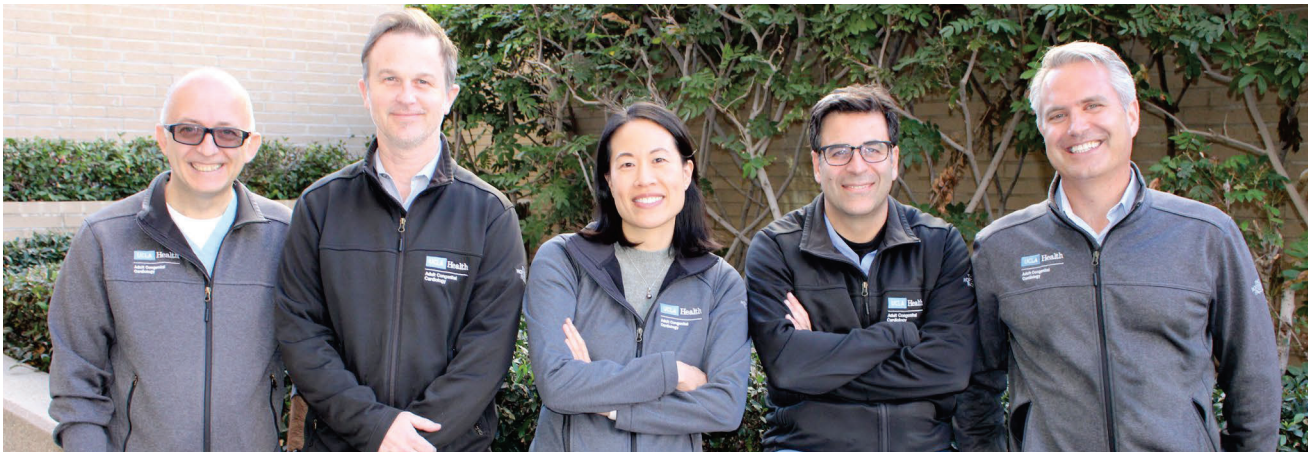
AHMANSON/UCLA ADULT CONGENITAL HEART DISEASE CENTER

NEWSLETTER WINTER 2024-25

UCLA Health | David Geffen School of Medicine

END OF YEAR GREETINGS FROM OUR DIRECTOR



The Ahmanson-UCLA Adult Congenital Heart Disease Center team pictured from left to right: Gentian Lluri, M.D., Ph.D.; Jeremy Moore, M.D.; Jeannette Lin, M.D.; Jamil Aboulhosn, M.D.; Leigh Reardon, M.D.

As 2024 comes to a close, the Ahmanson-UCLA Adult Congenital Heart Disease (ACHD) Center has had its busiest year ever, logging 8,082 outpatient visits so far. Since opening our dedicated ACHD clinic space on the 7th floor of the Vatche and Tamar Manoukian Medical Building in February, we have been able to streamline patient visits with a dedicated licensed vocational nurse and medical assistant. An on-site cardiac imaging suite with highly specialized sonographers has been essential for congenital echocardiograms.

In addition to our outpatient services, the ACHD team has performed 159 catheterization-based interventions and 72 cardiac surgeries and managed 38 high-risk pregnancies this year. Our team has also grown, with the addition of a new fellow and a physician's assistant.

The ACHD research program continues to rank among the busiest in the nation, contributing to a growing foundation of practice standards based on scientific evidence. This year, we are including UCLA Health patients in a clinical trial evaluating a new heart failure medication (see research and heart failure review below) and expanding clinical experience with a new transcatheter pulmonary valve.

We will commemorate the center's 45th anniversary in 2025. We also plan to recognize one of our earliest ACHD team members, Kevin Shannon, M.D., who will retire in July. Dr. Shannon defined and evolved the electrophysiology (arrhythmia) specialty for ACHD and is deeply admired and respected. We look forward to a fitting celebration in his honor.

Thank you, as always, for your continued interest and support. All of us at the ACHDC wish you and your families a happy, healthy holiday season!

