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Variations of Coronary Origin: A Case of Bland White Garland Syndrome via 64 Slice Cardiac CT Angiography

By Shah Azmoon, MD; Matthew Budoff, MD; FACC, FAHA, FSCAI; and David Atkinson, MD

Abstract

Background: Anomalies of the coronary arteries have been described as early as the 1800's. In 1933, Drs. Bland, White and Garland described the clinical syndrome of heart failure seen in infants with anomalous origin of the left coronary artery arising from the pulmonary artery. Mortality rates have been reported to be greater than 90% within the first year of life if left untreated. Two dimensional echocardiography with Doppler color flow mapping has been frequently used for initial diagnosis, whereas conventional coronary angiography remains the gold standard.

Case Presentation: Here we present a case of Bland White Garland Syndrome in which the left coronary arteries arise from the pulmonary artery in a symptomatic young child. Diagnosis was promptly made using 64 slice Cardiac CT Angiography after failed attempts in diagnosis using other modalities and the patient was sent for surgical correction. Using low dose protocols, the estimated radiation dose to the infant was 1 milliseivert.

Conclusion: The high spatial and temporal resolution of 64 slice cardiac CT angiography may provide an alternate imaging modality in the diagnosis of congenital coronary anomalies, avoiding invasive risks of conventional coronary angiography.

Manuscript

Early in fetal myocardial development the myocardium is nourished via myocardial sinusoids where persistence of the sinusoids may lead to coronary aberrancy and fistulae. Coronary vessel formation begins approximately at 32 days of gestation. In as many as 50% of the population, the right coronary artery (RCA) and its conal branch may originate separately (normally single origin from the right coronary cusp). Similarly, in approximately 1% of patients, the left anterior descending artery (LAD) and the left circumflex coronary artery (LCx) may originate separately (normally from the left main artery (LM) via the left coronary cusp). Location of these cusps on the aorta may vary, and while most may be inconsequential, a high origin of a coronary ostium may reduce diastolic coronary artery blood flow[1]. More importantly, origination of LAD or a single left coronary arising from the right sinus of Valsalva, and even origination of the RCA from the left side can have deleterious effects, as it may course between the pulmonary arteries and aorta prior to reaching the left ventricle.

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Vigorous expansion of these large vessels, especially with exercise can compress the coronaries leading to myocardial ischemia and even sudden cardiac death[2-4]. Despite the low incidence of anomalous coronary arteries (< 0.64% of live births[5]), such an anomalous course carries a high mortality due to sudden cardiac death (SCD) in young athletes under the age of 35 years (13%); only second to hypertrophic cardiomyopathy in this age group[6-8]. The most common anomalous origin of a coronary artery is the right-sided origination of the LCx and, when arising from the RCA, it usually has a posterior course to the left ventricle avoiding vigorous contact with larger vessels. Anomalous origin of coronaries has been associated with other congenital heart defects including Tetralogy of Fallot (9%)[9-10].

Anomalous origin of the coronaries from the pulmonary artery (PA) has been documented as far back as the 1800s[11-12]. While most coronary anomalies arise from the aorta, less than 0.4% of patients with congenital cardiac anomalies may have origination of their coronary vessels from the PA (~ 1 in 300,000 live births in the US)[13]. Abnormal division of the cono-truncus into the aorta and PA or abnormal involution and persistence of an endothelial bud on a pulmonary sinus may lead to an aberrant connection to the developing coronary artery. In 1933, Drs. Bland, White and Garland, who, at autopsy, diagnosed the syndrome of early progressive heart failure associated with anomalous origin of the coronary arteries arising from the PA in a 3-month old child, whose father would be the future chairman of radiology at Massachusetts General Hospital[14]. Most commonly, the anomaly is a single left coronary rather than the RCA and in some instances the LCx and LAD may have separate origins from the PA[15-18]. While the clinical presentation may vary depending on the coronary involved as well as its size and distribution, there exists significant hemodynamic compromise in most cases with ensuing myocardial ischemia and progressive ischemic cardiomyopathy. Untreated, mortality has been reported as high as 90% within the first year of life[18]. In the less common variant where both the left and right coronaries arise from the PA, prompt diagnosis and treatment is necessary as such a circumstance is not compatible with survival.

Symptoms of angina in an infant may include: feeding intolerance, irritability or respiratory distress when crying, diaphoresis, pallor, failure to thrive or even shock. Early symptoms may temporarily be masked by the presence of large collateral vessels and rarely child onset symptoms may include precordial pain, dyspnea, tachypnea, shortness of breath, exertional chest pain, syncope or even sudden cardiac death. Upon normalization of the physiologically-elevated neonatal pulmonary vascular resistance in the first four to six weeks of infancy, the low perfusion pressures and low oxygen saturation characteristic of the PA lead to myocardial ischemia and progressive cardiomyopathy. Demand ischemia due to anomalous coronaries from the PA is not noted prenatally because of the parallel circulatory system providing relatively equivalent oxygen concentrations as well as equivalent pressures between the aorta and main pulmonary artery through

"Use of color-flow Doppler imaging has largely avoided the need for cardiac catheterization by providing significantly increased diagnostic accuracy with demonstration of abnormal retrograde anomalous coronary flow, although dependent on PA pressures and development of collaterals."

the patent ductus arteriosus (PDA). With the formation of collateral blood supply between the right and left coronary systems, symptom onset may be delayed, however, coronary steal phenomenon may occur from the higher pressure collateral vessels and retrograde flow into the PA from the anomalous coronary should collaterals arise from normal coronaries[19-21]. A "step up" in oxygen saturation within the PA may be detected on cardiac catheterization in lieu of the left to right shunt, with the ratio of pulmonary blood flow (Qp) to systemic blood flow (Qs) ranging from 1-1.5. Untreated, coronary steal will exacerbate symptoms of heart failure and accelerate complications of ischemia, cardiomyopathy and pulmonary hypertension. Although anomalous coronary origination is usually an isolated defect, association with other congenital heart defects such as patent ductus arteriosus, coarctation of the aorta, ventricular septal defect, Terology of Fallot and Hypoplastic Left Heart Syndrome have been noted[22].

Here we present a case of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). J.O. was born in the Philippines at 8 lbs 2 oz, term without complications. There existed no family history of significant disease, maternal radiation exposure or intake of teratogenic drugs. At 9 months of age, she presented with symptoms of chronic cough and shortness of breath, and found to have cardiomegaly on routine chest X-ray. A two dimensional echocardiogram revealed dilated cardiomyopathy (CM) with a left ventricular (LV) ejection fraction (EF) of approximately 20% with LV thrombus. Patient's family was informed she would need to be on lifetime digoxin and captopril and diagnosed with a history of CM presumed secondary to viral myocarditis. Within the following year J.O.'s family immigrated to the US where she would run out of medications, and was unable to seek medical care for several months.

