

CONGENITAL CARDIOLOGY TODAY

Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

August 2017; Volume 15; Issue 8
International Edition

IN THIS ISSUES

Rare Case of a Left Superior Vena Cava Connection to the Left Upper Pulmonary Vein

By Sinéad L. Murphy; Kenneth Fang, MD; Tabitha Moe, MD
~Page 1

Medical News, Products & Information

~Page 7

MEDICAL MEETINGS

CSI at UCSF - Catheter Interventions in Structural, Valvular & Congenital Heart Disease, Atrial Fibrillation & Heart Failure

Sep. 8-9, 2017; San Francisco, CA USA
www.csi-congress.org

27th International ACHD Advanced Symposium, 2017

Sep. 14-16, 2017; Cincinnati, OH USA
cincinnatichildrens.org/service/a/congenital-heart/achd-annual-symposium

50th Anniversary Southeast Pediatric Cardiovascular Society Conference

Sep. 28-30, 2017; Atlanta, GA USA
choa.org/medical-professionals/professional-events/50th-anniversary-southeast-pediatric-cardiovascular-society-meeting

11th IPC Workshop

Sep. 28-30, 2017; Milan, Italy
www.workshopipc.com/main.php

33rd Annual Echocardiography in Pediatric and Adult Congenital Heart Disease Symposium

Oct. 8-11, 2017; Rochester, MN USA
cveducation.mayo.edu/marketing/echocardiography-in-pediatric-and-adult-congenital-heart-disease-case-studies--2#overview

CONGENITAL CARDIOLOGY TODAY

Editorial & Subscription Offices
16 Cove Road
Westerly, RI 02891

www.CongenitalCardiologyToday.com

Twitter: www.Twitter.com/ccardiology

Official publication of the CHIP Network

Rare Case of a Left Superior Vena Cava Connection to Left Upper Pulmonary Vein

By Sinéad L. Murphy; Kenneth Fang, MD; Tabitha Moe, MD

Keywords: Left superior vena cava, Congenital Heart Disease (CHD), Anomalous left upper pulmonary venous return, Thromboembolic stroke

Abstract

A persistent Left Superior Vena Cava (LSVC) is a relatively common, quiescent congenital anomaly in the general population, often with drainage into the coronary sinus and return to the right atrium. We present a rare case of persistent LSVC with connection to the Left Upper Pulmonary Vein (LUPV), creating potential for right-to-left shunting, in the absence of other congenital anatomical anomalies. This connection can result in cyanosis, increased risk for endocarditis and, as seen in this patient, paradoxical thromboembolism. A discussion of morphologic forms, diagnostic imaging, clinical significance, and potential corrective procedures is reviewed.

Case Report

We present the case of a 64-year-old female who initially presented to care in the Bahamas for an acute middle cerebral artery stroke, thought to be thromboembolic in nature. TPA was administered, and she was transferred to Arizona for management and further workup. In the search for thromboembolic origin, a transthoracic echocardiogram was performed, showing severe mitral regurgitation, mitral valve

“A persistent Left Superior Vena Cava (LSVC) is a relatively common, quiescent congenital anomaly in the general population, often with drainage into the coronary sinus and return to the right atrium. We present a rare case of persistent LSVC with connection to the Left Upper Pulmonary Vein (LUPV), creating potential for right-to-left shunting, in the absence of other congenital anatomical anomalies. This connection can result in cyanosis, increased risk for endocarditis and, as seen in this patient, paradoxical thromboembolism.”

prolapse and severe left atrial dilation. No

CONGENITAL CARDIOLOGY TODAY

CALL FOR CASES AND OTHER ORIGINAL ARTICLES

Do you have interesting research results, observations, human interest stories, reports of meetings, etc. to share? Submit your manuscript to: RichardK@CCT.bz

signs of shunting were observed on echocardiogram. With the working diagnosis of undetected atrial fibrillation, the patient was scheduled for operative replacement of the mitral valve.

Pre-operatively, a left internal jugular central venous catheter was placed for hemodynamic management during surgery. Initially, the catheter transduction demonstrated venous waveforms as anticipated, but subsequently changed to demonstrate an arterial pattern. The patient was hemodynamically stable, and the surgical team chose to proceed as planned. Careful dissection showed a persistent LSVC which drained into the LUPV which then returned normally to the left-sided atrium (Figure 1). The other pulmonary veins were identified and found to return to the left atrium. The right-sided superior vena cava was intact and patent. The central venous catheter was withdrawn into the venous system, and the anomalous systemic-pulmonary venous connection was ligated.

Post-operatively, the patient developed advanced atrio-ventricular block and profound bradycardia, which precipitated torsades de pointes on three occasions. As a result, she underwent transvenous placement via right subclavicular approach of a permanent pacemaker without further incident. She was discharged to inpatient rehabilitation with plans to return to the Bahamas.

