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An Example of Pectus Excavatum Causing Tricuspid Valve Compression

By Jeffrey Womble, MD; Robin Fountain-Dommer, MD; Marc T. Downing, MD; and Michael J. Leinwand, MD, FACS

Abstract

A 15-year-old girl with pectus excavatum was referred for surgical evaluation by her cardiologist due to tricuspid valve stenosis possibly secondary to compression of the tricuspid valve by the sternum. Also present was moderate to severe pulmonary hypertension. Historically, most studies have shown minimal physiologic benefit from repair of pectus excavatum. We present an interesting case showing significant improvement in the degree of tricuspid valve stenosis, the estimated pulmonary artery pressures, and the patient's symptoms following pectus excavatum repair.

Background

Pectus excavatum is a chest wall deformity that is not typically associated with physiologic abnormalities, though it often has psychosocial ramifications. Most patients are asymptomatic, but some patients complain of decreased exercise tolerance. Other related issues include pain along the costal cartilages and recurrent respiratory infections.

Though many patients report vague, subjective symptoms such as dyspnea on exertion, weakness, or poor exercise tolerance; few objective studies have been able to demonstrate significant physiologic benefits from repair. Pulmonary function tests performed with patients at rest have shown low-normal to normal results. Exercise pulmonary function tests, however, have demonstrated early fatigability with a restrictive pulmonary pattern. Additionally, some studies have shown postoperative improvement in maximum voluntary ventilation, maximum oxygen utilization, total lung capacity, and total exercise time.

“We present an interesting case showing significant improvement in the degree of tricuspid valve stenosis, the estimated pulmonary artery pressures, and the patient’s symptoms following pectus excavatum repair.”

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From a cardiac standpoint, there is a well-known association between pectus excavatum and mitral valve prolapse. In addition, there have been reports of compressive effects on the heart including tricuspid stenosis, compressed innominate vein syndrome, and right heart compression.

We present a patient with pectus excavatum and mild symptoms of exercise intolerance. She was found to have significant derangements in her cardiac hemodynamics, all of which improved or completely resolved following repair of her deformity.

Case Report

E.C. is a 15-year-old girl referred by her cardiologist for repair of a pectus excavatum deformity. At age 9, she was found to have supraventricular tachycardia and a mild pectus excavatum deformity. An echocardiogram showed mild mitral valve prolapse with mild mitral valve insufficiency, patent foramen ovale, and moderate tricuspid valve stenosis. At fifteen, she was found to have a more pronounced pectus excavatum deformity. She had become involved in competitive dance and complained of some shortness of breath with particularly long routines. Repeat echocardiography showed significant right atrial and left atrial enlargement consistent with restrictive cardiac physiology. Right ventricular pressures were estimated at 70-80% systemic, consistent with moderate to severe pulmonary hypertension. There was significant compression of the tricuspid valve annulus with moderate tricuspid valve stenosis. Also present was mitral valve prolapse.

The patient underwent open (modified Ravitch) pectus excavatum repair without complication. Three months later, her mild symptoms resolved. Echocardiography showed the tricuspid annulus size had increased from 1.2mm to 1.5mm with only mild tricuspid valve stenosis. There was improvement of the right and left atrial enlargement with normal estimated right ventricular pressures. (Table 1)

Discussion

Pectus excavatum deformities do not typically cause significant cardiac compromise. Repair is usually performed to correct the congenital deformity which can have significant psychosocial impact during adolescence. Though many studies have looked at various cardiac and pulmonary parameters, only a few have shown objective evidence of improvement after repair. In our patient, compression on the heart by the sternum, significantly narrowed the tricuspid annulus. This resulted in altered cardiac flow dynamics. Following open repair of the deformity, the size of the tricuspid annulus increased and estimated pulmonary artery pressures returned to normal. The patient's symptoms also resolved. This case demonstrates that the pectus excavatum deformity can occasionally result in cardiac compromise. Repairing the defect can resolve anatomic issues related to compression of the heart by the sternum and improve cardiopulmonary physiology.

