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FDA Clearance of Cardiac Devices for Children: A Primer and Call to Action

By Robert H. Beekman, III, MD

Interventional pediatric cardiology and cardiac surgery are highly technical specialties that are dependent upon the availability of appropriate medical devices. However, because of the relative infrequency and diversity of cardiac disorders in children (relative to adult cardiovascular disease), very few devices have been designed and approved explicitly for pediatric cardiovascular applications.[1]

Recent events have elevated the level of discourse concerning FDA device clearance processes. Using the Humanitarian Device Exemption (HDE) process, the FDA cleared the Melody Transcatheter Pulmonary Valve (Medtronic) in 2010, and the EXCOR VAD (Berlin Heart) in late 2011. Nevertheless, some cardiologists and third-party payers persist in believing that these two devices are still experimental. Even more recently, concern has arisen around episodes of cardiac perforation in patients treated with the Amplatzer Septal Occluder (St. Jude) – a device cleared in 2001 when the FDA approved its Pre-Market Approval (PMA) application without a randomized clinical trial. If pediatric cardiologists and cardiac surgeons are to be effective advocates for children in need of improved access better cardiac devices, it is imperative that they understand the processes used by the FDA to clear devices for marketing.

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FDA Clearance of Medical Devices

The FDA Center for Devices and Radiological Health (CDRH) is responsible for assuring the safety and effectiveness of medical devices in the United States. The FDA clears a device for marketing and shipment across state lines (the FDA "clears" rather than "approves" medical devices). In general, FDA clearance is dependent upon a manufacturer providing sufficient data to support a claim of safety and effectiveness for an intended application of a given medical device.[2]

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- Obstruction of the central veins;
- Clinical or biological signs of infection;
- Active endocarditis;
- Known allergy to aspirin or heparin;
- Pregnancy.

Potential Complications / Adverse Events: Potential procedural complications that may result from implantation of the Melody device include the following: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, pain at the catheterization site.

Potential device-related adverse events that may occur following device implantation include the following: stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, hemolysis.

For additional information, please refer to the Instructions For Use provided with the product.

The Melody® Transcatheter Pulmonary Valve and Ensemble® Transcatheter Delivery System has received CE Mark approval and is available for distribution in Europe. Additionally, a Medical Device Licence has been granted and the system is available for distribution in Canada.

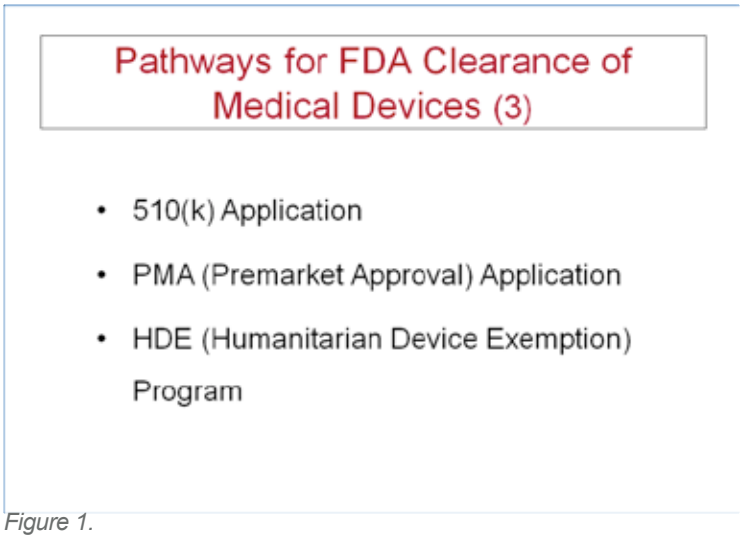

Figure 1.

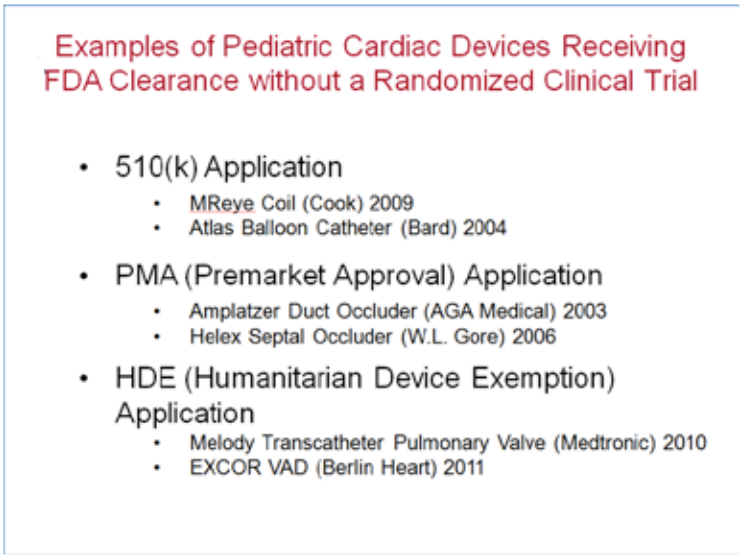

Figure 2.

Currently there are three pathways for FDA clearance of medical devices (Figure 1):

- The 510 (K) application;
- The PMA (pre-market approval) application; and
- The HDE (humanitarian device exemption) application.

Each of these will be discussed in some detail below.

A. 510 (K) Application

The 510 (K) application provides the least-burdensome pathway to FDA clearance for manufacturers of medical devices. This pathway is used for lower risk (generally Class II) devices such as guidewires, coils, and infusion catheters. The 510 (K) application requires that a new device be "substantially equivalent" in intended use and device performance to a "predicate device" that is already on the market. The 510 (K) application typically does not require a manufacturer to submit clinical trial data in support of the application.

Examples of devices cleared by the 510 (K) mechanism include the MReye coil cleared in 2009 (Cook), the Atlas balloon dilation catheter cleared in 2004 (BARD) and the Gianturco-Grifka vascular occlusion device cleared in 1994 (Cook) (Figure 2).

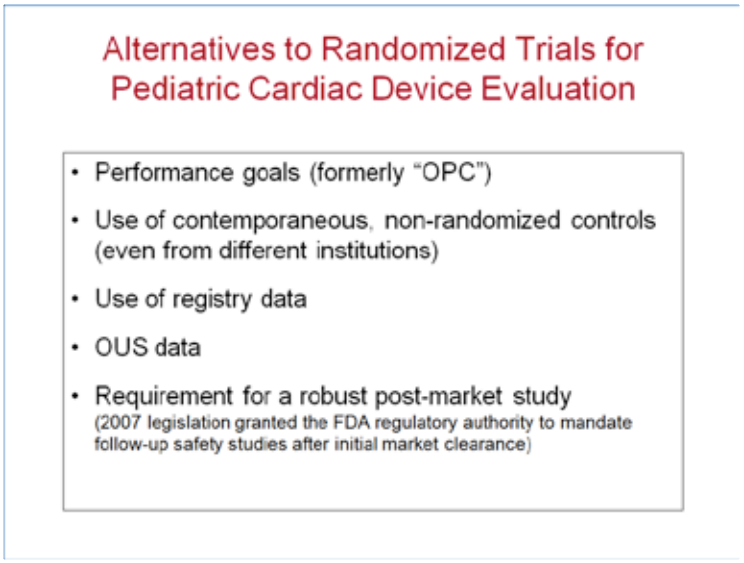

Figure 3.

B. PMA (Pre-Market Approval) Application

A PMA application is the most common mechanism used by industry to obtain FDA clearance for marketing of a new medical device. The PMA pathway is used for higher risk (Class III) devices such as septal occluders, vascular occluders, coronary artery stents and implantable defibrillators. Of the three FDA pathways (Figure 1), a PMA application requires the most rigorous data demonstrating device safety and effectiveness. A manufacturer typically is required to submit bench, animal and clinical trial data in support of a PMA application. Clinical data are obtained under an FDA-approved Investigational Device Exemption (IDE) protocol. Classically, these protocols have been randomized clinical trials, considered the gold standard for generating safety and effectiveness data in support of a PMA application.

However, *randomized clinical trials are exceedingly difficult to accomplish in pediatric cardiology and cardiac surgery*. In fact, it is exceptional to find any randomized trials of pediatric cardiac devices in the literature.[3] The challenges to successful randomized trials in pediatric cardiology and cardiac surgery are well-known. Congenital cardiovascular malformations are uncommon and exceedingly diverse in nature. Affected patients vary widely in size, often are expected to grow substantially after a device is implanted, and have life expectancies measured in decades (in contrast to the shorter life expectancies of many adult cardiac patients). Thus, it is exceedingly difficult to design adequately-powered randomized clinical trials for relevant pediatric cardiac devices. Moreover, the relative small numbers of pediatric cardiac patients limit market potential for industry and provide only small incentives for investment in research and development of devices targeted for this population.

