CONGENITAL CARDIOLOGY TODAY

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

February 2017; Volume 15; Issue 2
North American Edition

IN THIS ISSUE

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

Twitter:
www.Twitter.com/ccardiology

Official publication of the CHiP Network

Recruitment ads on pages:
4, 6, 14, 17, 19

Cyanosis from Portosystemic Shunt after Fontan Completion

Ege Ozdemir, MD; Saadeh B. Jureidini, MD; and Renuka E. Peterson, MD

Introduction

Heterotaxy may be associated with various forms of venous anomalies. We describe here anomalous venous drainage from a portosystemic communication that resulted in severe cyanosis in a heterotaxy patient after Fontan completion.

Case Report

A newborn male was diagnosed shortly after birth with Heterotaxy Syndrome, asplenia and Complex Congenital Heart Disease (CCHD). His cardiac anatomy consisted of dextrocardia, common atrium, unbalanced atrioventricular canal, double-outlet right ventricle with subpulmonary and pulmonary valve stenosis, and obstructed infradiaphragmatic Total Anomalous. Pulmonary. Venous Return

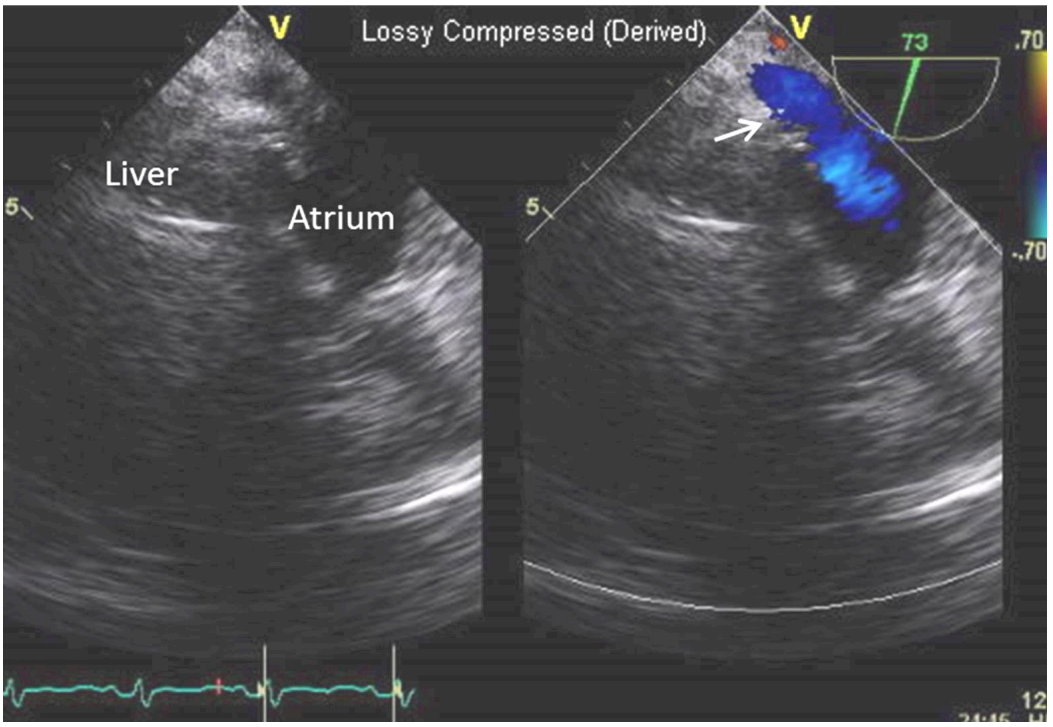


Figure 1. Transesophageal echocardiogram image showing residual venous drainage directly to the atrium (arrow) inferiorly; this drainage is separate from where the pulmonary veins drain.

(TAPVR). His systemic venous anatomy demonstrated a right-sided inferior vena cava with right hepatics draining to the inferior vena cava while the left hepatics drained separately to the common atrium. A left superior vena cava drained to the left superior portion of the common atrium; a right superior vena cava was absent.

He initially underwent repair of the anomalous pulmonary venous return in the neonatal period, and then subsequently had a left-sided bidirectional Glenn shunt with division of the main pulmonary artery. Fontan completion was achieved with an extracardiac, fenestrated conduit on the left with incorporation of the hepatics. Postoperatively, his saturations remained unusually low for a fenestrated Fontan, in the high 60s to low 70s. A postoperative echocardiogram suggested residual venous drainage inferiorly to the atrium (Figure 1), prompting cardiac catheterization.

The catheterization demonstrated a newly-opened right superior vena cava draining to the common atrium (Figure 2a). Prior catheterizations with injection in the innominate vein, and at surgery, had not shown any evidence of patency. Since an additional venous source of blood flow inferiorly was also suspected, the catheter was passed into the atrium from the right superior vena cava and the atrium was probed. An unusual accessory vein was shown by angiography draining directly to the inferior aspect of the common atrium (Figures 2b, 2c), and this vessel was remote from the pulmonary veins. Further catheterization by abdominal CT demonstrated that the accessory vein arose from the portal vein (Figure 3).

The right superior vena cava was occluded with a transcatheter device. The patient then underwent exploratory laparotomy and a large vessel from the portal vein was confirmed that drained to the inferior aspect of the atrium, again noted to be away from the pulmonary veins. The portosystemic vein was surgically ligated and the patient had improvement in his oxygen saturations to the high 80s to 90s on room air and was discharged.

Discussion

Heterotaxy Syndrome is a failure of lateralization during embryological development leading to bilateral right-sidedness (asplenia) or bilateral left-sidedness (polysplenia), often accompanied by CCHD. Proper cardiac surgical management of patients with Heterotaxy Syndrome is contingent upon recognizing associated complex venous anomalies. Variations in both systemic venous return and pulmonary venous return have been well-described in the literature.[1,2] Many patients with heterotaxy and asplenia have bilateral superior vena cavae; however, only a portion of these may be patent.[2] Our patient, despite documented absence of a right superior vena cava previously, displayed recanalization of this persistent embryologic structure, which accounted for some of his desaturation.

Since this patient had infradiaphragmatic TAPVR, the additional aberrant vessel connecting to the portal system was thought to possibly represent a persistent vertical vein, which has been described in the literature.[9-12] However, on cardiac catheterization, the vessel was easily engaged directly from the atrium as opposed to through the pulmonary venous anastomosis and the vessel was not seen on levophase or pulmonary capillary wedge injection. This is in contrast to prior reports of persistent vertical vein after TAPVR repair, including a persistent vertical vein post-Fontan[13] with right to left shunting where the vertical vein drained via a pulmonary vein to the atrium. On multiple imaging modalities and direct visualization, the aberrant vessel in our patient was remote from the pulmonary veins and entered separately at the base of the atrium.

In addition, at the time of catheterization, the pressure in the aberrant venous vessel was identical to the pressure tracing in the common atrium, whereas the direct pulmonary venous pressures were higher due to mild pulmonary vein obstruction bilaterally. We would expect that if the aberrant vessel were related to a persistent vertical vein, it should have the same pressures as the pulmonary venous system. If there was direct communication with the pulmonary venous system from this vessel, then shunting in this patient should be left-to-right given the higher pressure in the pulmonary veins. For these reasons, the aberrant vessel seems less likely to represent a persistent vertical vein.

Despite well-documented pulmonary and systemic venous variations in heterotaxy patients, the importance of anomalies involving the portal circulation has only more recently been recognized.[3-5] Normal venous embryological development consists of paired cardinal, vitelline, and umbilical veins which ultimately develop into the systemic, portal, and hepatic venous structures of the neonate.[6] The timing of development of the portal circulation coincides with that of cardiac

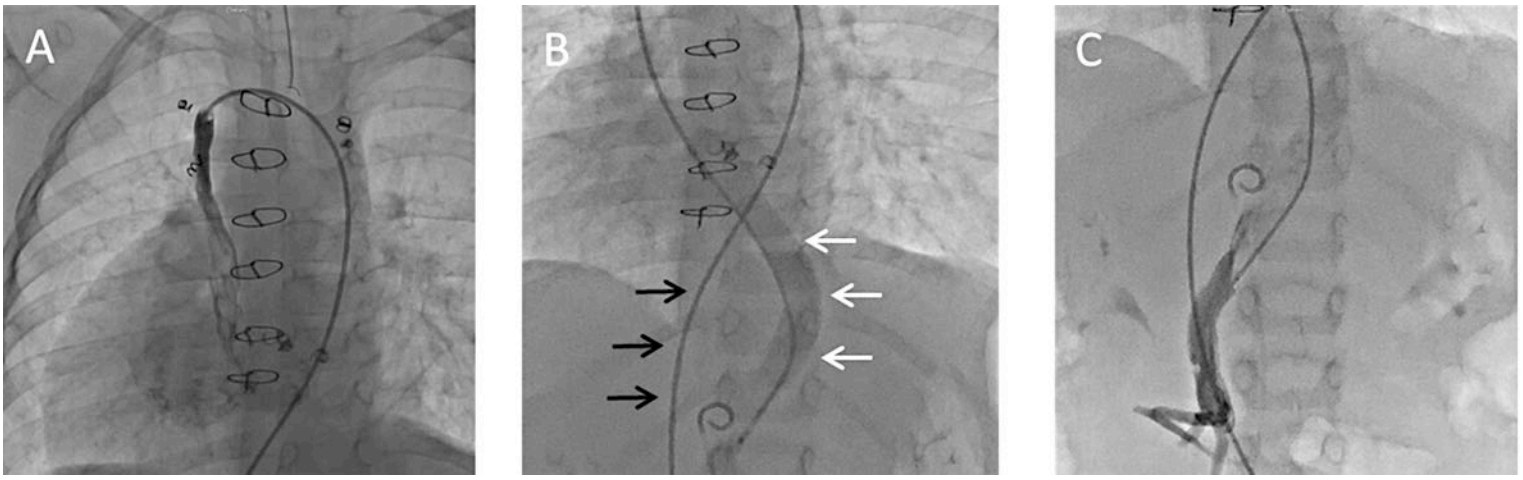


Figure 2: Catheter course through left-sided Fontan conduit, into innominate vein and passing through (A) recanalized right superior vena cava draining to the atrium, and then, (B) into abnormal venous drainage entering inferiorly to the atrium (light arrows) and separate from the Fontan conduit (dark arrows). (C) Injection more caudally into the abnormal vessel shows filling of vessels related to portal venous drainage.

