CONGENITAL CARDIOLOGY TODAY
Timely News & Information for Congenital/Structural Cardiologists & Cardiothoracic Surgeons Worldwide

International Edition
Vol. 18 - Issue 1

JANUARY 2020

PERCUTANEOUS AMPLATZER SEPTAL OCCLUDER DEVICE OCCLUSION OF A LARGE PORTAL VEIN TO PULMONARY VENOUS ATRIUM COLLATERAL RESULTING IN SEVERE SYSTEMIC HYPOXEMIA POST FONTAN

WRITTEN BY
Howard S. Weber, MD, FSCAI
Farrah Munir, MD
Bryan P. Stefek, MD
Frank C. Lynch, MD

INTRODUCTION

The development of decompressing veno-venous collaterals is common following the bidirectional Glenn shunt and Fontan operations which can result in varying degrees of systemic desaturation. This phenomenon is thought to be a biologic reaction to the increased systemic venous pressure changes associated with this type of circulation. Often, these vessels are of supradiaphragmatic origin such as the brachiocephalic veins which communicate with the inferior cardinal vein, pulmonary venous atrium or coronary sinus. Other potential locations include the azygous and hemiazygous venous systems. The portal venous system is rarely discussed as a potential for veno-venous collateralization, however, it is certainly a vascular territory at risk for dysfunction in the presence of Fontan physiology. We describe an unusual cause of significant systemic hypoxemia many years post extracardiac non-fenestrated Fontan completion secondary to the development of a large portal veno-venous communication with the pulmonary venous atrium that was successfully embolized utilizing an Amplatzer Septal Occluder device.

CASE PRESENTATION

The patient is a 17-year-old male with complex single ventricule physiology (dextrocardia with situs ambiguous, asplenia, complete atrioventricular canal, pulmonary valve atresia, right-sided IVC and left-sided SVC with infradiaphragmatic total anomalous pulmonary venous return). He underwent a modified left BT shunt and TAPVR repair soon after birth, followed by a bidirectional Glenn shunt at six months of age and extracardiac non-fenestrated lateral tunnel Fontan completion at four years of age. At five years of age, he underwent a routine post Fontan cardiac catheterization utilizing intravenous conscious sedation which demonstrated acceptable hemodynamics with a mean central venous pressure of 13mmHg and transpulmonary gradient of 4mmHg.

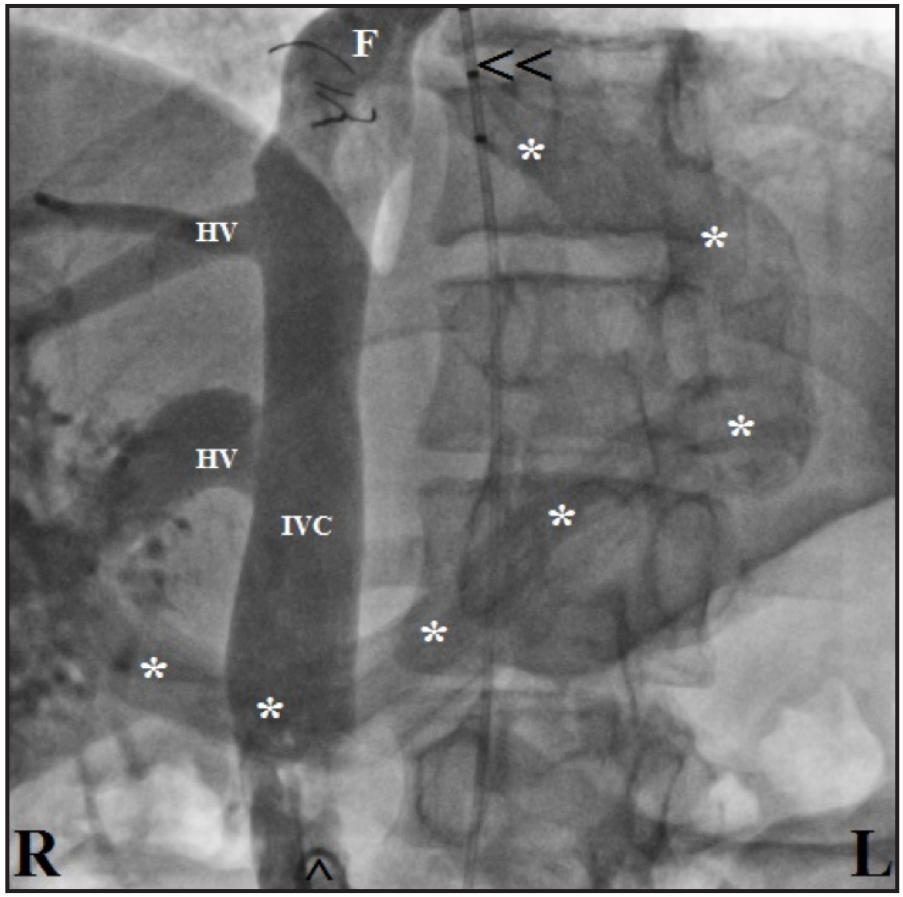


FIGURE 1 *Anterior-Posterior (AP) View IVC injection with opacification of hepatic veins (HV) and Fontan pathway (F). A hepatic vein-to-portal vein shunt (asterisks) is identified on late filling which communicates with a venous structure that ascends above the diaphragm and into the pulmonary venous atrium. (R: patient right; L: patient left; double carrot: sizing catheter in descending aorta)*

JANUARY 2020

International Edition Vol. 18 - Issue 1

INSIDE THE ISSUE

1 **PERCUTANEOUS AMPLATZER SEPTAL OCCLUDER DEVICE OCCLUSION OF A LARGE PORTAL VEIN TO PULMONARY VENOUS ATRIUM COLLATERAL RESULTING IN SEVERE SYSTEMIC HYPOXEMIA POST FONTAN**

WRITTEN BY Howard S. Weber, MD, FSCAI; Farrah Munir, MD; Bryan P. Stefek, MD & Frank C. Lynch, MD

8 **ANOMALOUS CONNECTION OF INFERIOR VENA CAVA TO LEFT ATRIUM**

WRITTEN BY Salwa M. Gendi, MD & Patricia O'Dierno, MD

14 **MEDICAL NEWS**

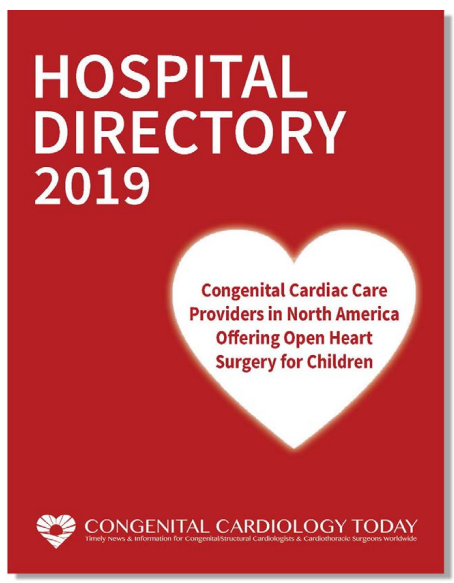
COMPILED BY Kate Baldwin & Tony Carlson

@CCardiology

HOSPITAL DIRECTORY 2019

IS NOW AVAILABLE ON CCT'S WEBSITE

Check It Out Today!



www.CongenitalCardiologyToday.com/2019/09/03/hospital-directory-2019

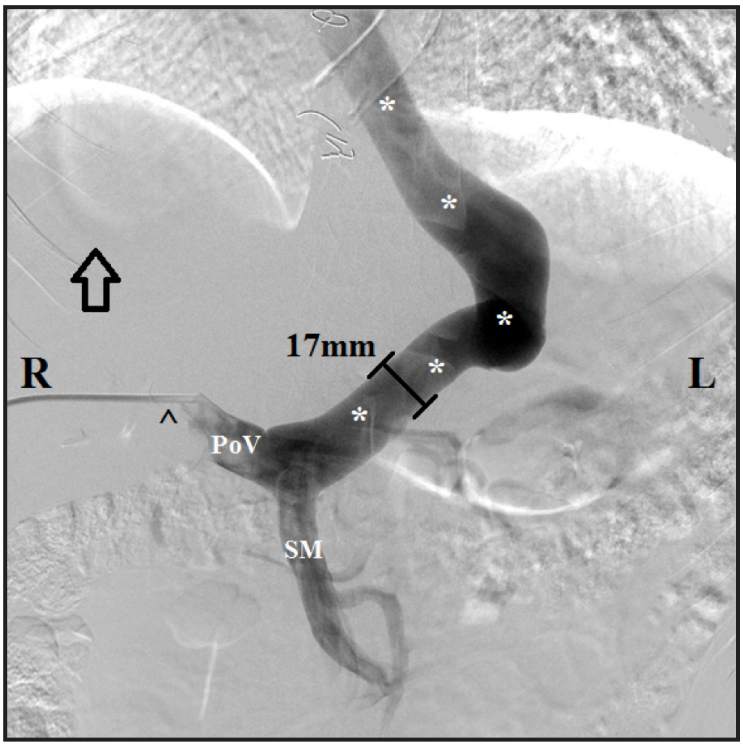


FIGURE 2 *AP Digital Subtraction Angiography* *Access to the portal venous system from a mid- axillary intercostal approach. Mid-portal vein (PoV) contrast injection demonstrates connection to a large venous collateral (Asterisks: 17mm diameter) continuing above diaphragm with opacification of the pulmonary venous atrium. Dextrocardia demonstrated with right-ward cardiac apex (arrow). (R: patient right; L.: patient left; SM: superior mesenteric vein; carrot: injection catheter)*