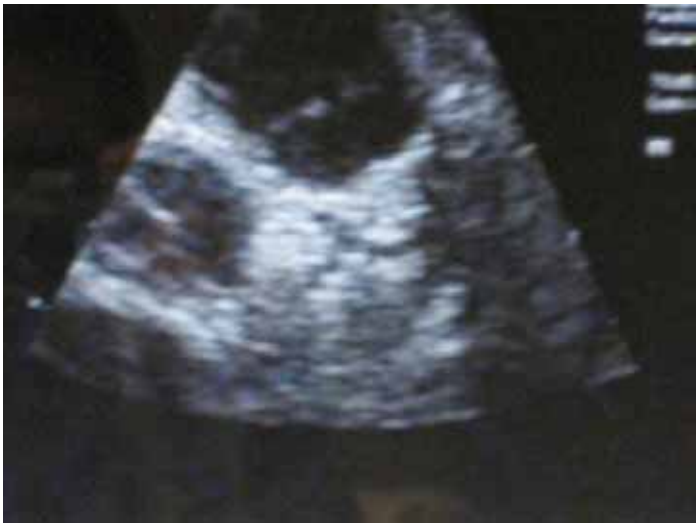

Image 1a: A trans-thoracic echocardiogram with the patient's left main coronary appearing to originate from the pulmonary artery.

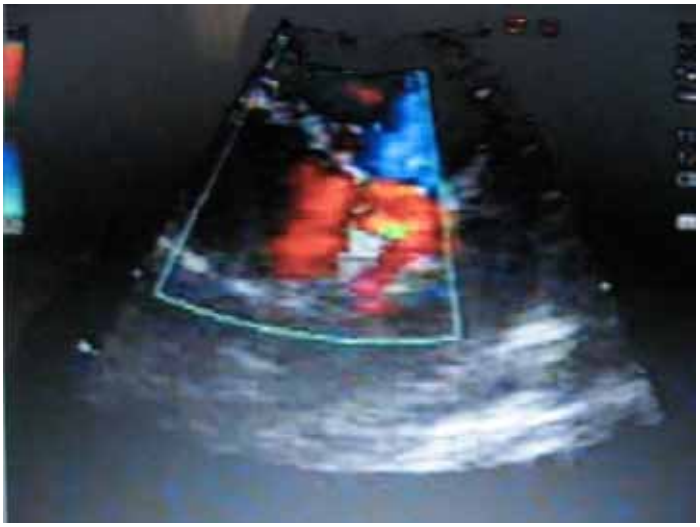

Image 1c: A trans-thoracic echocardiogram with retrograde flow (in red) through the patient's left coronary into the pulmonary artery.

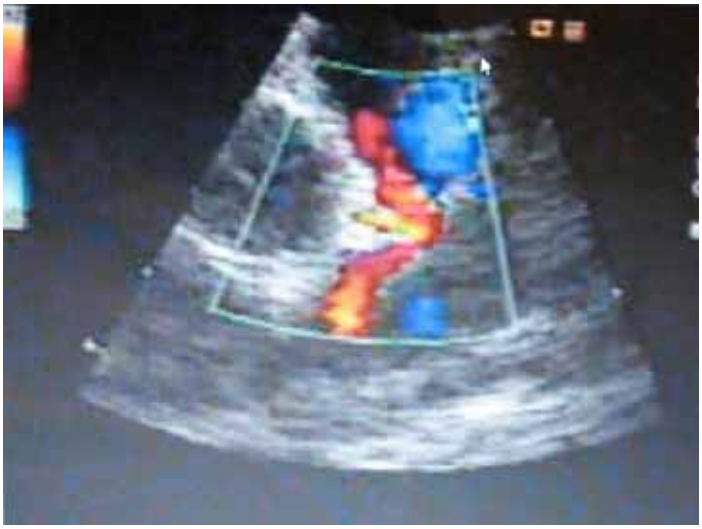

Image 1b: A trans-thoracic echocardiogram with apparent retrograde flow (in red) through the patient's left coronary.

In patients with ALCAPA chest roentegram may demonstrate cardiomegaly with or without pulmonary vascular congestion, although this is not diagnostic. Aside LV hypertrophy, abnormalities of repolarization detected as ST segment depression and/or inversion in the inferior and lateral leads may be noted on electrocardiogram (ECG) along with wide, deep Q waves in the lateral leads with poor R wave progression. On trans-thoracic echocardiogram (TTE) LV dysfunction, mitral regurgitation or mitral annular dilation and wall motion abnormalities in the setting of left heart dilatation may be seen. Enlargement of the proximal right coronary artery may reflect development of extensive collateralization. While most cases of ALCAPA may be diagnosed with echocardiography, two dimensional echo alone may be inadequate as the close course of the anomalous coronary to the aortic sinus may create a false impression of normal anatomic origin (images 1a-1c). Use of color-flow Doppler imaging has largely avoided the need for cardiac catheterization by providing significantly increased diagnostic accuracy with demonstration of abnormal retrograde anomalous coronary flow, although dependant on PA pressures and development of collaterals. However, echocardiographic diagnosis even with Doppler may be difficult should the anomalous coronary arise from a branch pulmonary artery. While retrograde flow into the PA is usually directed in an unusual orientation, improper diagnosis of a PDA shunt or coronary-cameral fistula can be erroneously made. Lack of collateralization may also make identification of ALCAPA by selective right coronary arteriography or aortography difficult in the cath lab, while stop flow angiography may result in a high rate of false-negatives due to incomplete occlusion of the PA. Use of trans-esophageal echocardiography is seldom necessary in infants.