10 YEARS.
10,000 PATIENTS.
1 LIFE-CHANGING VALVE.

Melody™
Transcatheter Pulmonary Valve Therapy

Designed Specifically For Pulmonary Valve Replacement

The Melody valve is the longest studied transcatheter valve and is proven to delay conduit replacement:

88.8%
freedom from reoperation at 7 years post-implant*

*US IDE Study

Melody-TPV.com

The Melody TPV System first received CE mark in September, 2006.
The Melody TPV System received Health Canada approval in December 2006 and US approval under an HDE on January 25, 2010 (H080002).
PMA approval received January 27, 2015 (P140017).

710 Medtronic Parkway
Minneapolis, MN 55432-5604
USA
Tel: (763) 514-4000
Fax: (763) 514-4879
Toll-free: (800) 328-2518

LifeLine
CardioVascular Technical Support
Tel: (877) 526-7890
Tel: (763) 526-7890
Fax: (763) 526-7888
rs.cstechsupport@medtronic.com

UC201607890a EN 11/2016

Medtronic
Further, Together

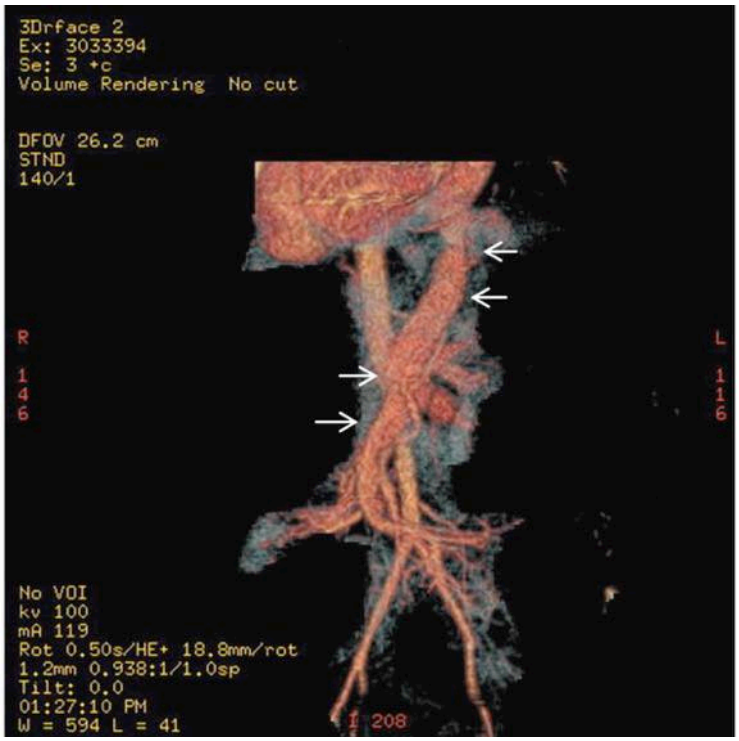


Figure 3. Three-dimensional CT reconstruction showing abnormal vessel that represents a congenital portosystemic shunt (arrows) draining from the portal circulation to the base of the atrium; this vessel is not connected with the pulmonary venous system.

embryogenesis and there has been a reported increased incidence of Congenital Heart Disease in patients with a congenital portosystemic shunt.[7] The potential embryologic causes for such shunts may include primary failure of critical anastomoses of the vitelline veins leading to complete absence of the portal system, persistence of the ductus venosus, or agenesis of the ductus venosus.[8] In heterotaxy patients, these shunts have been reported in patients with polysplenia and an interrupted IVC where the connection is generally between a renal vein and the portal vein.[3,5]

Although systemic and pulmonary venous circulations are routinely visualized during preoperative evaluation for single ventricle palliation, portosystemic shunts may be missed with echocardiography and even venography, unless there is a high index of suspicion. In our patient, multiple veins entering the atrium also made this difficult to diagnose preoperatively. This case points to the importance of considering congenital portosystemic shunts in heterotaxy patients. Evaluation by CT or MRI may be a useful adjunct to delineate the complex venous structures in such patients prior to surgical intervention.

References

1. Burton EC, Olson M, Rooper L. Defects in laterality with emphasis on heterotaxy syndromes with asplenia and polysplenia: an autopsy case series at a single institution. Pediatr Dev Patho 2014; 17(4): 250-264.
2. Rubino M, Van Praagh S, Kadoba K, Pessotto R, Van Praagh R. Systemic and pulmonary venous connections in visceral heterotaxy

with asplenia. Diagnostic and surgical considerations based on seventy-two autopsied cases. J Thorac Cardiovasc Surg 1995; 110(3): 641-650.
3. Mcelhinney DB, Marx GR, Newburger JW. Congenital portosystemic venous connections and other abdominal venous abnormalities in patients with polysplenia and functionally univentricular heart disease: a case series and literature review. Congenit Heart Dis 2011; 6(1): 28-40.
4. Sokollik C, Bandsma RH, Gana JC, Van den Heuvel M, Ling SC. Congenital portosystemic shunt: characterization of a multisystem disease. J Pediatr Gastroenterol Nutr 2013; 56(6): 675-681.
5. Newman B, Feinstein JA, Cohen RA, et al. Congenital extrahepatic portosystemic shunt associated with heterotaxy and polysplenia. Pediatr Radiol 2010; 40(7): 1222-1230.
6. Yagel S, Kivilevitch Z, Cohen SM, et al. The fetal venous system, part I: normal embryology, anatomy, hemodynamics, ultrasound evaluation and Doppler investigation. Ultrasound Obstet Gynecol 2010; 35(6): 741-750.
7. Guérin F, Blanc T, Gauthier F, Abella SF, Branchereau S. Congenital portosystemic vascular malformations. Semin Pediatr Surg 2012; 21(3): 233-244.
8. Yagel S, Kivilevitch Z, Cohen SM, et al. The fetal venous system, Part II: ultrasound evaluation of the fetus with congenital venous system malformation or developing circulatory compromise. Ultrasound Obstet Gynecol 2010; 36(1): 93-111.
9. Kumar RN, Dharmapuram AK, Rao IM, et al. The fate of the unligated vertical vein after surgical correction of total anomalous pulmonary venous connection in early infancy. The Journal of Thoracic and Cardiovascular Surgery 2001; 122(3): 615-617.
10. Kobayashi D, Forbes TJ, Delius RE, Aggarwal S. Amplatzer vascular plug for transcatheter closure of persistent unligated vertical vein after repair of infracardiac total anomalous pulmonary venous connection. Catheter Cardiovasc Interv 2012; 80(2):192-8.
11. Narula N, Wilson N, Kumar RS. Transcatheter closure of persistent unligated vertical vein after TAPVC surgery using the Amplatzer PDA device. Catheter Cardiovasc Interv 2007; 70(1):117-9.
12. Larsen B, Pophal S, Richardson R. Persistent unligated vertical vein following repair of total anomalous pulmonary venous return: an unusually late presentation. Congenital Cardiology Today 2015; 13(12): 1-8.
13. Mishaly D, Hegesh J, Frand M, Smolinsky A, Amir G. Unintended right-to-left shunt after Fontan operation: consequence of unligated vertical vein in a child with total anomalous pulmonary venous drainage, single ventricle, and heterotaxy. J Thorac Cardiovasc Surg 2003; 125(2): 421-423.

CCT

Corresponding Author

Ege Ozdemir, MD
Department of Pediatrics
Division of Pediatric Cardiology
SSM Health Cardinal Glennon Children's Hospital
1465 S. Grand Blvd.
St. Louis, MO 63104 USA
Phone: 314-577-5634
Fax: 314-577-2616
eozdemir@slu.edu

Saadeh B. Jureidini, MD
Department of Pediatrics
Division of Pediatric Cardiology
SSM Health Cardinal Glennon Children's Hospital and Saint Louis University School of Medicine
St. Louis, MO 63104 USA

Renuka E. Peterson, MD
Department of Pediatrics
Division of Pediatric Cardiology
SSM Health Cardinal Glennon Children's Hospital and Saint Louis University School of Medicine
St. Louis, MO 63104 USA

E-mail comments to:
CCT-Feb17-Cyanosis@CCT.bz

Image of the Month from the Archiving Working Group

Contributors: Diane E. Spicer, BS; Jeffrey P. Jacobs, MD; James St. Louis, MD; Jorge M. Giroud, MD; Charles W. Shepard, MD; Allen Everett, MD; Robert Anderson, MD; Vera D. Aiello, MD; Meryl S. Cohen, MD

International Paediatric and Congenital Cardiac Code (IPCCC) AEPC Derived Terms

1. Common atrioventricular valve with unbalanced commitment of valve to ventricles (06.07.36)
2. Common atrioventricular valve with unbalanced commitment of valve to ventricles: dominant right (06.07.37)
3. Common atrioventricular valve with unbalanced commitment of valve to ventricles: dominant left (06.07.38)
4. Ventricular myocardial noncompaction cardiomyopathy (07.08.50)
5. Mirror image atrial arrangement (atrial situs inversus) (01.03.01)
6. Discordant atrioventricular connections (01.04.01)

EACTS-STS Derived Terms

1. AVC (AVSD), Complete (CAVSD)-modifier, Common atrioventricular valve with unbalanced commitment of valve to ventricles (06.07.36)
2. AVC (AVSD), Complete (CAVSD)-modifier, Common atrioventricular valve with unbalanced commitment of valve to ventricles, Unbalanced commitment of valve to right ventricle (06.07.37)
3. AVC (AVSD), Complete (CAVSD)-modifier, Common atrioventricular valve with unbalanced commitment of valve to ventricles, Unbalanced commitment of valve to left ventricle (06.07.38)
4. Cardiomyopathy, Spongiform cardiomyopathy (Noncompaction cardiomyopathy) (Embryonal cardiomyopathy) (07.08.50)
5. Atrial situs inversus (Mirror image atrial arrangement) (01.03.01)
6. AV connection(s) = Discordant AV connections (biventricular) (01.04.01)

ICD10 Derived Terms:

1. Atrioventricular septal defect (Q21.2)
2. Other cardiomyopathies (I42.8)
3. Mirror-image atrial arrangement with situs inversus (Q89.3)
4. Discordant atrioventricular connection (Q20.5)

Commentary

The chosen figures *(see pages 8, 9 and 12*) illustrate, in different specimens, and in echocardiographic images, a particular presentation found in some hearts with Atrioventricular Septal Defects (AVSD) and common atrioventricular junction, the so-called unbalanced forms with a dominant right or left ventricle. The key morphological features of the hearts under discussion are the presence of a common atrioventricular junction, unwedging of the antero-superiorly located aorta, and shortening of the inflow ventricular dimensions so as to produce inlet-outlet septal disproportion. The great variability and complexity to be found in the group occurs due to the diverse morphology of the atrioventricular valvar orifices and leaflets, and to the presence, absence, and dimensions of the communications between the atriums and between the ventricles. Another significant variable found in a small proportion of cases, and discussed here, is the unequal commitment of the common junction to the ventricles, producing the so-called unbalanced Atrioventricular Septal Defects.

