Ahmanson / UCLA
Adult Congenital Heart Disease Center

NEWSLETTER FOR THOSE TOUCHED BY CONGENITAL HEART DISEASE | WINTER 2011/2012

Center Updates

Dr. John S. Child retiring in 2012 after 35 years at UCLA

After 35 years at UCLA, Dr. John S. Child, director of the Ahmanson UCLA Adult Congenital Heart Disease Center is stepping down. Dr. Child has been the Director of the Ahmanson/UCLA Adult Congenital Heart Disease Center since 2001. That same year he was named the "Streisand/American Heart Association Professor of Medicine". After completing his cardiology fellowship training at UCLA in 1974, he became the chief medical resident and adjunct assistant professor of medicine from 1974-1975. In 1975, he then went on active duty in the US Navy. He returned to UCLA in 1977 and assumed an attending cardiologist position at UCLA, working with Dr. Joseph Perloff in the newly formed adult congenital heart disease clinic.

By 1980, Dr. Perloff and Dr. Child formed the Adult Congenital Heart Disease Center at UCLA, which is recognized as the first and one of the largest adult congenital heart disease programs in the U.S. The diagnostic tool of echocardiography was first utilized in the1970's and Dr. Child became recognized as a world expert in this imaging technique, and is a pioneer in imaging congenital heart disease with echocardiography. He has multiple publications in the field of imaging, congenital heart disease, valvular heart disease,and hypertrophic cardiomyopathy. He has received numerous teaching awards as a master educator for our cardiology trainees. Besides directing the Ahmanson/UCLA Adult Congenital Heart Disease Program, he is the Director of the EchocardiographyLaboratory. With more than 30 years of expertise in ACHD, Dr. Child is internationally recognized as one of the founding cardiologists of this growing subspecialty.

Preparing for the Transition

John Child leaves behind an impressive legacy for the Center and for the specialty of adult congenital heart disease as a whole. The process of transitioning Dr. Child's patients over to our other two ACHD cardiologists, Dr. Jamil Aboulhosn and Dr. Leigh Reardon has begun in earnest. By February 2012, Dr. Child will wrap up his final patient appointments. He will then spend the final 4 months finalizing the transition of both the Center and the Echocardiography laboratory. Dr. Aboulhosn will assume the Director role for the Center, bringing his talents as both a master clinician in adult congenital heart disease and a fierce advocate for adults with congenital heart disease. The end of Dr. Child's tenure here at UCLA is bittersweet. His departure is a huge loss, but the Center will flourish based on the solid foundation and state-of-the-art care that he and Dr. Joseph Perloff modeled this Center around, and will carry the momentum of advances in congenital heart disease technology, treatments and research that have become the mainstay of congenital heart disease in the United States.

New Cardiologist – Dr. Leigh Reardon

Part of that momentum includes expanding our circle of doctors and nurses who are

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MELODY VALVE UPDATE

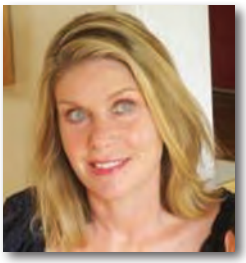
October 14, 2011, marked the one year anniversary of the UCLA Melody transcatheter pulmonary valve program. In that year, Dr. Jamil Aboulhosn and Dr. Dan Levi have successfully implanted over 30 Melody valves in adults and children. All patients are currently doing very well and most report a subjective improvement in symptoms. Further details of the Melody valve program at UCLA can be found on www.youtube.com by entering "Pulmonary Valve Replacement Without Surgery", an informative and educational video produced by UCLAHealth.

The transcatheter valve replacement field continues to grow with the ongoing COMPASSION trial (Congenital Multicenter Trial of Pulmonic Valve Regurgitation Studying the SAPIEN Interventional THV), sponsored by Edwards Lifesciences. This clinical trial, currently taking place at 5 Medical Centers in the U.S., will evaluate the use of another type of percutaneous valve, the Edwards Sapien valve, in the pulmonary position. The Sapien valve has the advantage of being a larger prosthesis than the Medtronic Melody valve and will hopefully allow patients with unsuitable anatomy for the Melody to undergo transcatheter pulmonary valve replacement.