Importantly, alternatives to the randomized clinical trial (RCT) design have been developed and used successfully for evaluation of pediatric cardiac devices (Figure 3). Performance Goals (PGs; formerly known as "objective performance criteria" or OPC's) are defined measures of device safety and effectiveness that are obtained from the literature and expert opinion. Performance Goals can be used to judge the performance of an investigational device, and serve as an alternative to data obtained from a randomized control group. Another alternative to the RCT is the use of contemporaneous but non-randomized control subjects (even from different institutions) to compare to subjects receiving the investigational device. Other alternatives to the RCT include the use of existing registry data, or data obtained from studies outside of the United States. Finally, pediatric device clearance can be

tied to a requirement that the manufacturer perform a robust post-approval study to evaluate longer-term device safety and effectiveness. The 2007 Pediatric Medical Device Improvement and Safety Act (PUB L N0. 110-85) granted authority to the FDA to mandate follow-up safety studies after initial market clearance.

There are several examples in the last decade of pediatric cardiac devices receiving PMA clearance from the FDA without a RCT. For example, the Amplatzer Duct Occluder (AGA Medical) received FDA PMA approval on May 14, 2003. The pivotal clinical study relied upon formal Performance Goals rather than control data from a RCT. An advisory panel to the FDA recommended the use of the following Performance Goals:[4]

- > 85% complete PDA closure at 12 months;
- > 95% clinical closure at 12 months; and
- major adverse event rate < 6%.

The FDA also required a five year post-approval study of the Amplatzer Duct Occluder.

The Helex Septal Occluder (W.L. Gore) provides another example of a pediatric cardiac device receiving PMA clearance without a RCT. The pivotal clinical study used historical and contemporaneous surgical control data obtained from non-randomized cases at the same institutions that implanted the investigational device.[5] The pivotal study demonstrated device non-inferiority in a composite clinical outcome measure at 12 months. The Helix device was cleared by the FDA with PMA approval on August 11, 2006. The FDA also required a five year post-approval study be performed.

C. HDE (Humanitarian Device Exemption) Application

An HDE application provides a pathway to FDA clearance for devices used to treat "rare" disorders that affect fewer than 4,000 patients in the United States annually. This clearance process was developed in recognition of the difficulty of performing adequately-powered trials in small patient populations, with uncommon disorders and limited treatment options. Clinical trials of patients with such "rare" disorders are unable to generate safety and effectiveness data as robust as trials performed in patients with more common conditions. Consequently, HDE clearance by the FDA is based on evidence demonstrating:

1. Safety, and
2. Probable Benefit.

Because of the small number of patients available to participate in these trials, an HDE is exempt from showing "proof of effectiveness" as is required for a PMA application.

An approved HDE application authorizes the manufacturer to market the device. Furthermore, the 2007 Pediatric Medical Device Improvement and Safety Act permits the manufacturer to earn a profit for HDE-cleared devices for children. Because an approved HDE is based on less robust data than a PMA, there is a requirement that each institution's IRB approve the use of the device. Nevertheless, this does not imply that an HDE-cleared device is investigational.

Recently, two important cardiac devices for pediatric use have been cleared by the FDA using the HDE process (Figure 2). The Melody Transcatheter Pulmonary Valve (Medtronic) was cleared by an HDE application approved by the FDA on January 25, 2010. The pivotal clinical data submitted in support of the Melody HDE application was a prospective, non-randomized evaluation of 99 subjects which assessed procedural success, safety and six-month effectiveness without a control group. The FDA also considered data submitted from outside the United States on a 68 patient cohort submitted by Dr. Philipp Bonhoeffer. The FDA approval of the Melody HDE application also included a requirement for two post-approval studies evaluating the device to a five-year endpoint. The first study included the patients from the initial IDE study, and the second was based on enrollment of 100 new patients also followed for five years.

The EXCOR Pediatric VAD (Berlin Heart) was cleared for marketing by the FDA when its HDE application was approved on December 16th, 2011. The EXCOR Pediatric VAD was evaluated in a prospective pivotal trial which was a multicenter study of 48 children at imminent risk of death from heart failure and who were listed for transplantation. Historical control data were obtained from the existing ELSO Registry of patients supported on ECMO between 2000 and 2007, and matched for age, weight and primary diagnosis with the EXCOR-supported patients. The FDA considered clinical data on 136 children supported on EXCOR at non-study sites in the United States, under a compassionate use protocol. The FDA also reviewed data on more than 100 patients from outside the United States. Finally, as part of the HDE approval, the FDA required that Berlin Heart perform a post-approval study to evaluate whether EXCOR safety and outcomes in the commercial setting are comparable to those reported in the pivotal study.

Next Steps: A Call To Action

The recent FDA clearance of the Medtronic Melody Valve and the Berlin Heart EXCOR VAD are important achievements for the health and welfare of children with serious cardiovascular disease. Neither device could be studied in a population large enough to support a robust RCT to support a PMA application, and therefore both devices were cleared for marketing by the FDA using the HDE pathway. *Nevertheless, important obstacles exist that block access to HDE-cleared devices for some children who need them*. These fundamentally derive from the nature of the HDE approval process itself, and specifically from:

1. the exemption from "proof of effectiveness" and
2. the IRB requirement -- both of which were incorporated into the HDE process by the FDA because of the difficulty studying small patient populations with uncommon disorders.

There appears to be poor understanding of the HDE process by some individual IRBs and third-party payers. For example, I recently obtained a confidential draft of a "policy statement" from one of the largest health insurance companies in the United States. This draft was dated July 14, 2011, a full 18-months after the FDA granted clearance to Medtronic to market the Melody Valve. In this "policy statement," the company reviewed existing data and noted that published literature consists of relatively small case series with evaluation of short-term outcomes only. It concluded, therefore, the device remains investigational. Importantly, the "policy statement" noted that there were no available practice guidelines or formal position statements from professional organizations to draw upon. Patients insured through this company, and there are many, currently are denied coverage for implantation of the Melody Valve.

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Figure 4.

These observations, particularly that there are no existing practice guidelines or position statements from medical societies to guide third party payers, strongly suggests some next steps to help increase access to current HDE-cleared devices (Figure 4). Pediatric cardiologists, cardiac surgeons and affiliated organizations should develop policy statements and practice guidelines to help educate third party payers of the following facts:

- HDE is the only FDA mechanism for clearance of devices used for uncommon disorders (< 4,000 patients per year in the United States);
- An HDE-cleared device is approved for marketing, and the manufacturer can earn a profit on such devices for children;
- An HDE-cleared device is not investigational.

Since the 2007 Pediatric Medical Device Improvement and Safety Act became law, the FDA has come a long way in its understanding of the unique challenges posed to device approval by pediatric cardiology and cardiac surgery. The relatively rare and diverse cardiac conditions that affect children are not amenable to evaluation in large randomized trials. Fortunately, the FDA recognizes these challenges and offers the HDE pathway as a mechanism to clear such devices for marketing. However, an apparent lack of understanding by third-party payers of the HDE clearance process has led to denial of coverage to families whose children stand to benefit from these devices. This situation creates a call to action for pediatric cardiologists and cardiac surgeons to lead the way in educating the public and third-party payers that HDE-cleared devices are truly approved and should be widely available to the children who need them.