Discussion

A persistent Left Superior Vena Cava (LSVC) is a relatively common, incidentally found congenital anomaly in the general population, with estimated prevalence of from 0.3% to 2%.¹ In patients with other forms of CHD the prevalence increases to 4.4%, and has been reported in association with heterotaxy syndromes, primum-type Atrial Septal Defects, clefing of the left-sided atrioventricular valve, atrioventricular canal defects, and coarctation of the aorta.² Most patients are asymptomatic, with 80%-90% of reported anomalies draining into the coronary sinus and right atrium.¹

The condition presents more significant risk and sequelae, rarely, when the LSVC connects to a left heart structure, creating the potential for a shunting defect. This can occur with persistent LSVC drainage through the left atrium via a direct connection to the left atrium, or through the Left Upper Pulmonary Vein (LUPV). These types of anomalies are

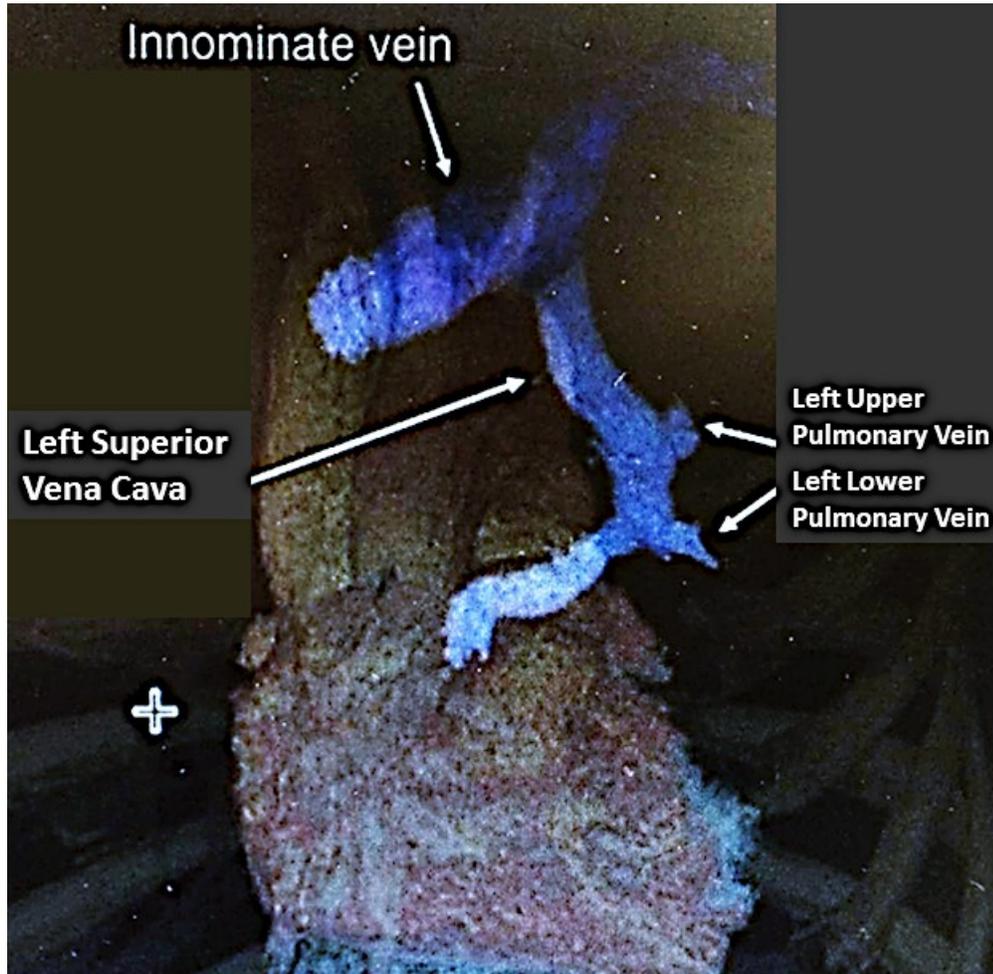


Figure 1. Angiography of left superior vena cava to left upper pulmonary vein connection. Informed consent: Informed consent was obtained from the individual participant included in the study.

associated with an increased risk of cyanosis, heart failure, intracerebral abscess,^{3,4} and embolic cerebrovascular events.⁵ Reports of these anomalies often follow embolic stroke work up, as seen in this patient.

The case we present is a very rare morph of persistent LSVC to LUPV connection with only a half dozen other cases reported in the literature to our knowledge.^{3, 5-8} Of these reported cases, only three have been in the absence of other congenital cardiac anomalies. Diagnostic modalities previously reported include chest radiograph,⁴ angiography⁴ and digital subtraction angiography,⁵ contrast enhanced computer tomography scan,⁵ transesophageal echocardiography with microbubbles,⁷ and open dissection. With the current imaging

modalities, the authors believe cardiac magnetic resonance imaging would also be a viable method of diagnosis. Intervention is not absolutely indicated, with medical management to avoid thromboembolic events a reasonable alternative, but necessary due to right-to-left shunting potential. Reported methods of closure include: surgical diversion of the LSVC to the right atrium via baffle in the case of absent right SVC,⁴ occlusion of the LSVC with the Amplatzer Vascular Plug,⁷ and surgical ligation of the LSVC. Post-operative side effects have included Superior Vena Cava Syndrome,⁴ and arrhythmia.

