

# CONGENITAL CARDIOLOGY TODAY

Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

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## Anomalous Origin of the Left Pulmonary Artery from the Ascending Aorta in a Patient with 22Q11.2 Deletion Syndrome

By Ryan Halas, DO; Christopher Schmeihil, MD; Devika Malhotra, MD; Ming-Sing Si, MD; Robin Fountain, MD

**Key Words:** Anomalous Origin of one Pulmonary Artery from the Ascending Aorta (AOPA), Anomalous Origin of the Left Pulmonary Artery from the Ascending Aorta (AOLPA) Hemitruncus, DiGeorge Syndrome, 22Q11.2 Deletion Syndrome.

### Introduction

Anomalous origin of one pulmonary artery from the ascending aorta (AOPA), also known as hemitruncus, is an extremely rare congenital heart malformation first described in 1868.<sup>1</sup> This defect manifests with early pulmonary hypertension by two unique mechanisms:

1. one lung receiving an obligate entire cardiac output from the right ventricle and
2. systemic pressure driving systemic arterial blood into the lung with the anomalous origin of the branch pulmonary artery.<sup>2</sup>

Early surgical repair with direct implantation of the anomalous artery into the main pulmonary artery has provided excellent results.<sup>2</sup> While anomalous aortic origin of either the right or left pulmonary artery from the ascending aorta can occur, the frequency and cardiac associations of each have led some to believe that they have separate embryologic considerations.<sup>3</sup> Anomalous aortic origin of the right pulmonary artery is approximately 4 to 8 times more common.<sup>4</sup>

There are only two reported patients with 22Q11.2 Deletion Syndrome, also known as DiGeorge Syndrome and Velocardiofacial Syndrome, and Anomalous Aortic Origin of the Left Pulmonary Artery (AOLPA), both described by Dodo Et al in 1995.<sup>4</sup> Here we present a third case of AOLPA associated with 22Q11.2 Deletion Syndrome.

### Case Report

A 3.99 kg, 7-week-old girl with recently diagnosed bronchiolitis and uncomplicated birth history presented for evaluation of heart murmur. ROS was positive for mild tachypnea, but otherwise negative. There was no family history of Congenital Heart Disease (CHD).

A physical exam was pertinent for a peripheral oxygen saturation of 95%, a grade III/VI systolic ejection murmur best heard at the left upper sternal border that radiated to the bilateral axilla and back and clear lung fields. An echocardiogram demonstrated the Left Pulmonary Artery (LPA) arising from the leftward aspect of the ascending aorta while the anatomy of the right pulmonary artery was normal. (Figure 1). The right ventricle was mildly dilated and hypertrophied with a flattened interventricular septum. The aortic arch was not well-imaged on initial imaging. The right ventricular pressures were estimated to be slightly sub-systemic.

The patient was referred for urgent surgical correction. She underwent surgical implantation of the LPA to the main pulmonary artery (MPA) with pericardial patch augmentation of the

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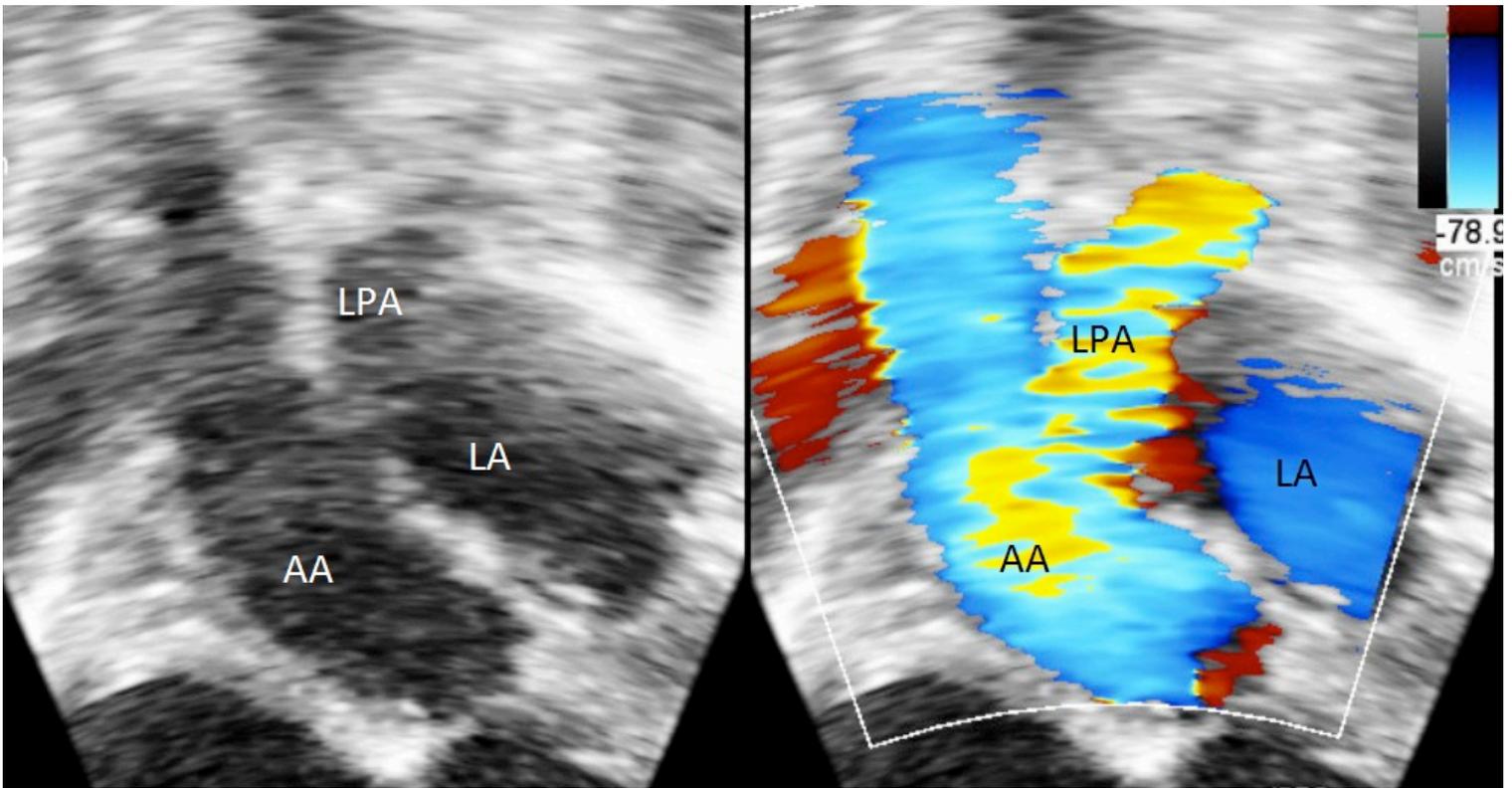


Figure 1. Pre-surgical echocardiogram with color flow, apical view with anterior angulation, reveals ascending aorta giving rise to left pulmonary artery. AA- Ascending aorta, LPA- Left Pulmonary Artery, LA- Left Atrium.