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Table 1: Comparison of Preoperative and Postoperative Echocardiography

Parameter	Preoperative Result	Postoperative (3 months) Result
Ventricular function	Normal	Normal
Atrial enlargement	Moderate	Mild
Mitral prolapse	Mild to Moderate	Mild
Mitral insufficiency	Trivial	Absent
Tricuspid size	1.2mm	1.5mm
Tricuspid inflow peak pressure gradient	8mmHg	3.5mmHg
RV to RA pressure gradient	71mmHg	29mmHg
RV pressure	70-80% systemic	35% systemic (normal)



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“In our patient, compression on the heart by the sternum, significantly narrowed the tricuspid annulus. This resulted in altered cardiac flow dynamics. Following open repair of the deformity, the size of the tricuspid annulus increased and estimated pulmonary artery pressures returned to normal. The patient’s symptoms also resolved.”

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- **BREAKOUT SESSIONS** for cardiovascular nurses and CV technicians.
- **MEET THE EXPERT SESSION**, held on Sunday, will give attendees the opportunity to discuss difficult cases with our renowned faculty.

ACCREDITATION The Society for Cardiovascular Angiography and Interventions is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to sponsor continuing medical education for physicians.

Abstract Submission Deadline is March 15, 2008.

For registration and abstract submission go online to www.picsymposium.com



Highlights from The Charleston Symposium on Congenital Heart Disease - May 2-4, 2007

By Girish S Shirali, MBBS, FAAP, FACC

The Children's Heart Program of South Carolina hosted the 15th Charleston Symposium on Congenital Heart Disease in the historic city of Charleston, SC, from May 2-4, 2007. The Symposium highlighted the central role of cutting-edge cardiac imaging in guiding our understanding of cardiac development and anatomy, and the impact of cardiac imaging on the management of patients with congenital heart defects. Imaging modalities that were highlighted including 3D echocardiography, CT angiography, MRI, ultrasound biomicroscopy, intracardiac echocardiography and 3D mapping tools for intracardiac electrophysiology. Over 150 attendees and 30 proctors and exhibitors came from locations that ranged from all over the USA and Canada to Germany and New Zealand. Attendees consisted primarily of pediatric cardiologists (~ 65%, consisting of those in practice as well as in fellowship training) and sonographers (~25%), with some surgeons, intensivists and nurses (~10%).

The meeting featured guest faculty from traditionally disparate sub-disciplines and a wide range of institutions: the Children's Hospital of Philadelphia (Drs. Gil Wernovsky, Tom Spray and Paul Weinberg), Great Ormond Street Hospital for Children (Prof. Robert H. Anderson, Martin Elliott and Andrew Taylor), the University

of Chicago (Dr. Ziyad Hijazi) and All Children's Hospital, Tampa (Diane Spicer). Faculty from the Medical University of South Carolina included Drs. Andrew Atz, Scott Bradley, Fred Crawford, Anthony Hlavacek, Tim McQuinn and Girish Shirali.

The teaching environment was unique in its degree of technical sophistication. It was designed as an interactive, hands-on workshop for attendees to learn 3D echocardiography cropping techniques. Over fifty PC workstations were loaded with identical datasets and 3DE software. Over 30 proctors provided personal





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attention to attendees at PC workstations. Between 3 and 4 attendees shared each workstation.

Each of the 3 days of the conference was positioned around a central theme (The Atrioventricular Junctions, The Outflow Tracts and Septation), which was explored from the perspectives of cardiac development, pathology, imaging, surgery and perioperative management, ending with a discussion of long-term outcomes. Every day, we started with Dr. McQuinn's demonstration of how the avian heart develops in early gestation. Dr. McQuinn's use of ultrasound biomicroscopy enables him to obtain spectacular moving images of the heart as it loops, and of the developing endocardial cushions and septums. This set the stage for Prof. Anderson, Paul Weinberg and Diane Spicer, who demonstrated heart specimens: normal as well as a wide range of pathology. Each heart specimen was matched with a 3D echocardiographic

dataset that demonstrated identical findings. Dr. Shirali manipulated the 3D dataset in real time, thus enabling attendees to develop familiarity with the roles of cropping and other techniques that typify 3D echocardiography. Next, a 'recipe' of the cropping maneuvers was displayed. Now, it was time for attendees to open up the identical 3DE datasets on their computers and manipulate them to achieve the identical result. The recipe and the proctors served as guides to help attendees to achieve this goal. This method enables attendees to develop hands-on familiarity with 3D echocardiography.