References

1. Tak T, Crouch E, Drake GB. Persistent left superior vena cava: incidence,

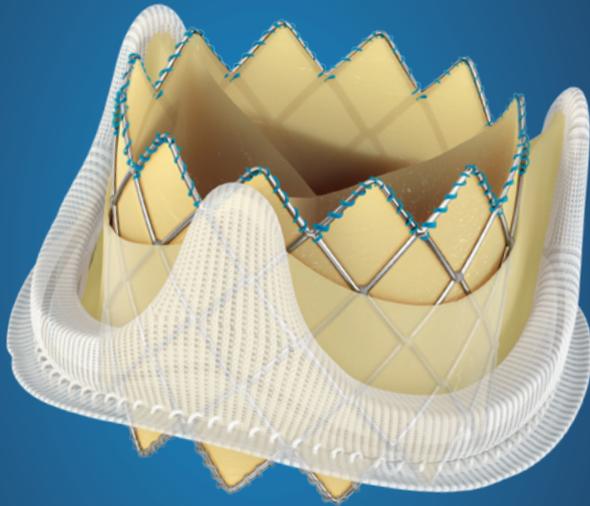
CONGENITAL CARDIOLOGY TODAY

CALL FOR CASES AND OTHER ORIGINAL ARTICLES

Do you have interesting research results, observations, human interest stories, reports of meetings, etc. to share? Submit your manuscript to: RichardK@CCT.bz

CE MARK RECEIVED

FOR USE IN
FAILED SURGICAL
BIOPROSTHETIC
PULMONARY
VALVES



Melody™

Transcatheter Pulmonary
Valve (TPV) Therapy

Reaching even
more patients
with Melody™ TPV

- The first commercially available TPV
- A breakthrough non-surgical option to treat failing pulmonary valve conduits
- Has treated more than 11,000 patients globally over the last 10 years

Melody-TPV.com

Medtronic

Further. Together

Melody™ Transcatheter Pulmonary Valve, Ensemble™ II Transcatheter Valve Delivery System

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.

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

LifeLine
CardioVascular Technical Support
Tel: (877) 526-7890
Tel: (763) 526-7890
Fax: (763) 526-7888
rs.cstechsupport@medtronic.com

Medtronic, Medtronic logo and Further, Together are trademarks of Medtronic. All other brands are trademarks of a Medtronic company.

Medtronic

Watch over 300 Live Case Videos, Presentations and Workshops Online from Leading Congenital and Structural Medical Meetings from Around the World

www.CHDLiveCases.com



- Transseptal Access Workshop from Cook Medical
- Workshop: Past Present and Future of Pediatric Interventions Cardiology - St. Jude & AGA Medical
- Symposium on Prevention of Stroke Clinical Trials at the Heart of the Matter - WL Gore Medical
- Imaging in Congenital & Structural Cardiovascular Interventional Therapies
- Morphology of The Atrial Septum
- Morphology of The Ventricular Septum
- Pre-Selection of Patients of Pulmonic Valve Implantation and Post-Procedural Follow-up
- Echo Paravalvular Leakage (PVL)
- ICE vs TEE ASD Closure in Children - PRO & CON ICE
- 3D Rotational Angiography - Why Every Cath Lab Should Have This Modality
- PICS Doorway to the Past - Gateway to the Future
- Follow-up From PICS Live Cases 2010 Presentation
- Intended Intervention - Transcatheter TV Implantation - *Live Case*
- Intended Intervention - LAA Closure Using Amplatzer Cardiac Plug Under GA & Real Time 3D
- Provided Intervention - LPA Stenting / Implantation of a Sapien Valve
- Intended Intervention - PV Implantation
- Intended Intervention - COA Stent Using Atrium Advanta V12 Covered Stent - *Live Case*
- Intended Intervention - ASD Closure - *Live Case*
- Intended Intervention - Transcatheter VSD Device Closure - *Live Case*
- Intended Intervention - COA Stenting Using Premounted Advanta V12 Covered Stent - *Live Case*
- Stunning Revelation - The Medical System is Changing - What Can You Do To Show Patients That Your Practice Does It Right? Patient Perspective
- Percutaneous Paravalvular Leak Closure Outcomes
- Intensive Management of Critically Ill Infants Undergoing Catheterization
- **and many more....**

Presented by **CONGENITAL CARDIOLOGY TODAY**



27th International Symposium on Adult Congenital Heart Disease

Sep. 14-16, 2017 | Cincinnati, Ohio USA



www.cincinnatichildrens.org/ACHDsymposium

“The case we present is a very rare morph of persistent L SVC to LUPV connection with only a half dozen other cases reported in the literature to our knowledge.^{3, 5-8} Of these reported cases, only three have been in the absence of other congenital cardiac anomalies.”

