

Children's Hospital  
LOS ANGELES



# imagine

SUMMER 2018

## WHO WILL JORDAN BE?

A CHLA 'pit crew' helped the 6-year-old overcome a range of serious ailments.

### *Also in this issue:*

#### **A LIVER FOR LEX**

A baby boy's harrowing journey to a lifesaving liver transplant.

#### **SETTING THE PACE**

Despite a rare breathing disorder, Jim Harrison is living life in full.

#### **THE SOUND OF SCIENCE**

Is this the end of hearing loss caused by chemotherapy?



# About Us

The mission of Children’s Hospital Los Angeles is to create hope and build healthier futures. Founded in 1901, CHLA is the top-ranked children’s hospital in California and among the top 10 in the nation, according to the prestigious U.S. News & World Report Honor Roll of children’s hospitals for 2018-19. The hospital is home to The Saban Research Institute and is one of the few freestanding pediatric hospitals where scientific inquiry is combined with clinical care devoted exclusively to children. Children’s Hospital Los Angeles is a premier teaching hospital and has been affiliated with the Keck School of Medicine of the University of Southern California since 1932.



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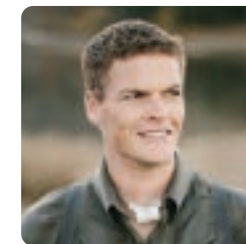
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## Welcome



I often state publicly how proud I am to work for Children's Hospital Los Angeles, an organization committed to advocating for children—individuals who cannot advocate for themselves or their own health care.

Earlier this year, I was relieved and heartened when Congress extended funding for the Children's Health Insurance Program (CHIP) for another 10 years. CHIP was originally enacted in 1997 to increase access to affordable health coverage, but it was at risk of losing funding. That would have left roughly 9 million children, including 2 million in California, uninsured.

CHIP provides important preventive services for children up to age 19 whose family income is too high to qualify for Medicaid. Together, the CHIP and Medicaid programs have cut the rate of uninsured children in California to the current historic low of 2.9 percent. The program has enjoyed bipartisan support from the start, but had become ensnared in the political clash over a government shutdown. Thankfully, temperate voices prevailed.

This year as well, I have redoubled my own efforts to advocate for the health care of children. In January 2018, I assumed the role of board chair for the California Children's

Hospital Association, and committed myself to ensuring that legislators in Sacramento prioritize access to the highest-quality medical care for children. I am also a member of the Public Policy Committee for the national Children's Hospital Association, helping to guide and steer conversations that matter to all children's hospitals and to families across the nation. Serving in these unique and crucial roles is of the utmost importance; together, we can promote children's continued access to and coverage for the health care they need.

In these roles, it helps to have stories of children who have benefited—even had their lives changed forever—because of the kind of unique, compassionate and comprehensive care that is available at a children's hospital. In this issue of Imagine magazine, you will read about research breakthroughs designed to solve hearing loss caused by chemotherapy, a toddler who received a liver transplant, and a young boy afflicted with Rubinstein-Taybi syndrome who has been supported by an array of CHLA's clinical experts stretching across Pulmonology, Orthopaedics, Gastroenterology and more.

These stories spotlight our patients' struggles and triumphs, as well as the physicians, nurses and researchers whose expertise and dedication help to heal them. Increasingly, I see my role as protecting patients' access to the expert teams here and at other children's hospitals like ours. I hope you'll be inspired by what you find in these pages and join me in advocating for them.

Warmest regards,

A handwritten signature in black ink that reads "Paul S. Viviano".

**Paul S. Viviano**  
President and Chief Executive Officer  
Children's Hospital Los Angeles

# A LIVER FOR LEX



BY SARA NAFIE

*Shortly after his first birthday,  
Baby Lex became a new man.*

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Lex Velarde Jr. was the picture of health at birth, but within just two weeks, his rosy pink skin had turned a disconcerting dark yellow.

A pediatrician assured his parents, Alejandra and Lex Sr., that jaundice was normal in newborns; they just needed to get Baby Lex some sun. More weeks went by, and still his skin looked unnaturally dark. The sun was having no effect.



Baby Lex seemed perfectly healthy when he was born in July 2016.

Alejandra's motherly instinct was strong: Something more serious than a vitamin D deficiency was affecting her baby, and it was time for a second opinion. The new pediatrician evaluated Lex and ordered an ultrasound and lab tests to determine liver function and the level of bilirubin—an orange-yellow pigment made during the normal breakdown of red blood cells—in his blood. Abnormal results can be an indication of liver damage or disease.

Lex's numbers came back "super high," Lex Sr. says. "The doctor said, 'Take him to the emergency room right now.'"

#### High hopes for a temporary fix

The Velarde family frantically drove to an emergency department near their San Diego home, where 6-week-old Lex underwent test after test. The doctors suspected a condition called biliary atresia, but additional testing was needed to confirm the diagnosis.