His systemic saturation was mildly reduced at 89% with a normal hemoglobin of 14.8 gm/dl. Angiography in the inferior vena cava demonstrated reversal of flow from the hepatic veins into the portal vein which then communicated with the pulmonary venous atrium via a large ascending venous structure. No intervention was performed at that time. He continued outpatient followup with a local pediatric cardiologist who recently documented significant systemic desaturation at rest (81%) and the development of a secondary polycythemia (hemoglobin: 18.9 gm/dl and hematocrit: 55%) with normal RBC indices. He was subjectively asymptomatic from a cardiovascular standpoint without hepatomegaly. Transthoracic echocardiography demonstrated subjectively normal biventricular systolic function with trivial insufficiency of the common atrioventricular valve and the caval connections appeared unobstructed. The patient did not report any clinical evidence of protein-losing enteropathy. He underwent repeat cardiac catheterization under general anesthesia in April 2019 which demonstrated excellent post-Fontan hemodynamics with a mean central venous pressure of 11mmHg and normal transpulmonary gradient of 5mmHg. The extracardiac Fontan conduit, caval connections and pulmonary arteries were widely patent. There was no echocardiographic evidence of micro pulmonary arterio-venous fistulas in either lung field via agitated saline injection. Angiography demonstrated flow reversal within the hepatic veins into the portal vein with significant opacification of the pulmonary venous atrium via a large ascending venous structure. This communication was clearly larger than first noted many years ago and accounted for the severe systemic desaturation (Figure 1). The patient was referred to interventional radiology for embolization of the portal veno-veno communication utilizing percutaneous techniques.

From a low intercostal approach in the mid axillary line, a 22-gauge Chiba needle was advanced through the liver obliquely and during slow withdrawal while contrast was being injected, opacification of a peripheral right portal vein was achieved. Eventually a #5 French sheath was placed into the portal vein through which digital subtraction angiography was performed. A large vein, measuring approximately 17 mm, was seen arising from the portal vein-splenic vein junction and drained above the diaphragm to the left-sided atrium (Figures 2,3). However, due to the large diameter of this venous structure, an Amplatzer vascular plug was not an option. Therefore, a 12mm Amplatzer Septal Occluder device (26mm left atrial disc and 24mm right atrial disc) was chosen. Following device implantation there was an immediate improvement in the patient's systemic saturation to 95% in room air (Figure 4). Additional Gianturco coils were then placed proximal to the septal occluder to prevent theoretical recanalization. Following embolization there was preserved hepato-pedal flow in the superior mesenteric vein and

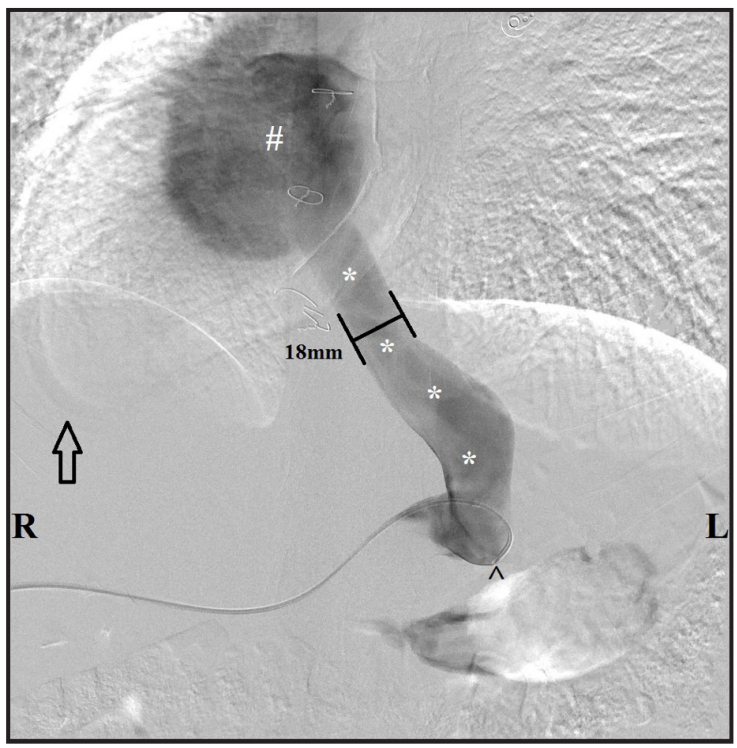


FIGURE 3 *AP Digital Subtraction Angiography (DSA)* *Contrast injection into the large venous collateral (Asterisks: 18mm diameter) continuing above diaphragm to pulmonary venous atrium (hashtag). (R: patient right; L.: patient left; carrot: injection catheter; arrow: right-ward cardiac apex)*

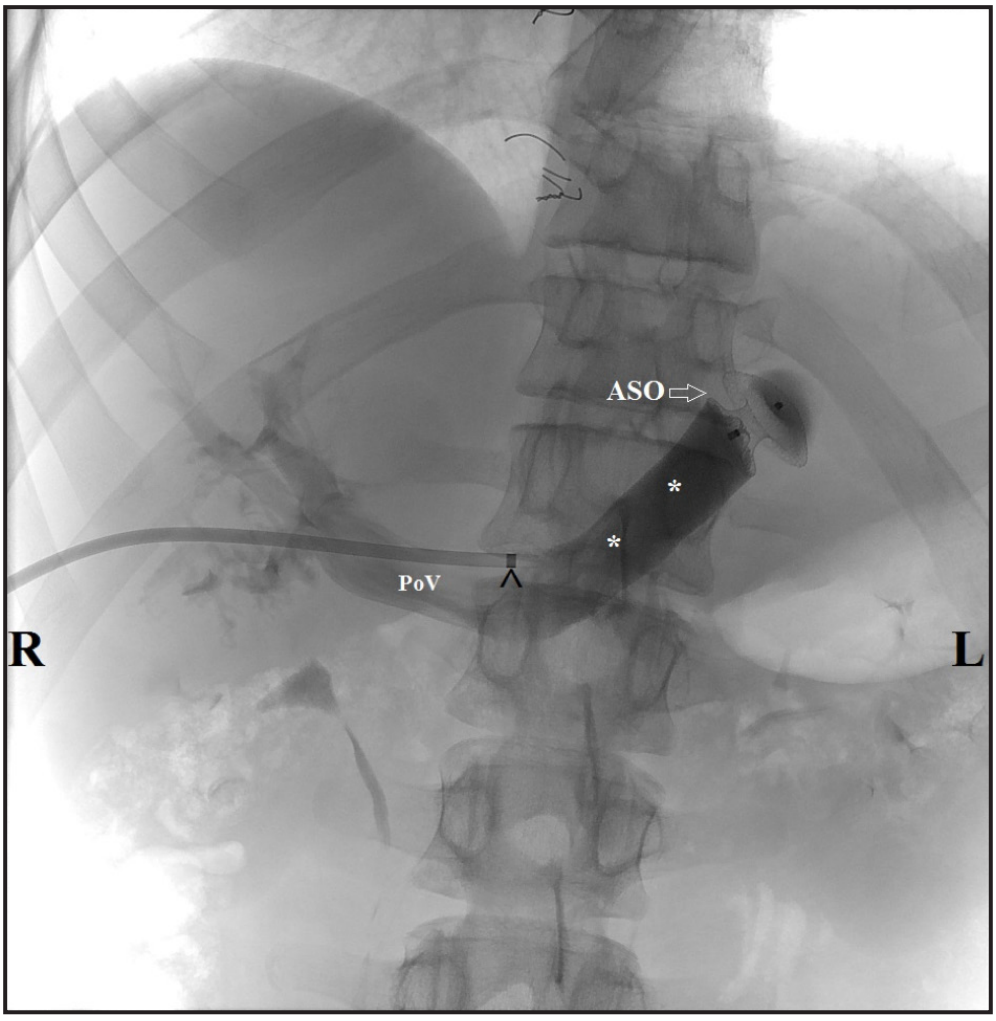


FIGURE 4 *AP Contrast Angiography within the porto-venous connection proximal to the 12mm Amplatzer Septal Occluder device (ASO) which has become elongated as expected. Contrast now injected in proximal collateral vein (asterisks) stops at lateral spine border with no continuation above diaphragm.*

within the main and intrahepatic portal veins. The direct portal vein pressure prior to embolization was a mean of 11 mmHg and, as expected, increased to 15 mmHg post embolization. He was discharged home the following morning with a normal systemic saturation of 95% in room and no cardiovascular or abdominal symptoms.

