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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

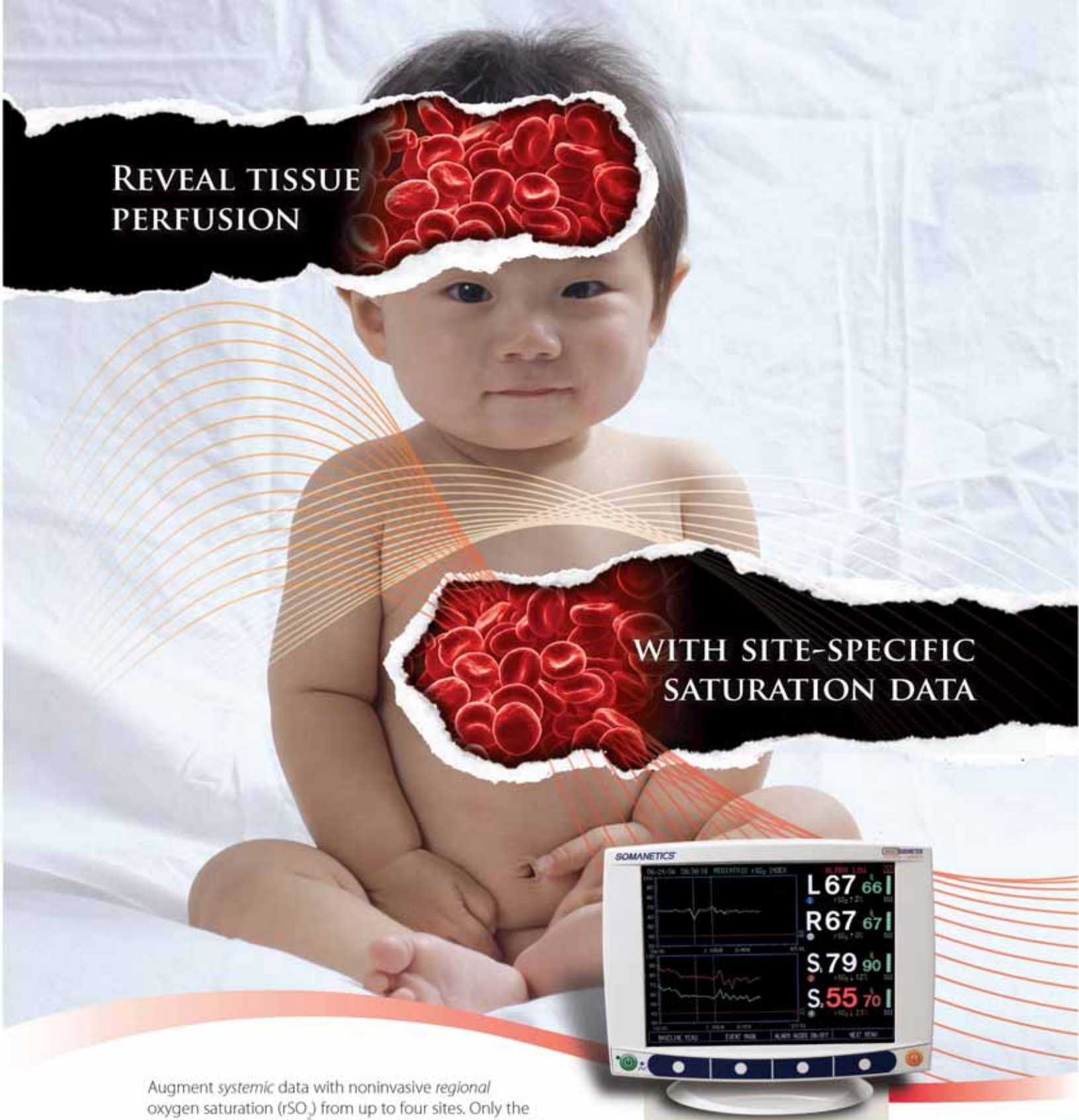
Background: Anomalies of the coronary arteries have been described as early as the 1800's. In 1933, Drs. Bland, White and Garland described the clinical syndrome of heart failure seen in infants with anomalous origin of the left coronary artery arising from the pulmonary artery. Mortality rates have 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

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"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.