"We were just going to the ER for one day, and we ended up staying for more than 22 days," says Lex Sr. "We were overwhelmed."

Biliary atresia is a liver disease that develops in infancy, and the exact cause is unknown. In healthy babies, bile ducts carry chemicals the body needs to dispose of—called bile—from the liver to the gallbladder and eventually into the small intestine. Biliary atresia causes these ducts to become inflamed, scarred and blocked, leaving noxious chemicals to accumulate in the liver. Severe scarring or cirrhosis can occur by 6 to 12 months of age and ultimately leads to liver failure.

Typically, biliary atresia is treated with a surgery called the Kasai procedure. The blocked bile ducts are removed, and the small intestine is then connected directly to the liver. Although it does not usually eliminate the need for a liver transplant, the procedure can significantly extend the time before transplant.

"We heard that some kids who get the Kasai don't need a transplant," says Lex Sr., "and obviously that was our hope." Lex had the Kasai procedure in September 2016 in San Diego, and for the next few months he seemed to be getting better. Then one day he had a bowel movement and it was black—a telltale sign of internal bleeding. Blood was not passing through his liver as it should, pressure had built, and the veins in his esophagus had become swollen and burst.



October 2016, after undergoing the Kasai procedure at another hospital

He was rushed into emergency surgery to stop the bleeding, and the surgeon didn't mince words: "That's it. He needs a liver right away."

On June 2, 2017, Baby Lex and his mom flew by helicopter to Children's Hospital Los Angeles while his dad drove to meet them.

#### Destination: CHLA's Liver Transplant Program

The Liver Transplant Program at Children's Hospital Los Angeles is consistently rated among the top pediatric liver transplant centers in the nation, with survival rates that outpace other regional centers and significantly higher volumes of pediatric patients than its peers. Its experts perform the highest number of transplants in Southern California for children under the age of 18, and many, like Lex, are referred to CHLA from long distances, including San Diego.

With more than 20 years of experience performing more than 330 liver transplants in children, an exceptional multidisciplinary team led by Hepatology Director Daniel Thomas, MD, and transplant surgeon Yuri Genyk, MD, would be fighting for Lex's life. Rohit Kohli, MBBS, MS, chief of the Division of Gastroenterology, Hepatology and Nutrition, and George Yanni, MD, director of the Transplant Hepatology Fellowship Program, would also take part in his care, along with a team of pediatric hospitalists and specialists from Interventional Radiology and the Pediatric Intensive Care Unit.

"What makes our program so successful, besides experience," says Thomas, "is that we are able to combine molecular and genetic testing, high-resolution imaging, state-of-the-art pathology service and an on-site laboratory to ensure a more exact and timely way to care for our patients with liver disease."

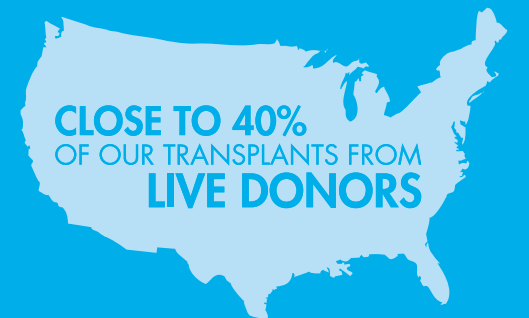
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## THE LIVER TRANSPLANT PROGRAM AT CHLA

### AVERAGE WAIT TIME TO TRANSPLANT



ONE OF THE TOP THREE LIVING-DONOR LIVER TRANSPLANT CENTERS IN THE COUNTRY



MORE THAN 300 PEDIATRIC LIVER TRANSPLANTS SINCE PROGRAM BEGAN IN 1998





Baby Lex with Daniel Thomas, MD

*“What makes our program so successful, besides experience, is that we are able to combine molecular and genetic testing, high-resolution imaging, state-of-the-art pathology service and an on-site laboratory to ensure a more exact and timely way to care for our patients with liver disease.”*

—Daniel Thomas, MD

### Fighting for Lex’s life

When a patient’s liver is failing, the patient is placed on the liver transplant list and ranked by severity. A child’s spot on the list is determined by his or her PELD (pediatric end-stage liver disease) score, which is assigned by the United Network for Organ Sharing (UNOS), a federally managed agency. The PELD score predicts how urgently a transplant is needed to avoid death within the next three months. The sicker the child, the higher the score, and for children 11 and younger, a score of 40 is required to be at the top of the list.

“Trying to get our baby on the list was so hard,” Lex Sr. recalls. “The results of his lab tests weren’t that bad—he’d score a 12 or 14—yet he was having bleeding episodes, a swollen belly and needed IVs [nutrition and antibiotics] to stay alive.”

Yanni says that in most cases, a child’s PELD score doesn’t reflect the severity of the illness. “It is the responsibility of the medical team to advocate for the child based on the clinical condition [including infection and other issues not evident in blood tests], and we can also write an appeal letter to UNOS.”



