CONGENITAL CARDIOLOGY TODAY
Timely News & Information for Congenital/Structural Cardiologists & Cardiothoracic Surgeons Worldwide
International Edition
Vol. 20 - Issue 8
August 2022

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Congenital Corrected Transposition of the Great Arteries

P. Syamasundar Rao, MD

Introduction

Over the years, I have contributed numerous papers to Neonatology Today and Congenital Cardiology Today in an attempt to educate the neonatologists, cardiologists, trainees in pediatric cardiology, neonatology and pediatric intensive care, and paramedical personnel in these subdisciplines. Initially, generalized topics in the care of neonates with Congenital Heart Disease were addressed.[1-7] This was followed by description of neonatal transcatheter interventions.[8-13] Subsequently, individual cardiac defects were addressed.[14-21] In this review, congenital corrected transposition of the great arteries will be addressed.

Congenital Corrected Transposition of the Great Arteries

Congenital Corrected Transposition of the Great Arteries (CCTGA) is an infrequent multifaceted cardiac defect. CCTGA was first documented by von Rokitansky in 1875.[22] Although only a few cases had been reported through the 1950's, some large case series have subsequently been published.[23-27] These papers defined the pathologic, clinical, and hemodynamic features of this heart defect. The prevalence of CCTGA is in the order of 0.5% of all Congenital Heart Defects (CHD).[28-29]

In the most usual type, the position of the atria is normal (atrial situs solitus), atrio-ventricular discordance exists, the Morphologic Left Ventricle (MLV) is located on the right side while the Morphologic Right Ventricle (MRV) is placed on the left side (ventricular inversion), and ventriculo-arterial discordance is present. Principally, the ventricles are inverted. The systemic venous blood from the right atrium is passed onto a right-sided MLV and then into the pulmonary artery, while the pulmonary venous blood from the left atrium is passed onto a left-sided MRV and then into the aorta. Consequently, the blood circulation is normal.[30-32] Since the aorta comes off the right ventricle and the pulmonary artery comes off the left ventricle, Transposition of Great Arteries is considered to exist. Yet, a normal blood flow pattern is kept intact, henceforth the term CCTGA, or corrected transposition for short. Usually, the aorta is situated to the right of the pulmonary artery; but, in this cardiac defect, the aorta is positioned to the left of the pulmonary artery; hence the term l-Transposition of the Great Arteries (l-TGA).

Associated Heart Defects

While patients without any other heart defects have been described,[33,34] the majority of CCTGA patients have significant heart defects, namely: Ventricular septal defect (VSD), pulmonary stenosis (PS), atrioventricular (AV) block and/or an Ebstein's-like abnormality of the left-sided, morphologic tricuspid valve.[30,31,35] The size of the VSD may be small, medium, or large. Obstruction of the pulmonary outflow tract, i.e., of the right sided MLV may be seen in 44 to 57% of cases.[31] The more usual causes of such narrowing are atresia or stenosis of the pulmonary valve, subvalvar membrane, and subpulmonary stenosis due to muscular malalignment and/or hypertrophy. Less frequent causes are fibrous tags or accessory valve tissue, aneurysm of the membranous septum, and intra-cardiac blood cysts.[31,35] If both a VSD and PS or pulmonary atresia coexist, right-to-left shunt occurs resulting in cyanosis.

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Complete AV block may develop spontaneously or may occur following catheter or surgical intervention. Ebstein's-like malformation of the left-sided, morphologic tricuspid valve may be mild and may not be recognized initially or may be severe resulting in presentation in early life.[35]

Clinical Features

Patients without associated heart defects are not usually detected until adulthood[34] unless an imaging study is performed for an unrelated reason. Patients with only a VSD exhibit a holosystolic murmur at the left lower sternal border. Patients with isolated PS (valvar or subvalvar) typically have an ejection systolic murmur at the left-upper or left-mid sternal borders. Children with both VSD and PS may present with symptoms shortly after birth, typically during the first month of life. Cyanosis, tachypnea and tachycardia are the common findings; these are similar to those seen in patients with Tetralogy of Fallot (TOF). An increased right ventricular impulse, a loud single second heart sound, and an ejection systolic murmur along the left upper sternal border may be detected on physical examination. Children with significant left AV valve insufficiency have a holosystolic murmur, heard best at the apex.[35] A slow heart rate may be the presenting finding in children with AV block.

Chest X-ray

Straightening of the left upper border of the heart caused by the ascending aorta of l-TGA may be visualized, unless obscured by the thymic shadow. Moderate to severe cardiomegaly may be perceived in subjects with moderate to large VSDs and those with moderate to severe left AV valve insufficiency. Babies with VSD and PS may have a normal-sized heart or mild cardiomegaly and decreased pulmonary vascular markings.

Electrocardiogram

Electrocardiogram usually demonstrates distinctive findings that include the absence of Q waves in the left chest leads (V5 and v6) and the presence of Q waves in V1; these indicate the reversal of septal depolarization **(Figure 1)**. Findings indicative of atrio-ventricular conduction block (first-, second- or third-degree heart block) may be seen in some patients.