DISCUSSION

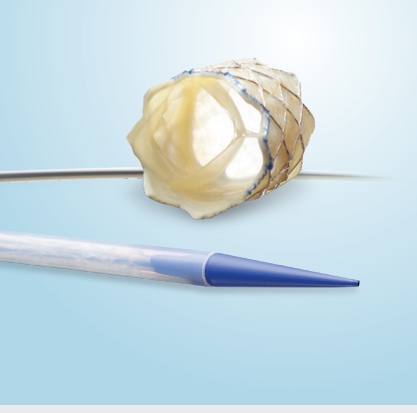
The development of veno-venous collaterals resulting in varying degrees of systemic desaturation is common in patients with Fontan physiology. Previous published reports have estimated that approximately 1/3 of patients will develop connections between the systemic and pulmonary venous systems following either a bidirectional Glenn shunt or Fontan completion[1,2]. This phenomenon is thought to be a biologic reaction to the increased systemic venous pressure changes associated with Fontan physiology. Some authors describe these vessels as 're-opened' embryologic pathways resulting in a pop-off for the abnormally elevated systemic venous pressures. Others have proposed that these vessels may result from primary angiogenesis[1]. Often, these vessels are of supradiaphragmatic origin such as the brachiocephalic veins draining either into the inferior cardinal vein, pulmonary venous atrium or coronary sinus[2]. Other potential locations include the azygous and hemiazygous venous systems. The portal venous system is rarely discussed as a location for veno-venous collateralization; however it is certainly a vascular territory at risk for dysfunction in the Fontan population[3,4]. Hsia et al described the potential for congestion in the hepatic-portal venous system, which may contribute to liver and gastro-intestinal dysfunction[5]. We speculate that similar physiology may have contributed to our patient's unusual hepatic vein-portal-vein shunt development.

A variety of transcatheter devices have been employed to occlude these veno-veno connections when hemodynamically or clinically significant[6]. However, in this case, the portal vein had to be directly accessed percutaneously in order to implant an occluder device. Although this is a rarely utilized technique by interventional pediatric cardiologists, it's not an uncommon approach for interventional radiologists. In patients with portal hypertension secondary to primary liver disease, the development of porto-pulmonary venous anastomosis leading to systemic embolization and stroke has been documented with resolution following successful embolization techniques[7,8]. To our knowledge, this is the first use of an Amplatzer septal occluder device for embolization of a large portal vein to pulmonary venous atrium collateral resulting in significant systemic desaturation in a postoperative Fontan patient. This entity should be considered in future patients with significant systemic desaturation when other etiologies have been excluded.

REFERENCES

1. Sugiyama H, Yoo SJ, Williams W, Benson LN. Characterization and treatment of systemic venous to pulmonary venous collaterals seen after the Fontan operation. Cardiol Young. 2003; 13(5):424-30.
2. Weber HS. Incidence and predictors for the development of significant supradiaphragmatic decompressing venous collateral channels following creation of Fontan physiology. Cardiol Young. 2001; 11(3):289-94.

RIGHT CHOICE.



Melody™

Transcatheter Pulmonary Valve (TPV) System

Not intended to constitute medical advice or in any way replace the independent medical judgment of a trained and licensed physician with respect to any patient needs or circumstances. Melody TPV is not suitable for all patients and ease of use, outcomes, and performance may vary. See the Instructions for Use for indications, contraindications, precautions, warnings, and adverse events.

Restoring lives for

12

years and counting.

The only transcatheter pulmonary valve specifically designed for RVOT conduits and bioprosthetic valves. The longest studied, with the largest body of clinical evidence at over 8 years post-implant.* Over 12 years of implants, more than 14,000 patients' lives have been changed.

Melody TPV — The Right Choice for Your Patients

*Melody Transcatheter Pulmonary Valve Study: Post Approval Study of the Original IDE Cohort.

©2019 Medtronic. All rights reserved.
UC201809495a EN 07/2019

Medtronic
Further, Together

Important Labeling Information for the United States

Indications: The Melody TPV is indicated for use in the management of pediatric and adult patients who have a clinical indication for intervention on a dysfunctional right ventricular outflow tract (RVOT) conduit or surgical bioprosthetic pulmonary valve that has ≥ moderate regurgitation, and/or a mean RVOT gradient ≥ 35 mm Hg.

Contraindications: None known.

Warnings/Precautions/Side Effects

- **DO NOT implant in the aortic or mitral position. Pre-clinical bench testing of the Melody valve suggests that valve function and durability will be extremely limited when used in these locations.**
- DO NOT use if patient's anatomy precludes introduction of the valve, if the venous anatomy cannot accommodate a 22 Fr size introducer, or if there is significant obstruction of the central veins.
- DO NOT use if there are clinical or biological signs of infection including active endocarditis. Standard medical and surgical care should be strongly considered in these circumstances.
- Assessment of the coronary artery anatomy for the risk of coronary artery compression should be performed in all patients prior to deployment of the TPV.
- To minimize the risk of conduit rupture, do not use a balloon with a diameter greater than 110% of the nominal diameter (original implant size) of the conduit for pre-dilation of the intended site of deployment, or for deployment of the TPV.
- The potential for stent fracture should be considered in all patients who undergo TPV placement. Radiographic assessment of the stent with chest radiography or fluoroscopy should be included in the routine postoperative evaluation of patients who receive a TPV.
- If a stent fracture is detected, continued monitoring of the stent should be performed in conjunction with clinically appropriate hemodynamic assessment. In patients with stent fracture and significant associated RVOT obstruction or regurgitation, reintervention should be considered in accordance with usual clinical practice.

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, blistering, or peeling of skin, pain, swelling, or bruising at the catheterization site. Potential device-related adverse events that may occur following device implantation include the following: stent fracture,* 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.

*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

For additional information, please refer to the Instructions for Use provided with the product or available on http://manuals.medtronic.com.

CAUTION: Federal law (USA) restricts this device to sale by or on the order of a physician.

Important Labeling Information for Geographies Outside of the United States

Indications: The Melody™ TPV is indicated for use in patients with the following clinical conditions:

- Patients with regurgitant prosthetic right ventricular outflow tract (RVOT) conduits or bioprostheses with a clinical indication for invasive or surgical intervention, OR
- Patients with stenotic prosthetic RVOT conduits or bioprostheses where the risk of worsening regurgitation is a relative contraindication to balloon dilatation or stenting

Contraindications

- Venous anatomy unable to accommodate a 22 Fr size introducer sheath
- Implantation of the TPV in the left heart
- RVOT unfavorable for good stent anchorage
- Severe RVOT obstruction, which cannot be dilated by balloon
- 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, swelling or bruising at the catheterization site. Potential device-related adverse events that may occur following device implantation include the following: stent fracture.* 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.

*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

For additional information, please refer to the Instructions for Use provided with the product or available on http://manuals.medtronic.com.

The Melody Transcatheter Pulmonary Valve and Ensemble II Transcatheter Delivery System has received CE Mark approval and is available for distribution in Europe.

medtronic.com

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

©2019 Medtronic. All rights reserved. Medtronic, Medtronic logo, and Further, Together are trademarks of Medtronic. All other brands are trademarks of a Medtronic company.

UC201809495a EN 07/2019

Medtronic

3. Layangool T, Kojaranjit V, Promphan W, Kirawittaya T, Sangtawesin C, Prachasilchai P. An unusual cause of progressive cyanosis post Fontan operation: congenital extra-hepatic porto-systemic shunt. J Med Assoc Thai. 2014; 97 Suppl 6:S176-81.
4. Lluri G, Levi DS, Aboulhosn J. Systemic to pulmonary venous collaterals in adults with single ventricle physiology after cavopulmonary palliation. Int J Cardiol. 2015 15; 189:159-63.
5. Hsia TY, Khambadkone S, Deanfield JE, Taylor JF, Migliavacca F, De Leval MR. Subdiaphragmatic venous hemodynamics in the Fontan circulation. J Thorac Cardiovasc Surg. 2001; 121(3):436-47.
6. Masura J, Bordacova L, Tittel P, Berden P, Podnar T. Percutaneous management of cyanosis in Fontan patients using Amplatzer occluders. Catheter Cardiovasc Interv. 2008 1; 71(6): 843-9.
7. Miura H, Yamagami T, Tanaka O and Yoshimatsu. Portopulmonary venous anastomosis detected at balloon occluded retrograde transvenous obliteration for gastric veins. J. Vasc Interv Radiol 2013; 24: 131-134.
8. Rossi UG, Gandini G and Cariati M. Portal-left atrium shunt leading to stroke. J. Vasc Interv Radiol 2014; 25: 363.

