CONGENITAL CARDIOLOGY TODAY
Timely News & Information for Congenital/Structural Cardiologists & Cardiothoracic Surgeons Worldwide
International Edition
Vol. 19 - Issue 12
December 2021

## Table of Contents



# Biatrial Connection of Superior Vena Cava: Unusual Presentations of a Rare Disease in Two Children

*Sulafa M. Ali, FRCPCH, FACC; Abdelrahman Atya, MD; Noha Karadawi, MD, MRCPCH; Amna Mamoun, MD*

## Abstract

Biatrial connection of the superior vena cava is a rare congenital anomaly leading to cyanosis and consequent complications. Diagnosis can be missed on routine echocardiography. We describe two children with this defect, one presented with early onset of heart failure and the second with a brain abscess. Diagnosis was missed on the first echocardiogram for both patients. The defect was associated with narrowing of the right superior vena cava to right atrium opening leading to predominant flow to the left atrium in both patients. Contrast echocardiogram confirmed the diagnosis in both cases. Medical and surgical management issues are discussed.

## Introduction

Anomalous connections of systemic veins are rare congenital heart defects. Biatrial right superior vena cava (RSCV) connection had been first described by postmortem examination in 1914.[1] In the 1960s to 1980s few cases were described, mostly in adults.[2,3,4,5] Up to 2020, only 12 cases (with six pediatric patients) had been described with this diagnosis.[6] The defect is typically located at the common wall that separates the RSVC from the right upper pulmonary vein, the area where sinus venosus atrial septal defect (ASD) is located. As the defect may overlap or co-exist with sinus venosus ASD, its true incidence may be difficult to identify. The most common presentation of this defect is unexplained cyanosis which can be mild leading to delay in diagnosis up to adulthood. Echocardiography (echo) diagnosis can be challenging, and routine transthoracic echo may miss this lesion. We describe two cases of biatrial connection of RSVC that were missed on the initial echo examination. Rare associations are described.

## Case 1

A 21-month-old boy was referred to our clinic with shortness of breath since the age of four months. He was initially seen by a pediatric cardiologist and diagnosed as having sinus venosus ASD. Physical examination revealed tachypnea, a respiratory rate of 40/minute with intercostal retractions and Harrison's sulci. The heart rate was 120 beats/minute. There were no dysmorphic features. His weight was 8.5K g (four standard deviations below the mean for his age) and oxygen saturation was 87% in room air. The left ventricle apex was felt at the 5$^{th}$ intercostal space lateral to the mid clavicular line, and there was a positive

# DECEMBER 2021

International Edition Vol. 19 - Issue 12

## TABLE OF CONTENTS

right ventricle heave. On auscultation, the first heart sound was normal and the second was loud. There was an ejection systolic murmur grade II/VI heard best at the left second intercostal space.

Chest x-ray showed cardiomegaly with RA and right ventricle enlargement and pulmonary plethora. Electrocardiogram showed right ventricle enlargement of volume overload pattern. Echo showed situs solitus, atriovenetricular and ventricuoloarterial concondrance. There was biatrial connection of the RSVC, mostly to the left atrium (LA). The RSVC/right atrium (RA) communication was narrow, measuring 5 mm, while the left atrium (LA) communication was 9 mm with a mean Doppler gradient of 0.8 mmHg. There was an ASD of sinus venosus type measuring 10 mm with left-to-right shunt **(Figure 1)**. The right upper pulmonary vein drained to the RSVC; the three other pulmonary veins drained normally to LA. The right ventricle was dilated with paradoxical septal motion. The LA was mildly dilated (LA: aorta ratio 1.5:1). There was no left SVC, the other anatomy and function were unremarkable.

An echo contrast study using agitated saline in the left cubital vein showed simultaneous appearance of the bubbles in both atria, LA having a denser contrast than RA and earlier filling of the left ventricle was demonstrated **(Figure 2)**. The case was referred for discussion with the surgeon.

## Case 2

A five-year-old boy, presented to the pediatrician with high grade fever and vomiting; brain computed tomography scan showed a brain abscess which was drained surgically. He was noted to be clubbed and cyanosed and referred for cardiac assessment. He did not experience any cardiovascular symptoms. Physical examination revealed no dysmorphic features and mild clubbing; the weight was 16 Kg (at the 5th percentile for his age) and oxygen saturation was 85% in room air.

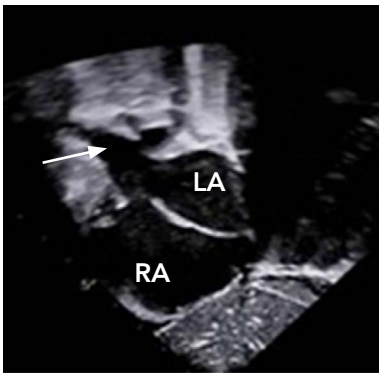



**FIGURE 1**
*Subcostal sagittal (bicaval) view of Patient 1 showing a sinus venosus ASD, RSVC (arrow) opening into both atria with narrow RSVC/RA junction. ASD: Atrial Septal Defect. RSVC: Right Superior Vena Cava. RA: Right Atrium.*

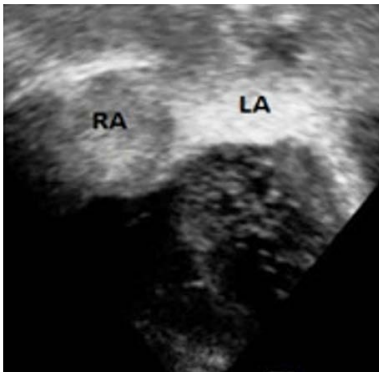



**FIGURE 2**
*Four chamber view of Patient 1 showing agitated saline injection into the left cubital vein with simultaneous contrast appearance in both atria (denser in LA and showing in left ventricle). LA: Left Atrium. RA: Right Atrium.*

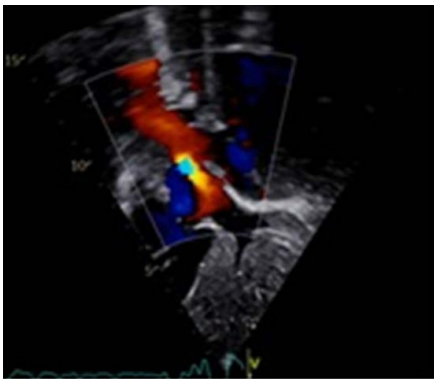


**FIGURE 3**
*Subcostal sagittal (bicaval) view with color Doppler of Patient 2 showing RSVC (arrow) opening into both atria with narrower RSVC/RA junction compared with Patient 1. RSVC: Right Superior Vena Cava. RA: Right Atrium.*