Echocardiogram

Echocardiogram is greatly helpful in the diagnosis; however, it is challenging for the beginner both to perform the study and to interpret it. Lack of continuity between the left AV valve and the aortic valve secondary to the presence of a conus on the left side (MRV) is an important echocardiographic feature. Examination of the AV valve attachment may be helpful in diagnosing ventricular inversion. In patients with normal ventricular relationship, the attachment of the tricuspid valve to the septum is at a higher level than that of the mitral valve **(Figure 2A)**. By contrast, in CCTGA, the attachment of the mitral valve is at a higher level on the right side when compared to the tricuspid valve attachment on the left side **(Figure 2B).**

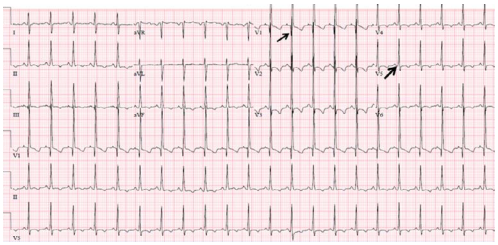

FIGURE 1
An electrocardiogram of a patient with corrected transposition showing Q waves in the right chest leads and no Q waves in the left chest leads. This is indicative of ventricular inversion, though such a pattern may also be observed in patients with severe hypertrophy of the right ventricle. The P waves are normal, suggestive of situs solitus. Reproduced from reference 36.

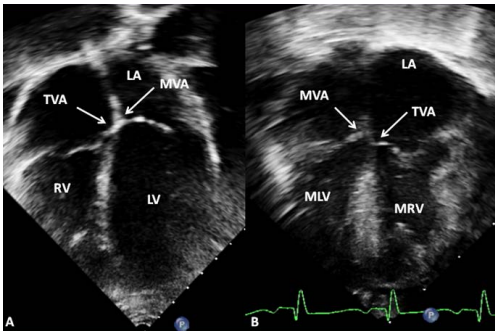

FIGURE 2
*Echocardiographic video frames from apical four-chamber views of two different patients. **A)** Normal interventricular relationship. A higher level of attachment of the mitral valve (MVA) on the left is seen compared to the tricuspid valve attachment (TVA) on the right. **B)** Valve attachments in a patient with corrected transposition (ventricular inversion). A higher level of attachment of the mitral valve (MVA) on the right when compared with the tricuspid valve attachment (TVA) on the left were demonstrated. LA, left atrium; LV, left ventricle; MLV, morphologic left ventricle; MRV, morphologic right ventricle; RV, right ventricle. Reproduced from reference 36.*

The size and location of the VSD and shunting across it can be demonstrated on echo. Enlargement of the left atrium (LA) and MRV can also be revealed by m-mode and 2D imaging and the degree of their enlargement is proportional to the magnitude of the shunt. Also, the type of pulmonary outflow tract obstruction (atresia or stenosis of the pulmonary valve, subvalvar membrane, subpulmonary stenosis due to muscular malalignment and/or hypertrophy, fibrous tags or accessory valve tissue, aneurysm of the membranous septum, and intra-cardiac blood cysts) may be shown on echo and the degree of PS may be quantitated by Doppler interrogation.

In patients with Ebstein's-like abnormality of the left-sided, morphologic tricuspid valve, the extent of downward dislocation tricuspid valve leaflets may be appraised in a four-chamber view **(Figure 3A)**. The extent of left AV valve insufficiency may also be shown on color Doppler study **(Figure 3B)**. Enlargement of the LA and MRV can also be illustrated by m-mode and 2D imaging and the degree of their enlargement is proportional to the magnitude left AV valve insufficiency.

Magnetic Resonance Imaging (MRI) and Computed Tomography (CT)

MRI and CT studies are not required for making a diagnosis or for evaluation of pathophysiology, particularly in infants and children. But, in patients with poor acoustic windows, particularly adolescents and adults, MRI and CT evaluation may be useful in confirming the diagnosis and in evaluating the severity of the defect.

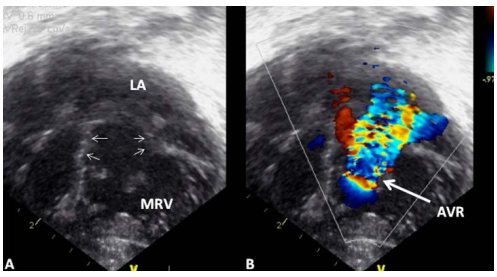

FIGURE 3
***A)** Modified apical view illustrating the downward displacement of the attachments of a left-sided morphologic tricuspid valve (small arrows) in a patient with l-TGA. **B)** Color flow Doppler image of the patient shown in A, demonstrating severe atrioventricular valve regurgitation (AVR). LA, left atrium; MRV, morphologic right ventricle. Reproduced from reference 36.*