Howard S. Weber, MD, FSCAI
Principle Author

Director of Interventional Pediatric Cardiology
Penn State Health Children's Hospital
500 University Drive
Hershey, PA, USA
hweber@pennstatehealth.psu.edu

Bryan P. Stefek, MD

Division of Pediatric Cardiology
Penn State Health Children's Hospital
Hershey, PA, USA

Farrah Munir, MD

Division of Pediatric Cardiology
Penn State Health Children's Hospital
Hershey, PA, USA

Frank C. Lynch, MD

Department of Radiology
Penn State Health
Milton S Hershey Medical Center
Hershey, PA, USA

ANOMALOUS CONNECTION OF THE INFERIOR VENA CAVA TO THE LEFT ATRIUM

WRITTEN BY
Salwa M. Gendi, MD
Patricia O'Dierno, MD

ABSTRACT

Anomalous drainage of the IVC into the left atrium is a very rare congenital anomaly of the heart. In our case report, we have a newborn female who had continued hypoxia after birth despite multiple interventions. An echocardiogram showed a bilateral SVC, with the right SVC and IVC draining into the right atrium and the left SVC draining directly into the left atrium, an absent innominate vein between the two SVCs, a small PFO, and an unroofed coronary sinus. The patient was discharged home with oxygen saturations ranging from 75-85%. During surgical correction at four months of age, it was found that her IVC entered the diaphragm at a normal position, but drained into the left atrium due to the malformation of the atrial septum. The atrial septum was reconstructed to redirect the IVC to drain into the right atrium. After surgical correction, she had normal oxygen saturations and was discharged home.

INTRODUCTION

The IVC is normally formed by the contribution of five venous systems and carries blood into the right atrium. In our case, the malposition of the septum primum caused the IVC to drain into the left atrium. When the septum secundum is absent, it can cause the septum primum to be displaced in either direction. This can cause the pulmonary veins to drain into the opposite atria, and can cause the IVC to drain in the opposite atrium as well. Many times, surgical treatment is warranted. There are three types of repairs. If there is AV concordance, a new septum can be constructed so that the systemic and pulmonary veins drain into the corresponding atria. If there is AV discordance with well-developed ventricles, the pulmonary and systemic veins can be rerouted to drain

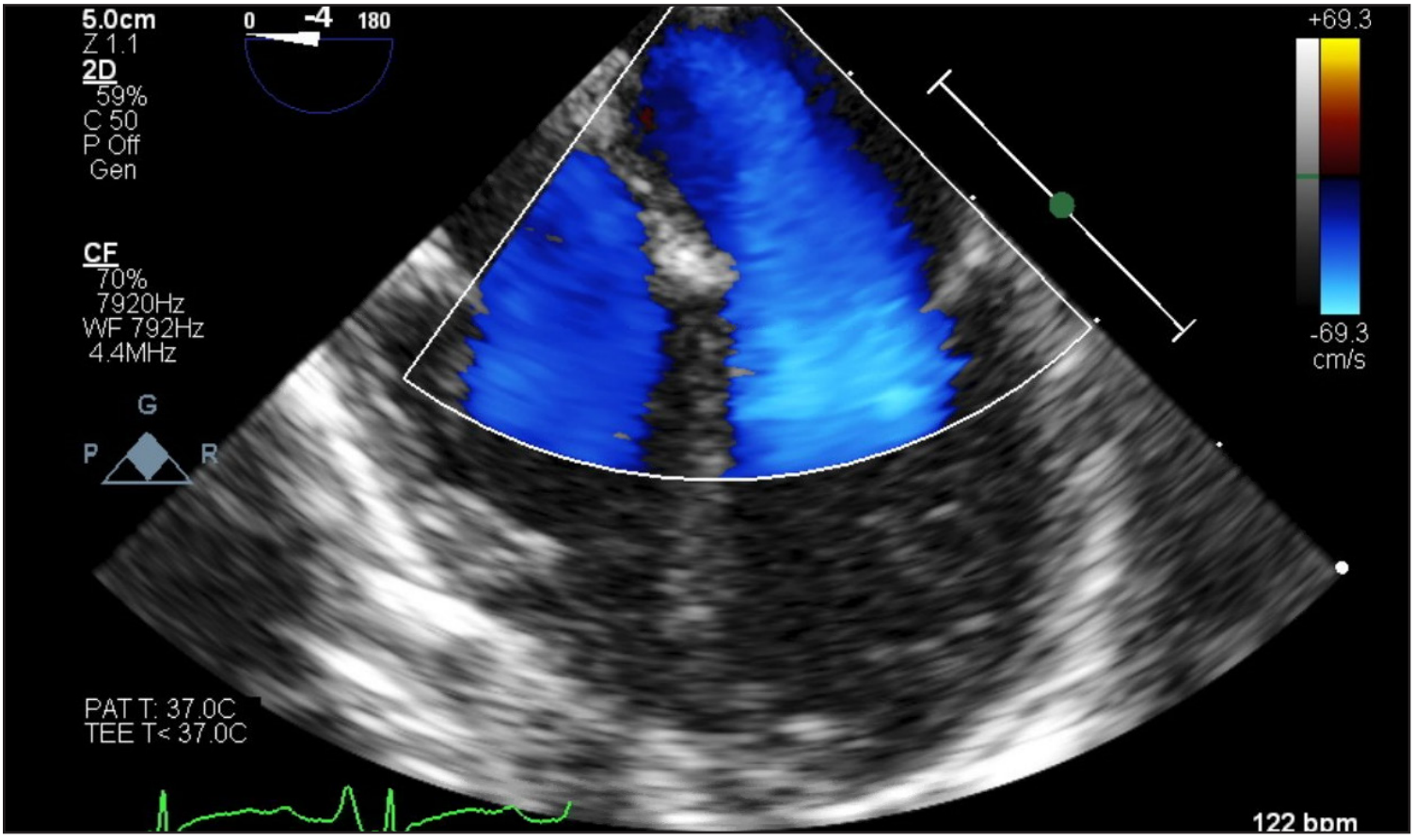


FIGURE 1 *Apical 4 chamber view.* *No abnormality detected.*

CONGENITAL CARDIOLOGY TODAY

- Written by doctors and their team
- Case studies, articles, research findings
- Submit on your schedule
- Print and electronic
- Published within 3 months of submission
- No fees

- In print and electronic monthly issue
- On our website
- In our monthly email blast
- No cost for CCT to create the ad
- Multiple sizes available

Subscribe Electronically
Free on Home Page

www.CongenitalCardiologyToday.com

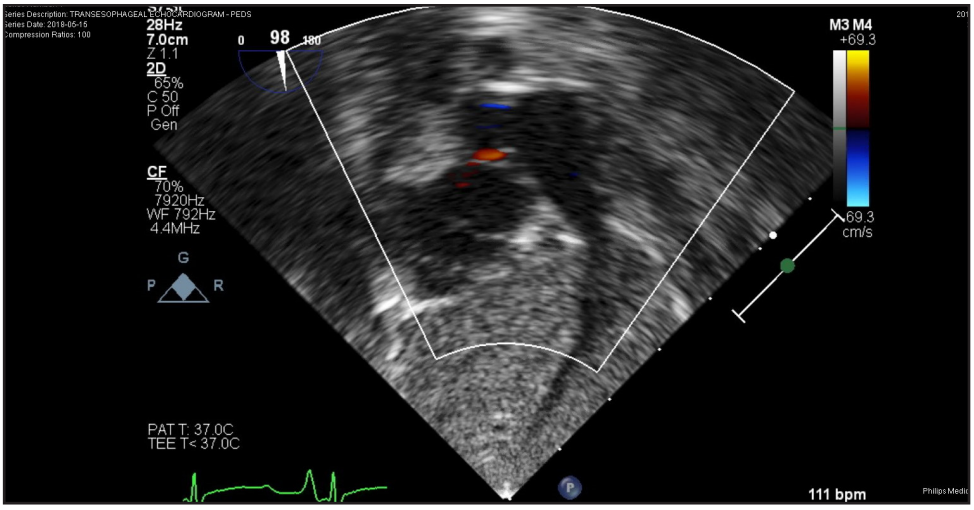


FIGURE 2a *Subcostal view showing IVC entering into LA to the left of the atrial septum. Small PFO noted.*

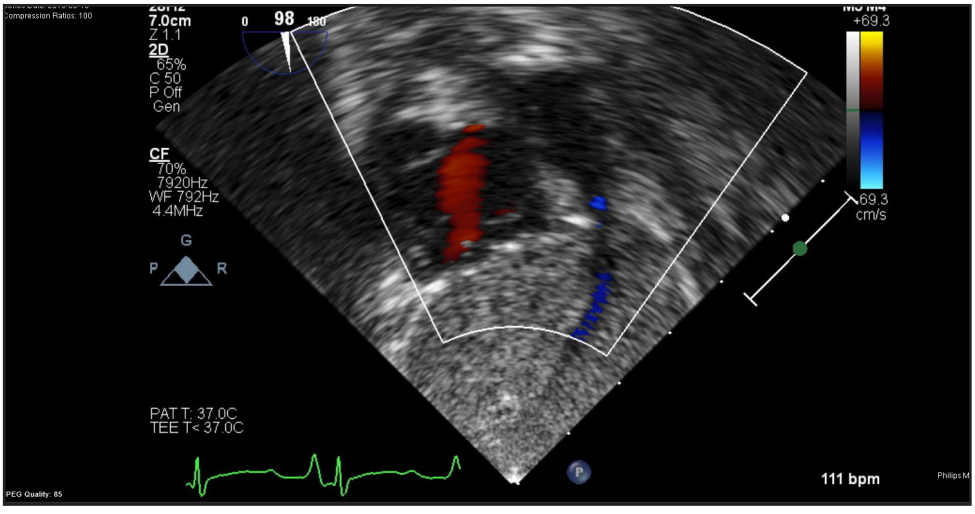


FIGURE 2b *Subcostal view showing flow into IVC going into LA.*

into the corresponding ventricles. If a biventricular repair cannot be achieved, it is generally due to a more serious congenital malformation. In the case of our patient, her septum was able to be reconstructed to cause her IVC to drain into her right atrium. Partial or complete drainage of the IVC into the LA can cause cyanosis, polycythemia, brain abscess and paradoxical emboli due to the right-to-left shunting[1].

