Pediatric Insights

BEST CHILDREN'S HOSPITALS U.S.News & WORLD REPORT CARDIOLOGY & HEART SURGERY 2020–21

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An Update From the Heart Institute at UPMC Children's Hospital of Pittsburgh

In This Issue

UPMC CHILDREN'S HOSPITAL OF PITTSBURGH

Catheterization Lab Update:

New Director to Lead Major Expansion of Program

The Heart Institute at UPMC Children's Hospital of Pittsburgh has embarked on a multiyear, multifaceted expansion of the pediatric cardiac catheterization laboratory that will introduce new technologies for patient care, expand treatment options, and enhance UPMC Children's research capabilities.

Leading the effort is the new Director of the Cardiac Catheterization Laboratory (CCL) and Interventional Cardiology Service Bryan H. Goldstein, MD. Dr. Goldstein joined UPMC Children's in December 2019 as the new director of the CCL. Dr. Goldstein takes over director duties for the CCL from Pediatric Cardiology Division Chief and Medical Director of the Heart Institute Jacqueline Kreutzer, MD, FAAC, FSCAI, who has been responsible for growing the program over the last decade to become a nationally and internationally recognized leader in pediatric cardiac catheterization procedures and research. Dr. Kreutzer will continue her catheterization practice and research but will be focusing more on expanding the academic and research programs of the entire Heart Institute at UPMC Children's.

"Under Dr. Kreutzer's leadership, the CCL has markedly expanded its capabilities and affords patients some of the highest quality standards and procedural outcomes in the field. The success of the cardiac catheterization program at UPMC Children's is one of the reasons our Heart Institute is now ranked #2 in the country by *U.S. News & World Report*," says Mark Sevco, president of UPMC Children's.

An associate professor of Pediatrics at the University of Pittsburgh School of Medicine, Dr. Goldstein most recently held appointments at Cincinnati Children's Hospital, where he was the Associate Director of the Cardiac

CCL Leadership. *L to R: Bryan Goldstein, MD; Sara Trucco, MD; Jacqueline Kreutzer, MD.*

Catheterization Laboratory. Dr. Goldstein earned his medical degree at the Boston University School of Medicine, followed by pediatric residency training at Boston Children's Hospital. Dr. Goldstein next completed fellowships in cardiology and interventional cardiology at the University of Michigan C.S. Mott Children's Hospital in Ann Arbor. In addition to his responsibilities at UPMC Children's, Dr. Goldstein is co-founder and vice president of the Congenital

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Catheterization Research Collaborative (CCRC), a multicenter academic collaborative that fosters research and quality improvement efforts amongst a group of 10 congenital heart centers in the United States. The CCRC focuses on conducting outcomes research following surgical and transcatheter interventions for congenital heart disease. Upon his arrival, UPMC Children's joined the CCRC as its tenth member.

"We are pleased to have Dr. Goldstein join the Heart Institute at UPMC Children's and expand upon the stellar work that has propelled the Institute to the highest levels of patient care and recognition in the United States and internationally. Dr. Goldstein will continue to build upon our clinically outstanding program, and further develop our academic and research platforms to spur the next generation of advances in pediatric cardiology," says Victor Morell, MD, chief of Pediatric Cardiothoracic Surgery, Heart Institute co-director, and UPMC Children's Surgeon-in-Chief.

Catheterization Lab Expansion Plans and New Technologies

The two existing CCLs at UPMC Children's are planned to undergo complete reconstruction and refitting with the latest technologies and equipment for cardiac catheterization procedures. In addition, a new third lab space will be constructed in collaboration with the Department of Radiology (led by Ashok Panigrahy, MD) to house a hybrid MRI/catheterization suite that will allow for both cardiac MRI and cardiac catheterization procedures to be conducted simultaneously on two patients, or combined at the same time for patients in need of both catheterization and MRI.

"One of our goals is to conduct radiation-free cardiac catheterization with MR guidance for both diagnostic and interventional procedures. This field is in its infancy, but we anticipate that it will grow over time; getting in on the ground floor makes sense as a national leader in congenital heart disease management. However, this technology will not only allow for radiation-free procedures, it also will allow us to develop novel programs that rely upon both interventional techniques and cardiac MRI imaging data. We are preparing for the future, but that future is near," says Dr. Goldstein.

