CONGENITAL CARDIOLOGY TODAY

Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

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A Cautionary Tale for Pediatric Cardiologists

By John W. Moore, MD, MPH

I received an email from Marty Stein, a senior developmental pediatrician at Rady Children's Hospital, asking me whether I had come across the book, *Immortal Bird* by Doron Weber. Marty said that it brought up a number of serious communication issues between the patient's family and the physicians who cared for him, as well as communication and relationship issues among the cardiologists and the cardiac surgeons. "Did the issues raised in the book ring true?" he asked me. I looked at reviews of the book which were mostly positive, read the book, and decided it could be an excellent teaching tool for our fellows and staff

The book begins with a quote from "Ode to a Nightingale" by John Keats. The first line of which is "Thou wast not born for death, Immortal Bird!" The very line embodies the deep love and profound loss felt by Doron Weber after the death of his son, Damon.

Immortal Bird was not written for a professional audience, so many medical details of Damon Weber's case are missing or obscure. It appears that the boy's underlying heart condition is AV canal defect, probably in the context of a Heterotaxy Syndrome. He was cared for at Columbia in the 1990's and early 2000's. Dr. Jan Quaegebeur performed his initial surgeries, including Fontan completion. From ages four to twelve, Damon grew normally, was medication-free and his cardiologist "marvels at his progress." But, his parents began to notice that his abdomen looked "paunchy" and that he stopped growing. At first, his cardiologist reassured Damon's parents that he was fine. Soon thereafter, Damon revealed to his father that he had swollen testicles and, ultimately, it became clear that he had a ruptured hernia, an enlarged liver and had developed Protein-Losing Enteropathy (PLE). Overtime, he was treated with medications and a special diet. His PLE progressed, and Dr. William Hellenbrand created a fenestration in the catheterization lab. Damon also received albumin and IVIG infusions. He was treated with steroids, and heparin therapy was considered. Although each of these therapies provided temporary improvements, the PLE was relentless.

Damon eventually was listed for and had a heart transplant performed by Dr. Jonathon Chen. The Columbia pediatric transplant team, given the pseudonyms of Drs. Mason, Davis, Becker and Sanford, orchestrated his pre- and post-transplant care. Damon survived transplantation and left the hospital. Soon thereafter he developed fever, other systemic symptoms, thrombocytopenia, and liver dysfunction. The transplant team treated him for early rejection with a boosted immunosuppressive regime, in spite of a negative biopsy and an Ebstein—Barr Virus (EBV) mismatch. He died shortly thereafter from something Weber calls fulminate Post-Transplant Lympho-Proliferative Disorder (PTLD), but as described, sounds more like viral sepsis and multi-organ failure. Damon lived 16 years.

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My feelings as I read this book were complicated. Doron Weber's name-dropping, sense of entitlement and privileges, initially put me off. I almost put the book down after reading only the first few chapters. I felt that Weber would have nothing to say about my patients from less affluent and less sophisticated middle and lower class families. I read further mainly because I wanted to respond to Marty. What unfolded was a deeply moving story of a father's love for his son, and of the father's arduous and unsuccessful battle to save his son's life. During this struggle, Weber lost trust in most of his son's doctors, and felt abandoned by or isolated from many of them. He contacted a number of additional institutions, looking for better treatment options. His attempt to manage his son's care did not build bridges with the physicians at Columbia. Ultimately, in spite of a heart transplant, all efforts were in vain and, eventually, Damon died. In the epilogue, Weber explained that he was so angry and so distraught about his son's care that he initiated "an all-out hunt for justice" in the form of a malpractice suit. Damon's medical records, shipped to an off-site facility were "lost," further fueling Doron's anger and grief. He pours his grief into the memoir.

I'm glad that I read *Immortal Bird.* The book provides a cautionary tale for pediatric cardiologists about how and what to communicate to cardiac patients and their families. Damon, for sure, is one of pediatric cardiology's archetype patients. His case was complicated; he needed careful follow-up; and eventually, he needed timely treatment for severe complications of single ventricle and Fontan palliation. His father, a former Rhodes Scholar and boxer, applied his pugilistic spirit to Damon's problems. Like any good parent, he was caring and involved. Unlike most, he felt responsible for his son's care, attending an international PLE medical symposium, making a list of all the known treatments for PLE based on his review of the medical literature and labeling this list his "battle plan." Doron Weber was also a hopeless doctor shopper with the means to take his son anywhere for consultation. He sought opinions from four different institutions, numerous individuals and even a Nobel Prize winner. To say the least, he was a complicated and difficult parent. Creating and maintaining a healthy or even an adequate professional alliance with Doron Weber would have been a challenge for any cardiologist.

In the memoir, we have a sad and angry retelling of the course of Damon's treatment. We also learn of a close and loving father's best attempts to cope with an overwhelming situation. We come to know Damon as a bright, talented kid with a flair for acting, who, when on stage, is "like a bird that can only display its full plumage in a native habitat." As for confronting his cardiac issues, his father's description of him as "a brave sixteen-year old boy with a lion's heart" couldn't be more apt. Doron's artful description of Damon, his life, his playfulness, and his implacable spirit engenders affection for him in any reader. His death is a sad ending capable of evoking intense memories of other patients lost.

The book is not without a few light moments; my favorite is when Doron Weber consults with Dr. Alvin Chin at Children's Hospital of Philadelphia (CHOP) to complain about Damon not being properly listed on the Columbia transplant list. Dr. Chin reminds Weber that everyone makes mistakes, and that he probably wouldn't do any better at CHOP, because half the CHOP staff trained at Columbia!

There are many important issues raised in this book: Who is responsible for Damon's outpatient care and for his care plan? Is it the "team," the primary cardiologist, and/or the surgeon? How should the child's family be prepared for possible future complications and his uncertain long-term prognosis? Who should educate the family? Furthermore, without agreed upon algorithms for surveillance or treatment, how should Damon's care be managed? How accessible should his cardiologists be to the patient and family? Should they give the family their cell phone numbers and emails? What are the responsibilities of the "on-call" cardiologist? What is the role of the cardiology fellow receiving calls from outpatient families or PCP's about patients? Should care be "handed off" between team members when the child's most identified cardiologist is away or otherwise unavailable? How should his cardiologists react to intrusions or recommendations by VIP's, authorities, or well-know pediatric cardiologists from other programs?

Programmatic and system issues are also raised: Does it make sense to use the ED as a place to evaluate well-known chronically ill cardiology

patients or post-transplant patients, or should they be admitted directly to the ward? What is the role of the PICU attending in the care of critically ill cardiac patients who are in the PICU? Who provides and who plans the care in the PICU? Who performs and is responsible for needed procedures (lines, tubes, etc.)? Should there be a CVICU staffed by cardiologist-intensivists? What is the role of the inpatient cardiology fellow? Are there sufficient inpatient cardiology beds available, such that patients like Damon can be properly accommodated? Is there an electronic medical record? How are communications between physicians caring for inpatients occurring? Is critical information exchanged well among practitioners (things like EBV mismatch between donor and recipient)? What is the role of consultants in care planning? Who provides strategic direction of care when the patient is sick and in the hospital? Who provides the key communications with the family and the patient? How can a hospital "lose" important patient records?

Immortal Bird is a powerful personal tale of the loss of a child, and it also provides important lessons for the congenital cardiology community. Damon's story should inspire us to strive for the best possible communications with families and with one another. Review and consideration of the medical issues raised in the book may lead to improvements in the way we care for and treat our patients. I highly recommend that all congenital cardiologists and cardiac surgeons read the book.

