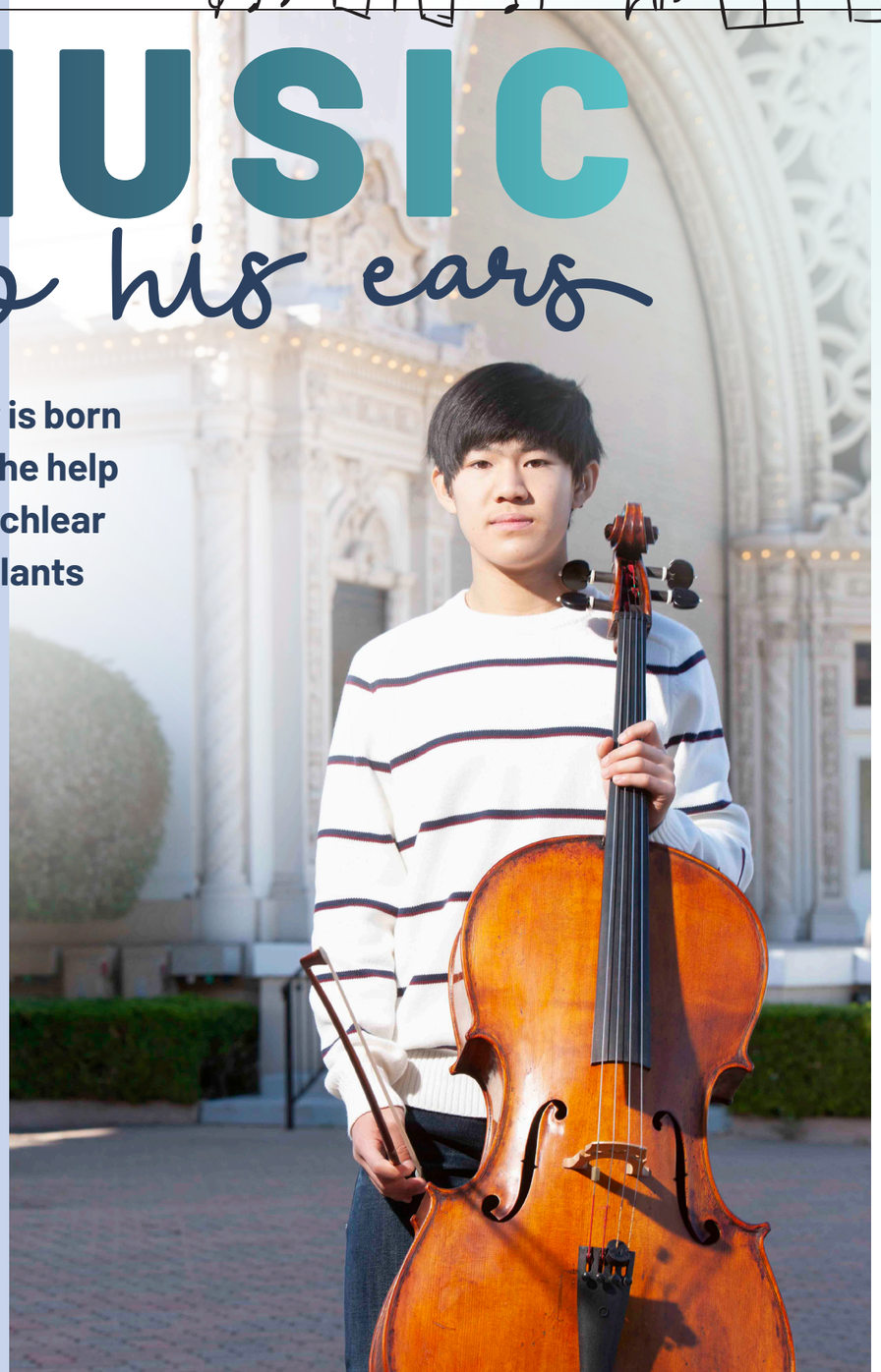
BY **CHRISTINA ORLOVSKY**

MUSIC to his ears

A star is born with the help of cochlear implants



Before Hayden Chen was born, his father, Chi-Yuan Chen, the principal viola of the San Diego Symphony, made sure his son was surrounded by music. Chi-Yuan dreamed of passing down to his firstborn the family's passion for music, so it came as quite a shock for the Chen family that Hayden was born with hearing loss.