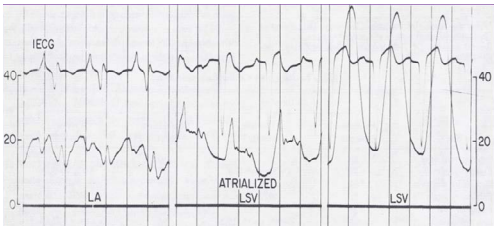

FIGURE 4
Intra-cardiac electrocardiogram (IECG) and pressures were recorded simultaneously as the platinum-tipped electrode catheter is gradually pulled from the left atrium (LA) to the left sided systemic ventricle (LSV). This ventricle is morphologic right ventricle. Left-most panel displays atrial pressure curve with atrial electrogram. The central panel demonstrates atrial pressure curve with ventricular electrogram when the tip of the recording catheter is in the atrialized section of the LSV. Right-most panel illustrates ventricular pressure curve along with ventricular electrogram when the tip of the catheter is in the LSV. Pressures are marked in mmHg. The pressure in LSV is damped due to small lumen of the catheter. Reproduced from reference 35.

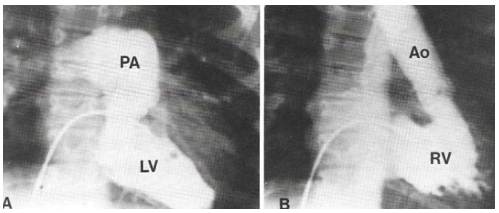

FIGURE 5
*Cine-angiographic frames in an infant with congenital corrected transposition of the great arteries. **A)** Right-sided, smooth-walled morphologic left ventricle (LV) which gives rise to the pulmonary artery (PA). The pulmonary valve is located inferior to aortic valve and rightward of aortic valve and posterior (in the lateral view – not shown) from the normal position. **B)** Coarsely trabeculated morphologic right ventricle (RV) on the left side, which gives origin to the aorta (Ao). The aortic valve is located superior to and leftward of pulmonary valve and anterior (in the lateral view – not shown) from the normal position. Angiogram B was secured via a catheter advanced from the right atrium to the left atrium via a patent foramen ovale and then into morphologic RV. Reproduced from reference 30.*

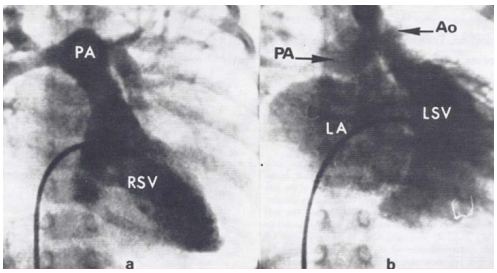

FIGURE 6
***A)** Right-sided ventricular (RSV) cine-angiogram, illustrating a smooth-walled morphologic left ventricle on the right side which gives rise to the pulmonary artery (PA). **B)** Cine-angiographic frame of the left-sided ventricle (LSV), showing a coarsely trabeculated morphologic right ventricle which gives origin to the aorta (Ao). Significant left-sided atrioventricular valve insufficiency with resultant opacification of the left atrium (LA) is shown. The PA is also opacified secondary to a left-to-right shunt via a ventricular septal defect (not marked). Reproduced from reference 35.*

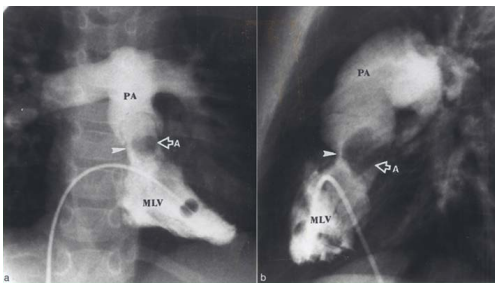

FIGURE 7
***A)** and **B)** Morphologic left ventricle (MLV) in postero-anterior (A) and lateral (B) views, illustrating a finely trabeculated ventricle which gives rise to the pulmonary artery (PA). The pulmonary valve is located posterior to (B) and inferior to and rightwards from (A) the normal position. The open arrows in (A and B) point to a radiolucent aneurysm in the sub-pulmonary area. The solid arrows in (A and B) point to a thin jet of contrast material ejected from the MLV to the PA, indicating severe sub-pulmonary narrowing. Reproduced from reference 31.*

Cardiac Catheterization and Selective Cineangiography

Catheterization and angiography are not generally needed; however, on rare occasions, when the diagnostic details can't be clearly established by non-invasive techniques, catheterization studies may be useful.

The diagnosis of Ebstein's anomaly of the tricuspid valve (in patients with normally positioned ventricles) by concurrent recording of pressures and intracavitary electrocardiograms across the tricuspid valve is a well-established method;[30] these were routinely used prior to the advent of echocardiography. Such a technique to establish diagnosis of Ebstein's like malformation of the left AV valve in subjects with CCTGA has also been employed.[35] Simultaneous recording of pressures and intracavitary electrograms across the left atrioventricular valve in an infant with angiographically proved CCTGA and left AV valve regurgitation is demonstrated in **Figure 4**.[35]

