can develop after you leave the hospital. Therefore, checking your baby's speech development over the 1st year is important.

Certain conditions increase the possiblity of a hearing loss developing later:

- Family history of permanent hearing loss from childhood
- Infections after birth such as bacterial Meningitis
- Head trauma
- Ear infections with fluid lasting at least three months
- Infections during pregnancy such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis
- Some syndromes associated with progessive hearing loss
- Neonatal indicators such as persistent pulmonary hypertension
- Unusual appearance of baby's head, face or ears

If your baby DOES NOT PASS the screening...

It does not mean that he or she definitely has a hearing loss. It may be because of fluid in the baby's ear or because the baby was too active during the test. There is also the possiblity of a hearing loss. Therefore, it is important to return to the hospital for another test within 2 to 4 weeks.

Many babies with hearing loss will startle to loud sounds. The only way to know for sure if your baby's hearing is normal is to have the testing done with special equipment.

The following is a list of things a child with normal hearing should be able to do.

Months of age	Average Developmental Milestones
2	· Quiet when hearing a familiar voice · Make vowel sounds like ahh, ohh
4	· Look for sounds with his eyes · Use sounds such as squeals, whimpers, chuckles
6	· Turn head toward sound · Babble ba-ba, ma-ma, da-da
9	· Imitate speech sounds of others · Understand no-no or bye-bye · Turn head toward soft sounds
12	· Correctly use ma-ma or da-da · Respond to singing or music

A child with a mild hearing loss may also meet these milestones. It is important to talk to your baby's doctor with any concerns.

If you have any questions or want more information, please call your doctor or the hospital nursery where your baby was born at this # _____

Before you leave the hospital, be sure you know the results of your baby's hearing screening and what to do next if your baby did not pass the screening.

TEST RESULTS

Right EAR:	Pass	Refer
Left EAR:	Pass	Refer

Rescreen Appointment: Date _____
Time _____

For more information about the Newborn Hearing Screening Program or for alternative formats, please contact:

Arizona Department of Health Services
Office of Newborn Screening
State Laboratory
250 N 17th Ave
Phoenix, AZ 85007
Local: (602) 364-1409
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Arizona Department of Health Services

Exámenes Auditivos Universales en el Recién Nacido

EHDI de Arizona
Programa de Detección
Auditiva Temprana e Intervención

Nunca es
Demasiado Temprano
para el Examen
Auditivo de su Bebé

NBS-EHDI01 (8/09)

El problema más común en los recién nacidos es...

La pérdida de oir. Los primeros tres años son los más importantes para aprender a hablar el lenguaje. Los bebés que no oyen muy bien tienen problemas para aprender a hablar. Si la pérdida de oir se identifica temprano, su niño/a va a tener mejor oportunidad para aprender a hablar bien y tener éxito en la escuela.

Sobre la prueba de escucha/audición...
Las pruebas de escucha/audición se les hacer a los niños en el hospital después de unas cuantas horas de nacer. La prueba toma solamente unos 10 a 15 minutos si el bebé está tranquilo o dormido. La escucha de un recién nacido se puede probar con aparatos especiales usando uno o los dos métodos rápidos:

Emisión Otoacústico (OAE) usa una sonda suave para medir el "eco" del oído.

Respuesta Auditiva del Vástago de Cerebro (ABR) utiliza sensores que miden las ondas cerebrales de su bebé para determinarse si los sonidos se oyen normalmente.

Si su bebé pasa el examen...

Quiere decir que la prueba de audición de su bebé era normal en este momento. Sin embargo, la pérdida de oir puede ocurrir después de salir del hospital. Por lo tanto es muy importante revisar el desarrollo del habla de su bebé durante el primer año.

Ciertas condiciones aumentan la posibilidad de desarollar una pérdida auditiva más adelante:

- Historia familiar de pérdida permanente de oir desde el niñez.
- Infecciones después del nacimiento como meningitis bacteriana
- Trauma del cerebro
- Infecciones del oído con flúido que permanece por lo menos tres meses.
- Infecciones durante el embarazo como cytomegalovirus, herpes, sarampión, sífilis y toxoplasmosis.
- Algunos síndromes asociados con pérdida progresiva de oir.
- Indicadores neonatales como hipertensión pulmonar persistente.
- Una aparencia de la cabeza, cara, u óidos del bebé que es fuera de lo común.

Si su bebé no pasa el examen...

No significa que él o ella tiene definitivamente una pérdida de oir. Puede ser por causa de flúido que tenía el bebé en el oído o porque el bebé era demasiado activo durante la prueba. Hay también la posibilidad de una pérdida de oir. Por lo tanto es importante volver al hospital para otro examen dentro de 2 a 4 semanas.

Muchos bebés con pérdida de oir se asustan con ruidos fuertes. Para estar seguro, la única manera de saber si el oir de su bebé es normal es hacer un examen con aparatos especiales.

La siguiente lista es de cosas que un niño que oye normal debe poder hacer.

Edad de meses	Las piedras milleras del desarrollo mediano
2	• Silencio al oir voces familiares • Hace sonidos vocales como ahh, ohh
4	• Busca los ruidos con sus ojos • Hace ruidos como chillidos, lloriqueos, risitas
6	• Voltea la cabeza hacia al ruido • Barbotea ba-ba, ma-má o pa-pá
9	• Imitar sonidos del habla de otros • Comprende no-no y ro-ro • Voltea la cabeza hacia sonidos suaves
12	• Usa correctamente mamá o papá • Responde a cantos o música

Un niño con pérdida leve auditiva también puede lograr estas piedras miliarias. Es importante hablar con el doctor de su bebé con cualquier preocupación.

Si usted tiene preguntas o desea más información, favor de llamar a su médico o a la guardería del hospital donde nació su bebé al # _____

Para saber que hacer si su bebé no pasa el examen, antes de salir del hospital, esté seguro de que sepa los resultados de las pruebas de oir de su bebé.

Resultados del Examen

Oído derecho: paso referir

Oído izquierdo: paso referir

Cita para el re-examen: Fecha _____

Hora _____

Para más información sobre el Programa de Detección para los Recién Nacidos u otros formatos, favor de comunicarse con:

Arizona Departemento de Salud
Oficina Análisis de Recién Nacidos
Estado de Laboratorio
250 N 17th Ave.
Phoenix, AZ 85007
Teléfono: (602) 364-1409
Teléfono: (800) 548-8381
Fax (602) 364-1495

Arizona Department of Health Services

Frequently Asked Questions Parent's May Ask (English and Spanish)

Why screen my baby's hearing?

Hearing loss is one of the most common conditions present at birth. It is easy to miss hearing loss because you usually can't see anything different. Without screening, hearing loss is often not detected until the baby is 2 years old and not talking. Early identification and intervention means that your baby won't fall behind other children in speech and language development.

¿Por qué hacerle una prueba auditiva a mi hijo?

La pérdida auditiva es una de las condiciones más comunes que se presentan en los recién nacidos. Es fácil no percatarse de su existencia porque uno no puede ver nada diferente en el bebé. Sin la prueba auditiva, es frecuente que la pérdida auditiva no se detecte hasta que el niño tiene 2 años y no habla. La identificación e intervención temprana hacen que su bebé no tenga un retrazo en su habla y desarrollo del lenguaje.

How do you check my baby's hearing?

OAE: Soft sounds are made into the baby's ear. If the ear is working normally, it will send back sounds that the computer can pick up and analyze. Your baby doesn't have to do anything other than be quiet.

ABR: Soft sounds are made into the baby's ear and electrodes or little sensors pick up the brain's response to the sounds.

¿Cómo le hace la prueba auditiva a mi hijo?

OAE: Por medio de una sonda se introducen sonidos suaves en el oído del bebé. Si el oído funciona normalmente, éste producirá sonidos que son detectados y analizados por la computadora. Su bebé no tiene que hacer nada solamente permanecer callado.

ABR: Por medio de una sonda se introducen sonidos suaves en el oído de su bebé. Electrodos localizados en la frente y en los lóbulos de las orejas detectan la respuesta del cerebro a estos sonidos.

What does Pass or Refer mean?

Pass means that your baby's ears are working normally today. However, some babies develop hearing loss later so if you are concerned, you should always talk to your baby's medical provider about getting a hearing test.

Refer means that your baby did not pass the hearing screening and needs additional testing.

¿Qué significa cuando mi bebé pasa/no pasa la prueba?

Si su bebé pasa la prueba, esto significa que los oídos de su bebé funcionan bien. Sin embargo, algunos bebés pueden desarrollar una pérdida auditiva después de la primera prueba. Si usted está preocupado debe hablar con la persona que provee los servicios de salud a su hijo sobre la posibilidad de hacerle otra prueba auditiva. Si su bebé no pasa la prueba esto significa que necesita exámenes adicionales.

What happens if my baby Refers?

If your baby refers a second time, it is very important that you make an appointment with a pediatric audiologist as soon as possible to have a complete hearing test called an Auditory Brainstem Response test or an ABR.

¿Qué pasa si mi bebé no pasa la prueba auditiva por segunda vez?

Si su bebé no pasa la prueba por segunda vez, es importante que haga una cita con un audiólogo pediatra lo más pronto posible para que realicen un examen que se llama ABR (por sus siglas en ingles).

How long does the hearing screen take?

Usually it takes 10 to 15 minutes depending on how quiet your baby is during the screening.

¿Cuánto tiempo toma hacer el examen?

Usualmente de 10 a 15 minutos dependiendo de que tan callado esté el niño durante la prueba.