Study Guide

Immortal Bird is so loaded with relevant medical and behavioral topics that I decided it merited being the focus of discussion for two sessions of our regular fellowship educational lecture series. I purchased three-dozen used books on Amazon, and provided a copy to each fellow, cardiologist, surgeon, and nurse practitioner in our program. I scheduled one-hour medical and behavioral sessions to occur two months after I distributed the books. I thought that the "readers" among us would finish the book quickly, and word of mouth would take care of the rest. I was right. After two months nearly everyone had read the book, and I was party to many informal discussions about it. By the time the scheduled sessions rose on the calendar, I expected and received large audiences. Marty Stein joined us as well.

Medical Session

In the medical session I reviewed the medical problems encountered by Damon Weber, including PLE, transplantation for failed Fontan, and PTLD. I found the following references to be the most useful and informative:

- *Mertens L, Hagler D, Sauer U, et al. Protein-losing enteropathy after the Fontan operation: an international multicenter study. J Thorac Cardiovasc Surg 1998;115:1063-1073.* (Background, much of my pre-review knowledge came from this report).
- Rychik J. Protein-Losing Enteropathy after Fontan Operation. Congenit Heart Dis. 2007;2:288-300. (A slightly dated review, with a sophisticated discussion of factors involved in pathophysiology).
- *Meadows J, Jenkins, K. Protein-losing enteropathy: integrating a new disease paradigm into recommendations for prevention and treatment. Cardiology in the Young 2011;21:363-377.* (The most recent comprehensive review, good references, also a summary of the Boston Children's Hospital experience with PLE).
- *Bernstein D, Naftel D, Chin C, et al. Outcome of Listing for Cardiac Transplantation for Failed Fontan. Circulation 2006;114:273-280.* (Multi-center review of results from pediatric transplant group, provides separate analysis of PLE patients).
- *Davies RR, Sorabella RA, Yang J et al. Outcomes after transplantation for "failed" Fontan: A single-institution experience. J Thorac Cardiovascular Surg 2012;143:1183-1192.* (Review of Columbia's experience, separates PLE patients, includes period when Damon received a transplant).
- *Manihiot C, Poliock-DarZiv S, Holmes C, et al. Post-transplant lymphoproliferative disorder in pediatric heart transplant recipients. J Heart Lung Transplant 2010;29:648-657.* (Current experience with PTLD, summary of characteristics, relationship to EVB).

During the medical discussion, my group expressed interest in developing and adapting a common algorithm for surveillance and maintenance care of single ventricle patients after Fontan. We noted that among eleven cardiologists, we had eleven slightly different approaches toward things like monitoring serum albumin and liver function, as well as chronic anticoagulation, use of diuretics and afterload therapy. There was a consensus among us that a more consistent approach could improve patient care for a variety of reasons.

Behavioral Session

The behavioral session was new territory for me. I had no guidance and little experience. My goal was simply to host an active discussion of issues raised by the author. My method was to raise issues for discussion by reading relevant quotations from the book, such as:

- Thinking his son's surgery was a "cure," and having pointed out Damon's paunchy belly, slowdown in growth, asthma-like symptoms, Daron Weber is shocked to hear his son has PLE. His son's doctor offers to send him a recent article. "*I thank Dr. Hayes, but asked why she never hinted to me before that anything like this could happen. Haven't I always asked for more information about my son?" - p. 54*
- Feeling frustration and possibly guilt that Damon's PLE has gone undiagnosed, his father laments, *"Because no one has checked Damon's albumin for several years, and because Shealagh and I have observed symptoms for an indefinite period, I don't know how long ago the illness started or how much of Damon's time it has already consumed." - p. 60*
- Contemplating a transplant for their son, Damon's parents meet with his transplant doctor to learn more about the procedure. His father finds the doctor can't give him specific statistics off the top of her head and concludes, "*She either doesn't have the facts, or won't discuss them with us."* The doctor tries to reassure him, *"Trust me, it's all good. She said. I begin to worry we are getting a sales pitch. Her language is vague and imparts no real information." - p. 214*
- Damon's parents call and get him listed for a transplant at Columbia three days before Christmas, *"I've phoned to announce our readiness, at long last, to hand over our son into the care of the pediatric cardiac transplant unit at Columbia—but there's no one on the other end to receive the news! Nor is anyone on hand to tell us about next steps or to provide the standard set of instructions." - p. 253.*
- Failing to hear from the transplant team for a week, Damon's father says, *"I become increasingly concerned about the lack of communications. I decide to send a stronger message to Dr. Davis. I title my e-mail 'Who's in Charge of Damon Weber?'" - p. 258*
- Damon is admitted to the hospital, and in spite of the fact that multiple doctors are involved in Damon's care, his father laments, *"No one is taking responsibility for Damon's care." - p. 312*
- Damon has an EBV infection, and develops PTLD. One member of the transplant team attributes his EBV to something he may have picked up from his siblings, parents or a friend. Another tells Doron Weber, *"Oh, of course, his donor was EBV positive!"* Shocked, his father feels betrayed, *"These words tossed out so casually, send me reeling. So she's known this all along?"* He wonders, *"Is it possible Dr. Mason did not communicate such vital information to her colleague?" - p. 324*

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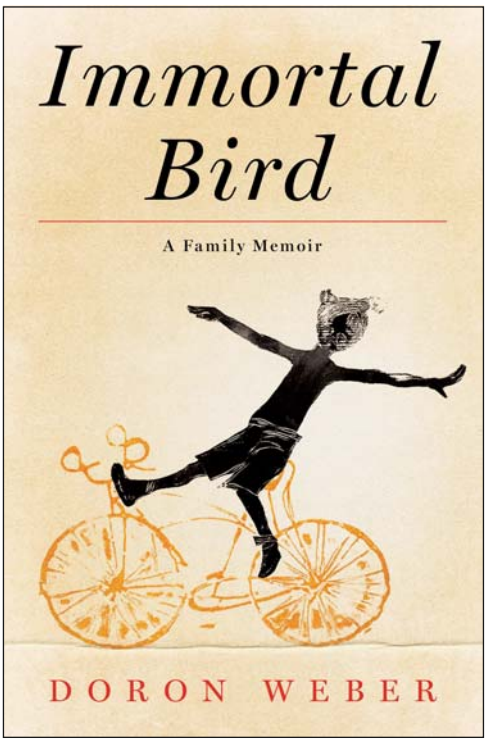
It turned out that the group needed little inspiration. After just a few quotes, multiple hands went up. It seemed that nearly everyone had opinions they wanted to express and to discuss. It was interesting that the case specifics in our discussions turned quickly from Damon Weber and Columbia to our patients and our program at Rady Children's Hospital.

John Lamberti, our Chief Surgeon, summarized the conversations by declaring, "I have dealt with similar intense family situations many times during my career!" And, as for the answer to Marty's question,



Immortal Bird: A Family Memoir by Doron Weber. Published by Simon & Schuster - www.SimonandSchuster.com.

"Yes, the issues raised in the book do ring true." It's too early to tell whether the book will stimulate positive changes in us or our program.

Hallway discussions are continuing….

CCT

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Successful Percutaneous Closure of Aortic Pulmonary Window Residual Leak

By Jorge Gomez, MD; Ruben Vieira MD; Andres Cuenca, MD

Introduction

Aortopulmonary Window (APW) is a communication between the ascending aorta and the main pulmonary artery. It represents only 0.2%–0.6% of all patients with Congenital Heart Disease. This communication is usually nonrestrictive, and is conventionally treated by surgery at an early age to prevent the development of Pulmonary Vascular Obstructive Disease.

There are several reports of percutaneous closure in the past two decades, including restrictive and non-restrictive native defects, or residual leaks.