In patients with ALCAPA chest roentegram may demonstrate cardiomegaly with or without pulmonary vascular congestion, al-

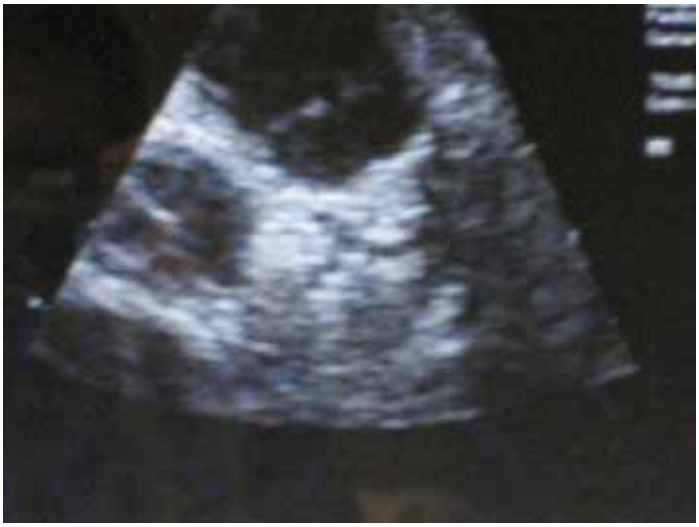

Image 1a: A trans-thoracic echocardiogram with the patient's left main coronary appearing to originate from the pulmonary artery.

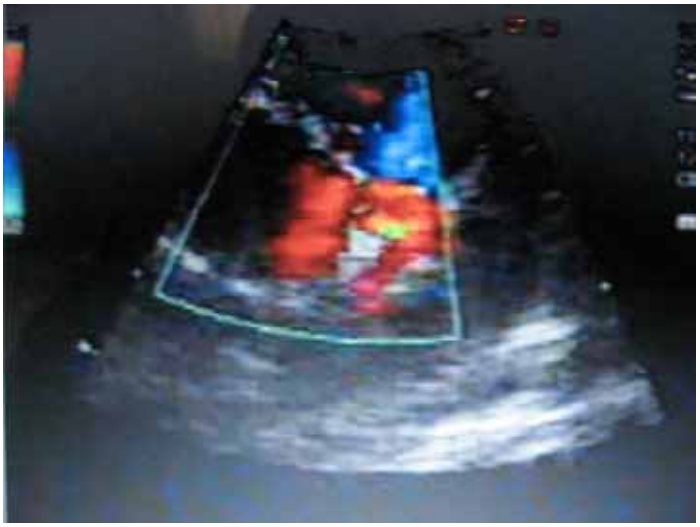

Image 1c: A trans-thoracic echocardiogram with retrograde flow (in red) through the patient's left coronary into the pulmonary artery.

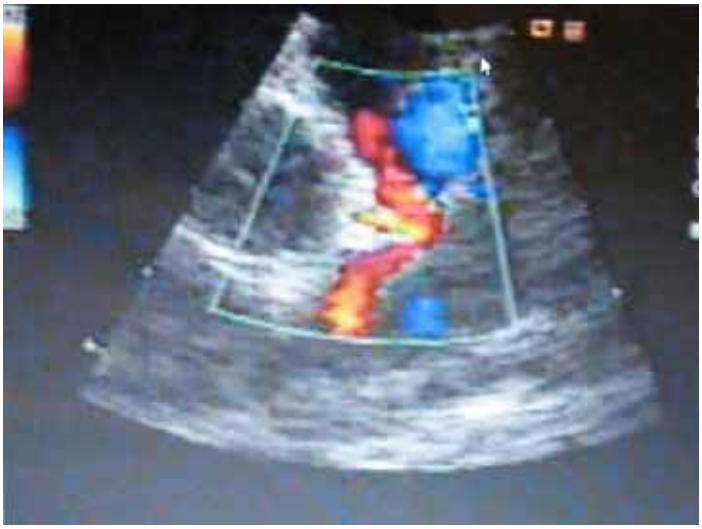

Image 1b: A trans-thoracic echocardiogram with apparent retrograde flow (in red) through the patient's left coronary.

though 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 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%