Oxygen saturation and pressure data reflect hemodynamic abnormalities related to the associated cardiac defects, but will not be reviewed since catheterization is not routinely performed at the present time. However, some angiographic examples will be presented in **Figures 5-8**. Smooth-walled MLV on the right side **(Figures 5A, 6A, 7)**[30,31,35] and trabeculated MRV on the left side **(Figures 5B, 6B, 8)**[30,31,35] can clearly be seen. The severity of left AV valve regurgitation may be semi-quantitated by the degree left atrial opacification **(Figure 6B)**. Angiographic ventricular morphology may also be defined in patients with dextrocardia **(Figure 9)**.[37,38]

Therapy

The need for treatment is largely dependent on the presence of associated cardiac defects and their hemodynamic significance and is reviewed elsewhere.[30,35,36,39] Patients with only a VSD (moderate to large) may not develop congestive heart failure (CHF) during the neonatal period; this is due to high neonatal pulmonary vascular resistance. Signs of CHF may manifest in a few weeks to months after birth. At that time anti-congestive medical management, including diuretics, afterload reducing agents and digoxin may be administered as deemed appropriate. In the absence of adequate control of CHF, surgical closure of the VSD becomes necessary. But, there is a risk for the developing complete heart block because of a reversed conduction system. In patients with CCTGA, the AV conduction bundle traverses along the anterior and superior rims of the VSD and courses along the right septal side of the MLV. This is well-known to most surgeons and appropriate precautions should be taken to prevent heart block while performing a patch closure of the VSD. If the location of the conduction bundles cannot be clearly defined, intra-cardiac mapping to identify the conduction system[40,41] should be pursued.

In patients with both a VSD and MLV outflow tract obstruction, the management is similar to that of TOF patients, with prostaglandin E1 (PGE1) infusion in neonates with ductal

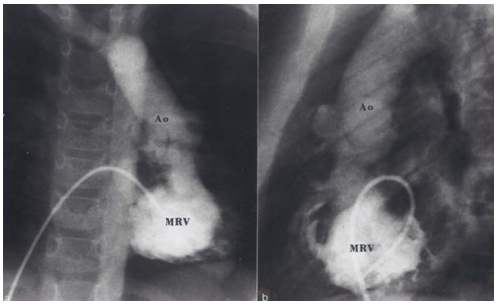

FIGURE 8
A) *and* ***B)*** *Morphologic right ventricular (MRV) angiogram in postero-anterior (A) and lateral (B) views, illustrating a coarsely trabeculated ventricle which gives rise to the aorta (Ao). The aortic valve is located anterior to (B), superior to (A and B) and leftwards (A) from the normal position. Reproduced from reference 31.*

dependent pulmonary blood flow, a modified BT shunt[42] in babies whose anatomy is unsuitable for complete surgical correction during the neonatal period, and total surgical correction between three and twelve months of life. The considerations for prevention of heart block are like those outlined in the preceding paragraph. If the MLV outflow tract obstruction is secondary to subvalvar membrane, fibrous tags or accessory valve tissue, aneurysm of the membranous septum, and intra-cardiac blood cysts, they should be resected along with closure of VSD.

In children with moderate to severe insufficiency of the morphologic tricuspid (secondary to an Ebstein's-like valve), afterload-reducing drugs such as Captopril or Enalapril and other anti-congestive measures may be employed first to treat CHF. If medical management is not able to control the CHF, surgery to repair or replace the morphologic tricuspid valve may be needed.[30,35,36]

Complete heart block may spontaneously develop, or it may be precipitated by catheter or surgical procedures; in such patients, pacemaker implantation may be needed.[43] Epicardial pacemakers in neonates and infants and transvenous pacemakers in children and adults may be used.

Because of relatively poor long-term outcome, predominantly related to allowing the MRV to sustain the systemic circulation, especially when associated with significant morphologic tricuspid valve regurgitation, a double-switch operation was developed.[44] In this procedure, an arterial switch procedure similar to that

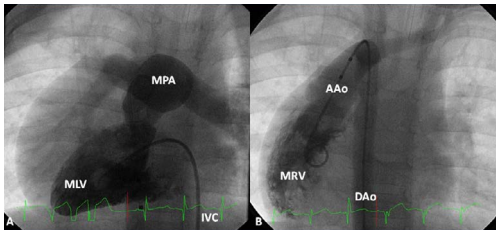

FIGURE 9
A) *Morphologic left ventricular (MLV) angiogram in a postero-anterior projection in a patient with dextrocardia, displaying a finely trabeculated left-sided MLV which gives origin to a dilated main pulmonary artery (MPA). The catheter in the inferior vena cava (IVC) is on the left-side of the spine.* ***B)*** *Morphologic right ventricular (MLV) angiogram, also a postero-anterior projection in the same dextrocardia patient shown in A, illustrating a coarsely trabeculated right-sided MLV which gives origin to the aorta (AAo). Note that the descending aorta (DAo) is on the right of the spine. The aortic valve is located superior to and rightwards from the pulmonary valve. It is also anterior in the lateral view (not shown). These data would suggest a d-loop of the ventricles and d-transposition of the great arteries in a patient with dextrocardia, a condition indicative of corrected transposition type of physiology. Reproduced from reference 37.*

used for Transposition of the Great Arteries[45] is performed; this would connect the MLV to the aorta and MRV to the pulmonary artery. Then, a venous switch procedure such as Mustard[46] or Senning[47] to counter the arterial switch[45] is undertaken. Initial results reported by Imai were good with a mortality of 9%. Subsequent reports[48,49] were also encouraging. Relative advantages of simpler corrective procedures vs. the double switch operation should be balanced while embarking on double switch procedure.

