

Featured Disorder

According to the Centers for Disease Control and Prevention, for every 1,000 babies born, 2-3 of them will have hearing loss. Children with typical hearing begin learning speech and language in their first six months of life. Furthermore, the most critical time for a child to learn their language (spoken or signed) is in the first 3 years of life when the brain is developing and maturing. This is why it is so important to screen and start interventions for hearing loss as early as possible.

When a baby does not pass their newborn hearing screen, parents are often scared and have many questions. Reassurance may be given to parents that hearing loss may be permanent, but it could also be temporary and easily corrected. There are many causes of hearing loss at birth – maternal infection, medical complications, extra fluid in the ears, or a family history of hearing loss. Educating parents to follow up with an audiologist for a complete hearing test is essential. Also, primary care providers need to practice timely follow-up, ensure the diagnostic evaluation takes place and coordinate care with other providers and interventionists as appropriate.

Missouri's Newborn Hearing Screening Program strives to ensure all newborns are screened for hearing loss using the 1-3-6 benchmarks recommended by Early Hearing Detection and Intervention. These benchmarks include screening for hearing loss before 1 month of age, an audiologist performing a diagnostic evaluation on any newborn who fails their screening by 3 months of age, and all infants diagnosed with permanent hearing loss are enrolled in an early intervention program by 6 months of age.

Missouri has a great early intervention program called "Missouri First Steps." Specialists work with families to foster communication, set goals, and monitor progress. The great thing about First Steps is that you do not need a provider referral; parents can refer their children if their child has confirmed hearing loss.

Resources:

Benchmarks for hearing screen, diagnosis and intervention services Missouri First Steps

Missouri Newborn Hearing Screening Program

More than 95% of newborns who are born deaf have parents with normal hearing.

Patient Spotlight

"Throughout my pregnancy, I remember constant worries as many first-time moms would: 'Will my baby be okay if I accidently ate this?', 'Will my delivery go well?'. I can't say the worries stopped once my daughter Anna was born. However, one concern that never crossed my mind was the one that would change our families' lives forever – the results of the newborn screen.

About a month after Anna was born, when we finally started to feel like we found our footing as parents, we received the news that our care team at Washington University was able to officially diagnose our baby with Pompe disease based on the results of her newborn screening. Pompe disease untreated can cause muscle loss. The purpose of having this condition on the newborn screen is to get her treatment before there is any damage to her muscles. Before testing for this, it would take, on average, twelve years for a person to receive a Pompe diagnosis. Our doctors explained

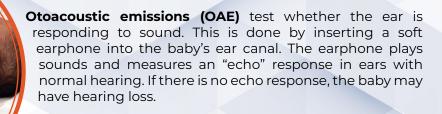
that Anna was currently asymptomatic and would not currently require treatment, only monitoring at this time. The anxiety that I thought was subsiding came back with a vengeance. The first few weeks trying to grasp this news, along with still being a parent to a newborn, were some of the hardest in my life. As time went on, I began to look at our situation in another way. I realized that I felt comfortable and confident in our care team and learned to trust them entirely. I learned that this diagnosis is a blessing, not a curse, and I had the information I needed to be proactive with my child's health. I started to appreciate all the insights I had on her wellbeing and took solace in the amount of knowledge I had. Most of all, I learned to take things day by day and enjoy my baby because our time with our children is so precious.

Today, Anna has met all her milestones and is incredibly healthy. Our care team does not think she will be symptomatic for years to come. I am so incredibly grateful for the newborn screening and the gift of knowledge it has given my family."

Robyn, Anna's Mom



There are $\mathbf{2}$ types of tests used to screen hearing in babies.





The second screening type is the **auditory brain stem response (ABR)** test. The auditory nerve and brain stem (which carries sounds from the ear to the brain) are tested for their response to sound during this test. The baby wears small earphones and has electrodes painlessly placed on the head. The electrodes record brain wave activity in response to sounds the baby should hear through earphones.



Provider Tip

Only complete a newborn hearing screening (NBHS) on a baby twice, if necessary, before discharge. Do not keep repeating the screening to get a pass. If the infant fails the second NBHS, schedule for an outpatient hearing screening to be completed within 7-10 days of age.

