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# Surgical Strategy for Thrombosed Unilateral Absence of a Pulmonary Artery


*By Mark Cartoski, MD; Jessica M. Parson, BS; Alexander Ellis, MD; Elliot Tucker, MD; Felix W. Tsai, MD*


## Introduction

Unilateral absence of a pulmonary artery (UAPA), first described by Frantzel in 1868, is a rare cardiac anomaly thought to develop from the persistence of the hilar components of the pulmonary artery to the distal sixth aortic arch with concurrent involution of the proximal portion of the same arch.[1,2] It is a rare defect with an estimated prevalence in adult males of 1 in 200,000.[1] It is often associated with other cardiovascular anomalies such as Tetralogy of Fallot, pulmonary atresia, and Ventricular Septal Defect (VSD); however, UAPA may occur as an isolated finding.[2,3] Left-sided defects, while UAPA is more often diagnosed with other cardiac defects while right-sided UAPA is more likely to be seen as a solitary finding.[4] UAPA treatment strategies have mostly been limited to older presentations from single institution case reports; none have discussed neonatal presentations or strategies.[1,4]

***"Unilateral absence of a pulmonary artery (UAPA), first described by Frantzel in 1868, is a rare cardiac anomaly thought to develop from the persistence of the hilar components of the pulmonary artery to the distal sixth aortic arch with concurrent involution of the proximal portion of the same arch.[1,2]"***

## Case Description

A previously healthy two-day-old term female infant was referred to our center for evaluation of Congenital Heart Disease following an episode of desaturation to 80% at 16 hours of life. An echocardiogram demonstrated only a single left pulmonary artery with a small ipsilateral Patent Ductus Arteriosus (PDA) with bidirectional flow. Following transfer to our hospital, the infant was started on prostaglandin (PGE) infusion. After several hours of PGE infusion and clinical stability with oxygen saturations of 95%, a repeat echocardiography was not able to show any discernible right pulmonary artery flow. No

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aortic collaterals were noted going to the right lung and no right-sided pulmonary venous flow could be seen. Right ventricular enlargement, PDA bidirectional flow, and interventricular septal flattening were all suggestive of elevated right heart pressures.

Prostaglandin therapy was continued. Computed tomographic arteriography was suggestive of a distal right pulmonary artery (RPA) that originated from an occluded right-sided PDA (Figure 1A and 1B). The lumen and size of the RPA could not be established as no contrast entered it. The right lung was otherwise well developed with normal pulmonary venous return, Cardiac catheterization with pulmonary venous wedge angiography confirmed the diagnosis of anomalous origin of the RPA from the innominate artery via an occluded right-sided ductus, as well as the presence of thrombus in the distal RPA (Figure 2A and 2B).