J.O. presented to our clinic with parental complaints of intermittent episodes of nighttime diaphoretic spells, and a history of viral CM. On physical examination she was found to be in no apparent distress, acyanotic, and 50th percentile for height and weight. Her blood pressure was 93-126/44-77 mmHg; she had

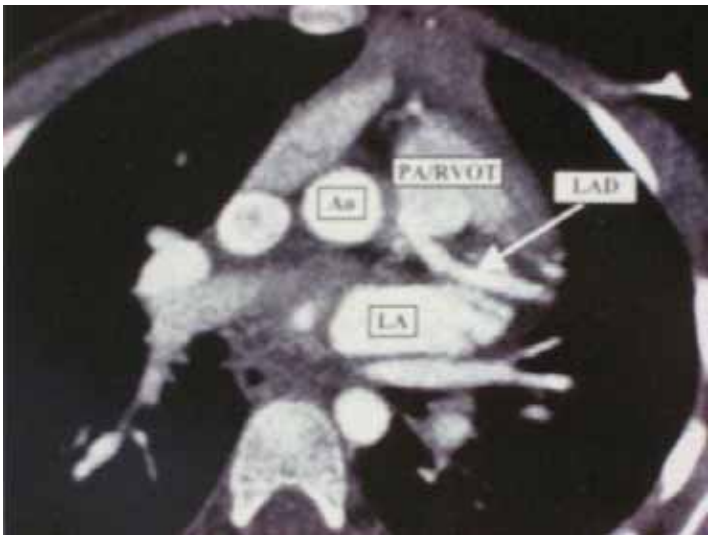

Image 2a: A cardiac CT angiography depicting patient's left main coronary anomalously originating from the pulmonary artery / right ventricular outflow tract.

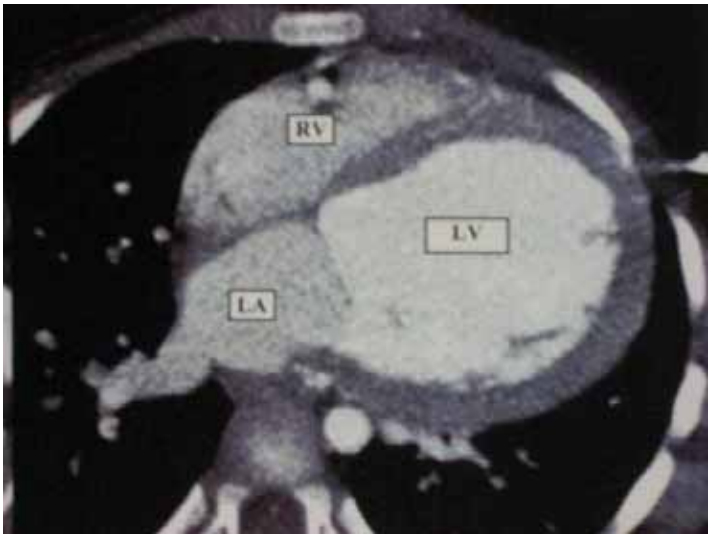

Image 2b: A cardiac CT angiography revealing our 2 year old patient's marked left ventricular dilatation.

a heart rate of 75-125 beats per minute, was afebrile, had a respiratory rate of 20-24, and an oxygen saturation level of 98% on room air. Cardiac examination revealed a visible and lateralized point of maximal intensity, normal rate and rhythm, a 4th heart

sound and II/VI holosystolic blowing murmur of mitral regurgitation. A soft continuous murmur may resemble a persistent PDA with flow from the aorta to the pulmonary circulation or via collateral vessels. Breath sounds were clear bilaterally, without hepatomegaly on abdominal exam and symmetric normal amplitude pulses in all extremities. Her chest radiograph was significant for cardiomegaly without evidence of acute venous congestion. Electrocardiogram revealed sinus tachycardia, Q waves in Lead I and aVL and inferior ST segment depression with T wave inversion suggestive of ischemia in the inferolateral distribution with abnormalities of repolarization. An initial TTE evaluation revealed an EF 15% with appearance of LV thrombus and spontaneous contrast, massively dilated CM with turbulent flow and a small pericardial effusion. The patient was admitted for anticoagulation and further workup of CM which also included a negative Gallium scan as well as mildly elevated PA pressures on right heart catheterization. Repeat TTE during the course of the admission showed resolution of thrombus, EF ~ 25% with appearance of left coronary in some views to arise from the aortic root with a coronary to main PA fistula while in other views it appeared the left coronary would originate directly from the PA. The patient was referred for cardiac CT angiography (CCTA) for better delineation of coronary anatomy (Images 2a-2b).

More than 35 years after diagnosis by Drs. Bland, White and Garland of what is now known as ALCAPA, Drs. Sabiston, Neil and Taussig first showed retrograde flow via an anomalous coronary into the PA with effective treatment by ligation of the anomalous coronary at the junction of the PA. Today, generally surgical intervention is recommended for treatment of anomalous coronary artery when increased risk of ischemia or serious ventricular arrhythmias exists with surgical mortality rates quoted at less than 5-10% by most congenital heart surgery programs[23]. In the case of anomalous coronaries arising from the PA, surgical correction is necessary to avoid further strain on the heart and progressive cardiomyopathy. Surgical procedure of choice remains the direct anastomosis of the anomalous coronary from the PA to the aorta, first described in 1970s. In those young patients in which the anomalous coronary position is not suitable for direct transfer creating an intrapulmonary aorto-coronary tunnel may be required, as described by Takeuchi and colleagues in 1979. However, this procedure may result in tunnel stenosis, aortic regurgitation or pulmonary artery stenosis[23]. When significant cardiac dysfunction is present evaluation for cardiac transplantation may be necessary. While coronary anomalies may also be corrected by coronary artery bypass grafting or stenting, precutaneous treatment options for coronary anomalies of pulmonary origin do not exist[24-25]. Prognosis after surgical correction is frequently excellent with improvement of global left ventricular dysfunction or mitral regurgitation, however patients are still susceptible to atheromatous as well as non-atheromatous coronary stenoses. Acute coronary takeoff with non-atheromatous ridge formation, coronary hypoplasia, myocardial bridging or surgical reimplantation of coronaries may lead to increased risk of coronary stenosis[26-28]. Coronary enlargement, which tends to increase with age, may also occur when collateral vessels are of normal origin and coronary steal is present[29].

While, traditionally cardiac catheterization has been used as the gold standard for diagnosis of coronary anomalies, angular restriction of angiographic projections and limitations by its planar imaging nature may render conventional angiography less useful for a more clear anatomic picture when compared to newer imaging modalities. Conventional angiography is also invasive and carries a morbidity and mortality rate of 1.5% and 0.15%, respectively[30]. With progressive improvement in both resolution and technical specifications, as well as multi-planar reconstruction with maximum intensity projections and volume rendering, cardiac CT angiography may be an adequate alternative diagnostic tool in the detection of coronary anomalies. While in younger patients a short investigation time and minimal after care provides practical usefulness, multi-angle assessment using three dimensional reconstruction can provide optimal vessel projection for evaluation of surgical intervention and thus avoid the repeated exposure to radiation and contrast required with conventional angiography. Clinical usefulness of CCTA in evaluation of infants with complex congenital heart diseases has been successfully evaluated[31], and published reports have also noted superiority of CCTA in defining ostial origins and proximal paths of anomalous coronaries when compared to conventional angiography[32]. More importantly, we hope to draw attention with this case of ALCAPA to the need for early recognition of coronary anomalies when presented with clinical symptoms of heart failure or ischemia in infants.