Thanks to the effort and skill of Drs. Levi and Aboulhosn and a growing number of interventional specialists around the country, a non-surgical option for pulmonic valve replacement is now a reality for a growing number of patients with congenital heart disease.

Center Updates (continued)

providing adult congenital heart disease specialty care. After completing his pediatric cardiology and adult congenital heart disease training at UCLA in July 2011, **Dr. Leigh Reardon** joined Dr. Child and Dr. Aboulhosn as the third congenital cardiologist for ACHDC. Dr. Reardon adds rich experiences and training to the Center, having completed his pediatric cardiology and adult congenital heart disease training at UCLA. As a child, Dr. Reardon underwent cardiac surgery to correct a congenital defect of his pulmonary valve, so his clinical expertise is coupled with personal experience that brings empathy and a warm bed-side manner Dr. Reardon is welcomed as a valuable member of the adult congenital heart disease team.



New Nurse Coordinator – Jennifer Doliner

Jennifer Doliner, RN joined ACHDC in July 2011 as a Nurse Coordinator. She serves a valuable role as the liaison between patients, the administrative scheduling staff, the nurse practitioners, and cardiologists. She provides clinical support to patients who call with medical issues and require feedback before their scheduled appointments. Jennifer has 20 years of clinical experience as an RN, and a special aptitude as a communicator, a problem solver, and an advocate when it comes to dealing with insurers, pharmacies, and the health care system as a whole. She is a great addition to the Center.

New Facility – UCLA – Cardiovascular Center, 100 UCLA Medical Plaza, Suite 630

The ACHDC outpatient facilities are also going through transition this year. Both the outpatient clinic and echocardiography laboratory are moving to the UCLA 100 Medical Plaza Building as a part of the "UCLA – Cardiovascular Center". This will be a comprehensive UCLA cardiology practice, including our Center, the Cardiomyopathy (Heart Failure) Center, Heart Transplant program, the Arrhythmia Center, and Cardiac Surgery. You will receive an announcement once this move takes place. The phone number remains the same: (310)794-9629, and the UCLA – Cardiovascular Center is staffed by 7 administrative assistants who can assist you with scheduling your appointments, taking messages, and connecting you to our Nurse Coordinator if needed.

Research Studies

Contraceptive Practices in Females with Congenital Heart Disease: We are the lead Center for this multi-center questionnaire study exploring the birth control practices of our female patients, including hormonal therapies and any associated side effects and complications. To date, there are no large studies documenting the risks of different contraceptive methods in women with congenital heart disease. The objective of this study is to begin to stratify risks for different birth control methods in women with CHD, based on the review of 500 questionnaires and corresponding medical records. If you are interested in participating in this study, please email Pam Miner (pminer@mednet.ucla.edu) and she will provide you with the link to the questionnaire. You will require a study ID number to complete the questionnaire, which Pam will provide for you.

Iloprost for the Treatment of Pulmonary Hypertension in Adults with Congenital Heart Disease: We are conducting a study on the effects of an inhaled pulmonary vasodilator named iloprost (Ventavis) on adults with Eisenmenger syndrome (see the newsletter article on Eisenmenger syndrome for more details about these new pharmaceutical treatments). The medication is self-administered via an inhaled dispenser 6 times a day for three months. Treatments generally take 10 minutes. The medication is provided at no cost to the patient, as is a pre and post- treatment exercise test. If you are interested in hearing more about this study, please contact Pam Miner (pminer@mednet.ucla.edu) or Linda Houser (lhouser@mednet.ucla.edu).

