

Ahmanson / UCLA Adult Congenital Heart Disease Center

FOR THOSE TOUCHED BY CONGENITAL HEART DISEASE

FALL 2012

Center Updates

First Annual Los Angeles Congenital Heart Walk a Success

The Ahmanson/UCLA Adult Congenital Heart Center was a proud cosponsor of the first annual Congenital Heart Walk held at the Johnny Carson Park in the studio district of Burbank, CA on July 15th. Over one hundred participants, including adults and children with congenital heart disease, their families, doctors, nurses and supporters turned out on a beautiful Sunday morning to join co-chairs Dr. Piper Calisante and Barbara DeMaria for the growing number of cities across the country in the campaign to promote awareness of congenital heart disease and raise funds for research, education and advancement of medical care.

Team Ahmanson/UCLA was one of the top three fund raising teams in the event, which raised over 40,000 dollars. The funds will be shared by the Adult Congenital Heart Association and the Children's Heart Foundation. As Ms. De Maria stated, "We are excited to finally have a walk of our own, uniting our community to fight congenital heart disease and support the missions of these two wonderful organizations."

For more information, visit www.congenitalheartwalk.org, or the Congenital Heart Walk Facebook page at www.facebook.com/CongenitalHeartWalkLA

The second annual Los Angeles Walk is tentatively slated for Spring 2013, so keep your calendars open!



Above: Team Ahmanson/UCLA



Dr. Leigh Reardon (left)
and Dr. Jamil Aboulhosn (right)

Insurance Update

Blue Shield has a new contract with UCLA!

The termination of the UCLA/ Blue Shield contract in January, 2012 has been challenging for the ACHDC patients and providers. We are pleased to report that effective September 1, 2012, the contract between UCLA Health System and Blue Shield will be reinstated. Patients who are insured by Blue Shield will again be able to receive in-network care from UCLA physicians and hospitals. Blue Shield members may immediately begin scheduling appointments at UCLA for dates of service after September 1, 2012.

Website Update

New ACHD Website:

Visit our Patient Resource Page
www.uclahealth.org/ACHDC

We have now opened a new section on our "Patient Resource" page of our website titled: "In Your Own Words". This section is dedicated to contributions from patients, which can include a personal story, or thoughts about how congenital heart disease has impacted your life. Posting content in this section is at the discretion of the Center's Faculty. Feel free to send your story to Pam Miner, NP at pminer@mednet.ucla.edu.

[Faculty and Contact Information](#)

Faculty

Ahmanson/UCLA ACHDC Faculty

Jamil Aboulhosn, MD

Director, Interventional Cardiology

John Child, MD Emeritus Director

Leigh Reardon, MD Congenital Cardiology

Jeannette Lin, MD, Congenital Cardiology

Daniel Levi, MD Interventional Cardiology

Kevin Shannon, MD Electrophysiology

Jeremy Moore, MD Electrophysiology

Kalyanam Shivkumar, MD Electrophysiology

Ravi Mandapati, MD Electrophysiology

Hillel Laks, MD Cardiothoracic Surgery

Brian Reemtsen, MD Cardiothoracic Surgery

Pamela Miner, NP Nurse Practitioner

Linda Houser, NP Nurse Practitioner

ACHDC Staff

Jennifer Doliner, RN Nurse Coordinator

Yvonne Jose Administrative Manager

Contact Us

How to Contact the Center

Patient Calls (310) 794-9629 or (310) 825-9011

Nurse Coordinator (310) 794-5636

Fax Numbers (310) 825-6346 or (310) 825-9012

Email: achdc@mednet.ucla.edu

Website: www.uclahealth.org/ACHDC

Information & Resources

Adult Congenital Heart Association (ACHA)

Website: www.achaheart.org

Call (888) 921-ACHA

Email: info@achaheart.org

International Society for Adult Congenital Heart Disease (ISACHD)

Website: www.isachd.org

Faculty Profile

Dr. Leigh Christopher Reardon

Dr. Reardon joined the faculty of our Center in July 2011, after completing his fellowship in pediatric cardiology and advanced training in adult congenital heart disease at UCLA. His residency at Cedars Sinai Medical Center prepared him for a career caring for patients across the lifespan, since he specialized in both pediatrics and internal medicine, and ultimately became board certified in both. His interest in congenital heart disease was also personal – he was born with a congenital heart defect of his pulmonary valve, and required open heart surgery at the age of 5.