More than the simple size, or volume of the ventricular cavities, several criteria have been described in order to define ventricular imbalance in the clinical setting, especially when the primary concern is the achievement of a biventricular surgical correction. When considering the right dominant form of the disease, multiple factors should be taken into account. Of particular importance is, first, the Atrioventricular Valve Index (AVVI), which measures how the valve is apportioned to each ventricle in the subxiphoid echocardiographic view, and helps define imbalance.[1] A second consideration is the Left Ventricular Inflow Index, which estimates how much blood flow enters the left ventricle.[2] The angulation between the inflows to the right and left ventricles then provides another surrogate of flow into the left ventricle.[3] Ahmad and co-workers,[4] using multi-planar echocardiographic interrogations, have also demonstrated recently that the angulation between the septal structures gives important information about the degree of imbalance. As shown in Figure 3, however, septal malalignment is not usually a feature of right ventricular dominance.

Malalignment between the atrial and ventricular septums, in contrast, is the major morphological feature in the setting of hearts with left ventricular dominance. This not only results in a diminished volume of the right ventricle, but also produces an anomalous infero-lateral location of the atrioventricular node, comparable to the situation seen in the setting of straddling tricuspid valve.[5] Since there can be an additional source used for pulmonary blood flow, however, the extent of right ventricular hypoplasia is less likely to influence the chosen surgical approach.

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.

References

1. Jegatheeswaran A, Pisarro C, Caldarone C, Cohen MS, Baffa JM, Gremmels DB, Mertens L, Morell V, Bacha E, Williams W, Blackstone EH, McCrindle BW, Overman D. Echocardiographic Definition of "Unbalanced" in Complete Atrioventricular Septal Defect: A Multi-institutional Study, Circulation 2010;122 [suppl 1]:S209-S215.
2. Szwast AL, Marino BS, Rychik J, Gaynor JW, Spray TL, Cohen MS. Usefulness of left ventricular inflow index to predict successful biventricular repair in right-dominant unbalanced atrioventricular canal. Am J Cardiol 2011;107:103-109.
3. Cohen MS, Jegatheeswaran A, Baffa JM, Gremmels DB, Overman DM, Caldarone CA, McCrindle BW, Mertens L. Echocardiographic features defining right dominant unbalanced atrioventricular septal defect: A multi-institutional study of the Congenital Heart Surgeon's Society. Circulation: Cardiovasc Imag, 2013;6:508-13.
4. Ahmad Z, Lim Z, Roman K, Haw M, Anderson RH, Vettukattil J. The angulation of the septal structures impacts ventricular imbalance in atrioventricular septal defects with a common atrioventricular junction. Cardiol Young. 2016;26(2):321-6.
5. Spicer DE, Anderson RH, Backer CL. Clarifying the surgical morphology of inlet ventricular septal defects. Ann Thorac Surg. 2013; 95: 236-41.

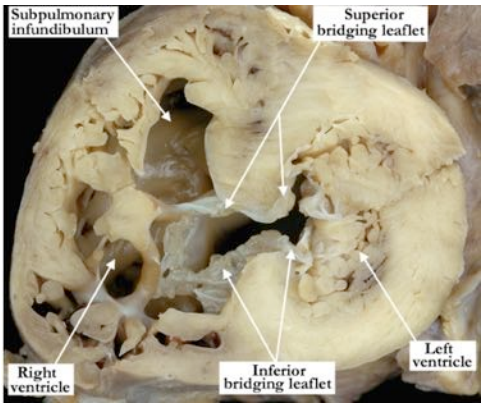


Figure 1.

- ***Common atrioventricular valve with unbalanced commitment of valve to ventricles: dominant right***
- *Modality: Anatomic specimen*
- *Orientation: Apical short axis view*
- *Description: This figure illustrates an unbalanced Atrioventricular Septal Defect with right ventricular dominance. Note the hypoplastic left ventricle and the close approximation of the papillary muscles within the ventricle.*
- *Contributor: Diane E. Spicer, BS*
- *Institution: Congenital Heart Institute of Florida, St. Petersburg & Tampa, FL USA*
- *Reference: ACHavsd*

"The chosen figures (see pages 8, 9 and 12) illustrate, in different specimens, and in echocardiographic images, a particular presentation found in some hearts with Atrioventricular Septal Defects and common atrioventricular junction, the so-called unbalanced forms with a dominant right or left ventricle."

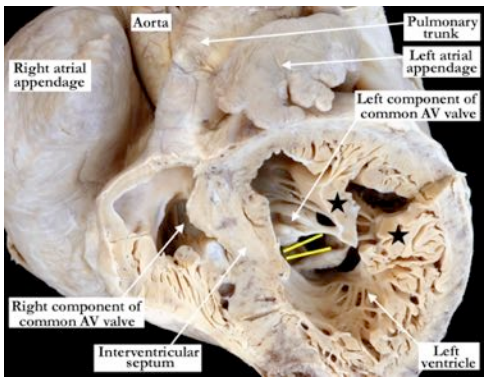


Figure 2.

- ***Common atrioventricular valve with unbalanced commitment of valve to ventricles: dominant left***
- ***Ventricular myocardial noncompaction cardiomyopathy***
- *Modality: Anatomic specimen*
- *Orientation: Apical short axis view*
- *Description: This figure demonstrates an unbalanced Atrioventricular Septal Defect with left ventricular dominance. The papillary muscles (black stars) are close together within the left ventricle and the zone of apposition between the superior and inferior bridging leaflets is marked with the yellow lines. The right ventricular cavity is hypoplastic as is the right component of the common atrioventricular valve. Additionally, there is exaggerated trabeculation, so-called "non-compaction," of the left ventricular myocardium.*
- *Contributor: Diane E. Spicer, BS*
- *Institution: Congenital Heart Institute of Florida, St. Petersburg & Tampa, FL USA*
- *Image source: Van Mierop Archive, University of Florida*
- *Reference: UF67-42*

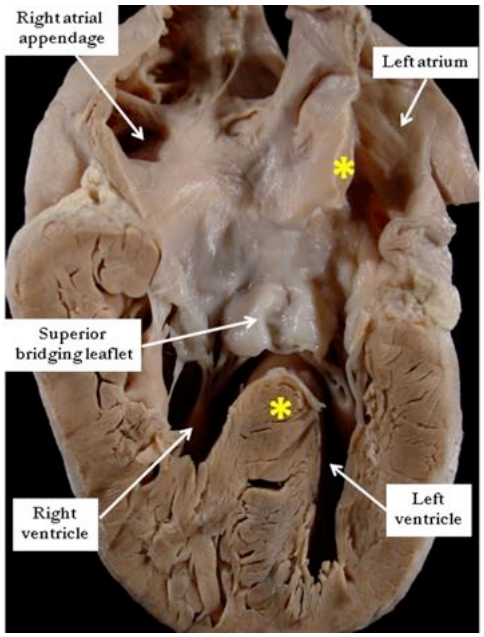


Figure 3.

- ***Mirror-image atrial arrangement (situs inversus)***
- ***Common atrioventricular valve with unbalanced commitment of valve to ventricles: dominant right***
- *Modality: Anatomic specimen*
- *Orientation: Four-chamber view section viewed from behind*
- *Description: This figure demonstrates a heart with mirror-image atrial arrangement (situs inversus), concordant atrioventricular connections and an Atrioventricular Septal Defect with right ventricular dominance. The superior bridging leaflet floats over the ventricular septum. There is alignment between the atrial and ventricular septums (asterisks).*
- *Contributor: Vera D. Aiello*
- *Institution and image source: Heart Institute (InCor), University of São Paulo School of Medicine, Brazil*
- *Reference: DSC03534*

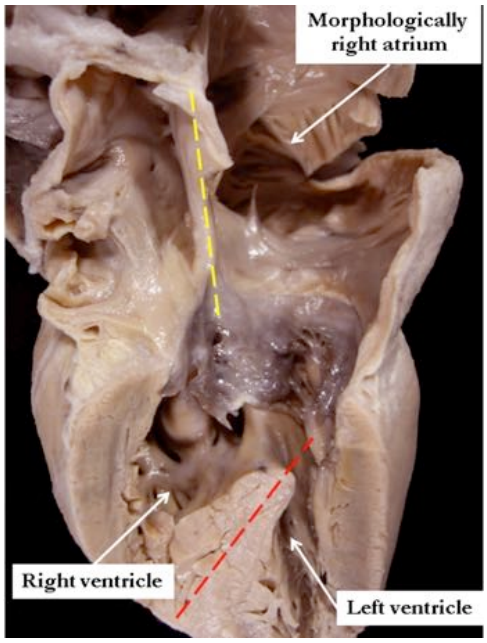


Figure 4.