Next, we moved into the realm of surgery and perioperative management. The surgeons embellished their descriptions of operations with spectacular intra-operative videos. These brought home many important surgical concepts, all tied to the theme of comprehensive preoperative diagnosis and customizing the imaging to the individual patient and defect. After the surgeons had detailed their approach, it was up to the intensivists to detail



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cutting-edge perioperative management techniques.

After the morning sessions, heart specimens were available for the rest of the day for interested attendees' hands-on learning. Each afternoon, we had two concurrent breakout sessions. One continued on the theme of cardiac imaging, employing 3D echocardiography, CT or MRI. The other breakout session focused on perioperative issues pertaining to the topic under discussion. The latter session included didactic presentations as well as case presentations with audience participation.

The final session each day focused on long-term management issues featuring, for example, outcomes after repair of AV septal defect or tetralogy of Fallot. This was also the time for some philosophical discussions: Martin Elliott's presentation, titled 'Should we do it just because we can?' was moving and thought-provoking.

Coming Soon....

The next Symposium will be held from November 16 to 19, 2008, in Charleston, SC at the Francis Marion Hotel. We plan a similar program with 60 PC's and a maximum of 200 attendees, with the following themes:

- Valve Repairs (Mitral, Tricuspid and Common AV valves)
- Double Outlet Right Ventricle
- Ventricular function

We shall feature breakout sessions that are geared to fellows in training, including career guidance and opportunities to network with potential employers.

Thanks to MUSC sonographers, fellows and junior faculty who helped out as Proctors, to Debbie Bryant who helped organize everything, and to our IT folks for helping put together an unprecedented event! For more information go to: www.musckids.com/heart/ or contact Debbie Bryant, Meeting Coordinator at byrantd@musc.edu; tel: 843-792-3286.

