

Date: May 6, 2024

From: South Dakota Department of Health, Newborn Screening Program  
State Hygienic Laboratory at the University of Iowa

RE: IMPLEMENTATION OF TESTING FOR A NEW DISORDER FOR NEWBORN  
SCREENING

### Screening for X-ALD and Hearing will begin June 3, 2024

On April 15, 2024, the South Dakota Department of Health Newborn Screening Program received approval through the administrative rule process to begin screening for X-Linked Adrenoleukodystrophy (X-ALD) as well as require newborn hearing screening as part of the newborn screening program.

Upon the adoption of the proposed Administrative Rule that includes the addition of X-ALD to the South Dakota Newborn Screening panel, the South Dakota Department of Health Newborn Screening Program will begin screening for X-ALD through the Iowa State Hygienic Laboratory's screening process on **June 3, 2024**. Additionally, all babies born in South Dakota will be required to have an initial hearing screening before leaving the hospital or by one month of age, also effective on June 3, 2024.

Effective **June 3, 2024**, all newborns born in South Dakota will be screened for X-ALD and hearing.

### What is X-ALD?

Adrenoleukodystrophy (X-ALD) is a rare, X-linked disease caused by a genetic mutation, and is more common in males than females. As a metabolic disease, X-ALD can lead to adrenal problems and potentially to more serious complications if not managed. There are three distinct presentations of X-ALD, which differ in regard to disease severity and age of onset. The symptoms and long-term outcome of each form vary widely.

To screen for X-ALD, laboratories use a machine called a tandem mass spectrometer to measure how much of the Very Long Chain Fatty Acids (VLCFA) is in the dried blood spots. High levels of VLCFAs mean a higher risk for X-ALD. When a newborn has high VLCFA levels, the baby needs more tests. The newborn screening program works with the baby's doctor to ensure the baby receives additional tests and/or visits a specialist to confirm if the baby had X-ALD.

As a reminder, this is a screening test. A false negative or a false positive result must always be considered when screening. Therefore, clinical findings and status should be considered whenever interpreting laboratory results.

**Notice of X-ALD Pilot:** During the time between June 3 and November 1, we want to be sure all components of the NBS system for X-ALD are not only in place but functioning effectively. During this time, the X-ALD results will not be reported. However, if there is a screen positive result, the results will be communicated to the SD Newborn Screening Follow-Up staff, and they will proceed with their protocols consistent with how abnormal results are handled for the other NBS conditions being screened for. The lab intends to formally begin reporting X-ALD results starting November 1, 2024.

### Purpose and Standard for Newborn Hearing Screening

Congenital hearing loss is the most common birth anomaly, affecting three in every 1,000 births in South Dakota. Hearing loss or deafness can occur when any parts of the ear are not working in the proper way. Hearing loss can affect a child's ability to develop speech, language, and social skills. However, with early identification and intervention, children with hearing loss are less likely to experience these challenges. Hearing tests can identify problems early so that children with hearing loss can be treated and get the help they need as soon as possible. [The CDC's benchmark](#) is that 95% of infants will receive an initial hearing screen by 1 month of age. Infants who do not pass the hearing screen with both ears should have a diagnostic audiology exam by 3 months of age.

Hearing screening for newborns can measure hearing without needing the child to cooperate or can be done while the child is sleeping. For example:

- **Automated auditory brainstem (aABR) tests** check the inner ear, the hearing nerve, and parts of the brain that are involved with hearing. These measure how the brain responds to the sounds.
- **Otoacoustic emissions (OAE) tests** check for damage in the hair cells in the cochlea. If the test shows little or no OAEs, a child may have sensorineural hearing loss.

Newborns and infants may "pass" or "refer" a hearing screening. Infants who "refer" in one or both ears need to be sent to a pediatric audiologist for a diagnostic exam no later than 3 months of age. See <https://EHDIPALS.org> for pediatric audiology providers in the state.

Both hearing screening and hearing diagnostic results should be reported to the South Dakota Department of Health Newborn Screening Program via the South Dakota Department of Health Electronic Vital Records and Screening System (EVRSS). Providers who do not have access to the EVRSS system may use this [form](#).

Since hearing loss may be associated with other health conditions, other referrals should be considered like ophthalmology and genetics.

Early intervention services are recommended for infants identified as deaf or hard of hearing no later than 6 months of age. Interventions may be medical, audiologic, or educational, including a range of assistive hearing technologies and communications modalities.