Will hearing screening hurt my baby?

No. Most babies sleep through the screen.

¿Le dolerá a mi bebé?

No. La mayoría de los bebés duermen durante la prueba.

Where is the hearing screening done?

Your baby's hearing can be screened at this hospital, as part of the newborn hearing screening program.

¿Dónde se realiza la prueba?

La prueba auditiva se puede realizar en este hospital, como parte del programa de pruebas auditivas de recién nacidos.

What can be done if hearing loss is detected?

Hearing loss cannot be determined by screening. Screening tells us if further testing by a pediatric audiologist is needed. If an audiologist finds that your baby has a hearing loss he or she will talk with you about what happens next.

¿Cuál es el siguiente paso si se sospecha la existencia de una pérdida auditiva?

Una pérdida auditiva no puede ser confirmada por la prueba auditiva, esta indica que un audiólogo pediatra necesita realizar más pruebas. Si un audiólogo diagnostica una pérdida auditiva, él o ella le dirán cual es el siguiente paso a seguir.

What if I choose not to allow the hearing screen?

You will be asked to sign a refusal form and your baby's doctor will be advised of your decision. We recommend that you think about the screening. Please ask questions about your concerns. Finding a hearing loss as early as possible is critical in order for children to develop normal speech and language.

¿Qué pasa si tomo la decisión de no permitir que se le haga a mi bebé la prueba auditiva?

Se le pedirá que firme un documento y se le comunicará al doctor de su bebé su decisión. Le recomendamos que piense su decisión. Por favor haga preguntas sobre sus preocupaciones. El diagnóstico de una pérdida auditiva los más temprano posibles es importante para que los niños desarrollen un habla y lenguaje normal.

NCHAM
National Center for Hearing Assessment and Management
Utah State University™
2800 Old Main Hill
Logan, UT 84321-9981

How Does Your Child Hear and Talk?

Here is a checklist that you can follow to check your child's hearing and understanding development. You should talk to your child's doctor about anything on the list that your child is not doing and you have checked "No".

Birth to 3 Months	**Yes**	**No**
Reacts to loud sounds	___	___
Quiets or smiles when spoken to	___	___
Seems to recognize your voice and quiets if crying	___	___
Increases or decreases sucking behavior in response to sound	___	___
Makes noise when talked to	___	___

4 – 6 Months	**Yes**	**No**
Moves eyes in direction of sounds	___	___
Responds to changes in tone of your voice	___	___
Notices toys that make sounds	___	___
Pays attention to music	___	___

7 Months to 1 Year	**Yes**	**No**
Enjoys games like peek-a-boo and pat-a-cake	___	___
Turns and looks in direction of sounds	___	___
Listens when spoken to	___	___
Recognizes words for common items like "cup", "shoe", or "juice"	___	___
Begins to respond to requests (e.g. "come here" or "want more"?)	___	___

Parents: This is simply a guide to help you check your baby's development at different ages and stages. It should not be used to replace a hearing test. If you ever have concerns about your child's hearing, contact your baby's doctor immediately.

A Rallying Cry: We Need All Your HANDS & VOICES

If you or your child/student talk *or* sign... if you talk *and* sign, or cue, or care, we need you. If you are directly, indirectly, or potentially involved with children who are deaf or hard of hearing and their families, we need you. We need to be united by our hands and our voices. We are parents, professionals, adult mentors, consumers, and others who are in so many ways connected to a cause and to each other through the community of the Deaf and Hard of Hearing. We are Hands & Voices.

Our philosophy of unbiased support makes Hands & Voices unique among Deaf/HH organizations. There are many wonderful organizations that provide support and information about methods and modes of communication, but at Hands & Voices, we rally around the need to improve educational outcomes for our kids, and to enhance their quality of life.

Our organization is parent-driven, but highly collaborative with professionals who are represented on our advisory board, and make up approximately one third of our membership. We work at both the local and national levels. Our state chapters provide local and regional support consistent with the Hands & Voices non-biased philosophy and mission. In addition to family support, we strive to be meaningfully involved at the systems development level.

Whether we're at work locally or nationally, Hands & Voices continues to be driven by the need to improve the educational outcomes and quality of life for children who are deaf or hard of hearing.

Be a part of this dynamic movement….become a Hands & Voices member today!

HANDS& VOICES

Arizona Hands & Voices

PO Box 50423
Phoenix AZ, 85076

ph #: 1-866-685-1050

Phoenix Area and Northern AZ please contact:
Helen Cartwright
hcartwright@azhv.org

Tucson and Southern AZ please contact:
Carmen Haber
chaber@azhv.org

www.azhv.org
register@azhv.org

National Information:

HANDS& VOICES

Hands & Voices

P0 Box 371926
Denver, CO 80237

Phone (303) 300-9763
Toll free (866) 422-0422

Email: parentadvocate@handsandvoices.org
Website: www.handsandvoices.org

Hands & Voices is a non profit 501(c)(3) organization

HANDS & VOICES

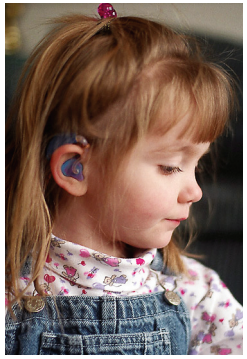
What Works for your Child is What Makes the Choice Right

Non-biased support for families of children who are deaf and hard of hearing

www.handsandvoices.org

Parent Support and Advocacy for Children who are Deaf and Hard of Hearing

Our Mission:



Families for Hands and Voices is dedicated to supporting families with children who are Deaf or Hard of Hearing without a bias around communication modes or methodology. We're a parent-driven, non-profit organization providing families with the resources, networks, and information they need to improve communication access and educational outcomes for their children. Our outreach activities, parent/professional collaboration, and advocacy efforts are focused on enabling Deaf and Hard-of-Hearing children to reach their highest potential.

What People say about Hands & Voices:

"Hands & Voices supported my personal decision about my son and how he would learn to communicate with the world as a deaf Person. Hands & Voices did not judge, but supported my choice."

Shirley Swope,
parent and advocate, Colorado

"*Hands & Voices is what our state has been waiting for. I had several requests for specific information which sent me scrambling for some appropriate resources and information. Hands & Voices was where I found it."*

Lorna Irwin,
parent and consultant, Idaho

"For too long we have debated what communication mode is best, rather than work to provide the variety of options necessary to meet the diverse needs of deaf/hh children. The broad vision of Hands & Voices comes at exactly the right time."

Lawrence Siegel,
Natl. Deaf Education Project

Hands & Voices Areas of Focus Include:

- Deaf Ed Reform - Educational Excellence
- "Medical Home" Initiative
- Communication Access for Children who are Deaf and Hard of Hearing
- Hearing Aid Coverage by Insurance Companies
- Universal Newborn Hearing Systems
- Parent/Professional Collaboration
- Deaf/HH Adult Mentoring
- Parent Education and Support
- Deaf Child's Bill of Rights
- Natural Environments and Children who are Deaf/HH
- Parent's right to make informed communication choices
- Meaningful Parent Involvement
- Educational Advocacy and Training

Hands & Voices Resources Available to Parents and Professional May Include:

- Website
- The Resource Guide for Parents
- Quarterly publication, "The Communicator"
- Lending Library
- IEP Facilitation/Advocacy
- Public Speakers Bureau
- State Chapters Affiliate Support
- "We are Hands & Voices" video
- Statewide workshops
- Legislative advocacy
- Parent perspective representation on statewide/national issues
- Regional Parent Coordinators
- Spanish Speaking parent coordinator
- Unilateral Hearing Loss parent Coordinator
- Teacher/professional pre-service and in-service training
- Deaf /HH Adult Role Models

Tell us Who You Are

We invite you to become a member of Hands & Voices. Your membership benefits include: the quarterly Hands & Voices Newspaper, THE COMMUNICATOR; reduced fees to attend workshops; advocacy assistance priority.

HANDS& VOICES

Annual Membership Dues

Sign up for:	Price
☐ Parent of deaf/ hh child*	$25.00
☐ Deaf or hard of hearing adult	$25.00
☐ Student	$25.00
☐ Professional	$40.00
☐ Organization / agency	$50.00
☐ Additional Donation	_____
Subtotal:	_____
*Scholarships available for parents check here	_____
Total:	_____

Name _____

Address _____

_____

_____

Children's Ages

_____

Phone

Method of Payment

☐ Check
☐ Cash
☐ Scholarship - Parent

Please include payment with registration

GUIDE BY YOUR SIDE™

FOR FAMILIES

Providing unbiased, emotional support and resources by trained Parent Guides to families families with children who are deaf, hard of hearing and hearing impaired.

Arizona Hands & Voices
Guide By Your Side
P.O. Box 64091
Tucson, AZ 85728
Toll free: 1-866-685-1050
Fax: 1-520 843-2070
Email: gbys@azhv.org

Guide By Your Side

provides. . .

- A Parent Guide to listen and share resources
- Unbiased support
- Compassion and knowledge from parents who have a child who has a hearing loss

Why is Parent Support Important?

- It helps families identify options without telling them which options to choose
- It offers experience from a Parent Guide who knows what it is like to raise a child who has a hearing loss
- It gives the family a chance to meet others who are traveling down the same path

CALL Guide by Your Side **TODAY**
toll free: 866-685-1050
or
520-331-3125
EMAIL Guide By Your Side
gbys@azhv.org
WRITE TO Guide By Your Side
P.O. Box 64091
Tucson, AZ 85705

"The best thing we did for our family was to get involved with the Guide By Your Side program. It made a huge difference in our knowledge & outlook regarding our son's future."