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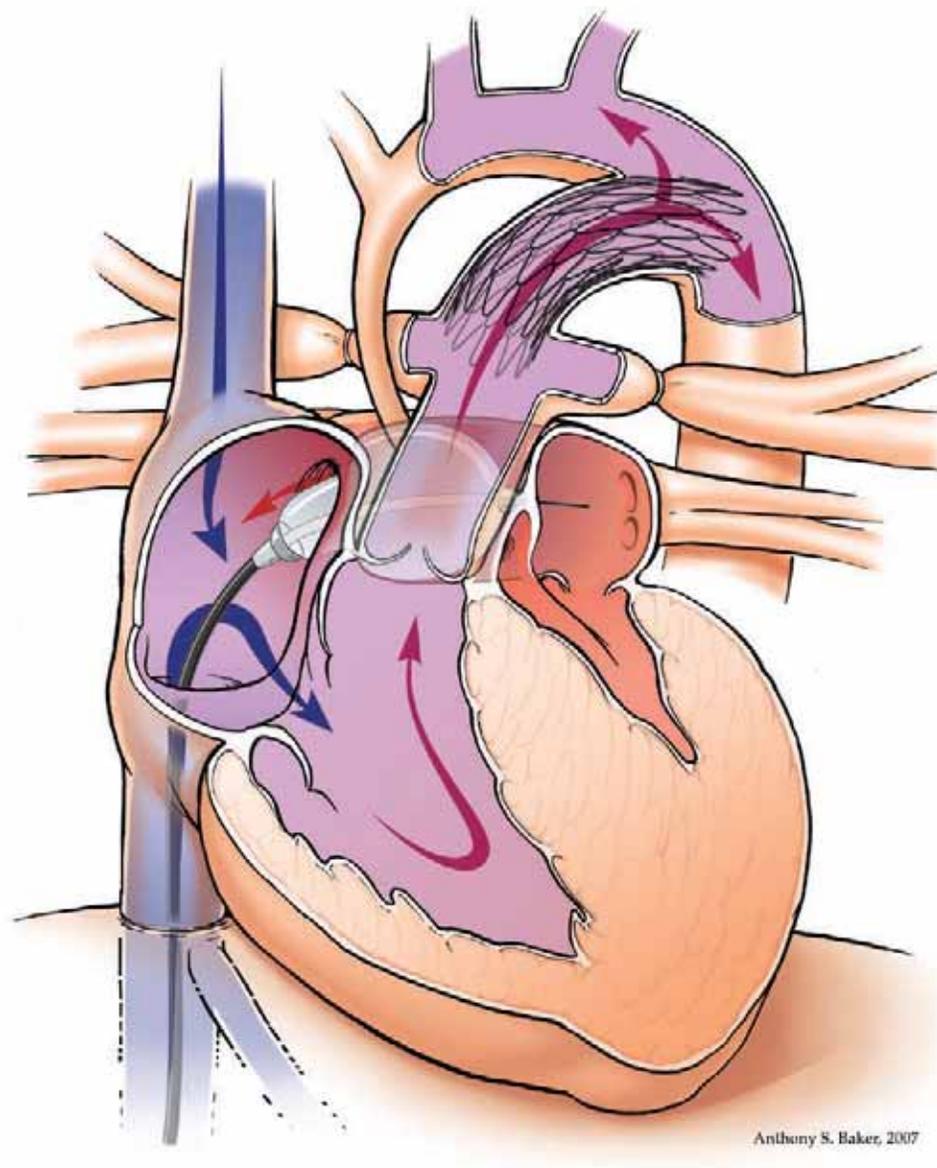
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Improved Patient Care Through Immediate Access to Useful Clinical Data: Epocrates® ACC Essentials for Cardiology

By Chi-Ming Chow, MDCM, MSc, FRCPC, FACC, FASE

Introduction

With the explosion of medical knowledge, it has been getting more and more difficult to stay up-to-date. There has also been an increasing need to bring the latest medical information to the bedside to help make clinical decisions for our patients. Fortunately, personal digital assistants (PDAs) have become indispensable clinical tools to help us stay current and serve as our "peripheral brains" in our day-to-day clinical activities. As an academic cardiologist with a busy inpatient and outpatient practice, I was one of the early adopters of PDAs and mobile medical software. I immediately noticed a significant difference in patient care and efficiency in information retrieval. I no longer have to interrupt a patient encounter or exit the exam room to thumb through heavy medical texts and drug reference books when I am seeing patients. In this article, I will address how the PDA reference has been a great help to me, by providing valuable point-of-care information at the bedside and how customized cardiology content improved efficiencies and accuracy in clinical practice.

Cardiology Goes Digital

After trying multiple mobile medical software and references on my PDA, I found most of what I need is in Epocrates. I have



Doctors with PDA

been using Epocrates® Essentials for Cardiology regularly to look up drug dosages, interactions among different drugs and alternative medications. In fact, nearly 60 percent of physicians reported avoiding one or more medical errors per week using Epocrates' references [1].

I can attest to the relevance of the cardiology tools as the developer and author of the CardioMath application featured in this software bundle. I conceived the idea of CardioMath when I was a cardiology fellow. In preparation for the board certifying examination, I had to remember many formulas used in echocardiography, hemodynamics, exercising stress testing, etc. When I entered into practice, I found it hard to remember all the formulas, and it was very time-consuming to do the calculations. Remembering the formulas and manually applying them created a barrier to operating a smooth and efficient practice and this problem became the impetus behind the creation of Cardio Math PDA and online calculators. A recent survey conducted among the online CardioMath calculators (www.csecho.ca/cardiomath), showed that 80% of the users thought that using CardioMath increased quantification in their clinical practice, and 80% thought that the program improved patient care[2]. To further support the growth and value of mobile technology, the American College of Cardiology (ACC) partnered with Epocrates® to create a customized and comprehensive set of cardiology appli-

"There has ... been an increasing need to bring the latest medical information to the bedside to help make clinical decisions for our patients. Fortunately, personal digital assistants (PDAs) have become indispensable clinical tools to help us stay current and serve as our 'peripheral brains' in our day-to-day clinical activities."