One of the clinical interests this new technology will allow Dr. Goldstein and colleagues to explore includes lymphatic interventions, a more recently identified major source of pathology and morbidity in patients with complex congenital heart disease.

"Plans for a lymphatic intervention program are in progress. We know we will be advancing more into this field to better treat many of our patients," says Dr. Goldstein.

The new imaging and intervention technologies will expand the Heart Institute's ability to comprehensively manage patients with single ventricle circulation, for which there are some current limitations in terms of treatment approaches. The hybrid suite will significantly reduce those limitations in the near future. This project also will allow for the expansion of invasive electrophysiology services, such as complex arrhythmia ablations and implantation of pacemakers and defibrillators in the CCL.

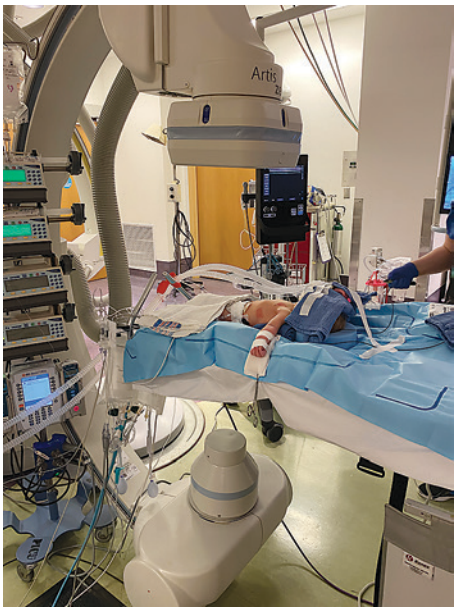
New Catheterization Programs for Infants with Heart Disease

Transcatheter PDA Stent for Palliation of CHD in Neonates

For those patients with cyanotic congenital heart disease, keeping the patent ductus arteriosus (PDA) open after birth is of great importance. For patients with ductal-dependent pulmonary blood flow — including patients with Tetralogy of Fallot, pulmonary atresia, and many other heart defects — transcatheter PDA stent implantation is a viable alternative to Blalock-Taussig (BT) shunt placement performed through an open surgical procedure.

Dr. Goldstein brings extensive clinical experience in catheter-based PDA stent procedures, and it has been a significant area of research for the CCRC, with numerous studies published by the group on the procedure over the last several years.

"The PDA stent procedure allows us to place a small coronary stent in the ductus arteriosus to maintain its patency — thereby allowing a patient to go home from the hospital — facilitating normal growth and bonding at home until the time of the next intervention when surgery would be performed. It affords the patient excellent palliation for their cyanotic congenital heart disease while also avoiding a major open-heart surgery as their first intervention in the high-risk neonatal period," says Dr. Goldstein.



PDA stent placement using the flip technique. *The patient is positioned backward compared to normal positioning to facilitate the procedure.*

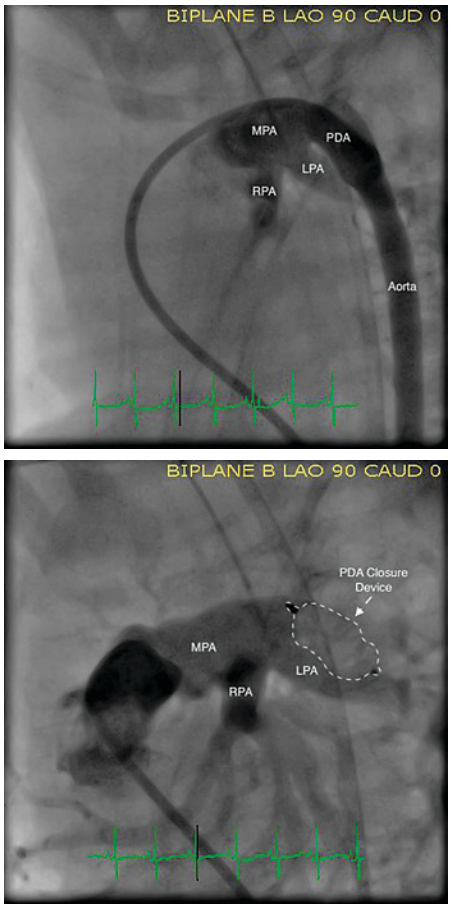
Catheter-based PDA Closure in Preterm Babies

A common comorbidity in very preterm neonates is persistent PDA. Neonates delivered before 30-weeks' gestation or who weigh less than 1500 grams at birth are at high risk for PDA. Having a PDA persist into postnatal life — in the setting of an otherwise healthy heart — can be significantly harmful to the premature infant, accelerating the development of chronic lung disease and increasing the risk of other complications, including pulmonary hypertension. For decades, older children with PDA who require closure have been successfully treated in the catheterization laboratory. Until recently, the only safe and viable option for very preterm neonates was surgical PDA ligation performed through a thoracotomy incision.