Lex celebrated his first birthday in the hospital, shortly before his liver transplant.



From left: Baby Lex’s sister, Alexia; Alejandra; George Yanni, MD; Baby Lex; and Lex Sr.

Multiple letters from the transplant team prompted UNOS to increase Lex’s PELD score to 40, enough to put him very high on the list. Feeling hopeful for the first time in weeks, Lex’s parents still had to wait for the right liver. The first two donor candidates were high risk, but Baby Lex was so sick that they had to be considered. Both were ultimately rejected by Genyk, who rushed by helicopter to inspect the potential donor livers firsthand.

Baby Lex’s family celebrated his first birthday in the hospital. Meanwhile, CHLA’s Liver Transplant Program was working to fast-track his parents for testing as potential living donors. Lex Sr. was a perfect match and was prepared to donate a portion of his liver to save his son’s life. Before that was necessary, though, another liver became available.

On July 25, 2017, Genyk transplanted the organ into Lex’s tiny body.

The transplant was a success. Baby Lex and his parents stayed in L.A. for three more weeks while he started a regimen of immunosuppressant drugs and was carefully



A father-son selfie during a post-transplant checkup, January 2018

monitored for complications. Today, the 21-month-old is back home in San Diego, where he is a happy and healthy toddler learning to speak his first words. And with his healthy liver, his skin has returned to its original rosy pink. “You’d never know he spent the first year of his life in the hospital,” says Lex Sr. “He’s beautiful. We love our Baby Lex and are so grateful to CHLA for saving his life.” 🌈



# THE SOUND OF SCIENCE )))

BY JEFF WEINSTOCK

*After a run of groundbreaking research trials, two CHLA physicians are closing in on eliminating one of the most debilitating side effects of chemotherapy.*

David Freyer, DO, hasn't had many encounters with *aha!* moments. The notion that research discoveries spring from revelations or sudden brainstorms draws a laugh from the Children's Hospital Los Angeles oncologist.

"Einstein probably had them," he says, "but for most of the rest of us, at least for me, it's very undramatic. It's just kind of hanging in there, working it out."

Since the late 1990s, Freyer's research has been occupied by a steadily gathering hypothesis: that sodium thiosulfate (STS), a rescue medication in cases of accidental cyanide poisoning, could subdue one of the harshest side effects of chemotherapy treatment in children: hearing loss. The theory started to pick up momentum a decade earlier, when Oregon Health & Science University neurosurgeon Edward Neuwelt, MD, began exploring STS' potential for preventing hearing loss caused by carboplatin, a medication used to treat brain tumors.

Neuwelt's work led to Freyer's extensive, landscape-altering four-year study on STS' ability to prevent hearing loss in young patients treated with cisplatin, a platinum-based chemotherapy like carboplatin, used against a variety of cancers. With Freyer serving as principal investigator, the National Cancer Institute-sponsored study brought together 38 hospitals from the Children's Oncology Group (COG), including CHLA. The results were published last year.

## Your hearing or your life?

For the majority of patients, use of cisplatin comes at immense cost. While going about exterminating cancer cells, cisplatin generates superoxide ions—free radicals that can travel into the inner ear and destroy the hair cells that recognize sound. Backing off the dose might spare the patient's hearing but jeopardize the cancer treatment; the alternative, however, means capitulating to an irreversible disability that affects many vital developmental functions, such as language acquisition, learning and memory.

"For decades that's how we approached it," Freyer says. "It was choosing between being alive and having some hearing loss, or dying of your cancer. That's been the paradigm we've been practicing under."

**Cisplatin** and similar platinum-based agents cause significant, **permanent hearing loss** in at least **50 percent of children.**



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STS seemed like a good candidate to disable cisplatin. A free-radical scavenger, it demonstrated in the lab that it would bind to and neutralize cisplatin before the medication could ravage the inner ear.

"It's a tricky thing," Freyer says. "You have to make sure you're not inactivating the cisplatin so that it doesn't do its job of killing the cancer cells."

To that end, STS, given intravenously, is not administered until six hours after cisplatin is given, allowing the medication time to confront the cancer cells.

The outcomes of the trial were transforming. Incidence of hearing loss was cut in half in children who received cisplatin plus STS, compared with those given cisplatin alone.

The effect was even larger in children under 5, who are most vulnerable to hearing impairment and have the most to lose from it. Seventy-three percent who did not receive STS suffered hearing loss; with STS, the number fell to 21 percent.

A longstanding assumption had been dispelled: Hearing loss was no longer an inevitability of chemotherapy. As further support, a concurrent European study of STS, called SIOPEL 6, produced similar data as the COG trial.

"This was a 'proof of principle' study," Freyer says. "It proved we have an agent that can prevent hearing loss. We don't have to accept this as a consequence of cancer treatment anymore."