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Robert H. Beekman, III, MD
The Heart Institute of the
Cincinnati Children's Hospital Medical Center
3333 Burnet Ave.
Cincinnati, OH 45229-3026 USA
Phone: 513-636-7072
Fax: 513-636-2410

Robert.Beekman@cchmc.org

The Archiving Working Group of the International Society for Nomenclature of Paediatric and Congenital Heart Disease: A Visual Encyclopedia Illustrating the Terms and Definitions of the International Paediatric and Congenital Cardiac Code

By Jorge M Giroud, MD; Vera D. Aiello, MD; Diane E. Spicer, BS; Robert H. Anderson, BSc, MD, FRCPath

"Classifications are theories about the basis of natural order, not dull catalogues compiled only to avoid chaos." ~ Stephen Jay Gould (1989)[1]

Introduction

The urge to name and classify is part of the scientific legacy of human history. Cardiac malformations have been described throughout the centuries, with descriptions of various cardiac malformations such as ectopia cordis, septal defects, single ventricle and others, many written as case reports.[2] In 1858, Peacock published 'On Malformations of the Human Heart', in which he grouped the various forms of cardiac defects based on a combined anatomical and embryologic classification.[3] Advances made during the late 19th, and early 20th, century reflected the increasing interest in congenital heart disease, with the descriptions of malformations by authors such as Fallot, Ebstein and Eisenmenger.[4] In 1936, following the advice of Osler to treat the subject "statistically," Abbott published the 'Atlas of Congenital Cardiac Disease,' in which she offered a list of over a thousand congenital cardiac lesions.[5]

Throughout much of the 20th century, nonetheless, the classification of pediatric cardiac disorders remained fragmented, inconsistent, and in many circumstances based on historical considerations and eponyms. It was not always constructed from logic, nor the best available scientific evidence. As these limitations became apparent, groups of experts in the field of pediatric cardiology, cardiac surgery, morphology, and pathology offered two systems of nomenclature. These were published almost simultaneously in 2000.

One system was the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of the European Association for Cardio-Thoracic Surgery (EACTS) and the Society of Thoracic Surgeons (STS). The second system of nomenclature was The European Paediatric Cardiac Code, produced on behalf of the Association for European Paediatric Cardiology (AEPC). It was quickly recognized that these systems, although slightly different in their main structure, were complementary. Thus, in October 2000, the International Nomenclature Committee for Paediatric and Congenital Heart Disease was created to reconcile these systems of nomenclatures. Fortunately, both of these systems were comprised of short and long lists. The initial result was the creation of a bidirectional cross-map of the short lists, which was presented at the First International Summit on Nomenclature for Congenital Heart Disease, held at the *Third World Congress of Paediatric Cardiology and Cardiac Surgery* in Toronto, Canada in May, 2001.

Four years later, the cross-map expanded to include the long lists. Now named the International Paediatric and Congenital Cardiac Code (IPCCC), it was presented at the *Second International Summit on Nomenclature for Pediatric and Congenital Heart Disease*, held at the *Fourth World Congress of Pediatric Cardiology and Cardiac Surgery* in Argentina, in September, 2005.[6] The most recent version of the IPCCC was presented at the *Third International Summit on Nomenclature for Pediatric and Congenital Heart Disease*, held at the *Fifth World Congress of Paediatric Cardiology and Cardiac Surgery* in Australia, in June, 2009.[7]

"Throughout much of the 20th century, nonetheless, the classification of pediatric cardiac disorders remained fragmented, inconsistent, and in many circumstances based on historical considerations and eponyms. It was not always constructed from logic, nor the best available scientific evidence."

The IPCCC is the responsibility of The International Society for Nomenclature of Paediatric and Congenital Heart Disease (ISNPCHD), which evolved from The International Nomenclature Committee for Paediatric and Congenital Heart Disease, and was incorporated in Canada in January, 2005.[6,7] The goal of the ISNPCHD is "to standardize and maintain an international nomenclature system to enhance global communication and facilitate patient care, teaching, and research into paediatric and congenital heart disease across disciplines."[8]

The ISNPCHD is composed of three working groups.[6,7,9] The oldest of the working groups is the International Working Group for Mapping and Coding of Nomenclatures for Paediatric and Congenital Heart Disease, also known as the Nomenclature Working Group (NWG). Currently, the NWG has cross-mapped the nomenclature of the International Congenital Heart Surgery Nomenclature and Database Project of EACTS and STS with the EPCC of AEPC. In addition, cross maps have been provided to the Fyler Codes of Boston Children's Hospital and Harvard University, the International Statistical Classification of Diseases and Related Health Problems of the World Health Organization (ICD-9, ICD-10), the Current Procedural Terminology (CPT), and Canadian Congenital Heart Disease Codes. In 2007, to complement the efforts of the NWG, the ISNPCHD created two additional committees or working groups. These are the International Working Group for Defining the Nomenclatures for Pediatric and Congenital Heart Disease, also known as the Definitions Working Group (DWG), and The International Working Group for Archiving and Cataloguing the Images and Videos of the Nomenclatures for Pediatric and Congenital Heart Disease, better known as the Archiving Working Group (AWG). The mission of the NWG is to continue to maintain, preserve, and update the IPCCC. The DWG writes definitions for the terms in the IPCCC, building on the previously published definitions from the NWG. The purpose of the AWG is to link images of all types to the definitions provided by the DWG, and the codes developed by the NWG. The images and videos include photographs of gross anatomic specimens, echocardiograms, angiocardiograms, computerized axial tomographic and

Figure 1. Archiving Working Group Website - http://ipccc-awg.net

magnetic resonance images, as well as intraoperative photographs and videos. The AWG currently has an active image and video archive, called the AWG Web Portal (Figure 1), which can be viewed at http://ipccc-awg.net/.

The AWG Web Portal

The AWG management and workflow structure follows a process similar to that used in a peer-reviewed publication. The identification of images and accompanying textual explanations, may be solicited or unsolicited. The members of the AWG project (Table 1), in particular the Senior Archivist, share in the responsibility of identifying suitable images that illustrate the various aspect of the codes and definitions of the IPCCC. The Senior Archivist, and the three Co-Chairpersons, work closely in the initial review and assignment of the codes and definitions of the images submitted. After the initial identification and assignment process is completed, a web page is modified or created, and the images, codes, definitions and explanatory text are posted to the AWG Web Portal (http://ipccc-awg.net). The initial publication of the images to the AWG Web portal is done with a 'pending' certification. Six times yearly, the members of the AWG review the posted images, codes, and text for accuracy, quality, and suitability. After this process is completed, the posted images, if approved, receive the official certification and rating, and the web page is updated to reflect the date of final approval.

The AWG Web Portal structure is a menu-driven, unidirectional, navigational system based on the IPCCC Short Lists. To navigate the web site, the user selects from drop-down menus to reach the areas of interest. The user clicks on the image and code of interest to access the web page, where the codes, images, and explanatory texts are displayed. The images and videos reflect a variety of modalities, including photographs of autopsy or intra-operative specimens, clips of echocardiograms, angiocardiograms, computerized axial tomograms, and magnetic resonance studies. The contributions are catalogued and stored using industry standard formats. Although the images displayed may have copyright obligations, a feature of the AWG Web Portal is that the contributing author has given permission for the portal visitor to download the images, and to use them for not-for-profit, instructional, or educational purposes.

Conclusion

The Archiving Working Group of the International Society for Nomenclature of Paediatric and Congenital Heart Disease has created a web-based archive of representative images that visually define

Table 1: (Members of the Archiving Working Group)

Co-Chairpersons • Vera D. Aiello (Cardiac Pathologist, Brazil) • Robert H. Anderson (Cardiac Morphologist, UK & USA) • Jorge M. Giroud (Pediatric Cardiologist, USA)	**Editorial Members** • Carl Backer (CV Surgeon, USA) • Meryl Cohen (Pediatric Cardiologist, USA) • Andrew Cook (Cardiac Morphologist, UK) • Allen D. Everett (Pediatric Cardiologist, USA) • J. William Gaynor (CV Surgeon, USA) • Marina Hughes (Pediatric Cardiologist/MRI, UK) • Marshall L. Jacobs (CV Surgeon, USA) • Amy Juraszek (Pediatric Cardiologist, USA) • Otto N. Krogmann (Pediatric Cardiologist, Germany) • Hiromi Kurosawa (CV Surgeon, Japan) • Leo Lopez (Pediatric Cardiologist, USA) • Bohdan Maruszewski (CV Surgeon, Poland) • Giovanni Stellin (CV Surgeon, Italy) • Paul M. Weinberg (Pediatric Cardiologist/Morphologist, USA)
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the codes and textual definitions of the International Paediatric and Congenital Cardiac Code.

The ISNPCHD has developed a system of nomenclature for multiple uses, including clinical, research, educational, and administrative purposes. The creation of a web-based visual 'encyclopedia' that illustrates the terms and definitions of the IPCCC helps to further this goal. It also adds the element of international inclusion, since the members of the AWG represent a cross section of the world's health care professionals interested in the cardiac diseases that affect children of all ages. Furthermore, the images and contributions to this project are open to all regardless of national or geographical origin. Please visit the AWG Web Portal (http://ipccc-awg.net/), and contribute your images and thoughts. We also invite you to review our 'Image of the Month' column, which debuts in this publication, and trust that you find it a valuable resource. We hope that this column becomes a part of your educational activities, and helps in furthering your understanding of the cardiac diseases which affect neonates, children, and increasingly adults with congenital cardiac disease.