How much does it cost?

- The Guide By Your Side program is **FREE** to families

THE EAR FOUNDATION OF ARIZONA

Arizona Department of Health Services

HANDS & VOICES™

GUIDE BY YOUR SIDE™
PARA FAMILIAS

Proporcionando apoyo emocional y recursos imparciales por Guías de Padres entrenados a las familias con niños que tienen pérdida auditiva

Arizona Hands & Voices
Guide By Your Side
P.O. Box 64091
Tucson, AZ 85728
Toll free: 1-866-685-1050
Fax: 1-520 843-2070
Email: gbys@azhv.org

Guide By Your Side
provee...

- Un Guía de Padres completamente entrenado quien escuchará, compartirá recursos y experiencias personales de manera compasiva y con conocimiento de causa
- Apoyo imparcial
- Guías de Padres que tienen experiencia de primera mano como resultado de criar a su propio niño con pérdida auditiva

Por qué es el apoyo a los padres importante?

- Ayuda a las familias a identificar opciones sin decirles cual opción escoger
- Ofrecen experiencias de Guías de Padres quienes conocen cómo es criar a un niño que tiene pérdida auditiva
- Da a la familia la oportunidad de conocer otras personas que viajan por el mismo camino

LLAME Guide by Your Side
sin pager: 866-685-1050
o
520-331-3125
EMAIL Guide By Your Side
gbys@azhv.org
ESCRIBA A Guide By Your Side
P.O. Box 64091
Tucson, AZ 85705

"La mejor cosa que hicimos por nuestra familia fue involucrarnos en el programa:
Guide By Your Side.
Hizo una diferencia enorme en nuestro conocimiento y en la perspectiva con respecto al futuro de nuestro hijo."

Cuanto cuesta?

- El programa Guide By Your Side es gratis para las familias

Mom, I just had my hearing screened and I need to have my ears re-checked in two weeks.

It is very important to bring me back to the hospital or to an Audiologist. I might have a hearing loss. Someone may be calling you to answer questions and help get my ears checked. If you don't want someone to call you, please call 1-866-685-1050.

Arizona Department of Health Services

HANDS & VOICES™

GUIDE BY YOUR SIDE™

THE EAR FOUNDATION OF ARIZONA

Arizona Department of Health Services is partnering with Guide By Your Side to assist families through the screening process. A Parent Guide will be contacting you. If you prefer not to be contacted, please call the Program Coordinator at 866-685-1050.

Mamá, acaban de examinar mi audición y necesito tener otra revisión en dos semanas.

Es muy importante que me traigas nuevamente al hospital o a visitar a un audiólogo. Puede ser que yo tenga perdida auditiva.
Podrían llamarte para contestar tus preguntas y ayudar a que examinen mi audición. Si no quieres que te llamen, por favor comunícate al 1-866-685-1050.

Arizona Department of Health Services trabaja con Guide By Your Side para assistir familias en el proceso de chequear. Un Guía de Padres le contactará a Ud. Si prefiera no ser contactado, favor de llamar el coordinador del programa 866-685-1050.

Hearing Aids – Resources for Parents

Hearing aids as well as other audiological services can be quite expensive and unaffordable for many families. Fortunately there are programs in Arizona that can help families and children that are in need of assistance.

Loaners:

Your first option would be to check with your audiologist about loaner hearing aids. Loaner hearing aids are also available to any Arizona family, through the HEAR for Kids Program with the EAR Foundation of Arizona. Loaners are usually available for up to six months. Longer loan periods are possible for special circumstances such as assessment for cochlear implantation. The program is funded by grants and donations.

Purchasing Equipment:

Research on how to purchase and pay for permanent hearing aids should begin as soon as possible. It may take several months to determine eligibility for some programs. The answers to some of the questions listed below may help you find the assistance that is right for your family.

Q. Is your child enrolled in an AHCCCS or KidsCare health plan?

NO ► You may be eligible for other programs as described below.

YES► Your child may be eligible for hearing services through Children's Rehabilitative Services (CRS). Information about CRS services and how to apply are on the next page.

Q. Does your health plan cover hearing aids?

NO➔ There may be an appeals process to challenge limits or restrictions on coverage. You may be eligible for other programs described below.

YES➔ Call your health plan Member Services and ask for information about your hearing service benefits.

Q. Are your financial resources limited?

NO➔ Ask your audiologist if the office or facility where you receive audiological services can arrange a payment plan.

YES➔ There are several programs available to those with financial needs.

Your audiologist may participate in some of the following programs, which are described in more detail on the next few pages. Your audiologist may be helpful in deciding which program/s best suit your needs. Please note that some programs may require you to change providers.

- HEAR for Kids
- UnitedHealthCare Foundation for Children (higher income levels)
- Starkey Foundation's HEAR Now Program
- Lions Affordable Hearing Aid Program
- Sertoma Clubs

Revised 10/2010 AzEHDI

Children's Rehabilitative Services (CRS)

CRS provides medical care and support services to children and youth who have certain chronic or disabling conditions. CRS recipients can get hearing related services in one of four MultiSpecialty Interdisciplinary Clinics (MSICs) located in Flagstaff, Tucson, Yuma and Phoenix.

The Arizona Health Care Cost Containment System has contracted with Arizona Physicians IPA (APIPA) to administer the CRS program, as APIPA-CRS.

Eligibility:

To be eligible for APIPA-CRS services you must:

- Have an eligible medical condition (Most hearing impairments are eligible conditions)
- Live in Arizona
- Be under age 21
- Be a U.S. citizen or qualified alien.

APIPA-CRS recipients must also be enrolled in an AHCCCS acute health plan or ALTCS (Arizona Long Term Care System) plan. You may call Member Services toll-free at 1-866-275-5776 for more information.

Hearing Aids and Cost to the Family

APIPA-CRS covered services are provided at no cost to recipients. Hearing services, including hearing aids, earmolds and fitting are covered for qualified APIPA-CRS recipients. Hearing aids may be replaced every three years or more often if there is a significant change in hearing. Hearing aids are covered for loss or damage by a two year replacement warranty. If the original hearing aid(s) are lost or damaged, families are encouraged to purchase insurance for the replacement aid(s).

For some types of hearing aids, prior authorization may be needed before APIPA-CRS can provide them. Your APIPA-CRS hearing services provider will take care of this for you.

For families who have ALTCS, APIPA-CRS and private insurance

APIPA-CRS will coordinate benefits with your private insurance and you will not be billed for any remaining cost, when you get hearing services at an MSIC or APIPA-CRS provider.

Additional Services

APIPA-CRS recipients who receive hearing services may also be eligible for additional medical specialty services like ENT, Genetics, Ophthalmology, among others at their MSIC.

Apply by Filling Out an Application:

To get an application or information:

- Call Member Services toll-free at: 1-866-275-5776.
- Visit the APIPA-CRS web site at www.myapipacrs.com,
- Call the Office for Children with Special Health Care Needs (OCSHCN) at 602-542-1860,
- Call 1-800-232-1676 and ask for the CRS Program, or
- Download an application from the OCSHCN web site at www.azdhs.gov/phs/ocshcn/crs/crs_az.htm.

HEAR for Kids

HEAR for Kids is a program of the EAR Foundation of Arizona. Most of the funding is provided by St. Luke's Health Initiatives and private donations as well as grants from Arizona Community Foundation, Nina Mason Pulliam Charitable Trust and others. Authorization is generally available within 48 hours of the application.

Eligibility:

Eligibility is based on family income, household size/dependent care, and expenses. If the child is covered by AHCCCS or KidsCare or other insurance that covers hearing aids, they are not eligible for HEAR for Kids. Children must be currently living in Arizona to be eligible.

- **Income:** Total Household income for the past 12 months: Wages/Salary, Pension, Social Security, Child Support and any other income.
- **Expenses:** Total Allowed Deductions for the past 12 months include medical/dental not paid for by health insurance or third party, annual rent or mortgage payments, annual payments for primary vehicle.
- **Dependent Care:** Use the following calculations:
  - Number of children in childcare_____ x $200 x number of months_____= $_____
  - Number of incapacitated adults receiving care____x$100x number of months_____=$____

Using the amounts you calculated above, find **Annual Income.** Take **Income** minus **Expenses** minus **Dependent Care** = **Annual Income**. Using the chart below, find the number of people in your family and the corresponding maximum **Annual Income** to be eligible. (current as of 3/2010).

Number in Family	**Annual Income**
1	$16,245
2	21,855
3	27,465
4	33,075
5	38,685
6	44,295

For each additional person, add $5,610

Cost to the Family:

As long as the family qualifies under the financial criteria above, there are no costs. Earmolds are covered for the first year as many times as needed. Earmolds maybe covered after the first year if the family continues to meet the financial criteria. Repairs or new aids are covered as needed if the family qualifies.

Hearing Aids:

Any aid selected by the audiologist will have a 1 or 2 year warranty. There is no restriction on type of hearing aid although the least expensive aid available with the features needed is generally purchased to ensure that the funding is available for all in need. Some repairs and replacement of cochlear implant parts may also be available.

Applications can be downloaded on the EAR Foundation website at www.earfoundationaz.com but must be submitted by the audiologist who will be helping select and fit the hearing aid(s).