Dr. Leigh Christopher Reardon

the San Diego Foundation for Cardiovascular Research and Education. He then obtained his medical degree at Virginia Commonwealth University in Richmond, Virginia where he was awarded the Ellen A. Holiman scholarship and served on the medical school admissions committee. In 2010, Dr. Reardon was given the Fellow Teaching Award in 2010 by the UCLA pediatric residency program for his dedication to resident and medical student education

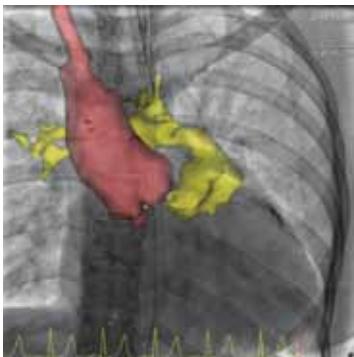
Now having completed his first year with our Center, Dr. Reardon continues to focus his efforts on the care of adults and children with congenital heart disease. Dr. Reardon is also spearheading the Transitional Care Program for Adolescents and Young Adults with Congenital Heart Disease. This is a renewed effort to confront the challenges of transitioning patients from the pediatric to adult setting. Working hand-in-hand with the program coordinator Mary Canobbio, RN, MN, FAAN – the reinvigorated transitional care program will offer tailored and developmentally appropriate care to patients age 12-20. Building on his experience with pediatric heart transplant and heart failure, Dr. Reardon is taking a particular interest in the adult congenital patient with worsening heart failure.

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3-D Imaging

Advanced 3-D Imaging Techniques Revolutionize Congenital Catheterization Procedures at UCLA

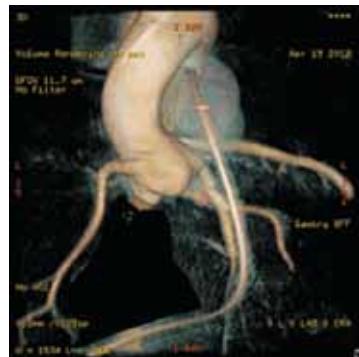
Cardiac catheterization is widely performed for diagnostic and interventional purposes in patients with congenital heart disease. Cardiac catheterization was first performed in 1929 and came into wide clinical use in the 1950's and 1960's with the availability of X-ray fluoroscopy to help guide catheters and image dye injected into the heart and circulation. Initially these techniques were used for purely diagnostic purposes. Over the past 40 years, a host of innovations have catapulted catheterization techniques to the fore-front of interventional treatment. Many patients who would have previously had surgical repairs are now more likely to undergo catheter based intervention, these include patients with ASD, pulmonary stenosis, prosthetic pulmonary valve dysfunction, coarctation of the aorta, among others. Improvements in catheter design, development of balloons and stents, and development of nickel/titanium devices, among many other advances have allowed this revolutionary transformation in the care of the patient with congenital heart disease. Catheter based interventions have a number of advantages when compared to traditional surgery, including shorter hospital stay, smaller skin incision, less pain, quicker healing, and less bleeding.



3-D MRI overlay live on the X-ray image to guide Melody valve placement.

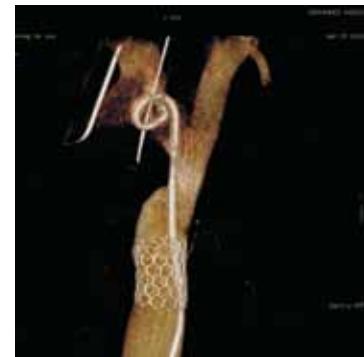
High quality imaging is of fundamental importance to the performance of catheterization procedures. The more and better we are able to visualize a problem, the more likely we are

to appropriately understand and treat it. The past few decades have seen improvements in X-ray fluoroscopic image quality and the wide use of intra-cardiac and trans-esophageal echocardiography to help guide catheter based interventions. Cardiac MRI and Cat Scan imaging is often used in pre-procedural planning but until recently could not be used to guide interventions in real time. Essentially, we were still working mostly in two dimensions with X-ray and ultrasound image guidance, and unfortunately, we were unable to actively use our best images from CT and MRI in real time.