- significance and clinical correlates. *Int J Cardiol* 2002;82:91-93.
- Fraser RS, Dvorkin J, Rossall RE, Eidem R. Left superior vena cava: a review of associated congenital heart lesions, catheterization data and roentgenologic findings. *Am J Med* 1961;31:711-716.
 - Erol I, Cetin, II, Alehan F, Varan B, Ozkan S, Agildere AM, et al. Brain abscess associated with isolated left superior vena cava draining into the left atrium in the absence of coronary sinus and atrial septal defect. *Cardiovasc Intervent Radiol* 2006;29:454-456.
 - Looyenga DS, Lacina SJ, Gebuhr CJ, Stockinger FS. Persistent left superior vena cava communicating with the left atrium through a systemic-pulmonary venous malformation. *J Am Coll Cardiol* 1986;8:621-626.
 - Yousaf M, Malak SF. Left Atrial Drainage of a Persistent Left Superior Vena Cava. *Radiol Case Rep* 2008;3:225.
 - Odman P. A persistent left superior vena cava communicating with the left atrium and pulmonary vein. *Acta radiol* 1953;40:554-560.
 - Recto MR, Sadlo H, Sobczyk WL. Rare case of persistent left superior vena cava to left upper pulmonary vein: pathway for paradoxical embolization and development of transient ischemic attack and subsequent occlusion with an amplatzer vascular plug. *J Invasive Cardiol* 2007;19:E313-316.

- De Geest B, Vandommele J, Herregods MC, Vanhaecke J, Gewillig M, Daenen W, et al. Isolated left sided superior vena cava draining into the left atrium associated with recurring intracerebral abscesses. A case report. *Acta Cardiol* 1994;49:175-182.

CCT

Corresponding Author



Sinéad L. Murphy
 Mayo Clinic College of Medicine
 200 1st St. SW
 Rochester, MN 55902
 Murphy.Sinead@mayo.edu



H. Kenith Fang, MD
 Phoenix Cardiac Surgery
 Phoenix, AZ, USA



Tabitha Moe, MD
 Adult Congenital Cardiology
 Phoenix Children's Hospital
 Phoenix, AZ, USA

CONGENITAL CARDIOLOGY TODAY

CALL FOR CASES AND OTHER ORIGINAL ARTICLES

Do you have interesting research results, observations, human interest stories, reports of meetings, etc. to share?

Submit your manuscript to:
 RichardK@CCT.bz

- Title page should contain a brief title and full names of all authors, their professional degrees, and their institutional affiliations. The principal author should be identified as the first author. Contact information for the principal author including phone number, fax number, email address, and mailing address should be included.
- Optionally, a picture of the author(s) may be submitted.
- No abstract should be submitted.
- The main text of the article should be written in informal style using correct English. The final manuscript may be between 400-4,000 words, and contain pictures, graphs, charts and tables. Accepted manuscripts will be published within 1-3 months of receipt. Abbreviations which are commonplace in pediatric cardiology or in the lay literature may be used.
- Comprehensive references are not required. We recommend that you provide only the most important and relevant references using the standard format.
- Figures should be submitted separately as individual separate electronic files. Numbered figure captions should be included in the main Word file after the references. Captions should be brief.
- Only articles that have not been published previously will be considered for publication.
- Published articles become the property of the *Congenital Cardiology Today* and may not be published, copied or reproduced elsewhere without permission from *Congenital Cardiology Today*.
- Please be sure any patient information such as name is removed from all figures.



**International Workshop
 on Interventional Pediatric
 and Adult Congenital Cardiology**

**September
 28th-30th 2017**

Crowne Plaza Linate



KINGSTON

FREELANCE CONSULTING

WE OFFER...

- Business Development
- Clinical Planning & Research
- Clinical Study Development
- Concept Development
- Coordination of Testing
- Design & Development
- Design Requirements
- Expert Assistance
- FDA Liaison
- Feasibility Evaluation
- Functional & Performance Studies
- Internal Compliance Audits
- Manage All Activities Related to Regulatory Approval & Compliance.
- Market Assessment (Global)
- Pre-Submission Meetings
- Post-Market Surveillance
- Project Management
- Risk Analysis & Management
- Risk-Benefit Analysis
- Report Writing
- Strategic Project Planning
- Supplier Audits

ARE YOU:

- + **LEADING AN EARLY STAGE PRIVATELY HELD HEALTHCARE ORGANIZATION?**
- + **A LARGE MULTINATIONAL ORGANIZATION?**

If you answered “Yes,” then you need a partner who understands your needs, and can effectively navigate and manage the challenges and risks associated with leading the clinical research priorities.



❖ AMBER KINGSTON ❖

OWNER

2305 WOODMONT CIRCLE

STE. K

MACUNGIE, PA 18062

TEL: 610.463.4964

AKKINGSTON@ICLOUD.COM

Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

Children's Hospital of Philadelphia and Mount Sinai Health System Mark Milestone in Fetal Medicine and Children's Heart Programs

Newswise — Children's Hospital of Philadelphia (CHOP) and the Mount Sinai Health System took another step forward at the end of May in their 18-month-old alliance with the official opening of their Fetal Medicine Program and the affiliation of the Mount Sinai Children's Heart Center with the Cardiac Center at CHOP.

Officials from both hospitals participated in a ribbon-cutting ceremony held today at the new facility at The Mount Sinai Hospital's Annenberg Building.