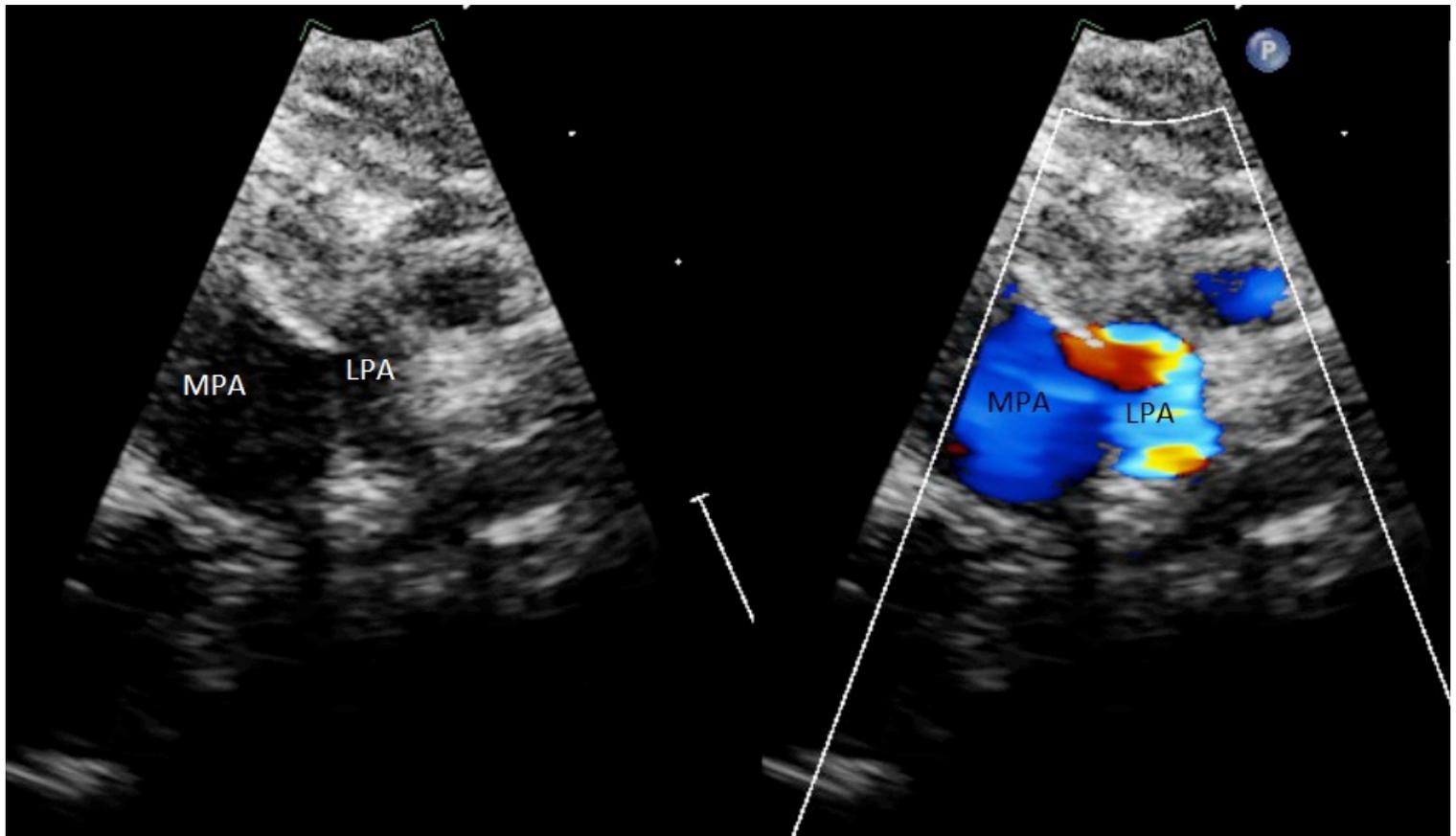


Figure 2. Post-surgical echocardiogram: parasternal short axis with color flow showing reimplantation of the left pulmonary artery to the main pulmonary artery.

connection (Figure 2). Inspection of the great arteries revealed a right-sided aortic arch with possible aberrant left subclavian artery and dilated MPA and RPA. No thymic tissue was visualized. After implantation of the LPA into the MPA, right ventricle and aortic systolic pressures were directly measured to be 25 and 75 mmHg, respectively. Approximately two-and-a-half weeks after the procedure she developed Post-Pericardiectomy Syndrome with pericardial effusion requiring a short course of steroids.

Genetic workup revealed 22q11.2 Deletion Syndrome. She continues to do well from a cardiopulmonary standpoint.

## Discussion

This is the third documented incidence of AOLPA in a patient with 22Q11.2 Deletion Syndrome. As with the other two cases described by Dodo, our patient presented in the first two months of life with respiratory symptoms and had a right aortic arch. In each case, there was good surgical outcome with the direct implantation of the anomalous left pulmonary artery to the main pulmonary artery. 22Q11.2 Deletion Syndrome is associated with conotruncal abnormalities including: Tetralogy of Fallot, interrupted aortic arch and truncus arteriosus, among other rare associations. We now encourage clinicians to consider AOLPA in this small group of rare associations.<sup>6</sup> It is important that clinicians recognize this pattern when AOLPA is suspected or confirmed, especially in combination with right aortic arch. This may allow for earlier diagnosis of 22q11.2 Deletion Syndrome, leading to earlier multidisciplinary care. Furthermore, hypocalcemia and hypomagnesemia are known complications of 22q11.2 Deletion Syndrome especially during times of biological stress and should be monitored closely in the perioperative period. Finally, 22q11 fluorescence in situ hybridization (FISH) testing may be warranted for patients with AOLPA.