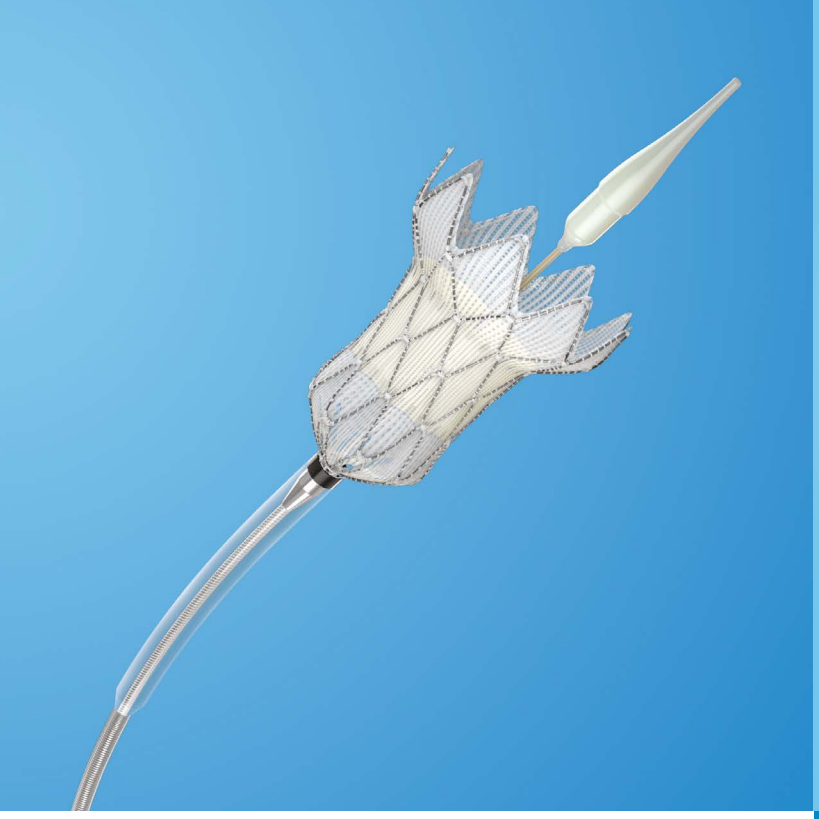
Echo was initially reported as normal, suspicion of pulmonary arterio-venous malformation led to performing a contrast echo study using agitated saline which revealed bubbles simultaneously filling both atria. Repeated echo revealed biatrial drainage of RSVC which mainly drains to LA. The RSVC to RA communication was stenosed with a mean gradient of 3.8 mmHg **(Figure 3)**. No clear ASD was seen. The right upper pulmonary vein was seen draining to the RSVC; the other three pulmonary veins were seen draining to the LA. LA was mildly dilated (LA: aorta ratio 1.5:1). There was no left SVC and the other anatomy and function were normal. Chest X-ray and electrocardiogram were normal. The case was referred for surgical opinion.

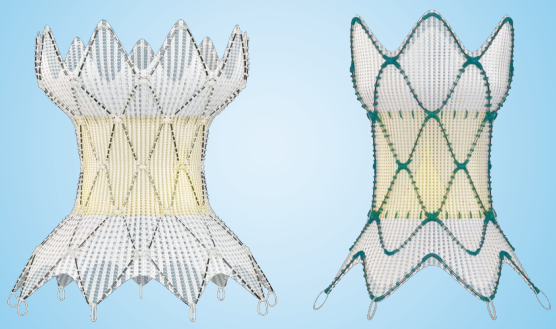
## Discussion

Biatrial drainage of RSVC has been described as a rare anomaly; more cases have recently been reported due to improvement in imaging modalities such as cardiac contrast-enhanced computed tomography and magnetic resonance. With improving echo techniques antenatal diagnosis has also been described.[6,7] However, as evident from our cases, transthoracic echo can miss the diagnosis and operators need to have a high level of suspicion, particularly if the cause of cyanosis is not obvious.

Although in sinus venosus ASD the RSVC overrides the atrial septum, the blood flow is usually directed to the RA; therefore, there is no cyanosis in this condition. In the current two patients there is narrowing of the RSVC/RA opening leading to predominant shunting of RSVC blood into the LA, and subsequently, desaturation. Similar findings had been described by Van Praagh.[9] If the RSVC to RA orifice is atretic, then the RSVC is directly connected to LA leading to a similar hemodynamic effect. This later condition had been reported in 45 cases including 26 children.[6]

Our first patient represents a rare occasion of early onset of heart failure in the setting of biatrial RSVC and sinus venosus ASD. In the latter lesion, heart failure typically occurs in young adults





# ONE. PARTNER.

Every step of the way

## Harmony™

Transcatheter Pulmonary Valve (TPV)

Backed by our commitment to providing solutions and support to you and your CHD patients, we offer dedicated clinical experts who can provide guidance in all stages of the therapy.

medtronic.com

Medtronic

**Indications**
The Harmony™ transcatheter pulmonary valve (TPV) system is indicated for use in the management of pediatric and adult patients with severe pulmonary regurgitation (i.e., severe pulmonary regurgitation as determined by echocardiography and/or pulmonary regurgitant fraction ≥ 30% as determined by cardiac magnetic resonance imaging) who have a native or surgically-repaired right ventricular outflow tract and are clinically indicated for surgical pulmonary valve replacement.

**Contraindications**
The following are contraindications for the use of this device: active bacterial endocarditis or other active infections, known intolerance to Nitinol (titanium or nickel), or an anticoagulation/antiplatelet regimen.

**Warnings**
*General:* Implantation of the Harmony TPV system should be performed only by physicians who have received Harmony TPV system training. The transcatheter pulmonary valve (TPV) is to be used only in conjunction with the Harmony delivery catheter system (DCS). This procedure should only be performed where emergency pulmonary valve surgery can be performed promptly. Do not use any of the Harmony TPV system components if any of the following has occurred: it has been dropped, damaged, or mishandled in any way, or if the use-by date has elapsed.

