CONGENITAL CARDIOLOGY TODAY
Timely News & Information for Congenital/Structural Cardiologists & Cardiothoracic Surgeons Worldwide
International Edition
Vol. 19 - Issue 7
July 2021

Table of Contents

Pulmonary Artery Intervention Following Transcatheter Pulmonary Valve (TPV) Implantation Can be Performed Safely Without Compromising the Previously Implanted TPV Function and Integrity

Sharib Gaffar, MD; Sanjay Sinha, MD; Michael R. Recto, MD

Keywords: percutaneous transcatheter pulmonary valve, pulmonary artery stenting, transvalvular interventions, tetralogy of Fallot, pulmonary atresia, pulmonary branch stenosis, RV to PA conduit stenosis

Abstract

Background: Transcatheter pulmonary valve implantation is a well-accepted method of addressing severe pulmonary valve insufficiency or stenosis following complete repair of tetralogy of Fallot (TOF) and similar lesions that require placement of a transannular right ventricular outflow tract patch or placement of right ventricle to pulmonary artery conduit. Many patients have benefitted from transcatheter pulmonary valve placement (TPV), but little information is known regarding TPV leaflet integrity following intervention to relieve distal conduit or main and branch pulmonary artery stenosis. Case: We describe six patients with either TOF or truncus arteriosus who had initially undergone complete surgical repair, followed by TPV implantation who then underwent either main or branch pulmonary artery angioplasty and stent implantation through previously implanted TPV. Results: Post intervention angiography showed competent TPV leaflet with normal leaflet function without evidence of pulmonary valve insufficiency. Post catheterization transthoracic echocardiography confirmed these findings. Conclusion: Multiple interventions across previously implanted TPV is a safe and effective procedure that can be successfully completed without damage to the transcatheter pulmonary valve leaflets.

Introduction

Transcatheter pulmonary valve (TPV) implantation has become accepted practice for patients of all ages following FDA approval of the Melody Valve (Medtronic Minneapolis, MN) in 2010 for Humanitarian Use device. The Sapien XT (Edwards Lifesciences, Irvine CA) TPV was FDA approved in March 2016 for use in dysfunctional RVOT conduits, and in March 2017 the FDA expanded approval of the Melody TPV for use in patients whose prior surgical valves have failed. More recently in August 2020, the FDA also approved the Edwards Sapien 3 Transcatheter Heart Valve that had previously been

JULY 2021

International Edition Vol. 19 - Issue 7

TABLE OF CONTENTS

approved for treatment of severe aortic valve stenosis to include treatment of severe pulmonary valve regurgitation and stenosis. The availability of the aforementioned TPVs has increased the number of patients that can be managed via the transcatheter approach avoiding open heart surgery to replace leaking or stenotic native pulmonary valves and conduits.[1,2] To date, there is limited information available regarding TPV leaflet integrity following transcatheter intervention through the previously implanted TPV. Little is known about the effect of interventions that require repeated or multiple crossings of the TPV on valve leaflet integrity. This case series describes six patients who safely underwent transcatheter interventions through previously implanted Melody and Sapien TPV while preserving the integrity and function of the pulmonary valve leaflets.

Case Series

Six patients (three females and three males) with mean age of 15 years (range 10-20 years), mean weight 54 kg (range 39-66), and mean body surface area 1.52m2 (range 1.45-1.75) underwent TPV implantation. Of the five patients with tetralogy of Fallot (TOF), three had severe pulmonary valve stenosis, one had pulmonary atresia with major aortopulmonary collateral vessels and one had absent pulmonary valve. The remaining patient had truncus arteriosus Type 1. All six patients had previously undergone complete repair with development of severe pulmonary valve insufficiency and right ventricular enlargement necessitating TPV implantation. Five of the patients underwent percutaneous transcatheter pulmonary implantation with a Melody valve (Medtronic, Minneapolis, MN, US) and one patient had a Sapien (Edwards Lifesciences, Irvine, CA, US) valve implanted. Following TPV implantation, six patients had evidence of either distal right ventricle to main pulmonary artery conduit or branch pulmonary artery stenosis. Two patients, one requiring distal conduit stenting **(Figure 1 A-D)** and another requiring left pulmonary artery stent re-dilation **(Figure 2 A-B)**, each underwent a second procedure 10.2 and 7.4 months after TPV implantation (early in our experience). The other four patients underwent branch pulmonary artery intervention during the same catheterization procedure after first undergoing TPV implantation.

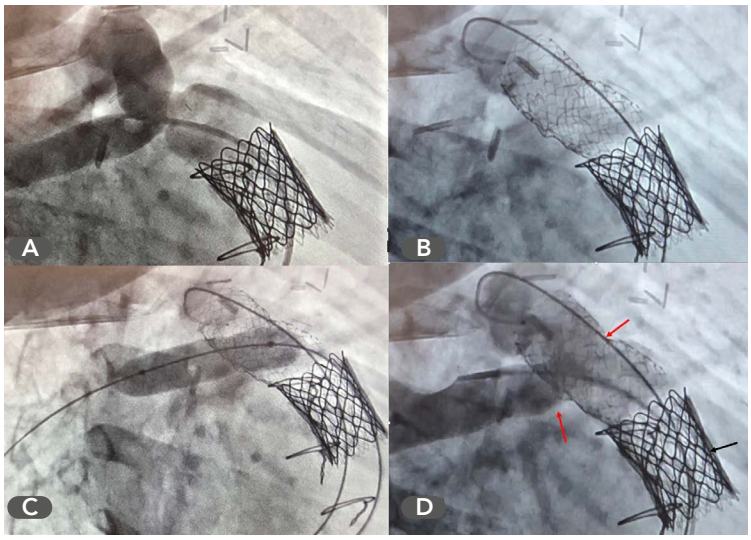


FIGURE 1 ***(A)*** *Angiography showing the distal conduit stenosis.* ***(B)*** *The stenosis responded well to stenting, with improvement in caliber.* ***(C)*** *The stenosed proximal right pulmonary artery was balloon angioplastied after advancing the balloon through a conduit stent strut.* ***(D)*** *Improvement in proximal right pulmonary artery caliber seen with post balloon angiography.*