The Fetal Medicine Program will offer access to an unprecedented scope of services. This program provides mothers carrying fetuses at risk for or identified with possible anomalies a "one-stop experience" that includes a comprehensive diagnostic evaluation and consultation. Specifically, in a single-day visit, the patient will undergo state-of-the-art diagnostic testing, which may include ultrasound, echocardiography, and fetal magnetic resonance imaging (MRI), and then meet with a team of Mount Sinai and CHOP experts to discuss the presumptive diagnosis and options for treatment. All diagnostic testing will be performed at The Mount Sinai Hospital; images will be read by specialists in fetal radiology at CHOP and Mount Sinai in coordination with Mount Sinai Maternal Fetal Medicine Specialists using telemedicine video links. Once the diagnostics are reviewed, and depending on the presumptive diagnosis, a conference with each patient and her family may include a maternal fetal medicine specialist, pediatric cardiologist, pediatric surgeon, geneticist, and other relevant pediatric subspecialists. CHOP subspecialists will participate in these family meetings by video conferencing. The program is the only one in New York City offering such convenience and level of services.

The new affiliation of the Mount Sinai Children's Heart Center and the CHOP Cardiac Center brings to New York access to unparalleled expertise and resources from one of the nation's leading pediatric cardiac centers. The affiliation includes the Fetal Heart Program, which aims to diagnose Congenital Heart Disease (CHD) as early as 12-14 weeks gestation utilizing fetal echocardiography. The results of the fetal imaging can then allow the clinical care team, comprised of providers at Mount Sinai and CHOP collaborating via telemedicine technology, to develop an optimal plan for care. The Children's Heart Center at Mount Sinai Hospital—overseen by the Divisions of Pediatric Cardiology and Cardiac Surgery—provides a continuum of care from fetal through adult life, and will now have access to the expertise of CHOP's Cardiac Center in areas such as pediatric cardiac intensive care and pediatric cardiothoracic surgery. The Children's Heart Center offers exceptional pediatric cardiology and cardiac surgical services, including echocardiography, exercise testing, Holter monitoring, interventional cardiology and angiography, and

cardiovascular genetics. The Mount Sinai and CHOP teams can provide joint expertise at all points of treatment through direct consultation and use of telehealth technology.

The alliance between Mount Sinai and Children's Hospital of Philadelphia was announced in the fall of 2015, and includes three services: Fetal Medicine, Pediatric Cardiac Care, and Pediatric Oncology. This collaboration is intended to give patients and their families access to the most advanced diagnostics and treatments delivered by Mount Sinai and CHOP experts close to home at select Mount Sinai locations.

"Mount Sinai is pleased to collaborate with CHOP, a world-renowned institution, to ensure excellent patient care," said Kenneth L. Davis, MD, President and Chief Executive Officer of the Mount Sinai Health System. "Our goal is to offer the highest quality maternal, fetal, and pediatric care to patients—especially those with complex needs—across a large health system and a fast-growing ambulatory care network. And together, we will be uniquely positioned to recruit and retain the best faculty in the region."

"CHOP is pleased to mark this milestone in the relationship between two health care institutions that are totally dedicated to caring for the health of children and their families," said Madeline Bell, President and Chief Executive Officer of Children's Hospital of Philadelphia. "The alliance with the Mount Sinai Health System continues to evolve and reflects both a shared vision to better serve families in the New York region and a mutual professional respect and admiration between our organizations," Bell said.

The Mount Sinai Health System is an integrated health system committed to providing distinguished care, conducting transformative research, and advancing biomedical education. Structured around seven hospital campuses and a single medical school, the Health System has an extensive ambulatory network and a range of inpatient and outpatient services—from community-based facilities to tertiary and quaternary care.

The System includes approximately 7,100 primary and specialty care physicians; 12 joint-venture ambulatory surgery centers; more than 140 ambulatory practices throughout the five boroughs of New York City, Westchester, Long Island, and Florida; and 31 affiliated community health centers. Physicians are affiliated with the renowned Icahn School of Medicine at Mount Sinai, which is ranked among the highest in the nation in National Institutes of Health funding per investigator. The Mount Sinai Hospital is in the "Honor Roll" of best hospitals in America, ranked No. 15 nationally in the 2016-2017 "Best Hospitals" issue of *U.S. News & World Report*. The Mount Sinai Hospital is also ranked as one of the nation's top 20 hospitals in Geriatrics, Gastroenterology/GI Surgery, Cardiology/Heart Surgery, Diabetes/Endocrinology, Nephrology, Neurology/Neurosurgery, and Ear, Nose & Throat, and is in the top 50 in four other specialties. New York Eye and Ear Infirmary of Mount Sinai is ranked No. 10 nationally for Ophthalmology, while Mount Sinai Beth Israel, Mount Sinai St.

Master Class in Congenital Cardiac Morphology

With world renowned cardiac pathologist Professor Robert Anderson, MD, FRCPath

Oct. 11 to 13, 2017 • Children's Hospital of Pittsburgh of UPMC • Pittsburgh, PA

This activity has been approved for AMA PRA Category 1 Credit™ • The University of Pittsburgh is an affirmative action, equal opportunity institution.

Learn more at www.chp.edu/MasterClassCCM



Children's
Hospital of Pittsburgh
of UPMC

Luke's, and Mount Sinai West are ranked regionally. Mount Sinai's Kravis Children's Hospital is ranked in seven out of ten pediatric specialties by *U.S. News & World Report* in "Best Children's Hospitals." For more information, <http://www.mountsinai.org>.