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## Pediatric Electrophysiologist and Noninvasive Imaging Faculty Positions

The Division of Pediatric Cardiology at the University of Utah School of Medicine and based at Primary Children's Hospital is recruiting BE/BC pediatric cardiologists with major interest and expertise in:

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The pediatric cardiologists will join a division of 29 faculty including 3 electrophysiologists and several noninvasive imagers as part of an active, growing, nationally ranked Heart Center. Approximately 500 surgeries are performed annually by 3 congenital cardiac surgeons. In addition to having a busy clinical program, a thriving fellowship training program provides opportunities for teaching. The division has an active research program with numerous opportunities for participation in basic, translational, and clinical research. The Division is one of the core participating centers in the Pediatric Heart Network funded by the NIH.

The successful candidates will receive a faculty appointment in the Department of Pediatrics on the Clinical or Tenure track at the academic level commensurate with experience and qualifications. The University of Utah and Department of Pediatrics offer an excellent benefits package that includes 20.2% retirement contributions that vest immediately and excellent health care choices. The Department offers an education loan repayment program, departmental research core with mentoring, as well as education and leadership opportunities. The area offers an excellent quality of life with immense cultural and recreational opportunities.

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**Biographical Sketch**

Ryan Halas is a third year Internal Medicine-Pediatrics resident at Western Michigan University Homer Stryker MD School of Medicine in Kalamazoo Michigan. He completed his undergraduate training at Michigan State University in Business Management before completing medical school at Michigan State University College of Osteopathic Medicine. After residency he is planning to pursue fellowship training in Pediatric Cardiology.

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# Heart Program



## Two Physicians Wanted to Join Our Heart Program's Division of Cardiac Critical Care Medicine

Nicklaus Children's Hospital (formerly Miami Children's Hospital), a 289-bed freestanding children's hospital and Level III trauma center, and Pediatric Specialists of America (PSA), the physician-led group practice of Miami Children's Health System, have an exceptional opportunity for two physicians to join our esteemed Heart Program's division of cardiac critical care medicine.

Our Cardiac Intensive Care Unit (CICU) was the first in the Southeast and provides care for newborns and children receiving treatment for congenital heart defects. With a long-standing tradition of excellence, our cardiac critical care team is currently comprised of six full-time attending physicians and six full-time nurse practitioners. We have an illustrious cardiology fellowship and have offered advanced training in cardiac critical care medicine for more than 20 years. The desired candidates should be board certified or eligible in pediatric critical care medicine or pediatric cardiology. Preference will be given to individuals with dual training in pediatric critical care and cardiology or those board eligible in either cardiology or pediatric critical care who have completed a minimum of one year of advanced training in cardiac intensive care medicine. Applicants should exhibit a strong interest in clinical care, education and academics. Nicklaus Children's Hospital is an affiliate of the Florida International University Herbert Wertheim College of Medicine. Candidates possessing all levels of experience shall be considered.

In October 2016, we moved into our new state-of-the-art Advanced Pediatric Care Pavilion, which houses a 34-bed cardiac in-patient unit with an adjustable acuity model that allows all rooms to accommodate critically ill patients with heart disease. The Heart Program offers a full range of services, including the management of patients following congenital heart surgery, interventional catheterization and invasive electrophysiology. Open heart surgical services are offered to patients as small as one kilogram through young adulthood. Our cardiac surgical program, led by Dr. Redmond Burke, is one of the most innovative in the world and the most transparent. It remains the only cardiovascular surgical program to offer real-time outcomes reporting (<http://www.pediatricheartsurgery.com/realtimeoutcomes/cvperformance.aspx>). Nicklaus Children's also plans to open a birthing center for at-risk fetuses with congenital heart disease. Construction began last fall.

Founded in 1950, the rebranded Nicklaus Children's Hospital is renowned for excellence in all aspects of pediatric medicine and has numerous subspecialty programs that are routinely ranked among the best in the nation. It is also home to the largest pediatric teaching program in the southeastern U.S. Many of the physicians on our staff have trained or worked at other leading medical institutions. Join a phenomenal team that brings lifelong health and hope to children and their families through innovative and compassionate care.

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### Interested candidates please send inquiries to:

Anthony Rossi, MD  
Section Chief, Cardiovascular Medicine  
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[anthony.rossi@mch.com](mailto:anthony.rossi@mch.com)



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# Q&A Interview with Dr. Henry (Heinz) Gelband - May 16<sup>th</sup>, 2016

By Bradley W. Robinson, MD



Dr. Henry (Heinz) Gelband, MD

## Preface

I interviewed Dr. Henry (Heinz) Gelband on Monday, May 16<sup>th</sup>, 2016. It was my sincere pleasure to interview him, (Known as HG below). He was the Fellowship and Division Chief at University of Miami from 1989-92, and still is my clinical and life mentor. Dr. Gelband was Pediatric Cardiology Division Chief at University of Miami between 1975-1996. He is a Chief's "Chief" of Pediatric Cardiology. Considered one of the founding fathers of Pediatric Electrophysiology, he has authored numerous papers and books on the subject. I caught up to Dr. Gelband at home this Spring.

## Q: When did you come to Miami as a faculty member?

**HG:** I joined as an Assistant Professor of Pediatrics in Pediatric Cardiologist in 1971. Dr. Mary Jane Jesse was the Division Chief. Drs. Dolores Tamer and Otto Garcia were on faculty already.

## Q: Who was your clinical mentor?

**HG:** Dr. Sidney Blumenthal, Head of Pediatric Cardiology at Columbia University. He was one of the first 6 original members that developed the Pediatric Cardiology Sub-Board of Pediatrics (it was also the first sub-board of Pediatrics) in the early 1960s. Dr. Blumenthal later came to University of Miami to be the Associate Dean of Medical School there.

## Q: Who was your research mentor?

**HG:** Brian Hoffman, Chairman of Dept of Pharmacology at Columbia. I spent 2 years doing research in his lab, doing basic Electropharmacology. He was a brilliant guy and one of fathers of Cardiac Electrophysiology. He gave credit to young investigators, and had 5-6 research labs going. All fellows did well that did research with him, and were successful all over the world. An example is Dr. Robert

Myerberg, who came to Miami as Chief of Cardiology at the VA Hospital.

## Q: When did you become interested in research?

**HG:** I always had a yen for research. I did research as an intern in 1963 looking at cardiopulmonary function in certain surgical patients to see who were high risk. While I was in the Public Health Service in San Diego, CA, the soldiers were using a solvent to clean their guns (trichloroethylene or TCE), which was absorbed into the skin and created neurologic symptoms. We published this finding when I was a resident in J of Public Health.

## Q: When were you in the military service?

**HG:** I was in the Marines, active duty from 1963-65. I was in the reserve and active duty 1959-65. In June 65, I was discharged just 2 months before the Marines went to combat in Vietnam.