- ***Discordant atrioventricular connections***
- ***Common atrioventricular valve with unbalanced commitment of valve to ventricles: dominant right***
- *Modality: Anatomic specimen*
- *Orientation: Four-chamber view section, viewed from behind.*
- *Description: This figure demonstrates a heart with usual atrial arrangement (situs solitus) and biventricular, discordant atrioventricular connections. There is gross malalignment between the atrial (yellow dotted line) and ventricular (red dotted line) septums, and the common valve connects predominantly with the left-sided morphologically right ventricle (right ventricular dominance).*
- *Contributor: Vera D. Aiello*
- *Institution and image source: Heart Institute (InCor), University of São Paulo School of Medicine, Brazil*
- *Reference: R0016882*

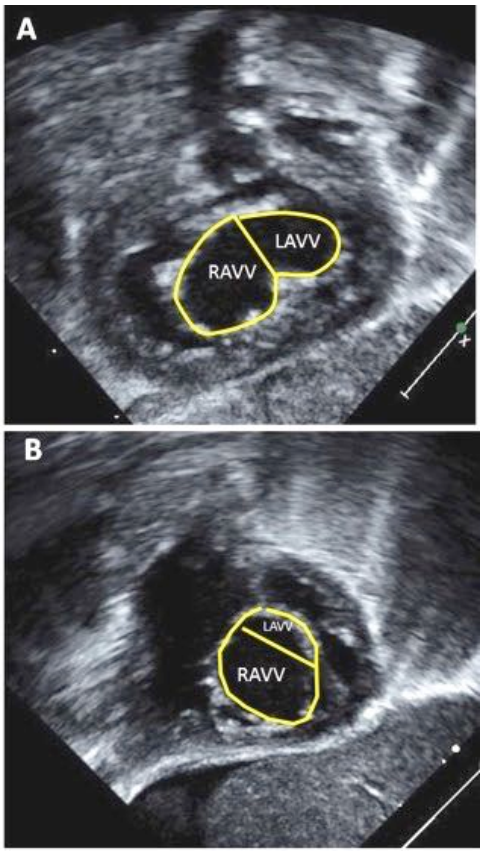


Figure 5.

- ***Measurement of the Atrioventricular Valve Index (AVVI)***
- *Modality: Echocardiographic image*
- *Orientation: Left anterior oblique view of the common AV valve in the open position en face.*
- *Description: The Atrioventricular Valve Index (AVVI) is measured in this view with the valve apportioned to each ventricle.*
  - *A) Mildly unbalanced with AVVI of 0.38*
  - *B) Severely unbalanced with AVVI of 0.25.*
- *RAAV- right atrioventricular valve*
- *LAVV- left atrioventricular valve*
- *Contributor: Meryl S. Cohen*
- *Institution: The Children's Hospital of Philadelphia, Philadelphia, PA*

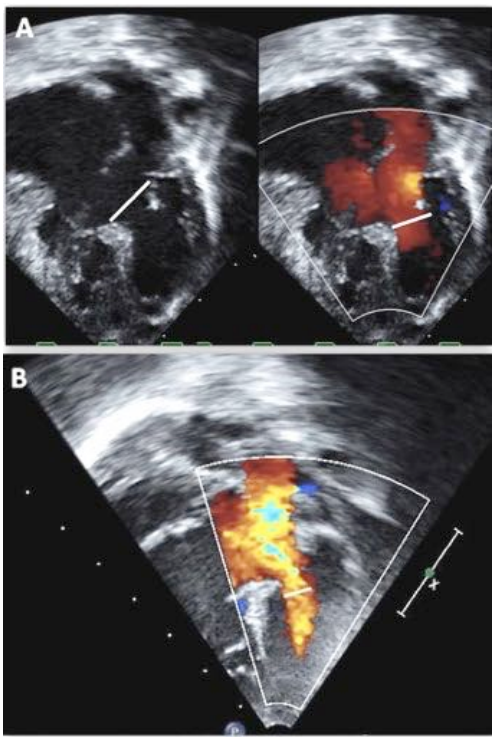


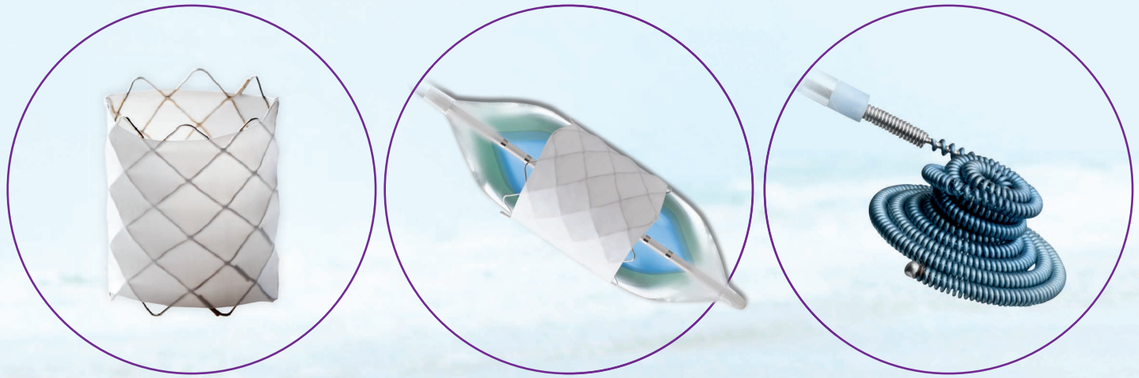
Figure 6.

- ***Measurement of the Left Ventricular Inflow Index (LVVI)***
- *Modality: Echocardiographic images*
- *Orientation: Apical four-chamber view*
- *Description: Measures of LVII (Left Ventricular Inflow Index) in two cases of unbalanced Atrioventricular Septal Defect.*
  - *A) The LVII is 0.65, suggesting that the inflow is adequate for biventricular repair.*
  - *B) Only color inflow is shown in this case but LVII measures 0.5, suggesting that the inflow may be inadequate for biventricular repair.*
- *Contributor: Meryl S. Cohen*
- *Institution: The Children's Hospital of Philadelphia, Philadelphia, PA*

"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."

Interventional Systems
B|BRAUN
SHARING EXPERTISE

MAKING A DIFFERENCE



CP STENT™ UNMOUNTED OR PRE-MOUNTED ON A BIB® CATHETER
FOR TREATMENT OF COARCTATION OF THE AORTA

NIT-OCCLUD® PDA COIL SYSTEM
FOR TREATMENT OF PATENT DUCTUS ARTERIOSUS

NuMED

pfmmedical

INDICATIONS FOR USE:
The CP Stent™ is indicated for use in the treatment of native and/or recurrent coarctation of the aorta involving a compliant aortic isthmus or first segment of the descending aorta where there is adequate size and Patency of at least one Femoral Artery and the balloon angioplasty is contraindicated or predicted to be ineffective. **WARNINGS / PRECAUTIONS:** Coarctation of the aorta involving the aortic isthmus or first segment of the descending aorta should be confirmed by diagnostic imaging. The CP stent has not been evaluated in patients weighing less than 20kg. As with any type of implant, infection secondary to contamination of the stent may lead to aortitis, or abscess. Over-stretching of the artery may result in rupture or aneurysm formation. Crimping the stent on a balloon catheter smaller than 12mm may cause damage to the stent. This device is intended for single use only. Do not resterilize and/or reuse it, as this can potentially result in compromised device performance and increased risk of cross-contamination. **CONTRAINDICATIONS:** Patients too small to allow safe delivery of the stent without compromise to the systemic artery used for delivery. Unfavorable aortic anatomy that does not dilate with high pressure balloon angioplasty. Curved vasculature. Occlusion or obstruction of systemic artery precluding delivery of the stent. Clinical or biological signs of infection. Active endocarditis. Known allergy to aspirin, other antiplatelet agents, or heparin. Pregnancy.

INDICATIONS FOR USE:
The Covered CP Stent™ is indicated for use in the treatment of native and/or recurrent coarctation of the aorta involving the aortic isthmus or first segment of the descending aorta where there is adequate size and patency of at least one femoral artery associated with one or more of the following: Acute or chronic wall injury; Nearly atretic descending aorta of 3 mm or less in diameter; A non-compliant stenotic aortic segment found on pre-stent balloon dilation; A genetic or congenital syndrome associated with aortic wall weakening or ascending aortic aneurysm. **WARNINGS / PRECAUTIONS:** Coarctation of the aorta involving the aortic isthmus or first segment of the descending aorta should be confirmed by diagnostic imaging. The CP stent has not been evaluated in patients weighing less than 20kg. As with any type of implant, infection secondary to contamination of the stent may lead to aortitis, or abscess. Over-stretching of the artery may result in rupture or aneurysm formation. Crimping the stent on a balloon catheter smaller than 12mm may cause damage to the stent. Excessive handling and manipulation of the covering while crimping the stent may cause the covering to tear off of the stent. This device is intended for single use only. Do not resterilize and/or reuse it, as this can potentially result in compromised device performance and increased risk of cross-contamination. **CONTRAINDICATIONS:** Patients too small to allow safe delivery of the stent without compromise to the systemic artery used for delivery. Unfavorable aortic anatomy that does not dilate with high pressure balloon angioplasty. Curved vasculature. Occlusion or obstruction of systemic artery precluding delivery of the stent. Clinical or biological signs of infection. Active endocarditis. Known allergy to aspirin, other antiplatelet agents, or heparin. Pregnancy.