The Children's Hospital of Philadelphia was founded in 1855 as the nation's first pediatric hospital. Through its long-standing commitment to providing exceptional patient care, training new generations of pediatric healthcare professionals and pioneering major research initiatives, Children's Hospital has fostered many discoveries that have benefited children worldwide. For more information, visit www.chop.edu.

All Heart Patients Have Some Liver Disease After Fontan Surgery - CHOP Researchers Report Universal Liver Fibrosis in Survivors of Operation for a Severe Heart Defect

Newswise — Patients who undergo the Fontan operation as children for a Complex Congenital Heart Defect are at risk of developing progressive liver fibrosis, a buildup of fibrous deposits, as a result of the circulation created by the surgery, according to a new study. A research team says their findings underscore the importance of improving ongoing medical surveillance, so that physicians can develop the most appropriate care for their patients.

The Fontan operation is a series of three-staged reconstructive surgeries in children born with single-ventricle disease, a life-threatening condition characterized by a severely underdeveloped ventricle, one of the heart's two pumping chambers. The palliative surgeries re-route blood to the lungs, but result in an abnormal physiology that puts many organ systems at risk. One such organ is the liver, in which fibrosis develops in response to elevated pressure within the veins. Over time this can lead to liver cirrhosis, a condition of significant liver impairment.

"We have known for some time that liver fibrosis is a complication of Fontan surgery, but this was an important study that demonstrated that the length of time after the Fontan operation is a significant contributor to the degree of fibrosis," said study leader David J. Goldberg, MD, a pediatric cardiologist at Children's Hospital of Philadelphia (CHOP).

Goldberg and colleagues published their study online April 26th in the *Journal of the American Heart Association* (<https://doi.org/10.1161/JAHA.116.004809>).

The research is a retrospective analysis of 67 children and adolescents (with a mean age of 17 years) evaluated at CHOP between 2009 and 2014 under the hospital's Single Ventricle Survivorship Program. Most of these patients were evaluated 10 to 15 years after their original Fontan operation.

Most of the patients had no overt symptoms of liver disease, which progresses very gradually, and were generally considered to be in good health. All 67 patients showed evidence of some liver fibrosis, measured by the quantity of collagen deposits found after needle biopsies of the patients' livers. The only risk factor the researchers found was time from Fontan—the degree of fibrosis increases over

time. The researchers added that there are likely other risk factors for fibrosis not measured in their analysis.

"Our finding that liver fibrosis is universal after the Fontan operation reinforces the need for ongoing surveillance of the liver in this patient population," said Goldberg. "However, it is important to find a less invasive method than liver biopsy to measure liver fibrosis, as well as to investigate possible medications that may reduce the rate of fibrosis. As we work to develop clinical guidelines for liver surveillance, we can better provide care tailored to our individual patients."

Children's Hospital of Philadelphia: Children's Hospital of Philadelphia was founded in 1855 as the nation's first pediatric hospital. For more information, visit www.chop.edu

Edwards SAPIEN 3 Valve Receives FDA Approval For Aortic, Mitral Valve-In-Valve Procedures

PRNewswire - Edwards Lifesciences Corporation, a global leader in patient-focused innovations for structural heart disease and critical care monitoring, announced June 5th it has received U.S. Food and Drug Administration (FDA) approval for aortic and mitral valve-in-valve procedures using the Edwards SAPIEN 3 transcatheter heart valve. The SAPIEN 3 valve is the first transcatheter heart valve approved in the U.S. for the treatment of both aortic and mitral patients who are at high risk for a subsequent open-heart surgery to replace their bioprosthetic valve.

"Expansion of the SAPIEN 3 device indication to include valve-in-valve procedures is a meaningful advancement for patients at high risk of an additional open-heart valve procedure, particularly for those in need of a safe alternative for mitral valve replacement," said Larry L. Wood, Edwards' Corporate Vice President, transcatheter heart valves.

This anticipated FDA approval of the indication expansion was supported by real-world data collected from the Society of Thoracic Surgeons and American College of Cardiology (STS/ACC) Transcatheter Valve Therapy (TVT) Registry. The TVT Registry includes information and outcomes on patients undergoing transcatheter valve replacement and repair procedures in the United States

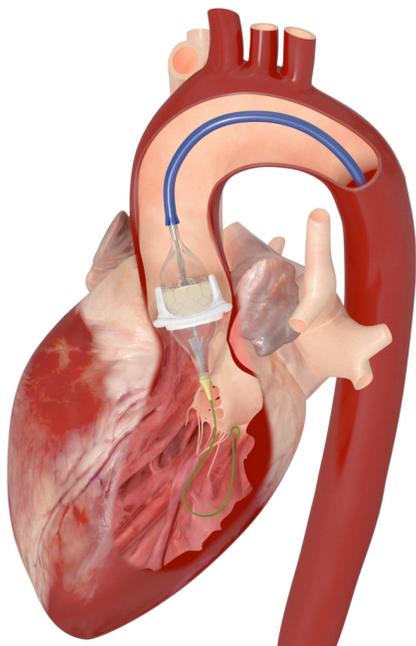
"This approval brings a safe and effective transcatheter therapy to patients who would do very poorly with repeat open-heart surgery," said John Carroll, MD, Professor of Cardiology at the University of Colorado School of Medicine and Director of Interventional Cardiology at the University of Colorado Hospital, Denver and member of the TVT Registry Steering Committee. "I am pleased to see that the FDA recognizes the value of the high-quality evidence generated by the STS-ACC TVT Registry and its ability to play an important role in assessing 'real-world' clinical results in specialty indications, such as valve-in-valve, and for particular patient groups, such as those needing replacement of a bioprosthetic mitral valve."

