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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 Schmehil, 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.[1] 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.[2]

Early surgical repair with direct implantation of the anomalous artery into the main pulmonary artery has provided excellent results.[2] 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.[3] Anomalous aortic origin of the right pulmonary artery is approximately 4 to 8 times more common.[4]

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.[4] 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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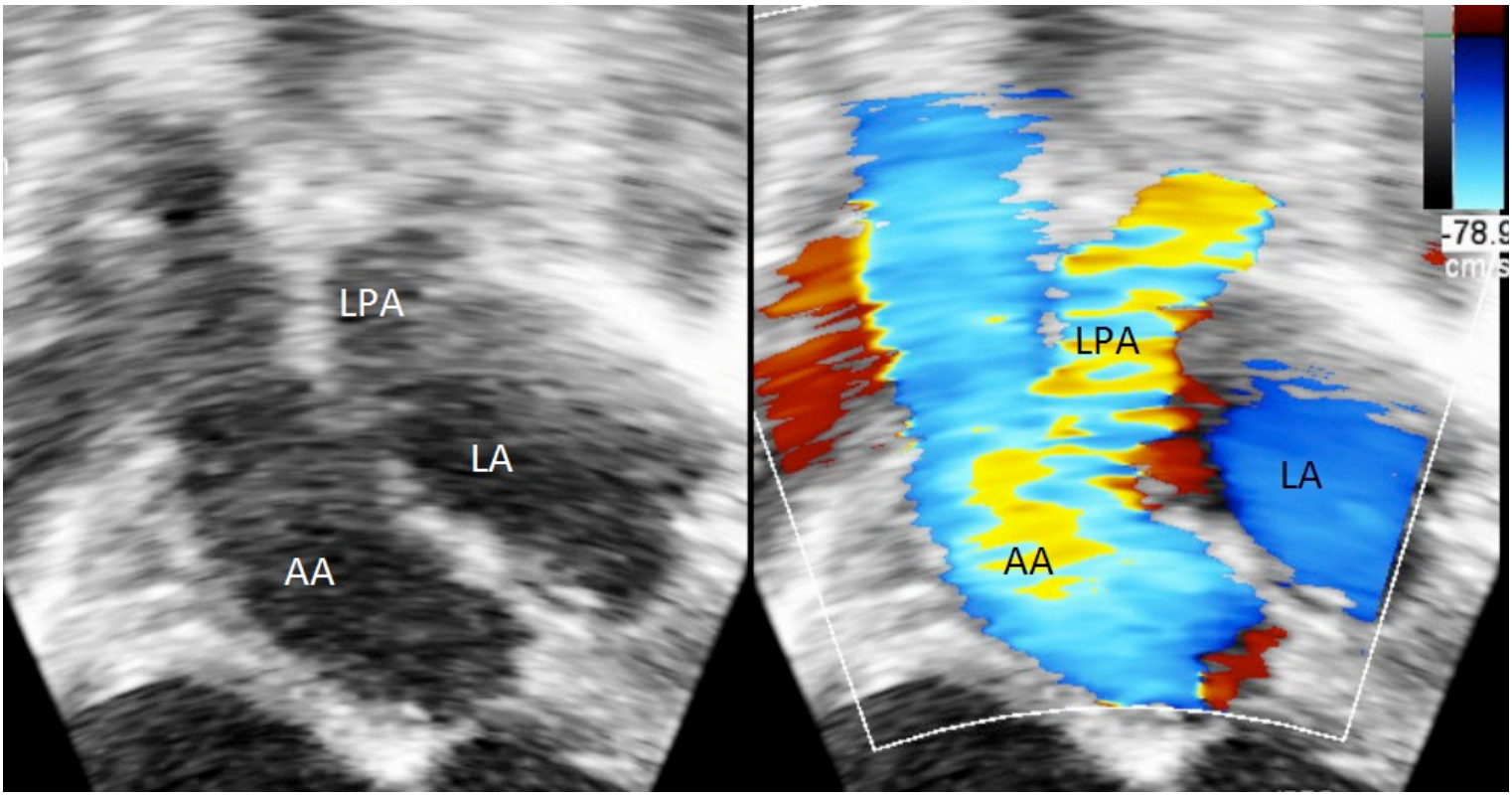

Figure 1. Pre-surgical echocardiogram with color flow, apical view with anterior angulation, reveals ascending aorta giving rise to left pulmonary artery. AA- Ascending aorta, LPA- Left Pulmonary Artery, LA- Left Atrium.