INDICATIONS FOR USE:
The Nit-Occlud® PDA coil is a permanently implanted prosthesis indicated for percutaneous, transcatheter closure of small to moderate size patent ductus arteriosus with a minimum angiographic diameter less than 4mm. Nit-Occlud Brief Statement: Do not implant the Nit-Occlud PDA into patients who have endocarditis, endarteritis, active infection, pulmonary hypertension (calculated PVR greater than 5 Wood Units), thrombus in a blood vessel through which access to the PDA must be obtained, thrombus in the vicinity of the implantation site at the time of the implantation or patients with a body weight < 11 lbs (5 kg). An angiogram must be performed prior to implantation for measuring length and diameter of the PDA. Only the pfm medical implantation delivery catheter should be used to implant the device. Administration of 50 units of heparin per kg body weight should be injected after femoral sheaths are placed. Antibiotics should be given before (1 dose) and after implantation (2 doses) in order to prevent infection during the implant procedure. Do not implant the Nit-Occlud PDA in an MR environment. Do not pull the Nit-Occlud coil through heart valves or ventricular chambers. Contrast media should not be injected through the implantation catheter. The catheter must not be connected to high pressure injectors. Patients may have an allergic response to this device due to small amounts of nickel that has been shown to be released from the device in very small amounts. If the patient experiences allergic symptoms, such as difficulty in breathing or swelling of the face or throat, he/she should be instructed to seek medical assistance immediately. Antibiotic prophylaxis should be performed to prevent infective endocarditis during first 6 months after coil implantation. Potential Adverse Events: Air embolism, Allergic reaction to drug/contrast, Apnea, Arrhythmia requiring medical treatment or pacing, Arteriovenous fistula, Bacterial endocarditis, Blood loss requiring transfusion, Chest pain, Damage to the tricuspid or pulmonary valves, Death, Embolization of the occluder, requiring percutaneous or surgical intervention, Endarteritis, False aneurysm of the femoral artery, Fever,Headache/migraine, Heart failure, Hemolysis after implantation of the occluder, Hypertension, Hypotension or shock, Infection, Myocardial infarction, Occluder fracture or damage, Perforation of the heart or blood vessels, Stenosis of the left pulmonary artery or descending thoracic aorta, Stroke/TIA, Thromboembolism (cerebral or pulmonary), Valvular Regurgitation, Vessel damage at the site of groin puncture (loss of pulse hematoma, etc.).

Refer to the Instructions for Use for complete indications, relevant warnings, precautions, complications, and contraindications.

Rx only CV9058 - 9/16

Distributed by:
B. Braun Interventional Systems Inc. | 824 Twelfth Avenue | Bethlehem, PA 18018 USA
Tel 877 836 2228 | Fax 610 849 1334 | www.bisusa.org

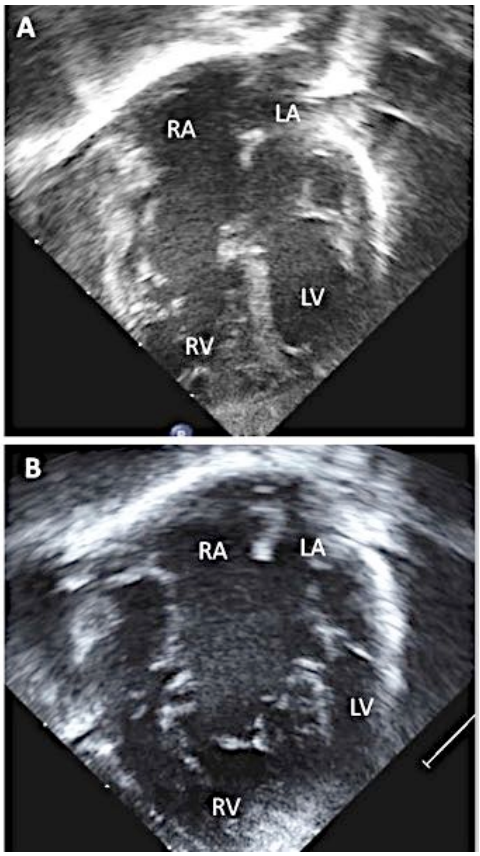


Figure 7.

- ***Ventricular unbalance with left ventricular hypoplasia***
- *Modality: Echocardiographic images*
- *Orientation: Apical four-chamber view.*
- *Description: The images are from two patients with unbalanced Atrioventricular Septal Defect.*
  - *A) The patient has mild unbalance at the ventricular level with mild left ventricular hypoplasia.*
  - *B) The patient has severe unbalance at the ventricular level with severe left ventricular hypoplasia.*
- *RA- right atrium; RV- right ventricle; LA-left atrium; LV-left ventricle.*
- *Contributor: Meryl S. Cohen*
- *Institution: The Children's Hospital of Philadelphia, Philadelphia, PA*

The authors would like to acknowledge the Children's Heart Foundation www.childrensheartfoundation.org

CCT

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

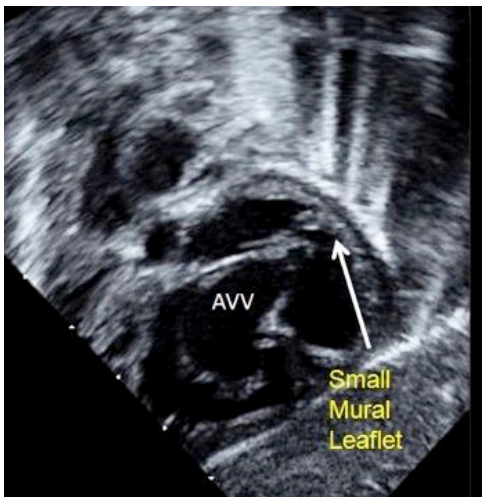


Figure 8.

- ***Small mural leaflet in unbalanced Atrioventricular Septal Defect***
- *Modality: Echocardiographic images*
- *Orientation: Subxiphoid left anterior oblique view*
- *Description: The figure demonstrates a common finding in unbalanced Atrioventricular Septal Defect, namely a small left-sided mural leaflet.*
- *AVV = atrioventricular valve*
- *Contributor: Meryl S. Cohen*
- *Institution: The Children's Hospital of Philadelphia, Philadelphia, PA*

Jeffrey P. Jacobs, MD
Archiving Working Group
Professor of Surgery, Cardiovascular and Thoracic Surgeon, Johns Hopkins Children's Heart Surgery,
All Children's Hospital and Florida Hospital for Children
Saint Petersburg, FL USA

James D. St. Louis, MD
Archiving Working Group
Professor and Surgical Director, Cardiac Transplantation
University of Missouri-KC School of Medicine
Children's Mercy Hospital
Kansas City, MO USA

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

Charles W. Shepard, MD
Archiving Working Group
University of Minnesota Amplatz Children's Hospital, Department of Pediatrics,
Minneapolis, MN USA

Allen D. Everett, MD
Archiving Working Group
Johns Hopkins University Helen B. Taussig Heart Center
Charlette R. Bloomberg Children's Center
Baltimore, MD USA

Robert Anderson, MD
Co-Chairman, Archiving Working Group
Institute of Genetic Medicine, Newcastle University, Newcastle upon Tyne, UK

Corresponding Author

Vera D. Aiello, MD
Co-Chairman, Archiving Working Group
Heart Institute (InCor), São Paulo University School of Medicine,
São Paulo, Brazil
email: anpvera@incor.usp.br

Meryl S. Cohen, MD
Archiving Working Group
The Children's Hospital of Philadelphia
Philadelphia, PA USA

And the members of the Archiving Working Group of the International Society for Nomenclature of Paediatric and Congenital Heart Disease

E-mail comments to:
CCT-Feb17-AWG@CCT.bz

Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

Imprecise Diagnoses - Genetic Tests for Potentially Fatal Heart Anomaly Can Misdiagnose Condition in Black-Americans

Newswise — Genetic testing has greatly improved physicians' ability to detect potentially lethal heart anomalies among asymptomatic family members of people who suffer cardiac arrest or sudden cardiac death.

But a study from Harvard Medical School published in the Aug. 18th issue of *The New England Journal of Medicine* shows that over the last decade these lifesaving tools may have disproportionately misdiagnosed one cardiac condition — Hypertrophic Cardiomyopathy (HCM) – in Black-Americans.

HCM, which affects one in 500 people, is an often-asymptomatic thickening of the heart muscle that can spark fatal arrhythmias in seemingly healthy young adults.

Get More HMS News

The notion that genetic tests could misread benign genetic alterations as disease-causing mutations is not entirely new, but this study is believed to be the first one to trace the root of the problem to racially biased methodologies in early studies that defined certain common genetic variants as causes of HCM.

Indeed, the analysis reveals that in the case of HCM, the false positive diagnoses stemmed from inadequately designed clinical studies that used predominantly white populations as control groups.

White Americans harbor far fewer benign mutations on several genes implicated in HCM than Back-Americans. The higher rate of benign alterations in the latter group can cause test results to be misread as abnormal, the researchers say.

Using statistical simulations, the HMS team demonstrated that including even small numbers of Black participants in the original studies would have improved test accuracy and, consequently, helped avert some of the false-positive diagnoses.

The findings, the researchers say, highlight the importance of interpreting genetic test results against diverse control populations to ensure that normal variations of genetic markers common in one racial or ethnic group do not get misclassified as disease-causing in another.

The team says their findings point to a pressing need to reevaluate decades-old genetic studies by using new racially diverse sequencing data.

"We believe that what we're seeing in the case of Hypertrophic Cardiomyopathy may be the tip of the iceberg of a larger problem that transcends a single genetic disease," said study first author Arjun Manrai, a research fellow in the Department of Biomedical Informatics at Harvard Medical School. "We hope our study motivates a systematic review of this issue across other genetic conditions.

"Ensuring that Genomic Medicine benefits all people and all populations equally is nothing short of a moral imperative, not only for scientists and clinicians but for political and health policy powers that be" —Zak Kohane

Aside from the emotional toll that a genetic misdiagnosis can take on individuals and families, the researchers say their findings represent a cautionary tale with a broad relevance to geneticists, clinicians and policy-makers alike.

"Our study powerfully illustrates the importance of racial and ethnic diversity in research," says Zak Kohane, Senior Investigator on the study and Chair of the Department of Biomedical Informatics at Harvard Medical School. "Racial and ethnical inclusiveness improves the validity and accuracy of clinical trials and, in doing so, can better guide clinical decision-making and choice of optimal therapy. This is the essence of Precision Medicine."

In the current study, the team analyzed more than 8,000 DNA samples stored in three national databases — the National Institutes of Health's Mendelian Exome Sequencing Project, the 1000 Genomes Project and the Human Genome Diversity Project.

Five genetic variants — each of them benign — accounted for 75% of all genetic variation across populations. However, the team found, these five mutations occurred disproportionately in Black-Americans.

Between 2.9% and 27% of Black-Americans harbored one or more such variants, compared with 0.02% to 2.9% of White-Americans.