This invasive procedure was accompanied by substantial risks of postsurgical morbidity, which limited its use. Recently, techniques and equipment have evolved that facilitate the use of catheter-based techniques for closure of the preterm PDA, even in the extremely low birth weight infant.

Dr. Goldstein's substantial clinical experience will allow UPMC Children's to develop a robust PDA closure program in preterm neonates. The ability to offer this patient population access to a much less invasive procedure — with lower rates of complications and postprocedural morbidities in a catheter-based, short procedure — stands to greatly improve care in select patients for whom the procedure can benefit.

"We now have the tools and technology to transition most of these patients who require PDA ligation to transcatheter device closure via a short procedure. The first catheterization procedure that I performed after arriving at UPMC Children's was a PDA closure on the smallest patient ever treated in the catheterization laboratory here — just under 800 grams," says Dr. Goldstein.



Angiograms from a PDA closure procedure in a preterm infant. *Top: Baseline. Bottom: After closure. The patient was a 900-gram 24-week preterm infant. MPA: Main Pulmonary Artery; LPA: Left Pulmonary Artery; RPA: Right Pulmonary Artery; PDA: Patent Ductus Arteriosis.*

CLL Team Members

Fortifying the Academic Footprint and Research Impact

Dr. Goldstein and colleagues are set to expand the academic development and research programs associated with the cardiac catheterization laboratory on three separate fronts. The first has been UPMC Children's joining the CCRC network (www.ccrcresearch.org) of high-impact congenital heart centers to take part in its multicenter studies. The hospital is also likely to join and contribute to other multicenter research collaboratives in the future. More information about the CCRC can be found in Petit CJ, et al. Comprehensive Comparative Outcomes in Children with Congenital Heart Disease: The Rationale for the Congenital Catheterization Research Collaborative. *Congenital Heart Dis.* 2019; 14: 341-349.

A second priority is to facilitate UPMC Children's close collaboration with industry partners to conduct clinical trials on novel devices to treat various forms of congenital heart disease.

"Not only will we be able to contribute our significant resources and expertise to test some of these new devices, but it will also allow us to provide our patients greater access to emerging technologies that could afford life-altering and life-saving benefits," says Dr. Goldstein.

Additionally, Dr. Goldstein and colleagues at the Heart Institute at UPMC Children's will work on assessing the experiences, novel techniques, and programs related to catheterization procedures as a way to drive clinical innovation, be it through improvement in techniques, novel uses of existing technology, or the development of new platforms and devices.

"One area of excitement that I, personally, am looking forward to is collaborating with colleagues at the McGowan Institute for Regenerative Medicine and other local collaborators on potential device development and research," says Dr. Goldstein. "I am honored to be surrounded by an incredible group of collaborators, clinicians, and resources here at UPMC Children's. I look forward to building upon the exceptional catheterization program that Dr. Kreutzer and others have already propelled to the highest levels of clinical and research excellence."

ACHD Program Expands with New Faculty Member

The Heart Institute at UPMC Children's Hospital of Pittsburgh is pleased to welcome **Anita Saraf, MD, PhD**, to the Institute. Dr. Saraf is fellowship-trained in adult congenital heart disease (ACHD) and joins the UPMC ACHD program, which is directed by Arvind Hoskoppal, MD. Dr. Saraf officially began her tenure at UPMC Children's in August. Dr. Saraf holds dual academic appointments in the Department of Pediatrics and the Division of Cardiology in the Department of Internal Medicine at the University of Pittsburgh School of Medicine. In addition to her position in the Heart Institute at UPMC Children's, she also is a member of the UPMC Heart and Vascular Institute.