## Q: What was history of the University of Miami training program?

**HG:** We started the training program in 1973. We started with one trainee/year, and increased to three trainees/year. Dr. Pedro Ferrer (at Yale) came down to run adult portion of SCOR grant (Specialized Center for Research on Atherosclerosis). We had an electrophysiology training grant from NIH grant for 10 years. Drs. Pickoff, Young, Casta, Villafane, McCormick and others trained under the EP grant, and continued their careers elsewhere (close to 30 fellows over a 10 years period). Jackson Memorial Hospital funded the clinical pediatric cardiology program. I had a training grant from the hospital and one from the American Heart Association. I received a career development award from AHA for my own lab which supported pre/post doctorate candidates who did clinical research/basic research.

## Q: What was success of program based on?

**HG:** The success of the program was based on good communication skills. I learned to "be honest with people." I rewarded faculty/fellows for what they did, and I gave appropriate credit to faculty/fellows.

## Q: What were the most important lessons you learned from from mentors?

**HG:** Never work for someone who is not as smart as you. Never work for someone who doesn't work as hard as you. Never work for someone who isn't able to give you resources to be successful.

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## Dr. Gelband's Advice to Follows:

**"If opportunity to do what you want exists, then GO DO IT: don't make excuses!"**

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## Q: How did this work out at the University of Miami?

**HG:** I was fortunate that we recruited people as smart or smarter than I, and I treated faculty fairly. We had great resources (from hospital/university) and the faculty. The faculty provided support, and we had grants to provide support (March of Dimes, AHA and The NIH). People had faith in what the division was doing, and the division produced grants. I had a great time and loved it.

**Q: What was your favorite part of being Chief?**

**HG:** First, I enjoyed the morning report 7:30-8:30 am Mondays, Wednesdays, Fridays with interaction with fellows. I read those reports because I wanted to be smarter than the fellows. (It was an ego thing). Second, I enjoyed Cardiac Cath Conference discussing complicated cases and observing interaction among specialists. I let division staff make their own decisions based on advice from the others. This made me happy and was what I wanted to do. If I was happy, I went to work happy and made others happy.

**Q: How did you recruit such a good faculty?**

**HG:** Drs. Dolores Tamer and Otto Garcia were at U Miami already, Dr. Dolores Tamer had trained in Philadelphia under Dr. William Rashkind, and was an expert in the cardiac cath lab. Dr. Otto Garcia worked with Dr. Agustin Castellanos (who was nominated for Nobel Prize for Medicine twice), performing some of the first cardiac catheterizations on children in the Western Hemisphere in Cuba. Dr. Grace Wolff developed clinical EP at U Miami, diagnosing and treating arrhythmias. She came from Albany, NY; we had known each other at Columbia. Dr. Pedro Ferrer, joined the faculty, and was an expert on non-invasive cardiology having been well-trained at Boston Children's Hospital and at Yale in New Haven. Word spread around about the U Miami program. I was chairing national sessions, so our program became known and attractive to potential fellows/faculty.

**Q: What would you tell a new pediatric cardiology fellow starting in 2016?**

**HG:** Decide if you want to be a general pediatric cardiologist with skills in noninvasive cardiology, or do you want to be hospital-based/academic-based. If you stay in Pediatric Cardiology, become proficient in a specialty to make you a marketable item after you finish your fellowship and start your career. For instance, develop a background in computer skills/technology of imaging MRI/CT. Develop expertise in Adult CHD. Become an Intensive care specialist to take care of critically ill cardiac patients. Become an electrophysiologist. Become a non-invasive specialist.

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***Dr. Gelband's Advice on Being True to Self: "Only one person knows how good you are and that is yourself (you don't need plaques or awards or honors). When you have to think every day, learn something everyday and laugh every day, then this is a good day for the physician."***

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**Q: Where do you see the field in the future in 10 years?**

**HG:** We are reinventing ourselves every 5-10 years. In Imaging, Echo has gone from M Mode to MRI to 3D reconstruction of heart. We will rely more on technology in therapy: robots helping with cardiac surgery. Computer programs are helping to take care of post-op CHD (Congenital Heart Disease). Pacemakers and advancements and transcatheter valves are driving forces. Molecular cardiology-genomic cardiology is a huge field - for example, SUD in Long QT Syndrome. Neurobehavior development in cyanotic CHD (lower IQ, more ADD), cardiomyopathies and transplants.



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**Q: What message would you tell your audience of pediatric cardiologists?**

**HG:** Enjoy it. This is the best thing I can tell you as the audience. It is a great field. It is an open door. It is beautiful. It is totally amazing. Pediatric Cardiology touches on everything.

**Q: What was your education/training, and how did you get into Pediatric Cardiology?**

**HG:** I went to Columbia HS in Maplewood, NJ. I was asked what I wanted to be in 9<sup>th</sup> grade to make class selection. I wanted to be: 1) a doctor, and 2) a teacher. I always wanted to be a pediatrician. I was fascinated with cardiology - the heart. I was always interested in arrhythmias that the pediatric cardiologist took care of. The field generated many questions that couldn't be answered. This fascinated me. I went to Washington and Jefferson College in Washington, PA from 1954-58. I had a full scholarship to college and a full medical school scholarship. I was very proud of this. I went to medical school at Thomas Jefferson University in Philadelphia, PA, 1958-1962. I did my Pediatrics training at Mt. Sinai Hospital in NYC for 2 years 1965-67, followed by specialty training in Pediatric Cardiology at Columbia University in NYC, followed by 2 years clinical training at Columbia University under Dr. Sidney Blumenthal, 1967-69. I did 2 years of research in Brian Hoffman's lab from 1969-1971.

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***Dr. Gelband's Career Advice: "Don't ever let the job interfere with love of kids, wife and family life!!! You have to be happy - separate the two. If there is a problem at work, don't bring it home. If there is a problem at home, don't bring it to work!"***

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**Q: How did you know the University of Miami was the right choice?**

**HG:** Dr. Manny Papper came from Columbia and he became the Dean of Medical School (he hired me). Sidney Blumenthal was Associate Dean at U Miami, and came to join Manny Papper (He was Division Chief of Pediatric Cardiology at Columbia). Mary Jane Jesse was Chief of Cardiology at U Miami, she came from Columbia. Robert Myerberg was Chief of Cardiology at the VA Hospital in Miami, and he had shared a lab with me, and was running a lab there (he did EP research at Columbia with Brian Hoffman). Gerard Kaiser, surgeon, came from Columbia - the youngest Chief of Surgery ever. Arthur Bassett came to the lab to do research. Everyone got along so well already; this continued once I got to Miami.