We report a case of a 38-year-old woman, asymptomatic, with a residual leak of an Aortopulmonary Window treated by surgery, and successfully closed with an Amplatzer Muscular Ventricular Septal Defect (MVSD) device.

Case Report

A 38-year-old woman was referred to our institution with a history of APW, who in 1973, at the age of 8 months, underwent surgery. She had always asymptomatic. After pregnancy, a routine Doppler echocardiogram revealed anomalous flow in ascending aorta.

A continuous murmur of 3/6 was heard at left upper sternal border; she had normal blood pressure, EKG had no significant findings, and chest X-ray showed a slightly increased cardiothoracic ratio. A treadmill test was stopped at 7 Mets due to dyspnoea.

Transesophageal 3D Doppler echocardiogram (3D TEE) study defined a residual leak of the APW, which measured 4 mm near semilunar valves.

Under general anesthesia and 3D TEE guidance, the patient was heparinised for interventional procedure. Qp/Qs calculations reported 1.6, and pulmonary artery pressure was 30/15.

Different angiograms in ascending aorta were performed to properly visualize and measure the defect. At 45° LAO and 2O° cranial angulations (Figure 1), the defect was perfectly assessed and measured 7mm of diameter. The proximity of the left coronary artery (LCA) was a concerning issue. The distance between the inferior rim and the ostium of LCA was 6 mm.

The defect was crossed from the arterial side with a 6 Fr Judkins left catheter (Cordis Corporation, PO Box 025700, Miami, Fl). We advanced a PTCA guidewire 0.014" x 300 cm. (Asahi Intecc C.O, Seto, Aichi Japan), which was snared in the left pulmonary artery to create an arterio-venous circuit. A 5F TorqVue LP delivery catheter (St. Jude Medical, Inc. One St. Jude Medical Drive St. Paul, MN, U.S.A.) was introduced from the venous side, and the tip was left in the descending aorta. We first attempted to close the residual communication using an Amplatzer Duct II Occluder 6/6 device (St. Jude Medical, Inc., One St. Jude Medical Drive, St. Paul, MN, U.S.A.) We were unable to stabilize the device, and it passed through the defect without resistance from the ascending aorta to the main pulmonary artery. Still attached to the cable, the device was retrieved and removed. The same procedure was repeated. An arterious venous loop was created, this time using an Amplatzer Noodle wire 0.035" x 300 cm. A 7 Fr Torq Vue delivery catheter was advanced over the wire from the venous side and deployed an Amplatzer MVSD device of 8 mm occluding the residual leak. Exit angiogram performed in ascending aorta showed an initial passage of contrast through the device and normal flow in LCA (Figure 2). TEE showed some disturbance of pulmonary flow and an estimated gradient of 10 mm Hg (Figures 3-4). No events were registered post-procedure course in hospital for 24 hours. She was treated initially with clopidogrel 75 mg/ 24 hs for 3 months, and then aspirin 100mg per day due to proximity of the device to LCA. At 1 year and 5 months, follow-up patient continues asymptomatic, with no residual leak.

"Aortopulmonary Window (APW) is a communication between the ascending aorta and the main pulmonary artery. It represents only 0.2%–0.6% of all patients with congenital heart disease."

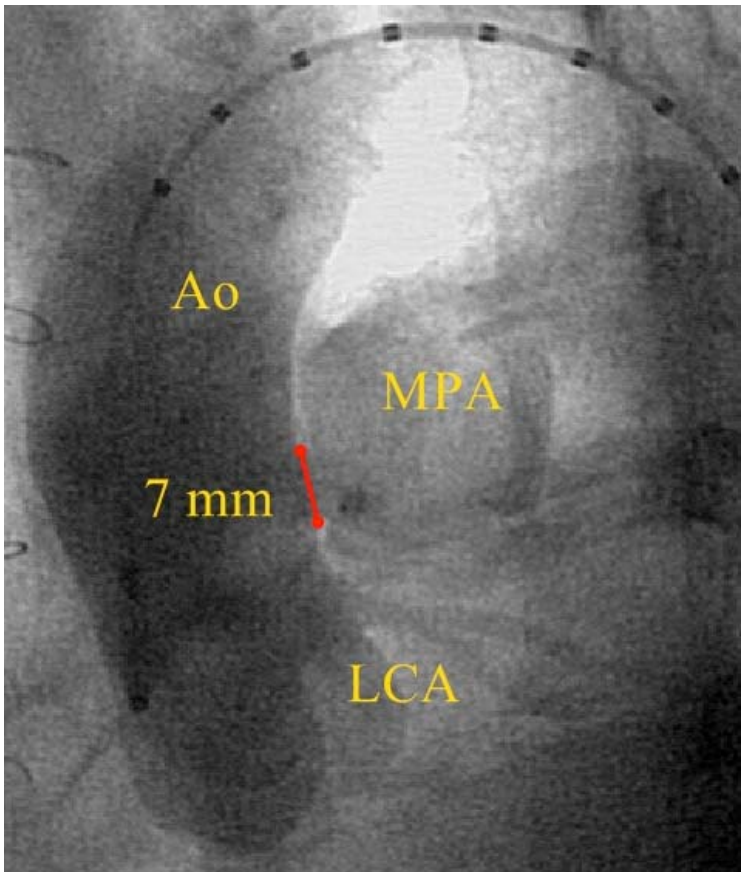

Figure 1. Aortic angiogram at 45° LAO and 20° cranial angulations. Residual defect measured 7 mm. Proximity of left coronary ostium is also visualized (Ao= Aorta, MPA=main pulmonary artery, LCA= left coronary artery).

Adult Congenital Heart Disease (ACHD) Specialist

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Candidates may submit their curriculum vitae to:
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Akron Children's Hospital
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Akron Children's Hospital

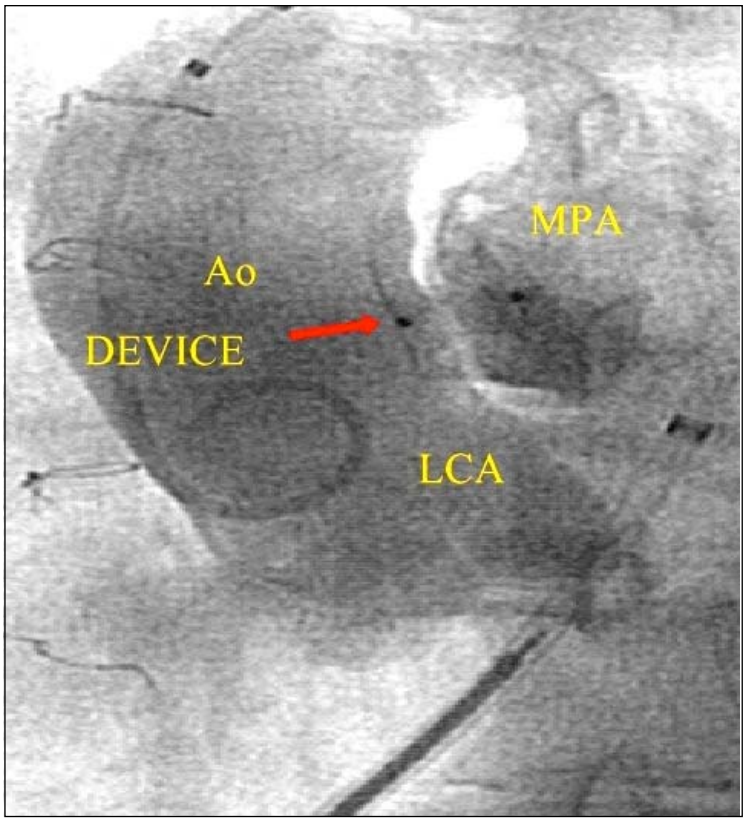

Figure 2. Aortic angiogram after device deployment indicated with a red arrow, some initial contrast is seen passing through the device (Ao= Aorta, MPA=main pulmonary artery, LCA=l left coronary artery).