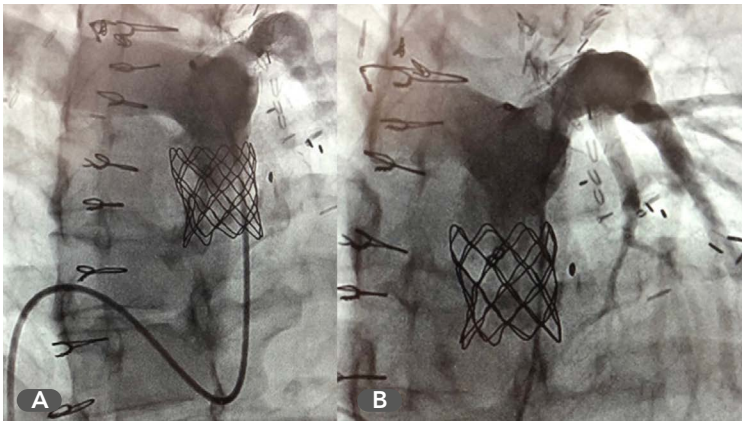


FIGURE 2 ***(A)*** *Proximal left pulmonary artery in-stent stenosis.* ***(B)*** *Improvement of the in-stent stenosis following balloon redilation.*

Cases

Areas of stenosis were documented in the distal main pulmonary artery or branch pulmonary arteries utilizing an Arrow Berman balloon-tipped angiographic catheter (Teleflex, Wayne, PA, US) that was advanced through the previously implanted TPV with the balloon inflated so as to minimize trauma to the previously implanted TPV leaflets. The Berman catheter was then replaced with an Arrow Balloon Wedge catheter (end hole catheter) that was advanced with the balloon inflated past the TPV into the distal branch pulmonary artery. An Amplatz Super Stiff wire (Boston Scientific) wire was then advanced through the wedge catheter into a distal right or left pulmonary artery branch. For the patients that required stent implantation of the distal main pulmonary artery or branch pulmonary arteries, a long transseptal sheath was initially positioned across the TPV to protect the TPV leaflets from the multiple wire and balloon exchanges that were required to deliver the stents **(Figure 3)**. For the patient that required re-dilation of a previously implanted stent **(Figure 4)**, the angioplasty balloon was delivered directly over the Super Stiff wire without utilizing a transseptal sheath and positioned across the stent. After the stent was re-dilated, the angioplasty balloon was deflated and removed over the Super Stiff wire. Post angioplasty angiography performed with a Berman angiographic catheter demonstrated competence of the TPV with normal leaflet function without evidence of pulmonary valve insufficiency, perivalvar leak or valve migration.

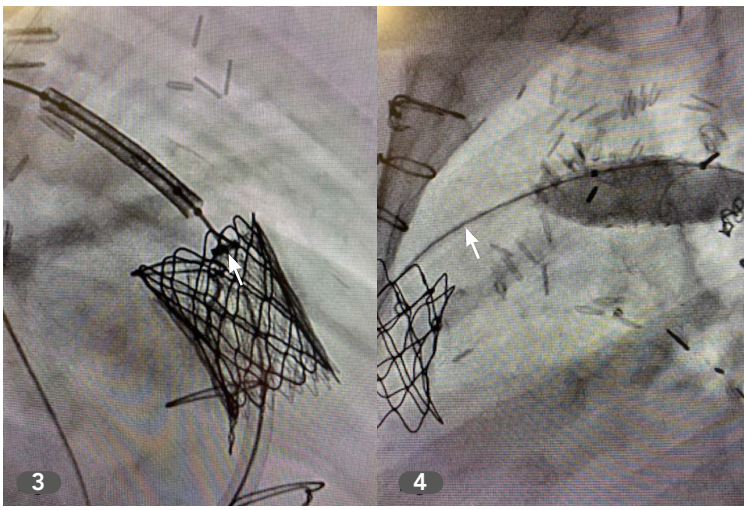


FIGURE 3 *The tip of the long transseptal sheath (white arrow) is positioned above the level of the pulmonary valve leaflets.*

FIGURE 4 *The balloon utilized to perform left pulmonary artery stent redilation was directly advanced over a super stiff wire (white arrow).*

Results

Please see **Table 1** for full results. Four of the six patients underwent stent implantation into distal right ventricle to main pulmonary artery conduit stenosis (one patient) and right or left branch pulmonary arteries (three patients) with Palmaz Genesis (Cardinal Health, Dublin, OH) or Max LD (Medtronic) stents. All six patients underwent branch pulmonary artery angioplasty with Atlas Gold (Bard Medical, New Providence, NJ, US), Vida (Bard), Powerflex (Cardinal), or Opta Pro (Cardinal) balloons. All patients underwent a minimum of two interventions (median 2.5, range 2-4) through the TPV. All six patients underwent successful distal right ventricle to main pulmonary artery conduit or branch pulmonary artery balloon angioplasty and/or stenting without complications. Post-intervention angiography showed competence of the previously implanted TPV with normal leaflet function and no evidence of pulmonary insufficiency, perivalvular leak, or valve migration. All patients then underwent post-catheterization transthoracic echocardiography demonstrating intact TPV leaflet integrity without pulmonic insufficiency. Post-catheterization course was uneventful for all patients.

Discussion

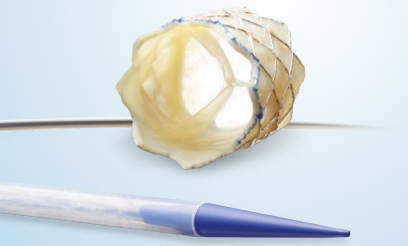
Transcatheter pulmonary valve implantation is now widely accepted as standard therapy for patients who have developed severe pulmonary valve insufficiency with subsequent development of right ventricular enlargement following repair of tetralogy of Fallot with transannular right ventricular outflow tract patch or following placement of right ventricle to main pulmonary artery conduits that have failed.[1,2] TPV is also now routinely utilized in patients whose prior surgical valves have failed (valve in valve procedure).[3] What is not well known or described is the effect of repeated interventions on TPV leaflet integrity following transcatheter intervention through previously implanted TPV. This case series describes six patients who underwent pulmonary artery stent implantation and balloon angioplasty through previously implanted Melody and Sapien S3 TPV **(Table 1)**. Early in our experience, two of the patients, one requiring distal conduit stenting and another requiring left pulmonary artery stent re-dilation, underwent a second procedure 10.2 and 7.4 months after their initial TPV implantation, respectively. The four other patients underwent balloon angioplasty and stent implantation immediately following TPV implantation. Five of the six patients underwent balloon angioplasty before undergoing stent implantation. One patient only required balloon angioplasty/stent re-dilation of a previously implanted left pulmonary artery stent. In all six patients there was no disruption of TPV leaflet integrity and in the four patients who had undergone balloon angioplasty and branch pulmonary artery stent implantation immediately following TPV, there was no dislodgement or movement of the TPV or development of increased TPV insufficiency on review of main pulmonary artery angiography following TPV implantation. In order to minimize trauma to the TPV leaflets, a long transseptal sheath was first positioned across the TPV. The angioplasty balloon and stents were then delivered through the transseptal sheath into the distal main pulmonary artery and branch pulmonary artery stenosis without injuring the TPV leaflets. In one case balloon angioplasty/stent re-dilation was performed over a Super Stiff wire without utilizing a long transseptal sheath without damage to the TPV leaflets. In retrospect, the operators feel that the balloon utilized for the stent re-dilation procedure should have been