"We hope that this column becomes a part of your educational activities, and helps in furthering your understanding of the cardiac diseases which affect neonates, children, and increasingly adults with congenital cardiac disease."

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Vera D. Aiello, MD
Co-Chairman, Archiving Working Group
Heart Institute (InCor). São Paulo
University School of Medicine, Brazil

Robert H. Anderson, BSc, MD, FRCPath
Co-Chairman, Archiving Working Group
Institute of Medical Genetics
Newcastle University, UK

Corresponding Contributor

Jorge M. Giroud, MD
Co-Chairman, Archiving Working Group
Congenital Heart Institute of Florida &
Pediatrix Medical Group
St. Petersburg & Tampa, USA
jorgemgiroud@gmail.com

Diane E. Spicer, BS
Senior Archivist, Archiving Working Group
University of Florida, Department of
Pediatrics-Cardiology, Gainesville, Florida
Congenital Heart Institute of Florida
St. Petersburg & Tampa, FL USA

Use of a Standardized Approach to Evaluation of Pediatric Chest Pain: Does It Make a Difference?

By Brian M. Cardis, MD

Chest pain remains a common reason for referral to pediatric cardiologists. Despite a benign and non-cardiac etiology in the overwhelming majority of patients, this complaint continues to generate anxiety and fear of serious disease amongst patients and their families.[1,2] In addition, overutilization of diagnostic testing by providers to rule out rare but serious underlying cardiac pathology has led to a significant increase in healthcare dollars spent. A recent study estimated $3000 spent per patient in the outpatient evaluation of chest pain.[3] Large retrospective studies of outpatient or emergency room evaluation of pediatric chest pain have only recently been reported.[3,4]

Within our practice, thirty-six physicians evaluate approximately two-thousand children annually with the chief complaint of chest pain. In October 2010, our practice instituted a standardized evaluation protocol (Figure 1) in an attempt to develop a comprehensive yet cost-effective method to identify cardiac causes of chest pain in children and to limit practice variability amongst physicians. Beginning in October 2010, all new patients evaluated in an outpatient clinic with a chief complaint of chest pain were required to complete a standardized "Chest Pain Questionnaire." This questionnaire included objective answers to descriptive questions regarding chest pain such as character, location, duration, associated symptoms, occurrence at rest or exercise, etc. Patients also completed a "Cardiac Risk Assessment Form" in order to identify risk factors for sudden cardiac death in the past medical or family history. A number of questions on both forms were arbitrarily identified by our group as "red flag" items, indicating potential for underlying cardiac disease. Completed forms were used in conjunction with electrocardiogram (performed on all patients) and physical examination. According to the protocol, abnormalities on the chest pain questionnaire, cardiac risk assessment form, electrocardiogram, or physical exam warranted further evaluation with echocardiogram. If abnormalities existed and there was compelling evidence of a non-cardiac cause of chest pain, the physician was allowed to deviate from protocol but was required to document why an echocardiogram was not performed.

A total of 2,416 patients were seen with a chief complaint of chest pain during the protocol period (October 2010-December 2011). Ages ranged from 2-26 years (median age of 13 years) with slightly more males than females (54.3 vs. 45.7%). Abnormal findings on either electrocardiogram or echocardiogram occurred in 98 patients (4%) (Table 1). Many of these were felt to be incidental findings in patients with a clear non-cardiac cause of chest pain. In fact, a cardiac etiology of chest pain was found in only 26 patients (1%) (Table 2). Further, potentially life-

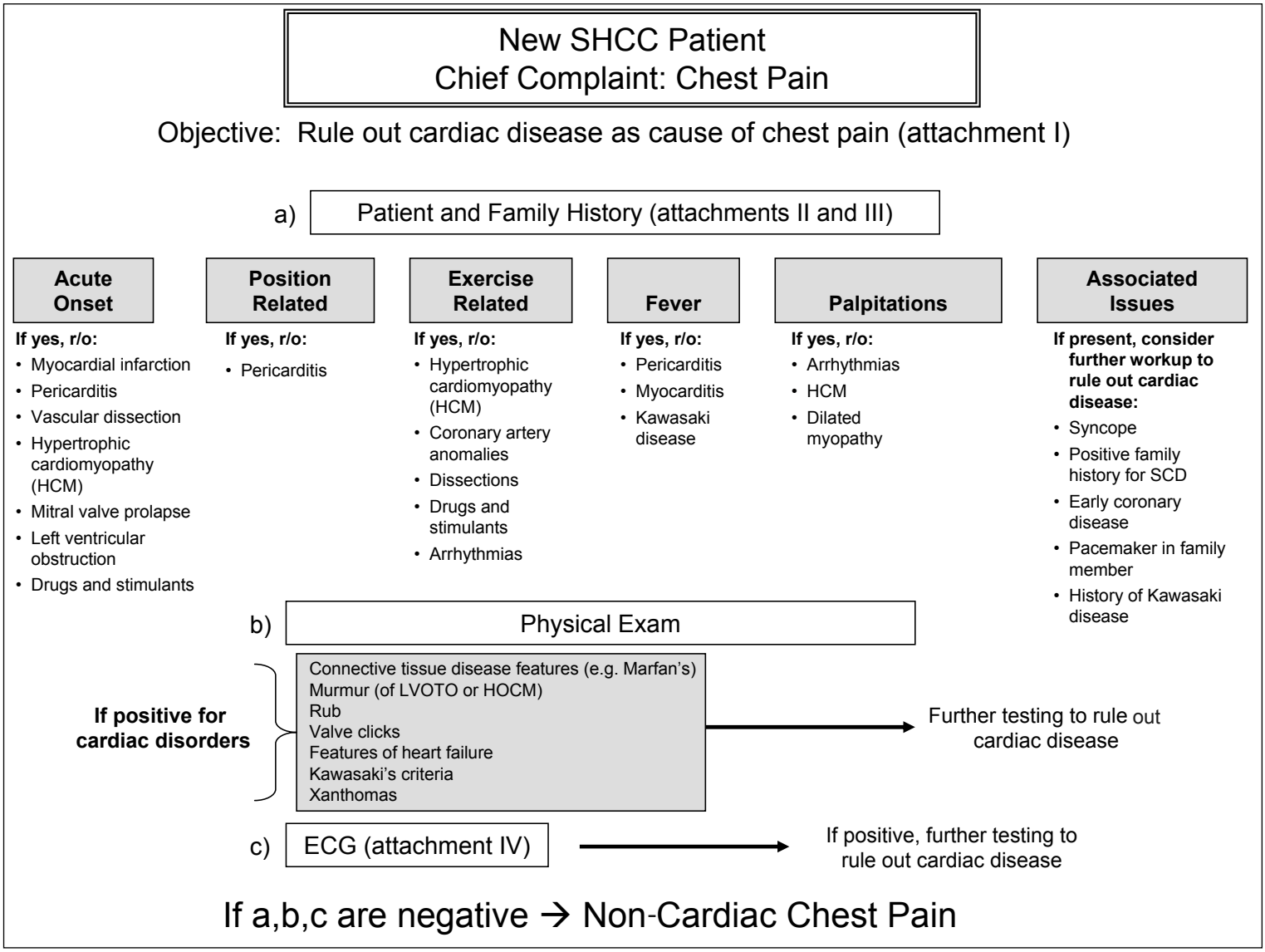

Table 1: Abnormalities Found by Either Electrocardiogram or Echocardiogram
98/2416 total patients identified

- Patent Foramen Ovale (16)
- Premature Ventricular Contractions (15)
- Wolff-Parkinson-White syndrome (8)
- Atrial Septal Defect (8)
- Mitral Valve Prolapse (7)
- Bicuspid Aortic Valve (6)
- Premature atrial contractions (5)
- Pericardial effusion (4)
- Mild pulmonary stenosis (3)
- Idiopathic dilated aorta (3)
- Supraventricular tachycardia (3)
- Patent Ductus Arteriosus – small (3)
- Left Ventricular Hypertrophy (3)
- Left Ventricular Noncompaction (2)
- Ventricular Septal Defect – small (2)
- Coronary Anomaly (2)
- Aortic valve disease (1)
- Hypertrophic Cardiomyopathy (1)
- Cleft Mitral Valve (1)
- Pericarditis (1)
- Coronary fistula (1)
- Right ventricular enlargement (1)
- Pulmonary hypertension – mild (1)
- Coarctation of the aorta (1)

threatening causes of chest pain were identified in only 17 patients (0.7%) and included: anomalous left coronary artery (n=1), anomalous right coronary artery (n=1), hypertrophic cardiomyopathy (n=1), Wolff–Parkinson-White Syndrome (n=7), mitral valve prolapse (n=6), and a combined finding of both Wolff-Parkinson-White syndrome and mitral valve prolapse (n=1). All 17 patients with potentially life-threatening cardiac disease had an abnormal physical examination or electrocardiogram.