The Ear Foundation can be contacted directly by calling 602-690-3975, or e-mail to: lylisolsen@msn.com

UnitedHealthCare Children's Foundation

The UnitedHealthCare Children's Foundation is a

An application must be submitted to the

501(c)(3)non-profit charity dedicated to facilitating access to medical-related services that are not fully covered by the available commercial health benefit plan. This "support" is in the form of a medical grant to be used for medical services not covered or not completely covered by commercial health benefit plans.

The applicant must be covered by a commercial health benefit plan and limits for the requested service are either exceeded, or no coverage is available and/or the copayments are a serious financial burden on the family. The UnitedHealthCare Children's Foundation requires a commercial health benefit plan. If your health plan is an AHCCCS, ALTCS or KidsCare plan, you will not be eligible for this grant, but you may be eligible for CRS. Read more about CRS on page 2.

Eligibility:
The applicant must be 16 years old or younger and live in the United States and receive and pay for care/items in the United States.

Financial need of the child's family will be evaluated and documented through information provided on the application and by submission of a photocopy of the most recently filed Federal tax return (Internal Revenue Service 1040, 1040-A, or 1040-EZ). The following scale will be used to determine financial eligibility:

Your Family Size As reported on your IRS 1040	**Adjusted Gross Income** As reported on your IRS 1040
2	$40,000 or less
3	$60,000 or less
4	$80,000 or less
5 or more	$100,000 or less

NOTE: Awards will NOT be granted to individuals in families whose Adjusted Gross Income (AGI) exceeds the scale.

Foundation prior to the receipt of services. The Foundation does not pay for past medical expenses

Exclusions:
The UnitedHealthcare Children's Foundation has a specific set of items that are excluded from grant consideration. For a complete list of exclusions see www.uhccf.org.

Hearing Aids:
There does not appear to be any restriction on types of hearing aids other than a limit of $5,000 or 85% of the fund balance, whichever amount is less per year. Awards to any one individual are limited to a lifetime maximum of $7,500.

If a grant is approved by the Regional Board of Directors for your child, the grant will help pay for approved medical services/items after your commercial health benefit plan submits payment, if any. The grant funds are not paid to you or the child outright - you work with the Foundation on submitting invoices/bills for approved medical services/items after your commercial health benefit plan submits initial payment (if any) to the health care provider

Applications:
Applications and further information on criteria and services can be found at the link below or by calling (952) 992-4459.
www.uhccf.org

Starkey Foundation's HEAR Now Program

HEAR Now is a national non-profit program committed to assisting those permanently residing the in the U.S. who are deaf or hard of hearing, who qualify under the National Poverty Guidelines for assistance and have no other resources to acquire hearing aids.

Eligibility:
All applicants are asked to call HEAR Now to discuss eligibility for the program. Please call 800-648-4327.

Income Guidelines: All income figures are based on take-home wages (net income) from all members of the household

Size of Family Unit	HEAR NOW Income Guidelines	Size of Family Unit	HEAR NOW Income Guidelines
1	$17,867	4	$36,137
2	$23,957	5	$42,227
3	$30,047	6	$48,317

NOTE: For family units with more than 6 members, add $6,090 for each additional member.

HEAR Now also considers family assets such as savings, retirement funds, life insurance and annuities.

Cost to the Family
The family is responsible for finding a hearing health care professional willing to work with them and the HEAR Now Program. HEAR Now does not provide a list of hearing health care professionals or make referrals to practitioners. Check the listings in your local phone book under "Audiologists" and/or "Hearing Aids" and call to ask if they are a HEAR Now provider. If they are a provider, ask if they can take you on as a new client. Most Pediatric Audiologists in Arizona are not participating providers and you may be responsible for the cost of the fitting and programming of the hearing aids.

The family is responsible for the cost of the evaluation/assessment and the non-refundable processing fee to HEAR Now. Once the aids are provided, the family is responsible for the purchase of batteries and extended warranty coverage for the aids.

The Hearing Aids are selected by the Foundation and will be Starkey products. Starkey makes behind-the-ear, in-the-ear and bone conduction hearing aids. A one year warranty is provided. Check with the audiologist to make sure that these hearing aids will meet your child's needs before pursuing this option.

Additional Information and Application:
This information is current as of March 2010. You can access the website link below for more current information and to access the application process or call 1(800) 328-9602.

www.sotheworldmayhear.org

Lions Affordable Hearing Aid Program (AHAP)

The Lions AHAP is rooted in a continuum of care

Sertoma Clubs

Sertoma stands for SERvice TO MAnkind.

model involving hearing care professionals and focuses on low-income beneficiaries. The hearing aid is one part of the solution to hearing impairment.

Also needed are the services of hearing care professionals to conduct tests, order ear molds, program the hearing aids and do follow-up care.

Currently Lions AHAP is distributing two hearing aids through a partnership with Rexton, Inc. in cooperation with participating Lions foundations, districts and clubs, and hearing care professionals. Lions clubs, districts and hearing programs are able to order one or both aids from Lions AHAP.

How does the Lions AHAP program work?

A Lions club decides the eligibility of a person, and the person is tested by a hearing care professional. The Lions club sends the order to Lions AHAP, who notifies the manufacturer. The hearing aid is shipped to the hearing care professional listed on the order form. The hearing care professional fits the hearing aid for the person. Individuals cannot apply directly to Lions Club International Foundation; he/she must work through his/her local Lions club.

Eligibility

The criteria for eligibility are income-based. This includes using the federal government's poverty guidelines and adjusting it to the local economy. The hearing aids are for the segment of the population who would never be able to purchase hearing aids.

Each club has their own criteria and application process. For further information on the program, contact the Lions AHAP office at (630) 468-6771 or e-mail LionsAHAP@lionsclubs.org
You can also contact a club near you by going to the Lions International website: www.lionsclubs.org

Sertoma's primary focus is on assisting the more than 50 million people with hearing health issues and educating the public on the issues surrounding hearing health. In order to achieve these goals Sertoma has undertaken a multi-faceted approach by launching programs that address both the treatment and prevention aspects of hearing health.

Communicative Disorders Scholarships

There is a $1,000 Scholarship for hard of hearing or deaf students that have clinically significant bilateral hearing loss. Graduating high school seniors or undergraduate students must be pursuing a four year degree.

There is also a $1,000 Scholarship for graduate students who have been accepted into a graduate level program in audiology or speech-language pathology at institutions in the United States. Sertoma provides more funds nationally for graduate level study in communicative disorders than any other single organization.

Other Services:

Sertoma Affiliates are non-profit hearing and speech facilities that have established a relationship with a Sertoma Club or have an independent relationship with Sertoma. This relationship results in greater service to people with communicative disorders by supporting the professional staff and programs of the affiliate.

There are 7 Sertoma clubs in Arizona. Locations and contact information can be found on their website or by calling 1 (816) 333-8300.

www.sertoma.org

Community Hearing Aid Program, Inc. (CHAP)

CHAP is an independent charity funded by St. Luke's Health Initiatives, BHHS Legacy Foundation,

This list was compiled to assist families in obtaining funding to cover the costs of

Board of Visitors, Virginia G. Piper Charitable Health Trust, Sertoma Clubs and private donations.

Eligibility

The criteria for eligibility are based on income,hearing loss, and residency status. Total household income for the past 12 months is taken into consideration. Income received from wages/salary, pension, Social Security, child support and any other income by any wage earner in the family is taken into consideration. Total Household income must be at or below 200% of the Federal Poverty Level (FPL).

Additional Cost to the Family

There is an application fee of $75 per hearing aid requested that is due along with the application and supporting documentation. There are no additional costs.

HEARING AIDS

The CHAP Preferred Audiologist who provides the fitting will select the most appropriate hearing aid for the patient's loss and listening demands. Hearing aids are both behind the ear and in the ear models. The majority of aids fit are new; however, there are a few very high end digital aids that have been certified to manufacturer specification that can be requested. All repairs to CHAP hearing aids are paid for by CHAP. If the hearing aid is old and/or deemed no longer repairable, an application for a new hearing aid would be required for a replacement aid.

Applications

Applications can be downloaded from the CHAP website www.communityhearingaidprogram.org

If you have questions you can call Kimberly Langer-Roedel @ 623-748-8814 or email theearlady@cox.net

hearing aids. The information should be verified before decisions are made as programs, funding and eligibility may change over time. Talk with your audiologist about other options.

If you have questions or would like to update the information you can contact:
Arizona Early Hearing Detection and Intervention State Coordinator:

Lylis E Olsen, MS, MPH
602-690-3975
lylisolsen@msn.com

Aparatos auditivos – Recursos para Padres

Los aparatos auditivos y otros servicios audiológicos pudieran ser demasiado caros e imposibles de pagar para muchas familias. Por suerte, hay programas en Arizona que pueden ayudar a las familias y a los niños que necesitan ayuda.

Prestados

Primero debería preguntarle a su audiólogo/a si tiene aparatos auditivos que le puedan prestar. También hay aparatos auditivos disponibles para cualquier familia de Arizona mediante el programa *HEAR for Kids* de la Fundación *EAR* de Arizona. Por lo común, puede usar los prestados por hasta seis meses. Pudiera usarlos por más tiempo en circunstancias especiales, tales como evaluaciones para implantes de cócleas. El programa se sufraga con subvenciones y donativos.

Compra de equipo

Debe comenzar a investigar lo más pronto posible cómo comprar y pagar los aparatos auditivos permanentes. Pudieran tardarse varios meses para determinar si reúne los requisitos de algún programa. A continuación se responde a algunas preguntas que pudieran ayudarles a conseguir la ayuda correcta para su familia

P. ¿Está inscrito/a su niño/a en el plan médico de *AHCCCS* o *KidsCare*?

NO ► Pudiera reunir los requisitos para participar en otros programas que se describen a renglón seguido.