TABLE 1 Demographics and Interventions

Patient	Sex	Age (years)	Weight (kg)	BSA	Diagnosis	Implanted PV	Balloon Angioplasty	PA Stenting	Total Interventions
1	F	10	51	1.45	ToF severe PS	Melody	3	1	4
2	F	20	55	1.54	ToF severe PS	Melody	2	0	2
3	F	16	65	1.75	ToF severe PS	Edwards	1	1	2
4	M	12	39	1.16	ToF PA MAPCAs	Melody	2	1	3
5	M	12	66	1.69	ToF absent PV	Melody	3	1	4
6	M	18	48	1.53	Truncus Arteriosus I	Melody	2	0	2
Average		14.667	54	1.52			2.167	0.667	2.833
Median		14	53	1.535			2	1	2.5

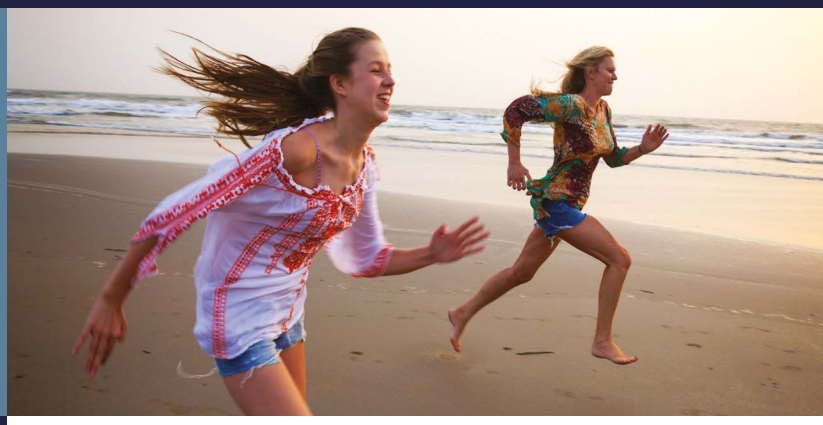
BSA = Body Surface Area; MAPCAs = Major Aortopulmonary Collateral Arteries; PA = Pulmonary Artery; PS = Pulmonic Stenosis; PV = Pulmonary Valve; ToF, Tetralogy of Fallot.

RIGHT CHOICE.



Melody™

Transcatheter Pulmonary Valve (TPV) System



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

Restoring lives for

13

years and counting.

The only transcatheter pulmonary valve specifically designed for RVOT conduits and bioprosthetic valves. The longest studied transcatheter valve, with the largest body of clinical evidence at over 10 years.* More than 16,000 patients' lives have been changed over 13 years, and counting.

Melody TPV — The Right Choice for Your Patients

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

UC201809495b EN 11/2020

Medtronic
Further, Together

Important Labeling Information for the United States

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

Contraindications: None known.

Warnings/Precautions/Side Effects

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

Potential procedural complications that may result from implantation of the Melody device include the following: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, blistering, or peeling of skin, pain, swelling, or bruising at the catheterization site. Potential device-related adverse events that may occur following device implantation include the following: stent fracture,* stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, hemolysis.

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

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

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

Important Labeling Information for Geographies Outside of the United States

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

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

Contraindications

- Venous anatomy unable to accommodate a 22 Fr size introducer sheath
- Implantation of the TPV in the left heart
- RVOT unfavorable for good stent anchorage
- Severe RVOT obstruction, which cannot be dilated by balloon
- Obstruction of the central veins
- Clinical or biological signs of infection
- Active endocarditis
- Known allergy to aspirin or heparin
- Pregnancy

Potential Complications/Adverse Events: Potential procedural complications that may result from implantation of the Melody device include the following: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, pain, swelling or bruising at the catheterization site. Potential device-related adverse events that may occur following device implantation include the following: stent fracture.* stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, hemolysis.

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

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

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

medtronic.com

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

LifeLine
CardioVascular Technical Support
Tel: (877) 526-7890
Fax: (651) 367-0918
rs.structuralheart@medtronic.com

UC201809495b EN 11/2020

Medtronic

delivered through a long transseptal sheath to minimize trauma from the uneven folding of the angioplasty balloon following stent re-dilation that could have damaged the TPV leaflets. All six patients underwent post procedure main pulmonary artery angiography demonstrating competence of the TPV. Transthoracic echocardiography performed the following day prior to discharge demonstrated absence to at most trace TPV insufficiency.

Conclusion

Transcatheter interventions consisting of balloon angioplasty and stent implantation can be performed safely and without injury to the pulmonary valve leaflets following TPV implantation. The operators recommend initially crossing the previously implanted TPV with balloon tipped catheters and once the TPV has been crossed so that a long transseptal sheath could be utilized to facilitate wire and catheter exchange through the TPV whenever possible. Once the transseptal sheath and wire are positioned across the TPV, wire and catheter exchanges can be performed through the TPV without risking damage to the leaflets of the TPV. In one case the operators did not utilize a transseptal sheath and an angioplasty balloon was directly advanced and subsequently withdrawn over the Super Stiff wire after performance of branch pulmonary artery stent re-dilation. While there was no damage to the valve leaflets, the uneven folding of the angioplasty balloon that can occur after the balloon is inflated could have damaged the TPV leaflets as there would have been direct contact between the rough uneven folds of the balloon and the valve leaflets. While this patient's TPV maintained leaflet integrity, the operators do not recommend performance of angioplasty procedures without first positioning a long transseptal sheath across the TPV.