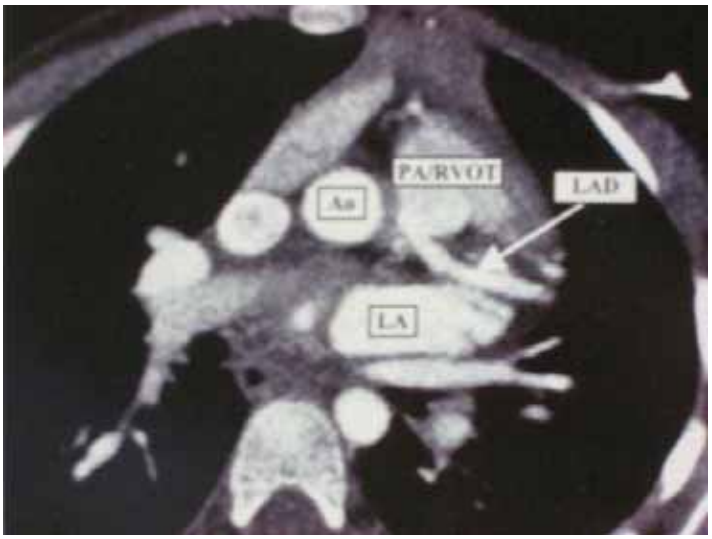

Image 2a: A cardiac CT angiography depicting patient's left main coronary anomalously originating from the pulmonary artery / right ventricular outflow tract.

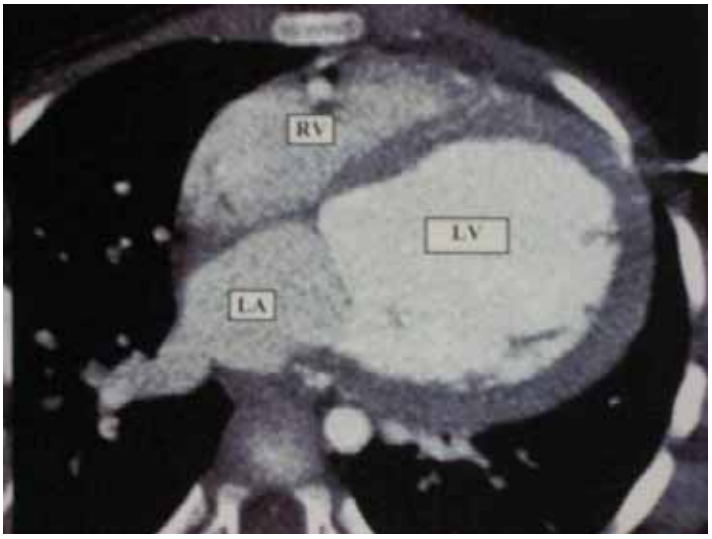

Image 2b: A cardiac CT angiography revealing our 2 year old patient's marked left ventricular dilatation.

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 persis-

tent 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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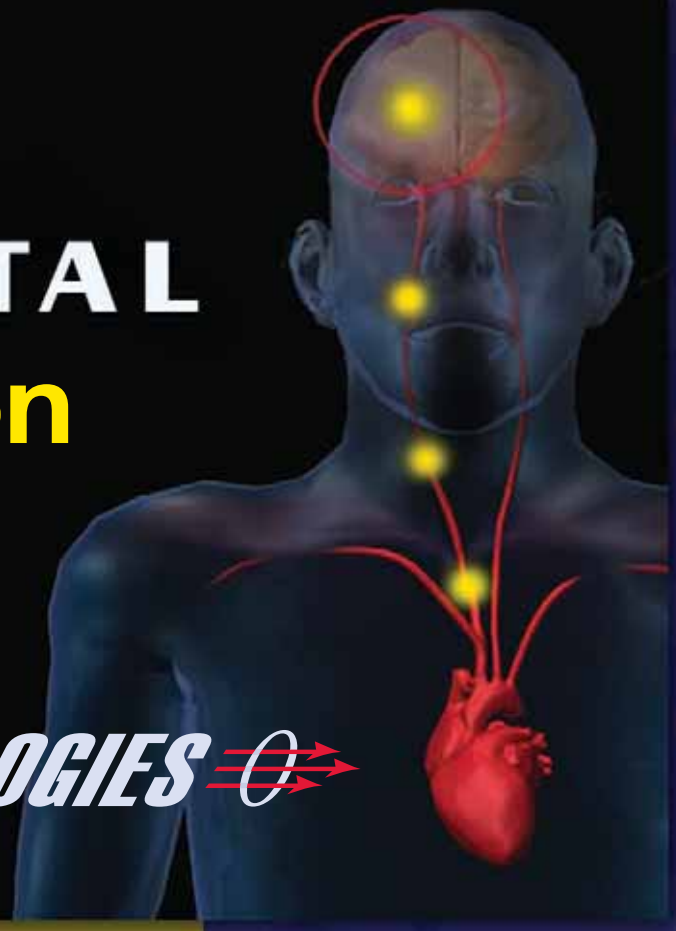