*Transcatheter pulmonary valve (TPV):* This device was designed for single use only. Do not reuse, reprocess, or resterilize the TPV. Reuse, reprocessing, or resterilization may compromise the structural integrity of the device and/or create a risk of contamination of the device, which could result in patient injury, illness, or death. Do not resterilize the TPV by any method. Exposure of the device and container to irradiation, steam, ethylene oxide, or other chemical sterilants renders the device unfit for use. The device is packaged with a temperature sensor. Do not freeze the device. Do not expose the device to extreme temperatures. Do not use the device if the arrow on the sensor points to the symbol that indicates that the temperature limit has been exceeded. Do not use the device if any of the following have occurred: the tamper-evident seal is broken, the serial number tag does not match the container label, the arrow on the sensor points to the symbol that indicates that the temperature limit has been exceeded, or the device is not completely covered by the storage solution. Do not contact any of the Harmony TPV system components with cotton or cotton swabs. Do not expose any of the Harmony TPV system components to organic solvents, such as alcohol. Do not introduce air into the catheter. Do not expose the device to solutions other than the storage and rinse solutions. Do not add or apply antibiotics to the device, the storage solution, or the rinse solution. Do not allow the device to dry. Maintain tissue moisture with irrigation or immersion. Do not attempt to repair a damaged device. Do not handle the valve leaflet tissue or use forceps to manipulate the valve leaflet tissue. Do not attempt to recapture the device once deployment has begun. Do not attempt to retrieve the TPV if any one of the outflow TPV struts is protruding from the capsule. If any one of the outflow TPV struts has deployed from the capsule, the TPV must be released from the catheter before the catheter can be withdrawn. Do not attempt post-implant balloon dilatation (PID) of the TPV during the procedure, which may cause damage to or failure of the TPV leading to injury to the patient resulting in reintervention.

*Delivery catheter system (DCS):* This device was designed for single use only. Do not reuse, reprocess, or resterilize the DCS. Reuse, reprocessing, or resterilization may compromise the structural integrity of the device and/or create a risk of contamination of the device, which could result in patient injury, illness, or death. Do not reuse or resterilize the DCS. If resistance is met, do not advance the guidewire, DCS, or any other component without first determining the cause and taking remedial action. Do not remove the guidewire from the DCS at any time during the procedure.

**Precautions**
*General:* Clinical long-term durability has not been established for the Harmony TPV. Evaluate the TPV performance as needed during patient follow-up. The safety and effectiveness of Harmony TPV implantation in patients with pre-existing prosthetic heart valve or prosthetic ring in any position has not been demonstrated. The Harmony TPV system has not been studied in female patients of child-bearing potential with positive pregnancy.

*Before use:* Exposure to glutaraldehyde may cause irritation of the skin, eyes, nose, and throat. Avoid prolonged or repeated exposure to the chemical vapor. Use only with adequate ventilation. If skin contact occurs, immediately flush the affected area with water (for a minimum of 15 minutes) and seek medical attention immediately. The TPV and the glutaraldehyde storage solution are sterile. The outside of the TPV container is nonsterile and must not be placed in the sterile field. The TPV and DCS should be used only in a sterile catheterization laboratory (cath lab) environment. Ensure that sterile technique is used at all times. Strictly follow the TPV rinsing procedure. For TPV 25: Ensure that all green sutures have been removed from the attachment suture loops on the TPV before loading onto the DCS. Prevent contamination of the TPV, its storage solution, and the DCS with glove powder. Verify the orientation of the TPV before loading it onto the DCS. The inflow end of the TPV with attachment suture loops must be loaded first. Do not place excessive pressure on the TPV during loading. Inspect the sealed DCS packaging before opening. If the seal is broken or the packaging has been damaged, sterility cannot be assured. Proper functioning of the DCS depends on its integrity. Use caution when handling the DCS. Damage may result from kinking, stretching, or forceful wiping of the DCS. This DCS is not recommended to be used for pressure measurement or delivery of fluids. Carefully flush the DCS and maintain tight DCS connections to avoid the introduction of air bubbles.

*During use:* The TPV segment is rigid and may make navigation through vessels difficult. Do not advance any portion of the DCS under resistance. Identify the cause of resistance using fluoroscopy and take appropriate action to remedy the problem before continuing to advance the DCS. Careful management of the guidewire is recommended to avoid dislodgement of the TPV during DCS removal. Once deployment is initiated, retrieval of the TPV from the patient is not recommended. Retrieval of a partially deployed valve may cause mechanical failure of the delivery catheter system or may cause injury to the patient. Refer to the section below for a list of potential adverse events associated with Harmony TPV implantation. During deployment, the DCS can be advanced or withdrawn prior to the outflow struts protruding from the capsule. Once the TPV struts contact the anatomy during deployment, it is not recommended to reposition the device. Advancing the catheter forward once the TPV struts make contact with the anatomy may lead to an undesired deployment or may cause damage to or failure of the TPV and injury to the patient. Refer to the section below for a list of potential adverse events associated with the Harmony TPV implantation. Physicians should use judgment when considering repositioning of the TPV (for example, using a snare or forceps) once deployment is complete. Repositioning the bioprosthesis is not recommended, except in cases where imminent serious harm or death is possible (for example, occlusion of the main, left, or right pulmonary artery). Repositioning of a deployed valve may cause damage to or failure of the TPV and injury to the patient. Refer to the section below for a list of potential adverse events associated with the Harmony TPV implantation. Ensure the capsule is closed before DCS removal. If increased resistance is encountered when removing the DCS through the introducer sheath, do not force passage. Increased resistance may indicate a problem and forced passage may result in damage to the device and harm to the patient. If the cause of resistance cannot be determined or corrected, remove the DCS and introducer sheath as a single unit over the guidewire, and inspect the DCS and confirm that it is complete. If there is a risk of coronary artery compression, assess the risk and take the necessary precautions. Endocarditis is a potential adverse event associated with all bioprosthetic valves. Patients should make their healthcare providers aware that they have a bioprosthetic valve before any procedure. Post-procedure, administer appropriate antibiotic prophylaxis as needed for patients at risk for prosthetic valve infection and endocarditis. Prophylactic antibiotic therapy is recommended for patients receiving a TPV before undergoing dental procedures. Post-procedure, administer anticoagulation and/or antiplatelet therapy per physician/clinical judgment and/or institutional protocol. Excessive contrast media may cause renal failure. Preprocedure, measure the patient's creatinine level. During the procedure, monitor contrast media usage. Conduct the procedure under fluoroscopy. Fluoroscopic procedures are associated with the risk of radiation damage to the skin, which may be painful, disfiguring, and long term.