A full cost analysis is currently in progress with results to be presented at the upcoming annual *Southeastern Pediatric Cardiology Society* (SEPCS) meeting in September. Preliminary analysis shows that echocardiogram was utilized in 1439/2416 (59.6%) of patients, an increase in usage when compared with patients evaluated prior to initialing the protocol (53.6%). Despite an overall increase in echocardiogram usage, this finding was not consistent throughout the group: 9/22 physicians who evaluated more than 20 patients with chest pain during the initial period had a lower utilization of echocardiogram after initiation of the protocol.

With the ultimate goal of identifying serious underlying cardiac pathology with minimal utilization of testing (echocardiogram), our group is currently working with the Georgia Institute of Technology to perform a detailed analysis of the chest pain questionnaire and cardiac risk assessment form. It is hoped that this will result in the ability to assign predictive values to each response with an eventual decrease in echocardiogram utilization. In addition, we are currently partnering with local pediatricians in order to adopt this protocol within the primary care setting. This may help to better select patients at risk for underlying cardiac conditions while eliminating unnecessary referrals for non-cardiac chest pain.

Table 2: Cardiac Causes of Chest Pain
23/2416 patients (1%)

- Mitral Valve Prolapse (7)
- Supraventricular Tachycardia (5)*
- Atrial Septal Defect – large (4)
- Pericardial Effusion (4)
- Coronary Anomaly (2)
- Hypertrophic Cardiomyopathy (1)
- Pericarditis (1)
- Coronary Fistula (1)
- Coarctation of the Aorta (1)

**Two patients with WPW were found to have SVT during subsequent electrophysiology study.*

To view the agenda or register for the *Southeastern Pediatric Cardiology Society* meeting, go to www.choa.org/sepcs.

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Brian M. Cardis, MD
Sibley Heart Center Cardiology / Children's Healthcare of Atlanta
1062 Forsyth St., Suite 3A
Macon GA 31201 USA
Phone: +1.478.743.4622

CardisB@kidsheart.com

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.

Closure.

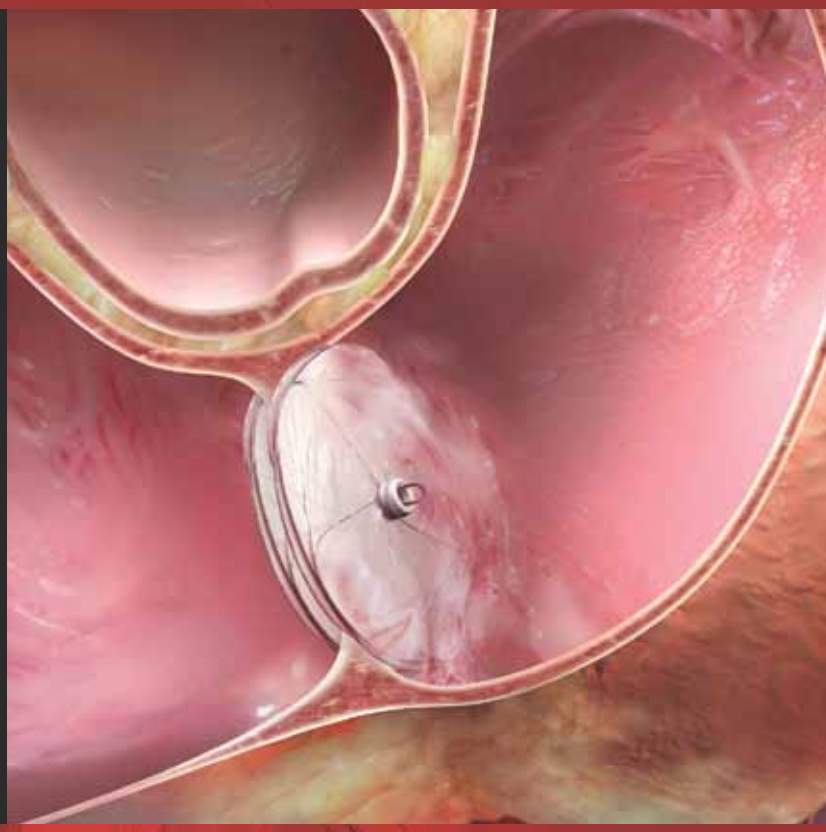

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Image of the Month - August 2012 - Presented by The Archiving Working Group

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

This is a special column that will be 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*.

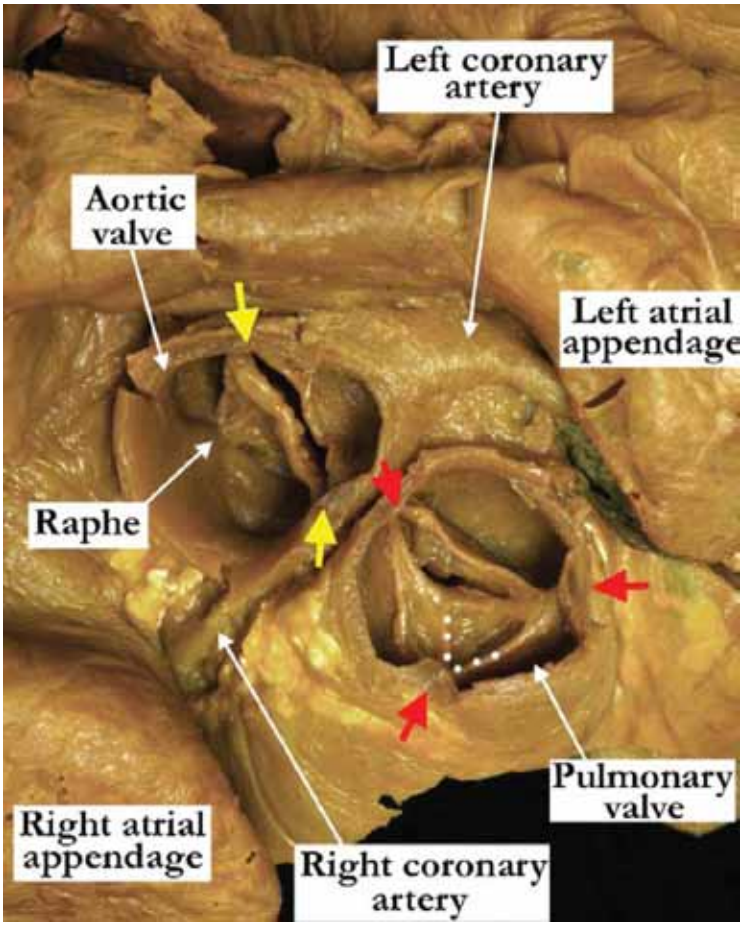

Figure 1. Description: This short axis view of the base of the heart shows the aortic and pulmonary valves. The aortic valve is bicuspid, with the attachments of the leaflets at the sinotubular junction marked with the yellow arrows. The right and left coronary arteries arise from separate sinuses. A raphe is present within the aortic sinus giving rise to the right coronary artery. The pulmonary valve is similarly bifoliate, and the valve is markedly stenotic. The white dots illustrate an area of fusion between two primordiums initially present during development, with the presence of an interleaflet triangle on the ventricular aspect supporting the notion of fusion during development.
Contributor: Diane E. Spicer, BS