Dr. Saraf earned her doctorate in bioengineering from William Marsh Rice University in Houston, Texas, and her medical degree from Baylor College of Medicine. Dr. Saraf then completed both her internal medicine residency and ACHD fellowship at Emory University School of Medicine in Atlanta, Georgia.

"As a physician-scientist, I wanted to join an institution where I could be surrounded by a robust research program where collaboration and leadership support are integral to its mission. Equally important to me is being part of a world-class clinical care program where patient outcomes are the focus and drive decision making across the institution. UPMC Children's is just such a place," says Dr. Saraf.

Clinical Practice

Dr. Saraf's clinical practice encompasses the full spectrum of ACHD, with a special focus on Fontan patients and women with adult congenital heart disease.

"We have a great opportunity with the UPMC ACHD program to expand its scope of services to better reach an ever-growing patient population who need complex, long-term care to optimally manage their condition," says Dr. Saraf.

Past and Future Research Priorities

As a physician-scientist, Dr. Saraf became interested in studying biomarkers in adult congenital heart disease during her medical training at Texas Children's Hospital and continued that line of research at Emory University during residency and fellowship. While at Emory University, Dr. Saraf and colleagues established a biorepository for Fontan patients to study biomarkers and biomarker expression in this complex single ventricle congenital heart disease.

Dr. Saraf's research using data collected in the Fontan biorepository led to a new paper published in April in the *International Journal of Cardiology* which, among other findings, is the first to uncover that Fontan patients exhibit a chronically elevated inflammatory profile, together with a number of other cytokines as clinical markers of systemic dysfunction. These biomarkers, if monitored over time, can help clinicians predict which patients are about to get sick, long before they show any clinical signs. Biomarkers research in ACHD will continue to be a focus of Dr. Saraf's research at UPMC Children's.

Additionally, Dr. Saraf also is interested in determining how these biomarkers affect heart function. She currently is developing translational models that may help discover new therapies to help ACHD patients live longer and healthier lives.

In addition to her work on biomarkers in ACHD, Dr. Saraf also has investigated and published findings related to neurocognitive function in ACHD, and outcomes in pregnancy for ACHD patients.

Select Publications for Further Reading

Saraf A, DeStaercke C, Everitt I, Haouzi A, Ko Y, Jennings S, Kim J, Rodriguez FH, Kalogeropoulos A, Quyyumi A, Book W. Biomarker Profile in Stable Fontan Patients. *Int J. Cardiology.* 2020 Apr 15; 305: 56-62

Raskind-Hood C, **Saraf A**, Riehle-Colarusso T, Glidewell J, Gurvitz M, Dunn JE, Lui GK, Van Zutphen A, McGarry C, Hogue CJ, Hoffman T, Rodriguez III FH, Book WM. Assessing Pregnancy, Gestational Complications, and Co-morbidities in Women With Congenital Heart Defects (Data From ICD-9-CM Codes in 3 US Surveillance Sites). *Am J Cardiol.* 2020 Mar 1; 125(5): 812-819

Saraf A, Book WM, Nelson TJ, Xu C. Hypoplastic Left Heart Syndrome: From Bench to Bedside and Back. *J Mol Cell Cardiol.* 2019, Oct; 135: 109-118. Review.

More About the UPMC ACHD Program

Decades of advances in pediatric cardiology and cardiothoracic surgery have produced an ever-growing population of adults living long, relatively normal lives with various forms of adult congenital heart disease.

However, the growing population of CHD patients surviving into adulthood requires lifelong, complex cardiac care. Meeting this patient population's needs means expanding services, developing new models of care, and introducing technological advances to lower or remove barriers to care. The UPMC Adult Congenital Heart Disease program is rising to the challenge with a multifaceted approach.

The ACHD program operates on a unified, tri-campus model with UPMC Children's, UPMC Presbyterian, and UPMC Magee-Womens Hospital serving as the hubs for clinical care for the entire spectrum of ACHD patients — younger or older, male or female.

Collaborations with the UPMC Heart and Vascular Institute and cardiology practices across the UPMC system and outreach to regional and community cardiology providers for referral needs and support are hallmarks of the program.

New to the UPMC ACHD program is a 24/7 consultation service for any patients admitted to the hospital for care of another condition or surgery, or for any ACHD patient who arrives through the emergency department for any reason.

Patient Referrals and Additional Information

Referring physicians can contact the UPMC Adult Congenital Heart Disease program at **412-692-3158** or by email at **achd@upmc.edu**.