**Potential Adverse Events**
Potential risks associated with the implantation of the Harmony TPV may include, but are not limited to, the following: ▪ death ▪ valve dysfunction ▪ tissue deterioration ▪ hematoma ▪ heart failure ▪ cerebrovascular incident ▪ perforation ▪ rupture of the right ventricular outflow tract (RVOT) ▪ compression of the aortic root ▪ compression of the coronary arteries ▪ sepsis ▪ pseudoaneurysm ▪ erosion ▪ stent fracture ▪ arrhythmias ▪ device embolization or migration ▪ pulmonary embolism ▪ occlusion of a pulmonary artery ▪ laceration or rupture of blood vessels ▪ device misorientation or misplacement ▪ valve deterioration ▪ regurgitation through an incompetent valve ▪ physical or chemical implant deterioration ▪ paravalvular leak ▪ valve dysfunction leading to hemodynamic compromise ▪ residual or increasing transvalvular gradients ▪ progressive stenosis and obstruction of the implant ▪ hemorrhage ▪ endocarditis ▪ thromboembolism ▪ thrombosis ▪ thrombus ▪ intrinsic and extrinsic calcification ▪ bleeding ▪ bleeding diathesis due to anticoagulant use ▪ fever ▪ pain at the catheterization site ▪ allergic reaction to contrast agents ▪ infection ▪ progressive pulmonary hypertension ▪ progressive neointimal thickening and peeling ▪ leaflet thickening ▪ hemolysis. General surgical risks applicable to transcatheter pulmonary valve implantation: ▪ abnormal lab values (including electrolyte imbalance and elevated creatinine) ▪ allergic reaction to antiplatelet agents, contrast medium, or anesthesia ▪ exposure to radiation through fluoroscopy and angiography ▪ permanent disability.

Please reference the Harmony TPV system instructions for use for more information regarding indications, warnings, precautions, and potential adverse events.

**Caution:** Federal law (USA) restricts these devices to the sale by or on the order of a physician.

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.structuralheart@medtronic.com



UC202112006 EN
03/2021

**medtronic.com**

Medtronic

where right ventricle volume overload is poorly tolerated. This early onset of heart failure could be explained by the volume overload of the left side of the heart caused by the RSVC adding to the right-side volume overload due to ASD. The presence of heart failure and low saturation in infants with ASD should alert the cardiologist to the presence of other lesions such as anomalous pulmonary or systemic venous drainage.

Brain abscess is a well-known complication of cyanotic heart disease. This complication had been reported in adult patients with biatrial SVC connection; however, it is exceedingly rare in children and, to our knowledge, only one case reported.[6] In our patient, the brain abscess was the presenting feature that led to the diagnosis, emphasizing the role of careful examination and detailed echo for such patients.

Echo diagnosis of biatrial RSVC needs a high index of suspicion and careful examination of subcostal bicaval view. If a standard echo is negative, a contrast study should be considered in patients with unexplained low oxygen saturation. Agitated saline is sensitive and has a documented role in diagnosis of anomalous systemic veins and other conditions such as unroofed coronary sinus.[10] Cardiac computed tomography and magnetic resonance imaging can delineate the detailed anatomy of the SVC and pulmonary veins,[7] their role in diagnosis supersedes invasive tools such as cardiac catheterization which is reserved for hemodynamic assessment of older patients.

Important clinical implications of biatrial RSVC include the need to avoid intravenous infusions using upper limb veins in order to prevent systemic thrombo embolism. In addition, an abnormal course during systemic venous cannulation or pacemaker insertion can lead to inadvertent positioning if the diagnosis was not established prior to the procedure.

The surgical management of this condition aims to redirect the pulmonary veins to the LA and the RSVC to the RA without causing obstruction to flow. Many techniques had been described, most commonly involving transection of the RSVC and anastomosing it to the RA appendage (Warden's procedure) as described for repair of superior sinus venosus defects with a high placement of the anomalously draining pulmonary veins. For a group of 23 patients who underwent surgical repair, the operative mortality remains high at 8.6%.[6] Recently, closure of sinus venosus ASD using covered stents had been introduced with acceptable results, which can potentially be applied to our first patient.[11]

## Conclusion

We described two rare presentations: early onset of heart failure and brain abscess in two children with biatrial connection of RSVC and dominant RSVC to LA drainage emphasizing the need for a high index of suspicion in cyanosed patients and the role of contrast echo in diagnosis.

## Learning Points

- Detailed echo including careful subcostal sweeping in patients with low O2 saturation
- Agitated saline contrast echo has a diagnostic in cases where low O2 saturation is unexplained
- Brain abscess can be secondary to rare congenital anomalies such as anomalous systemic venous drainage

**Conflict of Interest:** None declared

## References

1. Nützel H. Beitrag zur Kenntnis der Missbildungen im Bereiche der oberen Hohlvene. Frankf Z Pathol 1914;15:1–19. [Google Scholar]
2. von Hackensellner HA. Abnorme einmundung von lungenvenen in die vena cava cranialis. Virchows Arch. 1955;327(2):603-606.
3. Breitfelner VG. Zur Frage der ''physiologischen'' rechts- bzw.linksdrainage; bei einmundung von Lungenvenen in die obere hohlvene. Zentralbl Allg Pathol. 1963;105: 285-289.
4. Laboux L, Bouhour JB, Dupon H, Cornet E. Retours veineux anormaux. Arch Mal Coeur. 1968;61(4): 1035-1042.
5. Bharati S, Lev M. Direct entry of the right superior vena cava into the left atrium with aneurysmal dilatation and stenosis at its entry into the right atrium with stenosis of the pulmonary veins; a rare case. Pediatr Cardiol. 1984;5(8): 123-126.
6. Ujjwal Kumar Chowdhury, Robert H Anderson, Niwin George, Sukhjeet Singh, Lakshmi Kumari Sankhyan, Sushama B Gayatri et al. A Review of the Surgical Management of Anomalous Connection of the Right Superior Caval Vein to the Morphologically Left Atrium and Biatrial Drainage of Right Superior Caval Vein.World J Pediatr Congenit Heart Surg. 2020 Jul;11(4):466-484. doi: 10.1177/2150135120912677.
7. Mehul D. Patel, Sowmya Balasubramanian, Adam L. Dorfman, Aparna Joshi, Jimmy C. Lu, Maryam Ghadimi Mahani et al. Biatrial Drainage of the Right Superior Vena Cava: Imaging Findings. Radiol Cardiothorac Imaging. 2020 ; 2: e200414. doi: 10.1148/ryct.2020200414.
8. M Vassallo, M Pascotto, C Pisacane, G Santoro, D Paladini, M G Russo et al. Right superior vena cava draining into the left atrium: prenatal diagnosis and postnatal management. Ultrasound Obstet Gynecol 2006 ;27:445-8. doi: 10.1002/uog.2758.
9. Van Praagh S., Geva T., Lock J.E., Nido P.J., Vance M.S., Van Praagh R. Biatrial or left atrial drainage of the right superior vena cava: anatomic, morphogenetic, and surgical considerations–report of three new cases and literature review. Pediatr Cardiol. 2003;24(4):350–363. [PubMed] [Google Scholar]

10. Shetkar SS, Gupta SK, Gulati GS, Juneja R. Right superior vena cava to left atrium: importance of "bubbling it right." Echocardiography.2014;31: E161-E162.
11. Hussein A M Abdullah, Hussein A Alsalkhi, Khalid A Khalid. Transcatheter closure of sinus venosus atrial septal defect with anomalous pulmonary venous drainage: Innovative technique with long-term follow-up. Cardiovasc Interv. 2020 ;95:743-747. doi: 10.1002/ccd.28364.

