C O N G E N I T A L
C A R D I O L O G Y T O D A Y
Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

Volume 7 / Issue 12
December 2009
International Edition

IN THIS ISSUE

CONGENITAL CARDIOLOGY TODAY
Editorial and Subscription Offices
16 Cove Rd, Ste. 200
Westerly, RI 02891 USA
www.CongenitalCardiologyToday.com
Address Service Requested

UPCOMING MEDICAL MEETINGS

Evolving Concepts in the Management of Complex Congenital Heart Disease II
Jan. 14-16, 2010; San Diego, CA USA
www.rchsd.org/cme

Cardiology 2010
Feb. 10-14, 2010; Lake Buena Vista, FL USA
www.chop.edu/cardiology2010

8th Utah Conference on Congenital Cardiovascular Disease
Feb. 21-24, 2010; Snow Bird, UT USA
www.primarychildrens.com/pedscardiodisease

Masterclasess in Cardiac Morphology
Mar. 5-7, 2010; Charleston, SC USA
www.musckids.com/heart/conferences/

Image Gently and *Step Lightly* to Minimize Radiation Dose in Pediatric Congenital Imaging

By Kenneth A. Fetterly, PhD; James M. Kofler, PhD; Donald J. Hagler, MD

Introduction

The use of x-ray imaging modalities to diagnose and treat disease continues to increase in medical care in general, and the clinical specialty of congenital cardiology is no exception. X-ray imaging modalities, including computed tomography (CT) and x-ray fluoroscopy, nuclear medicine imaging, and chest radiography, have the potential to provide great benefit to the overall care provided to patients. However, the radiation dose associated with x-ray imaging also carries with it the potential for adverse effects. For young patients, the expected risk is that of a slight increase in the probability of developing cancer sometime later in life. For the relatively low radiation dose associated with medical imaging procedures, cancer risk is assumed to be directly proportional to the magnitude of the radiation dose received by a patient. The likelihood that a patient will develop a radiation-induced cancer later in life can be reduced by minimizing the patient's radiation dose.

Recently, several professional organizations recognized an opportunity to educate medical professionals and pediatric patient families regarding the benefits and potential risks of x-ray imaging procedures. The Alliance for Radiation Safety in Pediatric Imaging (The Alliance) was formed in 2006 and launched the *Image Gently* campaign in 2008 (www.imagegently.com).[1] The purpose of the *Image Gently* campaign is to share information regarding radiation dose for pediatric CT procedures and to assist providers with optimizing CT systems to minimize radiation dose. In 2009, The Alliance launched the *Step Lightly* campaign to educate and help minimize radiation dose associated with interventional radiology procedures. While there are some practical differences between general CT versus cardiac CT or interventional radiology versus interventional cardiology, there are enough similarities that both the *Image Gently* and *Step Lightly* campaigns are applicable to the specialty practice of pediatric congenital cardiology. This report will provide a general overview of radiation dose and radiation-induced cancer risks associated with cardiac CT and x-ray interventional procedures and will incorporate the foundations of radiation awareness and safety established by the *Image Gently* and *Step Lightly* campaigns. Suggestions for minimizing the radiation dose associated with pediatric cardiac CT and interventional fluoroscopy are included.

Radiation Dose and Patient Risk

The potential for biologic injury occurs when an x-ray photon(s) interacts with individual molecules in the nucleus of a cell.[2] The energy imparted by the x-ray photon(s) has the potential to break molecular bonds, potentially compromising critical sub-cellular structures, particularly DNA. Minor damage may be automatically repaired, but severe DNA damage may lead to cellular mutation,

Do you or your colleagues have interesting research results, observations, human interest stories, reports of meetings, etc. that you would like to share with the congenital cardiology community?

Submit a summary of your proposed article to Congenital Cardiology Today at:
RichardK@CCT.bz

Announcing Expanded Labeling

Improved Outcomes. Real-Time Data Accuracy.

Based on clinical evidence, INVOS® System labeling for site-specific blood oxygen monitoring has been expanded. This makes it the only commercially-available cerebral/somatic oximeter to be backed by the following claims.

- Improved outcomes after cardiac or major general surgery in patients > 2.5 kg
- On-label treatment of patients at any weight
  - o > 2.5 kg: real-time data accuracy
  - o < 2.5 kg: trend monitoring
- Noninvasive indication of O_2 changes in the cerebral and peripheral circulatory systems
  - o Often providing an early warning of O_2 deficits associated with impending shock and anaerobiosis

Labeling claims not applicable to other devices as data was derived using the INVOS System and its proprietary algorithm.

CEREBRAL/SOMATIC INVOS OXIMETER
REFLECTING THE COLOR OF LIFE®

"Recently, several professional organizations recognized an opportunity to educate medical professionals and pediatric patient families regarding the benefits and potential risks of x-ray imaging procedures."

potentially resulting in cancer. Because radiation-induced cancers have a latent period of at least several years, it is not possible to establish a direct link between a cancer and a previous exposure to radiation.

Radiation dose to a specific tissue or organ describes the amount of radiation energy absorbed per mass of tissue. It is given the special unit Gray (Gy), or more commonly, milliGray (mGy). Estimation of future radiation-induced cancer risk requires some knowledge or estimate of organ dose. Once known, individual organ dose values are multiplied by a numeric scaling factor representing the relative sensitivity of each organ. The weighted, organ-specific dose values are then summed over all organs to obtain an estimate of effective 'whole body' dose (E) with special unit Sievert (Sv).[3,4] Given that medical radiation is applied to only a portion of the body and therefore affects only a portion of the organs, the effective dose represents the whole body dose which would have resulted in the same overall cancer risk to the patient. This metric, therefore, allows comparison, in terms of risk for all types of medical radiation exposures, regardless of what part of the body was exposed. Note that most interventional x-ray systems report a cumulative entrance skin exposure (ESE, units gray (Gy)) value that is the product of the radiation exposure time (min) and rate (mGy/min) for the entire procedure. This value has some merit in that it can be used to avoid high skin dose levels for complex interventional procedures performed on adult patients. However, the ESE to pediatric patients is generally far below the threshold for skin injury and is not an effective surrogate for actual organ dose. Therefore, ESE is not useful for estimating risk to pediatric patients.

Generally, excess cancer risk from medical x-ray procedures is too small to be directly measured, even with large cohort patient studies. Notable exceptions include an increased risk of breast cancer in girls who were treated for scoliosis and children who were exposed to radiation in utero.[5] Most of the epidemiologic data from which cancer risk from medical procedures is estimated originates from the Japanese atomic bomb survivors from Hiroshima and Nagasaki. A comprehensive review of this data is included in the recent BEIR VII report, *Health Risks from Exposure to Low Levels of Ionizing Radiation (NRC 2006)*.[6] This report concluded that, for low radiation doses (<100 mSv), the risk of cancer from radiation is most reasonably modeled as being directly proportional to the dose. This linear, non-threshold dose response model forms the basis for discussions of relative risk and justification of efforts to minimize radiation dose.