**Q: How did you instill confidence and growth in your faculty/fellows?**

**HG:** First, you have to get younger faculty to be inquisitive and seek their interest in an issue and develop an idea. Second, I trusted fellows/faculty, but I knew what they were doing in the lab and in the clinic. And third, there were no barriers to get what they needed (equipment-wise or resource-wise), so this made them successful.



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## Heart Program

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Founded in 1950, the rebranded Nicklaus Children's Hospital is renowned for excellence in all aspects of pediatric medicine and has numerous subspecialty programs that are routinely ranked among the best in the nation. It is also home to the largest pediatric teaching program in the southeastern U.S. Many of our physicians have trained or worked at other leading medical institutions. Be part of a phenomenal team that brings lifelong health and hope to children and their families through innovative and compassionate care.

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  - regurgitation: ≥ moderate regurgitation, AND/OR
  - stenosis: mean RVOT gradient ≥ 35 mm Hg

**Contraindications:** None known.

### Warnings/Precautions/Side Effects:

- DO NOT implant in the aortic or mitral position. Preclinical bench testing of the Melody valve suggests that valve function and durability will be extremely limited when used in these locations.
- DO NOT use if patient's anatomy precludes introduction of the valve, if the venous anatomy cannot accommodate a 22 Fr size introducer, or if there is significant obstruction of the central veins.
- DO NOT use if there are clinical or biological signs of infection including active endocarditis. Standard medical and surgical care should be strongly considered in these circumstances.
- Assessment of the coronary artery anatomy for the risk of coronary artery compression should be performed in all patients prior to deployment of the TPV.
- To minimize the risk of conduit rupture, do not use a balloon with a diameter greater than 110% of the nominal diameter (original implant size) of the conduit for pre-dilation of the intended site of deployment, or for deployment of the TPV.
- The potential for stent fracture should be considered in all patients who undergo TPV placement. Radiographic assessment of the stent with chest radiography or fluoroscopy should be included in the routine postoperative evaluation of patients who receive a TPV.
- If a stent fracture is detected, continued monitoring of the stent should be performed in conjunction with clinically appropriate hemodynamic assessment. In patients with stent fracture and significant associated RVOT obstruction or regurgitation, reintervention should be considered in accordance with usual clinical practice.

Potential procedural complications that may result from implantation of the Melody device include the following: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, blistering, or peeling of skin, pain, swelling, or bruising at the catheterization site.

Potential device-related adverse events that may occur following device implantation include the following: stent fracture, \*stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, hemolysis.

\*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

For additional information, please refer to the Instructions For Use provided with the product.

**CAUTION:** Federal law (USA) restricts this device to sale by or on the order of a physician.

### Important Labeling Information for Geographies Outside of the United States

**Indications:** The Melody Transcatheter Pulmonary Valve is indicated for use in patients with the following clinical conditions:

- Patients with regurgitant prosthetic Right Ventricular Outflow Tract (RVOT) conduits with a clinical indication for invasive or surgical intervention, OR
- Patients with stenotic prosthetic RVOT conduits where the risk of worsening regurgitation is a relative contraindication to balloon dilation or stenting.
- Existence of a full (circumferential) RVOT conduit that was equal to or greater than 16 mm in diameter when originally implanted.

The intended lifetime for the Melody device is 2 years.

### Contraindications:

- Venous anatomy unable to accommodate a 22 Fr size introducer sheath; implantation in left heart.
- Unfavorable right ventricular outflow tract for good stent anchorage.
- Severe right ventricular outflow obstruction, which cannot be dilated by balloon.
- Obstruction of the central veins.
- Clinical or biological signs of infection.
- Active endocarditis.
- Known allergy to aspirin or heparin.
- Pregnancy.

**Potential Complications/Adverse Events:** Potential procedural complications that may result from implantation of the Melody device include the following: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, pain at the catheterization site.

Potential device-related adverse events that may occur following device implantation include the following: stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, hemolysis.

For additional information, please refer to the Instructions For Use provided with the product.

The Melody Transcatheter Pulmonary Valve and Ensemble II Transcatheter Delivery System has received CE Mark approval and is available for distribution in Europe.



## OPPORTUNITY IN PEDIATRIC CARDIOLOGY LAREDO, TEXAS

Driscoll Children's Hospital is advancing a comprehensive Heart Center to meet the healthcare needs of congenital heart patients in South Texas. The Center is recruiting a physician to support outpatient clinic activities in Laredo, TX. Sub-specialty board eligible or certification is required. Spanish speaking is preferred.

Pediatric Cardiology has been an integral part of Driscoll Children's Hospital since 1962. The Hospital and the Heart Center are committed to bringing state-of-the-art technology and quality service to 31 counties in South Texas. In 2013, the Heart Center saw 9,500 outpatient and satellite visits; 6,121 echocardiograms, including 500 fetal echos, and 192 heart catheterizations (82% interventional). The Laredo Clinic is a major clinic affiliated with Driscoll Children's Hospital. The Heart Center employees 8 physicians including 1 Electrophysiologist, 2 Interventional cardiologists, 1 MRI Imaging cardiologist, and 1 fetal cardiologist. Three pediatric cardio-thoracic surgeons deliver all aspects of surgical service including hybrid procedures.

The Laredo Clinic is a major clinic affiliated with Driscoll Children's Hospital. The cardiologist in Laredo will share a 1:2 call rotation. Physicians will see challenging, complex patients in a beautiful, well-staffed clinic with 2 sonographers. The qualified physician will enjoy a young, fast growing patient base.

Laredo offers a vibrant, multicultural population. With the mild weather, it is a haven for year-round outdoor activities, including golf, cycling, and tennis. South Texas offers world class hunting, fishing, sailing and wind surfing. The cost of living in south Texas is low, and there is no state income tax.

**If you are interested in more information on this excellent opportunity, please contact:**

John Brownlee, MD  
Cardiology Medical Director  
[John.Brownlee@dchstx.org](mailto:John.Brownlee@dchstx.org)

or

Annette Shook  
Physician Recruiter  
Driscoll Children's Hospital  
(361) 694-6807 or  
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## CONGENITAL CARDIOLOGY TODAY

**CALL FOR CASES AND OTHER ORIGINAL ARTICLES**

Do you have interesting research results, observations, human interest stories, reports of meetings, etc. to share? Submit your manuscript to: [RichardK@CCT.bz](mailto:RichardK@CCT.bz)

**Q: Where did you get your work ethic?**

**HG:** From my parents and my background. I was born in Vienna, Austria. My father was in Dachau in a Concentration Camp, but got out in 1939. He recognized it was best to leave Europe, and in 1939 we went to Cuba. We immigrated to US in 1941, and I came to Newark, NJ as a 5-year-old child (I did not speak English then). My mother passed away when I was age 13, and my father passed when I was 25, so I grew up independent. I helped with the family business at a young age. I had an older sister (1.5 years older) that helped raise me.