**COG Trial Results:**  
STS cut cisplatin-related hearing loss **by half.**



*"For decades ... it was choosing between being alive and having some hearing loss, or dying of your cancer. That's been the paradigm we've been practicing under."*



—David Freyer, DO

"There's still more work to be done," he adds, citing the segment of patients whose hearing wasn't saved by the introduction of STS. "It's not the end of the story, it's the beginning. There's still another 29 percent we need to work on."

#### Another drug emerges

That remaining portion of patients also concerned a colleague down the hall from Freyer, Etan Orgel, MD, MS. Freyer directs CHLA's Survivorship and Supportive Care Program at the Children's Center for Cancer and Blood Diseases; Orgel runs a component of it called Medical Supportive Care Services. Together they have made hearing loss a research priority. As it became evident that STS was not 100 percent effective, Orgel began to consider a successor trial.

"We're never satisfied with 'almost,'" he says. "We started looking at the next generation. We knew we had this other drug that may be a stronger version of STS."

Orgel believed he could repurpose N-acetylcysteine, or NAC, an antioxidant that came into use as an antidote for

Tylenol overdoses. He had reason to believe NAC would outperform STS. Both are antioxidants, but only NAC increases the body's production of another antioxidant, glutathione, which would protect the hair cells in the inner ear against cisplatin-related oxidant damage.

With funding from the American Cancer Society and subsequently the National Institutes of Health, Orgel launched an NAC trial at CHLA in spring 2016, to see whether the drug was safe and tolerable, and would not disrupt the effectiveness of cisplatin.

The study is ongoing, but thus far NAC has passed all the benchmarks. Though Orgel says a preliminary review of the drug's success at preventing hearing loss is underway, he'll have to take NAC into a larger, multicenter national trial to get a conclusive result, a move he is now planning.

"These two trials showed we can actually do something about this problem," Orgel says, echoing Freyer's view. "We don't have to say to patients and families, 'This is the way it is. We can cure your cancer, but you're going to have severe hearing loss.' We can change the paradigm and say it's not a zero-sum game."



Freyer (left) and Orgel have made preventing hearing loss a priority of CHLA's cancer research efforts.

### COG Trial Results in Children Under 5: HEARING LOSS



without treatment **73%**



with treatment **21%**

"Now that we know we can do it, we all want to find a drug that's even better. Whether it's 20 percent better or 50 percent better, it needs to be better."

Freyer has already led CHLA's participation in an early study of yet a third drug, OTO 104, which unlike the other two treatments is delivered by injection directly into the ear drum by an ear, nose and throat specialist. The study is now paused as Otonomy, the San Diego-based pharmaceutical company that developed the drug, reviews the data ahead of publishing it, which Freyer guesses will happen in a few months.

None of the three drugs have yet to receive federal approval; STS is closest. The Food and Drug Administration has fast-tracked it for review, recognizing the accumulation of good data behind it and the weight of the problem it addresses.

Freyer already sees the influence of CHLA's place at the head of these trials. He says it has established the hospital's cancer research program as a pacesetter in the area of hearing loss, and caught the interest of pharmaceutical companies as well as researchers and physicians who have to face this issue.

"This early work will have been the gateway. It will have opened the door to research. I think that five to 10 years from now, and beyond, we're not going to be dealing with cisplatin-induced hearing loss the way we are now. I predict the expectation will be that we can prevent it for the vast majority of children." 🦋





# HEADING FOR SUCCESS



FROM THE EXPERTS AT THE CHILDREN'S ORTHOPAEDIC CENTER'S SPORTS MEDICINE PROGRAM

*How to safely execute one of soccer's most exciting techniques.*

Heading is a tool in soccer used to control or redirect the flight of the ball. While heading is one of the game's flashier moves, it is also the riskiest part of soccer, sometimes leading to head injuries. Most of these injuries are not actually due to head-ball contact, but to athlete-to-athlete contact during heading. The doctors at the Children's Hospital Los Angeles Sports Medicine Program in the Children's Orthopaedic Center recommend some simple safety measures to protect young soccer players.

### New rules

The U.S. Soccer Federation has mandated new restrictions on heading:

- Heading is NOT permitted during ANY practices or games for athletes 10 or younger.
- Heading is LIMITED during training sessions and NOT permitted during games for athletes 11-13 years old.
- For athletes ages 14 and older, heading can be performed while focusing on correct technique.



### Does soccer headgear help?

Headgear for soccer is an impact-absorbing foam layer that may decrease head-to-hard-surface impacts. Some studies demonstrate the possibility of reduced head impacts.

However, headgear may provide a false sense of security and promote more aggressive play, or make players more of a target for others. Additionally, headgear increases head mass, which can actually increase propensity for concussion. Use headgear in soccer with caution.



Soccer is a great way to stay active, learn important life skills and begin a lifelong tradition of good health. Setting up young athletes with good habits from the very beginning will give them the tools they need to succeed.