Jamil Aboulhosn, M.D.
Streisand Chair in Cardiology
Director, Ahmanson-UCLA Adult Congenital Heart Disease Center
Professor of Clinical Medicine, David Geffen School of Medicine at UCLA

MEET THE NEW TEAM MEMBERS

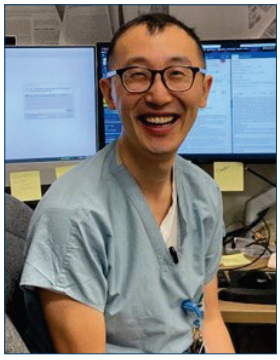
Mia Talamantes

Mia Talamantes joined the team in September as a physician's assistant (PA). She grew up in Los Angeles and completed her undergraduate degree at the University of Southern California before attending the Pace University-Lenox Hill Hospital PA program. Mia began her career at New York-Presbyterian Columbia University Irving Medical Center, focusing on interventional cardiology.

Mia's personal journey with congenital heart disease began at UCLA Health when she was diagnosed with a congenital heart defect at birth.

At age 8, she underwent surgery conducted by renowned cardiac surgeon Hillel Laks, M.D. This experience shaped Mia's career path and factored into her return to California to specialize in ACHD. Working with the ACHDC is her dream come true, and she looks forward to seeing her career evolve in this field. Mia is excited to contribute to the team and make a positive impact on the lives of patients.

Mia is a proud dog mom to a lovable mini dachshund and enjoys exploring thrift stores and indulging in guilty pleasures like watching *Love Island*. Please join us in welcoming Mia!



Hans Gao, M.D.

In July, Dr. Gao joined the ACHD team for a two-year fellowship. He started his medical training at the Warren Alpert Medical School of Brown University then conducted his residency at the University of California, San Francisco. Dr. Gao completed a cardiology fellowship at Cedars-Sinai Medical Center, where he had excellent mentors who introduced him to ACHD as a specialty. This led to a one-month visiting fellowship rotation at the ACHDC, during which Dr. Gao was nurtured by the team's cardiologists. In this time, he also developed his love of the patient population and the life-long patient-physician relationships that are unique to this cardiac specialty.

Dr. Gao's openness to learning from his patients and not missing "the forest for the trees" makes him easy to like and a welcome addition to our ACHD team. He understands the intellectual rigor of congenital cardiac pathophysiology and has eagerly applied himself for the long learning curve necessary to become an expert in this field.

On a personal note, Dr. Gao loves European-style boardgames and previously designed one based on H.P. Lovecraft lore. He loves hiking as well as reading science fiction and is currently a fan of the *Culture* series by Iain Banks. Dr. Gao reports that he has not yet encountered a food item that he could not stir fry, though he admits to some variance in palatability.

THE SUPER-FONTAN: UCLA STUDY DEFINES THIS OVERACHIEVING GROUP

The Fontan surgical procedure has become the mainstay for patients born with single ventricle physiology over the past 40 years. Long-term outcomes are variable, and exercise capacity is typically limited and worsens with each passing decade. Those with poor exercise performance tend to confront a higher risk of death, transplant and hospitalization. However, a subset of Fontan survivors enjoy nearly normal exercise capacity, fewer symptoms and complications, and improved prognosis.

The term "Super-Fontan" has been used to define these patients, and research efforts are now dedicated to identifying what factors contribute to this enviable Fontan survivor subgroup. If a combination of exercise and lifestyle interventions can protect children growing up with Fontan physiology from early mortality, it is well worth a detailed retrospective examination of what makes these Fontan adults "super."

Dr. Aboulhosn and his research team looked at 195 of their adult Fontan patients, and 27 were considered "high performers" based on cardiopulmonary exercise testing. Compared to their more typical Fontan counterparts, these high performers had significantly lower body mass indexes, were younger in age at the time of their Fontan surgeries and exercised more regularly and at a higher intensity.

Researchers then dug deeper by examining available diagnostic tests, including catheterization hemodynamic data, liver diagnostics and laboratory tests. Overall, the Fontan high performers demonstrated higher oxygen saturations and higher albumin levels that indicate better liver function. Interestingly, there was no difference in age of these high performers, pointing to the long-term benefits of maintaining a healthy body weight and regular exercise in aging Fontan patients. Catheterization data obtained in eight high performers showed lower Fontan pressures compared to their counterparts. Liver fibrosis is common in adult Fontan patients, and high performers were more likely to have less evidence of this compared to controls.