Summary and Conclusion

CCTGA is a rare, complex CHD and constitutes less than 0.5% of all CHDs. In this anomaly, there is: atrial situs solitus, atrio-ventricular discordance, left to right reversal of the ventricles and ventriculo-arterial discordance. Given both atrio-ventricular and ventriculo-arterial discordance, normal blood circulatory arrangement is preserved. While occasional patients may not have other cardiac defects, the majority of patients have associated defects. CCTGA and associated defects can easily be diagnosed by echo-Doppler studies with rare need for other imaging studies. Therapeutic interventions are largely based on the type of associated cardiac defects and the degree of hemodynamic abnormality that they produce and were detailed in the above review. Consideration for double-switch operation is appropriate for patients with significant morphologic tricuspid valve insufficiency. It was concluded that CCTGA can be effectively diagnosed with the currently available non-invasive and invasive investigative techniques and the defect can successfully be managed with the existing therapeutic medical and surgical techniques.

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Innovation in Congenital/Pediatric Interventional Cardiology: Exciting Developments Ahead

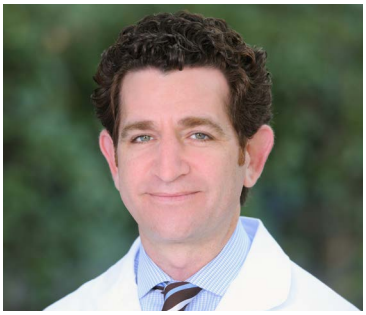
Kamel Shibbani, MD

As the pace of innovation in our field accelerates almost daily, we recently interviewed several of your colleagues involved with exciting up-and-coming devices, procedures, and imaging modalities on the horizon. What innovations are coming soon? What obstacles face us? How can The PICS Society encourage innovation, lessen impediments, and pursue best possible outcomes for patients? (Interested in being interviewed on this important topic? Email us at membership@CHDinterventions.org)

To understand all that and more, we recently were privileged to speak individually with Drs. Athar Qureshi and Darren Berman. Dr. Qureshi is a Professor of Pediatrics at Baylor College of Medicine and also serves as Medical Director of the CE Mullins Cardiac Catheterization Laboratory, Texas Children's Hospital. Dr. Berman recently moved to Los Angeles and joined the Heart Institute at Children's Hospital Los Angeles as the Director of the Cardiac Catheterization Laboratory. *(Note that references to specific devices should not be construed as The PICS Society or interviewee endorsements).*

KS: These discussions are especially timely, given that The PICS Symposium will celebrate its 25th Anniversary this September in Chicago, where looking into the future of our profession will be a major focus. Can you tell us about what advancements await us in the coming months and years?

Dr. Kamel Shibbani



Dr. Darren Berman

Dr. Athar Qureshi

"Our field has exploded with innovation in many areas where we can expect much growth in the near future."

– Athar Qureshi, MD, FPICS

AQ: I think this an exciting time to be a pediatric interventional cardiologist. Our field has exploded with innovation in many areas where we can expect much growth in the near future. Valve technology, for example, is one of the hottest areas in our field, with native outflow valves having evolved a great deal in the last 5-10 years. Despite this growth, there are still patients with a native RVOT that do not qualify for such valves. I think that in the short term, we can expect to see innovation in valve design to help many more patients. Along with that, I think we can expect to see development in imaging technology to allow us to customize valves to fit individual patient anatomy (or groups). In fact, we have seen some hints of that already, with custom-made stents being used in innovative new procedures such as sinus venosus ASD closure.

DB: When I think of innovation in our field, one category that comes to mind is bioabsorbable technology. One representative example of advancement of this is the reSept™ ASD Occluder by atHeart Medical. Current ASD devices do an excellent job at occluding atrial septal defects, but if we can do this with a device that eventually resorbs and leaves less of a footprint, this may turn out to be best for our patients, especially when we think of our growing adult congenital population that might need future left heart interventions. We are hopeful that we will see promising results from the ASCENT Trial in the coming years, https://atheartmedical.com/wp-content/uploads/2022/02/atHeart-Press-Release-FEB2022-FINAL.pdf.

Beyond this device, I think we are just scratching the surface with bioabsorbable technology, and I think it will play an important role moving forward. A big change over the past few years has been a better understanding of which metals might be more suitable for such devices. While refinement of this technology might take some time, I think we will see bioabsorbable devices playing a big role in our field in the future.

AQ: One area also experiencing a wave of innovation is heart failure therapy within congenital heart disease. The devices we typically use for heart failure in our pediatric population are ones that are approved for adults. Not only are we having to innovate in the way we use them in children, but we are also working with industry to miniaturize this equipment. There is also growing interest in developing stents that can be implanted in children and dilated to adult size.