"Hayden passed the newborn hearing test, and we never expected anything different to happen—he seemed like a happy, healthy, normal baby," Chi-Yuan says. "But then my wife, Hsin-Yee, started to notice something abnormal when he was around 4 months old. He wouldn't turn his head or react in any way to a loud noise. Sometimes his toys would drop on the floor and he would have no reaction."

The family made an appointment with Hayden's pediatrician.

"At the appointment, the doctor threw a heavy phone book on the floor. Hayden didn't even blink," his dad continues. "That's when we knew something was really wrong."

At 5 months old, Hayden visited an audiologist who delivered the devastating news that he was deaf.

"The moment we found out, it was like the entire world had collapsed. We truly couldn't believe it because no one in either of our families had any history of hearing loss and we don't know anyone with hearing loss or any other hearing disability," Chi-Yuan says. "It was especially hard for me as a musician—I really wanted him to appreciate music. It took us a long time to accept it, but fortunately, we had access to doctors who kept us very well informed. We learned that we had options."

> "the doctor threw a heavy phone book on the floor. Hayden didn't hear a sound.

EXPLORING COCHLEAR CANDIDACY

The Chens' next step was to uncover the root of Hayden's hearing loss in order to determine the appropriate intervention. They went back to their native Taiwan to see specialists and did more tests in San Diego with their doctors at Rady Children's. They learned that Hayden's hearing loss was related to the connexin 26 genetic mutation, meaning it was inherited (see sidebar). This discovery set the next steps in motion.

Knowing the cause of Hayden's hearing loss—and the potential for him to be a candidate for cochlear implant surgery—the family was referred to Daniela Carvalho, MD, director of the Rady Children's Hearing Program, a pediatric otolaryngologist and cochlear implant surgeon and a professor in the Department of Surgery at UC San Diego.

"Once we knew he could possibly have the surgery, we were more hopeful—we wanted him to at least be able to hear something," Chi-Yuan says.

Hayden was fitted for hearing aids at 5 months, which he wore while he continued to undergo additional tests.

"He had to go through all these tests and treatments to determine that he had profound hearing loss, but we were so lucky to be at Rady Children's. Everyone was so knowledgeable, and they don't just care about the medical needs of their patients. They care about how he feels. It's amazing how many patients they have, yet they can treat everyone like an individual with such personal care," he continues. "When we met with Dr. Carvalho, we were very fortunate to learn that he was a good candidate for cochlear implants. We were even more fortunate to work with Dr. Carvalho. Not only is she one of the premier surgeons doing bilateral implants (both ears at the same time), which no one could do in Taiwan, but she also explained everything thoroughly and treated us with such great empathy, like we were her relatives."

While not all patients are candidates for cochlear implants due to their neuro pathways, Hayden was one of the lucky ones.

TURNING UP THE VOLUME

At 1 year old, Hayden underwent implantation in both ears.

"With Hayden, we tried to get him implanted as soon as possible," explains Dr. Carvalho. "When they're implanted as babies, they don't know any difference. Families sometimes even call the implants their 'ears.' Hayden was implanted really young, and he's done amazingly well."

One month after the surgery, Hayden's implants were activated. It was a stressful month of wait-and-see for the Chen family.

"Over that month, you don't know if the surgery was successful," Chi-Yuan recalls. "It was a scary time. We still didn't know if after all that, he would even be able to hear."

But when they knew, they knew.

"It's amazing how many patients they have, yet they can treat everyone like an individual with such personal care.

THE GENETIC CONNECTION

Hayden Chen's hearing loss may have come as a surprise to his family, especially because neither of his parents nor other family members had any hearing loss. However, the majority of hearing-impaired children are born to hearing parents.

