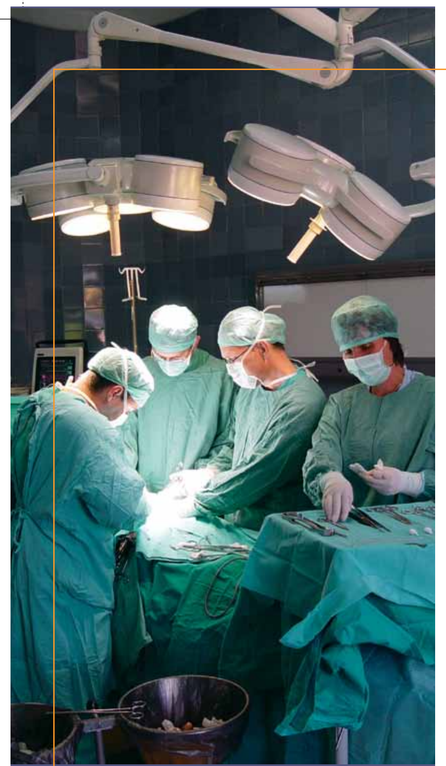


# THE FONTAN CIRCULATION:

## Current Options and New Management Strategies

Patients born with the congenital heart defect known as "single ventricle" generally undergo the Fontan operation in order to establish separate circulation pathways to the lungs and the body. However, because the single ventricle is ultimately used to pump blood exclusively to the body, blood flow to the lungs is dependent on passive flow and requires intrinsically higher pressure in the veins to drive the flow to the lungs. In some patients the higher pressure results in complications of decreased function of the single pumping chamber, abnormal heart rhythms (arrhythmias), chronic swelling and abdominal bloating, and liver or kidney problems. Management of these issues includes putting the patient on medications, having pacemakers implanted, and catheter procedures to eliminate arrhythmias. Sometimes a Fontan conversion surgery, in which older types of Fontans are converted into more streamlined newer versions of the operation or a heart transplant, is required.

For various reasons, a small subset of this patient population will ultimately be deemed unacceptable candidates for heart transplantation. Typically, this population has also failed other types of medical and surgical management. This growing patient population has resulted in increased research focused on ways to both prolong and improve quality of life in those Fontan patients with no further medical or surgical options. One of the most promising areas of research is looking at the use of implantable mechanical devices used to support either the circulation to the lungs or the circulation to the body. Ventricular assist devices (VAD), are commonly used in the conventional treatment of severe heart failure. VADs allow most of the oxygenated blood to bypass the failing pumping chamber (in structurally normal hearts, the left ventricle), providing better circulation to the body. Most recently, Providence Sacred Heart Medical Center has initiated a – Phases II Clinical Trial designed to assess the effect of VADs in Fontan patients. The study plans to enroll 20 patients and hopefully, the results will lead to a larger multicenter study. The goal is to validate a new therapy for a very challenging patient population.


**Ahmanson / UCLA**
Adult Congenital Heart Disease Center

Division of Cardiology
405 Hilgard Avenue
Box 951679, Room A2-237 CHS
Los Angels, Ca 90095-1679




## HAPPY HOLIDAYS TO ALL!!

**The Faculty and Staff of the Ahmanson/UCLA Adult Congenital Heart Disease Center wish you and your family a happy holiday season. We also wanted to pass along these tips to end 2010 on a heart healthy note, and to continue good eating and exercise habits through the New Year:**

- **Make smart choices:** Remember, there are no "good" or "bad" foods, only good and bad eating habits. Make sure that the mainstay of your holiday consists of fresh fruits and veggies, lean meats, low fat dairy products and grains. Visit the Food Pyramid website for more ideas on maintaining a healthy diet.

MyPyramid.gov
STEPS TO A HEALTHIER YOU!