References

1. Cheatham, J. P., Hellenbrand, W. E., Zahn, E. M., Jones, T. K., Berman, D. P., Vincent, J. A., & McElhinney, D. B. (2015). Clinical and hemodynamic outcomes up to 7 years after transcatheter pulmonary valve replacement in the US melody valve investigational device exemption trial. Circulation, 131(22), 1960-1970. doi:10.1161/CIRCULATIONAHA.114.013588 [doi]
2. Daily JA, Tang X, Angtuaco M, Bolin E, Lang SM, Collins RT. Transcatheter versus surgical pulmonary valve replacement in repaired tetralogy of fallot. Am J Cardiol. 2018;122(3):498-504. doi: 10.1016/j.amjcard.2018.04.028.
3. Cabalka AK, Asnes JD, Balzer DT, et al. Transcatheter pulmonary valve replacement using the melody valve for treatment of dysfunctional surgical bioprostheses: A multicenter study. J Thorac Cardiovasc Surg. 2018;155(4):1712-1724.e1. doi: S0022-5223(17)32772-1 [pii].

SHARIB GAFFAR, MD

Pediatrics Resident, PGY-3
CHOC Children's Hospital of Orange County
Orange, CA, USA
University of California Irvine
Irvine, CA, USA
sharibgaffar@gmail.com

SANJAY SINHA, MD

Pediatric Interventional Cardiologist
Assistant Professor, Pediatrics
CHOC Children's Hospital of Orange County
Orange, CA, USA

MICHAEL R. RECTO, MD

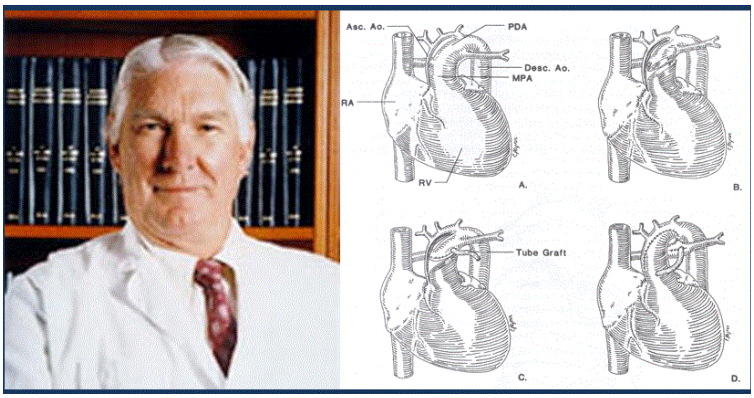
Medical Director, Cardiology
Medical Director, Cardiac Catheterization Lab
CHOC Children's Hospital of Orange County
Orange, CA, USA

In Memoriam: William I. Norwood Jr (1941-2020)

Edward J. Malec, MD, PhD & Marshall L. Jacobs, MD

With great sorrow, we acknowledge the passing of a dear friend and an esteemed mentor and colleague, William I. Norwood Jr.

On Sunday, December 13, 2020, Dr. William Imon Norwood, Jr, "Bill," loving husband, father of three children, grandfather to nine and great-grandfather to five, passed away at the age of 79. In reality, the "extended family" that mourn Bill's passing, and whose lives were touched in immeasurable ways by his life, numbers in the thousands, or likely tens of thousands. So many families in all parts of the world have been beneficiaries of Bill's contributions to the present-day understanding of congenital heart disease and its management. Many thousands of patients, in numerous countries, were direct recipients of Bill's outstanding care, having benefited directly from his incomparable surgical skill and his genius in harnessing the fundamentals of cardiovascular physiology to design surgical therapies and optimize outcomes. Many, many more were beneficiaries of Bill's gifts, through the work of countless other surgeons, cardiologists, anesthesiologists, nurses, and additional caregivers whose careers were directly impacted by his teachings, his mentorship, and the advanced understanding of Congenital Heart Disease that he promoted.



In 2010, the inaugural issue of the World Journal for Pediatric and Congenital Heart Surgery included an article by Dr. Norwood, titled "Our Roots, Our Future."[1] This text version of an address that Dr. Norwood had delivered at the 2009 meeting of the Congenital Heart Surgeons' Society recounted the contributions of some of his mentors and predecessors in the field of congenital heart surgery. Dr. Norwood emphasized the importance of innovation and discussed some of the inherent challenges that go along with it. Reflecting on the contributions of Robert Gross, C. Walton Lillehei, Clarence Dennis, Richard Varco, Aldo Castaneda, and others, Dr. Norwood concluded that "... the following are some of the characteristics of an innovator: confidence, conviction, creativity, courage." As he has done numerous times in numerous settings, he mentioned an aphorism attributed to the late Theodore Levitt, former professor at the Harvard Business School and editor of Harvard Business Review. Levitt, he said, put it simply: "Creativity is thinking up new things. Innovation is doing new things." In 2015, when the first Lifetime Achievement Award of the Congenital Heart Surgeons' Society was bestowed upon Dr. Norwood, it was accompanied by an expression of profound respect, but also one of gratitude for his having the "Curiosity, Determination and Genius that it takes to Think Up New Things ... for the Empathy, Faith and Courage that it takes to Do New Things ... and the Generosity to use those gifts to improve so many, many lives."

All who devote their careers to improving the lives of individuals with congenital heart disease have been influenced by Dr. Norwood's enormous contributions. His influence extends to individuals in diverse roles. Among them are practitioners who are engaged in virtually every aspect of the multidisciplinary management of congenital heart disease. Perhaps those who worked side-by-side with him in Boston, Philadelphia, Genolier, Wilmington, Krakow, Oslo, Rome, and Shanghai feel most tangibly a sense of indebtedness to Bill Norwood as teacher and mentor. It is the case, however, that surgeons (and their non-surgical colleagues) throughout North, Central and South America, Europe, Asia, Africa, and Oceania have benefited from Dr. Norwood's mentorship and are more versatile and more effective as a result of his teaching.