Discussion

APW, a defect in the aortopulmonary septum, is a rare congenital heart disease occurring in 0.2–0.6% of all patients with congenital heart disease. It is usually nonrestrictive and is conventionally treated surgically at an early age to prevent the development of pulmonary vascular obstructive disease. Several authors had reported different classifications.[1-3] Ho et als divided isolated APW in four types:[1]

- *Proximal* defect is between ascending aorta and main pulmonary artery having little inferior rim separating the APW from semilunar valves.
- *Distal* defect is between origin of right pulmonary artery and ascending aorta, having a well-formed inferior rim, but little superior rim, sometimes associated with anomalous origin of right pulmonary artery.
- *Confluent* defect, a combination of the first and second types with little superior and inferior rims.
- *Intermediate* type, which has adequate superior and inferior rims, usually restrictive and represents 10% of all APW.

Although surgery remains the treatment of choice for large APW, in the past two decades, different reports of percutaneous closure

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A nonprofit organization which seeks to improve the quality of life and extend the lives of congenital heart defect survivors.

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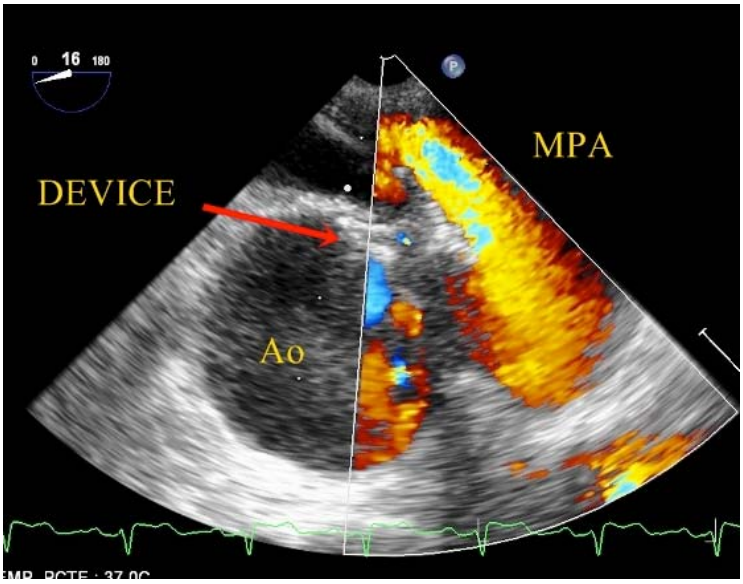

Figure 3. 2D TEE short axis view. Device position is indicated with a red arrow. Little color Doppler aliasing is visualized in the MPA (Ao= Aorta, MPA=main pulmonary artery LCA=left coronary artery).

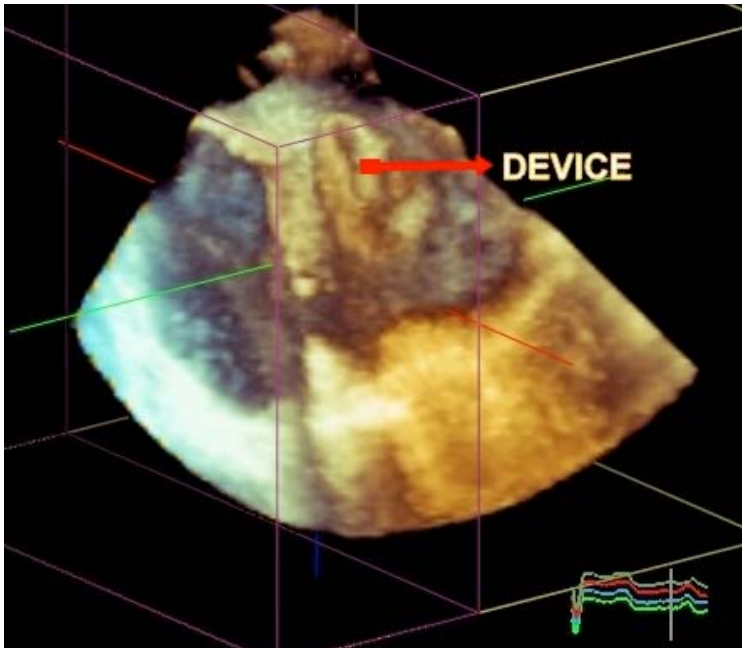

Figure 4. 3D reconstruction of short axis view. Device disc of pulmonary side is indicated with a red arrow and clearly visualized.

exist in the literature and transcatheter-device closure using several devices has been well described.[4-13] Therefore, this procedure should be considered after careful evaluation and selection of case.

"Because of the small sample of patients, statistical data is still unavailable. Percutaneous closure of APW is a feasible and secure procedure. In order to reach high standard results, every individual case has to be carefully evaluated before the procedure. Once performed, patients need an initial close follow-up for the first six months and then annually if no protocols were established."

Previous studies, such as transthoracic and transesophagueal echocardiography, should provide information not only of defect size and shunt direction, but also quality and length of the superior and inferior rims, proximity of other structures such as semilunar valves and coronary arteries have to be carefully examined. MNR or CT scans studies could also be considered.

Different types of devices for percutaneous closure of APW have been used.[4-13] Due to little available data, there are no standard rules for device selection. Low profile devices are preferred to avoid obstruction in ascending aorta or main pulmonary trunk, but these devices have larger discs that could jeopardize left coronary artery flow. In our case, we first selected an Amplatzer Duct Occluder II 6/6 device and failed to stabilize it, probably because it was not suitable for the size of the defect. Amplatzer MVSD of 8 mm was used instead and deployed successfully. The procedure under 3D TEE guidance (Figure 4) aids in providing information about normal function of semilunar valves and flow disturbance across the ascending aorta or main pulmonary artery. The left coronary artery is better evaluated with cineangiogram. Most operators have used venous approach, previously establishing an arterious venous loop. Even though arterial side deployment is possible, several reasons support venous approach delivery, such as: device configuration, delivery catheter size, control angiogram. Also, the possible need to reposition, retrieve, remove or exchange the device.

Conclusion

Because of the small sample of patients, statistical data is still unavailable. Percutaneous closure of APW is a feasible and secure procedure. In order to reach high standard results, every individual case has to be carefully evaluated before the procedure. Once performed, patients need an initial close follow-up for the first six months and then annually if no protocols were established.

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Letters to the Editor

Congenital Cardiology Today welcomes and encourages Letters to the Editor. If you have comments or topics you would like to address, please send an email to: LTE@CCT.bz, and let us know if you would like your comment published or not.

Pediatric Interventional Cardiologist

The Boston Children's Heart Foundation of Boston Children's Hospital and Harvard Medical School is recruiting a pediatric interventional cardiologist to join a large, academic, and innovative practice. Candidates should be at the instructor or assistant professor level, should be board certified in pediatric cardiology, and should have completed advanced training in congenital heart catheterization. This position will focus on clinical activity and will offer the opportunity to lead clinical research projects and train fellows. We are particularly seeking individuals with a track record of an active role in helping develop new devices/ procedures.

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HOW WE OPERATE

The team involved at C.H.I.M.S. is largely a volunteering group of physicians nurses and technicians who are involved in caring for children with congenital heart disease.

The concept is straightforward. We are asking all interested catheter laboratories to register and donate surplus inventory which we will ship to help support CHD mission trips to developing countries.