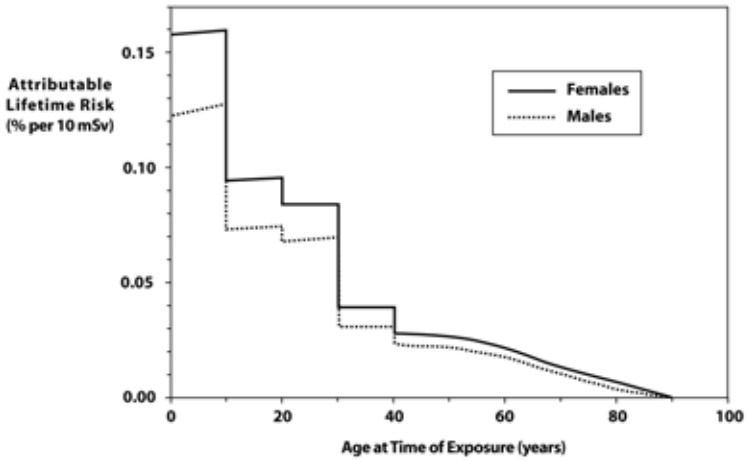


Figure 1. Adapted from ICRP Publication 60.

Figure 1, adapted from ICRP Publication 60, shows the expected attributable lifetime risk of cancer death from a single radiation dose of 10 mSv.[3] Including solid tumors and leukemia, the cancer risk to females is 35-40% higher for females than it is for males. This gender-specific discrepancy is due, in part, to an increased risk of ovarian and breast cancer. Also, young patients are at an increased risk of cancer compared to older patients. This is primarily because growth in childhood requires rapid cell division and because young patients are expected to live longer after exposure than are older patients, thereby providing more time for cancer to develop.[7] Both of these factors provide an increased chance for cellular mutation to develop into cancer. For our purposes, a radiation induced cancer risk of 0.15% per 10 mSv is a reasonable assumption for pediatric patients in the age range 0 to 10 years.[2] The risk decreases by a factor of about one-third for the age range 11 to 20 years. Note that, considering patients of all ages, risk is typically assumed to be 0.05% per 10 mSv.[2] As will be discussed below, patient-specific effective dose values are expected to be in the range of a few to a few tens of mSv for both cardiac CT and interventional x-ray procedures.

Cardiac CT and Dose

Computed tomography is an x-ray imaging system that can be used to acquire 3D images of patient anatomy. It has proven effectiveness in helping to diagnose structural heart disease and vascular anomalies in children.[8-10] Because CT uses x-rays to create images, it also delivers radiation dose, thereby presenting the possibility of cancer risk to patients. Effective dose estimates for pediatric patients undergoing CT of the heart and surrounding vessels have been reported in the literature. Published estimates vary substantially due to differences in patient size, CT technology, and scan protocol x-ray techniques used, including whether the acquisition and/or reconstruction is gated to the cardiac cycle via ECG monitoring. Hollingsworth et. al. reported phantom measurements and concluded that effective dose for ECG-gated CT angiography of 5 year-old patients was in the range 7.4 to 28.4 mSv.11 Herzog et. al. reported effective dose in the range 2.5 (± 2.1) to 6.3 (± 4.4) mSv for patients in the age range 1 day to 15 years. [12] It is important to note that there is not a universally accepted, clinically useful method to estimate individual patient dose and,

therefore, possible risk from CT. Because CT dose is highly dependent on equipment capabilities (or limitations) and site specific configuration, it is important that pediatric cardiologists work closely with radiologist and medical physicist colleagues to understand and minimize radiation dose for pediatric patients.

Interventional Fluoroscopy & Radiation Dose

Interventional x-ray systems use x-rays to produce real-time images of coronary and related vasculature to diagnose and treat disease. Similar to CT, there is not a standard method to estimate patient-specific effective dose from interventional x-ray in the clinical care setting. Also similar to CT dose estimates, published dose estimates for pediatric cardiac interventional procedures vary greatly and are, in part, influenced by x-ray system design and site-specific configuration. For interventional procedures, the complexity of each individual case can lead to substantial differences in radiation dose for a given patient. Rassow et. al. reported effective dose values in the range 2 to 18 mSv for infants undergoing cardiac catheterization procedures.[13] Bacher et. al. reported median effective dose values of 4.6 mSv (range 0.6 to 23.2 mSv) for diagnostic cardiac catheterization procedures and 6.0 mSv (range 1.0 to 37.0 mSv) for therapeutic procedures performed on patients in the age range 0.1 to 9.2 years .[14]

Image Gently for Cardiac CT

The Alliance for Radiation Safety in Pediatric Imaging launched the *Image Gently* campaign to raise awareness and to help practicing physicians to reduce radiation dose associated with pediatric computed tomography procedures. This campaign has been successful in helping to reduce radiation dose associated with pediatric CT in general, including cardiac CT. The *Image Gently* campaign encourages care providers to first consider whether the imaging task could be adequately accomplished without using ionizing radiation, such as with ultrasound or magnetic resonance imaging (MRI). Certainly, this is a very effective method to avoid radiation dose. If an alternative to CT is not appropriate or possible, then *Image Gently* suggests that "child-size" x-ray techniques be used for the CT scan. This ensures that the pediatric CT is performed with the lowest dose required to achieve adequate diagnostic images. Most importantly, *Image Gently* provides suggested protocols to reduce dose for pediatric patients of various age and size.[15]

Dose reduction for pediatric patients is most commonly accomplished by decreasing the current-time product (mAs) for the pediatric exams relative to a standard adult technique. *Image Gently* provides a table of mAs reduction factors based on the patient thickness in the posterior/anterior direction or based on approximate age. For example, suggested mAs reduction factors (RF) range from 0.42 for newborns to 0.73 for 15 year-old patients undergoing thoracic CT.[14] These RF values represent suggested initial starting points for dose reduction and specific scan protocols should be further optimized based on the imaging task (i.e., low contrast soft tissue visualization compared to high contrast bone imaging, etc). Thoughtful implementation of RF values for pediatric procedures helps to ensure that radiation dose is minimized while still maintaining clinically appropriate image quality for pediatric patients. Some CT scanners are able to achieve a similar effect automatically by using a technology called dose modulation, which is conceptually similar to automatic exposure control in radiography or automatic brightness control (ABC) in x-ray fluoroscopy. To understand dose modulation, one must first realize that a CT image is reconstructed from x-ray projection image data acquired from many different projection angles around the patient. If the x-ray technique is fixed for all angles, then projection images acquired for relatively thin anatomy (PA projection) would be of substantially higher quality than those acquired for relatively thick anatomy (lateral projection). The independent projection images are mathematically reconstructed to produce CT slice images. Ultimately, the quality of the resultant CT images is limited by the lowest quality angular projections. Therefore, the x-ray tube current (mA) used for the thin anatomy can be lowered (and dose decreased) without substantially affecting image quality. Hence, the goal of dose modulation is to equalize the quality of each projection, thereby maximizing the contribution of each x-ray photon to image quality. Proper modulation of the tube current is determined by the patient attenuation data obtained from the 2D scout view image that is acquired prior to acquisition of the CT projection images. This same scheme applies not only to projections through a single patient, but can also apply to patients of different sizes, with smaller patients automatically receiving less radiation than larger-sized patients. Compared to a fixed tube current, tube current modulation can result in radiation dose reduction without noticeable loss of image quality.[7] It may also be appropriate, with some guidance from the manufacturer or a physicist, to reduce the x-ray energies (kVp) used for a given CT scan protocol. Lower kVp settings provide improved contrast between organ boundaries or between tissues and contrast agents, and the improved contrast may allow for a subsequent reduction in x-ray tube current and patient dose.[16] However, the lower energy x-rays produced using lower kVp settings do not have the same penetrating capabilities as higher energy x-rays and therefore may not be appropriate for imaging larger children or adults. Additionally, the x-ray production changes non-linearly when the kVp setting is changed, therefore the appropriate mAs value will need to be determined for each kVp setting to be used. Another important consideration is to assure that the CT scanner has been properly calibrated at all kVps that are to be used clinically.