- **Go easy on the sugars, salt and fats:** Excessive weight gain during the holidays is usually due to eating too many of these types of foods and not enough of the "Smart" foods. It's ok to indulge in small amounts of tempting holiday foods and desserts, but in moderation. Snacks and desserts are not meals! In addition, excess salt can cause the body to retain water and can elevate blood pressure and fats and sugars can negatively impact cholesterol levels.
- **Don't arrive at the party hungry:** Don't fast before special events in anticipation of eating a big meal. Eat a light meal before you leave, preferably one that is high in protein. This will curb your appetite and make you less inclined to overeat at social occasions.
- **Don't supersize:** Eat smaller portions of food, particularly at buffets where you may want to try a little of everything.
- **Eat slowly:** It takes the brain about 20 minutes to realize the stomach is full, so savor each bite, monitor your hunger level, and stop eating when you feel full. The odds are good that you'll eat less.
- **Beverage tips:** Drinks add calories, particularly alcoholic beverages, fruit punches and sugary sodas. Choose diet sodas, ciders, and flavored waters instead.
- **Keep exercising:** All too often, exercise programs are sacrificed in favor of fulfilling other obligations and running errands. Don't let this happen to you! Commit to yourself. If you've been exercising, continue throughout the holiday season for both physical and mental benefits (exercise is a great stress-reducer). If you aren't getting enough exercise, now is a great time to start—and continue into the New Year.
- **Remember to get your flu shot.**


# Ahmanson / UCLA
Adult Congenital Heart Disease Center

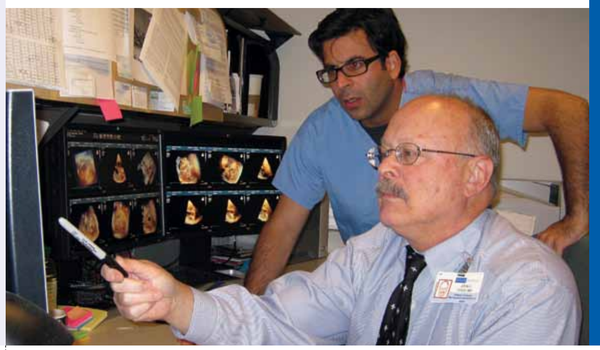
NEWSLETTER FOR THOSE TOUCHED BY CONGENITAL HEART DISEASE | FALL 2010


## Welcome to the Ahmanson/UCLA Adult Congenital Heart Disease Center Newsletter

Welcome to the Ahmanson/UCLA Adult Congenital Heart Disease Center Newsletter. The Ahmanson/UCLA Adult Congenital Heart Disease Center is the first of its kind and one of the largest facilities in the United States. The Center provides services to the growing number of patients with congenital heart disease who reach adulthood.

In addition to its emphasis on the highest quality of patient care, the Center is a major training and research facility for medical and pediatric house officers, medical and pediatric cardiologists, and visiting cardiologists from the United States and abroad. The Center offers an advanced fellowship for cardiologists with a career interest in congenital heart disease in adults.

We hope that you enjoy this newsletter. It provides an update on the Center, the Cardiothoracic surgery center, information regarding new technology, and a profile on a patient that we thought would be of interest to you.



## What's NEW



ACHA Adult Congenital Heart Association

# Cardiothoracic Surgery Faculty

In 2008, UCLA welcomed Dr. Brian Reemsten as the Chief of Congenital Heart Disease Surgery.

He joins internationally renowned congenital heart surgeon Dr. Hillel Laks, in providing world-class cardiac surgical care to our adults with CHD. Dr. Reemsten's training and experience are impressive, and began with a general surgery residency here at UCLA. He then did a cardiothoracic residency at the University of Washington, followed by an advanced fellowship in pediatric congenital heart surgery at Great Ormond Street Hospital in London from 2004 to 2005.

Dr. Reemsten joined Children's Hospital of Los Angeles in 2005 before being recruited to UCLA in 2008. His mentor at CHLA was Dr. Vaughn Starnes, with whom he developed his expertise in both pediatric and adult congenital heart surgery, as well as pediatric cardiac transplantation. We are privileged to have Dr. Reemtsen as part of our Ahmanson/UCLA Adult Congenital Heart Disease Center team.