**Q: What books do like to read (Dr. Gelband lives next to a library, and gets two books/month)?**

**HG:** I'm a mystery reader: Detective/CIA/ mysteries especially. I like Mickey Spillane, Robert Parker, Lincoln Lawyer, and some non-fiction, like *Unbroken*.

**Q: What movies do you like?**

**HG:** I'm a Robert DiNiro fan; I liked him in *Raging Bull*. I also like Al Pacino in *The God Father*. I saw Leonard DiCaprio in *The Revenant*.

**Q: What sports teams do you like?**

**HG:** I used to have season tickets to the Miami Heat. Dr. Myerberg and I used to share 4 season tickets. I like U of Miami Hurricane games, both basketball and football. And, I am a Dolphins fan.

**Q. What is your exercise routine now?**

**HG:** I swim at the Community Center 11 am-2 pm. They have an outdoor Olympic size pool. I swim 2.5 miles/week. I occasionally ride a bike, and I work out in the weight room.

**Q: Are you still working at the hospital?**

**HG:** On Tuesdays, I volunteer at University of Miami Jackson Hospital by reading to kids. I go to the Mailman Center, and play with kids which keeps me young. I attend Pediatric Grand Rounds at noon on Tuesdays.

**Q: Do you have any final words we haven't covered?**

**HG:** Being a medical doctor (even today with health care regulations) is a great life.

Being a pediatric cardiologist is extremely rewarding. I did it with a smile, and it was fun. It was a great ride, and I would do it all over again. I was so proud of the relationship with fellows and what those fellows accomplished. I was proud to influence people to carry on what I thought was a great thing.

*Thank you Dr. Gelband.*

**CCT**

**Biographical Sketch**

Dr. Brad Robinson is a graduate of the University of Miami Pediatric Cardiology program in 1992. He currently practices at The Nemours Cardiac Center at the Alfred I. duPont Hospital for Children in Wilmington, DE. He is Associate Professor of Pediatrics at the Sidney Kimmel Medical College at Thomas Jefferson University in Philadelphia, PA. His current interests are: exercise physiology, fetal cardiology and medical education of students and residents and fellows in pediatric cardiology.



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**PHOENIX CHILDREN'S Hospital**

**Advanced Heart Failure, Transplant Cardiologist (Pediatric)**

Phoenix Children's Hospital has an opportunity for a Pediatric Cardiologist with experience in Heart Transplant and Advanced Heart Failure Management to join one of the largest heart centers in the west. Join a thriving cardiac program with broad multidisciplinary expertise, three dedicated pediatric cardiothoracic surgeons and collaborate with 22 cardiologists to provide cutting edge cardiac care for children throughout the region. This transplant and heart failure focused position will join two subspecialty cardiologists, three nurse practitioners and a broad team supporting clinical, administrative and academic growth. With more than 20 pediatric heart transplants performed to date in 2016, the program continues its steady growth in complexity and volume. Targeting patients with advanced heart failure including those needing mechanical circulatory support and/or heart transplant care, opportunities abound for teaching, clinical leadership, as well as clinical and translational research.

**Job Roles**

- Provide subspecialty care in the inpatient and outpatient setting for targeted patient populations: advanced heart failure including those needing mechanical circulatory support, cardiomyopathy, the failing single ventricle and heart transplant candidates/recipients
- Follow hundreds of advanced heart failure and pre/post-transplant patients
- Work with a transplant team of 3 surgeons, 3 dedicated nurse practitioners, 2 transplant cardiologists and staff support program in a manner that fulfills the strategic goals of the Hospital while complying with state and federal laws and accreditation standards related to safety and risk management.
- Support outpatient VAD Program

**Desired Skills & Experience**

**Minimum Qualifications**

- Formal training or experience in pediatric heart transplantation and advanced heart failure
- Board Certified/Eligible in Pediatric Cardiology
- Willingness to apply for academic position with the University of Arizona

**Please send your resume to:**

Steven Zangwill, MD  
[szangwill@phoenixchildrens.com](mailto:szangwill@phoenixchildrens.com)



**Archiving Working Group**  
**International Society for Nomenclature of Paediatric and Congenital Heart Disease**  
[ipccc-awg.net](http://ipccc-awg.net)

# Sports Cardiology and Sudden Cardiac Arrest in the Young

By Anjan S. Batra, MD, FHRS; Mary E. Hickcox

On January 20<sup>th</sup>-21<sup>st</sup>, 2017, CHOC Children's, in affiliation with UC Irvine School of Medicine, will host the 4<sup>th</sup> Biennial Sports Cardiology & Sudden Cardiac Arrest in the Young Conference at Disney's Grand Californian Hotel in Anaheim, CA.

Dr. Anjan S. Batra, CHOC Children's Division Chief and Medical Director of Electrophysiology, as well as Vice Chair of Pediatrics at UC Irvine School of Medicine, will be the Program Chair. In previous years, more than 152 participants and faculty from all over the United States (Alaska, Arizona, Alabama, California, Colorado, Florida, Georgia, Iowa, Illinois, Louisiana, Massachusetts, Maryland, Maine, Michigan, Minnesota, North Carolina, Nevada, New York, Ohio, Oregon, South Carolina, Texas, Utah and West Virginia) have attended. Additionally, an international presence was represented from Australia, Canada and Puerto Rico.

The Pediatric & Congenital Electrophysiology Society (PACES) - <http://pediatricpsociety.org> is an international group of physicians and allied professionals dedicated to improving the care of children and young adults with cardiac rhythm disturbances. This nonprofit organization has endorsed this year's conference, and will host a special task force meeting at the event. The Task Force is an advocate in several key areas of Sudden Cardiac Arrest (SCA) prevention including CPR/AED training. Task Force Chairs Drs. Jack Salerno and Chris Erickson have been working on a position paper examining issues or problems with identification of those at risk for SCA. The group's primary mission is to foster high-quality collaborative research and exchange of ideas on arrhythmia topics that are particularly relevant to infants and children, or patients of any age with Congenital Heart Disease.