**SULAFA M ALI, FRCPCH, FACC**

Consultant Pediatric Cardiologist
Sudan Heart Center
Professor, University of Khartoum
Sudan
sulafaali2000@gmail.com

**ABDELRAHMAN ATYA, MD**

Pediatric Cardiology Fellow
Sudan Heart Center
Sudan

**NOHA KARADAWI, MD, MRCPCH**

*Consultant Pediatric Cardiologist*
Ahmed Gasim Cardiac Center
Sudan

**AMNA MAMOUN, MD**

Fellow Pediatric Cardiology
Sudan Heart Center
Sudan

Interventional Systems
B BRAUN
SHARING EXPERTISE


# INCREASE YOUR REACH

## with the sizing balloon catheter trusted for over a decade.

### What Clinicians Like:

- Comprehensive size offering with diameters ranging from 20mm – 40mm
- Four radiopaque marker bands to enable accurate defect sizing
- Low profile design for ease of deliverability and compatibility with D'VILL™ Introducer (65cm)
- Braided inner lumen for optimal pushability, trackability and kink-resistance

ESTABLISHED. PROVEN. EFFECTIVE.

# PTS-X™

## Sizing Balloon Catheter

Distributed by:
B. Braun Interventional Systems Inc. | Part of the B. Braun Group of Companies
Bethlehem, PA | USA | 877-836-2228 | www.bisusa.org

# The PICS Society Advocacy Program: Power in Numbers! Part 3

*Bharat Dalvi, MD, DM, FPICS; Natalie Poli, Ed.S; Kamel Shibbani, MD; Norm Linsky, MPA, MA*

As a global community dedicated to minimally invasive treatment of Congenital Heart Disease (CHD), do we have shared policy goals regardless of where we live and work? If so, how can we achieve those goals? As Dr. Shibbani says, "Do we have ***power in numbers?"***

In Part 1 (August 2021, CCT) Dr. Alejandro Peirone (Argentina) started our global dialogue about ADVOCACY—how we can advocate for safe, effective, affordable patient care. In Part 2 (September 2021, CCT) Drs. Ziyad Hijazi (Qatar) and John Cheatham (USA) continued the dialogue along with PICS Senior Patient Advocate Ms. Natalie Poli (USA). This month we continue our series through an interview with Dr. Bharat Dalvi (Mumbai, India), a PICS Board member and global leader. (Next month we conclude this series with an interview with another global leader, Dr. Hideshi Tomita in Tokyo, Japan).

***Dr. Shibbani:*** What are the most important advocacy issues facing you and your colleagues in India? Are there ways that PICS can help achieve them?

***Dr. Dalvi:*** One of the most pressing issues in India is the limited availability of patient care at all levels from primary to tertiary. We have a huge gap between supply and demand, with far too few centers to meet demand. In India 250,000 to 300,000 babies are born annually with CHD. Of those, 80,000-100,000 need some form of intervention (transcatheter/surgical), but our current socioeconomic environment allows for only about 25,000-30,000 to be treated; that's a painfully small number. Economics and affordability are the main issues (apart from social and educational factors) which delay or deny necessary treatment. This is true throughout the developing world.

**"My personal feeling is that globally 90% of the resources are used to treat 10% of the population. Organizations such as PICS should address this: How can we cut costs without sacrificing quality? We have to evolve strategies and protocols that are as safe as anywhere else in the world, but are less expensive. The hard reality: you can't pretend to have money you don't have."** ***—Bharat Dalvi, MD, DM, FPICS***

We have much to do in terms of education about CHD in three areas. First, our primary care colleagues often are not trained to detect CHD at an early age; this can be changed with continuing medical education programs emphasizing various protocols for detecting CHDs in newborns and early infancy. We must also improve awareness that we can treat CHD far more effectively and safely than in the past, with the majority going on to live productive and fruitful lives. This is especially critical for primary care providers in remote areas. Second, we must train more pediatric cardiologists, nurses, technicians, intensive care specialists, anesthetists and surgeons in India. Dr. Shibbani, you noted the 2021 PICS Symposium included a new program for interventional fellows and early career doctors. I am encouraged that the program was well-received and will be offered again in 2022. As a professional society we must continue training via every avenue possible. Partnering with national societies to train medical providers throughout their careers is so important. I envision selected PICS faculty teaming with faculty in my country to offer such programming online and in-person. *[Editor's Note: Subsequent to this interview, Dr. Dalvi created a highly successful partnership with the Pediatric Cardiac Society of India (PCSI) and provided such a joint program during the PCSI's Annual Congress in October 2021. Thank you to PCSI Congress Chair Prof. Ramakrishna and his distinguished team! Details in a future CCT column.]* Third, we must stress the importance of public education. Early detection is so important to improve quality of life and life expectancy. Too often in the "olden days" a diagnosis of CHD led to doctors advising there was little they could do. Now there ***is*** much we can do—the word needs to get out to the public and to those who hold the purse strings.

***Mrs. Poli:*** Dr. Dalvi you are so right, and I was so lucky. My CHD was undetected for many years. It wasn't until I had a stroke as an adult that I was properly treated. Thank you to my own physicians Drs. Ziyad Hijazi and Cliff Kavinsky! Creating awareness is a global issue. Early detection and treatment are vital everywhere in the world.

***Dr. Dalvi:*** Thank you Natalie. People like you are the role models. This is one of the best ways to spread awareness. No matter how hard we as physicians try, a patient advocate can be so critical in communicating with patients, families and the public. When patients tell their stories and successes, the impact is so real.

***Dr. Shibbani:*** Let's address the other issue you raised, not enough specialists in the developing world. Advocating for change is something PICS could address in partnership with PCSI. Many PICS members participate in medical missions to treat patients and train local providers in regions lacking access to specialized care. Is this an area where PICS can help?