### Ways to stay safe

- Help players strengthen their heading muscles. Begin heading practice with a lightweight, spherical object like a balloon, and work up from there in this order:
  1. Balloon
  2. Sponge (NERF-type) ball
  3. Play ball (the soft, air-filled balls sold from big bins in grocery, drug and toy stores)
  4. Volleyball
  5. Partially deflated soccer ball
  6. Properly inflated, appropriately sized soccer ball
- Do not force young athletes to head the ball, and avoid excess heading training in practices.
- Teach good heading technique:
  - o Eyes open
  - o Move toward the ball
  - o Bend at the waist, instead of the neck
  - o Use the legs to help launch the body forward and maintain a solid foundation on the ground
- Choose the right ball:
  - o Look for Fédération Internationale de Football Association (FIFA)-certified balls to ensure optimal quality.
  - o Avoid overinflated balls. During practice consider decreasing air pressure in the ball to minimize head impacts.
  - o Stitched is better than thermal molded.
  - o Correct size is important! Appropriate ball size is determined by age. 🎯





# WHO WILL JORDAN BE?

BY SARAH BROWN

*A mother and father are determined to give their 6-year-old son the chance to succeed in life. Specialists at CHLA have given them the hope to do so.*

If you are invited into his house, Jordan, age 6, will grab you by the hand and lead you to the family room. His father might be working at the computer, his grandma baking in the kitchen; his twin brother, Justin, and 9-year-old brother, Jadin, playing in the backyard. And as is often the case, mom runs the show. You will immediately feel at home.

Things have not always been so idyllic for Jordan, though. He spent two weeks in the neonatal intensive care unit when he was born, and doctors knew that something was not right, but they could not diagnose anything specific. He had crooked thumbs, larger than normal big toes, and breathing and feeding difficulties. The pediatrician's advice: "Don't Google anything. Just enjoy your baby for now, and when you are ready, take him to the best possible place, Children's Hospital."



Jordan at 4 months old, after a stay in the hospital for RSV



## Thumbs, and courage

"Our visit with Dr. Lightdale-Miric was weird," says Jordan's mom, Joy, with a tone of happy disbelief. (She is perhaps the most aptly named person in history.)

Nina Lightdale-Miric, MD, director of the Pediatric Hand and Upper Extremity Surgery Program in the Children's Orthopaedic Center at Children's Hospital Los Angeles, knew patients with Rubinstein-Taybi syndrome (RTS) when she saw them. The telltale crooked thumbs were one of the classic signs shared by most children with this rare genetic disorder. The family had already been to see Linda Randolph, MD, in CHLA's Division of Medical Genetics, and was waiting on test results to confirm the diagnosis. Jordan was found to have RTS, though his gene mutation was different from that of anyone else in the world who had been tested to date.

Joy and her husband, Dain, had been trying to get the twin brothers to stop fighting as their first visit began. Lightdale-Miric sat and watched the frustrated and embarrassed mother try to control Jordan, but to her, Jordan's "misbehavior" was completely developmentally appropriate for his age—and surprising for RTS.

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*“A large part of our independence—our ability to communicate, our ability to complete activities of daily living on our own independently—is focused on pinch and grasp in some function.”*

—Nina Lightdale-Miric, MD

“She told us, ‘He’s going places, and he’s going to need his thumbs,’” says Joy. “Those were her exact words.” And just like that, they had hope. “She gave us the courage to step up and be the parents he needed.”



While recuperating from thumb surgery, Jordan insisted on holding his own bottle.

Lightdale-Miric fixed his thumbs, reconfiguring Jordan’s triangular distal phalanx bone into its intended rectangular shape.

“A large part of our independence—our ability to communicate, our ability to complete activities of daily living on our own independently—is focused on pinch and

grasp in some function,” says Lightdale-Miric. “Who knows what he’ll be able to do with those hands and those thumbs and his intelligence?” Now Jordan’s hands could keep up with his mind. And with his mom.

Lightdale-Miric describes the mobile classroom that Joy folds out of a backpack at every clinic visit.

“Every time you open the clinic door you can expect that mom’s singing, talking to him and engaging him in developmental learning activities,” she says.

“We decided—nobody knows who he’s going to be, so we’re going to make him who he’s going to be,” says Joy. Her background as a nurse with a doctorate in clinical child psychology does not hurt.



Joy is giving Jordan every opportunity to reach his full potential.



### Vision for the future

Joy and Dain began to notice that Jordan was having vision issues as well. When they called for Justin, he would come running. But when Jordan was called, he would respond by looking in every direction, unable to decide which way to go. A visit to Angela Buffenn, MD, at The Vision Center at CHLA, determined that Jordan was actually legally blind due to high myopia (nearsightedness). The day he got glasses was another game changer.



Jordan is fitted for his first pair of glasses.

“I remember when John [the optician] put on his glasses, and he stood and he looked around. You could see that smile on his face, and he was just looking all over the place. He was just *seeing*,” says Dain.