Heart Institute Welcomes New Director of Cardiac MRI and CT

In August, **Adam B. Christopher, MD**, joined the Heart Institute at UPMC Children's Hospital of Pittsburgh as its new Director of Cardiac MRI and CT. Dr. Christopher also holds an appointment as an assistant professor in the University of Pittsburgh School of Medicine Department of Pediatrics. Dr. Christopher is fellowship-trained in both pediatric cardiology and advanced cardiac imaging. Dr. Christopher earned his medical degree from the University of Pittsburgh School of Medicine, followed by a pediatric residency at Yale-New Haven Children's Hospital.

After residency, Dr. Christopher returned to Pittsburgh to complete his pediatric cardiology fellowship at UPMC Children's Hospital. His fellowship in advanced cardiac imaging was conducted at Children's National Hospital in Washington, DC.

"I knew from my medical school training and fellowship that the Heart Institute at UPMC Children's offers top notch patient care, as well as support for faculty in their clinical practices and research interests. It is an honor to return to Pittsburgh and help lead the growth and expansion of the advanced cardiac imaging program," says Dr. Christopher.

Expanding Cardiac Imaging Services at UPMC Children's: Increasing Access, Improving Quality Care

Dr. Christopher will lead the existing cardiac MRI and CT imaging program and collaborate closely with the Department of Radiology and Heart Institute colleagues to expand capabilities of the imaging program in an effort to increase access for patients and provide clinicians with the diagnostic imaging data they require to optimize patient care.

"Cardiac MRI and CT have become increasingly important to the disciplines of cardiology and cardiothoracic surgery. Echocardiography will always be our first line imaging modality, but there are a growing number of indications for advanced imaging, and the better we are at harnessing the power of these newer technologies and interpreting the findings we obtain, the better our patient outcomes will be. That is the singular goal we all are pursuing: better patient care," says Dr. Christopher.

The program also will be expanding the number of days that cardiac MR and CT scans are conducted to improve accessibility and create shorter wait times for patients, particularly those cases that require sedation or general anesthesia.

New Interventional Cardiac MR Suite In Development

In conjunction with enhancements to the existing cardiac catheterization laboratories led by Bryan Goldstein, MD, the Heart Institute at UPMC Children's is in the planning phases to build a new interventional cardiac MR (ICMR) suite that has a conventional interventional catheterization table with fluoroscopy capabilities at one end of the room, and an MRI magnet on the other side.

This hybrid room will allow simultaneous catheterization procedures and MR procedures for separate patients. Also, catheterization patients and procedures that need cardiac MR before or during the procedure for guidance can simply be transferred to the other end of the room under the same anesthetic.

"The new suite will open up a host of research, clinical care, and quality improvement opportunities for our faculty and patients. We will have the ability to gain MR acquired data on patients during catheterization procedures for research, while at the same time reducing the radiation exposure to patients in certain clinical scenarios," says Dr. Christopher.

The hybrid suite will facilitate fewer sedated or anesthetized procedures for some patients because of the combined abilities and proximity of the interventional space with the MR imaging.

The new MR scanner also brings with it motion detection capabilities that can cancel out a patient's motion during imaging procedures, providing better image quality and data acquisition. Patient motion during imaging procedures is of general concern but magnified when performing imaging on children who have less capability to follow instructions for long periods of time, such as with breath holds during scans.

"Many of our patients are not MR candidates because of pacemakers or other metal objects in the body, so for them, CT will always be necessary. CT does provide higher resolution imaging than cardiac MR, and it will always have a role in our practice. The new CT technology that we will be integrating has better quality, faster image acquisition, and lower radiation exposure levels, which will provide many benefits for our patients. Each patient is unique and the risks and benefits of each imaging modality must be matched with nuanced clinical questions. That is where my expertise comes in to play — assisting our physicians, surgeons, and radiology partners in obtaining the critical diagnostic findings they need and interpreting those findings to better guide clinical decision making," says Dr. Christopher.

Research Interests

Dr. Christopher's imaging research interests lie in the use of anesthesia reduction technologies and motion detection in cardiac imaging procedures. His past work also has involved studying the noninvasive assessment of myocardial disease, including heart transplant rejection through MR imaging. At present, standards of care involve using invasive biopsies to sample myocardial tissues, but proving the efficacy of noninvasive imaging to assess myocardial disease with the same accuracy as tissue biopsy will require much more proof before it can become a trusted method of diagnosis and detection.