The Edwards SAPIEN 3 valve was approved by the FDA in 2015 for severe, symptomatic aortic stenosis patients at high risk for open-heart surgery, and, in 2016, received approval for the treatment of patients



Archiving Working Group
International Society for Nomenclature of
Paediatric and Congenital Heart Disease
ipccc-awg.net

who are at intermediate risk for open-heart surgery. The SAPIEN 3 valve builds on



Credit Edwards Lifescience

Top: (Anatomical image) An image of the deployed SAPIEN 3 transcatheter heart valve inside a surgical valve

Bottom: The SAPIEN 3 transcatheter heart valve is now approved for aortic and mitral valve-in-valve procedures in patients at high risk of a subsequent surgery.

Edwards' decades of experience in the development of tissue heart valves, and the proven benefits of the Edwards SAPIEN valves.

For more information on Edwards Lifesciences Corporation, visit www.Edwards.com.

CONGENITAL CARDIOLOGY TODAY

We Can Help You Recruit:*

- Pediatric Cardiologists
- Pediatric Interventional Cardiologist
- Adult Cardiologist
- focused on CHD
- Congenital/ Structural Heart
- Surgeons
- Echocardiographers,
- EPs
- Pediatric Transplant
- Cardiologist



Your Recruitment Advertising Includes:

- Full color recruitment ad in the issue(s)
- Your recruitment listing in the email blast for the issue(s) with a hot link
- 3-Step Special Recruitment Opportunity Website Section in three (3) areas of the website
- *We can create your recruitment ad at no extra charge!*

For more Information Contact:

Tony Carlson

+1.301.279.2005 or

tcarlsonmd@gmail.com

CONGENITAL CARDIOLOGY TODAY

© 2017 by Congenital Cardiology Today (ISSN 1554-7787-print; ISSN 1554-0499-online). *Published monthly. All rights reserved.*

www.CongenitalCardiologyToday.com

Publication Company Address:

11502 Elk Horn Dr. Ste. 201
Clarksburg, MD 20871 USA
Tel: +1.301.279.2005

Publishing Management:

- Tony Carlson, Founder, President & Sr. Editor - TCarlsonmd@gmail.com
- Richard Koulbanis, Group Publisher & Editor-in-Chief - RichardK@CCT.bz
- John W. Moore, MD, MPH, Group Medical Editor - JMoore@RCHSD.org
- Allan Berthe, Contributing Editor-Special Projects

Editorial Board:

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

Free Subscription to Qualified

Professionals: Send your name, title(s), hospital or practice name, work address and url, phone, fax and email to: sub@cct.bz.

Official publication of the CHIP Network

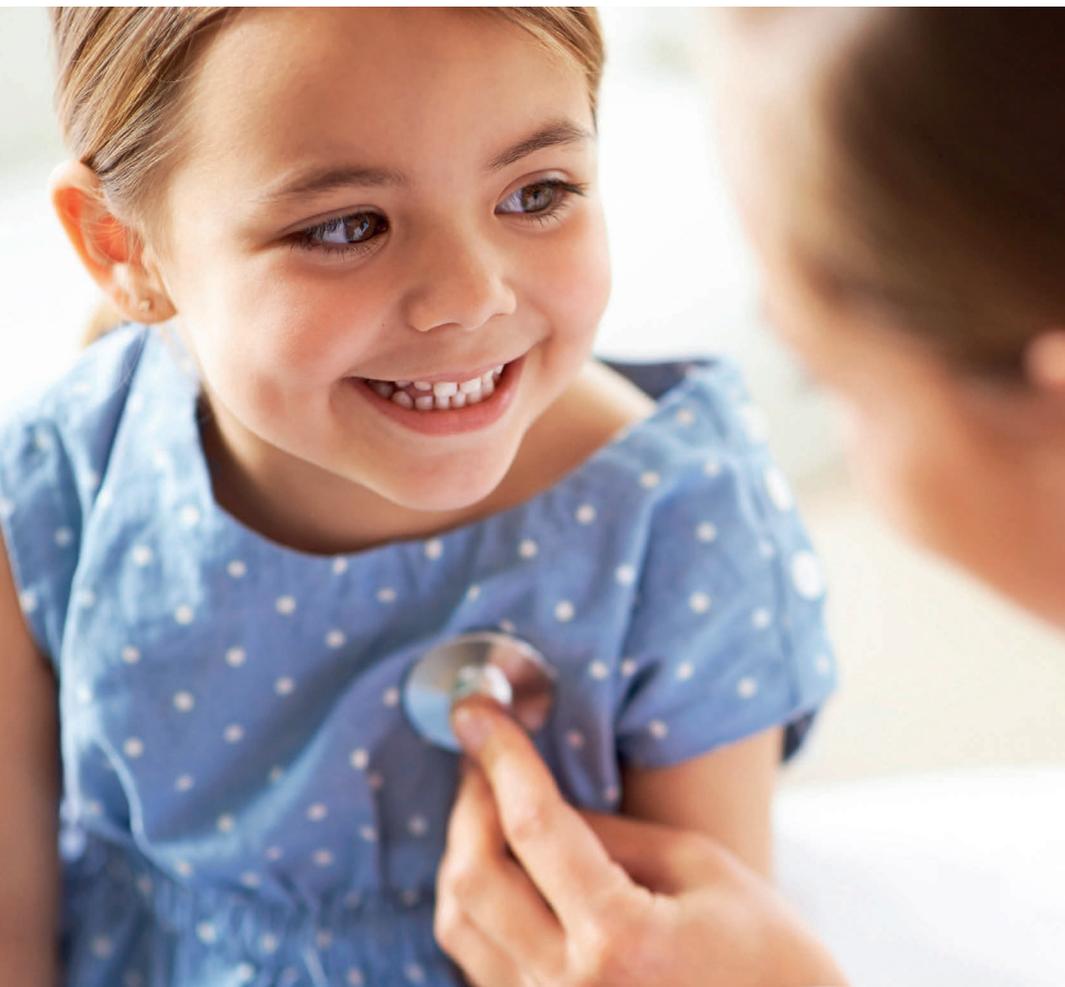
Statements or opinions expressed in Congenital Cardiology Today reflect the views of the authors and sponsors, and are not necessarily the views of Congenital Cardiology Today.