Dain remembers Jordan holding Joy’s face and seeing her clearly for the first time, and then crawling over to see his twin brother’s face. “He laughed the whole way home. His head was just on a swivel.” Soon after his first clear sight came his first steps.

“He would stand up, but he would never let go because he couldn’t see the floor,” says Joy. “A month after getting his glasses, he walked.”

Another obstacle removed, another milestone achieved.



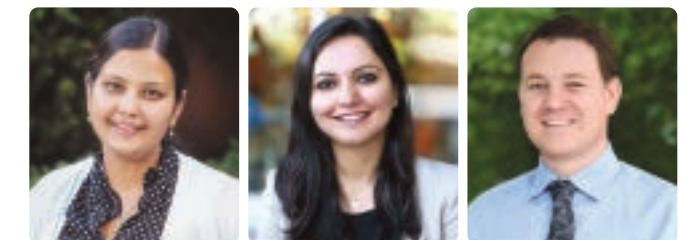
### A breath of fresh air

Jordan’s digestive and breathing issues were another challenge. He had trouble breathing and would aspirate into his airway; he also had frequent ear infections and ended up in the hospital almost like clockwork every spring and fall, with respiratory syncytial virus (RSV), aspiration pneumonia or some other ailment.

“Jordan used to fall asleep during his meals, because it was difficult to eat and breathe because his airway was so crowded,” says Joy.

Doctors had tried swallow studies and procedures to check the condition of his lungs; he had tubes placed in his ears and a frenulectomy to try to correct his swallowing issues. But nothing seemed to make much of a difference.

He eventually made his way to CHLA’s multidisciplinary Aerodigestive Program. Led by co-directors Manvi Bansal, MD, from Pulmonology; Vrinda Bhardwaj, MD, from Gastroenterology, Hepatology and Nutrition; and Christian Hochstim, MD, PhD, from Otolaryngology, the program brings together physicians and care providers from many disciplines to collaborate on diagnoses and care, and consolidate appointments and procedures for children with complex feeding and breathing issues.



Manvi Bansal, MD (left), Vrinda Bhardwaj, MD, and Christian Hochstim, MD, PhD, co-directors of CHLA’s Aerodigestive Program

While CHLA’s division head of Urology, Roger De Filippo, MD, planned an operation to fix a urological issue for Jordan, Bansal took the opportunity to perform a bronchoscopy to get to the root of his problems. Minimizing the number of times Jordan went under anesthesia was especially important, as his breathing issues made this an exceedingly risky endeavor.

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The results showed that his lungs were in great shape, but his tonsils and adenoids were enlarged and causing an obstruction in his airway. Six weeks later, Hochstim and Bhardwaj performed a combined endoscopy of the upper airway and GI tract to pinpoint the problem and find a solution. During that same procedure, Hochstim removed Jordan's tonsils and adenoids.

Yet another obstacle removed, quite literally this time.

The difference was night and day. Jordan stopped falling asleep during meals. He was able to breathe while he slept. And he started doing better in school. Although he'd previously tested into special education classes, his parents noticed improvement and asked that he be retested. The result: He could now join a mainstream kindergarten class.

"He now has more oxygen getting into his brain, which has increased his brain function and his ability to process the information," says Joy. "Isn't that crazy?"

"We often see that correlation with kids who are not breathing well at night with obstruction during sleep," says Hochstim. "They are more fatigued and tend to struggle in school. It makes a big difference for them if we can help improve their breathing. It gives them better sleep, and better energy and performance."



Jordan and his mom on a recent clinic visit with Melinda Braskett, MD

*"The largest impact on Jordan's life was having a group of people working together, like a 'pit crew,' that got him racing back to life and his family. The Aerodigestive team is a prime example of what health care should be."*

*—Joy, Jordan's mom*



#### More hurdles

The upper endoscopy had identified a condition called eosinophilic esophagitis (EoE), a chronic allergic inflammation of the esophagus.

"We like to call this asthma of the food pipe," says Bhardwaj, who is also director of the hospital's Eosinophilic Gastrointestinal Disorders Program for Gastroenterology. Melinda Braskett, MD, of the Division of Clinical Immunology and Allergy, is the program's director for Allergy. EoE can cause reflux, difficulty swallowing and vomiting, among other symptoms. This explained Jordan's problems with aspiration, his longstanding abdominal discomfort and his general lack of enjoyment related to food.

"EoE is often associated with atypical food allergies and best evaluated and treated by a multidisciplinary team including gastroenterologists, allergists and dietitians," says Braskett.

"He's severely allergic to the world," says Joy, though she is unperturbed. The family keeps an EpiPen on hand and has cut the main irritant, dairy, from Jordan's diet. The final pieces fell into place—Bhardwaj says his EoE is in remission.



Justin (left), Dain, Jadin, Joy and Jordan

"The largest impact on Jordan's life was having a group of people working together, like a 'pit crew,' that got him racing back to life and his family," says Joy. "The Aerodigestive team is a prime example of what health care should be."