***Dr. Dalvi:*** Yes! The need is great as is our collective ability to help. When I began my practice in 1994, there were perhaps five pediatric cardiologists in the entire country. Now we have about 500; progress, but for a population of 1.4 billion there are still too few. PICS can help. First, it can support efforts of PCSI and other organizations to plan medical missions to regions in need. Second, PICS can partner with those organizations to increase the knowledge base amongst pediatric cardiologists in India. This will give them expanded perspectives into patient management

and build relationships with peers globally. One thought among many: include India-based fellows and early career doctors in the annual PICS Fellows/Early Career Course. Giving them exposure to what is possible, what is excellence, learning from experienced providers would be invaluable to physicians-in-training.

***Dr. Shibbani:*** What would be topics we can advocate for globally?

***Dr. Dalvi:*** My personal feeling is that globally 90% of the resources are used to treat 10% of the population. This leaves us with 10% of the resources to care for 90% of the population. Organizations such as PICS should address this: how can we serve the lower part of the economic/access pyramid? How can we cut costs without sacrificing quality? The hard reality: you can't pretend to have money you don't have.

We have to evolve strategies and protocols that are as safe as anywhere else in the world, but are less expensive. That requires a lot of thinking and hard decisions. People like me who have grown up in this challenging environment have insights into how we can save money without sacrificing excellent care or causing infection.

In much of the developing world, of necessity there are protocols for identifying certain items that can be fully sterilized and re-used, with similar protocols to discard items that are one-time use. In many parts of the world, there simply is no other choice. This is a controversial area, which should be carefully explored through open dialogue, rigorous research and equally rigorous outcomes tracking.

There are many items that must never be re-used no matter how well re-sterilized. For certain other procedures this may be possible if the appropriate controls are in place. If the goal is to improve access to safe, effective care, there are many countries where the situation is dire. Guidance from PICS as a society with global standing could make a difference to hundreds of thousands in underserved regions.

***Dr. Shibbani:*** In that context, what are your thoughts regarding clinical guidelines?

***Dr. Dalvi:*** In the developing world we work hard to make decisions based on textbooks, journals and guidelines based on those materials. Authors frequently come from backgrounds with more resources than in the developing world. PICS has made important steps to diversify its leadership and committees—this is welcome and will continue. At the doctor-patient level, if a patient comes to me who has the resources, has researched the developed world literature and wants an expensive validated procedure, I have no problem with that. But if another patient comes to me and may have to spend his or her entire life savings on a procedure where less expensive, highly effective alternative treatments are available, a conversation needs to happen. This is the reality we face in our hospitals today.

***Dr. Shibbani:*** I have seen guidelines in the infectious diseases area where they factored in the difficult economic realities and choices based on whether one is in a developed vs a developing country. It is not an easy area.

***Dr. Dalvi:*** That's true and important in terms of developing guidance as to what practices should be minimum mandatory standards everywhere, and others which are state of the art. I am glad our community is starting the dialogue; we have much work ahead on this.

***Mrs. Poli:*** Thank you so much Dr. Dalvi. How do you recommend we move forward?

***Dr. Dalvi:*** In pursuing advocacy goals and related matters, partnership with respective national societies will continue to be vital and win-win. I have been in touch with the Pediatric Cardiac Society of India which agrees completely. We should link websites, share educational materials, be represented on one another's committees and writing groups, and jointly speak out on advocacy matters of mutual importance. In doing so, we can set the bar for partnerships with other national societies.

When a new society comes into being—and there are many such societies, most from North America and Western Europe—their emphasis, and I'm not being critical—is naturally on those regions. PICS on the other hand is fully global. Dr. Hijazi himself (and the entire PICS leadership) is completely dedicated to global health. Because of that, we now have a platform to bring that same global philosophy to the forefront addressing the challenges we discussed today. Natalie, as you said, we are doing this across all ages, socioeconomic strata, colors, genders and cultures. That should always be our focus and our goal.

***Mrs. Poli:*** I am so grateful to my own care team. If it weren't for doctors like you—and your superb nurses and technologists—I wouldn't be here today! I will never forget that. Thank you!

**BHARAT DALVI, MD, DM, FPICS**

Consultant Cardiologist
Glenmark Cardiac Centre
Sir H N Reliance Foundation Hospital
Mumbai, India

**NATALIE POLI, Ed.S**

*PICS Society Senior Patient Advocate*
Public Education Professional
Successfully treated interventionally for CHD, 2006
Mrs. Midwest International 2022
Former Captain of the NBA Chicago Bulls Dance Team

**KAMEL SHIBBANI, MD**

Advanced Pediatric Cardiology Fellow
University of Iowa
Iowa City, IA, USA

**NORM LINSKY, MPA, MA**

PICS Society Executive Director
Washington, DC, USA
nlinsky@CHDinterventions.org

# A Tribute to Gary

*Erwin Oechslin, MD; Jack Colman, MD; Rafa Alonso, MD*

Gary D. Webb, a jewel in the crown of Canadian cardiology, a giant in the specialty of Adult Congenital Heart Disease that he did so much to establish, passed away October 19, 2021, in Philadelphia.

Gary graduated from McGill, and after internship at the Royal Victoria Hospital, came to Toronto in 1968 for internal medicine and cardiology training, followed by a staff position at The Wellesley Hospital, and then at TGH.

Among his early responsibilities, he was director of post-graduate training in cardiology. He joined Peter McLaughlin and Peter Liu in the Adult Congenital Heart Disease (ACHD) program (founded in 1959), and by 1986 took over as its Director, a position he held till 2004, establishing a strong foundation for what became one of the largest and most influential programs in the world.

Gary either led or played a major role in essentially all the early international conferences and guideline committees that shaped the future of ACHD, including the first CCS Consensus Conference on Management of ACHD, published in 1996, for which he made masterful early use of his namesake, the World Wide Web, to develop Canadian guidelines with substantial international input without a single face-to-face meeting. Among his many accomplishments, he was the founding President of the Canadian Adult Congenital Heart (CACH) Network and of the International Society for Adult Congenital Heart Disease (ISACHD), he edited one of the most important textbooks in ACHD 'Diagnosis and Management of Adult Congenital Heart Disease' now in its 4th edition, he established the ACHD Learning Center, now part of Heart University, and he created and was editor-in-chief of the Congenital Heart International Professionals' (CHiP) Network.