SÍ ► Su niño o niña pudiera reunir los requisitos para recibir servicios de la audición mediante los Servicios de Rehabilitación de Niños/ *Children's Rehabilitative Services (CRS).* En las páginas siguientes se le informarán los servicios de *CRS* y cómo solicitarlos.

P. ¿Cubre su plan médico los aparatos auditivos?

NO ➔ Existe un recurso de apelación para oponerse a límites o restricciones de cobertura. Pudiera reunir los requisitos para otros programas que se describen a continuación.

SÍ ➔ Llame a los Servicios de Miembros de su plan médico y pida información acerca de sus prestaciones para servicios de audición.

P. ¿Tiene recursos económicos limitados?

NO ➔ Pregúntele a su audiólogo/a si el consultorio o la instalación en la que se le prestan servicios audiológicos puede ofrecerle un plan de pagos.

SÍ ➔ Hay varios programas disponibles para los que tienen necesidades económicas.

Su audiólogo/a pudiera participar en algunos de los programas que se describen con más detalles en las páginas siguientes. Le podrá ayudar a decidir qué programa(s) le convendrían más a sus necesidades. Por favor, tenga en cuenta que algunos programas pudieran conllevar que tuviera que cambiar de proveedores.

- *HEAR for Kids*
- Fundación para Niños de *UnitedHealthCare* (para ingresos más altos)
- Programa *HEAR Now* de la Fundación *Starkey*
- Programa de Aparatos Auditivos Asequibles de los Leones
- Clubes Sertoma

Revisado en octubre de 2010 AzEHDI

Servicios de Rehabilitación de Niños
Children's Rehabilitative Services (CRS)

El programa *CRS* proporciona atenciones médicas y servicios de apoyo para niños y jóvenes que tengan ciertas condiciones críticas o incapacitantes. Las personas que reciban servicios de CRS podrán conseguir servicios de la audición en una de las cuatro Clínicas Interdisciplinarias de Especialidades Múltiples (*MSIC*, por sus siglas en inglés) que se encuentran en Flagstaff, Tucsón, Yuma y Phoenix.

El plan médico público *Arizona Health Care Cost Containment System* (AHCCCS) contrató a la asociación de consultorios individuales *Arizona Physicians IPA* (*APIPA,* por sus siglas en inglés) para que administre el programa *CRS* con el nombre de *APIPA-CRS*.

Requisitos

Pare reunir los requisitos de los servicios de *APIPA-CRS*, debe:

- Tener una afección o condición médica que califique (la mayoría de los defectos de la audición son afecciones que califican)
- Vivir en Arizona
- Tener menos de 21 años de edad
- Ser ciudadano/a de los EE. UU. o extranjero/a calificado/a.

Los que reciban servicios de *APIPA-CRS* deberán inscribirse también en un plan médico de afecciones agudas de *AHCCCS* o en el plan del Sistema de Atenciones a Largo Plazo de Arizona (*Arizona Long Term Care System; ALTCS*, por sus siglas en inglés). Puede llamar gratis a Servicios de Miembros para mayor información al 1-866-275-5776.

Aparatos auditivos y costos para la familia

Los servicios que cubre *APIPA-CRS* se proporcionan gratis a los que los reciben. A los que reúnan los requisitos del programa se les cubrirán los servicios de la audición (entre ellos los aparatos auditivos, los aparatos conchas y los ajustes). Los aparatos pueden reemplazarse cada tres años o antes si hay cambio notable en la audición. Una garantía de reemplazo de dos años asegura las pérdidas o daños a los aparatos auditivos. Si se perdiera(n) o dañara(n) el o los aparato(s) auditivo(s) original(es), se anima a los familiares a que compren un seguro para los aparatos auditivos de reemplazo.

Pudiera hacer falta autorización de antemano para que *APIPA-CRS* proporcionara ciertos tipos de aparatos auditivos. Su proveedor(a) de servicios audiológicos de *APIPA-CRS* se encargará de esto por parte suya.

Para las familias que tengan *ALTCS, APIPA-CRS* y seguro privado

Cuando reciba servicios de la audición mediante un(a) proveedor(a) de una clínica *MSIC* o de *APIPA-CRS*, el programa coordinará las prestaciones con su seguro particular y no se le cobrará a usted por los costos restantes.

Otros servicios

Los que reciban servicios de audición de *APIPA-CRS* también pudieran reunir los requisitos para otros servicios médicos especializados en su clínica *MSIC*, tales como otorrinolaringología, genética, oftalmología y demás.

Rellene una solicitud para pedirlos

Obtenga así la solicitud o la información:

- Llame gratis a Servicios de Miembros al: 1-866-275-5776.
- Visite la página de Internet de APIPA-CRS al www.myapipacrs.com,
- Llame a la Oficina para Niños con Necesidades Especiales de Cuidado de Salud (*OCSHCN*) al 602-542-1860,
- Llame al 1-800-232-1676 y procure el programa CRS, o
- Descargue una solicitud de la página de Internet de OCSHCN al www.azdhs.gov/phs/ocshcn/crs/crs_az.htm.

HEAR para Niños
HEAR for Kids

HEAR for Kids es un programa de la Fundación *EAR* de Arizona. Las Iniciativas de Salud del Hospital San Lucas (*St. Luke's Health Initiatives,* en inglés) y los donativos particulares, junto con las subvenciones de la Fundación Comunitaria de Arizona (*Arizona Community Foundation*), el Fideicomiso Caritativo de Nina Mason Pulliam y otras organizaciones, sufragan la mayoría de los costos del programa. Por lo común, las solicitudes se aprueban a las 48 horas.

Requisitos

Se califica basándose en el ingreso familiar, así como en la cantidad de personas al igual que las atenciones de dependientes y los gastos del hogar. Si el o la niño/a cuenta con seguro de *AHCCCS, KidsCare* u otro seguro que cubra los aparatos auditivos, no reunirán los requisitos para *HEAR for Kids*. Para calificar, los niños deben vivir en la actualidad en Arizona.

- **Ingreso:** El ingreso del hogar por los 12 meses pasados: sueldos y salarios, pensiones, Seguro Social, pensión alimentaria de menores y todo otro ingreso.
- **Gastos:** Suma de deducciones que se permiten por los 12 meses pasados, incluso gastos médicos y dentales en dicho plazo que no pague un plan médico o terceros, alquiler o pago de hipoteca anual, pagos anuales para el vehículo principal.
- **Atenciones de dependientes:** Calcule lo siguiente:
  - Cantidad de niños en guarderías _____ x $200 x cantidad de meses _____= $_____
  - Cantidad de adultos incapacitados que reciban atenciones ____x $100 x cantidad de meses _____=$____

Use las cantidades que calculó para determinar el **ingreso anual. Ingreso** menos **Gastos** menos **Atenciones de dependientes** = **Ingreso Anual**. Use la tabla siguiente para determinar la cantidad de personas que componen a su familia y el **ingreso anual** máximo correspondiente para reunir los requisitos. (Actualizado para marzo de 2010)

Personas de Familia	**Ingreso Anual**
1	$16,245
2	21,855
3	27,465
4	33,075
5	38,685
6	44,295

Sume $5,610 por cada persona de familia que añada

Costo a la familia

Siempre que la familia reúna los requisitos económicos anteriores, no tendrán que pagar costos. Los aparatos conchas se cubren el primer año las veces que hagan falta. Pudieran seguirse cubriendo los aparatos conchas después del primer año si los familiares siguieran cumpliendo con los requisitos económicos. Se cubrirán los arreglos o los aparatos nuevos que hagan falta si la familia califica.

Aparatos auditivos

Todo aparato que escoja su audiólogo/a contará con una garantía de 1 ó 2 años. No hay limitaciones en cuanto al tipo de aparato auditivo, aunque por lo común se compra el aparato menos caro que cuente con las funciones necesarias a fin de asegurarse de que haya dinero para todos los que lo necesiten. También habrá disponibles algunos arreglos y reemplazos de piezas de implantes de cóclea.

Puede descargar las formas de solicitud mediante la página de Internet de la Fundación *EAR* al www.earfoundationaz.com, pero deberá presentarla su audiólogo/a que le ayudará a escoger y a ajustar su(s) aparato(s) auditivo(s).

Se puede comunicar directamente con la Fundación *Ear* por teléfono al 602-690-3975, o envíele un mensaje electrónico a: lylisolsen@msn.com

Fundación para Niños de *UnitedHealthCare*
UnitedHealthCare Children's Foundation

La Fundación para Niños de UnitedHealthCare es una sociedad caritativa constituida de conformidad con el apartado fiscal federal 501(c)(3) que se dedica a facilitar el acceso a los servicios médicos y afines que no cubran los planes médicos particulares disponibles. Este "suplemento" funciona como subvención médica para los servicios médicos que los planes particulares no cubran por completo o en absoluto.

La persona a cuyo nombre se solicite deberá tener un plan médico particular y o bien excederá o bien no contará con la cobertura, o bien los copagos resultarán una carga económica de gravedad para la familia. La Fundación para Niños de *UnitedHealthCare* exige que ya tenga un plan médico particular. Si su plan médico es de *AHCCCS*, *ALTCS* o *KidsCare*, no reunirá los requisitos para esta subvención, pero pudiera calificar para el programa *CRS*. Entérese mejor acerca del *CRS* en la página 2.