It is important though, to be mindful of the fact that the lives most directly impacted by the contributions of an innovator such as Bill Norwood are the patients—those individuals who might not otherwise have the opportunity to get up each morning and face the challenges of another day. In this journal, in 2016, a pediatric cardiac intensive care unit nurse who was born with hypoplastic left heart syndrome (HLHS) and had undergone her initial surgery by Dr. Norwood in 1987 shared a very personal perspective.[2] She said, "People have always asked me, both while I was growing up and since I have become an adult, "Was my life with congenital heart disease normal?" Well, what's normal? Was I sick a lot? Yes. Was I teased a lot for being small, for not having a lot of strength, for not being able to run? Yes. Were these things hard for me? Yes. However, I tried not to focus on what I could not do but would instead focus on what I could do. I could not play soccer, OK; I did not want to play soccer. I wanted to take Taekwondo! So, I did and soon earned my black belt! I wanted to be a cheerleader, so I tried out. I ended up being a flier who was at the top of the pyramids and did stunts. I did whatever I wanted, and I rarely asked for permission. I was intent on leading my life to the fullest without unmerited restrictions."

Now, several years after undergoing neo-aortic valve replacement, she continues to work in the Heart Institute Clinic. Of this work, she said, "Working in the clinic has allowed me to work closely with other heart patients and their families. I have loved getting to know these families on a personal level and seeing them on a consistent basis."

FIGURE 1 *Photograph taken during a picnic at the home of Dr. Norwood in Gladwyne, Pennsylvania, in the early 1990s.*

FIGURE 2 *Dr. Norwood and Dr. Malec at a gathering of families in Zakopane, Poland. The red T-shirts bear the inscription: "S/P Norwood, hope and miracle.*

Every patient's story is unique, but some common themes do emerge. In 2015, a young woman who was at that time a premedical student at college was asked what Bill Norwood's contributions mean to her. She had never met Dr. Norwood, though she was born in 1994 with HLHS and underwent what many call "the three stage Norwood Procedure." So, when asked what Dr. Norwood's contributions mean to her, she responded, "For my parents, it was the chance to have a family . . . But for me, it is every opportunity I have ever received. It's about being able to have dreams of things I'd like to do 5, 10 or 15 years from now." Where those dreams have taken her is to medical school, where she will soon graduate and pursue training in pediatric cardiology.

These anecdotes are but a few of the many stories that each of us could recount, by way of illustrating the countless lives that have been positively influenced by Bill Norwood and his immeasurable contributions to the understanding of congenital heart disease and the care of patients and families whose lives are affected by it. The accompanying photograph **(Figure 1)** was taken during a picnic at Dr. Norwood's home in Gladwyne, Pennsylvania, in the early 1990s. Each year, patients and their families were invited to get together in his back yard and enjoy one another's company. Without echocardiograms, one cannot be absolutely certain. But it is very likely that the older guy with white hair in the second row is the only person in the picture with two well-developed ventricles. As for the others, most are now beginning the fourth decade of their lives. In **Figure 2**, Dr. Norwood is seen with a gathering of families in Zakopane, Poland, in 2017. The yellow inscription on the back of the children's red T-shirts says: "S/P Norwood, hope and miracle." Surely, these "snapshots" suggest the tangible legacy of Dr. William I. Norwood Jr., MD, PhD.

References

1. Norwood WI. Our roots, our future. World J Pediatr Congenit Heart Surg. 2010;1(1): 127-131. doi:10.1177/2150135109360814.
2. Reed JM. Perspectives on HLHS from a pediatric cardiology nurse. World J Pediatr Congenit Heart Surg. 2016;7(5): 665-667. doi:10. 1177/2150135116662809.

This article was originally published in the World Journal for Pediatric and Congenital Heart Surgery. 2021;12(2): 161-163.
 DOI: 10.1177/2150135121992008.

EDWARD J. MALEC, MD, PHD
Department of Pediatric Cardiac Surgery
University Hospital Munster
Munster, Germany

MARSHALL L. JACOBS, MD
Department of Surgery
Johns Hopkins School of Medicine
1800 Orleans Street
Baltimore, MD, USA
marshall.jacobs@jhmi.edu

The PICS 25th Anniversary Symposium and Inaugural Fellows/Early Career Course: An Interview with the Program Directors

The PICS Society's Communications Staff recently interviewed Directors of the PICS Society's 25th Anniversary Symposium (September 1-4, 2021, in Las Vegas) and the inaugural PICS Fellows/Early Career Course (August 30-31, 2021 in the same location). Interview highlights:

The 25th Anniversary PICS Symposium

PICS staff: *Dr. Hijazi, we have heard you refer to the 2021 PICS Symposium as both the 25th anniversary and the first anniversary meeting! How is this possible?*

Ziyad M. Hijazi, MD, MPH, FPICS (PICS Society President & Symposium Founder/ Director): This is the 25th anniversary of the PICS Symposium AND the first anniversary for the new PICS Society. In 1997 our field didn't have its own focused educational meeting. We needed to change that! I explored the idea with my mentors Dr. Bill Hellenbrand and the late Dr. Charlie Kleinman, and with the great help of my first boss Dr. David R. Fulton. With their encouragement, in 1997 we held our first meeting and grew quickly from there. We constantly update the meeting while always focusing on excellence in education towards the best possible patient care.

PICS staff: Congratulations on the 25th Silver Anniversary! So, why is this also the first anniversary?

Dr. Hijazi: September marks the first anniversary of The Pediatric and Congenital Interventional Cardiovascular Society--The PICS Society--the professional home for our global community. Last year we looked at how best to represent the interests of interventional congenital cardiologists and our amazing teams. Until now, no organization has focused exclusively on our needs. With encouragement of colleagues worldwide, we formally launched The PICS Society late last year and are growing very fast!

PICS staff: Dr. Kenny, you have noted The PICS Symposium is the Society's flagship event, and that the Society as a membership organization is spearheading many additional areas.

Damien Kenny, MD, FPICS (PICS Society Vice President & PICS Symposium Director): The Symposium is a vital part of the PICS Society. It's exciting that the culture of **excellence and community** Dr. Hijazi alluded to continues to drive the Symposium and the Society overall. The PICS Society is now the dedicated home for passionate physicians who do amazing work for the equally amazing patients we treat, with many opportunities for our members to get involved--guidelines, advocacy, education, early career and more. Our culture is dedicated to learning from one another, constantly improving and advocating as a unified voice for our patients and colleagues.

PICS staff: Is it true that members of the PICS Society receive special recognition and benefits?