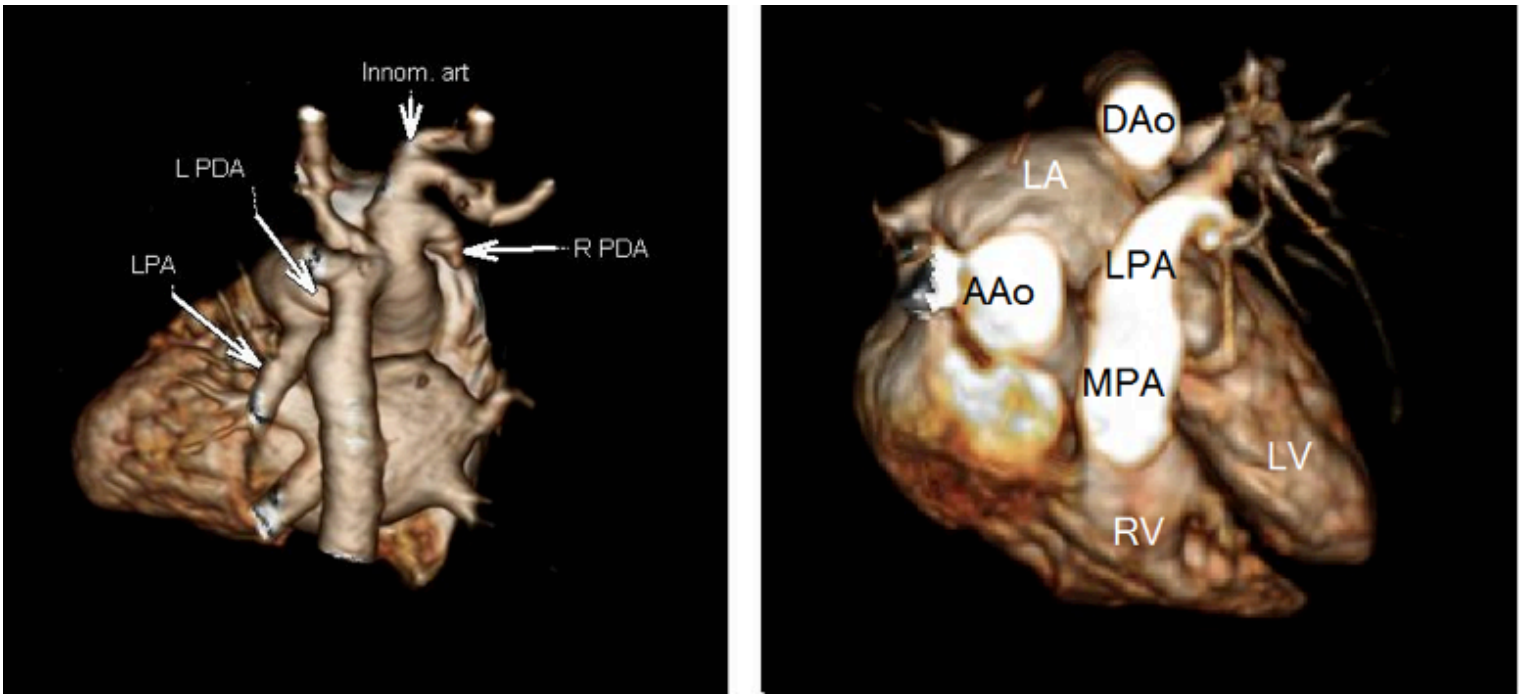
The next day, the patient was taken to the operating room to re-establish branch pulmonary artery continuity. The operation was performed through a median sternotomy without cardiopulmonary bypass. Upon opening the chest, the right lung was noted to be clearly paler than the left lung. A portion of the pericardium was harvested and formed into a 6 mm diameter tube using 7-0 Prolene suture. The patient was then systemically heparinized with 150 units/kilogram. In addition, PGE infusion was maintained and inhaled nitric oxide was started at 20 parts per million to avoid right ventricular failure during repair. Once snares were placed around the right-sided PDA, as well as the right upper and lower pulmonary artery branches, an incision was made in the right lower lobe branch. A Fogarty catheter was passed to remove all intraluminal clots within the RPA. The pericardial tube was then connected to the right pulmonary artery in an end-to-side fashion. The tube was clamped and the right sided snares were released, allowing continuous blood flow to the right lung via the right PDA. The pericardial roll was placed behind the aorta, trimmed to the appropriate length, and sewn in an end-to-side fashion to the main pulmonary artery after placement of a side biting clamp. Both right and left ducti were then ligated, which did not significantly change systemic or pulmonary blood flow. The chest was closed and the patient was transferred to the Intensive Care Unit in stable condition. Her post-operative course was uneventful and she was discharged six days after the operation on daily furosemide and prophylactic aspirin.