"I used to worry every morning I'd wake up if he'd be alive," says Joy. Oh, how times have changed. Dain shares proudly that Jordan plays baseball now, as well as soccer. He's doing well at school and loves books and playing with trucks and friends. He is also a daredevil on his Y Fliker scooter—just like other 6-year-olds.

Now Joy and Dain can focus on helping him learn and teaching him to be resilient—and also on being a normal family again. "We can go back to being a family," says Joy. "That's what we're supposed to do." 🦋



Jordan on his prized Y Fliker scooter



# SETTING THE PACE

BY KATIE SWEENEY

*Jim Harrison does not let a rare breathing disorder keep him from the important things in life: marriage, career and the perfect weekend hike.*

"Most people can breathe without thinking about it," Jim Harrison explains. "For me, breathing doesn't happen automatically."

It is a sunny Sunday morning in early spring, and the 29-year-old has already been on a bike ride through the canyon near his San Diego home—riding the trails alongside his wife, Brittany, on her morning run. That afternoon, they have planned a day trip to the desert and a hike to see some wildflowers.

"It's the weekend," Jim says with a grin. "Gotta pack it in."

For Jim, packing in the fun has always meant being outside and being active—whether hiking, mountain biking, skimboarding or playing a little basketball. And while there is nothing out of the ordinary about that, Jim has to go about those activities a little differently than most people: He has to remember to breathe.

## One breath at a time

Jim was born with congenital central hypoventilation syndrome (CCHS), an extremely rare condition affecting only about 1,200 people worldwide.

In CCHS, a genetic mutation causes abnormal development of the autonomic nervous system, the "life support" part of the brain that makes functions such as breathing, heart rate, blood pressure and digestion happen automatically.

The disorder can cause a variety of problems, but the biggest one—and the one that affects all patients—relates to breathing.



Jim, age 2

"Basically, their brains do not tell them to breathe deeply enough or frequently enough," explains Thomas Keens, MD, Jim's longtime pulmonologist in the Division of Pulmonology and Sleep Medicine at Children's Hospital Los Angeles. "They under-breathe. Therefore, we have to breathe for them."

Most CCHS patients need breathing support only while they're sleeping, but those with severe forms—like Jim—need 24-hour support.



Jim's backpack contains a transmitter, which sends signals to electrodes that stimulate his body to breathe, no matter where he is.

But if you think that means he is stuck in a hospital bed, think again. Although Jim uses a mechanical ventilator at home while he sleeps, the only thing he needs during the day is a simple, 3-pound backpack.

The backpack contains a battery-powered transmitter that sends signals to electrodes that have been surgically implanted on Jim's phrenic nerve (which controls breathing). Each signal stimulates his diaphragm to contract—resulting in a breath.

The system is called diaphragm pacing, and Jim has been using it since he was a toddler. Today, the pacers are set to give him 22 breaths a minute, which is plenty for everyday activities and for his job as a financial analyst in Del Mar. But hiking and biking are another story. That's where Jim has steadfastly trained himself to take extra breaths. Those are the ones he has to think about, one breath at a time.

"When I was bike riding this morning, as I went faster, I had to think, 'OK, breathe more. Breathe heavier,'" he explains. "I can't ever lose focus on that."

(continued on next page)



## 'A fighting stance'

Born and raised in Tucson, Arizona, Jim spent the first six months of his life in a hospital. At age 17 months, he was diagnosed with CCHS. His parents, Jan and Jim Harrison Sr., were reeling.



Jim with his mom, Jan

"Shock isn't a strong enough term," says Jan. "We were very afraid of the future. Fortunately, with family support, we quickly got into a fighting stance. We decided to do everything possible to make his and our family's lives as wonderful as possible.

"There were so many 'toughest things' I could say we faced," she adds. "But with each one there were blessings, because we learned, grew and found new strength."

One of those blessings came when Jim was 11, and his diaphragm pacer failed. The Midwest hospital where they'd been traveling for treatment didn't have an appointment for six months. Meanwhile, Jim would have to be hooked up to his home ventilator 24/7.

Unwilling to accept that, his parents called CHLA. An international leader in CCHS care and research and a major referral center for patients, CHLA is home to one of the world's largest pediatric home mechanical ventilation programs and is a pioneer in diaphragm pacing.

Keens, who has been with the program since it began in 1977, agreed to see Jim immediately. Shortly afterward, the faulty pacer was replaced in a successful surgery at CHLA. Jim has been seeing Keens and his team, including Nurse Care Manager Sheila Kun, RN, ever since.

Because there are no adult diaphragm pacing programs, Jim still comes to CHLA today. The program takes a multidisciplinary approach to CCHS care, drawing on pulmonologists, respiratory therapists, dietitians, social workers and nurse care managers, as well as specialists from Otolaryngology, Neurology, Surgery, Cardiology, and Gastroenterology, Hepatology and Nutrition.