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Medical News, Products and Information

Dr. Ziyad M. Hijazi Named President of the Society for Cardiovascular Angiography and Interventions (SCAI)

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 30 Gold

Biosound Esaote, a global leader in ultrasound technology, introduced the *MyLab30 Gold 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 MyLab30Gold 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 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.

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 re-

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ceived 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.

Workshop will Help Formulate Northern Ireland's Future Policy Development for Sudden Cardiac Death

Northern Ireland's Chief Medical Officer Dr Michael McBride said that a workshop into Sudden Cardiac Death will help inform future policy development.

The workshop, which was held in early March 2009 in Lisburn, brought together leading health experts in this field along with representatives from relevant Government Departments, Public Health Bodies, Sports Council, and Voluntary Organisations.

Speaking after the workshop, Dr McBride said that Sudden Cardiac Death has had a devastating impact on the lives of those who had lost a loved one. He said, "In September, the Health Minister, Michael McGimpsey, gave his commitment to the Northern Ireland Assembly to look at a way forward with this complex issue. This workshop is one of a number of things that have been organised in order to fulfil that commitment."

"The key aim of the workshop was to consider if anything more could be done to reduce the number of deaths that occur in Northern Ireland, especially with regard to our young people."

"Every young death is a tragedy, but when that death occurs in an apparently fit and healthy young person, the devastation is much greater because it is unexpected and comes without any kind of warning. Behind all the statistics, there is a child who had dreams and ambitions, and a family left to grieve."

McBride went on to say, "As well as organising this workshop, the Minister also asked me to write to the UK National Screening Committee asking if they can prioritise their planned policy review on research evidence on screening for hypertrophic cardiomyopathy. The committee, which advises the four UK Health Departments on all aspects of screening, had previously advised that the current evidence does not support the introduction of a screening programme.

In addition, Dr McBride said a Service Framework for Cardiovascular Health and Wellbeing is due to be published for consultation in May 2008. At the Minister's request, he said he has written to the chair of the group, asking for sudden cardiac death to be addressed within the framework and asked that account is taken of the chapter covering arrhythmia and sudden cardiac deaths in the Coronary Heart Disease National Service Framework developed for England.

Dr McBride continued: "Today's conference was extremely useful and informative. As well as hearing from leading experts, we also heard from voluntary and community groups who work closely with families who have been affected by tragedy.

"Sudden Cardiac Death is a very complex issue. Raising awareness is very important. It is right that the public has the information it needs to properly understand this issue. However, there is a balance which must be achieved."

Key speakers at the workshop in the Lisburn Civic Centre included:
Dr Pascal McKeown, a consultant cardiologist at the Royal Victoria Hospital and would be heavily involved with families affected by sudden cardiac death in Northern Ireland; Dr Brian Maurer, a consultant cardiologist and chair of the Department of Health and Children's special taskforce on sudden cardiac death. The report of the taskforce "Reducing the Risk: A Strategic Approach" was published in March 2006; Dr Deidre Ward, Consultant Cardiologist in Dublin will speak on her work on the management of inherited cardiac diseases; Dr Sanjay Sharma, Consultant Cardiologist and Honorary Senior Lecturer, King's College Hospital and Cardiologist for Cardiac Risk in the Young (CRY) to discuss the case for screening; Dr Frank Casey,Consultant Paediatric Cardiologist at the Royal Victoria Hospital; and Stephanie Leckey, Heartstart UK to discuss Resuscitation Support.