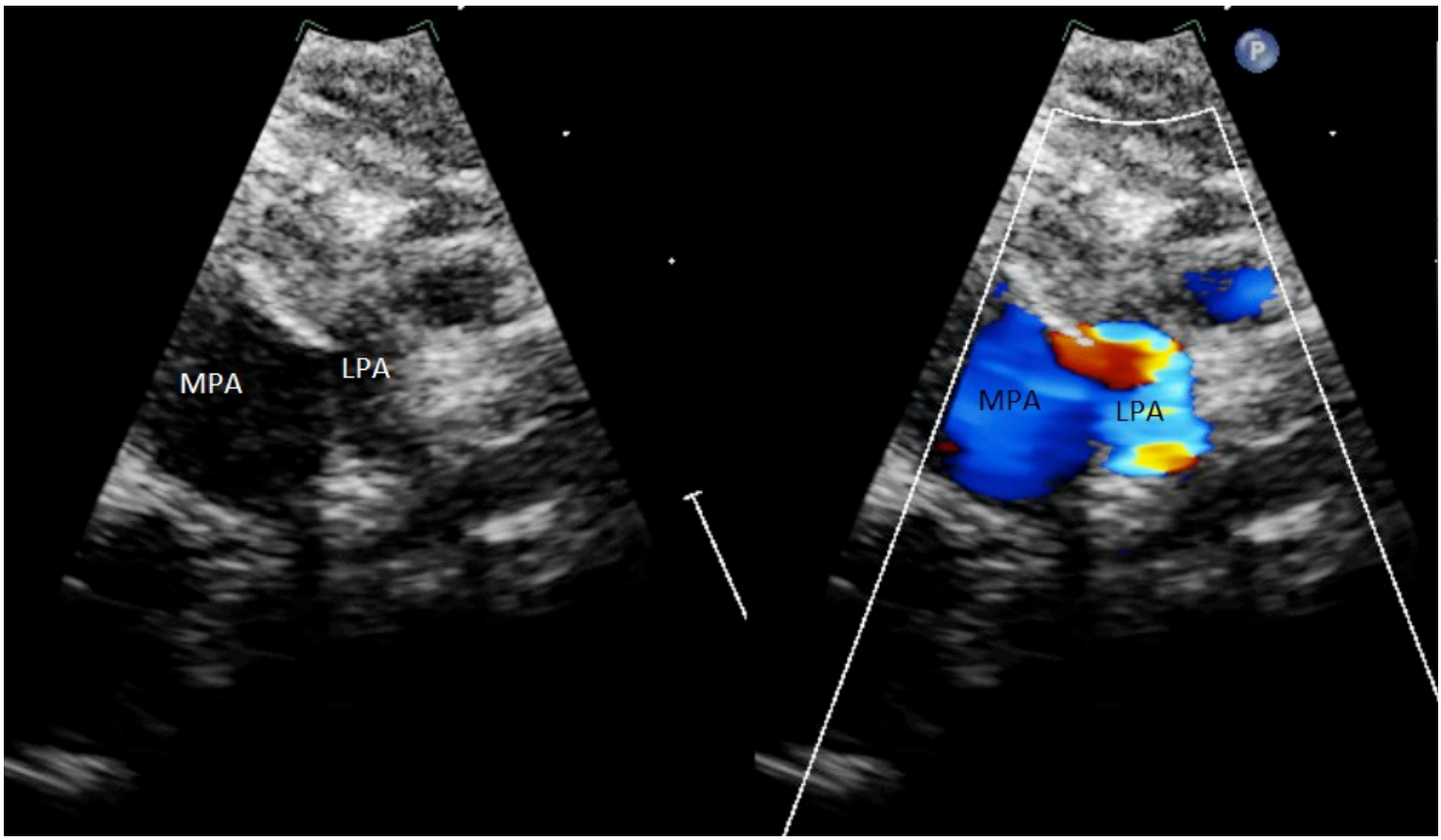

Figure 2. Post-surgical echocardiogram: parasternal short axis with color flow showing reimplantation of the left pulmonary artery to the main pulmonary artery.

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-Pericardiotomy 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.[6] 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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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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Q&A Interview with Dr. Henry (Heinz) Gelband - May 16th, 2016

By Bradley W. Robinson, MD

Dr. Henry (Heinz) Gelband, MD

Preface

I interviewed Dr. Henry (Heinz) Gelband on Monday, May 16th, 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.

Dr. Gelband's Advice to Follows: "If opportunity to do want you want exists, then GO DO IT: don't make excuses!"

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.

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."

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

HG: We are reinventing ourself 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.

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

HG: Enjoy it. This 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 9th 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.

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!"

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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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 catherizations (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.

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

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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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Sports Cardiology and Sudden Cardiac Arrest in the Young

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

On January 20th-21st, 2017, CHOC Children's, in affiliation with UC Irvine School of Medicine, will host the *4th 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://pediatricepsociety.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.

The Disney Grand Californian Hotel: Sequoia Ballroom.