Referrals should be made to the [South Dakota Department of Education Birth to Three Early Intervention Program](#).

Infants who pass their newborn hearing screening can develop hearing loss at any point after birth, especially those with risk factors. Continue screening throughout childhood per AAP guidelines.

If a parent/caregiver refuses the newborn hearing screening a [refusal form](#) must be completed and submitted to the South Dakota Department of Health Newborn Screening Program.

### Education for Providers and Families

In the coming weeks, revised brochures will be available for medical providers to order through the South Dakota Department of Health website. In the meantime, these details may be of interest to your practice:

#### X-ALD:

- The newborn screening fee for the bloodspot panel will increase from the current rate of \$98.00 to \$106 on **November 1, 2024, at the completion of the X-ALD pilot period.**
- There will be no charge for repeat specimens when requested by the South Dakota Department of Health or the State Hygienic Laboratory.
- Please check with your billing office regarding the CPT code that your facility will use for the newborn screening panel. The code for the newborn screening panel is S3620.
- Bloodspot specimen collection will not change from current procedures - nothing different needs to be done for the collection of the newborn blood spot specimen.
- Additional X-ALD information is available upon request. If interested, please contact the South Dakota Department of Health Newborn Screening Program at 1-800-738-2301.
- X-ALD information is also available from the US National Library of Medicine
- ([X-linked adrenoleukodystrophy: MedlinePlus Genetics](#)) and
- Baby's First Test [Adrenoleukodystrophy | Baby's First Test | Newborn Screening | Baby Health \(babysfirsttest.org\)](#)

#### HEARING:

- The required newborn hearing screen may or may not impact how your facility chooses to bill for this service. Common practices include the use of CPT codes 92558, 92588 and 92650.
- Newborn hearing screening results are shared with the SD Department of Health via entering them directly through the birth certificate process.
- A parental refusal form is available on the DOH website. Facilities using other forms for refusal documentation will need to update their forms to account for hearing screens.
- Newborn Hearing Screening information is also available from:
- Baby's First Test: <https://www.babysfirsttest.org/newborn-screening/conditions/hearing-loss>
- Joint Committee on Infant Hearing: <http://www.jcih.org>



## DIVISION OF FAMILY & COMMUNITY HEALTH

Child & Family Services | Disease Prevention & Health Promotion

- American Academy of Pediatrics (AAP): <https://www.aap.org/EHDI>
- AAP Article:  
<https://publications.aap.org/pediatrics/article/152/3/e2023063288/193755/Hearing-Assessment-in-Infants-Children-and>

The South Dakota Department of Health and the State Hygienic Laboratory intend to work closely with you to implement screening for X-ALD and hearing status, with the goal of saving the lives and improving outcomes for affected infants. Please feel free to contact us with any questions regarding these screens.

For questions for the South Dakota Newborn Screening Program, please contact:  
Amanda Peterson, Newborn Screening Coordinator  
Phone: 605-312-0978      Email: [amanda.peterson@sanfordhealth.org](mailto:amanda.peterson@sanfordhealth.org)

For questions related to screening follow up and treatment:  
Abby Seydel, Newborn Screening Program Specialist  
Phone: 605-312-0976      Email: [Abby.Seydel@sanfordhealth.org](mailto:Abby.Seydel@sanfordhealth.org)

For laboratory-related questions, please contact:  
Kenneth Coursey, Manager – Newborn Screening Laboratory  
Phone: 319-335-467-1022      Email: [kenneth-coursey@uiowa.edu](mailto:kenneth-coursey@uiowa.edu)

For newborn hearing screening-related questions, please contact:  
Shelby Jepperson, Project Director, South Dakota HRSA EHDI Grant  
Phone: 605-357-1544      Email: [Shelby.Jepperson@usd.edu](mailto:Shelby.Jepperson@usd.edu)

For all other questions, please contact:  
Kristy Jackson, CYSHN Director, South Dakota Department of Health  
Phone: 605-910-7091      Email: [Kristy.jackson@state.sd.us](mailto:Kristy.jackson@state.sd.us)

Sincerely,

Beth Dokken  
Division Director  
Family and Community Health Services  
South Dakota Department of Health  
605-773-3737  
[Beth.dokken@state.sd.us](mailto:Beth.dokken@state.sd.us)

Kenneth Coursey  
Manager  
Newborn Screening Laboratory  
Iowa State Hygienic Laboratory  
319-335-467-1022  
[kenneth-coursey@uiowa.edu](mailto:kenneth-coursey@uiowa.edu)