SKMC Pediatric Cardiac Surgery Team has Performed 120 Cardiac Operations for Babies within Ten Months

In March 2009, The Division of Pediatric Cardiac Surgery at the Sheikh Khalifa Medical City (SKMC) in Abu Dhabi, managed by

Cleveland Clinic announced it had successfully performed 120 cardiac operations since the division opened in April 2007 to March 2008.

The postoperative pediatric cardiac services at SKMC are comprised of a 5-bed pediatric cardiac ICU, conjoined with a high dependency Unit (HDU) of 4 beds. Medical and nursing staff exploits the unit's full capacity. In addition to many emergent cases, patients scheduled for elective procedures are admitted for operations on a daily basis. The PCICU/HDU is run by its own dedicated medical and nursing staff on a 24/7 basis.

Dr. Norbert Augustin, Chair, Department of Cardiac Sciences, stated that "the Congenital Heart Surgical Program provides full cardiac services to children in the UAE. It helps us meet the needs of all those children affected in the Emirates rather than sending them abroad for treatment resulting in a reduction of stress for parents. It is an advanced pediatric cardiac care for the Emirate of Abu Dhabi and the UAE. Furthermore, SKMC is committed to continuous improvement of care to ultimately improve the health of the population of the UAE. "

He added, "the goal of the department is to reduce the medical requirement for overseas travel, provide comparable cardiac services to all UAE residents and provide family members an opportunity to participate in the healing process before and after surgery. Health statistics show that the family is a vital part of the healing process and SKMC is happy to provide this service within the community. The Department of Cardiac Sciences at SKMC demonstrates the commitment of SEHA and SKMC to provide medical care that is advanced and meets internationally recognized standards of excellence."

From the Pediatrics Cardiology side, Dr Johannes Du Plessis, Consultant and Head of Pediatrics Cardiology, said "the Pediatrics Cardiac Surgery and Pediatric Cardiology divisions create a complete Pediatric Cardiac Team that manages the care of Pediatric patients at SKMC. The close collaboration is very vital between the surgeons and other members of the cardiollogy team to give an integrated service to our little children who deserve only the best. It is also worth to mention that our Cardiologists are available 24/7 for patients and doctors requiring pediatric cardiology advice."

Pediatric Cardiology, which existed previously at SKMC, is led by Dr. Johannes du Plessis a senior pediatric consultant cardiologist from South Africa with extensive experience in the specialty. He is ably assisted by a US-trained consultant pediatric cardiologist, Dr Haitham Talo, who is very experienced in echocardiography including fetal echocardiography and Dr. Stephen Shipton, a pediatric cardiac consultant from South Africa, who is in charge of the pediatric cardiac catheter laboratory. Dr. Shipton has an extensive experience in interventional procedures for babies and children with cardiac disease obtained both in South Africa and the UK. The division of pediatric cardiovascular surgery and cardiology have at their disposal a complete complement of sub-speciality pediatricians as warranted.

Pediatric Cardiology provides the following services at SKMC

- The whole spectrum of diagnostic procedures: from physical examination by specialists, echocardiography, electrocardiography, cardiac catetherization and cardiac computer tomography. The patient is thorougly worked up from first outpatient or ED visit till a comprehensive and holistic overview is developed, which includes extra cardiac anomalies. This enables the surgeon to perform the correct operation with a high degree of accuracy. Intraoperatively the pediatric cardiologists offer a Transesophageal echo service to perform immediate quality control on the surgery performed
- State-of-the-art interventional procedures in the new pediatric cardiac catheter laboratory will include balloon dilatation or stenting of obstructions, closure of anomalous communications, holes and defects with special devices.
- A 24/7 consultation service for patients and doctors requiring pediatric cardiology advice
- Preoperative and postoperative follow-up of CHS patients.
- A number of hybrid procedures have been performed where the invasive cardiologist and the surgeon operate simultaneously in theatre or the catheterisation laboratory. This is really cutting edge pediatric cardiology.
- Postoperatively pediatric cardiology is involved in assisting the critical care unit and the surgeons to monitor the patients for arrhythmias, structural and functional abnormalities of the heart and assist with the management in general.