Other radiation-conscious practices include reducing the scan field of view to cover the minimum possible patient volume and minimizing the number of unique CT scan passes (contrast vs. non-contrast, for example) performed on patients. Every healthcare facility that performs CT on pediatric patients should have body part and patient size-specific protocols to help minimize the radiation dose inherent to these procedures. It is important to note that CT continues to realize rapid advances in technology that affects both image quality and patient dose. State-of-the-art CT systems with wide x-ray detector arrays and exceptionally fast rotation speeds are especially useful for cardiac imaging due to the short acquisition times and the large coverage per rotation. One such technology uses two x-ray tubes and a very high pitch helical, or spiral, acquisition to scan the entire heart within a single cycle (assuming the heart rate is not too high) at a very low dose. This is also unique in that axial, or sequential, acquisitions are typically considered superior with respect to both image quality and dose for cardiac imaging. The pediatric cardiologist should be familiar with the technical capabilities (and limitations) of their CT equipment and

Table 1. System configuration and user specific technical parameters that can be adjusted to improve radiation dose efficiency of an interventional fluoroscopy system

	System Configuration	User Specific
Remove the anti-scatter grid		for small patients
Include Cu x-ray spectral filter	for all pediatric fluoro and cine imaging	
Lower the image receptor dose	for all pediatric fluoro and cine imaging	use low dose rate fluoroscopy
Collimate the primary beam		specific to the patient anatomy
Decrease the frame rate	set default to 15 fps	as low as clinically acceptable
Optimize x-ray geometry		for each projection

approximate radiation dose values associated with the cardiac scan protocols used at their site. The pediatric cardiologist should also consult with a medical physicist and the scanner manufacturer to ensure that appropriate steps have been taken to optimize the radiation dose to the imaging task.

Step Lightly for Interventional Cardiology

The Alliance for Radiation Safety in Pediatric Imaging launched the *Step Lightly* campaign to elevate radiation dose awareness and help practicing physicians reduce radiation dose associated with interventional radiology procedures. Given the similarities between the interventional radiology and cardiology practices, many of the concepts and principles suggested by *Step Lightly* are equally applicable to interventional cardiology. Generally, *Step Lightly* encourages practitioners to ensure: that non-radiation imaging alternatives are considered and that the x-ray procedure is appropriate, that the patient dose is monitored and recorded in the patient record, that the x-ray fluoroscopic system is properly configured to ensure clinically useful images using the least possible radiation dose, that all staff receive regular radiation safety training and use sound radiation safety practices for every procedure, and that patient care providers are prepared to speak to families about risks and benefits of radiation used in imaging.

There are several x-ray technical factors that contribute to dose-efficient use of the x-ray system. First, it must be ensured that the x-ray system is in good working condition and that it is set up to specifications required by state and federal regulations. It will likely be necessary to customize an interventional x-ray system to optimize it for pediatric imaging. Such customization should be managed by a qualified medical physicist working in conjunction with physicians and equipment manufacturer image quality experts. When changes are made, measurements should be acquired to determine the effects on both patient dose and image quality. Interventional x-ray systems require a fixed x-ray exposure to the image receptor (image intensifier or digital flat panel). The x-ray fluence from the x-ray tube is automatically adjusted by the automatic brightness control system to account for variations in x-ray absorption by the patient (or transmission through the patient). The ABC automatically adjusts both the x-ray beam energy (via kVp) and the x-ray tube current-time product (mAs) to ensure that the image receptor realizes the pre-set detector dose rate. As patient thickness increases, both the beam energy and current-time product are increased to compensate for increased x-ray attenuation by the patient. The opposite is true for small patients through whom a greater fraction of the incident x-ray photons transmit. As patient thickness decreases, the x-ray tube output and therefore patient skin dose rate is automatically lowered by the imaging system. In this manner, the ABC system automatically reduces the radiation dose rate delivered to small patients. It is of great importance to recognize the radiation safety implications of x-ray fluoroscopy versus cine acquisition mode imaging. X-ray fluoroscopy utilizes a relatively low dose rate x-ray beam to create low-quality images that are appropriate for verifying anatomic landmarks or navigating intravascular catheters. Fluoroscopy images are not automatically saved, however, many systems provide opportunity for users to manually store such images. Cine or acquisition mode imaging utilizes a relatively high dose rate that is typically 10x that of fluoroscopy and produces high-quality images that are appropriate for diagnostic purposes. For both fluoroscopy and cine mode imaging, there are several technical parameters that can and should be properly set to minimize dose to pediatric patients. These factors are listed in Table 1. Some parameters can be adjusted by individual users and some of them are controlled by the system configuration.

The purpose of the anti-scatter grid is to preferentially absorb x-ray photons that interact with patient tissue and are subsequently scattered in direction of the x-ray detector while transmitting primary (unscattered) x-ray photons. This reduction in the number of scattered x-rays reaching the x-ray receptor results in improved x-ray image contrast. However, the grid also reduces the overall photon fluence at the image receptor, requiring compensation by the ABC via increased x-ray tube output and higher radiation dose rate. For small patients, there is very little x-ray scatter and so the grid can be removed from the system without substantial loss of image quality. Our experience is that for small patients (<20 kg), removing the grid reduces patient radiation dose rate by about 35%. Dose rate savings for larger patients will be somewhat greater, and it may be reasonable to remove the grid for patients as large at 50 kg provided that the image quality is adequate. Use of the antiscatter grid may be necessary for larger patients or when additional image quality is required to properly view instruments or anatomy. Modern x-ray fluoroscopy systems allow for the use of a copper x-ray beam spectral filter for both fluoroscopic and cine mode imaging. The Cu filter preferentially removes low energy x-ray photons from the x-ray beam, thereby increasing the average x-ray energy of the beam. Because low energy x-ray photons preferentially contribute to tissue dose relative to image quality, removing the low energy photons from the beam serves to substantially reduce radiation dose. For pediatric imaging, ensure that the x-ray control configuration is set to include a copper x-ray beam spectral filter for cine mode imaging. Our experience is that introduction of a 0.1 mm Cu filter reduces x-ray dose rate by 40% while decreasing image contrast by 10%. Another x-ray system parameter that can be changed to decrease patient dose is the target image receptor dose. Most x-ray systems allow customization of this parameter in 15% or 20% increments. Decreasing the image receptor dose will result in a decrease in patient dose rate. However, it will also result in decreased image quality. For both fluoroscopy and cine acquisition modes, the image receptor dose should be set as low as possible while still maintaining clinically useful image quality. For a typical interventional fluoroscopy system, the radiation dose rate (mGy/min) increases when the x-ray magnification mode is increased (primary field of view decreased). Some modern systems provide for configuration of the change in x-ray dose rate for the various geometric magnification (field of view) modes. The relationship between dose rate and geometric magnification should be understood by the physician operator. Regardless of the field of view selected, the secondary x-ray field collimator blades should be used to confine the x-ray beam to the anatomy of direct relevance, thereby minimizing the patient surface area and tissue volume irradiated. The image acquisition frame rate can be customized to reduce patient dose. For interventional cardiac procedures, the frame rate default is typically set to 15 frames per second. For patients with a rapid heart rate, the frame rate may have to be increased to 30 fps to achieve appropriate temporal resolution. For other patients and procedures (or portions of a procedure) a lower frame rate of 10 fps or 7.5 fps, with proportionately lower x-ray dose rate, may be appropriate. Finally,