Requisitos

La persona a cuyo nombre se solicite deberá tener 16 años de edad o menos, vivir en los Estados Unidos, y recibir al igual que pagar por las atenciones y los artículos en los Estados Unidos.

Las necesidades económicas de la familia del o de la menor se evaluarán y se harán constar mediante la información que se incluya en la solicitud y con fotocopia de la declaración de impuestos federales más reciente que se haya radicado (formas de Servicio de Rentas Internas/*Internal Revenue Service* 1040, 1040-A, o 1040-EZ). Se usará la escala siguiente para determinar si reúne los requisitos económicos:

Personas en la familia que reportó en la declaración de impuestos federales IRS 1040	**Ingreso Bruto Ajustado** que reportó en la declaración de impuestos federales IRS 1040
2	$40,000 o menos
3	$60,000 o menos
4	$80,000 o menos
5 ó más	$100,000 o menos

NOTA NO se otorgarán subvenciones a las familias cuyos ingresos brutos ajustados (*AGI*, por sus siglas en inglés) excedan lo indicado.

Se remitirá una solicitud a la Fundación antes de que se reciban los servicios. La Fundación no pagará gastos médicos anteriores

Exclusiones

La Fundación para Niños de *UnitedHealthcare* enumera ciertos artículos en concreto que no se pueden tomar en cuenta para las subvenciones. La enumeración completa aparece en www.uhccf.org.

Aparatos auditivos

Parece que no hay restricciones en cuanto a qué clase de aparatos auditivos, aparte de un límite de $5,000 o del 85% del saldo del fondo, lo que resulte menos cada año. No se le otorgará más de $7,500 en la vida a persona alguna.

Si la Mesa Regional de Directores aprobara la subvención, la misma ayudará a pagar los servicios o artículos médicos aprobados después de que su plan médico particular presente sus pagos, de haberlos. El dinero de la subvención no se le pagará directamente a usted ni a su niño/a: usted colaborará con la Fundación para presentar las facturas por los servicios y artículos médicos aprobados después de que su plan médico particular efectúe los pagos iniciales (de haberlos) a su proveedor(a) de atenciones médicas.

Solicitudes

Consiga las solicitudes y mayor información en cuanto a requisitos o servicios si visita el enlace de Internet siguiente o si llama al (952) 992-4459.
www.uhccf.org

Programa *HEAR Now* de la Fundación Starkey
Starkey Foundation's HEAR Now Program

HEAR Now es un programa nacional sin fines de lucro que se dedica a ayudar a los que viven permanentemente en los EE. UU. y que son sordos o medio sordos, que reúnen los requisitos de las Pautas Nacionales de Pobreza para que les ayuden y que no tienen otros recursos para adquirir aparatos auditivos.

Requisitos

Se le pide a todos los que soliciten que llamen a *HEAR Now* para platicar acerca de cómo calificar para el programa. Llame por favor al 800-648-4327.

Pautas de ingreso: Todas las cifras de ingreso se basan en los sueldos netos (lo que le sobra para llevarse a casa) de todos los que vivan en el hogar.

Personas en el hogar	Pautas de ingreso de *HEAR NOW*	Personas en el hogar	Pautas de ingreso de *HEAR NOW*
1	$17,867	4	$36,137
2	$23,957	5	$42,227
3	$30,047	6	$48,317

NOTA: Si hubiera más de 6 personas en la familia, sume $6,090 por cada persona que añada.

HEAR Now también toma en cuenta bienes familiares, tales como ahorros, fondos de jubilación, seguros de vida y anualidades.

Costos a la familia

La familia se encargará de conseguir a un(a) profesionista médico/a de la audición que esté dispuesto/a a actuar con ellos y con el programa *HEAR Now*. El programa no proporcionará una lista de profesionistas médicos ni les referirá a los mismos. Revise las listas de su guía telefónica del área en la sección de "Audiólogos (*Audiologists*)" y hasta de "Aparatos Auditivos (*Hearing Aids*)" y llámelos para enterarse de si proporcionan servicios mediante *HEAR Now*. Si los proporcionan, pregúnteles si le podrían aceptar de cliente nuevo/a. La mayoría de los audiólogos pediátricos de Arizona no participan en el programa, y pudiera tener que encargarse del costo de ajustar y programar los aparatos auditivos.

La familia se encargará de la evaluación y de la tarifa de trámite no reembolsable para *HEAR Now*. En cuanto se le proporcionen los aparatos, la familia se encargará de comprar pilas y garantía de cobertura extensa para los aparatos.

La Fundación escogerá los aparatos, que serán productos Starkey. Starkey fabrica aparatos auditivos curvetas, conchas y vibradores de conducción ósea. Se proporciona una garantía por un año. Antes de acogerse a esta opción, asesórese con su audiólogo/a para asegurarse de que estos aparatos cumplirán con las necesidades de su niño o niña.

Solicitud y más información

Esta información se había actualizado en marzo de 2010. Para conseguir información más actual y para lograr acceso al proceso de solicitud, puede ir al enlace de Internet siguiente o llamar al 1(800) 328-9602.

www.sotheworldmayhear.org

Programa de Aparatos Auditivos Asequibles (*AHAP*) de los Leones
Lions Affordable Hearing Aid Program (AHAP)

El programa *AHAP* de los Leones se desprende de una gama continua de modelos de atenciones en los que participan los profesionistas audiológicos y se enfoca en personas de ingresos bajos. El aparato auditivo es una parte de la solución a los defectos auditivos.

También hacen falta los servicios de los profesionistas audiológicos para llevar a cabo estudios, pedir conchas auditivas, programar los aparatos y proporcionar atenciones de seguimiento.

El programa *AHAP* de los Leones distribuye en la actualidad dos aparatos auditivos en alianza con la Rexton, Inc. y cooperando con las fundaciones, distritos y clubes de Leones, aparte de los profesionistas audiológicos. Los clubes y distritos de Leones, así como los programas de audición, pueden pedir uno o ambos aparatos al programa *AHAP*.

¿Cómo funciona el programa *AHAP* de los Leones?

Cierto Club de Leones decidirá si la persona reúne los requisitos, y un(a) profesionista audiológico le practicará un estudio de la audición. El Club de Leones le envía el pedido al programa *AHAP*, quien a su vez le avisa al fabricante. Se envía el aparato auditivo al o a la profesionista audiológico/a que se mencione en el pedido. Tal profesionista ajusta el aparato a la persona. Los individuos no pueden solicitarle directamente a la Fundación Internacional de Clubes de Leones; debe actuar mediante su Club de Leones más cercano.

Requisitos

Los requisitos a reunir se basan en los ingresos. Entre otras, se aplican las pautas de pobreza federales y se ajustan a la economía local. Los aparatos auditivos son para el segmento de la población que jamás podría comprarlos.

Cada club tiene sus propios requisitos y trámite de solicitud. Comuníquese con la oficina del programa *AHAP* de Leones al (630) 468-6771 o envíe un mensaje electrónico a LionsAHAP@lionsclubs.org para mayor información. También puede comunicarse con un club cercano si visita la página de Internet de los Leones Internacionales: www.lionsclubs.org

Clubes *Sertoma*
Sertoma Clubs

La palabra *Sertoma* se compone de las siglas en inglés "*SERvice TO MAnkind*" (Servicio a la humanidad). *Sertoma* se concentra principalmente en ayudar a más de 50 millones de personas que padecen de males médicos de la audición y a enseñarle al público los asuntos que afectan la salud auditiva. A fin de lograr tales metas, *Sertoma* acometió una estrategia multifacética al inaugurar programas que atiendan tanto el aspecto del tratamiento como el de la prevención en cuanto a la salud auditiva.

Becas para trastornos de la comunicación

Existe una beca de $1,000 para los estudiantes semisordos o sordos que tengan pérdida auditiva bilateral de importancia. Los que estén por recibirse de preparatoria (*High School*) o los estudiantes de licenciatura deberán avocarse a estudiar una carrera de licenciatura.

También existe una beca de $1,000 para estudiantes de postgrado que se les admita a un programa de posgrado en audiología o patología del habla y de la lengua en instituciones estadounidenses. *Sertoma* proporciona más fondos por toda la nación para estudios de postgrado en trastornos de la comunicación que cualquier otra organización aparte.

Otros servicios

Las afiliadas de *Sertoma* son instalaciones sin fines de lucro para la audición y el habla que entablaron alianza con un Club *Sertoma* o que mantienen una relación independiente con *Sertoma*. La relación resulta en más servicios para las personas que tengan trastornos de la comunicación porque apoyan a los empleados profesionistas y a los programas de la afiliada.

Hay 7 Clubes *Sertoma* en Arizona. Consiga sus direcciones e información para comunicarse en la página de Internet (**www.sertoma.org**) o por teléfono al 1 (816) 333-8300.

Community Hearing Aid Program, Inc. (CHAP)

CHAP es una entidad caritativa independiente que se sufraga con las Iniciativas de Salud del Hospital San Lucas (*St. Luke's Health Initiatives*), La Fundación del Patrimonio BHHS (*BHHS Legacy Foundation*), *Board of Visitors,* el Fideicomiso Médico Caritativo Virginia G. Piper, los Clubes *Sertoma* y los donativos particulares.

Requisitos
Los requisitos a reunir se basan en los ingresos, la pérdida de la audición y la condición de residencia. Se tomará en cuenta el ingreso del hogar por los 12 meses anteriores. Se captarán los ingresos que se reciban de sueldos y salarios, pensiones, el Seguro Social, la pensión alimentaria de niños y otras entradas de todos los que devenguen sueldos en la familia. El ingreso total del hogar no deberá exceder el 200% del Nivel Federal de Pobreza (*FPL*, por sus siglas en inglés).