PICS-AICS
Pediatric and Adult Interventional Cardiac Symposium

VEGAS
MGM GRAND LAS VEGAS
SEPTEMBER 5-8, 2018

www.picsymposium.com



Repositionable and Retrievable
 Prior to Release

Tight and Compact Windings
 Ensure Efficient Occlusion

Designed to Match Individual
 Morphologies and Sizes

NIT-OCCLUD[®] Coil System

for PDA Closure

Designed For the Safe and Atraumatic Occlusion of the
 Congenital Heart Defect PDA (Patent Ductus Arteriosus)

INDICATIONS FOR USE:

The Nit-Occlud[®] PDA coil is a permanently implanted prosthesis indicated for percutaneous, transcatheter closure of small to moderate size patent ductus arteriosus with a minimum angiographic diameter less than 4mm.

NIT-OCCLUD BRIEF STATEMENT:

Do not implant the Nit-Occlud PDA into patients who have endocarditis, endarteritis, active infection, pulmonary hypertension (calculated PVR greater than 5 Wood Units), thrombus in a blood vessel through which access to the PDA must be obtained, thrombus in the vicinity of the implantation site at the time of the implantation or patients with a body weight < 11 lbs. (5kg). An angiogram must be performed prior to implantation for measuring length and diameter of the PDA. Only the pfm medical implantation delivery catheter should be used to implant the device. Administration of 50 units of heparin per kg bodyweight should be injected after femoral sheaths are placed. Antibiotics should be given before (1 dose) and after implantation (2 doses) to prevent infection during the implant procedure. Do not implant the Nit-Occlud PDA in an MR environment. Do not pull the Nit-Occlud coil through heart valves or ventricular chambers. Contrast media should not be injected through the implantation catheter. The catheter must not be connected to high pressure injectors. Patients may have an allergic response to this device due to small amounts of nickel that has been shown to be released from the device in very small amounts. If the patient experiences allergic symptoms, such as difficulty in breathing or swelling of the face or throat, he/she should be instructed to seek medical assistance immediately. Antibiotic prophylaxis should be performed to prevent infective endocarditis during first 6 months after coil implantation. Potential Adverse Events: Air embolism, Allergic reaction to drug/contrast, Apnea, Arrhythmia requiring medical treatment or pacing, Arteriovenous fistula, Bacterial endocarditis, Blood loss requiring transfusion, Chest pain, Damage to the tricuspid or pulmonary valves, Death, Embolization of the occluder, requiring percutaneous or surgical intervention, Endarteritis, False aneurysm of the femoral artery, Fever, Headache/ Migraine, Heart failure, Hemolysis after implantation of the occluder, Hypertension, Hypotension or shock, Infection, Myocardial infarction, Occluder fracture or damage, Perforation of the heart or blood vessels, Stenosis of the left pulmonary artery or descending thoracic aorta, Stroke/TIA, Thromboembolism (cerebral or pulmonary), Valvular Regurgitation, Vessel damage at the site of groin puncture (loss of pulse, hematoma etc.).

Nit-Occlud is a registered trademark of pfm medical Inc.

Rx only CV9064 - 5/17 ©2017 B. Braun Interventional Systems Inc.

Distributed by:

B. Braun Interventional Systems Inc.

824 Twelfth Avenue | Bethlehem, PA 18018 | USA

Tel 877 836 2228 | Fax 610 849 1334 | www.bisusa.org