"Imaging systems which use x-rays are an integral part of the modern medical practice and will remain so for the foreseeable future. Both the potential benefit and risk associated with x-ray imaging need to be well understood to make sound decisions that ensure both short-term and long-term patient health."

the geometry of the x-ray system with respect to the patient should be optimized to reduce patient dose. The x-ray dose rate at any point in space decreases at a rate that is inversely proportional to the square of the distance from the x-ray source. Therefore, the relative distance of both the patient and the x-ray detector from the x-ray tube can substantially affect patient radiation dose rate. Optimization of x-ray geometry to minimize patient dose requires only that the patient be moved as far away from the x-ray source as reasonably possible and that the x-ray detector is then moved as close as reasonably possible to the patient. Due to the strong influence of distance from the x-ray source on dose rate, seemingly modest changes in these distances can have a substantial impact on patient dose rate.

It should be recognized that, after the technical parameters controlling the fluoroscopic system are set, the performing physician can have a great deal of influence on the amount of radiation used during an interventional fluoroscopy procedure. To help minimize radiation dose, each procedure should start with a plan that is discussed with all members of the patient care team. The plan should include relevant details of anticipated x-ray system use. When available, previous imaging procedures should be reviewed. Reducing the duration of time that the x-ray beam is on is a fundamental ideal in the *Step Lightly* campaign. To this end, ensure that every second of beam on time provides clinically useful information. Though seemingly obvious, never activate the fluoroscopy foot switch if your eyes are not on the image display monitor. Also, step off of the radiation footswitch as soon as the relevant clinical information is acquired. As much as possible, navigate catheters using low dose rate fluoroscopy.

Managing Radiation Dose

X-ray based images, including CT, chest radiography, and x-ray fluoroscopy can be valuable tools for diagnosing, monitoring, and treating pediatric patients with congenital heart and/or vascular defects. However, with the possibility of multiple such procedures contributing to a lifetime cumulative radiation dose, the risk of these patients developing a radiation-induced cancer later in life should always be considered. As medical professionals, we should strive to ensure that x-ray modalities are used appropriately. First, consider whether an x-ray procedure is necessary or whether the information which it will provide could be obtained by another means. Consider the use of ultrasound or MRI, for example. Consider the frequency at which routine x-ray procedures, such as chest radiographs, are acquired and reduce the frequency to the minimum which is clinically appropriate. Once the need for an x-ray based procedure is confirmed, ensure that the radiation dose associated with that

EVOLVING CONCEPTS IN THE MANAGEMENT OF COMPLEX CONGENITAL HEART DISEASE II

Sponsored by

Rady Children's Hospital San Diego

In cooperation with

University of California San Diego School of Medicine

Thursday, Friday and **Saturday**

January 14, 15 and **16, 2010**

Hyatt Regency Mission Bay Spa and **Marina**
San Diego, California

COURSE COORDINATORS

John J. Lamberti, MD and
John W. Moore, MD, MPH

Rady Children's Heart Institute has organized and supported symposia dedicated to expanding our knowledge about the diagnosis and treatment of complex forms of congenital heart disease since 1984. The symposia are designed to educate attendees by combining formal presentations with lively discussions of controversial areas in the rapidly evolving science and clinical management of the neonate, child and adult born with congenital cardiovascular disease. For each program, the planners carefully assess prior attendees' comments and reviews,and invite renowned faculty from across the United States to present new data and ideas.

The three-day symposium on Pediatric and Congenital Cardiology and Cardiac Surgery will include lectures, roundtables and case discussions. We will focus on the latest techniques, including cutting-edge technologies such as the hybrid procedure and percutaneous heart valves. We expect to attract an enthusiastic audience of pediatric cardiologists, pediatric cardiology fellows, cardiac surgeons, adult cardiologists with interest in congenital heart disease and cardiac nurse practitioners, as well as some cardiac administrators.

We have pondered what quality measures are appropriate in 2010. We can assess the quality gap by observing adult patient outcomes and comparing those outcomes to the surgical procedures performed earlier in life. The title for this symposium emphasizes the ever-changing diagnostic and treatment modalities in the management of congenital heart defects. The analyses presented will benefit attendees in their medical and surgical practices by outlining the procedures most likely to produce optimal long-term patient outcomes. The planners hope that the Symposium will provide answers to the perennial question: ***"What is the right thing to do for this patient?"*** *Faculty panel and question-and-answer sessions will allow audience interaction as we jointly pursue the ideal treatment plan for each complex patient.*

We hope you will come warm yourself in beautiful San Diego in January!

Visit our website: **www.rchsd.org/cme** and click on Conferences & Seminars for more information or for registration and credit card payment form. We accept Visa and Mastercard

Planning Committee: John J. Lamberti, MD; John W.Moore, MD, MPH; R.R. Skoglund, MD, Immediate Past CME Director; Keith Vaux, MD, CME Director

Educational Design: Didactic presentations, with audio-visual support, are followed by panel discussion and audience question and answer sessions. A syllabus will include abstracts from faculty on their topics, with key references.

Objectives: At the completion of this activity, participants should be able to:

- Determine optimal management strategies for common diagnostic and treatment problems encountered in an outpatient office practice.
- Understand and manage the diagnosis and treatment of cardiomyopathy and pulmonary hypertension in infants, children and young adults.
- Optimize imaging modality selection to obtain a diagnosis in the most efficient and cost-effective manner.
- Diagnose and treat common and complex arrhythmias in patients of all ages.
- Utilize modern catheterization techniques as part of an integrated invasive treatment program for congenital heart disease.
- Integrate the Hybrid approach with modern variations of the Norwood and Fontan procedure to optimize care of infants born with HLHS.
- Utilize modern medical and surgical strategies to treat complex Congenital Heart Disease.
- Understand the role of “benchmark” data in assessing outcomes after treatment of pediatric and adult CongenitalHeart Disease.