Post-operative echocardiography showed persistent patency of the pericardial tube; however, she was lost to follow-up for several months. When she returned for care at 6 months, an echocardiogram revealed the interval development of some RPA stenosis. Cardiac catheterization revealed narrowing of the reconstructed RPA with the minimal diameter of 3-4 mm (Figure 3A). Initial RPA angioplasty with a 7 mm balloon resulted in a residual waist in the RPA. An 8 mm diameter, 18 mm length Valeo stent was placed. Initial dilation to 14 atmospheres of pressure resulted in residual stenosis. Subsequent dilatation with an 18 mm Conquest balloon taken to 23 mm atmospheres of pressure showed complete resolution of the waist, with angiographic equivalency of RPA and LPA flow. Prior to stent placement, right ventricular pressure was 27/5 mmHg and RPA pressure was 17/11 mmHg. Following angioplasty and stent placement, right ventricular pressure was 27/8 mmHg and RPA pressure was 20/8 mmHg. Repeat cardiac catheterization done 15 months postoperatively showed a widely patent RPA with a right ventricular pressure of 22/7 mmHg. She is doing well without clinical or echocardiographic evidence of recurrent RPA stenosis. She is off aspirin.

**Discussion**

The natural history of UAPA typically involves a progression of recurrent respiratory infections, hemoptysis due to congestion of collateral arterial circulation, and pulmonary hypertension. Signs and symptoms in infancy may include: murmur, failure to thrive, cyanosis, heart failure, and less commonly, respiratory distress or signs of pulmonary hypertension. As patients may remain asymptomatic or have vague symptoms, the diagnosis of UAPA can be difficult to make in infancy, which contributes to the later age at presentation with resultant difficulties in repair and effects on symptoms and outcomes.[5,6]

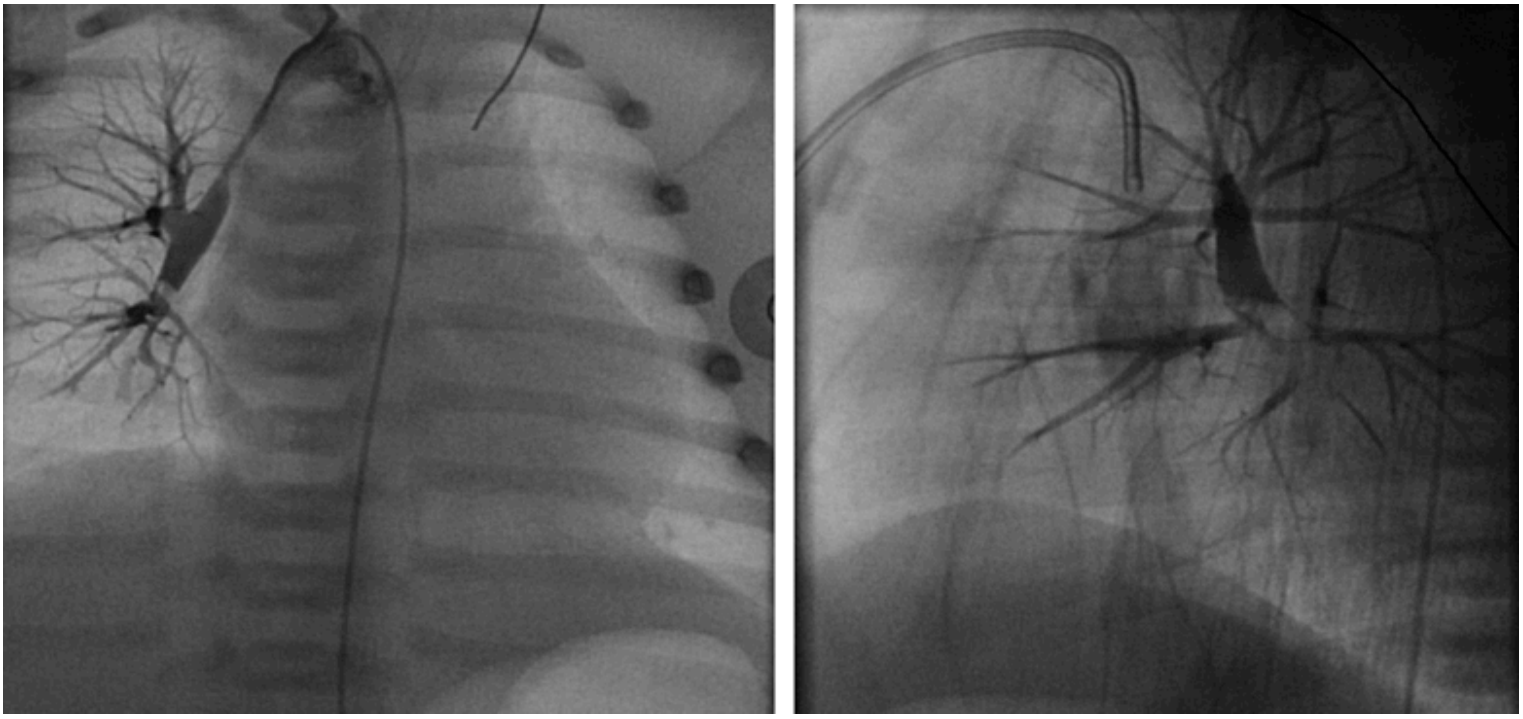
Anatomic repair can be delayed by using a temporary modified Blalock-Taussig shunt to re-establish pulmonary perfusion.[7]