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Saving Lives from Arizona to Texas and Across the Nation…Texas Cardiac Screening Pilot Program Task Force

By Sharon Bates and Laura Friend, Parent Heart Watch co-founders

Over seven years ago Anthony Bates, a Mountain Pointe High School graduate and Arizona native, died after a weight training football workout at Kansas State University. Anthony, age 20, was the only child of Sharon Bates and since his sudden cardiac death from undiagnosed Hypertrophic Cardiomyopathy (HCM), Sharon formed the Anthony Bates Foundation (ABF). The focus of ABF is education and awareness of heart disease in young people through the volunteer community based cardiac screening program. The life saving cardiac screening program has been offered in Kansas, Arizona, Nevada and Washington. Over 4400 young people have received free or low cost echocardiograms (ultrasounds), ECG and blood pressure tests. With the average abnormality rate well over 10%, Ms. Bates made a stark realization that more needed to be done.

Sudden Cardiac Arrest (SCA) in children is the # 2 killer next to cancer. One out of every 110 babies born today has a congenital heart defect (CHD). Many CHDs will go undetected. The current annual estimates from the CDC states the loss of children under age 18 range from 7,000 to 14,000. More young people die from SCA each year than teenagers in automobile accidents (under 6,000). We put seat belts in cars, helmets on young bicyclists, and pool fences in our back yards to save young life. What is happening in your school to prevent SCA?

The time is now to put preventative screenings at the forefront of children's health concerns. Other countries are doing just that with cardiac screenings in Japan for all children in kindergarten, seventh and ninth grades; Italy has been providing mandatory ECG screening on their athletes since 1984; the UK has instituted cardiac screenings for their athletes in competitive sports; Ireland has recently made great strides to build a cardiac screening program and the Olympic committee requires all Olympic athletes receive an ECG screening every other year. Today many of the professional athletes are getting some kind of cardiac screening, with the NBA requiring echocardiogram and ECG on all athletes and the MLB requiring ECG on all new players each year. Should we wait for a state or federal law to require cardiac screenings? Would you be willing to do more for your community or your own family?

Other states have laws requiring Automatic External Defibrillators (AEDs) in schools. Lives are being saved, and the proof is in the data. According to a recent article in *USA Today*, "Congress pushes for Defibrillators in Schools," 13 lives have been saved with school defibrillators since 2005 in Ohio. In New York state, 38 lives have been saved since 2002. Arizona passed a law requiring AEDs in all state buildings. On any given day, 20% of your community's population will be visiting, attending, or working at a local school. Do you know that legislators, state workers, and prisons now have AEDs, but schools are unprotected?

The Beat Goes On

The internet connected several families that had suffered similar loss of their children and wanted to do more to protect all children in this country. In the summer of 2005 with the generous help of Medtronic Foundation Rachel Moyer (PA), Linette Derminer (OH), Sharon Bates (AZ), and Laura Friend (TX) embarked on the creation of Parent Heart Watch. By early 2006 the formation of a national network of families who have lost a child to sudden cardiac arrest (SCA) was complete. PHW is a non-profit state-by-state network of parents dedicated to reduce the often disastrous effect of SCA. The first membership leader conference was held in January 2006. In attendance were 40 families with heart wrenching stories of loss. Shared at the first meeting were powerful ideas of sorrow, hope, togetherness and collaborative efforts to bring an end to SCA in young people. As of the January 2008 PHW conference, there are over 100 families of SCA victims and several dozen families of SCA survivors that have joined forces to raise the volume on this tragic issue.

As PHW members we spend so much time advocating for AEDs in schools, raising awareness of heart disease in young people, fighting for the lives of young people everywhere, and promoting better preventative cardiac screening programs across the US that we tend to forget to take in the victories of our work. Each one of the PHW members and associates are a huge part of each life saved, all the lives affected by our message, every AED placed in honor of a child lost and every heart screened to prevent SCA. To appreciate the victory of our work would mean we would have to acknowledge our own loss of our loved ones we hold so close to our hearts, and that continues to be hard. But, take a moment with us, and see how far we have come, and the lives we are affecting in our grassroots movement.

On October 10, 2007, Laura Friend, Parent Heart Watch (PHW) Development Director, and Sharon Bates, Anthony Bates Foundation (ABF) CEO/Founder and PHW affiliate member/co-founder, were in Austin, TX, at the initial Texas Cardiac Screening Task Force meeting hosted by Jeff Kloster, Associate Commissioner of the TEA, Texas Education Agency on Health and Safety. In attendance with the TEA, PHW and ABF were representatives from the Texas Governor's office, two Texas legislators' offices (Sen. Hourian & Sen. Janek's offices), two representatives of the American Heart Association and Dr. T. Souryal, one of the team doctors for the NBA Dallas Mavericks.

The background on this pilot program is simple. In June 2007, Texas passed legislation mandating AEDs be present in all high schools, at all sporting events, and that trained personnel are on each campus. There was $9 million appropriated to reimburse schools for these purchases. When the Texas legislators heard testimony from many supporters of their State Bill 7, including our PHW leader, Laura Friend, and the Texas teenage SCA survivor, Matt Nader, there was discussion that Texas could do more with their AED bill than just place AEDs. These senators decided to add a pilot program for cardiac screening that would offer echocardio-

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grams & ECGs tests to a portion of the 6th graders (12-year-olds) in Texas. They amended the State Bill 7 to incorporate a cardiac screening pilot program with intent to cycle in older students as the pilot program is proven and more funding is appropriated. Texas has become a leader in the cardiac screening for youth!

The good news is this new Texas pilot program has been backed with $1 million over a two-term period. The struggle in this scenario will be: who will do the screenings; how can the numbers be proven with cardiac screenings for 12,750 children by June 2009; where and how will it be done (the hope - all over Texas during the school day)? An RFP (Request for Proposal) had been authored by the TEA for this service request in late 2007. Hospitals and large medical groups were encouraged to submit their bid.

Each person in the room that day, and all the PHW membership across this nation have added to the successful opportunity to save lives in Texas. Now as we watch and wait for this program to grow in popularity, we can take a moment and breathe in the success of all the hard work, sacrifices, commitment and motivation that went into "moving mountains" in Texas.