Background

There are 25 million competitive athletes in the United States who are involved in competitive athletes involved in a network of sporting 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,[1] and it is at least 2.5-fold higher than that of the age-matched non-athlete population.[2] The risk for sudden cardiac death increases with increasing peak intensity of exercise and increasing level of competition.[3,4] 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.[5] 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).[11] Due to this updated report, Cardiologists, Sports Medicine physicians, ED physicians and general pediatricians need to review, discuss

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 *4th 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 20th, 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 21st.

Register now and receive the *Early Bird* rate! Visit www.choc.org/scaconference. Additional information can be obtained by calling 1-800-329-2900 or email choccme@choc.org.

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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. 5th 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

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.

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The University of California manages Berkeley Lab for the U.S. Department of Energy's Office of Science. For more, visit www.lbl.gov.

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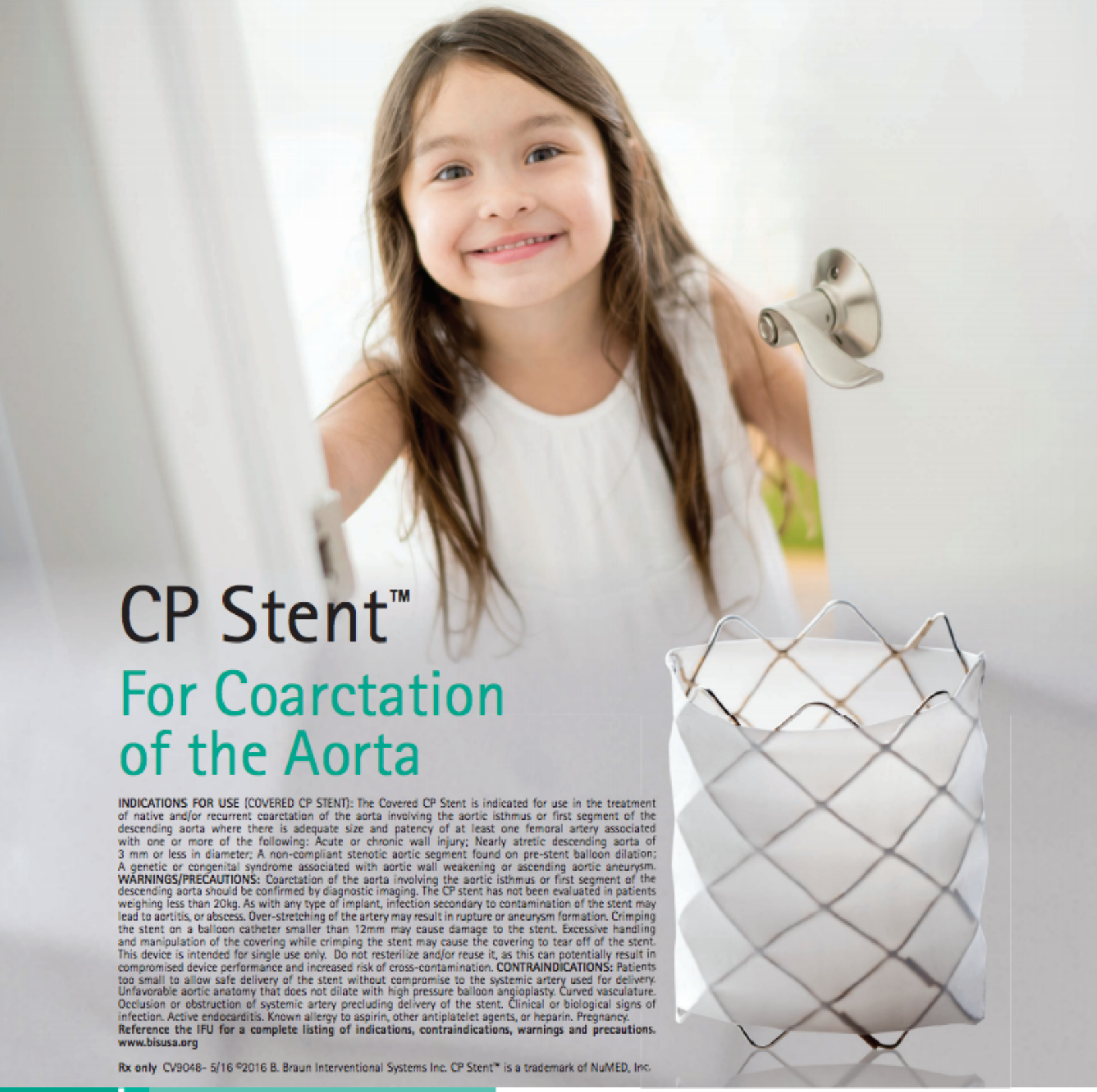

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