CASE REPORT

Our patient is a newborn female, born at 35 weeks four days via Cesarean section secondary to preeclampsia, who initially presented with hypoxia. A chest X-ray was normal. Initial transthoracic echocardiogram on Day 2 of Life showed normal anatomy and function. Hypoxia continued to persist on nasal CPAP despite treatment with Surfactant, Nitric oxide and Sildenafil. Due to continued desaturations with no known cause, an echocardiogram was repeated on Day 9 of Life. This repeat echocardiogram showed a bilateral SVC, with the right SVC and IVC draining into the right atrium and the left SVC draining directly into the left atrium, an absent innominate vein between the two SVCs, a small PFO, and an unroofed coronary sinus. Patient was discharged home on Day 15 of Life in stable condition, with oxygen saturations ranging from 75%-85%.

At her follow-up visit six weeks later, she was stable, with mild perioral cyanosis when agitated. The family elected to have her undergo surgical repair of the heart defect as soon as it was safe.

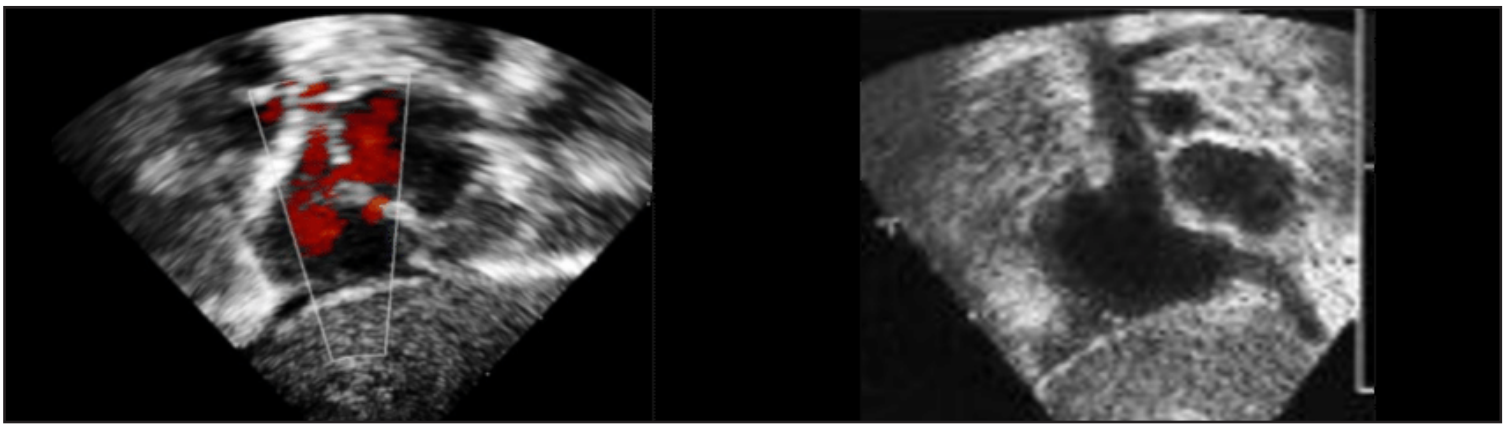


FIGURE 3 *Example of IVC draining normally into the RA to the right of interatrial septum.*

At four months of age, the patient underwent surgical correction. During the procedure, the IVC was noted to enter the diaphragm at the normal position, but was beneath the plane of the septum and draining into the left atrium. A bovine pericardial patch was used to reconstruct the atrial septum to redirect the IVC to drain superiorly into the right atrium. There was also repair of the anomalous systemic venous connections by connecting the left SVC to the right atrial appendage.

The patient had a complicated post-operative course, with a bilateral chylothoraces requiring chest tubes to be placed. She also had a left-sided internal jugular vein clot and left hemidiaphragm paralysis.

Patient's multiple post-operative echocardiograms showed normal structure and function. Patient was discharged home on Lasix and Diuril with normal oxygen saturations.

DISCUSSION

An abnormal IVC draining into the left atrium is a very rare defect. In fact, during our literature review, we could only find a few cases that were similar to ours. In a previous case report published, a 23-year-old male who presented with cyanosis was found to have an anomalous IVC connection to the left atrium with a secundum ASD. In that case, the septum primum was unroofed and closed so the IVC would flow anteriorly into the right atrium[2].

We could not find any other cases that were similar to ours. We did find a case of a 2-year-old girl with cyanosis that was found to have bilateral SVC with the left SVC draining into the right atrium and the right SVC draining into the left atrium. The SVCs were connected by an innominate vein[3].

Overall, our patient had a very rare abnormality of the atrial septum that caused the IVC to drain into the left atrium. After surgical correction, she was discharged home with normal saturations.

REFERENCES

1. Allen H.D., Shaddy R.E., Penny D.J., Feltes T.F., Cetta F. Moss and Adams' heart disease in infants, children and adolescents: including the fetus and young adults. Ninth edition 2013; 888-928.
2. Steele J, Erenberg F, Majdalany D, Prieto L, Yaman M.E. Anomalous Inferior Vena Cava To The Left Atrium. American College of Cardiology 2018. 71 (11) (Pubmed).
3. Alghamdi M.H, Elfaki, W., Al-Habshan F., Aljarallah A. Bilateral superior vena cava with right superior vena cava draining into left atrium. J Saudi Heart Assoc 2015. (2):123-126. (Pubmed).

Salwa M. Gendi, MD
Corresponding Author

Assistant Professor
Pediatric Cardiology
West Virginia University
Morgantown, WV, USA
salwa.gendi@hsc.wvu.edu

Patricia O'Dierno, MD

PGY3 Pediatric Resident
Pediatric Cardiology
West Virginia University
Morgantown, WV, USA

Interventional Systems
B|BRAUN
SHARING EXPERTISE

NuMED

Nano4Imaging
YOUR HEALTH - OUR VISION

pfmmedical

MAKING A DIFFERENCE

NuDEL®
CP Stent® Delivery System

EmeryGlide™
MR Conditional Guidewire

Nit-Occlud® PDA
Coil System For PDA Closure

NuDEL™ Indications for Use:
The NuDEL 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.

The NuDEL is indicated for use in the treatment of right ventricle to pulmonary artery (right ventricular outflow tract) conduit disruptions that are identified during conduit pre-dilatation procedures performed in preparation for transcatheter pulmonary valve replacement.

Caution: Federal (USA) Law restricts this device to sale by or on the order of a physician. **Contraindications:** Clinical or biological signs of infection. Active endocarditis. Pregnancy. **Contraindications (CoA only):** 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 or the stent. Known allergy to aspirin, other antiplatelet agents, or heparin. **Contraindications (RVOT only):** Patients too small to allow safe delivery of the stent without injury to a systemic vein or to the right side of the heart. **Warnings / Precautions:** Administer appropriate anticoagulation therapy to reduce potential thrombosis. If the patient is not appropriately anticoagulated, thrombus formation may occur. The sheath must be flushed with heparinized saline via the proximal side port prior to introducing the delivery system into the body. The inflated diameter of the stent should at least equal the diameter of the intended implant site. Excessive handling and manipulation of the covering while crimping the stent may cause the covering to tear off of the stent. Retracting the covered stent back into the sheath may cause the covering to catch and tear off of the stent. Do not exceed the RBP. An inflation device with pressure gauge is recommended to monitor pressure. Pressure in excess of the RBP can cause balloon rupture and potential inability to withdraw the catheter into the sheath. Confirm that the distal end of the introducer sheath is at least 2.5cm back from the most proximal image band before inflating the outer balloon. Failure to do so may stretch the outer tubing and severely hinder balloon deflation. Exercise caution when handling the stent to prevent breakage. The NuDEL system, especially at the stent, is rigid and may make negotiation through vessels difficult. The inflation diameter of the balloon used during stent delivery should approximate the diameter of the obstructive vessel and the intended implant site. If resistance is encountered upon removal, the whole system (balloon, guidewire and sheath) should be removed as a single unit, particularly if balloon rupture or leakage is known or suspected. **Warnings / Precautions (CoA only):** Coarctation of the aorta involving the aortic isthmus or first segment of the descending aorta should be confirmed by diagnostic imaging. The NuMED CP Stent has not been evaluated in patients weighing less than 20kg. The platinum/iridium stent may migrate from the site of the implant. 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. **Warnings / Precautions (RVOT only):** During the Premarket Approval study the Medtronic Melody valve was used for valve restoration. The safety and effectiveness of the Covered CP Stent for pre-stenting of the right ventricular outflow tract (RVOT) landing zone (i.e. prophylaxis or prevention of either RVOT conduit rupture or TPVR fracture; use as a primary RVOT conduit) in preparation of a transcatheter pulmonary valve replacement (TPVR) has not been evaluated. As with any type of implant, infection secondary to contamination of the stent might lead to endocarditis, or abscess formation. The Covered Stent can migrate from the site of implant potentially causing obstruction to pulmonary artery flow. Over-stretching of the RVOT may result in rupture or aneurysm of the RV-PA conduit or the native pulmonary artery.