## Background

There are 25 million competitive athletes in the United States who are involved in competitive activities, including 10 million high school and college athletes. We do not know exactly how many athletes are dying each year, as there is no national registry keeping track of these numbers. The risk for SCA for the young competitive athlete population is estimated to be 2 per 100,000 persons per year,<sup>1</sup> and it is at least 2.5-fold higher than that of the age-matched non-athlete population.<sup>2</sup> The risk for sudden cardiac death increases with increasing peak intensity of exercise and increasing level of competition.<sup>3,4</sup> Maron et al tracked sudden deaths in U.S. competitive athletes using a large registry over a 27-year period, and reported 82% deaths with physical exertion during competition/training in males, and only 11% deaths in females.<sup>5</sup> The most common cardiovascular causes were Hypertrophic Cardiomyopathy (36%) and congenital coronary artery anomalies (17%).

## Orange County, California, Experience

Orange County has a population of 3 million, making it the sixth most populous county in the United States. There are 504,072 students in Orange County public schools and 58,008 students in Orange County private schools. High school students comprise approximately one third of this population. Orange County has the largest number of NCAA and Olympic athletes per capita of any county in the United States. The precise incidence of life-threatening events in this population is unknown. However, within the last year, there have been three deaths of high school athletes reported by the media. In addition, there were other cases of life-threatening events that were not reported by the media because the athletes either completely recovered or went on to succumb to irreversible neurological injury.

The Life-Threatening Events Associated with Pediatric Sports (LEAPS) Initiative emerged in Orange County 4 years ago. This included an annual symposium, featuring local and national experts in the area, geared towards school educators, nurses, parents and physicians in the community. There were approximately 150 participants in each of the previous two symposiums. Task forces were created for screening strategies including: ECGs, implementation of an AED program in the schools, improved CPR training within schools, and an incident review team to provide feedback to the school and community after any incident involving a sudden death in a school setting. Each task force is comprised of school educators, coaches, nurses, parents, and physicians.

The efficacy of including ECGs for routine screening remains a highly debated controversial topic. Although the 2007 AHA screening recommendations do not endorse mandated national ECG screening for all competitive athletes, in no way do the recommendations discourage individual initiatives which offer ECG screening, such as the LEAPS Initiative in Orange County, California. More research is needed to evaluate the efficacy of a screening program that includes the ECG.

## Current Challenges

Cardiologists, Sports Medicine Physicians, ED physicians and general pediatricians need to have the most recent information about the impact of cardiovascular effects of athletic training on their patients. Also, they need to use the most recent data to facilitate proper diagnosis and management of the pediatric sports athlete who has inherited cardiac diseases. Lastly, they need to perform complete work-ups or follow recommendations for required screening of athletes to prevent sudden cardiac death. Only 45%-50% of physicians doing sports physicals were familiar with the AHA guidelines and recommendations, while less than 6% of physicians adhere to the full AHA Sudden Cardiac Death screening guidelines.

## Updated Recommendations

In late 2015, authors BJ Maron, DP; Zipes, and RJ Kovacs, wrote an update to the Bethesda #36 report (2005) addressing eligibility and disqualification criteria for competitive athletes with cardiovascular conditions. As with previous reports, its emphasis was toward student athletes (ages 12-25 years).<sup>11</sup> Due to this updated report, Cardiologists, Sports Medicine physicians, ED physicians and general pediatricians need to review, discuss



The Disney Grand Californian Hotel: Sequoia Ballroom.

and assess expert opinions on the 15 task forces' recommendations.

If you share these experiences or have an interest in this area, join us at Disneyland in January! Plan to attend the 4<sup>th</sup> Biennial Sports Cardiology & Sudden Cardiac Arrest in the Young Conference – January 20-21, 2017 in Anaheim, California. As in 2015, this biennial conference will be held at Disney's Grand Californian Hotel, Sequoia Ballroom, located at 1600 Disneyland Drive, Anaheim, California. Special room rates are available for conference registrants.

Participate in lively round table discussions, pro and con debates, and case-based review on the effects of athletic training, the diagnosis and management of individuals with inherited cardiac diseases, and strategies to prevent sudden death in the young.

Keynote Speaker - Michael J. Ackerman, MD, PhD, FAAC, a leading expert in genomics and genotype-phenotype relationships in inheritable cardiovascular diseases leading to sudden death, will join us on Friday, January 20<sup>th</sup>, 2017. You will want to be there to participate in his "State of Genetic Testing in Cardiac Channelopathies and Cardiomyopathies - Your Questions Answered!"

Of special note, we are including our first ever nursing-only track on Saturday, January 21<sup>st</sup>.

Register now and receive the *Early Bird* rate! Visit [www.choc.org/scaconference](http://www.choc.org/scaconference). Additional information can be obtained by calling 1-800-329-2900 or email [chocme@choc.org](mailto:chocme@choc.org).

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## Medical Director for Pediatric Cardiac Intensive Care Unit

The Congenital Heart Center at Levine Children's Hospital (LCH) and Sanger Heart & Vascular Institute (SHVI) announces a search for the Medical Director of the Cardiovascular Intensive Care Unit (CVICU). The director would oversee a well-established, multidisciplinary team in the management of pediatric patients with congenital heart disease. The successful candidate will have experience in the management of all forms of congenital heart disease and must excel working within a clinically integrated network of multidisciplinary teams.

The Congenital Heart Center at Levine Children's Hospital and Sanger Heart & Vascular Institute: Established in 2010, the Congenital Heart Center at LCH has consistently ranked as one of the top-50 pediatric heart centers in the country by U.S. News and World Report. Surgical highlights include > 95% Norwood survival, neonatal mortality rates well below national benchmarks, and an overall 30-day surgical survival rate of 98%. Case volume has expanded 15% yearly for 6 consecutive years; currently exceeding 500 cases.

The CVICU is a 10 bed, state-of-the-art unit staffed with experts in the field, including physicians trained in pediatric cardiology and critical care medicine; 24/7 acute care-trained nurse practitioners and dedicated critical care nurses, respiratory therapists, and a pharmacist who received specialized training in congenital heart disease.

The CVICU provides comprehensive care for complex medical and surgical patients, from newborns to adults. We are equipped to provide the highest complexity therapies including mechanical support with extracorporeal membrane oxygenation and ventricular assist devices (including the Berlin Heart, HeartMate, and Syncardia devices).

Candidate must be licensed MD/DO and Board Certified in Pediatric Critical Care Medicine with additional Pediatric Cardiology certification preferred. Those with critical care certification who have demonstrated an exceptional track record in the CVICU will also be considered.