Next, researchers examined records of more than 2,000 patients and family members tested at a leading genetic laboratory between 2004 and 2014. Seven patients received reports indicating they harbored disease-causing mutations that were subsequently reclassified as benign. Five of the seven patients were black, and two were of unspecified ancestry.

Researchers say it remains unclear how many of the seven patients had been re-contacted to communicate the change in test results.

The investigators caution that test results from a single genetic lab are not necessarily representative of the scope of the problem nationally, but say their findings likely point to a discrepancy that goes beyond a single laboratory and a single condition.

To trace the root of the misclassifications, researchers reviewed the five original studies that shaped early understanding of genetic variants and their role in the development of HCM. All of them, the researchers found, analyzed small population sizes, and none, indicated that black people were included in the control groups.

But, the investigators add, even small studies can avert misclassification of genetic variants as long they include racially diverse populations.

Using statistical simulation, the team showed that a sample of 200 people that included 20 Black participants would have only 50% chance of correctly ruling out a harmful mutation. The same sample would have more than an 80% accuracy, if a third of patients were black, and more than 90% accuracy, if half of them were black.

Investigators say the newly created Exome Aggregation Consortium — a compilation of data from various large-scale sequencing projects that includes DNA from more than 60,000 individuals — is well-powered to discern between harmful and benign mutations even for relatively rare genetic variants, and should help in the re-analysis of decades-old data.

The latest clinical guidelines urge physicians to interpret genetic test results by cross-referencing them against racially matched controls. However, with expanding efforts to sequence DNA from various ethnic and racial groups, researchers say more genetic variants will be reclassified in the next decade. Interpreting the meaning of test results within the context of such rapidly evolving knowledge will pose a serious challenge for clinicians.

One way to address the problem, the HMS team says, could be the use of point-of-care risk calculators to help clinicians and genetic counselors

more precisely gauge the significance of a given genetic variant. Such risk calculators would use algorithms that incorporate statistical probability, race, ethnicity and family history to help sift variant noise from truly pathogenic mutations.

"Ensuring that Genomic Medicine benefits all people and all populations equally is nothing short of a moral imperative, not only for scientists and clinicians, but for political and health policy powers that be," Kohane said.

The work was funded by the National Human Genome Research Institute under Grant 5T32HG002295-9, by the National Institute of Mental Health under Grant P50MH094267 and by the National Centers for Biomedical Computing under grant 5U54-LM-008748.

Other investigators on the research included: Birgit Funke, PhD; Heidi Rehm, PhD; Morten Olesen, PhD; Bradley Maron, MD; Peter Szolovits, PhD; David Margulies, MD; Joseph Loscalzo, MD, PhD.

Tiny Electronic Device Can Monitor Heart, Recognize Speech

Researchers from the University of Colorado Boulder and Northwestern University have developed a tiny, soft and wearable acoustic sensor that measures vibrations in the human body, allowing them to monitor human heart health and recognize spoken words.

The stretchable device captures physiological sound signals from the body, has physical properties well-matched with human skin and can be mounted on nearly any surface of the body, said CU Boulder Assistant Professor Jae-Woong Jeong, one of three lead study authors. The sensor, which resembles a small Band-Aid, weighs less than one-hundredth of an ounce and can gather continuous physiological data.

"This device has a very low mass density and can be used for cardiovascular monitoring, speech recognition and human-machine interfaces in daily life," said Jeong of the Department of Electrical, Computer and Energy Engineering. "It is very comfortable and convenient – you can think of it as a tiny, wearable stethoscope."

A paper on the subject was published Nov. 16th in *Science Advances*, a sister journal of *Science*. The other two co-corresponding

authors are Professors Yonggang Huang and John Rogers of Northwestern.

Listening in On the Body

"The thin, soft, skin-like characteristics of these advanced wearable devices provide unique capabilities for 'listening in' to the intrinsic sounds of vital organs of the body, including the lungs and heart, with important consequences in continuous monitoring of physiological health," said Rogers, the Simpson Querrey Professor of Materials Science and Engineering, Biomedical Engineering and Neurological Surgery. Rogers also is director of Northwestern's Center for Bio-Integrated Electronics.

The researchers say the new device can pick up mechanical waves that propagate through tissues and fluids in the human body due to natural physiological activity, revealing characteristic acoustical signatures of individual events. They include the opening and closing of heart valves, vibrations of the vocal cords and even movements in gastrointestinal tracts.

The sensor can also integrate electrodes that can record electrocardiogram (ECG) signals that measure the electrical activity of the heart, as well electromyogram (EMG) signals that measure the electrical activity of muscles at rest and during contraction.

While the sensor was wired to an external data acquisition system for the tests, it can easily be converted into a wireless device, said Jeong. Such sensors could be of use in remote, noisy places – including battlefields – producing quiet, high-quality cardiology or speech signals that can be read in real time at distant medical facilities.

"Using the data from these sensors, a doctor at a hospital far away from a patient would be able to make a fast, accurate diagnosis," said Jeong.

Talking to Machines

Vocal cord vibration signals also could be used by the military personnel or civilians to control robots, vehicles or drones. The speech recognition capabilities of the sensor also have implications for improving communication for people suffering from speech impairments, he said.

As part of the study, the team used the device to measure cardiac acoustic responses and ECG activity – including the detection of heart murmurs – in a group of elderly volunteers at Camp Lowell Cardiology, a private medical clinic in Tucson, Arizona collaborating with the University of Arizona, a project partner. The researchers also were able to detect the acoustical signals of blood clots in a related lab experiment, said Jeong.

Other CU Boulder study co-authors on the Science Advances paper include: Assistant Professor Jianliang Xiao and doctoral student Zhanan Zou of Mechanical Engineering and doctoral student Raza Qazi of Electrical Engineering.

The sticky, flexible polymer encapsulating the tiny device is stretchable enough to follow skin deformation, said study first author Yuhao Liu, who earned his doctorate at the University of Illinois-Urbana Champaign and now works at the Lam Research, headquartered in Fremont, California. The device contains a tiny commercial accelerometer to measure the vibration of the body acoustics, and allows for the evaporation of human sweat.

The researchers also showed vocal cord vibrations gathered when the device is on one's throat can be used to control video games and other machines. As part of the study a test subject was able to control a Pac-Man game using vocal cord vibrations for the words "up," "down," "left" and "right."

"While other skin electronics devices have been developed by researchers, what has not been demonstrated before is the mechanical-acoustic coupling of our device to the body through the skin," Jeong said. "Our goal is to make this device practical enough to use in our daily lives."

The study also included the Eulji University College of Medicine in Korea.

Unroofing Surgery Relieves Debilitating Symptoms of Heart Anomaly, Study Finds

A Stanford study shows that a type of surgery improves the quality of life for patients with myocardial bridging, a congenital condition caused by a major artery tunneling through heart muscle.

A little-known heart anomaly often confounds doctors, who don't know how to treat it. The condition results when a major artery runs through the muscle of the heart rather than resting on top of the organ.
Aksabir/Shutterstock

In 2010, Ingela Schnittger, MD, a cardiologist at the Stanford University School of Medicine, sat in her lab examining the echocardiogram of a young man who came to the heart clinic at Stanford Health Care complaining of chest pain. She spotted a curious motion of the heart on the computer screen, one that she'd seen before while examining these kinds of diagnostic tests.

"All of the sudden I had this flashback," she said. She remembered a young Physics professor at another institution who had suddenly dropped dead of a heart attack while running on a treadmill. During an autopsy, a little-known heart anomaly called a myocardial bridge was found. The term describes a condition in which a major artery runs through the muscle of the heart rather than resting on top of the organ.
"I was thinking, 'Wow, I wonder if this patient could have a myocardial bridge?'" she said. Six years later, Schnittger, Professor of Cardiovascular Medicine, has co-authored four research studies on myocardial bridges. A fifth, published online Oct. 13th in the *Annals of Thoracic Surgery,* finds that a procedure called surgical unroofing is safe and provides significant relief for patients with myocardial bridges who have incapacitating symptoms, such as chest pain, arrhythmias and fatigue, that are not helped by medication alone.

Schnittger is Senior Author of the study, and the Lead Author is Jack Boyd, MD, Clinical Assistant Professor of Cardiothoracic Surgery.

Long Considered Bbenign

Myocardial bridging remains a mystery to much of the medical community. It's a congenital anomaly that was discovered during autopsies almost 300 years ago, but it has long been considered benign, the study said.

Bridging continues to be little understood and is often misdiagnosed, Boyd said.

Ingela Schnittger

"It's not taught in medical school, and there is no agreed-upon treatment," he said.

That lack of understanding about the condition is what sent Schnittger along her path of investigation. She wanted to know more: Could bridging be dangerous? Did it cause symptoms? How should it be treated?

In 2011, Schnittger designed a study at Stanford to enroll patients with undiagnosed chest pain and examine them in the catheterization laboratory using diagnostic-imaging techniques, such as angiograms, to visualize the arteries of the heart and its chambers.

"Sometimes we saw myocardial bridging on the angiogram," she said. "Then we'd use an intravascular ultrasound to see them more clearly."

Since then, 150 participants in this ongoing study have been diagnosed with myocardial bridges. Research has involved measuring blood flow and blood pressure in the bridging vessel. Schnittger and her colleagues were able to show that compression of the heart muscle can reduce or cut off blood flow in the artery, resulting in serious problems, including angina, myocardial ischemia, acute coronary syndrome, left ventricular dysfunction and malignant ventricular arrhythmias.

For some, Symptoms Are Severe

"It hadn't before been proven with blood-flow studies that circulation got impaired enough to cause ischemia, inadequate blood flow and oxygen to the heart muscle," Schnittger said.

Many of these Patients Have These Heartbreaking Stories to Tell.

Most patients with the condition remain asymptomatic. Some have minor symptoms that can be controlled with medication, such as beta blockers and calcium-channel blockers. But a small portion have severe symptoms that greatly affect their daily lives. Some are left homebound. Many make repeated trips to emergency rooms complaining of heart-attack-like symptoms, only to get sent home with no answers.