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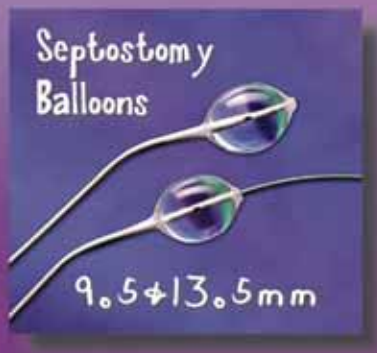

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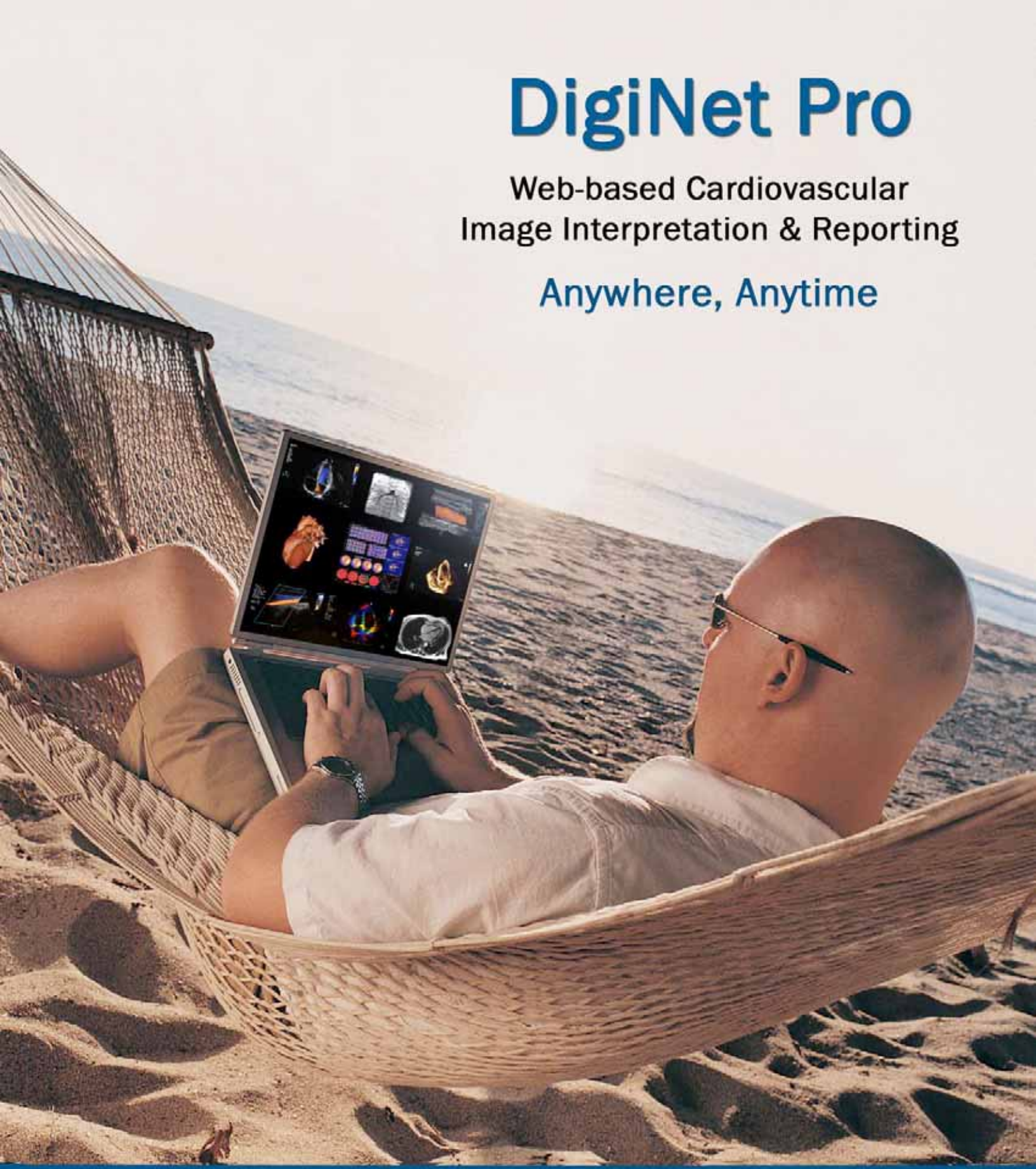