Costos para la familia
Se pide una tarifa de solicitud de $75 por aparato auditivo junto con la solicitud y los comprobantes. No hay más costos.

Aparatos auditivos
El o la Audiólogo/a Preferida por *CHAP* que le proporcione los ajustes escogerá el aparato auditivo más apropiado para las necesidades y pérdidas auditivas del o de la paciente. Se dispone de aparatos auditivos en modelos de curvetas y conchas. La mayoría de los aparatos que se ajusten serán nuevos; sin embargo, se pueden solicitar ciertos dispositivos digitales de precios elevadísimos que se certifique que cumplan con las precisiones de fábrica. El programa *CHAP* pagará por todos los arreglos que se le hagan a los aparatos de su programa. Si el aparato fuera viejo o hasta se estimara que ya no pudiera componerse, habrá que presentar una solicitud de aparato nuevo para que se reemplace.

Solicitudes
Puede descargar las solicitudes de la página de Internet de *CHAP* www.communityhearingaidprogram.org

Si tuviera dudas, pudiera comunicarse con Kimberly Langer-Roedel al 623-748-8814 o enviarle un mensaje electrónico a theearlady@cox.net

Se compuso esta lista para ayudar a las familias a obtener recursos para pagar los costos de aparatos auditivos. Deberá verificarse la información antes de tomar decisiones, puesto que los programas, fondos y requisitos pudieran cambiar con el tiempo. Hable con su audiólogo/a acerca de otras opciones.

Si tuviera dudas o quisiera actualizar la información, podría comunicarse con la Coordinadora Estatal de Detección e Intervención Temprana de Arizona:

Lylis E. Olsen, MS, MPH
602-690-3975
lylisolsen@msn.com

HEAR for Kids

THE EAR FOUNDATION® OF ARIZONA

Loaner aids

- No income eligibility requirements
- Standard loan is up to 6 months
- Longer loans for exceptional circumstances (such as cochlear implant trial)
- Loan requests are made online at earfoundationaz.com by the audiologist
- Youngest children first if there is a shortage
- Hearing aids are almost all digital and include BAHA, Bone conduction, AVR and others
- Participating audiologists donate their services

Vouchers

- See financial eligibility table below
- Legal residency status is not an issue
- Families who are enrolled in AHCCCS, KidsCare do not qualify
- Families eligible but still working on enrollment in AHCCCS or KidsCare may qualify
- Insured families may qualify if their insurance does not cover hearing
- Exceptions can be made for unusual circumstances especially if a child is "stuck"
- Vouchers cover behavioral, sedated or unsedated ABR, medical clearance for hearing aid
- Not for monitoring ongoing otitis media

Permanent Hearing Aids

- Same financial eligibility as the vouchers
- The managing audiologist chooses the most appropriate hearing aid for the child
- Colors and swirls are routinely approved
- Youngest children first if there is a shortage of money

Income

Total Household income for the past 12 months: $_____

Include: Wages/salary, Pension, Social Security, Child Support and any other income.

Expenses

Number of family members living in the household:_____

Total Allowed Deductions for the past 12 months: $_____

Include: Total medical/dental not paid for by health insurance or third party, Annual rent or mortgage payment, Annual payments for primary vehicle, Dependent Care. For dependant care, use the following calculations:

Number of children in childcare ____ x $200 x number of months ____=____

Number of incapacitated adults receiving care ____x$100 x number of months ____=____

Determine eligibility by subtracting the amount in the Expenses section from the amount in the Income section and reviewing the following chart (based on 150% of federal poverty, current as of 5/05)

Number in Family	*Annual Income*	*Number in Family*	*Annual Income*	*Number in Family*	*Annual Income*
1	*$14,355*	3	*$24,135*	5	*$33,915*
2	*$19,245*	4	*$29,025*	6	*$38,805*

For each additional person, add $4,890

Contact Lylis Olsen at lylisolsen@msn.com 602-690-3975 (phone) 602-296-0425 (fax)

Applications can be found at http://earfoundationaz.com/page_010_005.html

ADVICE
AT A GLANCE

For People Who Have Sickle Cell Trait (AS)
BE INFORMED: Here are 5 things to know

❶ You have one Sickle gene (S).

- **Genes** are what cause parents to pass traits (like eye color) or health conditions (like Diabetes) along to their children. The Sickle gene **(S)** affects the red blood cells.
- **Sickle Cell Trait (AS)** occurs when a person inherits a Normal gene **(A)** from one parent, and a Sickle gene **(S)** from the other.

Each parent has two genes,
and each parent passes one gene along to the baby.

❷ Sickle Cell Trait is usually a very mild condition.

- **Sickle Cell Disease** is a serious blood disease that can be very painful.
- **Sickle Cell Trait is NOT Sickle Cell Disease.** It does not make people sick the way Sickle Cell Disease does.

❸ Problems with Sickle Cell Trait are RARE.

- **Most people** with Sickle Cell Trait **(AS)** cannot tell that they have it. Millions of people have Sickle Cell Trait **(AS)**. They are fine, and they lead active lives.
- **A few people** with the Trait **(AS)** may have:
  - Blood in the urine, from time to time
  - Some pain and discomfort at high altitudes (like in the mountains or in certain cities that are high above sea level)
  - Problems with extreme exercise in hot, humid weather, when not drinking enough water.

❹ The Sickle gene (S) runs in many families.

The Sickle gene (S) is found in people from many different countries. It is found in:

- Africans, African-Americans, West Indians
- Latinos, Brazilians and in other people from Central and South America
- Italians, Greeks and other people from other Mediterranean countries
- East Indians, Asians and people from countries in the Middle East

❺ MOST IMPORTANT: You CAN have a baby with Sickle Cell Disease, in the future.

- **You can pass** your Sickle gene **(S)** along to any of your children in the future.
- **So, find out** if your partner also has a gene that can cause Sickle Cell Disease. It could be an **(S)** gene or another gene. If so, any of your children can be born with the disease. Ask, him or her to be tested to find out.
- **Most important** both parents should be tested.

Individuals with Sickle Cell Trait should inform their doctor about their condition. If you have further questions please contact:

Your local Sickle Cell Disease Organization
Quest to Cure at:
www.questtocure.org

Arizona Department of Health Services

Office for Children with Special Healthcare Needs
602-542-1860 or 1-800-232-1676

National Coordinating and Evaluation Center
Sickle Cell Disease and Newborn Screening Program

Contact National Coordinating and Evaluation Center:
410-528-1555 (Phone)
410-528-1495 (Fax)
1-800-421-8453 (Toll Free)
www.sicklecelldisease.net

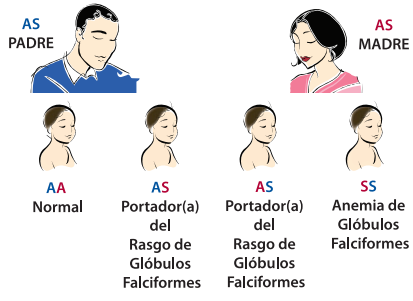
CONSEJO
DE UN VISTAZO

Para Personas Que Tienen El Rasgo De Glóbulos Falciformes. ENTÉRESE: 5 hechos a saber

❶ Usted porta un gen Falciforme (S).

- **Los genes** transmiten los rasgos (tales como el color de los ojos) o las condiciones de la salud (tales como la diabetes) de los padres a los hijos. El gen Falciforme **(S)** afecta a los glóbulos rojos de la sangre.
- **El Rasgo de Glóbulos Falciformes (AS)** sucede cuando la persona hereda un gen normal **(A)** de uno de los padres y un gen Falciforme **(S)** del otro.

Cada padre tiene dos genes, y cada padre le transmite uno de los genes a su bebé.



❷ El Rasgo de Glóbulos Falciformes suele ser una condición muy leve.

- **La Anemia de Glóbulos Falciformes** es un mal grave de la sangre que puede ser muy doloroso.
- **Ser portador del Rasgo de Glóbulos Falciformes** NO es padecer la Anemia de Glóbulos Falciformes. El rasgo no hace padecer a la gente de la forma que la Anemia de Glóbulos Falciformes enferma.

❸ Es RARO que el Rasgo de Glóbulos Falciformes cause problemas.

- **La mayoría** de los portadores del Rasgo de Glóbulos Falciformes **(AS)** no se dan cuenta del rasgo. Millones de personas son portadoras del Rasgo de Glóbulos Falciformes **(AS)**. Están bien, y llevan vidas activas.
- **Unos pocos** portadores del Rasgo **(AS)** pudieran presentar:
  - Sangre en la orina de vez en cuando
  - Ciertos dolores y molestias en elevaciones altas (tales como las montañas o ciertas ciudades muy sobre el nivel del mar)
  - Complicaciones al ejercitarse extremadamente en climas cálidos y húmedos si no se tomara suficiente agua.

❹ El Falciforme (S) se presenta en muchas familias.

El gen Falciforme (S) se encuentra entre familias de muchos países. Se encuentra entre:

- Africanos, Afro-estadounidenses, Antillanos
- Hispanoamericanos, Brasileños y otras personas de la América Central y del Sur
- Italianos, Griegos y otras personas de otros países Mediterráneos
- procedentes de la India, de otros países Asiáticos y hasta del Mediano Oriente

❺ LO MÁS IMPORTANTE: En el futuro PUDIERE tener un bebe con la Anemia de Glóbulos Falciformes.