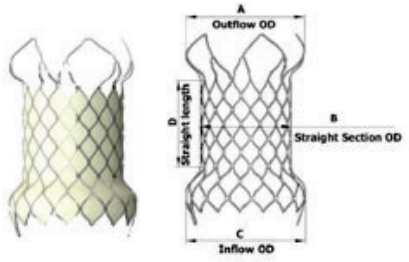
These findings demonstrate the importance of modifiable risk factors such as weight and exercise, but the small sample size makes accounting for all associated variables difficult.

Read the full study at **internationaljournalofcardiology.com/article/S0167-5273(23)00973-7/fulltext**.

RESEARCH TRIALS ENROLLING UCLA HEALTH ACHD PATIENTS

Proteus Trial for Transcatheter Pulmonary Valve Implantation

This is a prospective FDA-mandated trial of the Venus P valve for treatment of patients with pulmonary valve regurgitation. The Venus P valve is a self-expanding porcine transcatheter pulmonary valve platform that is placed in the femoral vein or the jugular vein without the need for open heart surgery. This valve comes in 10 different sizes, providing more options to better fit individual patients. The study is currently enrolling at UCLA, where Dr. Aboulhosn is the principal investigator and will be the implanting physician. To learn more about this study, please ask your cardiologist or email our research team at **ACHDResearch@mednet.ucla.edu**.



Illustrations of a percutaneous pulmonary valve

Table 1. Dimension when fully-expanded of Percutaneous Pulmonary Valve

Model	Specification	Diameter (Unit: mm)			D (Unit: mm)
		A	B	C	
L28P	P28-25	38.0	28.0	38.0	25.0
	P28-30				30.0
L30P	P30-25	40.0	30.0	40.0	25.0
	P30-30				30.0
L32P	P32-25	42.0	32.0	42.0	25.0
	P32-30				30.0
L34P	P34-25	44.0	34.0	44.0	25.0
	P34-30				30.0
L36P	P36-25	46.0	36.0	46.0	25.0
	P36-30				30.0

Fondap Study for Fontan patients with Heart Failure

The Fondap study is enrolling UCLA Fontan patients in collaboration with the Johns Hopkins Adult Congenital Heart Disease Center. This study is looking at the potential benefits of dapagliflozin (trade name Farxiga) in patients with Fontan physiology. Dapagliflozin is the newest medication used in patients with acquired heart failure and is one of the first medications to be effective in patients with heart failure with preserved ejection fraction. As such, this medication is one of the main tools used today in heart failure (see heart failure review below for more details). As with many other medical therapy treatment multicenter trials, ACHD patients were excluded from the initial trials of these medications. Therefore, further study is required to test how effective each medication is in complex ACHD patients, particularly single ventricle patients with Fontan physiology.

To learn more, you can discuss this with your cardiologist or email our research team at **ACHDResearch@mednet.ucla**.

PATIENT STORIES

JONAH, Age 37, Transposition of Great Arteries

Jonah and his dad

I'm afraid this story is going to be a tad short and sweet, but that's a testament to the excellent care from the fine folks at the ACHDC. I was born in 1987 with a transposition of the great arteries, which fortuitously came after a number of surgical advances earlier that decade.

I can't say I remember much of the corrective surgery, given that I was about three days old at the time, but — in the hands of the incomparable Dr. Laks — it went quite well.

In my early teens, I had a follow-up procedure to remove some internal scar tissue — again, a success. I've always been very fortunate in terms of how well my care and recovery have gone. I've never felt limited by the condition, never had issues with sports or physical exertion and have always been able to live a pretty darn regular life besides sporting a cool scar on my chest.

My father also has a corrected heart condition, tetralogy of Fallot, for which he receives care from UCLA Health, meaning the ACHD team has to put up with terrible jokes from two members of our family. (I'm pretty sure whatever we're doing in the photo of the two of us was his idea.) But I do think the fact that we both go to UCLA Health, and that Dr. Aboulhosn and the staff all graciously put up with both of us, really speaks to the breadth of care at the clinic.