DB: Having a stent that can be implanted in an infant and then further dilated to an adult size is definitely an unmet need in our patient population. Stent implantation to treat a stenotic vessel is a well-accepted therapy, yet our field has been limited in our ability to help infants and neonates because we have not focused on creating a stent with adult-size capabilities that can be delivered through a small sheath. The Renata Minima stent and delivery system, in my view, is a potential game-changing

technology and is something that we will hopefully see in the near future.

KS: How can the PICS Society help foster innovation?
AQ: I think The PICS Society has been great at advocating on our behalf and liaising between physicians and industry. Physicians that want to innovate have a particular skillset that might differ from the skillset required in industry and from that required for the regulatory environment. The role I see the PICS Society having is bringing everyone together, so the devices needed by our patients are not only available, but also approved.
DB: I think The PICS Society has a number of attributes that allow it to foster innovation. Two in particular come to mind. One, The PICS Society is truly an international society which immediately allows for collaboration and connection beyond any nation's borders. Two, is the fact that The PICS Society has established itself as the home of congential interventional cardiology. Whether your interest is in neonates or adults living with Congenital Heart Disease—or both--, this society provides a true home for us all. This creates a meeting place for us to work together to overcome obstacles facing us.

"The PICS Society has established itself as the home of congenital interventional cardiology. Whether your interest is in neonates or adults living with Congenital Heart Disease, or both, this society provides a true home for us all."

– Darren Berman, MD, FPICS

KS: This serves as a great segue to the next question. In your mind, what are some obstacles to innovation in our field and what can The PICS Society do to help manage them?
AQ: I think the biggest obstacle to innovation in our field is R & D funding. By virtue of the numerically smaller pediatric population, industry often sees a higher potential ROI by focusing on the more numerous adult population. In fact, we sometimes have to prove that a device needed in the pediatric population can be used in adults as well. One way The PICS Society can help is through securing R & D funds outside of industry. There are entities that focus on pediatric and congenital heart disease that understand that while it is not a very large space, it is a VERY important space. The PICS Society can help by connecting physicians with such entities. On a positive note, the FDA has recently become very receptive to different and more efficient/practical approval processes for congenital/pediatric applications of devices.

Another hurdle that must be overcome is translating an idea into a product. Our field is full of innovators, but going from a concept to an end-product is difficult. Here, too, I see a role for The PICS Society. I think The PICS Society can help overcome that through mentorship programs. By creating mentorship and research programs, The PICS Society can provide young interventionalists with tools needed to see their idea through completion. Innovation today must be followed up with longitudinal studies to assess the impact of new devices. This is a space where The PICS Society can help through creating and maintaining registries.
DB: Appropriately so, I think the regulatory hurdle is one that we often need to overcome when trying to innovate. However, I think this is a hurdle we are successfully navigating as we now see that the FDA wants to partner with us and help us achieve our innovations and goals in an appropriate and safe way. In the past, this felt like it was a hurdle that was difficult to overcome, but the work of The PICS Society has allowed for improved partnership with the FDA.

Importantly, this cooperation goes beyond our borders. Our regulators have been able to work with regulators from other countries such as Japan, to set standards appropriate for different countries and help the flow of ideas beyond our borders.

KS: Thank you so much for your time and your valuable insight, it's been wonderful looking at the future with you. We will check back with you next year!

Tribute to James S. Tweddell, MD

David M. Overman, President-Elect, Congenital Heart Surgeons' Society (CHSS)

It is with tremendous sadness that the CHSS observes the passing of its President, Dr. James Scott Tweddell after a heroic battle with brain cancer. Jim was a modern-day giant in congenital heart surgery. His skills, intellect, and innovations were inspirational to surgeons around the world, and his passing is an enormous loss to our community.

A graduate of Miami University, Oxford, OH and the University of Cincinnati College of Medicine, he completed his surgical training at NYU and Washington University, Jim joined Dr. Bert Litwin at the Children's Hospital of Wisconsin in 1993. Over the subsequent twenty years, Jim established himself as an elite surgeon with unmatched technical prowess, exemplary outcomes, and impactful innovation. He rose to become Professor and Chief for the Division of Cardiothoracic Surgery and S. Bert Litwin Chair of Cardiothoracic Surgery while in Milwaukee. He published extensively, sharing these insights with the worldwide community and was highly respected for his intellectual honesty. He moved back to his hometown in 2015, assuming the role of Executive Co-Director and Director of Cardiothoracic Surgery at Cincinnati Children's Heart Institute and Professor, University of Cincinnati College of Medicine. In the workplace, he was patient-centric and was devoted to the education and training of the next generation. He held many leadership positions during his career, including tenures as Secretary-Treasurer of the CHSS and its 10th President, Director, Society of Thoracic Surgeons, Director, American Board of Thoracic Surgery, and Associate Editor of the Journal of Thoracic and Cardiovascular. Jim was given the CHSS Lifetime Achievement award for his enormous contributions to our specialty on May 27, 2022.

Those who knew Jim well will remember him for his self-effacing humor, his ability to lead with grace and equanimity, his genuine humility, and his love of the banjo. A widely loved and respected colleague, his contributions will live on for years to come. He was a leader by example, a mentor, a role model, and a human being extraordinaire.