Rady Children's is pleased to welcome **Eric J. Devaney** as its Director of Cardiac Transplantation. This new program to begin providing transplants to children of San Diego County and beyond this year.

In addition to heart transplantation, Dr. Devaney specializes in congenital heart surgery and circulatory support for neonates through young adults. He comes to Rady Children's from the University of Michigan, where he completed his training in cardiothoracic surgery and pediatric cardiac surgery. Dr. Devaney developed his surgical practice over the last decade at C.S. Mott Children's Hospital at the University of Michigan.

Dr. Devaney is also Associate Professor of Surgery at the University of California, San Diego. He conducts basic research studies in the molecular biology of cardiac muscle function and heart failure, seeking possible therapies for the complications of congenital heart disease.

Providing transplants through Rady Children's Heart Institute will relieve a tremendous burden on families who until now have had to travel long distances for care.

Rady Children's Hospital San Diego

www.rchsd.org

Understanding VAD Management and Documentation Requirements

By Julie-Leah J. Harding, CPC, RMC, PCA, CCP, SCP-ED, CDIS

Recently in my acute care setting discussions of Ventricular Assist Device (VAD) management has been brought forward. VADs assist either the right (RVAD) or left (LVAD) ventricle, or both at once (BiVAD). A particular VADs use depends on the patient's underlying heart disease. VADs are intended for short-term use for patients who have had cardiac surgery, a recent heart attack or a bridge to transplant; long-term use for those patients with chronic congestive heart failure. For professional billing for the insertion or pump replacement, we report the following:

- 33975 Insertion of VAD; extracorporeal, single ventricle.
- 33976 Insertion of ventricular assist device; extracorporeal, biventricular
- 33977 Removal of VAD; extracorporeal, single ventricle.
- 33978 Removal of VAD; extracorporeal, biventricular
- 33979 Insertion of ventricular assist device, implantable intracorporeal, single ventricle.
- 33980 Removal of VAD, implantable intracorporeal, single ventricle
- 33982 Replace VAD Pump, Intracorporeal with Bypass (no global period).
- 33983 Replace VAD Pump, Intracorporeal no Bypass (no global period).

In 2010 a new Current Procedural Terminology (CPT) was introduced to allow us to report daily management:

- CPT 93750 Interrogation of VAD, in person, with physician analysis of device parameters (e.g. drivelines, alarms, power surges), review of device function (e.g. flow and volume status, septum status, recovery), with programming, if performed, and report; no global days.

NOTE: This code is not reported with any of the surgical implantation codes (33975, 33976, 33979, 33981-33983), but is typically reported in conjunction with an evaluation and management visit.

CPT 93750 requires the following documentation, either its own note, or its own paragraph within your daily progress note; the documentation is quite specific:

- Patients with a previously implanted VAD require periodic interrogation of the device. Code 93750 reports a diagnostic procedure that is performed in person, and includes a face-to-face assessment of all device functions, and must be stated in detail either in its own procedure note entry (separate from the rounding note) or in its own paragraph – separate from the daily rounding; if performed by a provider not a part of daily rounding, this entry must be on its own note. Components that must be evaluated include:
  - Device parameters (alarms, drivelines, clots, infection, overall assessment of augmenting cardiac output, and power surges) and,
  - A review of the device function (flow/volume status, septum status, and recovery).

Adjustments may not be needed each day, but each entry must support assessing the potential need. This code includes the physician analysis, review, and report. It also includes device programming, if performed. If the VAD management documentation is too limited, only the evaluation and management CPT should be reported.

Resource: AMA CPT Editorial Panel

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"VADs are intended for short-term use for patients who have had cardiac surgery, a recent heart attack or a bridge to transplant; long-term use for those patients with chronic congestive heart failure. For professional billing for the insertion or pump replacement, we report the following...."

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Archiving Working Group
International Society for Nomenclature of Paediatric and Congenital Heart Disease
ipccc-awg.net

Image of the Month #4 - February 2013 - Presented by The Archiving Working Group

Contributors: Diane E. Spicer, BS; Jeffrey P. Jacobs, MD; Jorge M. Giroud, MD; Robert Anderson, MD; Vera D. Aiello, MD

This is a special column that is published bimonthly in *Congenital Cardiology Today* with contributors and images from the Archiving Working Group (AWG) of *the International Society for Nomenclature of Paediatric and Congenital Heart Disease.*

Please visit us at the AWG Web Portal at http://ipccc-awg.net and help in the efforts of the *Archiving Working Group* and the *International Society for Nomenclature of Paediatric and Congenital Heart Disease*.

The authors would like to acknowledge the Children's Heart Foundation (www.childrensheartfoundation.org) for financial support of the AWG Web Portal.

IPCCC:
01.04.04, 01.03.00, 02.03.01, 02.06.02, 07.02.00, 07.10.00, 01.05.01, 09.29.31

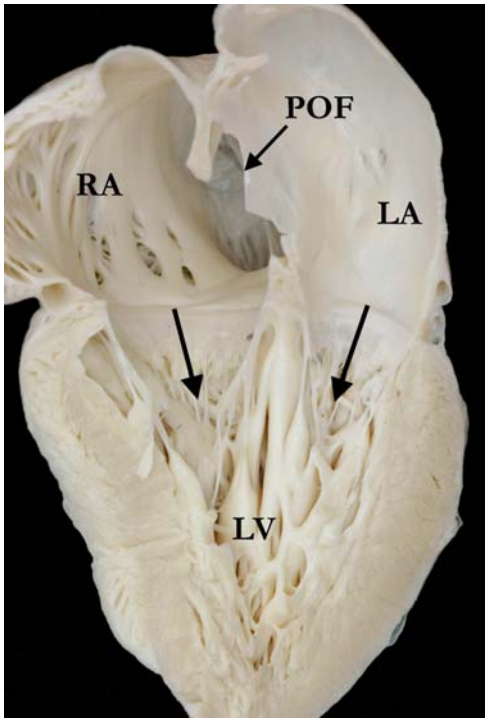
AEPC Derived Term:
Double inlet left ventricle (01.04.04)
Usual atrial arrangement (atrial situs solitus) (01.03.00)
Right hand pattern ventricular topology (D loop) (segmental nomenclature letter 2: 'D') (02.03.01)
Aortic orifice anterior right with respect to pulmonary orifice (02.06.02)
Right ventricular hypoplasia (07.02.00)
Ventricular septal defect (VSD) (07.10.00)
Discordant ventriculo-arterial connections (TGA) (01.05.01)
Interrupted aortic arch (09.29.31)

EACTS-STS Derived Term:
Single ventricle, DILV, {SDD}, Subaortic RV outlet chamber with VSD (Bulboventricular foramen) (01.04.04, 01.03.00, 02.03.01, 02.06.02, 07.02.00, 07.10.00, 01.05.01)
Interrupted aortic arch (IAA) (09.29.31)

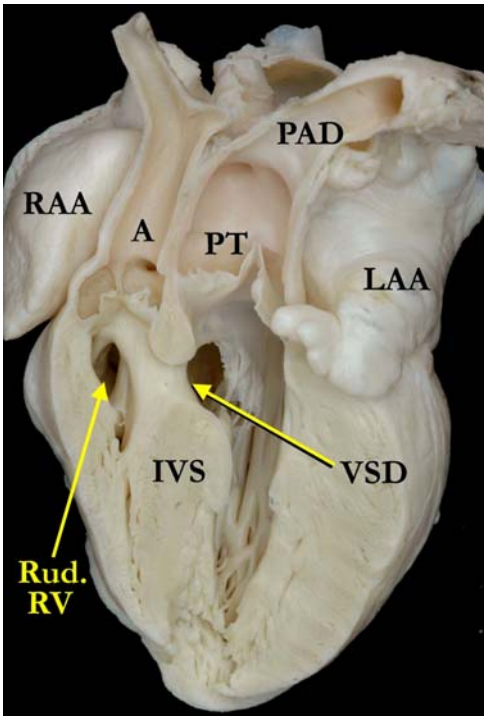
ICD10 Derived Term:
Double inlet ventricle (Q20.4)
Other congenital malformations of cardiac chambers and connections (Q20.8)
Ventricular septal defect (Q21.0)
Discordant ventriculoarterial connection (Q20.3)
Other congenital malformations of aorta (Q25.4)