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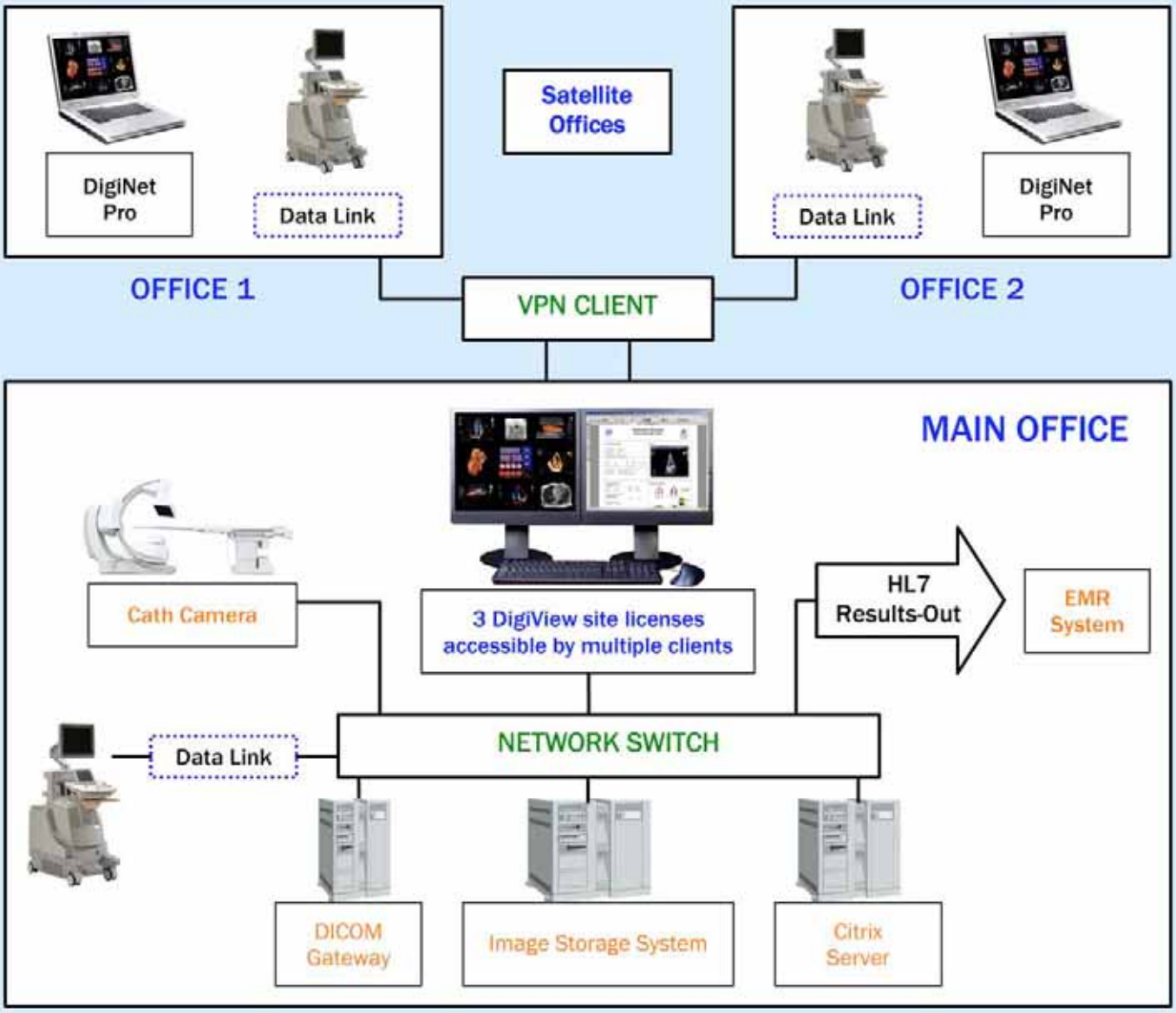

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