"I am also excited to support the many ongoing areas of research within our Heart Institute with cardiac imaging data. The radiation-free assessment of heart muscle size, function, and character offered by cardiac MRI is applicable to countless research questions both within and beyond the Division of Cardiology. I cannot wait to see all that we can learn by expanding this program and leveraging these tools to support the Heart Institute," says Dr. Christopher.

Examining Outcomes After Transcatheter Pulmonary Valve Replacement

In February, pediatric cardiology researchers taking part in the multicenter Congenital Catheterization Collaborative Project on Outcomes-Quality Improvement (C3PO-QI) published findings in the *Journal of the American College of Cardiology* on aspects and outcomes of transcatheter pulmonary valve replacement (TPVR) in a cohort of 530 patients who underwent the procedure between 2014 and 2016. Study data were derived from the prospective C3PO-QI registry.

The objectives of the investigation were to better characterize and define the risks of serious adverse events (SAE), the likelihood of reintervention after TPVR, and quantify the levels of ionizing radiation patients are exposed to during TPVR procedures. While some prior studies have looked at adverse events in TPVR, this study is the first to examine radiation exposure parameters and the incidence of reinterventions.

Bryan H. Goldstein, MD, (above left) director of the cardiac catheterization laboratory (CCL) at UPMC Children's Hospital of Pittsburgh, was the lead author of the study. Among other colleagues participating in the research was **Jacqueline Kreutzer, MD, FACC, FSCAI**, (above right) division chief of pediatric cardiology and co-director of the Heart Institute at UPMC Children's.

Summary of Key Findings

In the last decade, the advent of TPVR has revolutionized pulmonary valve replacement (PVR) for patients who are appropriate candidates for the procedure. TPVR can reduce the number of open-heart procedures a patient is subjected to over a lifetime, and it can increase the time spent with normal pulmonary valve function by lowering the threshold for PVR.

However, like virtually all surgical and nonsurgical interventions, TPVR entails the risk of complications. A more robust knowledge of the types of risks and those most susceptible is at the heart of improving outcomes in TPVR, be it through procedural modifications and advancements or improved patient selection for the procedure. Goldstein et al. add to the TPVR evidence base in three primary areas. *The length and detail of this study are extensive. The full findings of the published paper are available through the reference below.* What follows is a summary of findings related to the main study objectives.

Adverse Events and Serious Adverse Events

TPVR is associated with a relatively high percentage of adverse events. Adverse events in this study were categorized from level 1 to level 5, with level 1 being none and level 5 being catastrophic. In this study cohort, 26% of patients experienced some form of adverse event, while the risk for serious adverse event (SAE; level 3-5) was 13%. Peri-procedural mortality risk in the study cohort was low, at 0.8%, and largely reflective of pre-existing conditions.

While direct comparisons between types of implants were not a part of this study, the findings did point to variability in adverse events depending upon the implant substrate — homograft, bioprosthesis, or native right ventricular outflow tract (RVOT). The risk of a SAE was highest in patients with a homograft conduit RVOT substrate. In the 226 cases where homograft was used, 18% experienced an SAE. Bioprosthesis and native cases experienced SAE at rates of 14% and 15%, respectively. This discrepancy reflected the inclusion of RVOT conduit injury as a SAE.

Radiation Exposure

Given the finite lifespan of pulmonary valve replacements, many patients may have multiple repeat procedures over their lifetime to replace implants that have reached the end of their functional capacity. Furthermore, given that the procedures in use today use x-ray guidance — which exposes patients to significantly more ionizing radiation than would otherwise be normal — understanding radiation dose in TPVR procedures is essential. This is particularly true in younger patients who are still developing physically, and also because these individuals are more likely to need future diagnostic and procedural imaging throughout their lifetime.

Compared to other transcatheter procedures to treat congenital heart anomalies, TPVR procedures account for two times more radiation dose than the next highest-level procedure. In this study, the mean dose of radiation for TPVR was found to be 198 µGy-m2/kg. For comparison, the next highest level for an interventional cardiac procedure is balloon aortic valvuloplasty at 99 µGy-m2/kg.

