A Parent's Perspective

Two Sisters with Progressive Hearing Loss Enjoy the Gift of Hearing Again

Jessica Salzberg

On March 5th, 2019, my dream of becoming a mother came true. My husband, Matthew, and I welcomed our perfectly healthy baby girl into this world. Leah was truly everything we could have imagined and more. The first two years of her life were full of love, joy, and laughter. We were in awe watching our little girl bloom and blossom before our very own eyes. She even became a sister to her little sister, Sophie. She absolutely loved this new role of hers and took it on proudly.

The first two years of Leah's life, we watched her dance to music and clap her hands to the rhythm, dance, and babble. When Leah was two and a half years old, she began to attend a preschool in our community. She adapted and adjusted well and had a love for learning and exploration. About halfway through the school year, I noticed Leah's speech began to plateau and was then beginning to decline. Her articulation and pronunciation sounded muffled, her vocabulary was no longer expanding, and at times she would not respond when her name was called. Her teacher began to have the same observations, and as our concern grew, we took Leah to an ear nose and throat specialist.

Looking for Answers

She was found to have fluid in her ears, and we were told to treat it with saline spray and come back in a few weeks. We did as instructed and when we returned to the doctor we entered the sound booth for testing with an audiologist. She was unable to gather conclusive results, and explained it is rather challenging at this age as the results yielded fair reliability. We left the appointment feeling worried and defeated that we had no clear answers. At the time, we decided to begin speech therapy for Leah twice a week. During her sessions, she was not engaged or focused, which was not in line with her personality. Even at just two and a half, she was a very cooperative little girl and it was unlike her to not be participating in the therapy.

After three months, we were desperate for answers. We scheduled Leah for an auditory brain response test three days after her third birthday. After two hours of holding her hand while she was sedated, the audiologist had results. My husband and I will never forget her exact words, "I can't believe her speech given the degree of her hearing loss." That day, Leah was diagnosed with bilateral sensorineural hearing loss. We were told she would need hearing aids and educational and speech support, and together we headed home.

My husband and I took our heartbreak and grief and immediately sprung into action mode. We had her fit with hearing aids and followed protocol of scheduling appointments with a pediatric ophthalmologist and geneticist. The day came when Leah received her pretty pink sparkly hearing aids, which we called "super ears." The beginning was a challenge, but in time she adjusted beautifully and appreciated having the gift of hearing.

A Surprising Diagnosis for Our Family

A few months later, the geneticist called with results that we had never expected. He shared that Leah's hearing loss was in fact caused by a genetic mutation (MYO15a) causing progressive hearing loss in early life. We finally had an answer and although it did not change her care, it did mean we now needed to carefully monitor Sophie's hearing. I was pregnant with my third child at the time and we were aware that meant we would need to monitor our new baby as well. Sophie, who also was once a chatterbox who loved music and dancing, began to regress in her speech and language. Now knowing that there was a possibility she might also be affected by hearing loss, our concern grew. We took her to the geneticist and fitted her for hearing aids to be proactive.

A PARENT'S PERSPECTIVE

During the period waiting for results, our third daughter Mila was born. In a time of so much uncertainty, she brought our family love and light. The day she was born, we arranged for the geneticist to test her so we

could be prepared, either way. Three weeks later, the geneticist called my husband with results for both Sophie and Mila. It was found that Sophie had the same genetic hearing loss as Leah, and that Mila was a carrier, just like my husband and I, and therefore is not affected by hearing loss. We began the process all over again. Although it was difficult, we were ready to do whatever we need to do to ensure our girls would live normal, happy, and healthy lives.

Leah and Sophie both attended the school in our community and proudly wore their "super ears" to school. We couldn't have asked for more love or support from the school administrators or their teachers. We created a team of professionals and speech therapists so the girls received all the support and services that they needed to succeed. Leah and Sophie have speech therapy three days a week, and are such hard working and motivated little girls. We were comforted to know that the girls have each other through this journey, and their little sister as their biggest cheerleader.

Cochlear Implants for Leah and Sophie

We had frequent visits with our audiologist at the Children's Hearing Program to check their devices and monitor the degree of their hearing loss. Sophie wore her purple hearing aids for three months and Leah wore her pink ones for eight months before sound booth results showed both of them were experiencing a progression in hearing loss. We realized that both girls were now candidates for cochlear implants. At the time of diagnoses, Leah and Sophie presented with moderate to severe hearing loss. Within months, they were both in the severe to profound range.

After back-to-back sedated ABR tests and MRIs, they were both approved as candidates for cochlear implantation. My husband and I did a lot of research, asked a lot of questions, and met with many professionals and specialists. After educating ourselves, we felt in our hearts this was the best option for our girls. ACI Alliance's work providing resources for families and providers is so important for families during this decision-making process.

After a lot of debate and consideration, we decided to schedule Leah and Sophie's bilateral cochlear implant surgery

on the same day. On the morning of January 4th, 2023, the next chapter of our story began. Dr. Simon Angeli of University of Miami operated on our girls, back to back. After eight hours of surgery and a twelve-hour day at the hospital, we took them home to heal. We were amazed and inspired by their resilience and how well they recovered.

After three weeks, it was activation day. Dr. Christina Sanchez, our audiologist, activated the girls together. It was a day we will forever remember. Since that very day, our girls have exceeded every single expectation we ever had as their parents. We truly could not be prouder. Leah and Sophie attend the same community preschool and are shining superstars. There is truly nothing these girls can't do. They love to sing, dance, scooter down the street, play soccer, and do gymnastics. Anything they can dream of, they can do.

It has been the biggest honor and blessing to watch our little girls become who they are today, and we are forever thankful for this life changing technology and the medical experts who have taken care of our family. We know that this is only the beginning for Leah and Sophie, and that together they will change the world.

Editor's Note: The Salzburg's story was originally shared in People magazine in June 2024 at https://people.com/two-sisters-cochlear-implants-exclusive-8670766 The People coverage was an important opportunity to expand public awareness about hearing loss, genetic counseling, and cochlear implantation. We are grateful to the Salzbergs for supporting our mission to expand knowledge and understanding by sharing their journey. ■