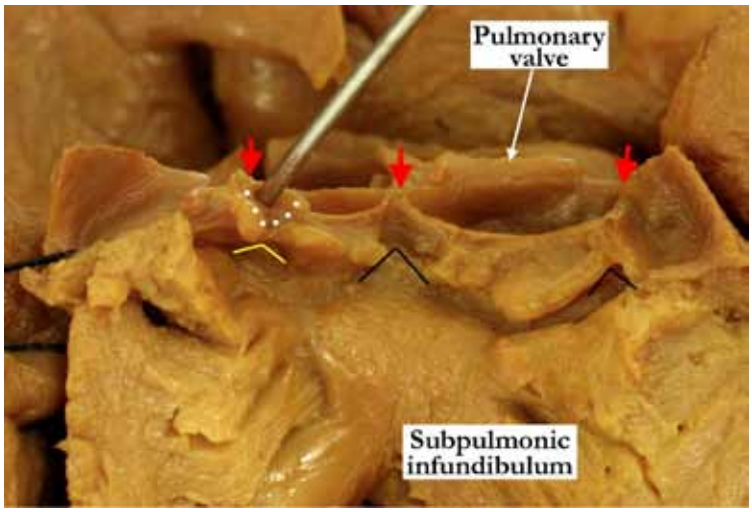

Figure 2. Description: The stenotic pulmonary valve is opened, demonstrating the thickened, nodular edges of the valve, along with the fused area (white dots) between two of the leaflets. The fused leaflets have been lifted to demonstrate the hypoplastic interleaflet triangle seen on the ventricular aspect (yellow lines). Note that this triangle is less well formed than the remaining two interleaflet triangles (black lines). The red arrows mark the attachment of the leaflets at the sinotubular junction.
Contributor: Diane E. Spicer, BS

IPCCC: 09.15.22, 09.05.32, 01.03.10

AEPC Derived Term

- Bicuspid aortic valve (09.15.22)
- Bicuspid pulmonary valve (09.05.32)
- Normal atrial arrangement (situs), atrioventricular and ventriculo-arterial connections (01.03.10)

EACTS-STS Derived Term

- Aortic valve pathology, Bicuspid (09.15.22)
- Pulmonary valve pathology, Bicuspid (09.05.32)
- Normal atrial arrangement (situs), AV & VA connections (01.03.10)

ICD10 Derived Term

- Congenital stenosis of aortic valve (Q23.0)
- Congenital insufficiency of aortic valve: Bicuspid aortic valve (Q23.1)
- Other congenital malformations of pulmonary valve (Q22.3)

"This series of images from the same specimen illustrate the use of the interleaflet triangles as a means of recognizing the overall nature of valves with two leaflets, and also those with only one leaflet (the so-called unicommissural and unicuspid variant)."

Comments

This series of images from the same specimen illustrate the use of the interleaflet triangles as a means of recognizing the overall nature of valves with two leaflets, and also those with only one leaflet (the so-called unicommissural and unicuspid variant). These images illustrate the bifoliate nature of the pulmonary and aortic valves of this specimen. Please note that the presence of three attachments at the sinotubular junction does not mean that the valve is trifoliate. The number of leaflets should be determined on the pattern of opening and closing of the valve, and these valves can only open and close in bifoliate fashion, since it has a solitary zone of apposition within the skirt of leaflet tissue. It is usual to find interleaflet triangles in the setting of bifoliate valves, but only rarely is the arrangement both bifoliate and bisinuate! Thus, the presence of the interleaflet triangle does not disqualify the valve from being bifoliate.

"Can You Identify the Malformations in Figure 3 for the October Column? Hint: Look at the aortic arch and log on to AWG Web Portal at ipccc-awg.net."

For Further Information on the Subject Please Refer to These References:

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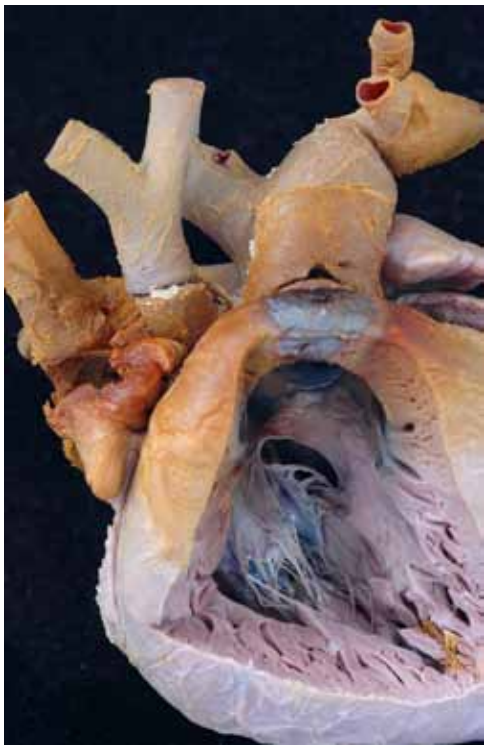
AWG Web Portal link for this series of images: http://ipccc-awg.net/Ao_Valve_Disease/Bicuspid_AO_PA_Valve_09_15_22/Bicuspid_AO_PA_Valve_09_15_22.html.



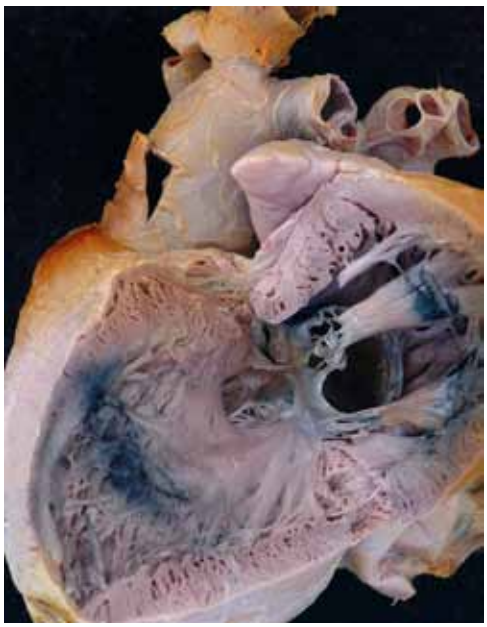


Figure 3. Images for the October column: 'Identify the malformations.'

Can You Identify the Malformations in Figure 3 for the October Column?

The answers for the malformations in Figure 3 will appear in the October issue of *Congenital Cardiology Today* in this bimonthly column. Hint: Look at the aortic arch and log on to AWG Web Portal at ipccc-awg.net.

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Vera D. Aiello, MD
Co-Chairman, Archiving Working Group
Heart Institute (InCor). São Paulo
University School of Medicine, Brazil

Robert H. Anderson, BSc, MD, FRCPath
Co-Chairman, Archiving Working Group
Institute of Medical Genetics
Newcastle University, UK

Corresponding Contributor

Jorge M. Giroud, MD
Co-Chairman, Archiving Working Group
Congenital Heart Institute of Florida &
Pediatrix Medical Group
St. Petersburg & Tampa, USA
jorgemgiroud@gmail.com

Diane E. Spicer, BS
Senior Archivist, Archiving Working Group
University of Florida, Department of Pediatrics-Cardiology, Gainesville, Florida
Congenital Heart Institute of Florida
St. Petersburg & Tampa, USA

Medical News, Products and Information

Scripps Doctors Study Novel New Device to Diagnose Irregular Heartbeat

A study conducted at Scripps Health has found that a novel new heart monitoring device helped emergency room patients avoid unnecessary follow-up care. Scripps Health electrophysiologist Steven Higgins, MD, presented findings of the study titled, "Prevalence of Arrhythmias in Emergency Department Patients Discharged Using a Novel Ambulatory Cardiac Monitor", today at the *Heart Rhythm Society's 33rd Annual Scientific Sessions* in Boston.

The study focused on the use of Zio® Patch, a single-use ambulatory cardiac monitor that looks similar to a 2- by 5-inch adhesive bandage and sticks to a patient's chest, that continuously monitors their heart rhythm for up to 14 days.

"The availability of this new heart monitor is exciting as it improves patient care. The patch is applied and when recording is done, the patient simply drops it in the envelope and returns it to us – it's like the Netflix of heart care," said Dr. Higgins, Chairman of the Department ofCardiology at Scripps Memorial Hospital La Jolla and a lead investigator. "Because they are infrequent, heart rhythm problems are often difficult to diagnose, even though they can be quite serious. The Zio Patch is a new digital advance that will allow us to better diagnose challenging cases so we can provide our patients the best care."

Scripps Memorial Hospital La Jolla was the only hospital in Southern California to participate in the study. Other study locations included Stanford Hospital and Scott & White Memorial Hospital in Temple, Texas.

The study followed 285 patients who had presented to emergency departments across the country with symptoms possibly related to arrhythmias, such as fainting, palpitations, dizziness and others. Patients received the unencumbering, wire-free Zio® Patch prior to being discharged from the emergency room and were instructed to wear the patch until it no longer adhered to their skin – up to 14 days duration. Devices were mailed back to iRhythm Technologies, Inc., the Zi® Patch's developer and service provider, using a pre-paid postage envelope, for analysis and reporting of results to the patient's physician.