Risk factors for receiving higher levels of radiation dose during TPVR procedure were shown to be:

- Older age
- Higher right ventricular pressure
- Use of 2 or more pre-TPV stents
- Presence of concomitant transcatheter intervention

Reintervention

Along with radiation exposure, this study is the first to examine the prevalence of reintervention after TPVR.

Analysis of the follow-up cohort of patients (a subset of the total study cohort consisting of complete follow-up individuals (68%) and partial follow-up individuals [84%]) found that 13.3% percent of patients had undergone a reintervention during the median follow-up period of one year from the date of their initial intervention.

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Cardiothoracic Surgery Update:

New Research Shows Efficacy for Cone Procedure After Starnes Procedure




Under the leadership of Victor Morell, MD, the pediatric cardiothoracic surgery program at UPMC Children's Hospital of Pittsburgh continues its pursuit of success and optimal outcomes for congenital heart patients, particularly in its treatment approaches for repairing complex congenital heart defects.

In May, **Jose Pedro Da Silva, MD**, (above, far left) director of the Da Silva Center for Ebstein's Anomaly at UPMC Children's Hospital of Pittsburgh, published new research in the journal *JTCVS Techniques* on the efficacy of the cone procedure after a Starnes procedure for the surgical treatment of Ebstein's anomaly.

Dr. Da Silva is the inventor of the cone procedure to repair defective tricuspid heart valves that, since its perfection in the 1990s, has become a worldwide standard of surgical care for the correction of congenital heart valve defects and Ebstein's anomaly. Since 1993, Dr. Da Silva and colleagues have performed the cone procedure on 208 patients. Since 2013, after Dr. Da Silva and colleagues modernized their overall care protocols, they have successfully performed the cone procedure on 74 non-neonatal patients without mortality.

With colleagues **Melita Viegas, MD**, (above, second from left) **Mario Castro-Medina, MD**, (above, third from left) and **Luciana Da Fonseca Da Silva, MD**, (above, far right) Dr. Da Silva published findings from the first two cases of neonatal patients who had previously undergone a Starnes procedure for surgical palliation of severe Ebstein's anomaly, and who subsequently had surgery using the cone procedure to repair the tricuspid valve of the right ventricle and restore the patient's biventricular physiology. A third successful case has since been performed after while this manuscript was in publication.

In the setting of Ebstein's anomaly, pulmonary atresia and circular shunt typically are managed with the Starnes procedure. The Starnes procedure is an effective surgical intervention for the critical neonate presenting with Ebstein's anomaly. However, after a Starnes procedure, the patient is usually committed to a single ventricle repair pathway. The single ventricle repair pathway subsequently leads to the long-term complications associated with Fontan palliation: ventricular failure, exercise intolerance, arrhythmias, coagulopathy, venous shunts, and others.

Because of these long-term morbidities, Dr. Da Silva and colleagues have searched for and developed a potential solution by demonstrating that right ventricular rehabilitation after the Starnes procedure is feasible. Their new approach of using the cone procedure after the Starnes procedure documented in the study results in a biventricular repair, which is likely to improve a patient's long-term outcomes compared against the outcomes associated with a single ventricle repair strategy.

"We recognize the Starnes procedure as an excellent initial operation for the rescue of critically ill neonates with Ebstein's anomaly. These patients usually present with a dilated right ventricle, which compresses the left ventricle, making its effective performance difficult. The procedure uses a patch with a small hole in its center to exclude the right ventricle. The severe reduction of the right ventricle's blood entrance reduces its size immediately, facilitating more space for the left ventricle, thereby improving its efficacy as a pump. Bearing in mind that the right ventricle undergoes progressive size reduction after the Starnes procedure, we reasoned that, at a point in this shrinkage process, the right ventricle might be a suitable pump for a biventricular repair. Based on this concept, we began to rehabilitate the right ventricle for a biventricular repair using the Da Silva cone technique. Theoretically, this new approach provides substantial long-term benefits versus a single-ventricle pathway," says Dr. Da Silva.

Findings and outcomes from the initial two patients to undergo cone procedure repair after the Starnes procedure show excellent promise for the technique. However, further research by Dr. Da Silva's team will be required to conclusively assess the benefits and long-term outcomes of this approach to treating severe cases of Ebstein's anomaly.