James S. Tweddell, MD
September 2,1959 – July 1, 2022

On behalf of the Executive Council of the CHSS and the staff of PRRI, we extend our sincere condolences to Jim's wife, Susie and their three daughters Sarah, Alison, and Caroline. In lieu of flowers, in consultation with the Tweddell family, the CHSS will establish a Travelling Fellowship in Congenital Heart Surgery in Jim's name. More details will follow about donations to this important legacy for Jim and his family.

This tribute was originally published on the Congenital Heart Surgeons' Society's website, https://chss.org/James-Tweddell.cgi. It is republished with permission.

BiVACOR Has Developed a Total Artificial Heart (TAH)

Heart failure is usually treated with a combination of medications. When medications fail, there are not enough donors to keep pace with potential recipients. People die waiting for a heart.

Houston-based BiVACOR has developed a Total Artificial Heart (TAH) designed to be a long-term device that can replace the total function of the patient's native heart. The small, compact device uses rotary blood-pump technology to provide the required cardiac output.

The BiVACOR system comprises a magnetically levitated rotor located between opposing pump casings. The key feature that enables this device to support both the left and right sides of the heart is the left and right impeller blades, which are mounted on either side of the rotating hub. The hub is levitated and rotated via an electromagnetic motor and bearing arrangement on top of the pump casings. The dedicated hydraulic design of the impellers, combined with state-of-the-art magnetic levitation (MAGLEV) technology, permits control of the circulation to be fine-tuned by means of a differential fluid output.

An external controller and batteries provide power to the internal device via a percutaneous driveline.

- Powerful: The centrifugal pumps can provide high flows over 12lpm for dynamic activity.
- Smart: Smart controllers adapt the pump operation to changes in the patient's activity.
- Durable: The anticipated device life is up to 10 years or more.
- Small: The device is small enough for a child, powerful enough for an adult.
- Portable: A small external controller and batteries to give patients freedom.

American Society of Echocardiography Welcomes Nine New Board Members

The American Society of Echocardiography (ASE) is pleased to announce that its membership has elected nine new board members to serve the Society starting July 1, 2022.

The 2022 ASE Executive Committee welcomes newly elected Vice President (one-year term) Theodore Abraham, MD, FASE, University of California at San Francisco, San Francisco, CA, and Secretary (two-year term) Kelly Thorson, DHSc, MSRS, ACS, RDCS, FASE, Lucile Packard Children's Hospital Stanford, Palo Alto, CA.

In addition to the new officers, the following new Board of Directors members were elected to serve two-year terms: Jose Banchs, MD, FASE, FACC, University of Colorado Anschutz Medical Campus, Aurora, CO (Member at Large); Akhil Narang, MD, FASE, Northwestern Medicine, Chicago, IL (Member at Large); Fadi Shamoun, MD, FASE, Mayo Clinic Arizona, Scottsdale, AZ (Council on Circulation & Vascular Ultrasound Steering Committee Chair); Neha Ringwala Soni-Patel, MEd, RCCS, RDCS (AE/PE), FASE, Cleveland Clinic Children's, Cleveland, OH (Member at Large); and G. Monet Strachan, ACS, RDCS, FASE, UCSD Medical Center, San Diego, CA (Council on Cardiovascular Sonography Steering Committee Chair). Sujatha Buddhe, MD, MS, FASE, Seattle Children's Hospital, University of Washington, Seattle, WA (Leadership Academy Representative) and Arthur Labovitz, MD, FASE, Naples Cardiac & Endovascular Center (Retired), Naples, FL (Council on Critical Care Echocardiography Steering Committee Chair) will each serve a one-year term.

Previously elected members of the 2021-2022 ASE Executive Committee transitioning to a new position on the 2022-2023 Board are: Stephen Little, MD, FASE, Houston Methodist Hospital, Houston, TX (President); Benjamin Eidem, MD, FASE, Mayo Clinic, Rochester, MN (President-Elect); and Raymond Stainback, MD, FASE, Texas Heart Institute, Houston, TX (Immediate Past President). Cynthia Taub, MD, FASE, Dartmouth Hitchcook Medical Center, Lebanon, NH, will continue serving as Treasurer through June 2024, and Keith Collins, MS, RDCS, FASE, Northwestern Medicine, Chicago, IL, will join the 2022-2023 ASE Executive Committee for a two-year term as the Council Representative.

Directors continuing with their final year of service include: Carolyn Altman, MD, FACC, FAHA, FASE, Baylor College of Medicine, Houston, TX (Council on Pediatric & Congenital Heart Disease Steering Committee Chair); Anthony Gallagher MHA, FASE, Baptist Health Lexington, Lexington, KY (Member at Large); Leo Lopez, MD, FASE, Stanford Children's Hospital, Palo Alto, CA (Member at Large); G. Burkhard Mackensen, MD, PhD, FASE, University of Washington, Seattle, WA (Council on Perioperative Echocardiography Steering Committee Chair); Susan Mayer, MD, FASE, Saint Luke's Mid America Heart Institute, Kansas City, MO (Member at Large); Kian-Keong Poh, MA, MMed, FRCP, FAMS, FAsCC, FACC, FASE, National University Heart Centre, Singapore (International Representative); Thomas Ryan, MD, FASE, The Ohio State University, Columbus, OH (Past President Member); and Vandana Sachdev, MD, FASE, National Institute of Health, Bethesda, MD (Member at Large).