Faculty: Zahid Amin, MD; Anjan S. Batra, MD; Daniel Bernstein, MD; ; Jane C. Burns, MD; John P. Cheatham, MD, FAAP, FACC, FSCAI; John S. Child, MD; Joseph A. Dearani, MD; Howaida G. EL-Said, MD; Frank L. Hanley, MD; Dunbar Ivy, MD; Jeffrey P. Jacobs, MD, FACS, FACC,FCCP; Joel Kirsh, MD; Steven E. Lipshultz, MD, FAHA, FAAP; James Lock, MD; Audrey C. Marshall, MD; Gerald Ross Marx, MD; Peter Pastuszko, MD; James C. Perry, MD, FAAP, FACC, FHRS; Beth Feller Printz, MD, PhD; Mohan Reddy, MD; Kevin Shannon, MD; Thomas L. Spray, MD; Vaughn A. Starnes, MD; Lloyd Y. Tani, MD; James S. Tweddell, MD; George F. Van Hare, MD; Victoria L. Vetter, MD; Gary Webb, MD; Gil Wernovsky, MD, FACC, FAAP

procedure is minimized. This requires some planning and work well before the patient reaches the procedure room. For both CT and fluoroscopic imaging, minimizing radiation dose includes using x-ray techniques that are appropriate for the patient size, minimizing the volume of tissue irradiated, and minimizing the duration of time that x-ray beam is on. For every patient, a comprehensive record of all x-ray based procedures should be maintained. This information should be integrated into the patient record and be immediately accessible to all care providers. A clear and concise radiation history in the patient record will help raise the radiation awareness of everyone involved in patient care. The Alliance website provides suggestions for discussing x-ray radiation and related issues with families, and it can be expected that the x-ray procedure history may be of interest to family members. When discussing radiation risks with families, it is important that the risks and benefits of performing versus not performing a procedure which uses ionizing radiation are well appreciated.

Conclusion

Imaging systems which use x-rays are an integral part of the modern medical practice and will remain so for the foreseeable future. Both the potential benefit and risk associated with x-ray imaging need to be well understood to make sound decisions that ensure both short-term and long-term patient health. Based on the linear non-threshold cancer response model and considering that pediatric congenital cardiac patients may undergo several CT or interventional fluoroscopy procedures during their childhood, these patients are expected to have a slightly increased lifetime risk of cancer. Minimizing the radiation dose for each procedure helps to minimize this risk. Through the *Image Gently* and *Step Lightly* campaigns, The Alliance for Radiation Safety in Pediatric Imaging continues to have a positive influence on the use of ionizing radiation in the care of pediatric patients. As promoted by these campaigns, pediatric radiation dose reduction can be achieved through decreased use of x-ray imaging procedures and through dose-efficient use of x-rays when such a procedure is indicated. Most importantly, these reductions can be realized without compromising image quality or patient care.

References

1. Sidhu, M., B. D. Coley, M. J, Goske, B. Connolly, J. Racadio, T. T. Yoshizumi, T. Utley, and K. J. Strauss, Image Gently, Step Lightly: increasing radiation dose awareness in pediatric interventional radiology, Pediatr. Rad. 2009; 39:1135-1138.
2. Hall, E. J., Radiation biology for pediatric radiologists. Pediatr. Rad. 2009; 39:S57-S64.
3. ICRP Publication 60, 1990 Recommendations of the International Commission on Radiologic Protection, Ann. ICRP. 1990; 21.
4. ICRP Publication 103, The 2007 Recommendations of the International Commission on Radiologic Protection, Ann. ICRP. 2008; 37.
5. Linet, M. S., K. Kim, P. Rajaraman, Children's exposure to diagnostic medical radiation and cancer risk: epidemiologic and dosimetric considerations. Pediatr. Rad. 2009; 39:S4-S26.
6. National Research Council, 'Health risks from exposure to low levels of ionizing radiation: BEIR VII - Phase 2,' 2006, Washington, DC.
7. Rice, H. E., D. P. Frush, D. Farmer, J. H. Waldhausen, APSA Education Committee, Review of radiation risks from computed tomography: essentials for the pediatric surgeon. J. Ped. Surg. 2007; 42:603-607.
8. Goo, H. W., D. S. Suh, Tube current reduction in pediatric non-ECG-gated heart CT by combined tube current modulation. Pediatr. Rad. 2006; 36:344-351.
9. Greil, G. F., M. Schoebinger, A. Kuettner, J. F. Schaefer, F. Dammann, C. D. Claussen, M. Hofbeck, H. P. Meinzer, and L. Sieverding, Imaging of aortopulmonary collateral arteries with high-resolution multidetector CT. Pediatr. Rad. 2006; 36:502-509.
10. Lerner, C. B., D. P. Frush, D. T. Boll, Evaluation of a coronary-cameral fistula: benefits of coronary dual-source MDCT angiography in children. Pediatr. Rad. 2008; 38:874-878.
11. Hollingsworth, C. L., T. T. Yoshizumi, D. P. Frush, F. P. Chan, G. Toncheva, G. Nguyen, C. R. Lowry, and L. M. Hurwitz, Pediatric cardiac-gated CT angiography: Assessment of radiation dose. Am. J. Roent. 2007; 189:12-18.
12. Herzog, C., D. M. Mulvihill, S. A. Nguyen, G. Savino, B. Schmidt, P. Costello, T. J. Vogl, and U. J. Schoepf, Pediatric cardiovascular CT angiography: radiation dose reduction using automatic tube current modulation. Am. J. Roent. 2008 190:1232-1240.
13. Rassow, J., A. A. Schmaltz, F. Hentrich, and C. Streffer, Effective doses to patients from paediatric cardiac catheterization. Brit. J. Rad. 2000; 73:172-183.
14. Bacher, K., E. Bogaert, R. Lapere, D. De Wolf, H. Thierens, Patient-specific dose and radiation risk estimation in pediatric cardiac catheterization. Circulation 2005; 111:83-89.
15. 15. The Alliance for Radiation Safety in Pediatric Imaging. 2009; Retrieved October 15, 2009, from American College of Radiology web site http://www.pedrad.org/associations/5364/files/Protocols.pdf
16. Kalender, W. A., P. Deak, M. Kellermeier, M. van Straten, and S. B. Vollmar, Application- and patient size-specific optimization of x-ray spectra for CT. Med. Phys. 2009; 36:993-1007.