Dr. Hijazi: Yes! Members receive a significant registration discount for the PICS Symposium if they stay in the Aria Las Vegas hotel-- we have negotiated an EXTREMELY low room rate for this world class hotel. Those who apply for membership by August 1 will be **Founding Members** of the Society. For those who qualify for the FPICS designation (Fellow of the PICS Society), FPICS wall certificates will be presented at a Symposium ceremony recognizing the highest level of achievement in our profession. This will be a historic moment.

PICS staff: Let's talk about safety. The world is starting to open up after an incredibly challenging year.

Dr. Hijazi: We take safety extremely seriously. COVID-19 has been devastating and we are a global organization. We are working closely with the hotel to ensure a safe, enjoyable gathering. Regarding social distancing, masking, sanitizing and the like, we are monitoring all policies, rules and guidelines--and will insist on adherence to them. Our staff and contractors are all experienced in these matters. As medical professionals, we believe in the importance of vaccination and urge attendees to do so.

Last year's Symposium was fully virtual to great success under difficult circumstances. This year we recognize some colleagues still will be unable to travel. There will be a hybrid component this year, with selected sessions streamed or archived for future viewing.

There is nothing better than an in-person meeting where you will interact with your colleagues. Networking is so important--we have all missed this so much during the past year. We are social beings, and we have all been starved for social contact for so long. We would like our attendees to come and have a wonderful experience in a great venue.

PICS staff: Tell us about opportunities to interact with industry partners at the Symposium.

Dr. Hijazi: This is crucial. CEO's or their representatives will be there to learn our needs. Take time to introduce yourself, interact, learn and avail yourself of opportunities for hands-on demonstrations. The relationship between industry and medical professionals is extremely important; we have the ideas about what we need to better treat patients. Industry has the engineers, the scientists and the resources to meet those needs.

Dr. Kenny: In the exhibit hall, industry will display the latest products and the tried-and-true. Everything will be on display, with those who developed these products available for discussion. Industry demonstrations in the exhibit hall and expert presentations during industry lunchtime sessions will focus on

advances in imaging and new interventional devices.

PICS staff: What will be the focus of this year's Symposium?

Dr. Kenny: The lynchpin will be live case demonstrations from around the world. Seeing how teams perform procedures provides invaluable educational benefit. We will have taped moderated cases with instructor/attendee interaction. We will have didactic lectures, vigorous debates and demonstrations. Finally, we will be providing a full 30 hours of CME, very important for board certification or recertification and, of course, for one's own professional development.

Dr. Hijazi: Whether you are new to the field or an experienced operator, there is always something new to learn. I can assure you 100% you will return home better informed to treat your patients--and that's our goal!

The Inaugural PICS Fellows and Early Career Course

Goal of this new course. Darren Berman, MD, FPICS, Course Co-Director: For those

in advanced training or early years of practice, there is a keen desire to constantly learn and improve. Our goal is to fill this niche: to provide a relatively small, focused program for those early in their careers. For fellows, the course will complement their home institution program relatively early in their interventional training. For early career interventionalists, this course will complement what they have learned. For both groups, they will be able to build networks that will benefit them throughout their careers.

It sounds exciting! Tell us more. Vivian Dimas, MD, FPICS, Course Co-Director:

What's exciting is that we will have a very full two days focused on this small group of attendees, who will then hopefully continue to expand on many of the topics covered by participating in the full Symposium. The faculty, attendees, live cases and taped moderated cases are all global, with core faculty eager to interact with attendees. There will be lively back-and-forth including case-based learning. "What have we done well? Where can we improve? What have we learned from our mistakes?" That's how we will teach this course.

How do fellows & early career physicians apply for this course? Dr. Dimas: Space is limited, so apply early! We have made the application process simple: Go to www.picsymposium.com and click on "registration" then "fellows course."

How will you serve two audiences--fellows and early career-- in one course? Dr. Berman: Think about it: an interventional fellow and a first-year attending are separated by one day! One day you're a fourth year. The next day you begin your first year in practice. The lines are blurry and your first day as an attending can be a little nerve wrecking-I know mine was!

Dr. Dimas: In those training and early years, there is much to learn, especially from faculty who have made mistakes themselves and want to share those lessons learned. Call it wisdom if you want to. These two groups of attendees are going to align very well.

Collaboration with industry. Dr. Dimas: We so appreciate their support and partnership, without which this course would be impossible to offer. We faculty are all volunteers, but this course does require resources. For this course and for our field as a whole, we are indebted to our industry partners and cannot thank them enough for their ongoing collaboration and partnership. This course will facilitate working relationships between attendees and industry, which will benefit our field's development well into the future.

Networking. Dr. Berman: The small size of this meeting creates the right environment for interactive discussion in the classroom and relationship building outside of the conference. This short time together can set the foundation for a lifetime of interactions. The nature of this focused course will allow a seamless way to start creating those relationships and bonds.

Why are you doing this? Dr. Berman: Great question! Each of us wants to contribute beyond the clinical work we do each day. It's important to find those things that really light the fire in each of us. For me, this opportunity to help create and build this course from scratch lays the foundation for teaching, learning, creating new friendships, and gives me the opportunity to start "paying it forward" by helping the next generation of great congenital interventional cardiologists. I'm grateful for the opportunity to help do this, and I know that Dr. Dimas and the entire faculty feels the same way.

Researchers Identify New Way to Improve the Diagnosis of Fetal Heart Defects

UC San Francisco researchers have found a way to double doctors' accuracy in detecting the vast majority of complex fetal heart defects in utero - when interventions could either correct them or greatly improve a child's chance of survival - by combining routine ultrasound imaging with machine-learning computer tools.

The team, led by UCSF cardiologist Rima Arnaout, MD, trained a group of machine-learning models to mimic the tasks that clinicians follow in diagnosing complex Congenital Heart Disease (CHD). Worldwide, humans detect as few as 30-50% of these conditions before birth. However, the combination of human-performed ultrasound and machine analysis allowed the researchers to detect 95% of CHD in their test dataset.

The findings appear in the May issue of Nature Medicine.

Fetal ultrasound screening is universally recommended during the second trimester of pregnancy in the United States and by the World Health Organization. Diagnosis of fetal heart defects, in particular, can improve newborn outcomes and enable further research on in utero therapies, the researchers said.

Typically, the imaging includes five cardiac views that could allow clinicians to diagnosis up to 90% of Congenital Heart Disease, but in practice, only about half of those are detected at non-expert centers.