The researchers found that 59% of the symptomatic patients who presented to the emergency rooms did not have arrhythmia and may not require any further work-up. "Thus, the new device has the potential to save the health care system millions of dollars," said Higgins. "We were also surprised to learn that there was 100% compliance by the patient with the process, which is an amazing finding for an emergency department study."

Monitoring Leads to Possible Lifesaving Procedure

One patient who benefited from the Zio Patch is La Jolla resident, Kenneth Curzon, who fainted while at work in March. Curzon continuously wore the Zio Patch for two weeks and then mailed it back to iRhythm, where the information was downloaded and formatted into a report for Dr. Higgins to review.

The recording showed Curzon was experiencing prolonged pauses in his heart rhythm of over three seconds as well as other episodes of rapid heart beats. On April 6, he received an implantable cardiac defibrillator to correct the problem and was back to his management job within five days.

"The Zio® Patch allowed me to diagnose and determine the most appropriate therapy for Ken," said Higgins.

I like to think of the whole experience as an adventure," Curzon said. "Most of the time I didn't even realize I was wearing a heart monitor, and when I peeled it off, I just put it in an envelope and sent it off in the mail. It was a very simple process."

Zio® Patch vs. Holter monitor

In addition, Dr. Eric Topol is leading a new related study at Scripps Green Hospital examining whether the Zio Patch does a better job of detecting heart arrhythmias than the Holter monitor, which has been the gold standard for rhythm monitoring since the early 1960s.

The portable Holter monitor collects its data through a series of wired electrodes that adhere to the chest. Because the device can be difficult to wear and can get in the way of normal activities such as showering, exercising and sleeping, continuous use of the monitor is typically limited to one or two days. In contrast, the Zio® Patch is a small, unobtrusive device that is indicated for up to 14 days of wear, and allows the patient to exercise and shower on their normal schedule, without the hassle of a bulky monitor and multiple wires.

"This is a great opportunity to compare these two side-by-side for use in diagnosing important heart rhythm disturbances," said Dr. Topol, a cardiologist who directs the Scripps Translational Science Institute and serves as Chief Academic Officer of Scripps Health. "We are trying to determine if the Zio Patch will have an increased diagnostic yield."

The study is currently enrolling about 150 Scripps Green and Scripps Clinic adult patients who have been seen by their doctors for arrhythmia. Each of the participants will wear a Holter monitor and a Patch for up to 48 hours and then continue wearing the Zio Patch for up to 14 days. More information about the study is available at www.clinicaltrials.gov.

Dr. Topol and his research associates will compare the data gathered from each device and report their findings later this year.

Arrhythmias affect millions of Americans each year and, if left untreated, may lead to serious consequences including stroke or sudden cardiac death.

Both studies are an extension of Scripps Health's leadership in heart care and research. Scripps is currently building the $456 million Prebys Cardiovascular Institute, a center for innovation that will bring together top researchers, physicians and staff. The institute will incorporate leading-edge wireless technologies and individualized medicine for the best in patient care when it opens in 2015. Annually, more than 55,000 patients receive their cardiovascular care from Scripps, making it San Diego County's largest heart care provider. Scripps is the region's only cardiovascular program consistently recognized by U.S. News & World Report as one of the best in the country.

More information can be found at www.scripps.org.

ACC and ACP Expand Performance Measures Detailed in PINNACLE-PC Registry To Provide Access to Primary Care Internists

The American College of Cardiology (ACC) and the American College of Physicians (ACP), announced in early June plans to launch a pilot program, PINNACLE Primary Care, that will expand the PINNACLE Registry to assess performance measures for primary care practices.

The PINNACLE Registry currently collects data from cardiology practices to calculate performance measures related to atrial fibrillation, hypertension, coronary artery disease and heart failure. Made up of more than 4.2 million patient encounters, PINNACLE is the nation's largest cardiovascular outpatient quality-improvement registry. PINNACLE Primary Care, which will be known as PINNACLE-PC, will measure performance on breast and colon cancer screenings, influenza and pneumonia immunizations, lower back pain, diabetes, depression, weight management, and chronic obstructive pulmonary disease.

The pilot will begin this summer with five to 10 internal medicine practices. Data from pilot practices will be collected through December 2012 and results will be analyzed in early 2013. If the pilot is successful, the ACC and ACP will make the PINNACLE Registry available to members of the American College of Physicians.

"Registries are excellent quality improvement tools," said ACC President Dr. William Zoghbi, MD, FACC. "Registry participants receive quick feedback that can help practices ensure that they are providing consistent, evidence-based care. Expanding the PINNACLE Registry to include primary care is a natural extension of this great resource, which will ultimately improve care."

"We are very enthusiastic about our collaboration with ACC and this program to test the PINNACLE Registry in primary care internal medicine practices," said ACP President David L. Bronson, MD, FACP.

ACC Senior Vice President of Science & Quality and PINNACLE Chair William J. Oetgen, MD, MBA, FACC, said expanding PINNACLE to primary care has been a goal for the registry since its inception.

The PINNACLE registry assists practices in understanding and improving care through the production and distribution of quarterly performance reports for data-submitting practices and physicians. The reports cover all valid patient encounters and detail adherence to 25 cardiovascular

clinical measures at the physician, location, and practice levels across the diagnoses of coronary artery disease, hypertension, heart failure, and atrial fibrillation. Thirteen additional performance measures will be added to PINNACLE to create PINNACLE-PC.

As part of the National Cardiovascular Disease Registries (NCDR) suite, the PINNACLE Registry provides clinicians with quality measurement solutions needed in today's health care environment. The registry provides a centralized system for clinical practices to promote practice innovations and achieve clinical excellence.

Skewed Results? Failure to Account for Clinical Trials Drop-Outs Can Lead to Erroneous Findings in Top Medical Journals

Newswise - A new University at Buffalo study of publications in the world's top five general medical journals finds that when clinical trials do not account for participants who dropped out, results are biased and may even lead to incorrect conclusions.

Published recently in the *British Medical Journal*, the methodological study consisted of a systematic analysis of 235 clinical trials published in the world's top five general medical journals between 2005 and 2007 that claimed a statistically significant effect.

"We found that in up to a third of trials, the results that were reported as positive – in other words, statistically significant – would become negative – not statistically significant, if the investigators had appropriately taken into consideration those participants who were lost to follow-up," says Elie A. Akl, MD, MPH, PhD, lead author, and Associate Professor of Medicine,Family Medicine and Social and Preventive Medicine at the University at Buffalo School of Medicine and Biomedical Sciences and School of Public Health and Health Professions. He also has an appointment at McMaster University.

"In other words, one of three claims of effectiveness of interventions made in top general medical journals might be wrong," he says.

In one example, a study that compared two surgical techniques for treating stress urinary incontinence found that one was superior. But in the analysis published this month, it was found that 21% of participants were lost to follow-up. "When we reanalyzed that study by taking into account those drop-outs, we found that the trial might have overestimated the superiority of one procedure over the other," Akl says.

According to Akl, it has always been suspected, but never proven, that loss to follow-up introduces bias into the results of clinical trials. "The methodology we developed allowed us to provide that proof," he says.

The methodology that he and his coauthors developed consists of sensitivity analyses, a statistical approach to test the robustness of the results of an analysis in the face of specific assumptions, in this case, assumptions about the outcomes of patients lost to follow-up.

"This study gives us a better understanding of the problem of loss to follow-up in clinical trials and provides us with better tools to address it," Akl says.

"This methodology will allow those who conduct the trials and those who use their results, including clinicians, other scientists, developers of clinical guidelines, policymakers and bodies like the Food and Drug Administration (FDA), to better judge the risk of bias," concludes Akl.

The studies that were analyzed had previously been published in Annals of Internal Medicine, British Medical Journal, the Journal of the American Medical Association, Lancet and the New England Journal of Medicine. To be included, the trials that were studied had to have reported a significant effect.

Akl led this major study, which took three years to complete. His co-authors, 20 clinical epidemiologists, are from the following institutions: McMaster University; University Hospital Basel; Kaiser Permanente Northwest; Hospital for Sick Children in Toronto; Institute for Work and Health, Universitè de Sherbrooke; University Children's Hospital Tuebingen; Pontificia Universidad Catolica de Chile;

Tel Aviv University; the University of Ottawa; the University of Freiburg and the University of Oxford.