Update to this story: On February 14, 2008, TEA announced in a press conference, the winners of the RFP: Championship Hearts Foundation (formerly Austin Heart Foundation) In Collaboration with Children's Cardiology Associates, Austin, affiliate of Pediatrix Medical Group, Children's Medical Center, Dallas and Texas Children's Hospital, Houston. Now the work begins, and a cost-effective program is created that will save lives and make a big difference to many! The challenge for all of us is: what can we do and/or what are we doing in our communities to raise awareness of heart disease in young people, protect children from SCA, and save lives? For more information visit: www.ParentHeartWatch.org; www.AnthonyBates.org; www.tea.state.tx.us; or www.austinheartfoundation.com

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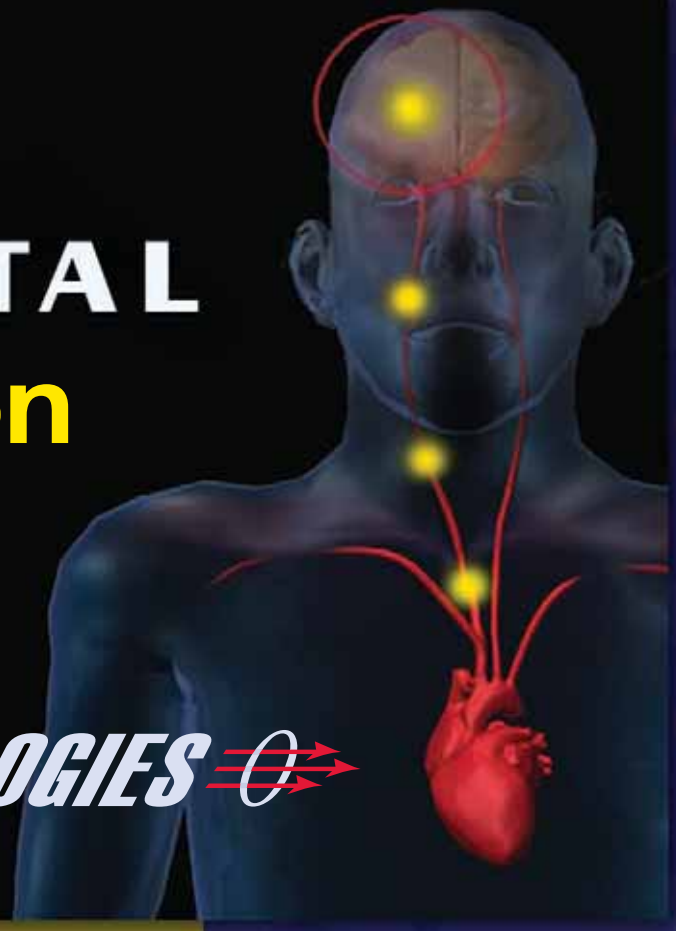

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* J of Neuroimaging, Vol 14, No 4, Oct 2004.

Medical News, Products and Information

Dr. Ziyad M. Hijazi Named President of Society for Cardiovascular Angiography and Interventions

Dr. Ziyad M. Hijazi, MPH, has been appointed as the 31st president of the Society for Cardiovascular Angiography and Interventions (SCAI). A pioneer in the nonsurgical repair of congenital heart defects, Hijazi is an interventional cardiologist who specializes in treating congenital heart disease in both children and adults and is also the director of the Rush Center for Congenital and Structural Heart Disease and professor of Pediatrics and of Internal Medicine at Rush University Medical Center, Chicago. Raised in Jordan, Hijazi is SCAI's first foreign-born president, and is also the first pediatric cardiologist to head the society.

Both factors influence the goals Hijazi has set for his presidency. One of his top priorities is to enlist every pediatric and congenital interventional cardiologist as a SCAI member. He also plans to expand the society's international membership, noting the advantages of broadening collaboration and partnerships worldwide. "The more diverse the membership, the stronger the society," says Hijazi, noting that SCAI is already an international organization devoted to all interventional cardiologists regardless of their subspecialties. "Diversifying our membership even further is good for the profession and good for patients."

In addition, Hijazi plans to expand SCAI's focus on structural heart disease including heart problems such as mitral valve regurgitation that are acquired rather than being present from birth. The society will launch a new committee focused specifically on the needs of physicians who use interventional cardiology procedures to treat structural heart disease. The society will also develop structural heart disease guidelines, training recommendations, educational programs, and criteria for device approval. "I want the field to flourish under SCAI," says Hijazi.

Dr. Hijazi began his training by completing a medical degree and internship in Jordan, and then earned a masters degree in public health at the Yale University School of Medicine. He remained at Yale for a residency in pediatrics and a fellowship in pediatric cardiology. Since then, he has had a distinguished career as both an academician and a practitioner. He spent eight years teaching at Tufts University School of Medicine before moving in 1999 to the University of Chicago, where he served as Chief of Pediatric Cardiology. He assumed his current position at Rush University Medical Center in 2007.

A renowned clinical investigator, Hijazi has written more than 200 articles, 25 book chapters, and three books. His work focuses on developing techniques and devices to address congenital heart problems without open-heart surgery. As a result of his research, the US Food and Drug Administration approved the first device for closing atrial septal defects in children in 2001. In addition to his work on non-surgical repair of defects, Hijazi was also the first to demonstrate the use of intracardiac echocardiography to guide transcatheter closure of atrial septal defects and another problem called patent foramen ovale.

Hijazi is course director of the annual Pediatric Interventional Cardiac Symposium (PICS) cosponsored by SCAI. Dr. Hijazi has also served as a member of SCAI's Board of Trustees and is an editorial board member for the society's public education Web site www.seconds-count.org.

Biosound Esaote Introduces the MyLab 30Gold

Biosound Esaote, a global leader in ultrasound technology, introduced the *MyLab30Gold product at the ASE show in June. The technology of the

successful MyLab30 CV product has been extended with the introduction of a new standard in cardiovascular ultrasound systems, the MyLab30 Gold. This new platform reflects the evolution of ultrasound users' needs: high performance and reliability, coupled with compactness and portability.

Some of the system's updated features include:

- Redesigned ergonomic keyboard
- User-friendly LCD screen with anti-glare high definition
- Battery operation for extreme portability
- Top-of-the-line advanced hardware for extreme performance

"The MyLab30 Gold system represents the next level of excellence in portable ultrasound imaging," said Claudio Bertolini, President of Biosound Esaote. "The Gold platform offers the latest technological innovations with an added ease of use, and represents the continuation of Biosound Esaote's commitment to the market and its customers." For more information on the MyLab30 Gold system and other MyLab ultrasound systems, visit www.biosound.com.