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cations for practicing physicians, known as Epocrates® ACC Essentials for Cardiology. This specialized product combines Epocrates® Essentials' premium drug, disease and diagnostics guide, with relevant cardiology applications including cardiac calculations, an obesity tool and a cardiology news channel.

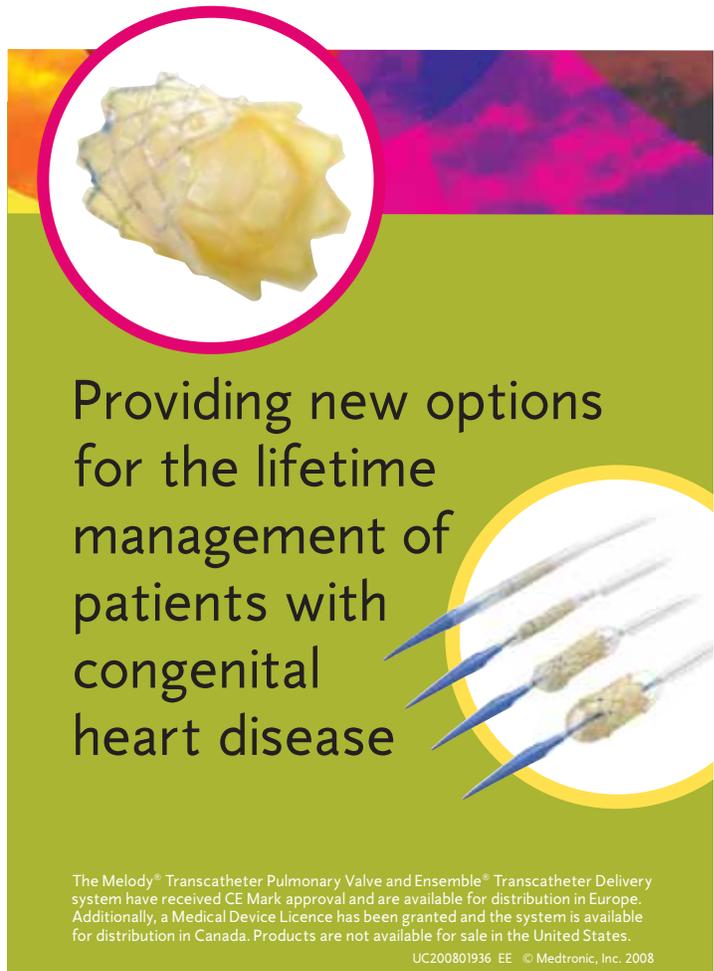
Cardiology-Specific Application

1. **Epocrates® Essentials** – A premium guide that includes information about more than 3,300 drugs; over 1,200 diseases and conditions; hundreds of lab test panels; and decision support tools such as drug interaction checking, symptom assessments and treatment recommendations. In addition, there is a database of more than 400 alternative medications. For example, you can look up interactions between green tea, ginseng or warfarin with a prescription drug. In addition to the USA, the drug database also covers medications available in Canada, the UK, and Germany.
2. **CardioMath®** – A suite of 50 commonly-used formulas in cardiovascular medicine. The application was designed to provide a way to perform day-to-day calculations associated with echocardiography, ECG, epidemiology, exercise stress test, general medicine, hemodynamics and unit conversion (SI and US), both accurately and efficiently. Each area includes the formulas, as well as supporting information such as reference values and the clinical use. For example, you can calculate the Duke Treadmill Score without leaving the testing area and look up the patient's prognosis as well. In addition, you can calculate the Qp/Qs by using the echocardiogram or catheterization data.
3. **Body Mass Index (BMI) and Obesity Application** – During the past 20 years, obesity among adults and adolescents has risen significantly in the North America. To support cardiologists with patient diagnosis, education and treatment, this mobile application features a detailed weight management tool. The application, using body mass index (BMI) and waist circumference measurements, provides recommendations, weight loss guidelines, a Basal Energy Expenditure calculator and other useful information.
4. **Clinical Messaging Alerts** – Timely medical news and interesting journal abstracts are delivered to cardiologists on their mobile device to keep them up to date. ACC Cardio-source is a valuable contributor to the cardiology messaging channel by providing access to relevant headlines such as, "Ten Facts to Remember about Atrial Septal Defects in the Adult" and "Influenza Vaccination as Secondary Prevention for Cardiovascular Disease."

No Cardiologist Left Behind

Cardiologists who do not own a mobile device, are not bound to the traditional textbooks, which are outdated as soon as they are printed. Most of Epocrates® clinical content is also available online. In the hospital where I work, Internet access is available on all our exam room desktop computers and in inpa-

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Formulary Coverage on Palm Device