Use of the drug bosentan in patients with single ventricle physiology/ Fontan repairs. This study aims to evaluate changes in pulmonary blood flow in response to the medication Bosentan, in patients with Fontan repairs. The protocol requires a baseline cardiac MRI (cannot include patients with pacemakers) followed by repeat MRI 3 months later on the medication. This would also require a monthly blood test and brief clinical evaluation. All expenses pertaining to the drug, MRI, and lab testing are covered in the study. If you would like more information, please email Pam or Linda at achdc@mednet.ucla.edu

PRESCRIPTION FOR A SUCCESSFUL DOCTOR VISIT:

1. **Be prepared (Part I).** The week prior to your appointment, keep a diary of your symptoms and pay close attention to intensity, duration, location, triggers, and effective measures for relief.
2. **Write it down!** Doctor visits can be stressful. It is easy to draw a blank on your important issues when sitting in the office. Take time beforehand to write your concerns and questions down, in the order of importance, so that they can be thoroughly covered during the visit.
3. **Be Honest.** This is your doctor, not your mother, so don't be shy about providing details about your lifestyle and habits that can potentially impact your health and your healthcare. This includes an accurate account of your exercise routine, your alcohol intake, use of prescribed and over the counter drugs, supplements, sexual activity, and any illicit drug use.
4. **Speak up.** Your top concerns should be brought up at the start of the visit, not when your doctor has one foot out the door! Start the discussion with the most important issues to avoid rushing. Don't leave with unspoken questions or concerns.
5. **Know your medications.** If you can't recall the names and dosages, always carry an up-to-date list with you, including your understanding of the purpose of each drug. Remember, vitamins and supplements are medications too, and can interact with traditional "Western" medications, so list them as well.
6. **Be prepared (Part II)** Provide your medical records and images. If you are seeing a new doctor, always come prepared with a copy of your medical records, and any images that are pertinent to the specialist that you are seeing, and current enough to be relevant to your care. Being prepared can mean the difference between a valuable and meaningful consultation versus one that is incomplete and ultimately unsatisfactory to both patient and practitioner. Medical records can now be easily retrieved using PDF documents and attachments to a flash drive, for easy storage in your home files. Ask your doctor to send you a paper or e-copy of their evaluation, so you can keep your medical records readily accessible to you at home.
7. **Don't be afraid to disagree.** Not every treatment plan is a perfect fit, and can sometimes even be a little confusing. Success depends on one's understanding of the reasons for the treatment and one's ability and willingness to comply. Feel free to ask for clarification or even for an alternative plan (if, for example, a medication or treatment is expensive or causing uncomfortable side effects). Don't hesitate to ask your doctor's office to verify your insurance authorization before proceeding with expensive testing or procedures.
8. **And finally….**Be upfront if you did not comply with the treatment plan. Rest assured, no one is going to be angry or upset with you, but medical professionals are not mind readers. If your doctor believes you were taking a prescribed medication but you were not, it is impossible to give safe advice regarding further treatment changes.

How *Can* You Help?

The Ahmanson/UCLA Adult Congenital Heart Disease Center 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.achdc.med.ucla.edu an clicking on "ways to give". You can also call 310-825-2019 if you would like to consider making a tax deductible donation to the Center, or if you have questions about specific gift options in the future.

Friends of the Ahmanson/UCLA Adult Congenital Heart Disease Center
With special thanks to our Bronze level donors:
Daniel Anaya Virginia Ettinger Judith McCormack

New Treatment Modalities for Pulmonary Hypertension

(Eisenmenger syndrome)

Children born with certain congenital heart defects, such as large holes or large connections between the great arteries, are at risk for developing a progressive blood vessel disease in their lungs if their heart defects are not repaired early in life. These types of congenital heart defects allow excessive blood flow to the lungs. Over the first 7 years of life these blood vessels can become very restrictive in response to this abnormal influx of blood flow. If this restriction of blood flow progresses to the peripheral blood vessels in the lungs, it is called pulmonary vascular disease. This usually marks the onset of "permanent" pulmonary hypertension (or pulmonary high blood pressure), and this type of blood vessel disease is not known to be reversible. To avoid the life-long effects of pulmonary hypertension, these large defects usually are repaired in the first 5-7 years of life.