"On the one hand, heart defects are the most common kind of birth defect, and it's very important to diagnose them before birth," Arnaout said. "On the other hand, they are still rare enough that detecting them is difficult even for trained clinicians, unless they are highly sub-specialized. And all too often, in clinics and hospitals worldwide, sensitivity and specificity can be quite low."

The UCSF team, which included fetal cardiologist and senior author Anita Moon-Grady, MD, trained the machine tools to mimic clinicians' work in three steps. First, they utilized neural networks to find five views of the heart that are important for diagnosis. Then, they again used neural networks to decide whether each of these views was normal or not. Then, a third algorithm combined the results of the first two steps to give a final result of whether the fetal heart was normal or abnormal.

"We hope this work will revolutionize screening for these birth defects," said Arnaout, a member of the UCSF Bakar Computational Health Sciences Institute, the UCSF Center for Intelligent Imaging, and a Chan Zuckerberg Biohub Intercampus Research Award Investigator. "Our goal is to help forge a path toward using machine learning to solve diagnostic challenges for the many diseases where ultrasound is used in screening and diagnosis."

Developing a Lead Extraction Program for Patients with Complex CHD

Wilson W. Lam, MD, FACC; Douglas Mah, MD; Jeffrey J. Kim, MD, FACC

Pacemaker and defibrillator lead extraction plays an important role in lead management strategy and complements novel technology, such as leadless pacing and subcutaneous implantable cardiac defibrillators. Indications and recommendations for lead extraction have been established.[1,2] Pediatric and adult congenital heart disease (CHD) centers may have lower case volumes than adult lead extraction centers. However, CHD adds anatomic complexity in a younger population of smaller patient size, where limited transvenous access, prior operations, and baffles and conduits may alter lead position. The 2014 PACES/HRS Consensus Statement recommends that device-based procedures be performed in a regional ACHD center laboratory with appropriate personnel and equipment, by an electrophysiologist with expertise in CHD.[3] Guidelines have identified necessary components of a CHD lead extraction program, and we propose modifiers that demonstrate CHD expertise **(Table 1)**. In experienced centers, procedural success remains high (~95%), with reported complication rates in the range of approximately 3-4%,[4-7] with no mortality – similar to adult series.[8,9] Due to anatomic complexity and potential for adverse outcomes which may be life threatening, centers performing these procedures should have a designated team familiar with the CHD management to maximize procedural success and safety.

TABLE 1 Suggested CHD Lead Extraction Operator Training/ Expertise

	Guideline Recommendation	Suggestion
Lead extraction specialist	Electrophysiologist or CT surgeon Initial: 30 lead extractions Annual: 20 extractions	Primary operator: training in pediatric or adult congenital heart disease or Co-operator: training in pediatric electrophysiology
Cardiothoracic (CT) surgeon	Continuing education in surgical management of lead complications Immediately available for emergent sternotomy or thoracotomy within 5-10 minutes	Additional congenital heart disease training
Cardiovascular anesthesiologist	Procedures performed under general anesthesia in most centers which facilitates intraprocedural TEE, and eliminates urgent intubation	Additional congenital heart disease training
Tools	Simple manual traction Lead locking stylets Telescoping sheaths Femoral snares Mechanical cutters Laser sheaths	For leads that have been in place for a longer time ("dwell time"), the likelihood of requiring snares is higher
Intraoperative imaging	Strong recommendation for transesophageal echocardiogram (TEE) or intra-cardiac echocardiography (ICE) based on operator familiarity and comfort with image interpretation	ICE - less fluoroscopic interference; better right heart visualization; relies on the extractionist for interpretation. TEE - less costly upfront; image interpretation independently performed by an echocardiographer.
Access	Some centers routinely place femoral arterial and venous sheaths for rapid cannulation if bypass is necessary	Venous occlusion and interrupted inferior vena cava with azygos continuation may limit maneuverability
Vascular tear preparation	Some centers include interventional radiologists or vascular surgeons to assist with percutaneous management of vascular tears	Routine preparation of tamponade balloon. Coordination with interventional cardiology (if use of covered stent required).

Where and who is to perform lead extractions for anatomically complex cases need to be determined. Both the Heart Rhythm Society (HRS) and European Heart Rhythm Association (EHRA) suggest that competency is conferred with the performance of at least 30-40 lead extractions as the primary operator, requiring 20 lead extractions annually to maintain competency.[10,11] Large adult referral centers perform an average of 9-16 cases (17-29 leads) per month[12,13] compared to pediatric and adult centers that perform approximately 0.2-1 case (0.3-2.4 leads)/month, albeit with an anticipated volume increase due to an aging ACHD population. Though lead dwell time is similar (~6-7 years), CHD centers treat cases with more complex anatomy, including 23% who had undergone atrial switch operations for d-transposition of the great arteries.[4-7] These differences in volume and anatomic complexity make it challenging to train extraction experts to care for Congenital Heart Disease patients.

Since the launch of our dedicated pediatric and congenital lead extraction program, we have identified strategies to assist with complex CHD **(Table 2)**:

TABLE 2 Strategies to Assist with CHD Extraction and Lessons Learned

	Recommendation	Comments
Pre-procedural		
Strategy	Aggressive lead management to remove dysfunctional leads and infected hardware	Institutional commitment to achieve this (e.g., equipment, staffing, operating room schedule, and surgical backup availability)
Case review	Multidisciplinary discussion between electrophysiologist, surgeon, and anesthesiologist	Review of prior imaging and past chest operations can assist with setup, access, approach, and surgical rescue planning
Protocols and simulation	Preparation for possible complications	Given rarity of emergencies, established routines and simulated practice may reduce mortality
Intraoperative		
Anesthesia	General anesthesia for all cases	Reduces patient movement and discomfort in anticipation of prolonged procedures and optimizes hemodynamics
Access	Arterial access obtained in all cases Venous access obtained in all cases	Real time blood pressure monitoring in case of emergency Immediate availability for percutaneous cannulation and rapid infusion of volume or blood products if superior vena cava (SVC) flow is obstructed when the tamponade balloon is deployed
Tamponade balloon	Access with stiff guidewire across SVC and innominate veins and 12F peelaway sheath	Tamponade balloon equipment prepped and readily available for immediate use
Imaging	Real-time intracardiac or transesophageal echocardiogram	Immediate detection of pericardial effusion and intraoperative assessment of leads. ICE may be beneficial in evaluating prosthetic valve function (difficult to image prosthetic pulmonary valves with TEE in cases of endocarditis).[14,15] TEE requires a separate echocardiographer for interpretation.
Pacing	Backup transvenous and transcutaneous pacing (in case of dislodgement) should be tested before extraction, if needed	Heart block without adequate escape rate may require additional venous access
Case differences		
High percentage of atrial switch operations	Majority of extractions in Mustard and Senning patients require concurrent baffle interventions	Coordination with congenital interventional cardiologist to optimize procedural yield
Lead dysfunction rather than infection	More aggressive attempts to salvage functional leads to limit collateral damage	Familiarity with techniques and potential complications may result in improved salvaging of functional leads
More snare techniques utilized	Longer lead dwell time and patient youth (<40 years of age) are associated with more challenging extractions[16]	Familiarity with femoral snare techniques and steerable sheaths can assist with challenging cases. Minority of cases are successful with manual traction alone (readiness to escalate to advanced methods and powered sheaths).
Complications	Anatomic concerns, cumulative instrumentation (e.g., number of generator changes over a lifetime), infection risk, and anticoagulation	Outcome Transparency and measurement of key metrics; Regular Data review to promote quality initiatives and improve outcomes

Conclusion

Lead extraction can be performed successfully in CHD with comparable results as non-CHD patients. However, differences exist between most adult institutions and those that care for patients with CHD. Additional training and experience are necessary for anatomically complex and challenging cases. Multi-disciplinary collaboration with focused attention on the nuances related to CHD is critical to ensure procedural success and safety.

References

1. Kusumoto FM, Schoenfeld MH, Wilkoff BL, et al. 2017 HRS expert consensus statement on cardiovascular implantable electronic device lead management and extraction. Heart Rhythm 2017;14:e503-e551.
2. Bongiorni MG, Burri H, Deharo JC, et al. 2018 EHRA expert consensus statement on lead extraction: recommendations on definitions, endpoints, research trial design, and data collection requirements for clinical scientific studies and registries: endorsed by APHRS/HRS/LAHRS. Europace 2018;20:1217a-1217j.
3. Khairy P, Van Hare GF, Balaji S, et al. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease: developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Heart Rhythm 2014;11:e102-65.
4. Gourraud JB, Chaix MA, Shohoudie A, et al. Transvenous lead extraction in adults with congenital heart disease: insights from a 20-year single-center experience. Circ Arrhythm Electrophysiol 2018;11:e005409.
5. Cecchin F, Atallah J, Walsh EP, Triedman JK, Alexander ME, Berul CI. Lead extraction in pediatric and congenital heart disease patients. Circ Arrhythm Electrophysiol 2010;3:437-44.
6. Fender EA, Killu AM, Cannon BC, et al. Lead extraction outcomes in patients with congenital heart disease. Europace 2017;19:441-46.
7. McCanta AC, Kong, MH, Carboni MP, Greenfield RA, Hranitzky PM, Kanter RJ. Laser lead extraction in congenital heart disease: a case-controlled study. PACE 2013;36:372-80.
8. Brunner MP, Cronin EM, Duarte VE, et al. Clinical predictors of adverse patient outcomes in an experience of more than 5000 chronic endovascular pacemaker and defibrillator lead extractions. Heart Rhythm 2014;11:799–805.
9. Wazni O, Epstein LM, Carrillo RG, et al. Lead extraction in the contemporary setting: the LExICon study: an observational retrospective study of consecutive laser lead extractions. J Am Coll Cardiol 2010;55:579–86.
10. Zipes DP, Calkins H, Daubert JP, et al. 2015 ACC/AHA/HRS advanced training statement on clinical cardiac electrophysiology (a revision of the ACC/AHA 2006 update of the clinical competence statement on invasive electrophysiology studies, catheter ablation, and cardioversion). J Am Coll Cardiol 2015;66:2767-2802.
11. Deharo JC, Bongiorni MG, Rozkovec A, et al. Pathways for training and accreditation for transvenous lead extraction: a European Heart Rhythm Association position paper. Europace 2012;14:124–34.
12. Tarakji KG, Saliba W, Markabawi D, et al. Unrecognized venous injuries after cardiac implantable electronic device transvenous lead extraction. Heart Rhythm 2018;15:318-25.
13. Tsang DC, Perez AA, Boyle TA, Carrillo RG. Effect of prior sternotomy on outcomes in transvenous lead extraction. Circ Arrhythm Electrophysiol 2019;12:e007278.
14. Sadek MM, Cooper JM, Frankel DS, et al. Utility of intracardiac echocardiography during transvenous lead extraction. Heart Rhythm 2017;14:1779-85.
15. Østergaard L, Vejlstrup N, Køber L, Fosbøl EL, Søndergaard L, Ihlemann N. Diagnostic potential of intracardiac echocardiography in patients with suspected prosthetic valve endocarditis. J Am Soc Echocardiogr 2019;32:1558-64.
16. El-Chami MF, Sayegh MN, Patel A, et al. Outcomes of lead extraction in young adults. Heart Rhythm 2017;14:537-40.

AUGUST

27-30

ESC Congress 2021 – The Digital Experience
Virtual
https://www.escardio.org/Congresses-&-Events/ESC-Congress

30-31

First Annual PICS Fellows & Early Career Course
Las Vegas, NV, USA
Kimberly_ray@chdinterventions.org

SEPTEMBER

01-04

PICS Society Symposium 2021
Las Vegas, NV, USA
https://picsymposium.com/

23-24

EPIC Live – Emory Practical Intervention Course
Virtual
https://www.epicsec.org/

OCTOBER

01-03

Echo in Congenital Heart Disease: Adult and Pediatric Cases, Including Multimodality Imaging
Rochester, MN, USA
https://cveducation.mayo.edu/store/echo-in-congenital-heart-disease-adult-and-pediatric-cases-including-multimodality-imaging

07-09

The Genetics of Heart & Vascular Disease
Nashville, TN, USA
https://cveducation.mayo.edu/store/the-genetics-of-heart-vascular-disease

CONGENITAL CARDIOLOGY TODAY

CORPORATE OFFICE
11500 Elk Horn Drive
Clarksburg, MD 20871 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.