Commentary: Controversy still attaches to the lesion illustrated in this month's column. There can be no question but that the malformation is well described as showing double inlet left ventricle. It is equally clear from the images that the heart does not have a single ventricle. Tradition has dictated, nonetheless, that the entity was the exemplar of "single ventricle".[1] This problem is one of linguistics rather than anatomy, and is circumvented simply by describing the anomaly as being one example of the functionally univentricular heart.[2] The only reason that the entity can be justifiably described as having a single ventricle is if the smaller chamber is denied ventricular status. Some still take this stance, having argued that the small chamber is no more than an infundibulum.[3] The morphological evidence, however, points to the chamber being an incomplete right ventricle, lacking its inlet component, which self-evidently is committed to the dominant left ventricle.[4] If the small chamber were truly an infundibulum, then the entirety of the septum separating it from the dominant ventricle would perforce be the infundibular septum. As can be seen from the images, the septum interposing between the big and small chambers has two discrete components. It is now well established that the apical of these two components carries the ventricular conduction tissues,[4] and is nourished by septal perforating arteries.[5] It is a true rudimentary ventricular septum. This fact also relates to whether the small chamber should be described as an incomplete as opposed to a rudimentary right ventricle. Both terms are appropriate. The ventricle is incomplete because it lacks its inlet component.[6] It is rudimentary because it

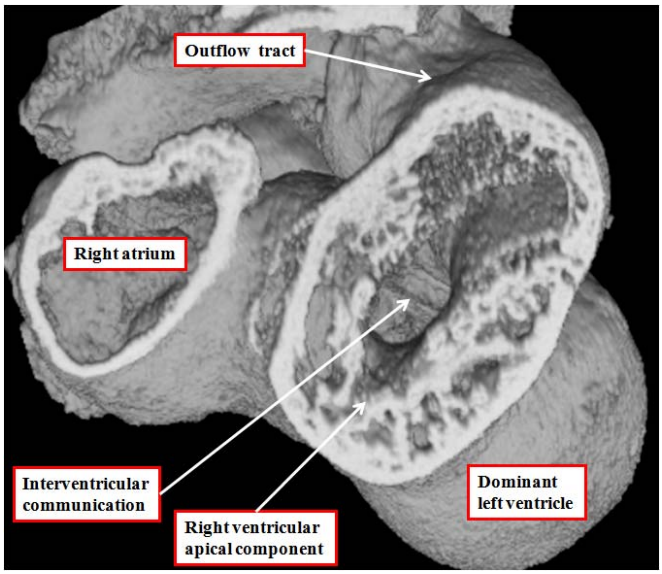
AWG Web Portal link for this series of images:
http://ipccc-awg.net/Single_Ventricle/DILV_Int_AO_Arch_01_04_04/DILV_Int_AO_Arch_01_04_04.html



Description: This heart is cut in the four chamber echocardiographic plane to demonstrate the double inlet atrioventricular connection (arrows). The atrial chambers are both connected to the dominant left ventricle (LV), which is the only ventricular chamber visible. There is an incomplete and rudimentary right ventricle positioned anterosuperiorly, as shown in the companion image. The right ventricle is incomplete because it lacks its inlet component, which is connected to the dominant left ventricle. (PFO-patent oval fossa, RA-right atrium, LA-left atrium). Contributor: Diane E. Spicer, BS.



Description: The heart, shown in the image above, has been cut at right angles to the four chamber section. This cut shows the incomplete and rudimentary right ventricle (RV) lying superiorly and in anterosuperior position. It supports the aorta (A), while the pulmonary trunk (PT) arises from the dominant left ventricle, so that the ventriculoarterial connections are discordant. The aortic arch is interrupted (not well imaged) and the arterial duct (PDA) is widely patent. There is a muscular ventricular septal defect (VSD). (IVS-interventricular septum, RAA-right atrial appendage, LA-left atrial appendage). Contributor: Diane E. Spicer, BS.



Description: The image shows a developing mouse heart at embryonic day 11.5. The heart has been prepared using the technique of high resolution episcopic microscopy, and the dataset sectioned so as to show the equivalent of the echocardiographic oblique subcostal cut. As can be seen, the developing right ventricle already possesses its apical trabecular component, and supports the ventricular outflow tract. At this stage, however, its inlet is through the embryonic interventricular communication, the atrioventricular canal being exclusively supported by the developing left ventricle. The right ventricular chamber at this stage is analogous to the incomplete and rudimentary anterior chamber found when there is double inlet left ventricle. The dataset was prepared by Dr Tim Mohun, Medical Research Council, London, United Kingdom, and the image is reproduced with his kind permission. Contributor: Robert H. Anderson, MD.

unequivocally represents the developing right ventricle, as can be seen by the image from the developing mouse heart, in which, like the incomplete right ventricle found in the setting of double inlet left ventricle, the developing right ventricle has yet to acquire its inlet component, but shows obvious apical and outlet parts.

The developers of the EACTS-STS derived version of the IPCCC agree with these concepts, and further agree that the perceived problem is one of linguistics rather than anatomy. They acknowledge that, in the EACTS-STS derived version of the IPCCC, the term "single ventricle" means "functionally univentricular heart", defined as describing a spectrum of congenital cardiovascular malformations in which the ventricular mass may not readily lend itself to partitioning that commits one ventricular pump to the systemic circulation, and another to the pulmonary circulation.[7] It follows that a patient may have a heart deemed to be functionally univentricular either because of its anatomy, or because of the lack of feasibility or lack of advisability of surgically partitioning the ventricular mass. Common lesions falling into the category include: double inlet right ventricle, double inlet left ventricle, tricuspid atresia, mitral atresia, and Hypoplastic Left Heart Syndrome. Other lesions which sometimes may be considered to be a functionally univentricular heart include: complex forms of atrioventricular septal defect, double outlet right ventricle, congenitally corrected transposition, pulmonary atresia with intact ventricular septum, and other complex cardiovascular malformations. Whenever possible, however, the lesions should be catalogued using specific diagnostic codes, and not the term "functionally univentricular heart." (Source: The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD) [www.ipccc.net].

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Medical News, Products and Information

Digisonics Named Best in KLAS for Fifth Consecutive Year

Digisonics has received the Best in KLAS Award in the Cardiology market segment for the fifth consecutive year, an unparalleled distinction.

KLAS, an independent healthcare research firm, conducts interviews with healthcare executives, managers and clinicians from thousands of facilities to monitor and evaluate the performance of healthcare IT software and services vendors. The annual Best in KLAS Awards Report names the best vendors in each category based on customer ratings. Vendor scores are compiled from customer evaluations in categories including implementation and training, functionality and upgrades, sales and contracting, and service and support.

Digisonics has again maintained the highest overall score across all vendors in the Cardiology market segment of the 2012 Best in KLAS Awards: Software & Services Report.