The combination of an unrepaired heart defect and pulmonary hypertension has been termed "Eisenmenger syndrome". If Eisenmenger syndrome has set in, closing the heart defect can be potentially dangerous rather than beneficial. Accordingly, children with Eisenmenger syndrome confront the lifelong effects of this disease, without a surgical repair option. The symptoms that accompany this problem include breathlessness with activity, bluish discoloration of the skin (particularly nose, lips, gums, and fingers), and several other blood and organ changes that occur with aging in the setting of low oxygen levels. The low oxygen levels are the result of blood passing across the defect in the heart without first picking up oxygen in the lungs.

Historically, it has been reported that patients with Eisenmenger syndrome confront early mortality, with 77% survival at 15 years of age and 42% survival at 25 years of age. The most common cause of death in these patients is sudden heart rhythm problems, heart failure, or lung hemorrhage. With advances in the recognition of risks confronted by adults with Eisenmenger syndrome, and evolving modalities to minimize the risks of bleeding, and minimize the evolution of heart failure, many of these adults are living well into their 40's, 50's, and even into their 60's. In the past, it was believed that the use of medications to reduce pulmonary blood pressure would be too risky in patients with Eisenmenger syndrome, since they would be susceptible to a decrease in their body's blood pressure, which could make their oxygen levels drop dangerously. However, studies over the past decade have dispelled this myth and have led to a widespread use of new drugs focused at reducing pulmonary blood pressure. Current studies have shown that these advanced therapies both improve quality of life, and may have a beneficial impact on survival.

The specialized drugs that are used to treat pulmonary hypertension fall into three main categories and may be used separately or in combination:

1. Phosphodiesterase-5 enzyme inhibitors, such as *revatio or adcirca*.
2. Endothelin antagonists, such as *tracleer or letairis*.
3. Prostanoids, which comes in an intravenous form called *flolan*, a skin delivery system called *remodulin*, and an inhaled version such as *ventavis or tyvaso*.

The Ahmanson/UCLA Adult Congenital Heart Disease Center is conducting a research study to evaluate the impact of inhaled ventavis on exercise capacity in our Eisenmenger syndrome patients. This is a 3 month trial, in which the medication and all associated testing is provided free of cost. If you are interested in learning more about these advanced therapies, or participating in the ventavis study, please call or email (310-794-9629 or achdc@mednet.ucla.edu).

"NOTHING ABOUT ME, WITHOUT ME!"

REFLECTIONS ON THE ACHA CONFERENCE

By Kay Deeney, MLS

I recently attended the Adult Congenital Heart Association (ACHA) Conference in Los Angeles. This conference is particularly unique and interesting because people with the disease, patients, and family members along with the health care practitioners attend together. This community engagement is useful for both the health care providers and the people impacted by the disorder. The Conference drew nearly 400 participants from more than 32 states, the District of Columbia and Canada. Of these, about 250 were patient or family members and about 150 registered for the professional/research programming.

At the Conference, there were general sessions for both health professionals and people with congenital heart disease, with special sessions for each group. Some of the sessions for patients were defect-specific, including topics such as tetralogy of Fallot or truncus arteriosus. Other patient topics were health insurance, exercise, disability rights, the Affordable Care Act and preexisting conditions. Sessions on aging for men and women were extremely popular! Many of the health care providers presented programs for the patient groups. The talks for the health care professionals were on latest technologies such as stent heart valve implantation and heart transplant issues, heart failure and translational research and patient-centered care.

One of the final sessions involved patients and health care providers. One gentleman in his fifties talked about having surgery when he was 20. "This should give you five years!" he was shocked to be told. He was ecstatic to have beaten the odds. "I will die one of these days, but so will all of us!" he said. Several of the health care professionals talked about the relationships they had with their patients over the years. Most of the time, it is gratifying for the doctors to see their patient population thrive, yet sometimes it is frustrating when patients pursue risky behavior or disregard advice.