## For more information, please contact our Physician Recruiter:

Lisa.Webster@CarolinasHealthCare.org  
or 704-631-1126



Carolinas HealthCare System

# Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

## For Normal Heart Function, Look Beyond the Genes

Newswise — Researchers have shown that when parts of a genome known as enhancers are missing, the heart works abnormally, a finding that bolsters the importance of DNA segments once considered “junk” because they do not code for specific proteins.

The team, led by scientists at the Department of Energy's Lawrence Berkeley National Laboratory (Berkeley Lab), examined the role of two heart enhancers in the mouse genome, showing that the loss of either one resulted in symptoms that resemble human cardiomyopathy, a disease in which the heart muscle often becomes enlarged or rigid. In humans, the disease often leads to heart failure.

The findings appeared in a study published Oct. 5<sup>th</sup> in the journal *Nature Communications*.

In that same paper, the researchers provided a comprehensive genome-wide map of more than 80,000 enhancers considered relevant to human heart development and function. The two heart enhancers that they tested were the mouse equivalent of enhancers chosen from among that catalog.

"The cardiac changes that we observed in knockout mice lacking these enhancers highlight the role of noncoding sequences in processes that are important in human disease," said study co-senior author Axel Visel, Senior Staff Scientist and one of three lead researchers at the Mammalian Functional Genomics Laboratory, part of Berkeley Lab's Environmental Genomics and Systems Biology (EGSB) Division. "Identifying and interpreting sequence changes affecting noncoding sequences is increasingly a challenge in human genetics. The genome-wide catalog of heart enhancers provided through this study will facilitate the interpretation of human genetic data sets."

Study lead author Diane Dickel, Project Scientist, and co-senior author Len Pennacchio, Senior Staff Scientist, both work with Visel at Berkeley Lab's Mammalian Functional Genomics Laboratory.

### DNA Dark Matter

When scientists sequenced the human genome, they discovered that less than 5 percent of our DNA were genes that actually coded for protein sequences. The biological functions of the noncoding portions of the genome were unclear.

Over the past fifteen years, however, there has been a growing appreciation for the importance of these noncoding regions, thanks in large part to the efforts of individual labs and, more recently, large international efforts such as the Encyclopedia of DNA Elements (ENCODE) project.

What became clear from this work is that there are many elements of the genome, including enhancers, that are involved in regulating gene



## **AIMed: Artificial Intelligence in Medicine**

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The Ritz-Carlton  
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## **Sports Cardiology & Sudden Cardiac Arrest in the Young**

January 20-21, 2017

Disney's Grand Californian Hotel  
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## **NeoHeart: Cardiovascular Management of the Neonate**

March 22-25, 2017

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expression, even though they do not encode for proteins directly.

This realization meant that there were vast sections of the genome that needed to be explored and understood. Dickel noted that there are about 20,000 genes in the mouse genome, and in many cases, scientists have a fairly good understanding of what will happen if any one of them is disabled. In contrast, there are 80,000 candidate heart enhancers in the human genome, and it is still unclear how important they are for human development.

"In genetic studies, the way you establish whether a gene is important is you delete it from the genome and see what happens," said Dickel. "In many cases, there are genes that, if disabled, make it difficult for the organism to survive. For enhancers, it's less known what the consequences are if they are damaged or missing. To use a car analogy, if we took the battery out of a car, it wouldn't start. That's a critical component. A missing or damaged enhancer could be essential like a battery, or more similar to a missing passenger seat in the car. It's not as nice, but it's still possible to drive the car."

#### Mapping and Testing the Enhancers

To assess the function of heart enhancers, the researchers first compiled a single road map to guide them. They used results from a technology called ChIP-seq (chromatin immunoprecipitation sequencing) to identify the likely heart enhancers in the human genome.

The researchers say this map will become an important tool as advances in genomics usher in a new era of personalized medicine.

"This compendium of human heart enhancers will be a valuable resource for many disease researchers who have begun adopting whole genome sequencing of patients to look for disease-causing mutations in both the coding and noncoding portion of the genome," said Dickel.

Using the map, the researchers picked two enhancers located near genes associated with human heart disease. They then determined their equivalent enhancers on the mouse genome and disabled them in mice.

They compared the mice with the disabled enhancers with control mice that had no mutation and saw very large changes in gene expression in the test mice.

Echocardiograms used to image the hearts from the two groups of mice confirmed that the heart tissue of mice with a disabled enhancer was pumping with less power than normal, consistent with the signs of human cardiomyopathy.

"Prior to this work, no study had looked at what happens to heart function as a result of knocking out the heart enhancers in the genome," said Dickel. "What was surprising to me was that outwardly, the knockout mice seemed fine. If you just looked at them, you wouldn't necessarily see anything wrong."

With so many enhancers to test, the map could help scientists prioritize which ones to assess in animal studies and in disease research, the researchers said.

The National Institutes of Health provided funding for this research.

The University of California manages Berkeley Lab for the U.S. Department of Energy's Office of Science. For more, visit [www.lbl.gov](http://www.lbl.gov).

DOE's Office of Science is the single largest supporter of basic research in the physical sciences in the United States. For more information, please visit [science.energy.gov](http://science.energy.gov).

## CONGENITAL CARDIOLOGY TODAY

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## Upcoming Medical Meetings

### AIMed

Dec. 12-15, 2016; Laguna Niguel, CA USA  
[Aimed-mi3.com](http://Aimed-mi3.com)

### Pediatric and Adult Interventional Cardiac Symposium (PICS 2017)

Jan. 16-19, 2017; Miami Beach, FL USA  
[www.picsymposium.com](http://www.picsymposium.com)

### Sports Cardiology & Sudden Cardiac Arrest in the Young

Jan. 20-21, 2017; Anaheim, CA USA  
[www.choc.org/events/sudden-cardiac-arrest-young-2016/](http://www.choc.org/events/sudden-cardiac-arrest-young-2016/)

### PEDIRHYTHM VII:

### Pediatric & Congenital Rhythm Congress

Feb. 4-7, 2017; Thessaloniki, Greece  
[www.pedirhythm.org/](http://www.pedirhythm.org/)

### Cardiology 2017

Feb. 22-26, 2017; Orlando, FL USA  
[www.chop.edu/events/cardiology-2017#.V-WXtaO-L5U](http://www.chop.edu/events/cardiology-2017#.V-WXtaO-L5U)



**PICS-AICS**  
Pediatric and Adult Interventional Cardiac Symposium

**MIAMI**

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