"Expectations for a top Cardiology system are daunting. Our users expect a combination of superior interfaces for smooth workflow between disparate systems together with structured, clinical reporting, administrative reporting, and image management features matched to their site. Digisonics is exceptionally pleased to be recognized again as the best vendor in this challenging CVIS market," said Digisonics President and CEO, Dr. Diana McSherry. "To have received this award for five straight years is a testament to Digisonics' goals of smart solutions, close attention to customer needs, and nimble responses."

The DigiView Image Management and Structured Reporting System, ranked Best in KLAS in the 2008-2012 Top 20 Best in KLAS Awards: Software & Professional Services reports for the Cardiology market segment, combines high performance image review workstations, a powerful PACS image archive, an integrated clinical database, comprehensive measurements and calculations package, and highly configurable reporting for all cardiovascular modalities. The DigiNet Pro add-on option provides users with fully functional web-based access to their cardiovascular studies from anywhere at any time. For more information, please contact: www.digisonics.com

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THE CONGENITAL HEART INTERVENTION MISSION SUPPORT

HOW WE OPERATE

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The concept is straightforward. We are asking all interested catheter laboratories to register and donate surplus inventory which we will ship to help support CHD mission trips to developing countries.

Ordinary Heart Cells Become "Biological Pacemakers" with Injection of a Single Gene

Newswise — Cedars-Sinai Heart Institute researchers have reprogrammed ordinary heart cells to become exact replicas of highly specialized pacemaker cells by injecting a single gene (Tbx18)–a major step forward in the decade-long search for a biological therapy to correct erratic and failing heartbeats.

The advance was published in the Jan 8th issue of *Nature Biotechnology,* and is available on the journal's website www.nature.com.

"Although we and others have created primitive biological pacemakers before, this study is the first to show that a single gene can direct the conversion of heart muscle cells to genuine pacemaker cells. The new cells generated electrical impulses spontaneously and were indistinguishable from native pacemaker cells," said Hee Cheol Cho, PhD, a Heart Institute research scientist.

Pacemaker cells generate electrical activity that spreads to other heart cells in an orderly pattern to create rhythmic muscle contractions. If these cells go awry, the heart pumps erratically at best; patients healthy enough to undergo surgery often look to an electronic pacemaker as the only option for survival.

The heartbeat originates in the Sinoatrial Node (SAN) of the heart's right upper chamber, where pacemaker cells are clustered. Of the heart's 10 billion cells, fewer than 10,000 are pacemaker cells, often referred to as SAN cells. Once reprogrammed by the Tbx18 gene, the newly created pacemaker cells – "induced SAN cells" or iSAN cells – had all key features of native pacemakers and maintained their SAN-like characteristics even after the effects of the Tbx18 gene had faded.

But the Cedars-Sinai researchers, employing a virus engineered to carry a single gene (Tbx18) that plays a key role in embryonic pacemaker cell development, directly reprogrammed heart muscle cells (cardiomyocytes) to specialized pacemaker cells. The new cells took on the distinctive features and function of native pacemaker cells, both in lab cell reprogramming and in guinea pig studies.

Previous efforts to generate new pacemaker cells resulted in heart muscle cells that could beat on their own. Still, the modified cells were closer to ordinary muscle cells than to pacemaker cells. Other approaches employed embryonic stem cells to derive pacemaker cells. But, the risk of contaminating cancerous cells is a persistent hurdle to realizing a therapeutic potential with the embryonic stem cell-based approach. The new work, with astonishing simplicity, creates pacemaker cells that closely resemble the native ones free from the risk of cancer.

For his work on biological pacemaker technology, Dr. Cho, the article's last author, recently won the Louis N. and Arnold M. Katz Basic Research Prize, a prestigious Young Investigator Award of the American Heart Association.

Children's Hospital of Pittsburgh | of UPMC

Opening for a 2nd or 3rd year Pediatric Cardiology Fellow for July 2013

Children's Hospital of Pittsburgh of UPMC

The Division of Pediatric Cardiology at the Heart Institute at Children's Hospital of Pittsburgh of UPMC is expanding its fellowship educational opportunities. We are increasing the number of permanent ACGME pediatric cardiology fellowship positions from seven to eight. In addition to recruiting three candidates to begin 7/1/2014 as first year fellows, we are making available a general pediatric cardiology position for a qualified, current 1st or 2nd year fellow interested in transferring from a current ACGME pediatric cardiology fellowship program into our program starting 7/1/2013.

The Heart Institute provides comprehensive pediatric and adult congenital cardiovascular services to the tri-state region. The group consists of 18 pediatric cardiologists, four pediatric cardiothoracic surgeons, five pediatric cardiovascular intensivists, 12 physician extenders and a staff of over 100.

Children's Hospital of Pittsburgh of UPMC has been named to *U.S. News and World Report's* 2012-2013 Honor Roll of Best Children's Hospitals, one of only twelve hospitals nationally to earn this distinction. Consistently voted one of America's most livable cities, Pittsburgh is a great place for young adults and families alike.

Please submit CV, personal statement, and three letters of reference to Dr. DeBrunner. Inquiries may be made by contacting **Lynda Cocco** at **412-692-3216**, or **lynda.cocco2@chp.edu**.

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Pediatric Cardiologist

The Department of Pediatrics of the University of Texas Health Science Center at San Antonio is seeking a pediatric cardiologist to join the UT Children's Heart Network. UTHSCSA, in an exciting partnership with Vanguard Health Systems and the Children's Hospital of Philadelphia, is entering an exciting period of expansion. Plans are underway for a new, free-standing Children's Hospital in the South Texas Medical Center. The new Children's Hospital will anchor a pediatric healthcare delivery network that promises to transform children's healthcare in San Antonio and throughout South Texas.

The candidate must be fellowship trained, board certified/board eligible in pediatric cardiology, and either possess or be able to easily obtain an unrestricted Texas medical license. The candidate will join an established academic clinical practice with 6 pediatric cardiologists and 3 congenital heart surgeons. The new Children's Hospital will be a 225 bed facility providing services across the spectrum of needs for all of the children of South Texas. The School of Medicine has 230 medical students at each level. Cardiology faculty are engaged in the training of these medical students and 49 pediatric residents. Candidates with interests in general cardiology, fetal imaging, echocardiography, advanced cardiac imaging, and pediatric heart failure are encouraged to apply.

Please submit a letter of interest, curriculum vitae, and three letters of recommendation to:

Steven R. Neish, M.D., S.M.., Professor &
Vice Chairman, Department of Pediatrics,
Chief, Pediatric Cardiology,
The University of Texas Health Science Center
at San Antonio,
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"This is the culmination of 10 years of work in our laboratory to build a biological pacemaker as an alternative to electronic pacing devices," said Eduardo Marbán, MD, PhD, Director of the Cedars-Sinai Heart Institute and Mark S. Siegel Family Professor, a pioneer in cardiac stem cell research. A clinical trial of Marbán's stem cell therapy for heart attack patients recently found the experimental treatment helped damaged hearts regrow healthy muscle.

If subsequent research confirms and supports findings of the pacemaker cell studies, the researchers said they believe therapy might be administered by injecting Tbx18 into a patient's heart or by creating pacemaker cells in the laboratory and transplanting them into the heart. But additional studies of safety and effectiveness must be conducted before human clinical trials could begin.

The study was supported by the Cedars-Sinai Board of Governors Heart Stem Cell Center, the Heart Rhythm Society, the Heart and Stroke Foundation of Canada, the American Heart Association (12SDG9020030), the National Heart, Lung, and Blood Institute (1R01HL111646-01A1), and the Mark S. Siegel Family Professorship. The authors report that they have no conflicts of interest.