The only little side note I would add is that, once I entered my mid-20s, I started to have some back and spine pain, which a number of physical therapists have suspected is related to the earlier surgical procedures, including the lingering scars. So, if you've also had some symptoms of that nature, I'd be happy to compare notes about what sort of stretches and strengthening are helpful. Send me an email at **ucla@jbjbjb.anonaddy.com**.

DUSTIN, Age 37, Tetralogy of Fallot with Pulmonary Atresia

Two combatants wearing thick, padded medieval jackets, protective armor covering the neck and hands and fencing helmets circle each other while wielding large two-handed steel longswords. One would be forgiven for thinking the confrontation is at a renaissance era

re-enactment show. However, this is a normal Thursday night in San Jose, Calif., for practitioners of historical European martial arts. I am one of the two combatants. My opponent has their sword down low on their left side. Good, I expect they will attack with an ascending cut up, and I move my longsword to my left side over my shoulder. This is known as "Posta di Donna" in Italian longsword traditions.

We have been sparring for some number of minutes now. My adrenaline is flowing, and my heart is pumping blood in my veins. My opponent moves as I expected, throwing their cut upward, and I swiftly step to my right side while moving my sword to the middle of my body. I then move my sword out, pull my pommel up and force the blade downward to displace my opponent's sword with a satisfying "ting!" I push my pommel back down, and the tip of my sword is now at my opponent's chest. This bout is mine. However, winning the exchange is no longer on my mind. I am forced to recognize that my heart is beating very quickly, and suddenly the fight is in my mind as I battle fear. Is my heart beating a "normal" fast rate? Did I push too far, and this is something more serious?

While my opponent gets ready for the next exchange, I am in my own head, no longer enjoying the moment but spiraling into negative thinking where I fear for the worst. For me, this thought process is a constant problem, especially when physically exerting myself. My initial reaction is to stop the fight, stop something that I love doing. How did I get like this? When it comes to sparring with fear, hopefully sharing and reflecting on my story will help.

I was born in January 1987. My parents were young when they had me, and I am their first child. When I was about 3 months old, my parents recognized that I was sick and took me to the pediatrician. I wasn't doing well, and a congenital problem with my heart was discovered. I can't imagine how scary that must have been for my parents to hear.

Over the next few years, I had a series of three corrective surgeries. Aside from the surgeries, I feel that I had a fairly normal childhood. A lot of credit is due to my parents, who did not shy away from letting me participate in activities I was interested in. I was involved with scouting, camped a lot and even played a year of soccer. I would never consider myself the best at any physical activity, but I was able to keep up with my friends and hold my own.

In the summer between junior high and high school, I needed to have another open heart surgery to replace the donor pulmonary artery that I had outgrown from previous surgeries. The surgery went well, and I even recovered quickly enough to go to a water park before fall started. While I was in a lazy river at the water park, I felt that my heart was beating funny and had to sit down. Concerned, my dad took me to the park's first aid station, and the first responders discovered that my heart rate was irregular and very fast — like over 200 bpm fast. I was rushed to the nearest hospital and ended up getting electrical cardioversion to correct the arrhythmia.

What happened next was an ablation procedure, and an internal cardiac defibrillator (ICD) was

implanted. I carried on the rest of the school year and, in the summer, went on a long-term camping trip with the Boy Scouts near Big Sur, Calif. While I was attempting to pass a swim test in a very cold lake, my heart rhythm started to feel odd again. Back at the dock, my new ICD delivered a small electrical shock and then several more with increasing power. I have never been as scared for my life as in that moment. I was airlifted to the hospital. Later, I found out that my heart rate naturally went above the ICD's programmed target rate and that it delivered therapy as a result. I understand now, having the ability to look back, that this was a moment that changed my life. Eventually, I developed an anxiety about activities that would get my heart rate up and started having panic attacks with some regularity. It has been a challenge to learn how to live with that trauma and how to adapt and be brave.

Circling back to the sparring match, I briefly hold up my hand to stop the fight so I can re-assess what my physiology is actually telling me. It hasn't always been easy to deal with the fear in these situations, but over time I have had a lot of support and love from family and friends in order to learn methods to work through the panic. In moments of exercise, like this sparring match, it is much easier for me to remember that what I am feeling while exerting myself is different but also normal. I also found that if I have something to hold on to, like a sword, it helps to ground my attention with tactile feeling of something outside my body. In this case, it gets me back into the match, and I am able to beat the fear, keep up with my opponent and enjoy what I love about the art of sparring.