# Updates from the Center

This year, the Ahmanson/UCLA Adult Congenital Heart Disease Center celebrates its 30th year of caring for the growing number of patients with congenital heart disease who have reached adulthood. This is a significant milestone for the Center, which is the first of its kind in the U.S and still among the largest in the country.

The next decade of adult congenital heart disease care at UCLA will continue to bring world-class care for adults with congenital heart disease through new technologies and treatments. Dr. John Child, the Director of the Center, and Dr. Jamil Aboulhosn, the Co-Director of the Center, recognize that the Center's core mission includes the provision of state of the art ACHD patient care, conducting cutting edge ACHD research, leading the nation in novel management strategies and interventions, and providing education to trainees, other health care providers, and patients and families.

### THE CENTER'S GOALS FOR THE COMING YEAR INCLUDE:

- Continue research to evaluate the effects of new medications for pulmonary hypertension and to determine whether there is a beneficial effect on survival for those patients taking the new medications.
- Improve cardiac imaging techniques by using new 3-dimensional echocardiographic technologies.
- Inaugurate the use of catheter-based valve replacements at UCLA, with the potential for eliminating future cardiac surgeries in some ACHD patients.
- Host the 2011 Adult Congenital Heart Association national meeting, at which new research in ACHD will be presented. The national meeting will also be a forum for the provision of education to health care professionals and patients and families.

# 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 and to pursue the goals of the Center. 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 and clicking on "Ways to Give." You can also call 310-794-4746, if you have any questions about making a gift to the Center.

# Music to our Patients' Ears:

## The Medtronic Melody Valve Comes to UCLA

On October 14th, the Ronald Reagan UCLA Medical Center joined a select group of medical establishments across the nation that began post-clinical trial pulmonary valve replacement with the Medtronic Melody transcatheter pulmonary valve. The team was led by UCLA congenital heart disease interventional experts Dr. Daniel Levi and Dr. Jamil Aboulhosn. Five patients (3 adults and 2 children) were the first to receive the Melody valve at UCLA.

### The Pulmonary Valve

The pulmonary valve is one of the four heart valves. It is located between the right ventricle and the pulmonary artery. The valve opens and closes as the right ventricle contracts and relaxes, sending a one-way flow of blood to the lungs to receive oxygen, which is then pumped to the body. A large percentage of patients with congenital heart disease are born with either a narrowing or total absence of this valve, requiring early open heart surgery, later surgeries to insert artificial conduits (tubes with valves sewn inside), or tissue valves. Over time, however, these valves narrow or become leaky, resulting in patient symptoms of palpitations, shortness of breath, fatigue and exercise intolerance. Traditionally, the valves have required surgical replacement every 5-15 years. As a result, these patients undergo multiple open heart surgeries over the course of a lifetime.

### Brief History of Transcatheter Valve Replacement

The concept of replacing the pulmonary valve via a method that did not involve open-heart surgery was first conceived in the late 1990s by Philipp Bonhoeffer, M.D., a pediatric cardiologist in Paris, France . Working with children and young adults with diagnoses such as tetralogy of Fallot and pulmonary atresia (total absence of the pulmonary valve) as well as many other congenital pulmonary valve disorders, Bonhoeffer observed that repeat surgeries were often delayed in order to avoid subjecting these young people to the discomfort, prolonged disability, and increased risks of a repeat operation. Existing catheter interventions to deal with valve stenosis such as opening up the valve with a balloon, or placing bare metal stents often left new problems in their wake, mainly severe regurgitation or leaking.

Ultimately Dr. Bonhoeffer developed the idea of attaching a valve to a stent or collapsible metal "scaffolding" that could be guided by a catheter to the heart through a vein in the leg and deployed in the area of the degenerated valve. The new valve and stent would be held in place by the walls of the old valve and/or conduit. Dr. Bonhoeffer established a partnership with medical device manufacturers Medtronic and NuMed. The type of valve that was ultimately chosen was the bovine jugular venous valve (a valve from the neck vein of a cow) and the stent was composed of a durable yet flexible platinum iridium material that could be crimped down to fit into a catheter delivery system. The Melody Valve —so named because of Dr. Bonhoeffer's love of music—was born.