"Many of these patients have these heartbreaking stories to tell," Schnittger said. "They can't hold a job, they can't travel, they can't take care of their families. Most cardiologists are completely at a loss. They know myocardial bridges exist, but they have been taught they are benign and never cause problems.

"When these patients go to the ER, and they go there a lot, all the cardiology tests come back normal. They're told, 'Here's a little Valium. I think you're anxious.' They get belittled, not taken seriously, and they get really depressed."

Unroofing

Unroofing the myocardial bridge entails cutting through the heart muscle to uncover the tunneled artery, thus relieving compression on the artery caused by the bridging.

The surgery is known to be effective, Boyd said. However, concerns that healthy heart muscle could be damaged during the operation have slowed its adoption. "At Stanford, we use new imaging techniques to map the bridge muscle very precisely, and we perform the unroofing with conservative surgical techniques to safeguard the healthy heart muscle," he said.

In the past, the surgery was done only as a treatment of last resort, Schnittger said. "You took a patient very, very occasionally to surgery when everybody had tossed their hands up," she said. "Stanford probably did no more than one surgery a decade in the past."

But with Schnittger and her team, including study co-author Jennifer Tremmel, MD, Assistant Professor of Cardiovascular Medicine, and Ian Rogers, MD, Clinical Assistant Professor of Cardiovascular Medicine and of Pediatric Cardiology, helping to guide the surgeries, the number performed at Stanford Medicine has increased substantially. More than 80 have been performed at SHC and Lucile Packard Children's Hospital Stanford in the past five years.

For the new study, the researchers examined 50 adult patients who between 2011-15 underwent the unroofing procedure because of severe symptoms that medication had failed to manage. The first 35 were performed using a heart-lung machine for life support. The last 15 were performed off-pump on a beating heart.

Studying the Condition in the Lab

"We studied the patients' hearts condition in the lab, proving there was ischemia when we simulated a stress test by infusing drugs that increased the heart rate," Schnittger said. "Anxiety, sleep deprivation — anything that drives up the heart rate would also cause ischemia."

Prior to surgery, study participants filled out the Seattle Angina Questionnaire, a three-page survey describing their symptoms and ranking their quality of life. Then six months after the surgery, they filled out the survey again.

Our team wants to spread the word, to educate the medical community that this is a real thing.

This is a comprehensive survey often used in cardiac research to ask how much pain the patient has, how frequently they have it, how much it limits their life.

Results showed that the average ranking of quality of life prior to surgery by the patients was 25%. After surgery, that figure jumped to 78%.

"Our patients come back after surgery so grateful," Schnittger said. "They have suffered for so long — finall, they've found doctors who understand them.

"One patient, a mother of five, was so symptomatic before surgery she had arranged her whole life around her symptoms. She didn't play with her children outside; she couldn't even pick them up at the playground. She was basically housebound. She couldn't do laundry or go grocery shopping. After surgery, she could do all those things — laundry, play outside with her children, go for a walk. It's a life-changer."

"Our team wants to spread the word, to educate the medical community that this is a real thing," she added, noting that a myocardial bridge clinic has been established at Stanford Health Care.

The work is an example of Stanford Medicine's focus on precision health, the goal of which is to anticipate and prevent disease in the healthy and precisely diagnose and treat disease in the ill.

Other Stanford co-authors are: postdoctoral scholar Vedant Pargaonkar, MD; resident David Scoville, MD; Takumi Kimura, MD, PhD; Shigemitsu Tanaka, MD, PhD, and Ryotaro Yamada, MD, PhD, all members of the Stanford University Cardiovascular Core Analysis Laboratory; Michael Fischbein, MD, PhD, Associate Professor of Cardiothoracic Surgery; and R. Scott Mitchell, MD, Professor Emeritus of Cardiothoracic Surgery.

New Report Outlines Ten Measures for the Prevention of Sudden Cardiac Death

A new report presents 10 quality and performance measures that are intended to help stakeholders—including health systems, legislative bodies, and nongovernmental organizations, as well as healthcare practitioners, patients, families and communities—in the effort to prevent Sudden Cardiac Death. The joint report from the American College of Cardiology and the American Heart Association was published online recently in the *Journal of the American College of Cardiology and Circulation: Cardiovascular Quality and Outcomes.*

Sudden cardiac death is an unexpected death due to the sudden cessation of cardiac activity, which is also known as sudden cardiac arrest. The two phrases are often used interchangeably; however, Sudden Cardiac Death (SCA) should only be used to describe fatal events.

"This is the first comprehensive measure set in the area of Sudden Cardiac Death prevention," said Sana Al-Khatib, MD, FACC, Co-Chair of the Writing Committee and Professor of Medicine at Duke University. "Our vision is that these measures will be developed, tested, and implemented in clinical practice and that implementation will improve patient care and outcomes."

In the United States, approximately 356,500 out-of-hospital cardiac arrests occur each year. Many of the sudden deaths occurring in the United States may be prevented by implementing evidence-based and guideline-endorsed recommendations for primary or secondary prevention of sudden cardiac death. However, sudden cardiac death can occur in people who do not appear to be at risk for this outcome and accurate risk stratification is not achievable in most people.

"While some people—such as patients with heart failure—are known to be at risk of sudden cardiac death, others are not. We need initiatives to improve the quality of care for those with a known risk, but also for the victims of sudden cardiac arrest," Al-Khatib said.

Sudden Cardiac Arrest is one of the leading causes of death in the United States. Even when a patient survives, the condition may have a devastating impact on the patient's quality and length of life. This clinical outcome also imposes a heavy economic burden through healthcare costs.

Although guidelines exist for the prevention of sudden cardiac death, there has been an underutilization of public health initiatives, treatments and device therapies for at-risk patients. The writing committee attempted to identify performance measures that can assess the quality of care for the prevention of sudden cardiac death. Although sudden cardiac arrest and sudden cardiac death can affect people of all ages and demographics, the performance measures focus on adults. No limitations or restrictions for other demographics, such as sex, race/ethnicity, or socioeconomic status, were applied.

The performance measure set is intended to assist clinicians and help them provide better care for their patients at risk of SCA, and thereby, to improve care and outcomes.

Performance and Quality Measures for the Prevention of Sudden Cardiac Death

- Smoking cessation intervention in patients who suffered SCA, have a life-threatening ventricular arrhythmia, or are at risk for sudden cardiac death.
- Screening for Family History of Sudden Cardiac Death
- Screening for asymptomatic left ventricular dysfunction among individuals who have a strong family history of cardiomyopathy and sudden cardiac death.
- Referring for Cardiopulmonary Resuscitation (CPR) and Automated External Defibrillator (AED) education those family members of patients who are hospitalized with known heart conditions that increase the risk of Sudden Cardiac Arrest.
- Use of Implantable Cardioverter Defibrillators (ICDs) for prevention of SCA in patients with heart failure and reduced ejection fraction who have an anticipated survival of more than one year.
- Use of guideline-directed medical therapy for prevention of sudden cardiac death in patients with heart failure and reduced ejection fraction
- Use of guideline-directed medical therapy for prevention of Sudden Cardiac Death in patients with heart attack and reduced ejection fraction.
- Documenting the absence of reversible causes of ventricular tachycardia/ventricular fibrillation cardiac arrest and/or sustained ventricular tachycardia before a secondary-prevention ICD is placed
- Counseling Eligible Patients about an ICD
- Counseling first-degree relatives of survivors of SCA associated with an inheritable condition about the need for screening for the inheritable condition.

For more information on The American College of Cardiology, visit acc.org.

To learn more about The American Heart Association, visit heart.org.

New Sensor System Predicts Heart Failure Events

Newswise – A suite of sensors can predict heart failure events by detecting when a patient's condition is worsening, according to Dr. John Boehmer, Professor of Medicine, Penn State College of Medicine, who presented the findings at the *American Heart Association* annual meeting in New Orleans.

Heart failure is responsible for more than 1 million hospitalizations each year and more than $20 billion in costs. The new technique could help prevent costly hospitalizations and poor health outcomes including death.

Current efforts to manage heart failure by monitoring weight and symptoms have not significantly reduced hospitalizations. More than one in five patients are readmitted within 30 days after being hospitalized for heart failure.

An international team of researchers set out to investigate if implantable devices already used in heart failure patients could be retrofitted with sensors to track their condition. Their results werel also published in *JACC Heart Failure.*

Nine hundred heart failure patients were followed for up to one year. At the beginning of the study, the researchers uploaded software to each patient's implanted defibrillator, a battery-powered device that delivers an electric shock if the patient's heart stops beating.

The software allowed the defibrillators to also act as sensors, monitoring the patients' heart rate, activity, breathing, heart sounds and electrical activity in the chest.

Over the study period, the suite of sensors detected 70% of heart failure events in patients. This detection was often more than a month before the events occurred. Sensitivity at this level far exceeded the researchers' goal of greater than 40% detection.

While there were false positives, the number was within an acceptable range.

"If you're going to monitor a hundred patients, it becomes a fairly manageable number of alerts that you have to deal with," said Boehmer, a cardiologist at Penn State Health Milton S. Hershey Medical Center.

Boston Scientific developed the system and funded the study. They named the system HeartLogic.

"This is a new and clinically valuable measure of worsening heart failure, and it combines a number of measures of the physiology and heart failure much like a doctor will look at a patient," Boehmer said. "Doctors look at all their signs and symptoms, get some tests and put it all together and make a decision about how well or ill the patient is. HeartLogic does it similarly. It integrates a number of measurements of what's going on with the patient, including breathing, activity and heart sounds, and puts that all together to give us an index that we believe is both sensitive and specific for heart failure."

Boehmer said the technology can help monitor the patient's condition so heart failure events can be prevented before they happen.

"It's like having high blood sugar if you're managing diabetes," Boehmer explained. "The doctor doesn't need to know about every high blood sugar and every high blood sugar doesn't result in a hospitalization. But you want to treat it before it gets very high and the patient becomes so symptomatic they become ill and end up in the hospital. This is the same concept."

A pilot study and intervention trials to test the system's safety, physician acceptance and use and patient outcomes are planned to investigate benefits to patients.