tient wards, so there is no trouble accessing the online version. Epocrates® Online provides information about medications and alternative medications similar to the PDA version. The MultiCheck function is also very easy to use and remembers my last search. I can also print out potential drug-drug interactions for my patients and my students.

Epocrates® Online has two new features that I find particularly helpful: the pill identifier and the patient handouts. Since a large proportion of my patients are non-English speaking immigrants, it is often hard for them to recall the names and dosages of the medications that they are taking. I have been using the pill picture function to help identify the medications and dosages that my patients are taking. I have many patients who take warfarin. I can bring up the pictures of the different dosages of warfarin from Epocrates® Online. Plus, the patients in identifying the dosage of their medication by the color of the pill that they are taking. In addition, there is a

new patient education handout section in Epocrates® Online, which is available in English or Spanish. I have been printing these handouts for patients who have questions on how to take the prescribed medications. This feature really saves a lot of time and space for me to keep all the patient handouts on medications, and allows my patients to go home with answers to many of their questions on

“As technology advances, the use of heavy paper-based medical references will be a thing of the past. A plethora of ever more powerful PDAs and ultra-mobile personal computer (UMPC) that are linked to the Internet in real time are becoming ubiquitous in the clinical setting.”

one sheet of paper. Epocrates® Online well-complements the PDA version and I have been using both of these versions frequently.

Summary

As technology advances, the use of heavy paper-based medical references will be a thing of the past. A plethora of ever more powerful PDAs and ultra-mobile personal computer (UMPC) that are linked to the Internet in real time are becoming ubiquitous in the clinical setting. By having clinical information readily available at the point-of-care, cardiologists can make better-informed deci-

sions for the patients. Epocrates® ACC Essentials for Cardiology is a useful collection of mobile references and tools that can help us stay current, be more efficient, and potentially provide better care for our patients. Epocrates® ACC Essentials for Cardiology is available for both the Palm OS device as well as for Windows Mobile / Pocket PC devices. Epocrates® for PDAs and Epocrates® Online are available at: www.epocrates.com.

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Medical News, Products and Information

Three Patients, Age 14, 28 and 72, Receive Heart Valve Replacements Without Surgery Using High Tech Investigational Device

CHICAGO - Interventional cardiologists at Rush University Medical Center now offer a minimally-invasive transcatheter valve replacement procedure for patients with congenital heart disease that doesn't involve open heart surgery.

Rush is one of three sites taking part in the investigational device exemption (IDE) feasibility study of minimally-invasive pulmonic valves and successfully implanted the first three patients enrolled in the trial on April 17, 2008.