- **Pudiere transmitirle** su gen Falciforme **(S)** a cualquiera de sus niños futuros.
- **Por lo tanto, entérese** de si su pareja también porta un gen que pudiera causar la Anemia de Glóbulos Falciformes. Pudiera ser el gen Falciforme **(S)** o algún otro gen. De ser así, cualquiera de sus niños pudiere nacer con el mal. Pídale a su pareja que se haga un estudio para enterarse.
- **Lo más importante** es que ambos padres se hagan la prueba.

Comuníquese con la organización de Anemia de Glóbulos Falciformes o clínica más cercana:
www.questtocure.org

Arizona Department of Health Services

La Oficina de Niños con Necesidades Especiales de Cuidado de Salud
(602) 542.1860 o 1 (800) 232.1676

National Coordinating and Evaluation Center
Sickle Cell Disease and Newborn Screening Program

Comuníquese con nuestra oficina nacional:
Sickle Cell Disease and Newborn Screening Program
Contact National Coordinating and Evaluation Center at SCDAA National Headquarters
231 E. Baltimore Street Suite 800
Baltimore, MD 21202
410.528.1555 (Phone)
410.528.1495 (Fax)
1.800.421.8453 (Toll Free)
www.sicklecelldisease.net

Cystic Fibrosis Newborn Screening (CF NBS)
Frequently Asked Questions (FAQ)

1. What is a "positive CF NBS" and what does it mean?

   It means that the immunoreactive trypsinogen assay (IRT) tested above the 97.8%ile or the day of the test. This triggered DNA analysis for 46 common CF genes in our population. That test was positive for at least 1 CF gene mutation. **This test is only done on the first sample.**

2. Does that mean my patient has CF?

   That means that child possesses at least 1 common gene for CF gene mutation. It also means one of their parents also must carry the gene. Your patient is at least a carrier of one CF gene mutation.

3. I thought you needed 2 gene mutations to have CF?

   This is true. CF is an autosomal recessive disease. **A child could have one common CF gene mutation and one uncommon CF gene mutation.** Only the common CF gene mutation would be detected in the newborn screen. This panel of 46 common CF gene mutations accounts for about 92% of CF disease. At this time there are over 1400 known CF gene mutations. That's why the positive NBS needs further evaluation.

4. Why don't we just send a genetic test for DNA?

   Unless you test for all the genes with expanded testing (which takes weeks and is very expensive) you will not have a definitive answer. Sometimes after the genetics are obtained the answers still remain unclear. The patient may have a new novel gene mutation and may even involve testing other family members as well. It is an expensive process that can delay an accurate diagnosis.

5. So the patient has a +NBS; what should I do now?

   At a minimum the child needs to obtain a sweat chloride test at a qualified testing center facility. Currently, those centers are at Phoenix Children's Hospital and Saint Joseph's Hospital in Phoenix and University Medical Center in Tucson.

6. What will happen at the NBS clinic visit?

   After you receive a call from the CF center regarding the +NBS, a test can be arranged, including follow up management of the patient and testing. The child may be seen at the time of the sweat chloride exam. During this visit the child is assessed for early subtle signs of CF. The family is also given accurate information about CF and given genetic counseling for family regarding the CF gene mutation. This service includes information back to your office about the testing and any further implications for your patient and their family. Providing accurate and rapid testing for your patient and information to you and your patient's family will follow.

Please have your referral coordinator obtain an authorization for both a visit/consultation and testing. The ICD-9 code is 796.6.

7. If a child had a positive CF NBS what should we do about a sibling?

   The CF NBS in Arizona started in 10/07. If a child has a + NBS for CF, any siblings that have not been previously screened (because they were born before Arizona began screening or were born in a state that didn't screen for CF) should have a sweat chloride.

8. Does the NBS detect all CF patients?

   CF NBS detection rate is about 92%. *Meaning 8 % of children could be lost to care through a variety of reasons*. The program detailed above is an attempt to limit some of the extraneous factors causing a delay to diagnosis or misinformation about CF.

9. Now that there is a CF NBS can I take CF out of a patient differential diagnosis?

   Absolutely not, as mentioned above CF NBS could miss 8 % of diagnoses. This means that any child with, for example, failure to thrive, recurrent pneumonia, chronic diarrhea or malabsorption it should be left high in the differential. *CF is a common illness present in 1/3,100 in the Caucasian population.* Please obtain a sweat chloride for that child; it is an easy, inexpensive test that can aid your diagnostic process.

10. If I have further questions who can I call?

    • Pediatric CF and NBS director at Phoenix Children's Hospital, Department of Pediatric Pulmonology (602) 546-0985.

    • Dr. Wayne Morgan, Tucson Cystic Fibrosis Center at (520) 626-7780.

    • A pediatric pulmonologist of your choice

Another source of accurate information regarding CF is the Cystic Fibrosis Foundation at www.CFF.org.

Thanks to Dana Valletta from PCH CF Center for creating this valuable resource. She can be reached at 602-546-0985

Arizona Hemoglobin Bart's
Fact Sheet for Health Care Providers

Hemoglobin Barts

Your patient has been found on the Arizona Newborn Genetic Screen to have a hemoglobin electrophoresis pattern consistent with "FA Bart's". The acronym stands for the hemoglobin species present in the baby's blood in descending order of prevalence. The F designates fetal hemoglobin (a2 y2), A denotes hemoglobin A (a2ß2) and Bart's represents hemoglobin Bart's, a tetramer of y-globin molecules (y4). Hemoglobin Barts (y4) appears in the newborn when one or more of the 4 human a-globin genes are missing. The relative over abundance of y-globin molecules leads to y4 production and the diagnosis of Hemoglobin Barts.

Alpha thalassemia is caused by deletions of the alpha globin genes on chromosome 16. Normal individuals have 4 copies of the gene with 2 on each chromosome. It is possible to lose 1 to 4 of these genes. The presence of hemoglobin Bart's on newborn screen usually suggests that the infant is missing at least 1 alpha gene.

*Usual Genotypes	Alpha-Globin Gene Deletions	Clinical Features
αα/αα	0	Normal
-α/αα	1	Silent Carrier
--/αα or -α/-α	2	α-thalassemia trait
--/-α	3	Hb H Disease
--/--	4	Fetal Hydrops

* α indicates presence of α-globin gene. – indicates deletion of α-globin gene

The silent carrier: One deleted Alpha Gene

Neonates and children with three functional alpha genes have a complete or nearly completely silent phenotype. The red cell indices are normal and remain so for life. When only one a gene is non-functional, the hemoglobin Barts percentage is usually 1-2% in the newborn, and is not detectable when the fetal hemoglobin synthesis stops at 6 months of age. As the newborn matures, the red cells can rarely exhibit a reduced MCV, MCH, but will show normal HBA2 and F levels if the hemoglobin electrophoresis is repeated. This condition is clinically benign, and these patients require no other evaluation or special health care.

Alpha thalassemia trait: Two Deleted Alpha Genes

Neonates and children with only 2 functional alpha genes will show alpha thalassemia trait. In the newborn period, Hemoglobin Barts is detected in the range of 5-6%. Most alpha thalassemia carriers will be mildly anemic, won't respond to iron therapy, and show mildly abnormal red cell indices. There is no specific treatment for the carrier state, but genetic counseling should be considered later in the child's life as it is possible that offspring could develop Hemoglobin H or hydrops. Individuals from SE Asia will often have the genes deleted from the same chromosome (*cis* mutation) while those of African heritage will have genes deleted from both chromosomes (*trans* mutation).

Hemoglobin H Disease: Three Deleted Alpha Genes

Hemoglobin H disease occurs when there is only one functional gene, and these individuals can have a mild to moderate thalassemia. Most patients are anemic and develop splenomegaly although the clinical manifestations can be variable. These children would require care at a pediatric hematology/oncology center for moderate to severe symptoms.

Fetal Hydrops: All Four Alpha Genes Deleted

This is also known as alpha thalassemia major. This is rarely compatible with life unless detected early. Those patients would be transfusion dependent, but death usually occurs in utero or in early infancy unless detected prenatally.

Follow-Up Recommendations:

At 2-3 months, examine infant for splenomegaly and do a CBC. If normal and hemoglobin electrophoresis did not report abnormal hemoglobin other than Bart's then Hemoglobin H disease is unlikely and no further work-up is necessary until 9-2 months. If CBC or exam are abnormal, please consult a pediatric hematologist.
Between 9-12 months check CBC and reticulocyte count. If this is normal, then the child likely is a silent carrier. No further work-up of the child is necessary. If infant is microcytic, do iron studies and treat with 3-6 months of iron if found to have iron deficiency. If the microcytosis resolves with iron therapy, then the child is likely a silent carrier and needs no further evaluation. If the patient has persistent microcytosis, then they likely have alpha thalassemia trait.
The parents of a child with hemoglobin Bart's should also have CBC's performed, particularly if parents are of Asian ancestry. If the parents are microcytic, then further genetic testing and counseling should be done as there may be a risk of Hemoglobin H disease or hydrops in future children.

A diagnosis of a silent carrier or alpha thalassemia does not necessarily constitute a reason to refer your patient to a pediatric hematology/oncology center. However, should questions or other circumstances arise in the lives of the affected children, **for consultation contact Dr. Brenda Wittman or the pediatric hematologist on call at the University of Arizona School of Medicine at (520) 626-8278 or through Physician's Resource Service at (520) 694-5868.** Parents of this child may be directly referred for free counseling, screening, and educational services from the Sickle Cell Anemia Society of Arizona.