In 2004 he left Toronto for Philadelphia, following his wife Anne Phillips, who took a senior executive position in the pharmaceutical industry. He was director of the ACHD program at Penn until 2009 when he moved on to establish the ACHD program at Cincinnati Children's Hospital Heart Institute. In 2017 he reactivated his Toronto appointment, and closed his clinical career at TGH, again seeing Toronto ACHD patients and teaching Toronto ACHD fellows.

Gary has been one of the pre-eminent pioneers in ACHD, a young subspecialty that owes much of its development to his personal vision and efforts. His national and international reputation reflects the global impact of his leadership. He has been a global authority in ACHD clinical care, in guideline development, in knowledge transmission, and in creating an enduring global network of like-minded individuals working towards the goal of improved understanding of ACHD and improved care for those born with this life-long chronic condition. The future of the global ACHD community is built on his legacy.

The Toronto ACHD group is planning to create a Gary Webb Fund for Education in ACHD. More details will be sent for those who want to contribute once they are available.

*This tribute was originally published by Heart University. CCT thanks Heart University and the authors, Drs. Erwin Oechslin, Jack Colman and Rafa Alonso, for allowing CCT to share their heartfelt tribute with our readers.*

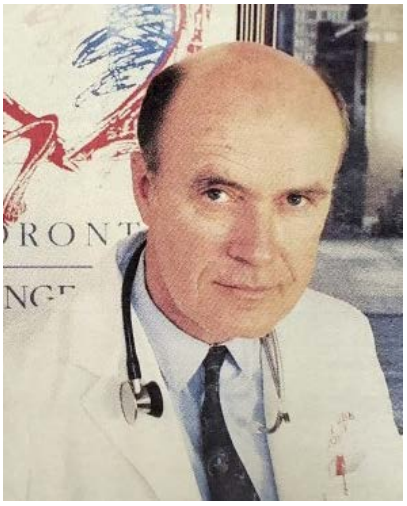
# Obituary for Gary Douglas Webb, MD, FRCPC, FACC

Gary Douglas Webb of Radnor, PA, passed away on October 19, 2021.

Born January 3, 1943 in Montreal, Quebec, he was the son of the late John Douglas Webb and the late Jeannie (Penny) Hardie Penman.

He leaves behind the love of his life and adoring wife of 41 years, Anne Michelle Phillips. He was the much-loved father of Lindsay (Stephen), Geoffrey (deceased July 1, 2017 (Jacqueline), Laura (Adam), Natalie (Quinn), brother to Blair (Linda), Barbara (Steve, deceased 2020) and Ross (deceased 1978). He was also a beloved fur father of Butters, Bueller and Tripp. He passed away at home surrounded by his family after a valiant battle with brain cancer.

Gary graduated from McGill University in Montreal and after an internship at Royal Victoria Hospital he came to Toronto in 1968 for internal medicine and cardiology training. He had staff positions first at Wellesley Hospital and then at Toronto General Hospital. Among his early responsibilities he was director of post grad training in Cardiology. He joined the Adult Congenital Heart Disease Program (ACHD) and took over as Director in 1986, a position he held until 2004, establishing a strong foundation of what became one of the largest and most influential programs in the world.

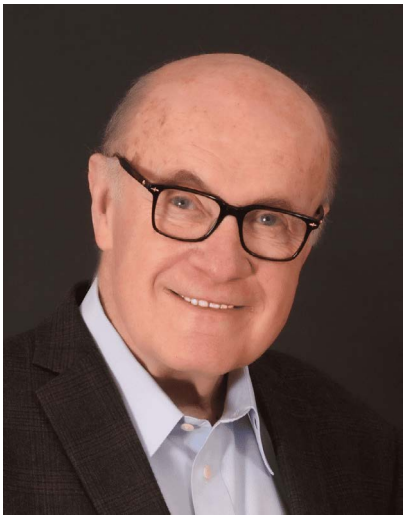


Gary either led or played a major role in essentially all of the early international conferences and guideline committees that shaped the future of ACHD. He also edited one of the most important textbooks in ACHD, now in the 4th edition. He established the ACHD Learning Centre, now part of Heart University, and was Editor-in-Chief of the Congenital Heart International Professions' (CHiP) Network.

In 2004, Gary left Toronto for Philadelphia, following his wife Anne, who took a senior executive position in the pharmaceutical industry. He was the director of ACHD program at University of Pennsylvania until 2009, and then he moved on to establish the ACHD program at Cincinnati Children's Hospital. In 2017, he reactivated his Toronto appointment, and began again seeing Toronto ACHD patients and teaching Toronto ACHD followers.

Gary has been one of the pre-eminent pioneers in ACHD, a young subspecialty that owes much of its development to his personal vision and efforts. His national and international reputation reflect the global impact of his leadership. The future of the global ACHD community is built on his legacy.

While Gary had an incredibly impressive career, his greatest source of pride and sense of achievement came from his family, both immediate and extended. His world was his family, and he was the bedrock of it.



His two constant mantras to his family were "Work Hard, Play Hard" and "Be Good to Yourself, Be Good to Others."

His focus was on time spent together, memories of wonderful family trips and extended family holidays on Shadow Lake and Gibson Lake in Ontario. He and his family returned to Toronto every Christmas to see both sides of his family, whom he loved more than anything. He was thrilled to just hang out, watch sports and movies, eat great soups, enjoy fabulous food and laugh. He also loved to watch his kids 'and nephews' sporting events (hockey, rowing and lacrosse). He was an amazing hockey player growing up, and he loved to see how the kids and their teams progressed year over year.

Gary loved fun, and all the kids loved him. He was like the Pied Piper – kids followed him blindly on whatever adventure he proposed – at the drop of a hat he'd take them go-karting, tubing on the lake and white-water rafting, "driving" the car (on his lap), rides on top of his shoulders, dizzying flips in his arms, all accentuated by his wild exclamations of "Hola-Bola" which he used as frequently as he possibly could.

## CHiP NETWORK

### CONGENITAL HEART INTERNATIONAL PROFESSIONALS

Gary was a devoted husband, father and the quintessential family man. It brought him great joy to walk his daughter Laura down the aisle on April 10th, 2021 at her wedding to Adam. He loved family parties and the communal Sunday night family dinner was a tradition that continued until he passed.

Gary's family was the pride and joy of his life. He loved to laugh and had a great sense of humor. Anne and Gary loved and laughed together throughout their wonderful life together.

Congenital Cardiology Today (CCT) and the CHiP Network have been long time partners. They began working together in 2013 when Tony Carlson, Founder and Senior Editor of CCT, helped Gary Webb, Founder and Editor-in-Chief of the CHiP Network, build his initial subscriber base. The CHiP Network grew along with their personal and professional relationship with CCT.