"We were able to successfully implant the Edwards SAPIEN transcatheter heart valve percutaneously in the first three patients treated in this trial. All of the patients are recovering and are expected to go home today," said Dr. Ziyad M. Hijazi, Director of the Rush Center for Congenital and Structural Heart Disease, Chief of the Section of Pediatric Cardiology and Professor in the Departments of Pediatrics and Internal Medicine at Rush University, Chicago. "Patients with congenital right ventricular outflow tract problems typically face the burden of multiple open-heart surgeries throughout their lives, either to replace their 'native' diseased valves or, as they age, their bioprosthetic replacement valves."

Dr. Hijazi, an interventional cardiologist and pioneer in nonsurgical repair of the heart, and his colleagues, Dr. Clifford J. Kavinsky and Dr. Zahid Amin, used a bovine pericardial heart valve that can be compressed onto a balloon to the approximate diameter of a pencil, threaded from the leg into the circulatory system and deployed across the patient's pulmonary valve. The valve re-

placement is accomplished as a "beating heart" procedure, without requiring cardiopulmonary bypass or an open-chest incision.

Edwards Lifesciences Corporation of Irvine, Calif., makes the Edwards SA-PIEN transcatheter valve that was implanted.

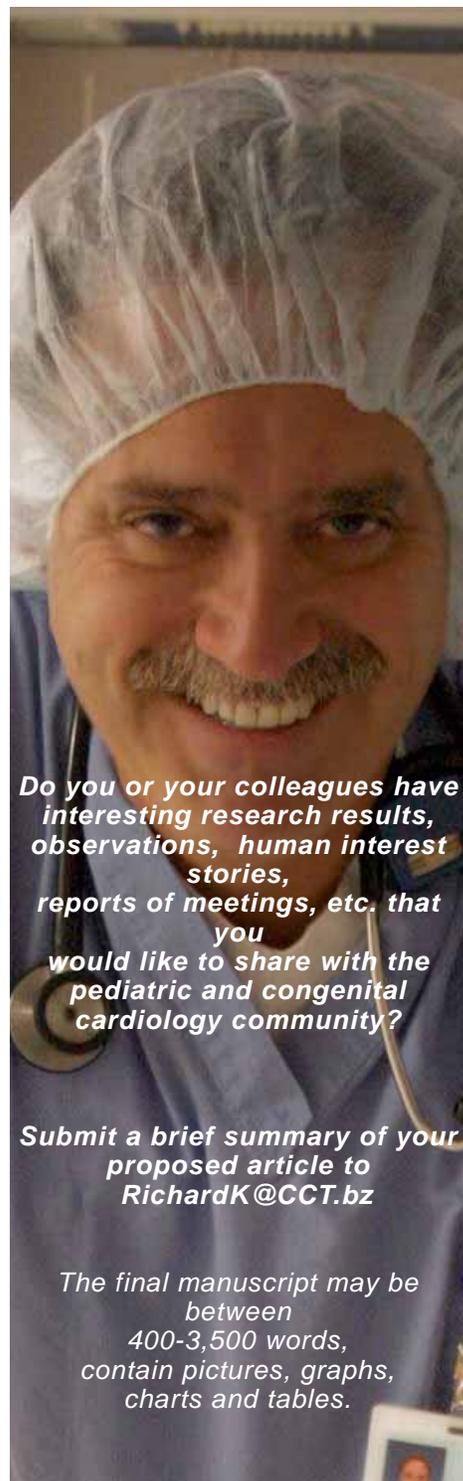
"We can replace heart valves in high-risk patients with severe pulmonary stenosis (abnormal narrowing in a blood vessel) who might not be candidates for conventional valve replacement surgery. Instead, these patients can benefit from a transcatheter valve replacement procedure done minimally-invasively, without cardio-pulmonary bypass, that has the potential to shorten recovery time," said Hijazi. "This clinical study will enable physicians to offer a minimally-invasive alternative to symptomatic patients with a regurgitant, or leaky, pulmonary valve conduit, giving them the opportunity to recover and resume their normal activities."

"My team is proud to be able to address this serious unmet patient need and to offer the chance to take one or more surgeries out of the treatment course for these patients," said Hijazi.