The ACC and the CRF Announce a Multi-Year Partnership to Sponsor the 2009 ACC i2 Interventional Cardiology Meeting

The American College of Cardiology (ACC) and the Cardiovascular Research Foundation (CRF) have announced a multi-year partnership to jointly sponsor the ACC i2 interventional cardiology meeting in Orlando, FL, USA, March 28–31, 2009, during ACC.09, the ACC's 58th Annual Scientific Session. This will be the beginning of a five-year partnership committed to collectively developing the interventional component of the ACC Annual Scientific Session.

The ACC's Innovation in Intervention: i2 Summit, in partnership with the Cardiovascular Research Foundation in Orlando, will deliver in-depth and cutting-edge science provided in a forum for both general cardiologists and interventional specialists, educating and guiding physicians, as well as cardiac care team members, to the next level of knowledge and practice. The meeting will emphasize the translation of evidence-based science and clinical trial data into daily interventional practice.

"We are honored to partner with the ACC to organize and enhance the preeminent interventional cardiology meeting of the spring," said Gregg W. Stone, MD, Chairman of the Cardiovascular Research Foundation and Professor of Medicine at Columbia University Medical Center/New York-Presbyterian Hospital. "The annual scientific session of the American College of Cardiology

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Watch Live Cases from Milan, Italy; Columbus, OH, USA; Toronto, Canada, performed by experts in the field:

Perventricular Muscular VSD - *Drs. Emile Bacha, Ziyad M. Hijazi and Daniel Rowland,*
Perventricular Membranous VSD - *Drs. Hakan Akintuerk, Zahid Amin and Qi-Ling Cao*
Hybrid Stage I Palliation for HLHS PA Bands and PDA Stent - *Drs. Mark Galantowicz and John P. Cheatham*
Intraoperative Aortic Stent for CoA - *Drs. Redmond Burke and Evan Zhan*
Intraoperative LPA Stent Using Endoscopic Guidance - *Drs. Alistair Phillips, Ralf J. Holzer, and Vincent Olshove, CCP*
Creation of ASD after PA Bands & PDA Stent for HLHS in a Preemie - *Dr. John P. Cheatham, Sharon L. Hill, ACNP*
Perventricular Implant of Edwards Valve Stent in the Pulmonary Position - *Drs. Ziyad M. Hijazi and Jinfen Lin*
Closure of Septal Defect Using Real Time 3D Echo Guidance - *Drs. Nikolay V. Vasilyev and Qi-Ling Cao*
High Frequency Ultrasound Creation of ASD - *Drs. Nikolay V. Vasilyev and Qi-Ling Cao*
PmVSD Closure - *Dr. Mario Carminati*
Transcatheter Implantation of Implantable Melody Valve- *Dr. John Cheatham*
Percutaneous Closure of ASD(s) with TEE or ICE Guidance - Percutaneous Valve Implantation - *Drs. Eric Horlick and Lee Benson*
Perimembranous VSD Closure with Amplatzer Membranous VSD Occluder - *Drs. M. Carminati, J. Bass, G.F. Butera, D. Hagler and M. Carrozza*

New live cases will be added in the following months.

If you would like to be notified when additional live cases have been added, please send an email to: LiveCases@CHDVideo.com.
For more information on the symposiums that produced these live cases, and how to attend, please visit:

- **ISHAC** (*International Symposium on the Hybrid Approach to Congenital Heart Disease*) **www.hybridsymposium.com**
- **PICS-AICS** (*Pediatric and Adult Interventional Cardiac Symposium*) **www.picsymposium.com**
- **International WorkshopIPC** (*International Workshop on Interventional Pediatric Cardiology*) **www.workshopipc.com**

www.Live-Cases.com

represents a unique opportunity to introduce the latest advances in interventional medicine to the greater cardiology community. It is our hope that CRF's expertise in cutting edge medical education will help appropriately translate evidence-based science into clinical practice."

"The i2 Summit will offer unsurpassed evidence-based content, presented by leaders in interventional cardiology, and this collaboration brings together the strengths and expertise of both the ACC and CRF to deliver an unparalleled program that appeals to all cardiovascular professionals," said ACC President Douglas Weaver, MD, "The ACC is committed to providing more value to attendees at our meeting by having a wide venue of educational opportunities for physicians including state-of-the-art updates, late breaking clinical trials and education required for credentialing and recertification."

The program committee will assemble a broad-based group of domestic and international interventional cardiologists and other heart disease specialists to provide a forum for interventional cardiology experts to discuss new developments within the field as well as a focused venue to train, educate and guide their colleagues in intervention and cardiac care. The program will feature: late-breaking clinical trials, case-based education, and analysis of advances in treating diseases of the coronary, vascular, and peripheral vessels, as well as pediatric and structural heart disease.

Hybrid Cardiac Catheterization Suite Combines Non Surgical and Surgical Treatments for Heart Defects

A new cardiac treatment facility that couples the benefits of interventional cardiology with cardiothoracic surgery for critically ill newborns, children and adults has opened at Rush University Medical Center.

The new bi-plane hybrid cardiac suite, which is one of only three facilities of its kind in the United States and the only one in Chicago, is equipped with the latest in continuous, real-time imaging technology and radio frequency identification (RFID) technology which allows "all-in-one-room" care.

The suite allows collaboration between the surgeon and interventional cardiologist on complex heart problems. For example, fixing a very large hole in the heart can be done by inserting a catheter through a small incision in the chest rather than relying on major surgery to open the chest to reach the heart.

"Now, interventional cardiologists and cardiothoracic surgeons working together in this suite will reduce the amount of time required to correct complex heart problems and reduce the emotional and physical stress placed on a patient and their family – which translates into less pain, less scarring and a faster recovery time," said Dr. Ziyad Hijazi, Director of the new Rush Center for Congenital and Structural Heart Disease.

The hybrid suite is equipped with the latest technology for minimally invasive interventional cardiology that involves the use of a catheter and an image-guidance system to thread tiny instruments through blood vessels to repair the heart. Through these special catheters, physicians at Rush can implant stents, artificial heart valves and insert patches for holes in the heart.

In many complex cardiac cases, patients who would otherwise have no other option but to undergo open-heart bypass surgery can now have minimally invasive procedures that would otherwise not be available to them.

"We can now communicate with colleagues and obtain their expertise in real time for very complex situations," said Hijazi. "If physicians decide another procedure is needed, even surgery, the suite can be converted into an operating room and the surgical team can be assembled in the new suite."