The genetic condition Hayden had is actually quite common. Connexin 26, a protein in the GJB2 gene, is responsible for allowing cells to communicate with each other. Without enough of this protein, the levels of potassium in the cochlea in the inner ear can become too high; this can lead to hearing loss. Mutations of Connexin 26 are the most common cause of hearing loss in newborns, resulting in as many as 50 percent of all congenital hearing loss.

This is not the only genetic cause. According to the Centers for Disease Control and Prevention, 50 percent or more instances of hearing loss in newborns is genetic, while 25 percent or more hearing loss is environmental—or due to infections in the mother while the baby is in utero or complications after birth. The CDC states that about 70 percent of genetic mutations that cause hearing loss are what's called non-syndromic, meaning they don't come with any other symptoms—like in Hayden's case. The remaining 30 percent of genetic mutations are part of a syndrome that has other symptoms, like blindness, for example.

The only way to determine if a child's hearing loss is due to a genetic mutation is to do genetic testing early on. Rady Children's offers the benefit of genetic testing as part of its comprehensive hearing program. The tests must wait until the child is born—they cannot be done in utero.

"Some parents are using 23andMe and other in-home genetic testing to test for genes they know to be responsible for hearing loss. If both parents are positive for certain genes, you know the baby will have a 25 percent chance of having hearing loss," says Dr. Carvalho.

Still, while genetic testing has come a long way, there's still much more to discover.

"We have genetic panels for 200 genes that we can test for hearing loss," Dr. Carvalho adds. "But there are thousands more we just don't know about yet."

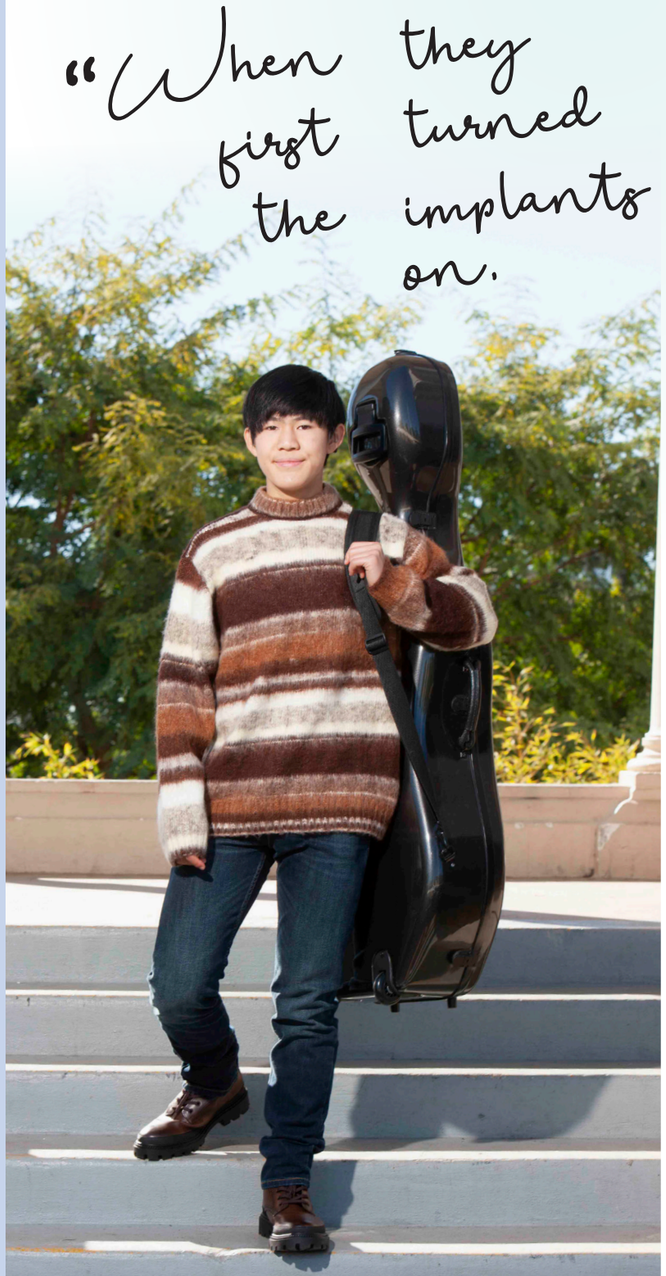
WHO IS A COCHLEAR IMPLANT CANDIDATE?