The US Food and Drug Administration (FDA) conditionally approved the investigational device exemption (IDE) clinical trial in late 2007.

Genetic Test Offers Clues About Cardiac Hypertrophy in Children

A mechanic uses diagnostic tests to determine why your engine is making strange sounds before lifting a wrench to fix the problem. Pediatric cardiologists would love to take a similar approach with patients experiencing cardiac hypertrophy—a thick-



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 pediatric and congenital cardiology community?

Submit a brief summary of your proposed article to RichardK@CCT.bz

The final manuscript may be between 400-3,500 words, contain pictures, graphs, charts and tables.

+++ Live Case Demonstrations from Frankfurt, Birmingham, Milan and Toronto +++ Live Case Demonstrations from Frankfurt, Birmingham, Milan and Toronto +++



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ening of the heart muscle. Rational treatment requires understanding the underlying causes of disease. But doctors know little about the causes of cardiac hypertrophy in children, so no diagnostic tests have yet been developed.

A new study, which appears online April 9, 2008 in the *New England Journal of Medicine* might change that. Working with a team of researchers, Harvard Medical School Professors Christine Seidman and Jonathan Seidman discovered that some children with unexplained cardiac hypertrophy harbor mutations in the same 10 genes responsible for the condition in many adults.

"Labs have done work on the genetic underpinnings of cardiac hypertrophy in adults, but few thought that the research applied to children," says Christine Seidman, who is also a member of the HMS-Partners HealthCare Center for Genetics and Genomics and an investigator with the Howard Hughes Medical Institute. Her lab is located at Brigham and Women's Hospital. "For years, doctors assumed the two conditions were clinically distinct."

According to Christine Seidman, 3 to 4% of adults have cardiac hypertrophy. Data from the Framingham Heart Study suggest that mutations in 10 genes-identified by the Seidmans and others-account for 1/5 or 1/6 of those cases. Most of the genes implicated encode "sarcomere" proteins, which make up the heart's contractile apparatus. They literally help the muscle tighten and relax to pump blood.

"Cardiac hypertrophy increases your risk of all types of negative cardiovascular outcomes, including heart failure and sudden death," says Jonathan Seidman. "Although the condition is rare in children, the prognosis is even worse. Kids with cardiac hypertrophy are often candidates for heart transplantation."

The Seidmans worked with Amy Roberts, assistant professor of pediatrics at Children's Hospital Boston, and Jeffrey Towbin, a professor of pediatric cardiology at Baylor College of Medicine, to find children with cardiac hypertrophy. They extracted DNA from 84 individuals diagnosed with the condition before age 15.

Just 33 of those children had family histories of cardiac hypertrophy.

Using a novel chip technology developed by molecular geneticist Heidi Rehm and others at the Harvard-Partners Center for Genetics and Genomics Laboratory for Molecular Medicine, they sequenced the 10 suspect genes.

The chip allowed them to read tens of thousands of nucleotides-of letters of the DNA "alphabet"-for a fraction the cost of traditional technology.

The team identified mutations in 25 of the 51 children without family histories and in 21 of the 33 children with family histories.

"I think it's remarkable that we found mutations in nearly 50% of the kids who didn't have family histories," says Christine Seidman.

The team took a closer look at the genes of these patients' parents. Eleven pairs of parents agreed to participate in the study. In 7 cases, one parent harbored the same mutation as his or her child. Though these adults assumed their hearts were fine, echocardiograms revealed thickening of the muscle in some cases.

"We still don't know why the children presented symptoms so much earlier than their parents," says Jonathan Seidman. "We suspect that other genes must influence the disease presentation."

Further genetic testing of children and their parents could shed light on this and help doctors choose appropriate treatments.

"This study demonstrates that kids who present with sporadic cardiac hypertrophy deserve the same genetic test as adults," says Christine Seidman.

This research was supported by grants from the Howard Hughes Medical Institute, the National Heart Lung and Blood Institute, the NFL Charities Foundation and the Children's Cardiomyopathy Foundation.

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