Nit-Occlud® 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. (5kg). 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 bodyweight should be injected after femoral sheaths are placed. Antibiotics should be given before (1 dose) and after implantation (2 doses) 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 IFUs for a complete listing of indications, contraindications, warnings and precautions. www.bisusa.org
NuDEL is a trademark of NuMED, Inc. Nit-Occlud is a registered trademark of pfm medical, inc.
Rx only CV-9112 Rev. 8/19 ©2019 B. Braun Interventional Systems Inc.

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

MEDICAL NEWS

COMPILED BY
Kate Baldwin
Tony Carlson

LURIE CHILDREN'S OFFERS KIDS VIRTUAL ESCAPE FROM INTENSIVE CARE UNIT

From Scuba Diving to Snowboarding, Patients in the Pediatric Intensive Care Unit Leave the Hospital Behind with Virtual Reality

For the first time in a Pediatric Intensive Care Unit (PICU), patients get a chance to scuba dive, snowboard, and go on a safari or other adventures, all from their hospital bed. The 360 degree immersions into virtual environments were extremely well received by PICU patients and their parents, according to results from a pilot study from Ann & Robert H. Lurie Children's Hospital of Chicago that were published in *Pediatric Critical Care Medicine*. All 32 study participants, ages 3-17 years, reported that they enjoyed using virtual reality. All of their parents agreed, with over 80% reporting that virtual reality experience calmed their child.

"We conducted this study to make sure that it is feasible to introduce virtual reality into a pediatric intensive care setting and that kids respond well to it," says senior author Marcelo Malakooti, MD, from Lurie Children's who also is Assistant Professor of Pediatrics-Critical Care at Northwestern University Feinberg School of Medicine. "We are now introducing virtual reality more broadly to critically ill children on the unit who are often alert, but stuck in bed just passively watching TV. Such minimal engagement with their environment over prolonged hospitalization can lead to delirium or other cognitive and emotional impairments. We hope that the stimulation and interaction that virtual reality offers will mitigate that risk and improve outcomes for these children."

Based on the positive results of the pilot study, Dr. Malakooti, lead author Colleen Badke, MD, and colleagues at Lurie Children's are now conducting a larger study to examine how virtual reality use in the PICU impacts pain, anxiety and physical factors like blood pressure and heart rate variation, among others.

INFOBIONIC'S REVOLUTIONARY MOME KARDIA EARNS TOP TECHNOLOGY LEADERSHIP AWARD FROM FROST & SULLIVAN

Mome® Kardia's Innovative and Disruptive Cardiac Monitoring Technology Means Better Outcomes for Patients and Doctors

Boston-based InfoBionic, developer of the MoMe® Kardia remote cardiac monitor and software, won this year's Technology Leadership Award from Frost & Sullivan. The award is presented to the technology firm that best embodies commitment to innovation and creativity, serves as an incubator and developer of new technology, and demonstrates a track record of commercial success[1].

InfoBionic's MoMe® Kardia, is the only full-disclosure remote monitor currently on the market that allows doctors immediate and 24/7 access to cardiac data on demand. It captures a clinical-grade electrocardiogram (ECG) and provides true continuous cardiac monitoring in near real time, which the doctor can access on their tablet, smartphone, or on any Internet-connected computer in the world.

"We're grateful and honored to have received this year's Technology Leadership Award," said Stuart Long, CEO of InfoBionic. "We're very excited to be helping transform the cardiac monitoring industry, and look forward to helping providers implement this potentially life-saving technology into their own work flows."

Each year, Frost & Sullivan presents Best Practices Awards to companies that are expected to encourage significant growth in their industries, have identified emerging trends before they became the marketplace standard, and have created advanced technologies that will catalyze and transform industries in the near future. This year's awards were presented at the Hyatt Regency in La Jolla, California.

ARRHYTHMIA

MoMe® Kardia is designed to help doctors detect and diagnose cardiac arrhythmia, which is characterized by irregular electrical activity in the heart that causes it to beat too rapidly, too slowly, or erratically.

BENEFITS OF THE MOME® KARDIA

InfoBionic's innovations in data analytics, in artificial intelligence, and in secure, HIPAA-compliant telecommunications enabled it to bring a device to market that is poised to transform the entire cardiac monitoring industry. MoMe® Kardia's documented benefits include:

- The physician has access to all data at any given point of time. Full data disclosure sets MoMe® Kardia apart from market competition because all other devices only share event triggers with the patient. Full data has to be requested, and even then it may take time to receive. By giving physicians complete access to patients' health data, InfoBionic empowers them with true 24/7 monitoring and faster intervention.
- It has vast improvements in data retention. In the past, 95% to 99% of cardiac data is lost in transmission. Independent diagnostic and testing facilities servicing monitors may send a 20 or 30 second tape of a detected arrhythmia. However, doctors too

often missed critical onset and offset data in the minutes prior to the cardiac event—this data is vital to diagnosis.
- The time-to-diagnosis cycle is improved, and in some cases by days. Doctors do not have to get bogged down in faxes, emails, and phone calls from third-party monitoring services vendors. They can easily access the digital ECG tape itself, and scroll forward and backward to any part of the day to verify arrhythmia data and view onset and offset information.
- The physician can conveniently gain access to monitoring data any time of the day and can remotely switch between testing modes (MCT, Holter, or Event) as needed. The system's remote transition capability eliminates the need for patients to visit a hospital and the physician can decide the treatment based on the arrhythmia detection relevance. This effectively replicates the practice of in-hospital monitoring, making patients unencumbered while they are studied in their natural state of activity.
- Doctors using the MoMe® Kardia can bill globally for both clinical and technical services.

NO PATIENT SELF DIAGNOSIS REQUIRED

Among the most important innovations: The MoMe® Kardia provides a continuous data feed for up to 30 days, and it does not rely on a patient clicking a button to indicate they are feeling symptoms. This is a significant improvement over legacy technology because, with this, patients either have to manually activate monitors after they begin experiencing symptoms or they just do not activate the monitor when there is an arrhythmia they do not feel. This causes doctors to miss important onset data and makes it much more difficult to assess and treat arrhythmia properly. The MoMe® Kardia's continuous feed, coupled with on-demand 24/7 access to every heartbeat, makes it much easier for physicians to capture critical data, analyze it, diagnose it, and initiate treatment.

CLINICAL IMPACT

Michael Mazzini, MD, a cardiologist who uses the MoMe® Kardia in his own practice, describes his experiences: "Last week I put Mobile Cardiac Telemetry (MCT) on a patient on a Friday. It picked up 10 to 14 second asystolic events the following morning, and I was able to get a pacemaker in him that morning," relates Dr. Mazzini. "No hospital telemetry stay, no emergency department visit, no unnecessary workups, no delay in diagnosis… Less than 24 hours from MCT hookup to definitive treatment. That's tremendous value to patients, clinicians, hospitals, and payers," he said.

"Patients and physicians are now equipped with greater control over heart monitoring for improved outcomes," said Frost & Sullivan in a release announcing the award decision[2].

SOURCES

1. Frost & Sullivan, Frost & Sullivan Recognizes Industry Leaders at Excellence in Best Practices Awards Ceremony. March 28, 2019, Web.
2. Frost & Sullivan, Technology Leadership Award – The Remote Cardiac Monitoring Industry, North America. 2018.