The title quote, "Nothing about me, without me!" refers to the fact that the research agenda for congenital heart disease should involve the patient as well. Because congenital heart disease is a chronic disease, the patient needs to understand current research that may affect their situation. At times, delaying surgery can free up opportunities for treatments that were not available a year before. Sometimes tests are done just to get a baseline for this patient. The patients need to feel that their concerns are being considered seriously. With an emphasis on community engagement, this patient and provider centered Conference will continue and probably extend to other groups as well. Genuine involvement of the congenital heart disease community and its health care providers should lead to the improvement of health for all.

Ahmanson/UCLA ACHDC Faculty

John Child, M.D. (Director)
Jamil Aboulhosn, M.D. (Co-Director, Interventional Cardiology)
Leigh Reardon, MD (Attending physician)
Daniel Levi, M.D. (Interventional Cardiology)
Kevin Shannon, M.D. (Electrophysiology)
Jeremy Moore, M.D. (Electrophysiology)
Kalyanam Shivkumar, M.D (Electrophysiology)
Ravi Mandapati, M.D. (Electrophysiologist)
Hillel Laks, M.D. (Cardiothoracic Surgery)
Brian Reemsten, M.D. (Cardiothoracic Surgery)
Pamela Miner, NP (Nurse Practitioner)
Linda Houser, NP (Nurse Practitioner)

Ahmanson/UCLA ACHDC Board of Advisors

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Kenneth Boyko
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Deborah Spander

How to Contact the Center:
Patient Call Center Triage (310) 794-9629
Administrative Issues (310) 825-2019
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Web: www.achdc.med.ucla.edu

Information and Internet Resources:

Ahmanson/UCLA Adult Congenital Heart Disease Center:
Web: www.achdc.med.ucla.edu
Call: (310) 794-9629
Email: achdc@mednet.ucla.edu

Adult Congenital Heart Association (ACHA):
Web: www.achaheart.org
Call (888) 921-ACHA
Email: info@achaheart.org

ACC/AHA Guidelines for Management of Adults with Congenital Heart Disease:
http://circ.ahajournals.org/cgi/reprint/CIRCULATIONAHA.108.190690

Camp Del Corazon:
http://www.campdelcorazon.org/

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Flu Facts:

- It's that time of year again! In the midst of Halloween costumes, roasting turkeys, sharing gifts and singing holiday songs, many of us will encounter an age-old enemy—the flu.
- The flu – influenza – is a contagious respiratory illness caused by several types of influenza viruses. These viruses spread mainly by droplets made when people with the flu cough, sneeze, or even talk. The droplets can land in the mouths or noses of people who are nearby. A person might also get the flu by touching a surface or object that has flu virus on it and then touching their own mouth, eyes or possibly their nose. **(The average person touches their face hundreds of times during the day, up to 3-5 times every waking minute!)**
- Signs and symptoms of the flu include a fever, chills, cough, sore throat, runny nose, muscle aching, headaches, fatigue, and sometimes vomiting and diarrhea (more common in children than adults).
- The best way to prevent the flu is by getting a flu vaccine each year. Since February 2010, the CDC has recommended that everyone 6 months and older receive an annual flu vaccine. The 2011-2012 flu vaccine will protect against the three influenza viruses that research indicates will be most common during the season. This includes an influenza A (H1N1) virus, an influenza A (H3N2) virus, and an influenza B virus.
- Some people should not be vaccinated against the flu without first consulting their physician. These include people who have a severe allergy to eggs, people who have had a severe reaction to the flu vaccine in the past, children younger than 6 months of age, people with moderate or severe illness with a fever (wait until symptoms are resolved), and people with a history of an illness called Guillain-Barre Syndrome.
- Day to day strategies for avoiding the flu also include covering your nose and mouth with a tissue when you cough or sneeze and washing your hands frequently with soap and water or using alcohol based hand rub. Avoid touching your eyes, nose and mouth, and try to avoid close contact with sick individuals.
- **REMEMBER: Stay home if you are sick to avoid spreading flu to others! Your co-workers will thank you!**

Wishing everyone a happy and HEALTHY new year!