CCT

Corresponding Author

Kenneth A. Fetterly, PhD
Assistant Professor of Radiology
Division of Cardiovascular Diseases
College of Medicine
Mayo Clinic
200 1st St. SW
Rochester, MN 55905 USA
Phone: 507-266-4384; fax 507-255-7688

fetterly.kenneth@mayo.edu

James M. Kofler, PhD
Department of Radiology
Mayo Clinic
200 1st St. SW
Rochester, MN USA

Donald J. Hagler, MD
Division of Pediatric Cardiology
Mayo Clinic
200 1st St. SW
Rochester, MN USA

Case Report: Fatal Thrombo-embolism in a Child with Noncompaction of Ventricular Myocardium

By Sulafa KM Ali, MD, FACC, FRCPCH

Introduction

Noncompaction of the ventricular myocardium (NCVM) is a cardiomyopathy that is characterized by prominent myocardial trabeculations and deep intertrabecular recesses with variable degrees of myocardial dysfunction. It has variable clinical presentations and in children had been associated with benign asymptomatic course, as well as severe myocardial dysfunction.[1]

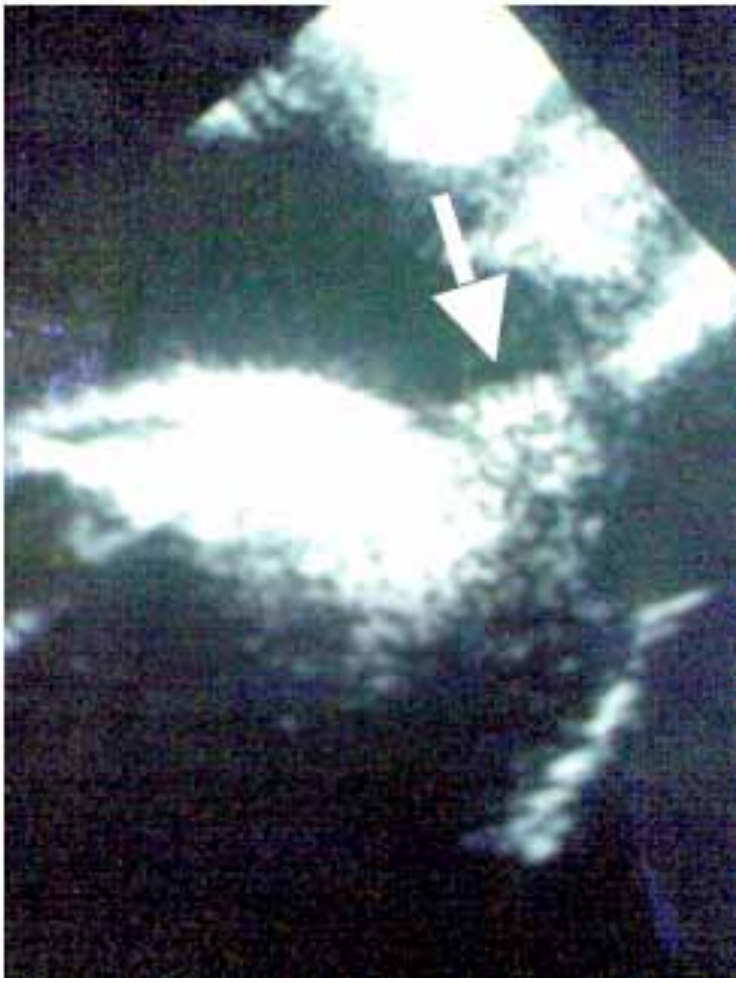


Figure 1. Suprasternal long axis view of the aortic arch showing a thrombus (arrow) extending between the left common carotid and the exit of the left subclavian artery.

In adults, NCVM is well known to be associated with thrombo-embolism.[2] In the pediatric age group, we observed thrombo-embolism with noncompaction in 9% of patients with the isolated form.[3] In this report we describe a further case of fatal thrombo-embolism in a child with NCVM.

Case History

A Sudanese boy 12 years of age presented two weeks shortness of breath. Physical examination revealed a severely distressed child with a respiratory rate of 40 cycles/minute, heart rate of 120 beats/minute. Chest examination showed fine bilateral crackles. Cardiovascular system examination revealed that the apex was displaced to the 6th intercostal space lateral to the mid-clavicular line. Normal first and second heart sounds were heard with a third heart sound. No murmurs were audible. The liver was enlarged 3 cm below the right costal margin. On presentation all pulses were palpable. The patient was seen in the echocardiography (echo) clinic and a limited echo showed normal heart anatomy, dilated left ventricle (diastolic dimension of 5 cm) with poor contractility, ejection fraction was 25%. The patient was admitted and started on intravenous furosemide and oral spironolactone. A few hours later blood pressure was found to be 145/90. Re-examination of the cardiovascular system revealed absent femoral and left brachial pulses. The patient was started on carvedolol with suspicion of 'missed' coarctation of aorta. A second detailed echo was done and revealed a large thrombus (2X3 cm) in the aortic arch opposite to the left subclavian artery (Figure 1). There was

"This case demonstrates a fatal course due to aortic obstruction and probably hypertensive or hemorrhagic brain injury. Transcatheter thrombectomy would have been a reasonable option in this patient because of the high risk of open heart surgery. The routine use of anticoagulation in patients with NCVM without cardiac thrombi is controversial, but in the presence of severely reduced ejection fraction this treatment should be considered."

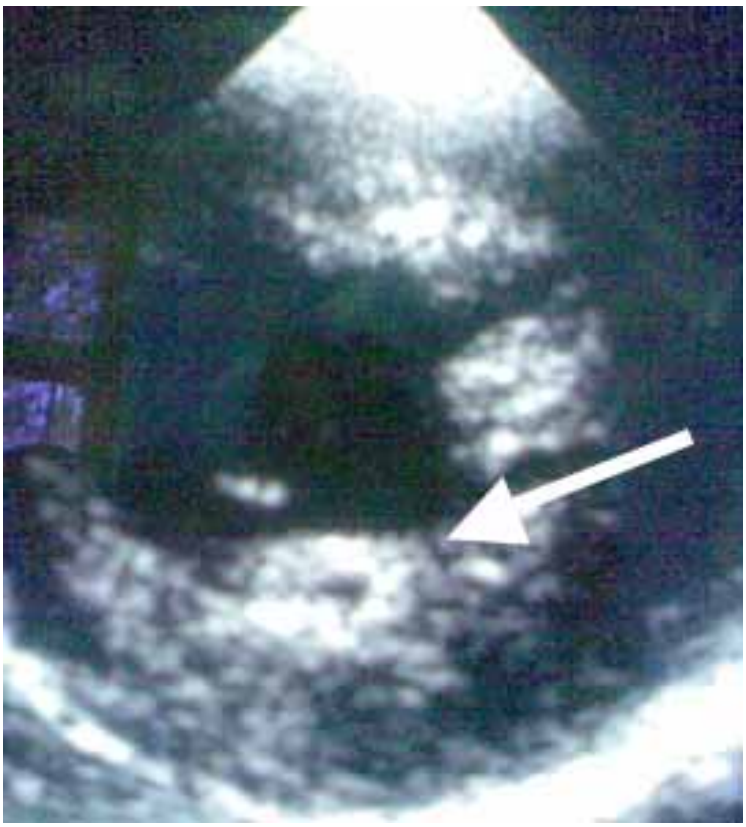


Figure 2. Short axis view showing trabeculated, 2- layer appearance of left ventricle myocardium with an echogenic mass (arrow).

scanty flow distal to the thrombus and in the abdominal aorta, no gradient could be measured. The left ventricle was noted to be noncompacted, the noncompacted: compacted layer thickness ratio was 2:1. A well-demarcated mass which looks like a thrombus was noted in the left ventricle trabeculations distal to the papillary muscles. (Figure 2). The ejection fraction was noted to be better than the previous study (45%). The blood pressure increased over two days to 200/100. Consultation with the cardiac surgeon was done and the joint opinion was to start systemic thrombolytic therapy initially in an attempt to dissolve the thrombus and avoid the high risk surgery. The patient was managed with streptokinase and intravenous hydralazine he developed sudden deterioration of consciousness and arrested suddenly.