HOW YOU CAN HELP

We reach out every year to remind our most dedicated supporters that, for those lives touched by ACHD, there is an amazing opportunity for charitable giving. All funds donated to the center are used to forward the science of ACHD with valuable research, in addition to securing funding to train future generations of care providers. Beyond that, there are opportunities to support signature research efforts or technological advances that are crucial to the interests of our most complex patients and those who love them. Most of us aim to make year-end charitable donations once all our mandatory financial obligations are met. But even small monthly donations are important for the ongoing work of our research and clinical goals.

A smaller subset of our patient population may benefit from focused wealth planning and attention to tax-deferred strategies with respect to estate planning. Since our ACHD patients are living longer and reaching all the financial milestones important to retirement, charitable giving is a meaningful option for a planned legacy at UCLA.

For more information about planned giving or to explore opportunities for philanthropy, please contact:
Lindsey Walton
Director of Development
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424-946-6102 | LSWalton@mednet.ucla.edu

Understanding "Heart Failure" and New Medications That Are Changing Outcomes

Some of you are familiar with the term "heart failure," which has the unfortunate negative connotation that those afflicted have reached the "end of the road." The reality is much less dismal in the current era of advancing medication options, and the expanding acceptance that early and ongoing screening and proactive treatment is changing the trajectory of heart failure.

Heart failure, also called congestive heart failure (CHF), refers to the symptoms caused by some degree of impairment in ventricular (pump) function. The most common symptoms are shortness of breath and edema (water retention), but these can vary from mild to severe, and some patients may not have any clinical awareness of them. The impairment in ventricular function varies widely and refers not only to the squeezing power of the ventricle (ejection fraction), as this can be normal in some with CHF, but also to the compliance of the ventricular muscle.

An echocardiogram is a valuable diagnostic tool to evaluate several heart function parameters including ejection fraction. A simple blood test called a brain natriuretic peptide can also be used for screening and surveillance of heart failure. Beyond the variables of ventricular function and symptom burden, adding the heterogeneity of congenital heart disease to the mix makes heart failure one of the greatest management challenges in the ACHD population, in addition to being the most common cause of heart-related mortality.

The field of ACHD and managing heart failure has always revolved around making inferences from large-scale clinical trials focused on acquired heart disease patients. As is the case in any advancing science in the world of ACHD, interpreting large clinical trials and making careful individualized decisions about their utility in ACHD patients is why these patients need to be in hands of highly specialized ACHD cardiologists. These specialists are tasked with understanding complex physiology, applying evidence-based practice stemming from national ACHD management guidelines and maintaining a rigorous learning curve based on smaller research trials in subsets of the ACHD population.

Until the late 1980's, treatment of heart failure was focused on symptom management, but the emergence of angiotensin-converting enzyme inhibitors in 1987, combined with beta blockers, changed that focus to improving survival. In 2015, the medication sacubitril/valsartan (trade name Entresto) changed the playing field for those with more severe heart failure by improving ventricular function and decreasing both hospitalizations and deaths.

In 2020, sodium-glucose cotransporter 2 (SGLT2) inhibitors, commonly known by the trade names Farxiga and Jardiance, became the most recent class of drugs to get FDA approval for the treatment of heart failure. SGLT2 inhibitors work by stimulating the kidneys to remove excess glucose and sodium from the body and were first approved for treatment of type 2 diabetes. The effect of removing excess sodium was found to be significantly beneficial to heart failure patients, leading to further study in this population.

Large clinical trials of more than 8,000 patients confirmed that SGLT2 inhibitors significantly

reduce the risk of heart failure events and hospitalizations and of cardiovascular death. Additional benefits include reducing the decline in kidney function and managing blood glucose levels in type 2 diabetics. As in any large clinical trial, ACHD patients were not part of this original patient cohort, so additional studies are underway. In April 2024, the *Journal of the American College of Cardiology* published the experience of 174 patients from four major European ACHD centers with SGLT2 inhibitors. Researchers found a significant reduction in heart failure hospitalization between six months before and six months after starting SGLT2 inhibitors. Side effects (the two most frequent being low blood pressure and urinary tract infections) resulted in 10% of the cohort stopping the medication.