Ebstein's anomaly with a circular shunt is a condition prompted by the combination of pulmonary valve leakage and tricuspid regurgitation, causing continuous retrograde ductal flow and blood circulating over and over inside the heart, which results in low tissue perfusion and a life-threatening condition.

"Effective treatment of fetuses and neonates with Ebstein's anomaly with a sizeable circular shunt is an urgent task that should begin soon after fetal diagnosis. We manage neonates with Ebstein's anomaly who present with a circular shunt according to their clinical condition. Often these patients must undergo the Starnes procedure as the initial surgical intervention. In this situation, it appears possible to follow this approach with a cone repair, ultimately accomplishing a two-ventricle repair," says Dr. Da Silva.

Additional details and videos clips from the study and surgical procedures are available at the journal website and reference below or by visiting https://www.jtcvs.org/article/S2666-2507(20)30232-7/fulltext.

Reference

Da Silva JP; Viegas M; Castro-Medina M; Da Fonseca Da Silva. The Da Silva Cone Operation After the Starnes Procedure for Ebstein's Anomaly: New Surgical Strategy and Initial Results. *JTCVS Techniques.* 2020; 3: 281-283.

More About the Da Silva Center for Ebstein's Anomaly at UPMC Children's

Founded in 2016, the Da Silva Center for Ebstein's Anomaly at UPMC Children's provides world-class care for children and adults who have congenital defects of the heart valves, including Ebstein's anomaly.

Referring physicians may contact the Da Silva Center for Ebstein's Anomaly for a referral or consultation by calling the Heart Institute at UPMC Children's at 412-692-5540.

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Transcatheter Pulmonary Valve Replacement *Continued from Page 6*

Risk factors for reintervention were shown to include smaller patients, individuals with truncus arteriosus or pulmonary atresia and intact ventricular septum, a smaller baseline RVOT diameter, and a higher baseline RV-PA gradient.

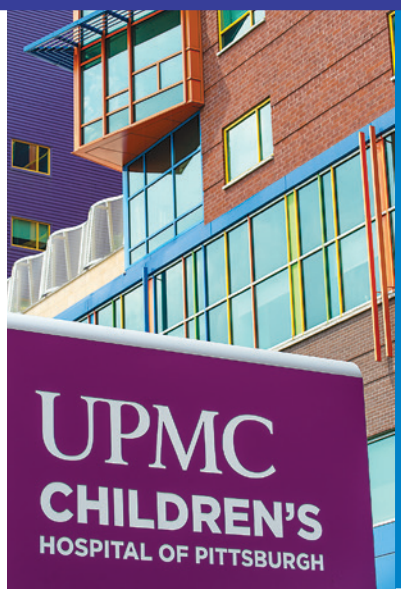
Conclusion

While TPVR has established itself as a formidable, successful, and minimally invasive procedure for the treatment of congenital heart disease with RVOT dysfunction, it is not without significant risks for adverse events, exposure to ionizing radiation, and potential reintervention in the early follow-up period. Fortunately, mortality is an uncommon complication with TPVR. As Dr. Goldstein and colleagues write, future research and procedural advancements should address strategies and modifications to reduce radiation exposure to patients, limit and further study the nature of adverse events within certain patient populations, and address the high level of reinterventions through advances in technique and new implant technology, as well as through optimization of patient selection for TPVR.

Reference

Goldstein BH, et al. Adverse Events, Radiation Exposure, and Reinterventions Following Transcatheter Pulmonary Valve Replacement. *J Am Coll Cardiol.* 2020; 75: 363-376.

Affiliated with the University of Pittsburgh School of Medicine and ranked among the nation's best children's hospitals by *U.S. News & World Report.*



About UPMC Children's Hospital of Pittsburgh

Regionally, nationally, and globally, UPMC Children's Hospital of Pittsburgh is a leader in the treatment of childhood conditions and diseases, a pioneer in the development of new and improved therapies, and a top educator of the next generation of pediatricians and pediatric subspecialists. With generous community support, UPMC Children's Hospital has fulfilled this mission since its founding in 1890. UPMC Children's is recognized consistently for its clinical, research, educational, and advocacy-related accomplishments, including ranking 15th among children's hospitals and schools of medicine in funding for pediatric research provided by the National Institutes of Health (FY2019) and ranking on *U.S. News & World Report's* Honor Roll of America's Best Children's Hospitals (2020–21).