Other researchers on this project were: Ramesh Hariharan, University of Texas Physicians, EP Heart, Houston, TX; Fausto G. Devecchi, Cardiac Arrhythmia Service, Lutheran Health Network, Fort Wayne, IN; Andrew L. Smith, Emory University, Atlanta, GA; Giulio Molon, Cardiology Dept, Sacro Cuore Hospital, Negrar, Italy; Alessandro Capucci, Università Politecnica delle March, Ancona, Italy; Qi An, Viktoria Averina, Craig M. Stolen, Pramodsingh H. Thakur, Julie A. Thompson, Ramesh Wariar and Yi Zhang, all at Boston Scientific, St. Paul, MN; and Jagmeet P. Singh, Massachusetts General Hospital Heart Center, Boston, MA.

Radiology Professionals Connect at RSNA 2016

The Radiological Society of North America's 102nd Scientific Assembly and Annual Meeting (*RSNA 2016*), held in Chicago's McCormick Place from November 27th to December 2nd, highlighted RSNA's ongoing commitment to improve patient care through radiology education, research and technological innovation.

"The 2016 theme was 'Beyond Imaging,'" said RSNA Executive Director, Mark Watson. "The meeting offered attendees a wealth of opportunities to broaden their perspectives on both the current state of radiology and the challenges and opportunities the future holds."

RSNA 2016 featured a number of hot topics in radiology including machine learning and 3D printing for medical applications.

Advances in machine learning and artificial intelligence offer an exciting suite of technologies that are now being applied to medical imaging with compelling results. *RSNA 2016* provided a variety of events related to machine learning, including scientific and education sessions, a hands-on workshop and the "Eyes of Watson" interactive demonstration of IBM's Watson technology platform.

To showcase the increasing clinical significance of 3D printing and its connection to medical imaging, in addition to many hands-on courses, the Learning Center included presentations on 3D printing along with demonstrations throughout the week.

Throughout the week at McCormick Place, professional attendees could choose from an abundance of scientific presentations, education courses, plenary sessions, education exhibits and scientific posters, as well as special presentations and performances at the Discovery Theater, part of the new RSNA Connections Center.

Upcoming Medical Meetings

Adult Congenital Heart Disease Quality Care in the Era of Certication and Accreditation – A Practical Approach
Feb. 10-11, 2017; Los Angeles, CA USA
www.congenitalcardiologytoday.com/Ads/2016_ACHD_Brochure.pdf

Cardiology 2017
Feb. 22-26, 2017; Orlando, FL USA
www.chop.edu/events/cardiology-2017#.V-WXtaO-L5U

CSI Asia-Pacific 2017 Catheter Interventions in Congenital, Structural & Valvular Heart Disease
Mar. 2 - 4, 2017; Bangkok, Thailand
www.csi-congress.org/csi-asia-pacific.php

NeoHeart: Cardiovascular Management of the Neonate
Mar. 22-25, 2017; San Diego, CA USA
www.choc.org/neoheart

51st AEPC Annual Meeting
Mar. 29 - Apr. 1 2017; Lyon, France
www.aepc2017.org/en/

Catheter Interventions in Congenital, Structural & Valvular Heart Disease
Jun. 28 - Jul. 1, 2017; Frankfurt, Germany
csi-congress.org

7th World Congress of Pediatric Cardiology & Cardiac Surgery
Jul. 16 - 21, 2017; Barcelona, Spain
wcpccs2017.org/en

27th International Symposium on Adult Congenital Heart Disease
Sep. 14-16, 2017; Cincinnati, OH USA
www.cincinnatichildrens.org/ACHDsymposium

"A highly visible new feature for 2016 was the Connections Center," Watson said. "The RSNA Services area was completely redesigned to enhance the attendee experience. The Connections Center provided not only important RSNA services, but also entertainment, networking functions, expanded lounge seating and digital support."

The technical exhibition was 412,000 square feet and housed 663 exhibits, including 104 first-time exhibitors.

The *RSNA 2016 Virtual Meeting* offered attendees on-demand access to more than 90 sessions from RSNA 2016, as well as Cases of the Day, digital education exhibits and scientific posters. The virtual meeting attracted 5,645 attendees from 98 countries.

Official registration figures for R*SNA 2016* reported total registrant numbers at 54,037, including 26,988 professional registrants of which 1,813 were virtual-meeting only.

RSNA 2017, RSNA's 103rd Scientific Assembly and Annual Meeting, will take place November 26th to December 1st in Chicago. The meeting's theme is "Explore. Invent. Transform."

E-mail comments or questions medical news to: CCT-Feb17-News@CCT.bz

CONGENITAL CARDIOLOGY TODAY

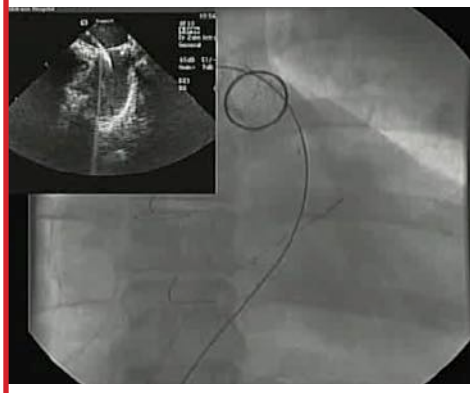
Publication Company Address:
11502 Elk Horn Dr. Ste. 201
Clarksburg, MD 20871 USA
Tel: +1.301.279.2005

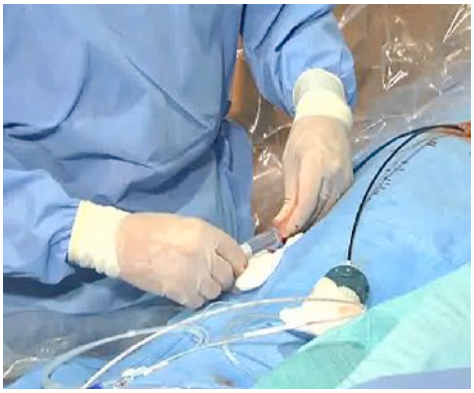
Publishing Management:
- Tony Carlson, Founder, President & Sr. Editor - *TCarlsonmd@gmail.com*
- Richard Koulbanis, Group Publisher & Editor-in-Chief - *RichardK@CCT.bz*
- John W. Moore, MD, MPH, Group Medical Editor - *JMoore@RCHSD.org*
- Allan Berthe, Contributing Editor-Special Projects

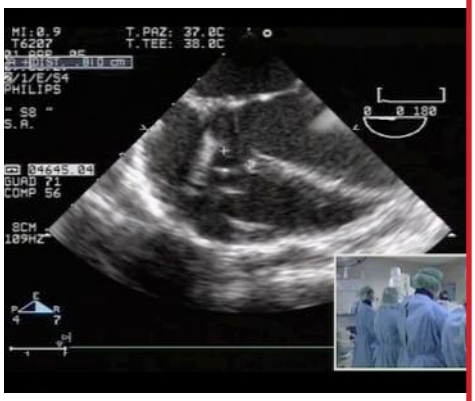
Free Subscription to Qualified Professionals: Send your name, title(s), hospital or practice name, work address and url, phone, fax and email to: sub@cct.bz.

Watch Over 300 Live Case Videos, Presentations and Workshops Online from Leading Congenital and Structural Medical Meetings from Around the World

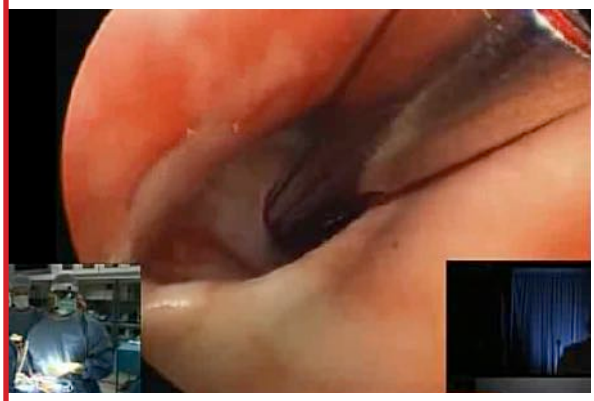
www.CHDLiveCases.com

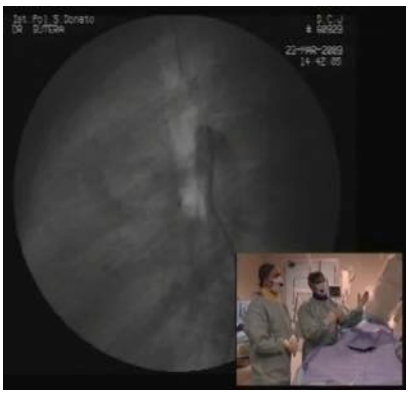


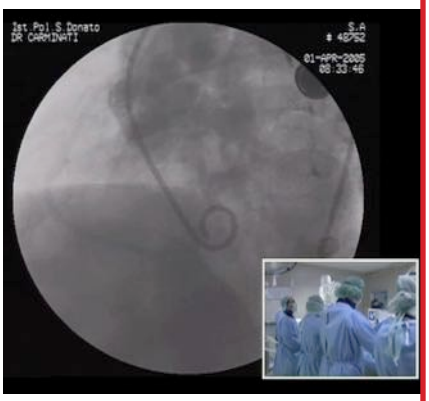




- Transseptal Access Workshop from Cook Medical
- Workshop: Past Present and Future of Pediatric Interventions Cardiology - St. Jude & AGA Medical
- Symposium on Prevention of Stroke Clinical Trials at the Heart of the Matter - WL Gore Medical
- Imaging in Congenital & Structural Cardiovascular Interventional Therapies
- Morphology of The Atrial Septum
- Morphology of The Ventricular Septum
- 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
- **and many more....**







Presented by CONGENITAL CARDIOLOGY TODAY

ONE PARTNER, MANY SOLUTIONS

Only Medtronic offers a broad portfolio of solutions to manage your patients with Congenital Heart Disease throughout their lifetime.

Transcatheter and Surgical Heart Valves | RVOT Conduits | Ablation Technologies | ICDs | Oxygenators and Filters | Cannulae | Pacemakers | Pulse Oximetry Monitoring for CCHD Screening | Cerebral/Somatic Monitoring

Medtronic
Further, Together