Discussion

Thrombo-embolism is rarely reported in children with NCVM. In our previous cases one patient developed a cerebrovascular accident leading to reversible hemiparesis.[3] Embolic cerebrovascular events had also been reported in adults with NCVM.[4] Left ventricle clots are probably related to stagnant blood flow in the intertrabecular recesses in the presence of myocardial dysfunction. Although some authors reported a frequency as high as 25%[2] others regarded thromboembolism a rare event in NCVM.[5] This case demonstrates a fatal course due to aortic obstruction and probably hypertensive or hemorrhagic brain injury. Transcatheter thrombectomy would have been a reasonable option in this patient because of the high risk of open heart surgery. The routine use of anticoagulation in patients with NCVM without cardiac thrombi is controversial, but in the presence of severely reduced ejection fraction this treatment should be considered. We believe that thrombo-embolism is an important and life-threatening complication of NCVM that should be promptly diagnosed and managed.

References

1. Sulafa K. M. Ali, Michael J. Godman. The variable clinical presentation of, and outcome for, noncompaction of the ventricular myocardium in infants and children, an under-diagnosed cardiomyopathy. Cardiol Young 2004; 14: 409–416.
2. Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. J Am Coll Cardiol. 2000;36:493-500.
3. Sulafa KM Ali. Unique features of non-compaction of the ventricular myocardium in Arab and African patients. Cardiovasc J Afr 2008; 19: 195–199.
4. Sahin S, Karsidag S. An unusual cause of cardioembolic stroke: isolated left ventricular noncompaction. Neurologist. 2009;15:51.
5. Stöllberger C, Finsterer J. Thrombi in left ventricular hypertrabeculation/noncompaction--review of the literature. Acta Cardiol. 2004 ;59:341-4.

CCT

Sulafa KM Ali, MD, FACC, FRCPCH
University of Khartoum
Faculty of Medicine,
Department of Paediatrics and Child Health
PO Box 102
Khartoum Erkaweit, Sudan

AND

Department of Pediatric Cardiology
Sudan Heart Center
PO Box 11917
Khartoum Erkaweit, Sudan

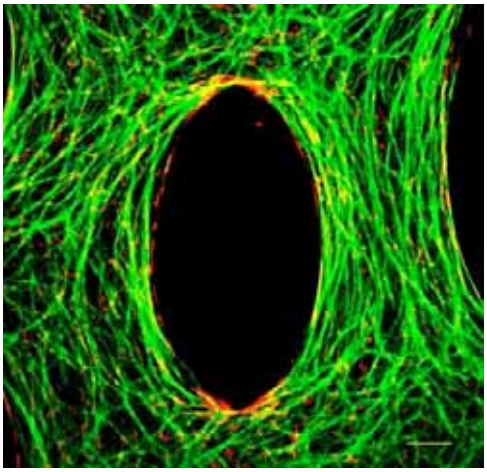
Tel: +24 (918) 3232133/3232137; Fax: +24 (918) 3232135

sulafakhalid2000@yahoo.com

Do you or your colleagues have interesting research results, observations, human interest stories, reports of meetings, etc. that you would like to share with the congenital cardiology community?

Submit a summary of your proposed article to Congenital Cardiology Today at: RichardK@CCT.bz

New Strategy for Mending Broken Hearts?



This immunofluorescence staining image shows the cardiomyocytes in green and the fibroblasts interspersed around them in red. The cells are aligned around the central pore.

By mimicking the way embryonic stem cells develop into heart muscle in a lab, Duke University bioengineers believe they have taken an important first step toward growing a living "heart patch" to repair heart tissue damaged by disease.

In a series of experiments using mouse embryonic stem cells, the bioengineers used a novel mold of their own design to fashion a three-dimensional "patch" made up of heart muscle cells, known as cardiomyocytes. The new tissue exhibited the two most important attributes of heart muscle cells -– the ability to contract and to conduct electrical impulses. The mold looks much like a piece of Chex cereal in which researchers varied the shape and length of the pores to control the direction and orientation of the growing cells.

The researchers grew the cells in an environment much like that found in natural tissues. They encapsulated the cells within a gel composed of the blood-clotting protein fibrin, which provided mechanical support to the cells, allowing them to form a three-dimensional structure. They also found that the cardiomyocytes flourished only in the presence of a class of "helper" cells known as cardiac fibroblasts, which comprise as much as 60% of all cells present in a human heart.

"If you tried to grow cardiomyocytes alone, they develop into an unorganized ball of cells," said Brian Liau, graduate student in biomedical engineering at Duke's Pratt School of Engineering. Liau, who works in the laboratory of Assistant Professor Nenad Bursac, presented the results of his latest experiments during the annual scientific sessions of the Biomedical Engineering Society in Pittsburgh.

"We found that adding cardiac fibroblasts to the growing cardiomyocytes created a nourishing environment that stimulated the cells to grow as if they were in a developing heart," Liau said. "When we tested the patch, we found that because the cells aligned themselves in the same direction, they were able to contract like native cells. They were also able to carry the electrical signals that make cardiomyocytes function in a coordinated fashion."

"The addition of fibroblasts in our experiments provided signals that we believe are present in a developing embryo," Liau said. The need for helper cells is not uncommon in mammalian development. For example, he explained, nerve cells need "sheathe" cells known as glia in order to develop and function properly.

Bursac believes that the latest experiments represent a proof-of-principle advance, but said there are still many hurdles to overcome before such patches could be implanted into humans with heart disease.

"While we were able to grow heart muscle cells that were able to contract with strength and carry electric impulses quickly, there are many other factors that need to be considered," Bursac said. "The use of fibrin as a structural material allowed us to grow thicker, three-dimensional patches, which would be essential for the delivery of therapeutic doses of cells. One of the major challenges then would be establishing a blood vessel supply to sustain the patch."

The researchers plan to test their model using non-embryonic stem cells. For use in humans, this is important for many reasons, both scientifically and ethically, Bursac said. Recent studies have demonstrated that some cells from human adults have the ability to be reprogrammed to become similar to embryonic stem cells.

"Human cardiomyocytes tend to grow a lot slower than those of mice," Bursac said. "Since it takes nine months for the human heart to complete development, we need to find a way to get the cells to grow faster while maintaining the same essential properties of native cells."

If they can use a patient's own cells, the patch would also evade an immune system reaction.