Citation: Nature Biotechnology, "Transcription factor-driven conversion of quiescent cardiomyocytes to pacemaker cells," online Dec. 16, 2012; print publication in issue dated Jan. 8, 2013.

Children with Heart Devices and Their Parents Struggle with Quality of Life

Children with implanted heart-rhythm devices and their parents suffer from a lower quality of life compared with their healthy counterparts and may benefit from psychotherapy according to new research in *Circulation: Arrhythmia & Electrophysiology*, an American Heart Association journal.

Researchers at the Cincinnati Children's Hospital Medical Center studied 173 children with either a pacemaker (40 patients) or implanted defibrillator (133 patients) to assess their quality of life compared to other children with Congenital Heart Disease and to healthy children. The children, ages 8 to 18 years old, and their parents completed quality of life questionnaires.

Compared with healthy children and their parents, children with heart devices and their parents reported significantly lower quality of life scores. Likewise, their scores were also lower than those of children with mild congenital heart disease. However, their quality of life scores were similar to those for children with more severe heart disease but no device.

For children, self-perception, self-worth, and athletic capability affected quality of life. For parents, their child's behavior was the biggest factor related to quality of life. Also, children with an implantable defibrillator tended to have lower quality of life scores than those with pacemakers.

"These findings should encourage us to consider the negative impact of devices, particularly defibrillators, on pediatric patients; and to develop strategies to mitigate these effects," said Richard J. Czosek, MD, study author and Assistant Professor of Pediatrics at the Cincinnati Children's Hospital Medical Center, Heart Institute in Ohio. "Whether these effects on quality of life can be reduced through the use of psychotherapy needs to be assessed."

Co-authors are: William J. Bonney, MD; Amy Cassedy PhD; Douglas Y. Mah, MD; Ronn E. Tanel, MD; Jason R. Imundo, MD; Anoop K. Singh, MD; Mitchell I. Cohen, MD, Christina Y. Miyake, MD; Kara Fawley, BS; and Bradley S. Marino, MD.

Author disclosures and sources of funding are on the manuscript.

Cartoons Reduce Anxiety in Children Undergoing Anesthesia

Newswise – Letting children watch a favorite cartoon is an effective and safe way to reduce anxiety before anesthesia and surgery, concludes a study in the November issue of Anesthesia & Analgesia, official journal of the International Anesthesia Research Society (IARS).

"Cartoon distraction" is an "inexpensive, easy to administer, and comprehensive"

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technique for reducing anxiety in young children before induction of anesthesia, according to the new research, led by Dr Joengwoo Lee of Chonbuk National University Hospital, South Korea.

The study evaluated the use of cartoons to reduce anxiety in 130 children, aged three to seven, undergoing routine surgical procedures—most commonly tonsillectomy. In a holding area, one group of patients were allowed to choose an animated movie to watch before induction of anesthesia. The children watched the movie on a tablet or laptop computer; a "Power Rangers" cartoon was the most popular choice.

Another group of children were asked to bring a favorite toy, which they were allowed to play with before induction. A third group received no special treatment. Measures of anxiety—as rated by the parents and judged by the child's behavior—were compared among groups.
In the holding area, anxiety scores were lower for the children who played with a favorite toy. However, after the children were moved to the operating room, anxiety was lowest for the children who watched cartoons.

According to both parent ratings and behavioral measures, only a few children in the cartoon group had increased anxiety once they went into the operating room. In contrast, nearly all of the children in the other groups had higher anxiety scores in the operating room. Anxiety was rated low or absent for 43% of children who watched cartoons, compared to 23% of those who brought a toy and 7% with neither treatment.

Many techniques have been tried to reduce preoperative anxiety in children, with inconsistent results. Treatment with a sedative (midazolam) is probably the most common approach, but this drug has the potential for side effects.

Watching cartoons might provide a simple way of alleviating anxiety before anesthesia by distracting the children. They write, "Preschool children generally enjoy watching animated cartoons , and they can become sufficiently engrossed to their surroundings and disregard verbal and tactile stimuli." Playing with a familiar toy may be comforting as well.

The results suggest that letting children watch cartoons "is a very effective method to alleviate preoperative anxiety," according to Dr. Lee and colleagues. By providing children with a distraction during preparations for anesthesia and surgery, cartoons are an "inexpensive, easy to administer, and comprehensive method for anxiety reduction."

It may seem like a small matter to reduce anxiety by showing children cartoons. But anxiety before surgery can be a significant problem, causing emotional trauma for both the parents and children. In some cases, preoperative anxiety can lead to lasting behavioral problems, such as separation anxiety, aggressiveness, and nightmares.

The study confirms what many parents already know, according to an editorial by Drs Franklyn P. Cladis and Peter J. Davis of University of Pittsburgh: "Trying to interact with our children when "Cars" or "SpongeBob" is on television is futile." Distracting children by letting them "tune into their favorite alternative realities" seems to lower anxiety responses to surgery. Drs Cladis and Davis note that more research would be needed to determine whether reduced anxiety at induction leads to fewer behavioral problems after surgery.

"We need to take a hard look at the cost, access, quality and safety issues related to present government and private insurer medical imaging policies and find ways to maximize the value, role and efficiency of radiology as health care systems evolve. The Neiman Institute will provide much needed information to ensure that future imaging policies benefit patients and make efficient, effective use of health care resources," said Duszak.

For more information about The Harvey L. Neiman Health Policy Institute, please visit neimanhpi.org.

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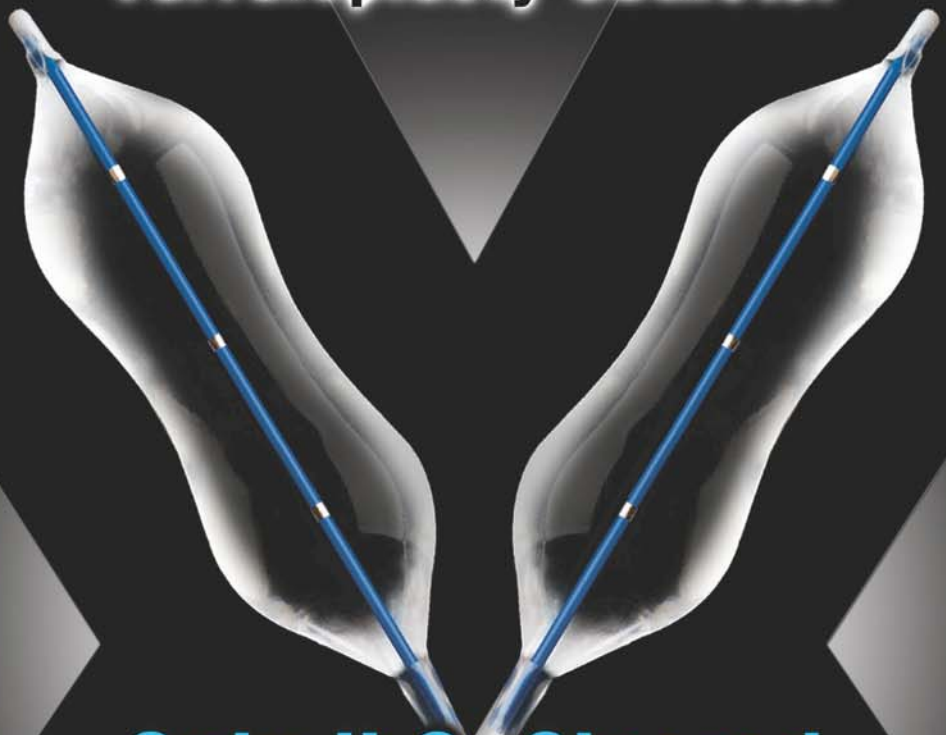

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