3-D reconstruction of aortic root angiogram with simultaneous inflation of a large diameter balloon within a pulmonary artery conduit. This is performed to test for compression of the coronary arteries, no compression is noted here.

In late 2011, the Ahmanson Foundation graciously awarded the Ahmanson/UCLA Adult Congenital Heart Disease Center a sizeable grant to upgrade the congenital catheterization laboratory with the latest GE Innova-3D and Heart Vision workstations, hard-ware, and software to allow for 3-D imaging and procedural guidance using MRI and CT techniques. The upgrades were installed and went into use in April of 2012 and the 3-D imaging techniques are being used regularly. After working in 3-D for just a few months, we cannot help but feel that we are in the era of yet another revolution in the care of the patient with congenital heart disease. We are actively collecting data on all studies performed and developing an



Rotational angiography of the aorta with 3-D reconstruction demonstrating residual narrowing of the thoracic aorta, stent migration inferiorly, and an aneurysmal outpouching of the aorta at the superior edge of the stent.

understanding of when and where these techniques are most necessary and most useful. In this regard, we are helping spearhead this 3-D revolution, and we have the generosity of the Ahmanson Foundation to thank for that. We hope that these advances in imaging will help make catheterization procedures even safer for our patients.

Research in Progress

Inhaled Iloprost for the treatment of pulmonary hypertension in adults with CHD: three-month trial of inhaled medication, self-administered six times a day.

Bosentan for reduction in pulmonary pressures in adolescents and adults with Fontan/single ventricles: three-month trial of oral medication twice a day

Gene study for patients with Shone's syndrome: one-time blood test

For more information regarding any of the above studies, please email achdc@mednet.ucla.edu or call (310)794-5636.

Three UCLA Heart Patients Share Their Stories



Vanessa, age 28 yrs

When new parents consider moving to a small rural town away from the city, they consider factors like crime rate, schools, and having a yard. One terrifying reality no parent wants to be confronted with is their new home's proximity to a top-rated pediatric cardiologist. But that is what my parents were forced to do as they raced down Highway 101 towards foggy San Francisco and UCSF, while I was put in a helicopter making my own, faster trek towards the famed hospital.

No one noticed that I was slightly blue in the beginning, and everyone just thought I was a great baby because I never cried. It took until my two-week wellness check-up for anyone to realize there was a problem. A major one.

I was born with Transposition of the Great Arteries, a Ventricular Septal Defect and Patent Ductus Arteriosus. However, unlike most patients, I never received the arterial switch operation. Instead, in 1984, Dr. Paul Ebert repaired the Patent Ductus, and performed a Senning procedure that rerouted the blood through the upper chambers of my heart while leaving the transversed arteries in place. My heart then stayed in unnaturally-working order for the next twelve years, and I was left to grow and play to my heart's content.

The one aspect of my childhood that I am most thankful for is the support I received from my family. My parents have been there every step of the way, even if that meant my dad had to sleep in our Suburban in the UCSF parking garage for six weeks because the hospital only allowed one parent to sleep in a hospital room. But more than that, my parents never treated me as "breakable". I was allowed to climb trees, play soccer, and go to sleepovers. Though I would get tired much

quicker than all of the other kids, I never felt different. I will be eternally grateful that I was not raised in a plastic bubble.

There have been bumps along the way, though. In middle school, I needed two more surgeries to place pulmonary bands to help strengthen one side of my heart in hopes that I could one day undergo a more radical "late" arterial switch operation. Luckily the pulmonary artery banding helped my heart so much that my badly leaky valve and weakened heart chamber improved enough that no further surgery warranted the risk. I resumed an active lifestyle with very few limitations. At age twenty-five, I made several trips to UCLA for ablations to put a stop to my fast heart rhythm, called Supraventricular Tachycardia.