Children's National Medical Center and Baby's First Test Join Forces to Advance - Screening for Critical Congenital Heart Disease in Newborns

The Children's National Medical Center and Baby's First Test released two videos on screening for critical congenital heart disease (CCHD) using pulse oximetry (pulse ox) in newborns. The videos, produced for parents and clinicians respectively, were designed to forward knowledge about the test.

CCHD screening is quickly becoming a standard of care across the country. Nine states have passed legislation surrounding CCHD screening of newborns, and many more have legislation or executive committees examining the implementation of the procedure. In September 2011, CCHD screening was endorsed by the Department of Health and Human Services to be included in the recommended uniform screening panel for newborns in the U.S.

Children's National is a national leader in research and advocacy on screening newborns for CCHD. "The videos we are releasing today are essential to our efforts to educate parents and clinicians about critical congenital heart disease screening. We have learned that the pulse ox test can be used to identify newborns with CCHD before discharge from birthing facilities. We know that approximately 3 of every 1,000 babies born are affected by CCHD and early detection of serious forms of the disease may improve health outcomes for those babies," said Gerard Martin, MD, Senior Vice President of the Center for Heart Lung and Kidney at Children's National.

Baby's First Test, which is operated by the non-profit group Genetic Alliance, is the nation's newborn screening. Newborn screening (NBS) is the process of testing newborn babies for some serious, but treatable, conditions. NBS can include a heel stick, hearing screen, and pulse oximetry. The conditions that newborn babies are screened for varies by state.

"We are thrilled to offer these videos to parents who want to understand what tests their babies are getting, and why; and to clinicians who want to learn more about the use of pulse ox in the newborn nursery," said Natasha Bonhomme, Director of Baby's First Test.

To view the videos, please visit www.ChildrensNational.org/pulseox or www.babysfirsttest.org.

Baby's First Test, the nation's newborn screening information center, is dedicated to educating parents, health professionals, and the public about the newborn screening system. Baby's First Test provides information and resources about screening at the local, state and national levels. This initiative is funded through a cooperative agreement with the Genetic Services Branch of the Maternal and Child Health Bureau of the Health Resources Administration to Genetic Alliance, a 25-year old non-profit organization that seeks to improve health through the authentic engagement of communities and individuals.

CONGENITAL CARDIOLOGY TODAY

Corporate Offices: 8100 Leaward Way, Nehalem, OR 97131 USA
Mailing Address: PO Box 444, Manzanita, OR 97130 USA
Tel: +1.301.279.2005; Fax: +1.240.465.0692
Editorial and Subscription Offices: 16 Cove Rd, Ste. 200, Westerly, RI 02891

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- Pre-Selection of Patients of Pulmonic Valve Implantation and Post-Procedural Follow-up
- Echo Paravalvular Leakage (PVL)
- ICE vs TEE ASD Closure in Children - PRO & CON ICE
- 3D Rotational Angiography - Why Every Cath Lab Should Have This Modality
- PICS Doorway to the Past - Gateway to the Future
- Follow-up From PICS Live Cases 2010 Presentation
- Intended Intervention - Transcatheter TV Implantation - *Live Case*
- Intended Intervention - LAA Closure Using Amplatzer Cardiac Plug Under GA & Real Time 3D
- Provided Intervention - LPA Stenting / Implantation of a Sapien Valve
- Intended Intervention - PV Implantation
- Intended Intervention - COA Stent Using Atrium Advanta V12 Covered Stent - *Live Case*
- Intended Intervention - ASD Closure - *Live Case*
- Intended Intervention -Transcatheter VSD Device Closure - *Live Case* Intended Intervention - COA Stenting Using Premounted Advanta V12 Covered Sten - *Live Case*
- Stunning Revelation - The Medical System is Changing - What Can You Do To Show Patients That Your Practice Does It Right? Patient Perspective
- Percutaneous Paravalvular Leak Closure Outcomes
- Intensive Management of Critically Ill Infants Undergoing Catheterization
- Off Label Device Usage - Careful!
- Evolving Hybrid Programs Communications is the Key
- Anti-Coagulation in Pediatric Interventions - Evidence Please! Are we Flying Blind?
- Shaping Your Structural Heart Interventional Career: How Do I Get the Necessary Training and Start a Program?
- Closure of Post-Infarct VSDs Limitations & Solutions
- Percutaneous Interventions During Pregnancy
- Percutaneous PVL Closure Techniques & Outcomes
- Percutaneous Therapy for MR
- Update on Percutaneous Aortic Valve Replacement: Time to Open the Flood Gates?
- 2011 PICS/AICS Achievement Achievement Award Presented To Horst Sievert, MD.
- The Cases I Learn The Most From
- If I was Starting Now I Would... Contemplating the Question.
- 10 Tips for the Trainee
- The Perfect CV
- Why I Left The Cath Lab
- Intended Intervention Occlude Right High Flow Superior Segment PAVM
- Intended Intervention Edwards Sapien XT Transcatheter Heart Valve - 3D Guidance - *Live Case*
- Closure of the Silent PDA - Indicated or Not
- Transcatheter Tricuspod Valve Replacement
- Recommendations For Device Closure of MVSDs
- Intended Intervention Diagnostic Angio/First Intervention Occlude Left Lower Lobe - *Live Case*
- Intended Intervention PFO Closure using Gore Helex Septal Occluder Under GA and TEE Guidance - *Live Case*
- Pulmonary Valvuloplasty - Pulmonary Oedema Presentation
- Intended Intervention LAA Closure Using the Amplatzer Cardiac Plug - *Live Case*
- Transcatheter Alternatives in Sick Neonates With Coartation of the Aorta; Balloon Angioplasty vs Surgery for Native Coartation of the Aorta in Newborns
- **and many more…. Plus, live cases from other major meetings**

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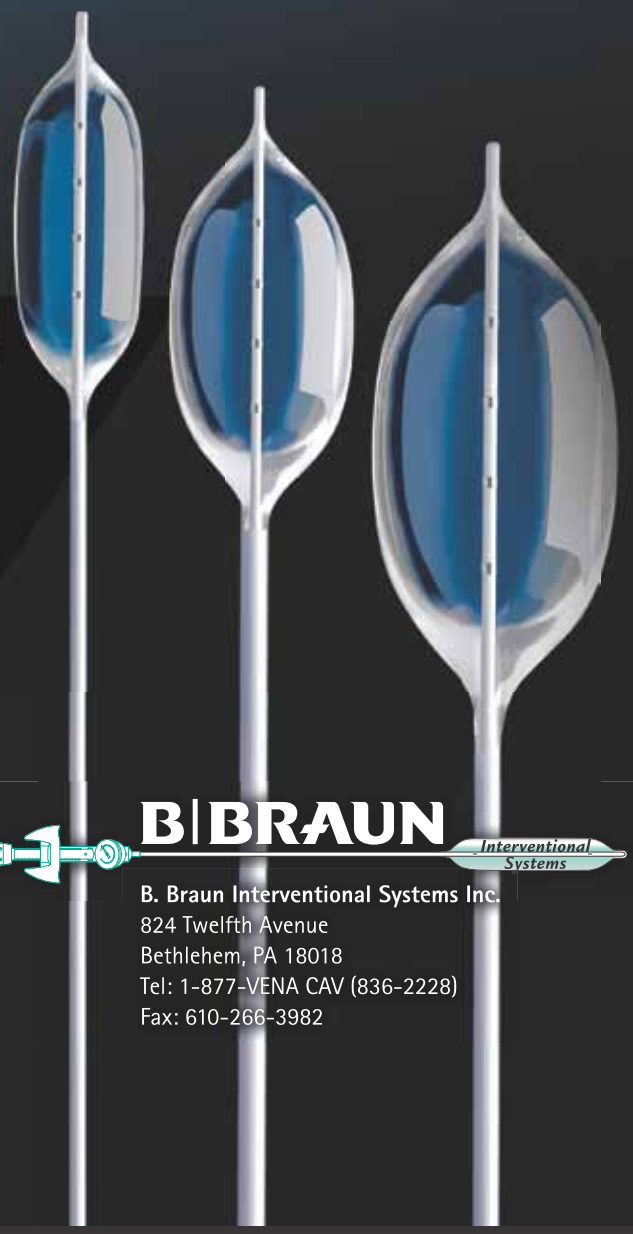

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B. Braun Interventional Systems Inc.
824 Twelfth Avenue
Bethlehem, PA 18018
Tel: 1-877-VENA CAV (836-2228)
Fax: 610-266-3982

www.bisusa.org