Unlike the typical set-up at most hospitals where the patient would have to be moved from one area of the hospital to another if additional surgical procedures were needed, patients at Rush will stay in one place in the new hybrid cardiac suite where all the imaging technology and implantable devices that might be needed are stored and located. Thus, patients undergo shorter and safer procedures that require less recovery time and rehabilitation, which can vastly improve patient outcomes. Not having to move the patient to another location, having to administer anesthesia a second time, or having to wait to schedule a second procedure is beneficial to the patients.

"The hybrid suite provides us with a single, state-of-the-art location for diagnosing our patients and providing them with the most advanced non-surgical treatments available for heart defects," said Hijazi. "The additional ability it gives us to provide surgical treatments allows us to provide the most comprehensive care in the most sensitive manner for patients with often extremely fragile conditions."

The new hybrid cardiac catheterization suite has the most advanced imaging technologies, and can still get a precise, optimal image of any region of the heart, regardless of the size or complexity of congenital heart disease. The imaging system also features eight-inch cardiac flat panel detectors designed to deliver distortion-free images.

In addition, the suite includes intravascular ultrasound machines, which take real-time images to allow physicians to see the progress of the procedure taking place inside the patient's body.

A high-tech, automated clinical resource management system located in the suite stores and tracks the medication, surgical tools, medical devices, and implantable devices and supplies using the latest RFID enabled technology.

Study Finds Benefits of ICDs in Children and Inappropriate Shocks for Some

More and more children with congenital heart disease (CHD) are receiving implantable cardioverter-defibrillators (ICDs) to maintain proper heart rhythm. ICDs were first introduced for adults in the 1980's, but little is known about how well they work in children, who account for less than 1% of the recipients. A report in the April 29, 2008 *Journal of the American College of Cardiology* summarized the largest pediatric experience to date. It found the devices to be life-saving, but also suggested that they tended to deliver more inappropriate shocks to children than to adults, making it important to watch children with ICDs closely.

The researchers, led by Charles Berul, MD, a cardiac electrophysiologist at Children's Hospital Boston, analyzed data from 443 patients who received implants between 1992 and 2004 at one of four pediatric centers. The study included both pediatric heart patients and adults with CHD. The median age was 16 years (range, 0 to 54).

Among the 409 patients for whom shock data was available, 26% had received appropriate shocks from their ICD, averaging four per patient. However, 21% had received inappropriate shocks - an average of six per patient. When broken down by age, 24% of children under 18 had received inappropriate shocks, versus 14% of adult patients. While such shocks aren't life-threatening, they are very unpleasant - "like being kicked in the chest," says Berul.

One reason for the inappropriate shocks was that children often have spikes in heart rate that aren't normally seen in adults. "Children are more active and get their heart rates up faster," Berul says. "The devices sometimes have trouble differentiating normal increases in heart rate from abnormal rhythms."

In addition, 14% of inappropriate shocks were due to failure of the ICD wires (leads). This was a particular problem in children, who live much longer with their ICDs than adults do, and whose leads can stretch due to activity and growth. Lead failure also requires periodic surgery to replace the wires.

Berul's team is working with device manufacturers to change ICD programming to minimize unnecessary shocks in children, and to provide more durable, longer-lasting leads.

Overall, the study shows a benefit of ICDs, but suggests that physicians should give more attention to determining which children with congenital heart disease really need the devices, since not all are at risk for life-threatening arrhythmias. For example, patients who had experienced a prior cardiac event had a higher likelihood of appropriate shocks than those receiving the devices preventively (32 vs.18%), improving the overall cost-benefit ratio.

Berul notes that the current study is by far the largest to date involving the newer generation of ICDs. It confirms the findings of earlier studies, which have looked at older-generation pacemakers and devices implanted only after children were resuscitated from cardiac arrest.

This study was funded in part by Medtronic, Boston Scientific, and St. Jude Medical.

The Division of Pediatric Cardiology at the University of Utah School of Medicine and Primary Children's Medical Center is recruiting additional pediatric cardiologists with major interests in:

- **Noninvasive imaging**
- **Outpatient cardiology**
- **Transplant/heart failure**
- **Cardiac intensive care**

Candidates should be BC/BE in pediatric cardiology and should have a strong clinical background in pediatric cardiology with expertise and interest in at least one of the areas listed above. Candidates will join a 21-member Division of Pediatric Cardiology. The Division has a very active, growing clinical program. The Division also has a very active clinical research program and is one of the participating centers in the Pediatric Heart Disease Clinical Research Network funded by the NIH. Protected time and mentoring for clinical research will be available within the Division for clinical research studies.

Successful candidates will receive faculty appointments at the University of Utah. The Pediatric Cardiology Division is based at Primary Children's Medical Center, a tertiary referral center for a three-state area located on the hills overlooking Salt Lake City. The area offers an excellent quality of life with immense cultural and recreational opportunities readily available. The University of Utah is an Equal Opportunity Employer and welcomes applications from minorities and women and provides reasonable accommodations to the known disabilities of applicants and employees.

Interested individuals should send or email a cover letter and curriculum vitae to:
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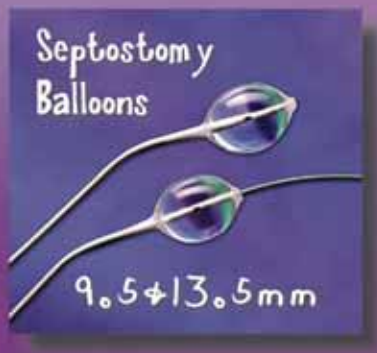