The first patient to receive the Medtronic Melody valve was a 12-year old Parisian boy with pulmonary atresia and a conduit between his right ventricle and pulmonary artery that had degenerated after 8 years. The child did very well after the procedure. Thereafter, clinical trials commenced and, to date, approximately 1200 patients worldwide have undergone the transcatheter pulmonary valve procedure. The Melody valve has demonstrated durability and stability in patients for 5 years in Europe and 3 years in the United States. In addition, several additional Transcatheter valves are being developed and trialed by other companies.

The first transcatheter valve approved for clinical use in the United States, the Melody Valve was given the green light in January 2010 through the FDA's Humanitarian Device Exemption program, which allows the agency to approve a device for limited use in patients with conditions that affect fewer than 4,000 individuals in the United States each year.

As recently noted by Dr. Aboulhosn, "This new procedure will revolutionize the care of the patient with pulmonary valve disease." UCLA is honored and excited to be participating in what is arguably the most significant breakthrough in interventional congenital cardiology in the past decade.

### So Far a Success

One of the patients that received a Melody Valve was John K., who is featured below.

# MELODY VALVE PATIENT PROFILE: JOHN K.

John has been a patient of the Ahmanson/UCLA Adult Congenital Heart Disease Center for the past several years. He was born with pulmonary atresia, or absence of the pulmonary valve (the gateway between the right ventricle and the pulmonary artery) preventing blood from reaching the lungs via this route. As an infant and child, John underwent two major cardiac surgeries to enable adequate blood flow to reach the lungs. Finally, 10 years ago, at the age of 16, he underwent surgical pulmonary valve replacement.

An athletic and active young man, one of John's major interests is professional motorcycle riding, and he travels extensively in the southwestern United States to engage in this competitive activity. Over the past year, he has been feeling more tired and breathless, and can no longer practice and compete to the degree that he desires. Investigation into the cause of these symptoms ultimately identified a deterioration of the 10 year old pulmonary valve accompanied by a decrease in the function of John's right ventricle, a sign that John's heart was not handling the pulmonary valve leak. Placement of a new pulmonary valve was therefore recommended.

Fortunately for John, the Medtronic Melody valve offers him an alternative to a repeat open heart surgical procedure. He was selected as a candidate and in mid October, underwent successful pulmonary valve replacement with the Medtronic Melody transcatheter valve. As of this writing, John is doing very well and anticipating a return to riding in the near future!

If you would like to share your story in future newsletter, please email us at achdc@mednet.ucla.edu.

## Ahmanson/UCLA ACHDC Faculty

John Child, M.D. (Director)
Jamil Aboulhosn, M.D. (Co-Director, Interventional Cardiology)
Daniel Levi, M.D. (Interventional Cardiology)
Kevin Shannon, M.D. (Electrophysiology)
Kalyanam Shivkumar, M.D (Electrophysiology)
Hillel Laks, M.D. (Cardiothoracic Surgery)
Brian Reemtsen, M.D. (Cardiothoracic Surgery)
Pamela Miner, NP (Nurse Practitioner, email at pminer@mednet.ucla.edu)
Linda Houser, NP (Nurse Practitioner, email at lhouser@mednet.ucla.edu)
Leigh Reardon, MD ( ACHD Fellow)

## Ahmanson/UCLA ACHDC Board of Advisors

Nora McGarry Arian
Kenneth Boyko
Delphone Lee
Douglas Praw
Thomas A. Bliss
Judah Hertz
Michael McNamara
Lori Yonan

**How to Contact the Center:**
Patient Call Center Triage (310) 794-9629
Administrative Issues (310) 825-2019
Fax Number (310) 825-6346
Email: achdc@mednet.ucla.edu
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/*