ASE thanks the following eight Board members who will complete their service on June 30, 2022: Meryl Cohen, MD, MSEd, FASE, FACC, FAHA, Children's Hospital of Philadelphia, Philadelphia, PA (Council Representative); Judy Hung, MD, FASE, Massachusetts General Hospital, Boston, MA (Immediate Past President); Danita Sanborn, MD, MMSc, FASE, Massachusetts General Hospital, Boston, MA (Member at Large); Vincent Sorrell, MD, FASE, FACP ASIM, FACC, FCMR, University of Kentucky Gill Heart & Vascular Institute, Lexington, KY (Member at Large); Jordan Strom, MD, MSc, FASE, Beth Israel Deaconess, Milton, MA (Leadership Academy Representative); Ritu Thamman, MD, FASE, University of Pittsburgh Medical Center, Pittsburgh, PA (Member at Large); Matt Umland, ACS, RDCS, FASE, Aurora Health Care, Muskego, WI (Secretary); and Bryan Wells, MD, FASE, Emory Healthcare, Atlanta, GA (Council on Circulation & Vascular Ultrasound Steering Committee Chair).

The current 2021-2022 Board of Directors met for the final time at ASE's 33rd Annual Scientific Sessions in Seattle, WA, from June 10-13, 2022.

Delivery Catheter Recalled

Recalled Product

- Product Name: Harmony Delivery Catheter System
- Product Codes and Batch Numbers: See database entry, https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfRES/res.cfm?id=192430
- Distribution Dates: April 7, 2021 to January 26, 2022
- Devices Recalled in the U.S.: 665
- Date Initiated by Firm: March 2, 2022

Device Use

The Harmony Transcatheter Pulmonary Valve (TPV) System is used to treat a leaky native or surgically repaired right ventricular outflow tract (RVOT), which is the part of the heart that carries blood to the lungs. The Harmony TPV System consists of a transcatheter pulmonary valve and a delivery catheter, the Harmony Delivery Catheter, that is used to implant the replacement valve without open heart surgery.

The system is indicated for use in children and adults who have severe pulmonary regurgitation, which is when blood flows backward from the RVOT into the right lower chamber (right ventricle) of the heart and require replacement of the pulmonary valve.

Reason for Recall

Medtronic is recalling the Harmony Delivery Catheter because it is possible that the bond holding the capsule at the end of the delivery catheter may break during a procedure to place the TPV.

A capsule bond break could cause procedure delays while the device is replaced with a new one or it may require the patient to undergo additional surgeries. Additionally, a capsule bond break while in use during a procedure could cause serious harm to the patient. Those risks include preventing blood flow and/or completely blocking (embolization or occlusion), tearing and/or splitting (perforation or dissection), or other types of damage to the patient's blood vessels.

There have been six reported complaints from clinical cases, one injury, and no deaths associated with the use of these devices.

Who May be Affected

- Health care personnel who plan to implant the Harmony TPV into patients with severe pulmonary regurgitation
- People who are candidates for valve replacement using the Harmony TPV system

What to Do

On April 6, 2022, Medtronic issued an Urgent Medical Device Recall notice to implanting physicians and customers recommending immediate suspension of use for Harmony TPV's delivery catheter.
Instructions for customers included:

- Removing all unused products from use and returning them to Medtronic.
- Pausing new clinical cases involving the Harmony TPV System.

Customers were also asked to complete a form that was enclosed with the letter to confirm receipt of the recall and to report the number of unused devices currently on hand.

The letter also specified that the recall is:

- Specific to the Harmony Delivery Catheter only, and not the Harmony TPV.
- The break has only occurred in the delivery catheter during delivery of a TPV, no actions are needed for patients who have already been successfully implanted with a Harmony TPV.

Contact Information

Customers with questions or concerns about this recall should contact Medtronic Customer Service at 800.854.3570.

Additional Resources:

- Medical Device Recall Database entry, https://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfRES/res.cfm?id=192430

How do I report a problem?

Health care professionals and consumers may report adverse reactions or quality problems they experienced using these devices to MedWatch: The FDA Safety Information and Adverse Event Reporting Program using an online form, mail, or fax, https://www.accessdata.fda.gov/scripts/medwatch/index.cfm?action=reporting.home.

SEPTEMBER

05-06

2nd Annual PICS Fellows & Early Career Course
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PICS Symposium
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OCTOBER

01

12th Annual UCLA Fetal Cardiac Imaging and Evaluation Symposium
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Hospital Directory 2022-2023

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Published Mid-August

- Send updates to Kate Baldwin, Kate.f.Baldwin@gmail.com
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- Hospitals that Offer Open Heart Surgery for Children in North America
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ISSN 1554-7787 print. ISSN 1554-0499 electronic.
Published monthly.