"I remember Gary talking to me about his vision of creating a Congenital Heart Professionals Communications Network in mid 2013. His vision for The CHiP Network was officially launched in 2014 with the support of 28 congenital heart and pediatric cardiac institutions and organizations from around the world. Gary helped shape the future of ACHD worldwide. He will be dearly missed." *—Tony Carlson, Founder & Senior Editor of Congenital Cardiology Today*



# Dr. Richard Kovacs Named ACC Chief Medical Adviser/Chief Medical Officer

*In Inaugural Position, Kovacs Will Provide Clinical Council Across Staff and Leadership*

Richard "Dick" Kovacs, MD, MACC, was named Chief Medical Adviser/Chief Medical Officer of the American College of Cardiology—a role in which he will work with physician leadership and staff to provide clinical advice in fulfilling the College's mission of transforming cardiovascular care and improving heart health. Kovacs is a past president of the College and has been an ACC member for over 30 years.

"Dick has been an exemplary leader in the College throughout his tenure as an ACC member, leading not only at the highest roles, but also in various committees and task forces ranging from finance to science and quality," said ACC CEO Cathleen C. Gates. "His history of leadership in medicine and his breadth of experience throughout the different areas of the College make him a perfect fit to serve as CMA/CMO and bring his clinical expertise and knowledge to the ACC staff."

In his role as CMA/CMO, Kovacs will counsel ACC staff on emerging clinical issues and help assess complex medical and scientific data. As a member of the senior executive staff, he will also work closely with partner cardiovascular societies, medical specialty organizations and external collaborators, including regulatory and governmental agencies, payers, and credentialing and licensing organizations.

"I've devoted a large portion of my professional career to volunteer member service to the ACC to help advance the mission and propel the field of cardiology forward," said Kovacs. "I'm honored to expand my involvement as ACC Chief Medical Adviser and Chief Medical Officer and more closely work with the College staff and physician leaders to transform cardiovascular care on a global scale."

Kovacs is the Q.E. and Sally Russell Professor of Cardiology at Indiana University (IU) School of Medicine and served as the cardiology service line leader of IU Health Physicians. He also served as the clinical director of the Krannert Institute of Cardiology.

Kovacs received his medical degree from the University of Cincinnati School of Medicine and completed an internship and residency at IU Medical Center. His fellowship training was also at IU, where he served as chief fellow and chief medical resident. He joined the IU School of Medicine in 1986 as an assistant professor, subsequently serving as the medical director and CEO of Methodist Research Institute. He then worked as a senior clinical research physician at the Lilly Research Laboratories of Eli Lilly and Company.

Kovacs returned to the IU School of Medicine faculty in 2003 and has since served as the Associate Dean for Clinical Research and associate director of the Indiana Clinical and Translational Sciences Institute.

Kovacs began his role as CMA/CMO on November 1, 2021.

# ACC, Wondr Medical to Create New Digital Educational Channel Enabling Interactive Learning Opportunities for Global Cardiovascular Community

*Channel Will Provide Access to World-Class Content, Support for Under-Supported Audiences*

The American College of Cardiology (ACC) and Wondr Medical are collaborating to create a new digital channel on the Wondr Medical Platform that will amplify the global reach of ACC's educational content on a dynamic new platform. The partnership also provides access to ACC's world-class content to the existing clinical users of Wondr Medical.

"As part of the ACC's vision of a world where innovation and knowledge optimize cardiovascular care and outcomes, our partnership with Wondr is an exciting opportunity to increase access to ACC's phenomenal educational content outside of those who have traditionally accessed it," said Neal Kovach, MBA, Vice President, Global Innovation and Clinical Transformation. "By increasing the ACC's global reach we can have a more active role in providing education and support for health care communities around the world caring for underserved populations and helping to reduce the global cardiovascular disease burden. Together with Wondr, we hope to create an interactive educational community where we can share our knowledge and better serve each other and our patients."

The ACC and Wondr Medical will strive to create a digital channel that pairs ACC's expertise and knowledge in creating high-quality cardiovascular care education with Wondr's mission to democratize health care content globally through connection and social networking. ACC educational resources, such as NCD Academy, will now be available as part of Wondr's content for medical professionals around the world, free-to-air and streaming on any device. In the coming months, the ACC and Wondr partnership will focus on creating and accelerating educational access and content for underserved audiences such as women in cardiology and more.

"We started Wondr with a vision to deliver the finest medical education to medical professionals wherever they were. This partnership with ACC is the clearest possible example of that vision becoming a reality. Now our users in 175 different countries can access the ACC's resources, starting with on-demand clinical education, on the platform. This bold partnership brings new voices into the ACC family and will accelerate improvements in the delivery of cardiovascular care across the world," said Justin Davies, MD, Founder, Wondr Medical.

Available today, ACC's digital channel on Wondr features free, on-demand courses for cardiovascular professionals and primary care clinicians to explore a wide variety of expert analysis of practice-changing research, clinical guidelines and topics relevant to everyday practice while also earning Continuing Medical Education (CME) credit. Access the channel: **https://wondrmedical.net/ch/acc**.



## FEBRUARY

### 17-19

**CATCH: Caring for Adults and Teens with Congenital Heart Disease**
Oahu, Hawaii, USA
https://www.hawaiipacifichealth.org/CATCH

### 26-01

**CRT22**
Washington, D.C., USA
https://crtmeeting.org/Default.aspx?mkt_tok=MjcxLVJPVS0xMjQAAAGARtmr7GGCAKCWH-KPGINB9OBz_gWE70ZTgkzti5b8yjBY_CBNiT3RArF_35KpicybtR-8YhHFysgwOThFq5ffpeaYr01wsngBVdWx

## MARCH

### 15-17

**ALICE 2022 – Advanced Live Interventional Course of Essen**
Essen, Germany
https://alice-the-course.info/

### 26-27

**CSI Focus LAA & PFO**
Tokyo, Japan
https://www.csi-congress.org/laa-pfo

## APRIL

### 02-04

**ACC22**
Washington, D.C., USA
https://accscientificsession.acc.org/

### 29-01

**Heart Rhythm 2022: Bringing the World of EP Together**
San Francisco, California, USA
https://heartrhythm.com/

CONGENITAL CARDIOLOGY TODAY

CORPORATE OFFICE
PO Box 52316
Sarasota, FL 34232 USA

# CORPORATE TEAM



# EDITORIAL BOARD



# OFFICIAL NEWS & INFORMATION PARTNER OF

PICS Society
Pediatric *and Congenital* Interventional Cardiovascular Society




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