NEW GUIDELINE PUBLISHED FOR EVALUATION OF VALVULAR REGURGITATION AFTER CATHETER-BASED VALVE INTERVENTIONS

Valvular regurgitation is a prevalent cardiac disorder, in which one or more of the heart's valves "leak," often leading to extra burdens on the heart muscle and requiring treatment. Catheter-based interventions to treat Valvular Heart Disease (VHD) have increased over the past few years with the advent of Transcatheter Aortic Valve Replacement (TAVR), edge-to-edge mitral valve repair, and other investigative devices to repair or replace diseased valves. Guidelines to assess the results of these interventions are lacking, but a new document supplements the previously published guideline *Recommendations for Evaluation of Prosthetic Valves with Echocardiography and Doppler Ultrasound*.

www.asecho.org/wp-content/uploads/2014/05/2009_Evaluation-of-Prosthetic-Valves.pdf

Guidelines for the *Evaluation of Valvular Regurgitation After Percutaneous Valve Repair or Replacement: A Report* from the *American Society of Echocardiography (ASE)* developed in collaboration with the *Society for Cardiovascular Angiography and Interventions, Japanese Society of Echocardiography,* and *Society for Cardiovascular Magnetic Resonance*, provides a resource to guide clinicians in best practices for approaching valvular regurgitation after repair or replacement of a valve.

www.asecho.org/wp-content/uploads/2019/04/Percutaneous-VR_2019.pdf

ASE's Chair of the writing group, William A. Zoghbi, MD, MACC, FASE, of Houston Methodist DeBakey Heart & Vascular Center in Houston, Texas, commented, "This new guideline is timely, as cardiologists and Valvular Heart Disease specialists need consensus on how to evaluate results of catheter-based valve repair or replacement — novel approaches that help many patients with valve disease." Echocardiography is essential in the evaluation of valvular regurgitation after percutaneous interventions and is the first-line tool for evaluation of procedural results. Its assessment, however, is more difficult than in native valvular regurgitation because of the multitude of procedures and hardware involved. This highlights the need for an integrative approach of all information gleaned from various parameters.

The document outlines, in detail, the technical considerations and imaging techniques, as well as the value that 3D echocardiography and cardiac magnetic resonance imaging can add to the diagnostic process. It delves into specific issues with each type of regurgitation,

namely mitral regurgitation (MR), aortic regurgitation (AR), tricuspid regurgitation (TR), and pulmonary regurgitation (PR). The document includes nine useful tables summarizing techniques and advantages of each modality, as well as 23 figures to illustrate various concepts.

In conjunction with the publication of this guideline, Dr. Zoghbi conducted a live webinar. All ASE-hosted guideline webinars are available at: https://aseuniversity.org/ase/.

The full guideline document is available on the *Journal of American Society of Echocardiography (JASE)* website: www.onlinejase.com/. This document and all ASE Guideline documents are also available to the medical community at www.asecho.org/guidelines-search/.

HEART FAILURE DEATHS RISING IN YOUNGER ADULTS

Black Men Under 65 Have Biggest Increase in Premature Heart Failure Deaths

NORTHWESTERN UNIVERSITY

- 6 million adults in US have heart failure
- Rise is likely due to obesity and diabetes epidemics
- Life expectancy in US is dropping, possibly due to heart failure rise
- Heart failure is number one reason adults are admitted to hospital

Death rates due to heart failure are now increasing, and this increase is most prominent among younger adults under 65, considered premature death, reports a new Northwestern Medicine study.

The increase in premature death from heart failure was highest among black men under age 65.

This study is showing for the first time that death rates due to heart failure have been increasing since 2012. The rise in deaths comes despite significant advances in medical and surgical treatments for heart failure in the past decade.

The study was published in the *Journal of the American College of Cardiology* in May.

"The success of the last three decades in improving heart failure death rates is now being reversed, and it is likely due to the obesity and diabetes epidemics," said Dr. Sadiya Khan, assistant Professor of Medicine at Northwestern University Feinberg School of Medicine and a Northwestern Medicine cardiologist. "We focused on patients with heart failure because they have the highest mortality related to cardiovascular death. They have a prognosis similar to metastatic lung cancer."

"An estimated six million adults in the US have heart failure. It's the number one reason older adults are admitted to the hospital," Khan said.

"Given the aging population and the obesity and diabetes epidemics, which are major risk factors for heart failure, it is likely that this trend will continue to worsen," she said.

Recent data that show the average life expectancy in the US also is declining, which compounds Khan's concern that cardiovascular death related to heart failure may be a significant contributor to this change.

The study used data from the *Centers for Disease Control and Prevention's Wide-Ranging Online Data for Epidemiologic Research* data, which includes the underlying and contributing cause of death from all death certificates in the US between 1999 to 2017 for 47,728,569 individuals. Researchers analyzed the age-adjusted mortality rate for black and white adults between the age of 35 to 84 years who died from heart failure.

Simply put, heart failure is when the heart muscle doesn't function properly in its squeezing or relaxing functions. It causes symptoms like shortness of breath and swelling. When the heart can't adequately squeeze to pump blood, it's called heart failure with reduced ejection fraction; when the heart can't relax it's called heart failure with preserved ejection fraction.

"To combat this disturbing trend, we need to focus on improving the control of risk factors, including blood pressure, cholesterol and diabetes," Khan said. "Healthy lifestyle changes promoting a normal body mass index also can protect from developing heart failure as well as engaging in regular physical activity and consuming a healthy, well-balanced diet."

In future research, Khan said she wants to better understand what causes the disparities in cardiovascular death related to heart failure.

CHiP NETWORK
CONGENITAL HEART INTERNATIONAL PROFESSIONALS

The congenital heart professionals network exists to facilitate communications between congenital heart professionals locally, regionally, and globally.

JOIN TODAY www.chip-network.org

Cincinnati Children's Heart Institute

Funded by Cincinnati Children's Heart Institute

PICS-AICS

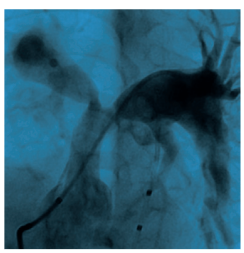
Pediatric and Adult Interventional Cardiac Symposium

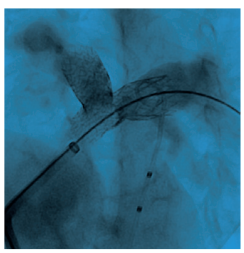
SAVE THE DATE

SEPTEMBER 8-11, 2020

BOSTON

THE WESTIN BOSTON WATERFRONT





LIVE CASE DEMONSTRATIONS | TAPED CASES | HOT DEBATES

ABSTRACT SESSIONS | BREAKOUT SESSIONS

MY NIGHTMARE CASE IN THE CATH LAB

WWW.PICSYMPOSIUM.COM

MEETING CALENDAR

FEBRUARY

7

SUDDEN CARDIAC DEATH IN THE YOUNG IN OHIO: THE POSTMORTEM INVESTIGATION

Akron, OH, USA
asuncire@akronchildrens.org

12–16

CARDIOLOGY 2020

Lake Buena Vista, FL, USA
https://chop.cloud-cme.com/default.aspx?P=5&EID=928

20–21

CTO SUMMIT 2020

New York, NY, USA
https://www.tctmd.com/node/29798

MARCH

25–28

PICS–AICS ISTANBUL

Istanbul, Turkey
http://picsistanbul.com/

28–30

ACC.20

Chicago, IL, USA
https://accscientificsession.acc.org/Information-Pages/future-meetings

MOUNT SINAI DISCOVERS PLACENTAL STEM CELLS THAT CAN REGENERATE HEART AFTER HEART ATTACK

Researchers at the Icahn School of Medicine at Mount Sinai have demonstrated that stem cells derived from the placenta known as Cdx2 cells can regenerate healthy heart cells after heart attacks in animal models. The findings, published in the May 20 issue of *Proceedings of the National Academy of Sciences (PNAS)*, may represent a novel treatment for regenerating the heart and other organs.

"Cdx2 cells have historically been thought to only generate the placenta in early embryonic development, but never before were shown to have the ability to regenerate other organs, which is why this is so exciting. These findings may also pave the way to regenerative therapy of other organs besides the heart," said principal investigator Hina Chaudhry, MD, Director of Cardiovascular Regenerative Medicine at the Icahn School of Medicine at Mount Sinai. "They almost seem like a super-charged population of stem cells in that they can target the site of an injury and travel directly to the injury through the circulatory system and are able to avoid rejection by the host immune system."

This team of Mount Sinai researchers had previously discovered that a mixed population of mouse placental stem cells can help the hearts of pregnant female mice recover after an injury that could otherwise lead to heart failure. In that study, they showed that the placental stem cells migrated to the mother's heart and directly to the site of the heart injury. The stem cells then programmed themselves as beating heart cells to help the repair process.

The new study was aimed at determining what type of stem cells made the heart cells regenerate. The investigators started by looking at Cdx2 cells, the most prevalent stem cell type in the previously identified mixed population and found them to comprise the highest percentage (40%) of those assisting the heart from the placenta.

To test the Cdx2 cells' regenerative properties, the researchers induced heart attacks in three groups of male mice. One group received Cdx2 stem cell treatments derived from end-gestation mouse placentas, one group received placenta cells that did not express Cdx2, and the third group received a saline control. The team used magnetic resonance imaging to analyze all mice immediately after the heart attacks, and three months after induction with cells or saline. They found that every mouse in the group with Cdx2 stem cell treatments had significant improvement and regeneration of healthy tissue in the heart. By three months, the stem cells had migrated directly to the heart injury and formed new blood vessels and new cardiomyocytes (beating heart muscle cells). The mice injected with saline and the non-Cdx2 placenta cells went into heart failure and their hearts had no evidence of regeneration.