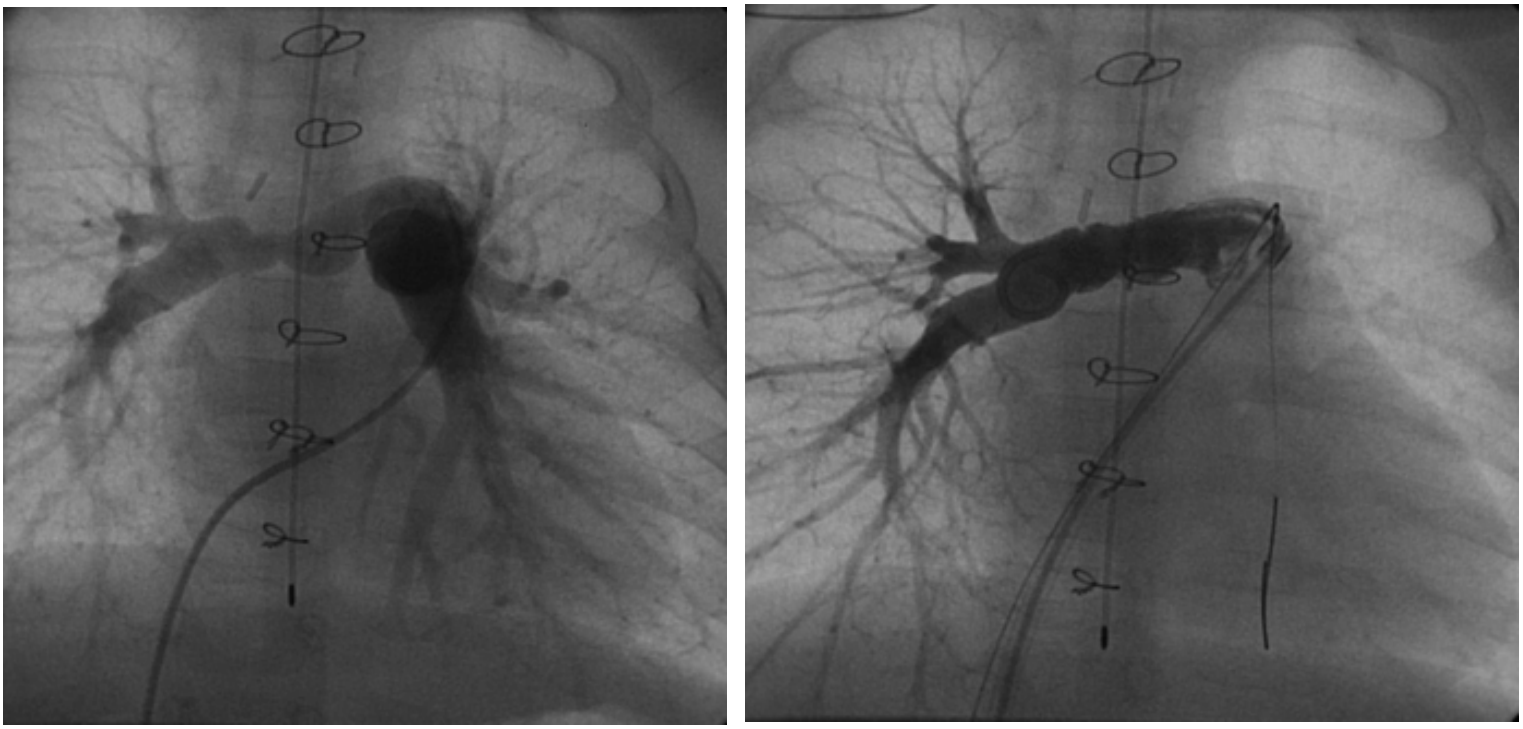

*Figures 1. Computed tomography angiogram illustrating complete absence of proximal RPA and presence of occluded right ductus arteriosus (R PDA). Left pulmonary artery (LPA), MPA (main pulmonary artery), AAo (ascending aorta), LA (left atrium), RV (right ventricle), LV (left ventricle).*

Previous case reports describe surgical palliation of UAPA utilizing artificial conduits such as polytetrafluoroethylene (PTFE) initially or repair with native tissue, such as a main pulmonary artery flap or pericardium to augment the anterior wall.[2,3] The present case differs in that we describe a single-stage surgical approach for repairing UAPA utilizing only an autologous pericardial tube.

An artificial graft in our patient would have been unable to grow with the patient, a critically important issue in the pediatric population. In a previous study, the diameters of PTFE grafts used in infant UAPA repairs measured 5 mm-8 mm, whereas the diameter of an adult right or left pulmonary artery is at least 15 mm.[3] Using a graft that does not enlarge over time requires a child to undergo subsequent staged operations to ensure they have a normally-sized PA and attendant adequate blood flow to the affected lung. An autologous pericardial tube, however, has the demonstrated potential to grow with the patient.[8] In addition, although anatomic placement of the pericardial roll behind the aorta may result in external compression, pericardial rolls have demonstrated enough compliance to withstand transcatheter ballooning and stenting as needed.[8]



*Figures 2. Cardiac catheterization with anterior (Figure 2A) and lateral (Figure 2B) views illustrating right ductus arteriosus off innominate artery giving rise to RPA with thrombus present in right lower pulmonary artery. Figure 2A has right PDA opened with #3 French pigtail catheter.*



*Figures 3. Cardiac catheterization at nearly 7 months of life showing anterior views before (Figure 3A) and after (Figure 3B) stent placement. Figure 2A demonstrates 3-4 mm narrowing of the RPA prior to stent placement.*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*"As patients may remain asymptomatic or have vague symptoms, the diagnosis of UAPA can be difficult to make in infancy, which contributes to the later age at presentation with resultant difficulties in repair and effects on symptoms and outcomes.[5,6]"*

Our patient presented with cyanosis and imaging evidence of an UAPA associated with acute distal RPA thrombus, and concerns for the subsequent development of an isolated right lung and pulmonary hypertension. Revascularization was achieved with thrombectomy and placement of a native pericardial tube to reconnect the main pulmonary artery and RPA. The Valeo stent placed is capable of dilatation to 18 mm, minimizing the need for future staged surgical intervention as our patient grows.