While children with mild to moderate hearing loss may benefit from hearing aids, the intervention for those with severe to profound hearing loss is a bit more complex. Thanks to Rady Children's exceptional Cochlear Implant Program and ever-evolving technology, more children receive the gift of hearing at an early age. An alternative to hearing aids, cochlear implants deliver sound information directly to the auditory nerve.

"Our Cochlear Implant Program is so comprehensive that the tool we developed to assess people for candidacy for implants is what is used by California Children's Services centers. It takes into consideration all the needs of the child and how we can best assess the level of hearing loss to get the best outcomes with the implant," Dr. Carvalho explains.

To determine candidacy, patients see a multidisciplinary team of specialists on the CHAT team over a four-hour visit. Babies as young as 6 months can receive implants.

"We aim to follow the Joint Commission on Infant Hearing guidelines, which say we should screen by 1 month, diagnose by 3 months and intervene by 6 months—whether that is fitting them with hearing aids, connecting them with resources or implanting them," Dr. Carvalho continues. "The sooner you can give kids access to sound—good sound—the better it is for their brains to develop normal speech and language that is on par with their peers. The longer you delay access to sound, the more delayed they'll be. That's why we're adamant about making sure these kids get what they need as soon as possible. It can truly make a huge difference."



"When they first turned the implants on.

"When they first turned the implants on, we could tell immediately from his eyes—all of a sudden there was something there," Chi-Yuan continues. "His eyes started to brighten, and we saw his facial expressions change as he was hearing for the first time. It gave his mom and I a lot of hope and strength."

That hope continued to grow as Hayden's implants were adjusted and he started to improve and recover.

"It's not like you put it on and it's immediately perfect," Chi-Yuan explains. "Mom did a lot of work to train him and play with him and talk to him, even just when he was eating. We followed all the doctor's instructions, which were so important, and I give her so much credit for giving him endless input for him to be able to listen, react and finally talk. He had a wonderful reaction and started making sounds just three to four months later. He started talking at 18 months."

GROWING UP HEARING

Hayden is now 15 and has spent the last 14 years of his life in the hearing world. He has no memory of not being able to hear.

"I definitely don't remember the time before my implants because it was so long ago, but because of technology, my hearing is actually really good," he says. "I can do everything my peers do and carry on conversations with them just fine. In some ways, my disability is even an advantage, because I can turn the implants off if I don't want to hear things or be distracted!"

Hayden's success is due in large part to the support he had from the hearing program at Rady Children's. His early childhood included speech language therapy three times a week. That way, he never fell behind. Living in a bilingual household also helped Hayden progress. He was exposed to both English and Mandarin from birth and is now fluent in both. Best of all, says his musician dad, Hayden is now a musical virtuoso.

"I wanted to expose him to a lot of different sounds to see what he was interested in," Chi-Yuan says. "I had my friend play cello for him, and I introduced him to the piano when he was about 5 or 6. He started to play the cello around the same time."

The cello stuck, and now Hayden is on his way to play the instrument in a showcase at the legendary Carnegie Hall in New York City, as a winner of the American Protégé international music competition.

"I love piano because I can make sounds I can hear and appreciate them," Hayden says. "But once I started cello, I started being able to play pieces that I recognized, and it became really interesting to me. I also knew other people appreciated the music I was playing, so I wanted to play for them."