Researchers noted two other properties of the Cdx2 cells: they have all the proteins of embryonic stem cells, which are known to generate all organs of the body, but also additional proteins, giving them the ability to travel directly to the injury site, which is something embryonic stem cells cannot do, and they appear to avoid the host immune response. The immune system did not reject these cells when administered from the placenta to another animal.

"These properties are critical to the development of a human stem cell treatment strategy, which we have embarked on, as this could be a promising therapy in humans. We have been able to isolate Cdx2 cells from term human placentas also; therefore, we are now hopeful that we can design a better human stem cell treatment for the heart than we have seen in the past," explained Dr. Chaudhry. "Past strategies tested in humans were not based on stem cell types that were actually shown to form heart cells and use of embryonic stem cells for this goal is associated with ethics and feasibility concerns. Placentas are routinely discarded around the world and thus almost a limitless source."

"These results were very surprising to us, as no other cell type tested in clinical trials of human heart disease were ever shown to become beating heart cells in petri dishes, but these did and they knew exactly where to go when we injected them into the circulation," said first author Sangeetha Vadakke-Madathil, PhD, postdoctoral fellow in Medicine (Cardiology) at the Icahn School of Medicine at Mount Sinai.

NOVEL TECHNIQUE REDUCES OBSTRUCTION RISK IN HEART VALVE REPLACEMENT

The Transcatheter Approach Increases Treatment Options for High-Risk Patients

Researchers at the National, Heart, Lung, and Blood Institute (NHLBI), part of the National Institutes of Health, have developed a novel technique that prevents the obstruction of

NEONATOLOGY TODAY
Peer Reviewed Research, News and Information in Neonatal and Perinatal Medicine

Neonatology Today is interested in publishing manuscripts from Neonatologists, Fellows, NNPs and those involved in caring for neonates on case studies, research results, hospital news, meeting announcements, and other pertinent topics.

Please submit your manuscript to: **LomaLindaPublishingCompany@gmail.com**

blood flow, a common fatal complication of transcatheter mitral valve replacement (TMVR). The new method, called LAMPOON, may increase treatment options for high-risk patients previously ineligible for heart valve procedures. The *Journal of the American College of Cardiology* published the findings online in May.

TMVR is used to treat mitral valve stenosis, a narrowing of the valve that restricts blood flow into the main pumping chamber of the heart. It also treats regurgitation, which occurs when the valve leaks and causes blood to flow back through the valve. Untreated, these conditions can cause pulmonary hypertension, heart enlargement, atrial fibrillation, blood clots, and heart failure.

For elderly or frail patients, TMVR offers a less invasive alternative to open heart surgery. During TMVR, doctors replace the mitral valve by delivering an artificial valve through a long, thin, flexible tube, called a catheter, through blood vessels and into the heart. But in more than 50% of patients, the heart's anatomy gets in the way. The heart leaflet is pushed back and blocks blood flow. This is known as left ventricular outflow tract (LVOT) obstruction, a common and the most life-threatening complication of TMVR.

"These patients have a failing mitral valve, are not able to undergo open heart surgery, and are now rejected as candidates for TMVR because of the very high risk of left ventricular outflow tract obstruction," said study author Jaffar M. Khan, MD, clinician at NHLBI.

To increase the availability of TMVR for this subset of patients, Khan and colleagues at NHLBI and Emory University developed a procedure that makes an intentional laceration of the anterior mitral leaflet to prevent left ventricular outflow tract obstruction, dubbed LAMPOON.

In the LAMPOON procedure, the operator inserts two catheters through the patient's groin, and then through the blood vessels until it reaches the heart. The doctor then uses an electrified wire the size of a sewing thread woven through the catheter to split open the leaflet. At that point, the patient is ready to undergo TMVR.

"Surgeons cut out the leaflets when they replace valves. They can do it, because they have cut open the chest and the heart and can clearly see the problem. LAMPOON is designed for patients who need a new mitral valve, but can't, or may not want to undergo open heart surgery," said Khan.

According to the researchers, other preventive strategies have had largely suboptimal outcomes.

Between June 2017 and June 2018, the LAMPOON study enrolled 30 patients, median age 76, considered at high risk for surgical valve replacement and at prohibitive risk of LVOT obstruction during TMVR.

All patients survived the procedure and 93% reached the 30-day survival mark, which compares favorably to a 38% reported with other methods. The primary outcome of the study, which combined a successful LAMPOON, followed by a successful TMVR without re-intervention, was achieved in 73% of the patients.

The researchers hope the technique will eventually help reduce the number of deaths from heart valve disease. Every year, approximately five million people in the United States are diagnosed with heart valve disease, and more than 20,000 Americans die of the disease each year, according to the American Heart Association.

WHAT IS KNOWN -- AND NOT KNOWN -- ABOUT HEART MUSCLE DISEASES IN CHILDREN

Cardiomyopathies (heart muscle diseases) in children are the focus of a new scientific statement from the *American Heart Association* that provides insight into the diagnosis and treatment of the diseases as well as identifying future research priorities. It will be published in the *American Heart Association's* journal.

When a child has certain types of cardiomyopathy, their heart is not able to pump blood efficiently. Symptoms may include difficulty breathing, heart rhythm abnormalities, dizziness, swollen hands and feet or other indications.

Although rare, cardiomyopathies in children can be life threatening and often result in either a child needing a heart transplant or premature death. There are many causes of cardiomyopathies including genetic variations that affect basic heart functions, systemic diseases, such as infections, treatments for medical conditions that injure the heart and many others, some of which are not yet understood.

According to the statement:

- Nearly 40% of children who are diagnosed with cardiomyopathies that produce symptoms receive a heart transplant or die within the first two years after diagnosis.
- The percentage of children with cardiomyopathy who received a heart transplant has not declined over the past 10 years.
- Cardiomyopathy remains the leading cause of transplantation for children over one year of age.

"This statement is designed to give medical professionals an overview of what we currently know about cardiomyopathies in children. Although we are able to provide effective treatments in many cases, research is urgently needed to better understand the causes of the diseases so we can help children with cardiomyopathies live their best lives," said Steven E. Lipshultz, MD, the Chair of the Writing Group and the A. Conger Goodyear Professor and Chair of the Department of Pediatrics at the Jacobs School of Medicine and Biomedical Sciences at the University at Buffalo.

Dr. Lipshultz is also the Chair of the Medical Advisory Board of the *Children's Cardiomyopathy Foundation*, which partners with the *American Heart Association* on funding pediatric cardiomyopathy research grants.

CONGENITAL CARDIOLOGY TODAY

CORPORATE OFFICE

11502 Elk Horn Drive
Clarksburg, MD 20871 USA

CORPORATE TEAM

FOUNDER & SENIOR EDITOR
Tony Carlson
Tony@cct.bz

CO-FOUNDER & MEDICAL EDITOR
John W. Moore, MD, MPH
Dr.John@cct.bz

EDITOR-IN-CHIEF
Kate Baldwin
Kate@cct.bz

STAFF EDITOR
Loraine Watts

EDITOR-IN-CHIEF EMERITUS
Richard Koulbanis

STAFF EDITOR & WRITER
Virginia Dematatis

EDITORIAL BOARD

Teiji Akagi, MD
Zohair Al Halees, MD
Mazeni Alwi, MD
Felix Berger, MD
Fadi Bitar, MD
Jacek Bialkowski, MD
Mario Carminati, MD
Anthony C. Chang, MD, MBA
John P. Cheatham, MD
Bharat Dalvi, MD, MBBS, DM
Horacio Faella, MD
Yun-Ching Fu, MD
Felipe Heusser, MD
Ziyad M. Hijazi, MD, MPH
Ralf Holzer, MD
Marshall Jacobs, MD
R. Krishna Kumar, MD, DM, MBBS
John Lamberti, MD
Gerald Ross Marx, MD
Tarek S. Momenah, MBBS, DCH
Toshio Nakanishi, MD, PhD
Carlos A. C. Pedra, MD
Daniel Penny, MD, PhD
James C. Perry, MD
Shakeel A. Qureshi, MD
P. Syamasundar Rao, MD
Andrew Redington, MD
Carlos E. Ruiz, MD, PhD
Girish S. Shirali, MD
Horst Sievert, MD
Hideshi Tomita, MD
Gil Wernovsky, MD
Zhuoming Xu, MD, PhD
William C. L. Yip, MD
Carlos Zabal, MD

OFFICIAL PUBLICATION OF
THE CHiP NETWORK

Statements or opinions expressed in Congenital Cardiology Today reflect the views of the authors and sponsors, and are not necessarily the views of Congenital Cardiology Today.

© 2020 by Congenital Cardiology Today

ISSN 1554-7787 print. ISSN 1554-0499 electronic.
Published monthly.

All rights reserved.