I have been given as much care and expert treatment at UCLA as an adult as I was given at UCSF as a child. But the one thing that has changed since aging into the adult program at UCLA is that for the first time, I feel heard. My concerns, my assessments, and my gut feelings about what is going on in my own body are taken into account. Finally, I feel like I have a voice, and that it counts in the discussion.

Because my parents have always trusted me not to push my limits too hard, and therefore never held me back, my hobbies include rock climbing, motorcycle racing, and my newest – surfing. I have been working in the film industry for the past four years doing special effects and stunt work, and my cardiologist, the one-and-only Dr. Kevin Shannon, says that I am one of the healthiest, physically capable people that he's ever seen with my condition. I don't say this to boast, but instead to show others in my situation that you don't have to live life scared of what might happen, or by the rules of what you can't do.

Jeanie, age 63 yrs

I was born in 1949, a bit premature. I was diagnosed with a slight heart murmur which doctors advised my parents I would probably outgrow. The murmur was not mentioned or detected again until 1963 when I underwent a physical to enroll in high school. I was sent to UCLA at age fourteen for a cardiac catheterization, however results were inconclusive.

I continued with my life, unfazed by any medical issues until the birth of my fifth child in 1979. During some routine testing following childbirth, my internist wanted to follow up a little more on the murmur. After more intensive testing available at that time, I was diagnosed with a benign or physiological murmur. A sound that was present, but had no known cause or effect on my overall health.

I continued to live my life as if I had no health issues. At age 42, while undergoing a routine physical, I was referred for the first time to a cardiologist. It was then that I was diagnosed with "congenitally corrected transposition of the great arteries", which means my pumping chambers were reversed as well as the valves that drain into those chambers. I still had no physical symptoms of heart disease, but I began my journey with cardiologists, ultrasounds, extensive testing and medications.

Within a few years, I would undergo valve replacement. I have since had two pacemakers, an ablation and numerous cardiac procedures. I am now 63 years old and while I am extremely grateful for the advancements and treatments in medicine that have enabled me to live a longer and more comfortable life since 1995, I cannot help but think I was somewhat lucky that my condition was not diagnosed in my youth. I may have lived my life very differently. I may not have had the children that I have and therefore not my eight grandchildren, whom I adore. I may not have enjoyed or attempted the many experiences that I did. That being said, I firmly believe in early detection and prevention.



I believe that all of us benefit with continued research and new skills in the treatment of all

congenital and acquired disorders. I thank the staff at the UCLA Adult Congenital Clinic for their continued efforts. I have always been treated with respect, efficiency and the highest degree of competence from the entire staff, professional through clerical. I believe they make it possible for a better quality of life for many of us.



Rachael, age 30 yrs

There is a fine art to maneuvering through life with a complex congenital heart defect, and while it'll be a while yet before anyone starts calling me Rembrandt, I do all right for myself. Born with transposition of the great arteries and ventricular septal defect with a heart that sits center-right stage (we're kind of a showboat), and has gone through both a Fontan operation and a pacemaker, I am not your typical cardiac patient. I'm slightly bombastic, a bit cheeky and more than a dash vainglorious. Thirty years of being poked, prodded, studied and talked at has left me just a touch defensive and I have worked tirelessly to construct a perfectly hardened exterior.

I struggled greatly to come up with a topic to write about because the possibilities seemed endless, with the only goal being that I didn't want this to wind up being a piece of fluff, painting an image of myself I do not necessarily resemble all of the time.

On one hand, I wanted to be irreverently humorous, something I can excel at given the right mood and circumstance. Making people

laugh has been a not-quite altruistic goal of mine, as I use laughter to deflect the seriousness of my health. Even amidst my first full-blown episode of tachycardia, which occurred during school hours in the first grade, I was cracking jokes as the nurses office became ground zero of frantic phone calls, panicked voices and forced calm for my benefit.