Another key part of the team? Families.

"We can provide the medical tools," Keens notes. "But the families are the ones who create meaningful lives for their children, by using these tools in a creative and courageous way."



Sheila Kun, RN (left), Thomas Keens, MD, and Jim and Brittany Harrison

## Living life to the fullest

Walking through Jim and Brittany's sun-filled home in a quiet, hilly neighborhood outside of Poway, you'd be hard-pressed to find signs of CCHS.

"We can tour the house; it doesn't look like a hospital," Jim says. "The bedroom upstairs has equipment in it, but that's it." What you do find, though, is wood—evidence not of CCHS, but of another one of Jim's passions: woodworking.

"He made the chairs in the living room, my nightstand, the spice rack, the key rack, the bird feeder," Brittany says. "He's made a lot of stuff."

"A lot of unnecessary stuff," Jim quips with his usual deadpan humor, then laughs. "But I like it. I like working with my hands."

He knows he makes CCHS look easy, but it wasn't always this way. Growing up, it was tough to be different, to not be able to play soccer with his friends, and to come home with excruciating headaches because he "fell behind" on his breathing.

But the older he got, the more he learned what worked for him—short, 20-minute "bursts" of exercise rather than sustained cardio—and how to say "no" to more strenuous activities.

By the time he entered the University of San Diego, he was hitting his stride. His senior year, he met Brittany. They had endless fun going to the beach and hiking and biking, and Jim doted on her, carving her a wooden jewelry box and other gifts.

"I fell in love with the fact that he always wants to live life to the fullest," Brittany says. She wasn't afraid of his CCHS. "The way we were happy together far outweighed anything else."

They were married in November 2016; both Keens and Kun attended their wedding.

"I was rejoicing," says Kun. "Jim has never let his condition stop him from anything. He doesn't let it define him."

For that resilience, Jim credits his parents and the support from CHLA.

"I'm really grateful for everything I've been able to do," he says. "Now I just want to stay healthy, continue to build our careers and have a family. I just want to keep the good times going." 🦋



Jim and Brittany were married in November 2016.

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# TAKING ACTION ON ASTHMA



BY KATIE SWEENEY

*Pediatricians in the CHLA Health Network are helping children with asthma—and their parents—breathe easy.*

For one Los Angeles mom, going on a business trip had always meant coming home to find her son's asthma out of control.

But not anymore. Thanks to the new Asthma Action Plan provided by her CHLA Health Network affiliate pediatrician, her husband can now confidently manage their child's asthma in her absence. When she returned home from her latest trip, their son was breathing easy.

The family's experience is one of many patient success stories from the CHLA Health Network, a premier network of pediatricians affiliated with Children's Hospital Los Angeles. Launched in 2016, the network has quickly grown to more than 130 independent physicians caring for 260,000 patients across Los Angeles County and surrounding areas.

Selected for their outstanding reputations, affiliate physicians enjoy close access to the world-class specialists in the CHLA Medical Group and work to deliver innovative, seamless care aligned with the latest treatment protocols and research developed at CHLA.

"Our affiliate pediatricians are committed to providing the highest-quality, integrated care for their patients," says Bhavana Arora, MD, medical director for the CHLA Health Network. "The goal is to continually raise the level of care for children in the communities where they live."

## Empowering families

One area where the network is already making a difference is asthma care. Asthma affects more than 1 million children in California, nearly 60 percent of whom had an asthma flare-up in the last year—a situation that often sends kids to the emergency department.

"Many parents don't have enough knowledge about this condition," Arora notes. "They may not understand the importance of their child's medicines and how they work, or how to manage their child's asthma at home."

Participating pediatricians receive a wealth of resources to empower their patients—including materials to help parents and kids better understand asthma and its treatments, and a simple, one-page Asthma Action Plan outlining how parents should respond to specific symptoms at home.



Affiliate pediatricians also share best practices with each other and learn the latest evidence-based treatment protocols from CHLA specialists. In addition, CHLA Health Network staff provide training and support to help practices implement new asthma tools, including patient questionnaires, action plans and hands-on instructions for using inhalers.

"This education can make a world of difference for patients," says Cindy Loth-Wiggins, quality improvement coordinator for the CHLA Health Network. "Pediatricians are telling us that parents love it."

"The goal is to keep kids out of the hospital—and we are making progress toward that goal," says Arora.

## Well-child visits, anxiety and more

Asthma isn't the network's only area of focus. Some practices are working to increase and improve well-child checks. And the network will launch a new initiative in July to enhance care for children and adolescents with anxiety, depression and ADHD.

"The physicians choose what they want to focus on, and we support them," Arora says. "But the true beneficiaries are the children of Los Angeles."

**To find a CHLA Health Network affiliate pediatrician in your neighborhood, go to [CHLA.org/HealthNetwork](https://www.chla.org/HealthNetwork).**





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