SGLT2 inhibitors are now strongly recommended in the acquired heart disease population for heart failure with reduced ejection fraction and have been included in national guidelines. Farxiga can be used in combination with the drug Entresto if tolerated, along with a beta blocker such as carvedilol. Small ACHD studies are encouraging and show this class of drug to be generally well tolerated, so the benefit of reduced hospitalizations and deaths far outweigh the risk of side effects.

As noted in the research trial announcement above, the ACHDC has embarked on a study in collaboration with Johns Hopkins ACHD Center to evaluate the benefit of SGLT2 inhibitors in Fontan patients. Consider talking to your cardiologist about whether this class of medication is right for you. It's a good opportunity to learn more about your pump function/ejection fraction and to become better informed.

ACHDC Transition Program Update

The UCLA ACHD Transition Program is a comprehensive educational program for young adults with congenital heart disease. Led by Nurse Coordinator Nicole Antonville with Leigh Reardon, M.D., Supporting Health Independence for Families Transitioning — or "SHIFT" — to ACHD from Pediatric Cardiology will help patients have a smooth transfer from pediatric to adult care.

ACHDC has also collaborated with ACHA to provide a support group for the emotional aspects involved with this change. During the monthly group sessions, co-moderated by Antonville and nurse Rosyln Rivera, young adults share their experiences. Topics have included relationships to pediatric providers, creating a new relationship with adult providers, feeling empowered to take over medical care from parents, going to college and talking to friends about heart conditions. While changing from a pediatric cardiologist to an adult provider can be hard, sharing experiences enables people to feel more comfortable with the transition process and less alone.

A Tribute to Ken Boyko (1951-2024)

We mourn the loss of Ken Boyko, a beloved, longtime patient. Ken was born with tetralogy of Fallot at a time when there was little hope for survival past adolescence. Ken first came to UCLA Health in the early 1980's, seeking the expertise of Joseph Perloff, M.D., and learned about the new clinic specializing in adult survivors with congenital heart disease. He was part of the evolution of the ACHDC, having two major reconstructive heart surgeries and ultimately a heart transplant in the expert hands of Dr. Laks. Although Ken had a clear personal interest in furthering the science of ACHD, his devotion to Dr. Perloff, Dr. Laks and UCLA motivated meaningful philanthropy and, ultimately, a role as a founding member of the center's board of advisors.

Through a combination of fortuitous timing and true grit, Ken prevailed and enjoyed a full life, never letting congenital heart disease define him. He was a successful entrepreneur, had a loving marriage and was a father. He lived long enough to see both his children get married and to become a grandfather.

We are grateful for the important role Ken played in the evolution and success of the Ahmanson-UCLA Adult Congenital Heart Disease Center. Because of him and the science that advanced due to so many pioneering early patients, many children born today with tetralogy of Fallot can expect to live long, healthy lives. Perhaps there is no better fitting tribute to a life well lived. Thank you, Ken.

To donate in his memory, please visit the Kenneth Boyko memorial site at **engage.ucla.edu/fundraiser/5671346**.

UCLA ACHD PATIENT AND FAMILY CONNECTIONS SUPPORT GROUP

Through the UCLA ACHD Patient and Family Connections meetings, the center aspires to connect ACHD patients and their support system members with other patients to provide effective peer support to this growing population. Connections meetings take place virtually via Zoom on the last Thursday of each month. ACHD social worker David Highfill, LCSW, facilitates the group. He is also an ACHD patient who underwent a modified Fontan procedure in 1990 and received a heart transplant in 2012, both with UCLA Health. Connections meetings are open to all ACHD patients and their family members. To obtain the Zoom link for the monthly Connections meeting, please contact Office Manager Yvonne Jose at 310-825-2019 or YJose@mednet.ucla.edu.

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SEASON'S GREETINGS FROM THE ACHDC TEAM!

ACHD TEAM AND CONTACT INFORMATION

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Kevin Shannon, M.D.
Electrophysiology

Kalyanam Shivkumar, M.D., Ph.D.
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