Yet another direction I debated was that of profound earnestness; philosophizing the various coping techniques my peers and I adopted, my own being in the form of a blog. It is easier to mentally digest the chaos my life can be caught up in when I have an outlet to, pardon my crudeness, vomit up my emotions. All of the preposterous, hyperbolic, vindictive and utterly raw thoughts and feelings I possess have a special place of their very own. It may not be agreeable for some, but for me it's the ultimate catharsis.

Waffling between these options has forced me to accept and now openly admit a hard truth; a truth I realized once I started volunteering with Camp del Corazon and again when I attended the ACHA conference last year. I keep up the hardened exterior to mask my vulnerability. Not unlike a wounded tiger, I sit alone and surly, watching the rest of the jungle from my dark corner, the façade of pride and judgment. I want to partake, not just the philosophizing joker with a bag of tricks designed to twist your attention from my wounds, but keeping a certain level of detachment has helped me not let my CHD define me as a whole person.

So maybe I'll spin a grandiose tale about butting heads with a medical official, or repeat something funny that transpired with TEAM RACHAEL! (my medical entourage), but you may not see the true depths of my grievances; the isolation of my childhood, the struggles of a teenager who could not rebel even as her friends did, the young adult who struggled to find her footing in adulthood, and the woman who continues to mourn the loss of her uterus despite not living with regret about making the choice to have it removed in the first place. No, you won't see these things because I am too quick with my slight of hand. Did you see what I did there?



How Can You Help?

The Ahmanson/UCLA Adult Congenital Heart Disease Center (ACHDC) relies in large part upon donations in order to meet its needs to pursue the goals of the Center. Tax-deductible contributions made to the Center directly support:

- Patient programs focused on enhancing quality of life
- Research programs aimed at improving life expectancy
- Training programs integral to preparing future providers to carry on the highest level of care

You can learn more about how to support the ACHDC by visiting www.uclahealth.org/achd and clicking on "ways to give."

You can also call (310) 825-2019 if you would like to discuss specific gift options, or to get more information.

Spotlight

The Fontan Operation

What is it?

The Fontan operation is a surgical procedure performed for single ventricle physiology. The normal heart has two pumping chambers—one to pump blood with oxygen to the body, and one to pump blood without oxygen to the lungs. Patients with single ventricle physiology have hearts with only one effective pumping chamber, and this results in mixing of the two circulations (blood with and without oxygen), leading to an overall lowered blood oxygen content. Uncorrected, this leads to long term cyanosis (or blueness), and associated issues such as poor exercise ability, elevated blood pressures in the lungs, abnormally high red blood cell counts and bleeding disorders. The Fontan operation allows for separation of the two circulations by re-routing oxygen poor blood from the body directly to the blood vessels that go to the lungs (the pulmonary arteries), and allows the single ventricle to receive and pump only oxygen rich to the body. Blood reaches the lungs by passive flow, not with the assistance of a pumping chamber. This creates a more normal circulation, and reduces the complications described above.

How is the operation performed?

The first Fontan operations involved directly connecting the right atrium (the heart chamber that receives all of the blood without oxygen back from the body) to the pulmonary artery. This was called a right atrium to pulmonary artery Fontan (or a “Classic” Fontan). Over the years, modifications of the operation have developed in order to exclude the right atrium from the circuit, since it was found that over time this chamber became very stretched out and enlarged, leading to problems with abnormal heart rhythms and blood clots. The two types of Fontan operations done today that effectively do this are the lateral tunnel Fontan and the extracardiac Fontan, and are usually performed at around 4 years of age. A procedure known as a Glenn shunt often precedes this, and is usually done at around 6-12 months. Many patients with the “classic” type of Fontan operation have undergone conversion to either the lateral tunnel or (more commonly) the extracardiac Fontan in order to decrease the complications associated with the older operation.

How is the Fontan circulation different from normal circulation?