Not only is he a star cellist, but the 10th grader is also the club president of the Fortissimo

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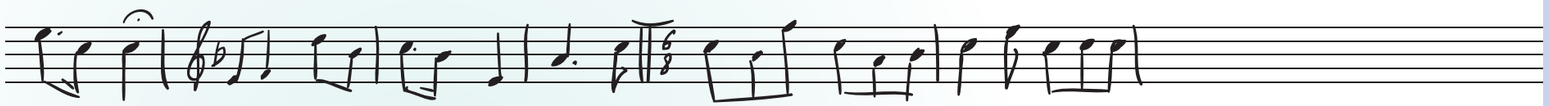

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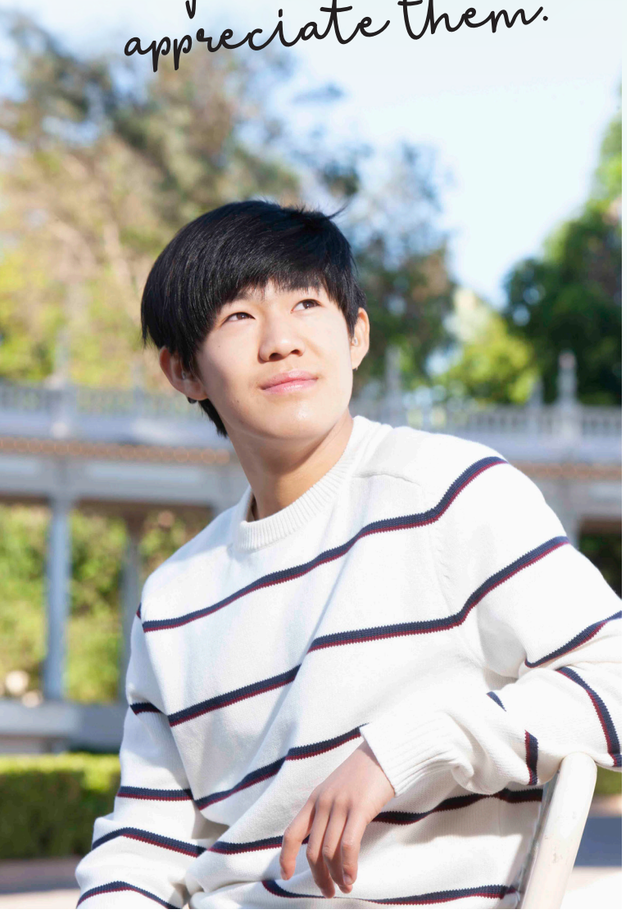

Challenge at Del Norte High School. He loves math and coding, and he plays basketball and golf. His next step, after Carnegie Hall, is to take his musical ability and give back to the hearing-impaired community. The Chens already performed a charity concert in Taiwan to raise funds for the Children's Hearing Foundation, with father playing viola and son playing cello.

"I really look forward to playing more music in the future," he says. "I started a club with my friends to play chamber music at local venues, share the joy music brings to other people and raise money to help others with disabilities gain more opportunities to thrive and be who they can be. I want to inspire others to work hard on their goals and achieve their full potential. Perhaps I can offer consultations with families going through emotional hardships and uncertainties."

His proud father is amazed at all his son has achieved and is thankful to the team at Rady Children's. He encourages others facing the life-changing diagnosis of childhood hearing loss to not delay in getting tested and exploring treatment options. Most of all, he urges, don't lose hope.

"We were very fortunate to receive the help from Dr. Carvalho and the whole Rady Children's team, and we did our best to not just start the surgical intervention early, but also do the work early so Hayden wouldn't be delayed," he says. "The surgery is just a start for families. Once you receive that benefit, it's up to you to continue to work hard to expose your kids to sound. The team's work is important, but the family is also the key to success. I encourage any families with a child with hearing loss to keep up the faith and keep up the work. There are many paths to hearing."

ABOUT COCHLEAR IMPLANTS

A cochlear implant is a surgically placed electronic device that transmits sound from an external microphone directly to the auditory nerve. It works by bypassing damaged portions of the ear. Sounds pass through a processor and are converted into digital signals. That information is then sent to a receiver/stimulator placed under the skin and muscle near the ear. Electrodes collect the signals and send them to the auditory nerve, which then sends them to the brain.