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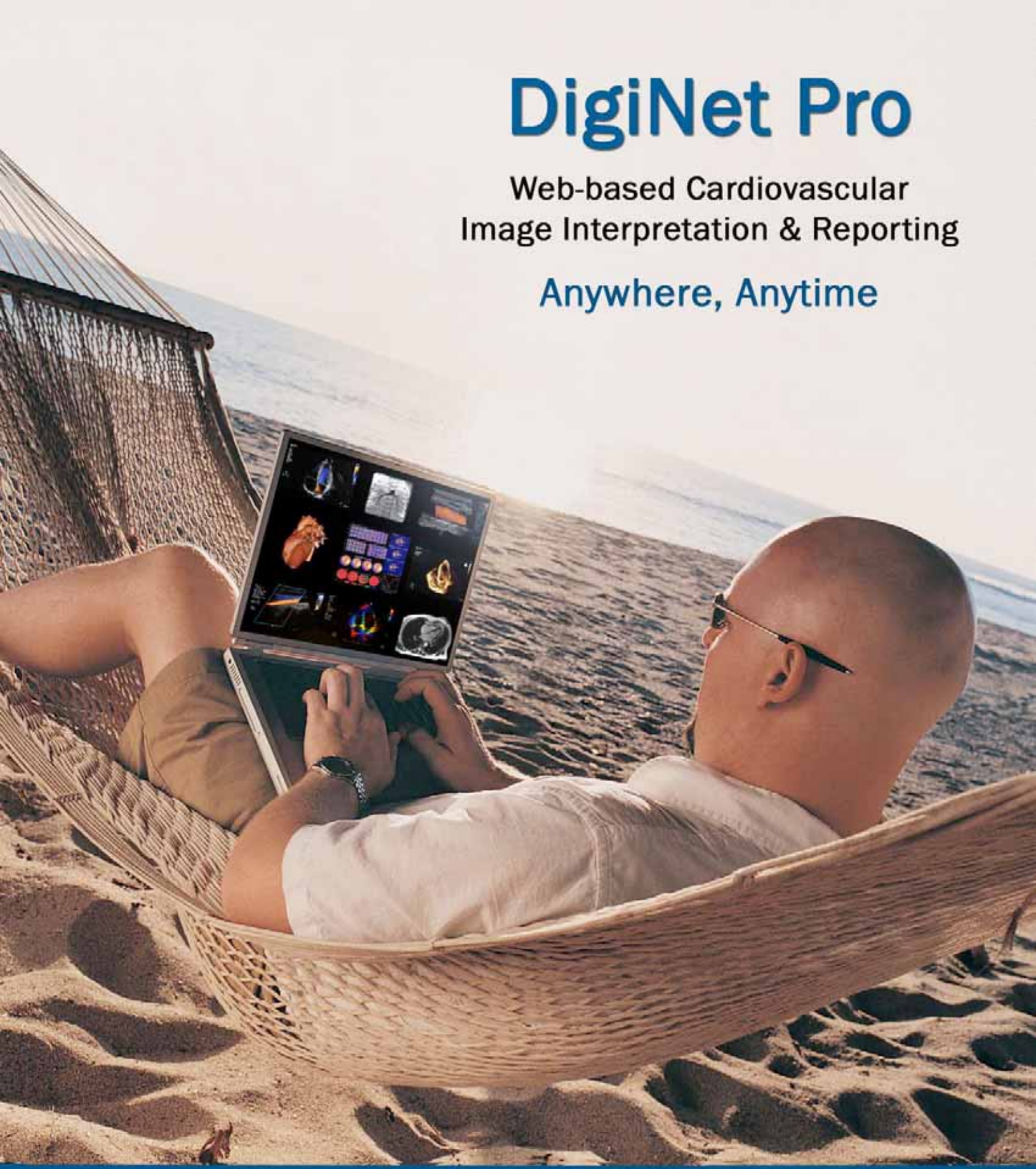

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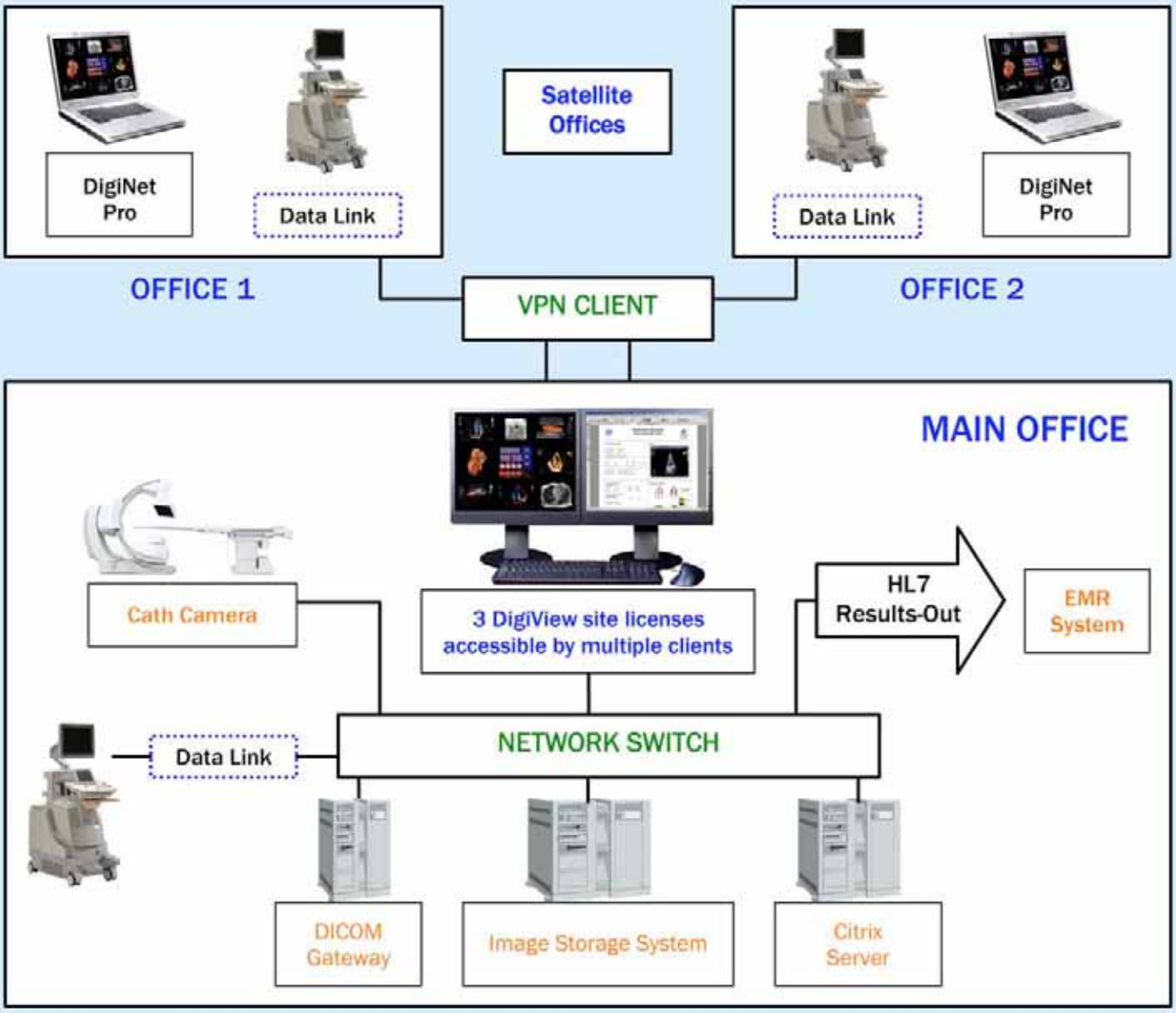

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