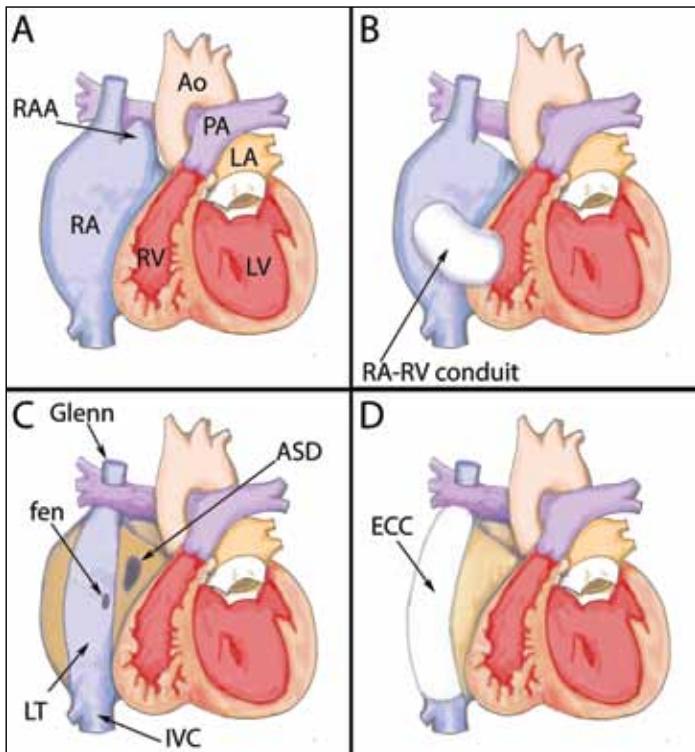
As described above, the immediate treatment in the Fontan circulation, no pumping chamber to the lungs exists. The blood flows passively to the lungs and is influenced by pressure changes caused by normal breathing and the development of higher pressures in the veins.

Are there any long term consequences related to having higher pressure in the veins?

Yes, some of these can include varicose veins in the lower legs, and congestion in the liver. The latter issue is followed very closely by adult congenital heart specialists, and if present, can often be helped by medications. In some cases, long-term congestion of the veins in the liver can lead to liver damage, so this is an important part of lifelong cardiac surveillance, and a major focus of current research into therapies to prevent these liver changes.

Are there any other complications or long term consequences of having a Fontan Operation?

Some patients can develop irregular heart rhythms (or arrhythmias) that may require treatment with medications. At times, a pacemaker is needed to help correct slow



heart rhythms or to allow for medication treatment of faster heart rhythms. Patients can have difficulty with fluid retention in the lower legs or, more commonly, the abdomen. In some patients the single ventricle can weaken over time, and may ultimately require heart transplantation.

Is exercise permitted after a Fontan?

Regular exercise is not only permitted, but encouraged. Most patients who have undergone a Fontan operation can engage in low to moderate levels of aerobic exercise such as walking, jogging, cycling, yoga, swimming and elliptical training without restriction. Light weight lifting is also acceptable, with focus on higher repetitions and lower weights. Limit your exercise to what feels comfortable for your body. Stop if you experience any symptoms that concern you such as extreme fatigue, abnormally high heart rates that don't slow down when you do, excessive breathlessness, or dizziness.

Can I have children if I've had a Fontan?

Many women who have undergone the Fontan operation have had successful pregnancies. However, advance planning is very important. Women wishing to start a family should discuss this topic with their adult congenital heart disease team long before they attempt to conceive so that appropriate testing can be done to better assess the overall risk. For both men and women with Fontan procedures (single ventricle physiology), there is a higher risk of passing congenital heart disease on to their biological offspring. The risk for this is estimated to be between 4-15%, and tends to be in the higher range if the parent with a single ventricle/Fontan is the mother.

Additional things to know:

Patients who have undergone the Fontan operation require lifelong care at a specialized Adult Congenital Heart Disease facility. It is very important that you work with your team to learn the details of your heart condition and prior operations, understand the medications you may be taking, and carry information about your heart condition with you at all times, either in the form of the Adult Congenital Heart Association Health Passport, paper records, or PDF files stored on a small flash drive. This will better enable outside medical personnel to treat you appropriately and connect with your primary team should you find yourself with a medical issue while in unfamiliar surroundings.