The research was supported by National Institutes of Health, the National Heart Lung Blood Institute and Duke's Stem Cell Innovation program. Other Duke members of the research team were Weining Bian and Nicolas Christoforou.

Quintet of Proteins Forms New, Early-Warning Blood Test Before Heart Attack Strikes

Newswise — A team of Johns Hopkins biochemists has identified a mixed bag of five key proteins out of thousands secreted into blood draining from the heart's blood vessels that may together or in certain quantities form the basis of a far more accurate early warning test than currently in use of impending heart attack in people with severely reduced blood flow, or ischemia.

The work, involving more than a dozen scientists and taking more than a year to perform, is believed to be the largest protein analysis ever done at Hopkins. It was based on 76 arterial blood samples from 19 men and women taken immediately before and after a period of medically induced ischemia lasting as long as 45 minutes.

All had ischemia induced through accelerated pacing of the heart's main chambers. Blood samples were provided by cardiologists at the University of Texas Southwestern.

Key to the researchers' selection criteria for which proteins to analyze from among tens of

thousands in the blood was what they call "a pipeline approach."

"From the start, we knew that we were looking for rare, almost unique biomarkers that bore some direct relationship with ischemia," says study senior investigator Jennifer Van Eyk, PhD, whose first step was to remove from the analysis common blood proteins, such as albumin and globulins. That left batches of 400 proteins for in-depth measure of any changes before and after ischemia.

Their analysis was presented Nov. 9, 2009 at the at the American Heart Association's (AHA) annual Scientific Sessions in New Orleans, found that only the five proteins were present in significantly increased amounts after ischemia occurred, with at least a doubling in the blood concentration, compared with those recorded during healthy blood flow. These were lumican, semenogelin, angiogenin, extracellular matrix protein, and so-called long palate, lung and nasal epithelium carcinoma-associated protein 1.

All of the proteins are believed to originate in the heart, but they can also be found in other tissues varying from the corneas of the eyes (lumican) to semen. Semenogelin, as it is known, has never before been seen in the heart, while others, such as angiogenin, are more predictably found in growing blood vessels and muscle tissue, and are actively involved in tissue repair. Little is known about the remaining two, which ironically have the longest names: extracellular matrix protein, secreted in a rare inflammatory disease; and long palate, lung and nasal epithelium carcinoma-associated protein 1, thought to play a role in innate immunity.

The Johns Hopkins biochemists say the presence of all or even a selected set of these proteins in a simple, rapid blood test could aid emergency paramedics and physicians during the critical 12- to 24-hour window before ischemia causes substantial heart tissue damage or death from heart attack.

A positive reading on a blood test incorporating these proteins, they add, could provide first responders with advance warning to take urgent action, such as using blood thinners like aspirin to prevent clotting, or performing cardiac catheterization to check for any more blockages in the blood vessels feeding the heart, which may in turn prompt more aggressive treatment. Further actions could involve angioplasty, in which a balloon device is threaded into the heart's surrounding blood vessels and then expanded to widen the arteries, or even surgery.

"Our results lay the foundation for a first-of-a-kind, early-warning system that could save tens of thousands of people on the brink of a heart attack," says Van Eyk, a professor at the Johns Hopkins University School of Medicine and its Heart and Vascular Institute. "People experiencing chest pain too often come to the emergency room, with subsequent electrocardiogram, also called EKG, readings not showing any evidence that a heart attack has occurred, but still leaving open the question of whether or not a heart attack is imminent and about to happen or has already happened," adds Van Eyk, Director of the Johns Hopkins NHLBI Proteomics Group and the Proteomics Center at Johns Hopkins Bayview Medical Center, where the protein analysis took place.

Van Eyk says, "People frequently have symptoms of chest pain, shortness of breath and dizziness, with pale or clammy skin coloring, while arterial blood is constricted, but not yet closed. But this myriad of complaints can just as easily be mistaken for the more everyday, less-serious problems of heartburn, stomach cramps or gas."

"A new test based on these five proteins," says Van Eyk, "could provide a 'more definitive answer' to the question 'how serious is it?' much earlier than existing assays for heart attack, such as tests for troponin proteins I and T."

Van Eyk says that commercially available tests for cardiac troponin, which is released into the blood in telltale patterns for heart attack, provide results "too late to take preventive action," and "after some damage has already occurred." Troponin lab tests also depend on the heart muscle dying first, which can take hours to detect, "So a negative reading is unreliable and can still mean that an ischemic problem is about to happen or has already happened," she says.

In the study, the protein analysis was conducted by mass spectrometry machines that can measure the presence of proteins in minute amounts. The machines, operated six days a week for six months, consumed more than 3,700 hours of spectrometric analysis.

Researchers next plan to verify the presence of the five proteins in a larger study with at least 150 participants, and more than 1,000 blood samples. Simultaneously, they plan further analysis of the proteins to map their molecular structures, so that an antibody can be identified to bind to one or several of the proteins, laying the basis for a blood test for ischemia. And they will conduct tests to verify that their study findings also apply to ischemia in stroke.

Funding support for this study was provided by Inverness.

Other Johns Hopkins researchers who took part in this study were Qin Fu, PhD; Simon Sheng, MSc; Steven Elliott, MSc; and Miroslava Stastna, PhD. Additional support was provided by James de Lemos, MD, at the University of Texas Southwestern.

CONGENITAL CARDIOLOGY TODAY

Headquarters
9008 Copenhaver Dr. Ste. M
Potomac, MD 20854 USA

Publishing Management
Tony Carlson, Founder & Senior Editor
TCarlsonmd@gmail.com
Richard Koulbanis, Publisher & Editor-in-Chief
RichardK@CCT.bz
John W. Moore, MD, MPH, Medical Editor/ Editorial Board
JMoore@RCHSD.org

FREE Subscription
Congenital Cardiology Today is available free to qualified professionals worldwide in pediatric and congenital cardiology. International editions available in electronic PDF file only; North American edition available in print. Send an email to Subs@CCT.bz. Include your name, title, organization, address, phone and email.

Contacts and Other Information
For detailed information on author submission, sponsorships, editorial, production and sales contact, current and back issues, see website or send an email to: INFO@CCT.bz.

MELODY®

Transcatheter Pulmonary Valve

Providing new options for the lifetime management of patients with congenital heart disease

A nonsurgical breakthrough in treating RV to PA conduit dysfunction

The Melody® Transcatheter Pulmonary Valve and Ensemble® Transcatheter Delivery system have received CE Mark approval and are available for distribution in Europe. Additionally, a Medical Device Licence has been granted and the system is available for distribution in Canada. Products are not available for sale in the United States.

Medtronic

Alleviating Pain · Restoring Health · Extending Life

TINY HEARTS INSPIRED HYBRID LABS WITH ACCESS FOR BIG TEAMS.

Fixing a heart from birth through adulthood takes big teams working together. So we examined the needs of leading clinicians when designing our hybrid solutions. The result: our Infinix™-i with 5-axis positioners and low profile detectors, stays out of the way, but right where needed, providing the best possible access to patients. To lead, you must first listen.

medical.toshiba.com

TOSHIBA
Leading Innovation