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**CCT**


*Mark Cartoski, MD, AAP*
*Chief Resident*
*Associate Instructor*
*Eastern Virginia Medical School*
*Children's Hospital of the King's Daughters*
*Department of Pediatrics*
*601 Children's Lane*
*Norfolk, VA 23507 USA*
*P: (757) 668-9759; F: (757) 668-9625*
*Mark.cartoski@chkd.org*

*Jessica M. Parson BS*
*Eastern Virginia Medical School*
*Norfolk, VA 23501 USA*

*Alexander Ellis, MD*
*Eastern Virginia Medical School*
*Children's Hospital of the King's Daughters*
*Department of Pediatrics*
*Department of Cardiology*
*Norfolk, VA 23507 USA*

*Elliot Tucker, MD*
*Eastern Virginia Medical School*
*Children's Hospital of the King's Daughters*
*Department of Pediatrics*
*Norfolk, VA 23507 USA*

*Felix W. Tsai, MD*
*Department of Cardiac Surgery*
*Children's Hospital of the King's Daughters*
*601 Children's Lane*
*Norfolk, VA 23507 USA*

# An Unusual Case of Totally Anomalous Pulmonary Venous Connection

*By Shazia Bhombal MD and Grace C. Kung, MD*

## Introduction

Totally Anomalous Pulmonary Venous Connection (TAPVC) is defined as failure of the development of the common pulmonary vein and, as a result, drainage of the pulmonary veins into the systemic venous system. TAPVC is commonly classified by the location of the end point of drainage, i.e. supracardiac (Type I, most common), cardiac (Type II), infracardiac (Type III), or mixed (Type IV).[1] Thoroughly defining the course of each anomalous vein along with a confluence, if present, is important to determine surgical approach and to ensure normal physiology. We present an interesting case of TAPVC with pulmonary veins that entered a large confluence which drained via a vertical vein below the diaphragm, then ascended superiorly back into the chest and inserted into the innominate vein. Although the initial vein descended below the diaphragm, the ultimate site of drainage was to the innominate vein, thus classifying it as a Type I.

To our knowledge, this has not been previously described.

## Case Report

The patient is a full-term female neonate with a prenatal diagnosis of a single ventricle and pulmonary stenosis. Initial oxygen saturations were 20% on 100% oxygen. An echocardiogram was concerning for obstructed infracardiac TAPVC, and she was transferred to a higher care facility for further evaluation and management. Echocardiogram on admission to our institution demonstrated an unbalanced atrioventricular canal with hypoplastic left ventricle and dilated right ventricle, mild common atrioventricular valve insufficiency, common atrium, double outlet right ventricle with L-transposition of the great arteries, mild subpulmonary narrowing, and four pulmonary veins that drained into a vertical confluence. This constellation of findings was consistent with those associated with Heterotaxy Syndrome. The vertical confluence coursed below the diaphragm into the liver (Figure 1), then made a hairpin turn and drained upwards into the innominate vein. (Figure 2). The patient was taken to the operating room shortly after arrival to our institution and underwent repair of TAPVC, placement of 3.5 modified Blalock-Taussig shunt, and ligation of main pulmonary artery. Obstruction of the pulmonary veins were not definitively seen by transthoracic echocardiogram, nor by transesophageal echocardiogram in the operating room, however, initial sats were as low as 20% on 100% FiO2 and CXR on arrival to our hospital demonstrated bilateral diffuse edema, clinically consistent with obstruction. Abdominal ultrasound demonstrated a transverse liver and asplenia, consistent with heterotaxy syndrome. The infant underwent Ladd's procedure for malrotation during the initial hospitalization and was discharged home tolerating full enteral feeds. She underwent dilations of her pulmonary vein anastomotic site in the catheterization lab twice due to obstruction. She has had her bidirectional Glenn palliation and has been doing well.

## Discussion

The incidence of TAPVC has been reported at 0.008% of live births, with an incidence of approximately 2.2% of patients with congenital heart disease.[2] It is more frequent in heterotaxy patients, occurring in 65-90% of patients.[3] Type I (supracardiac, 45%) involves drainage to the systemic venous system by a left-sided vertical vein (an embryological remnant of the left cardinal system) most commonly to the innominate vein or directly to the right superior vena cava (RSVC). Type II

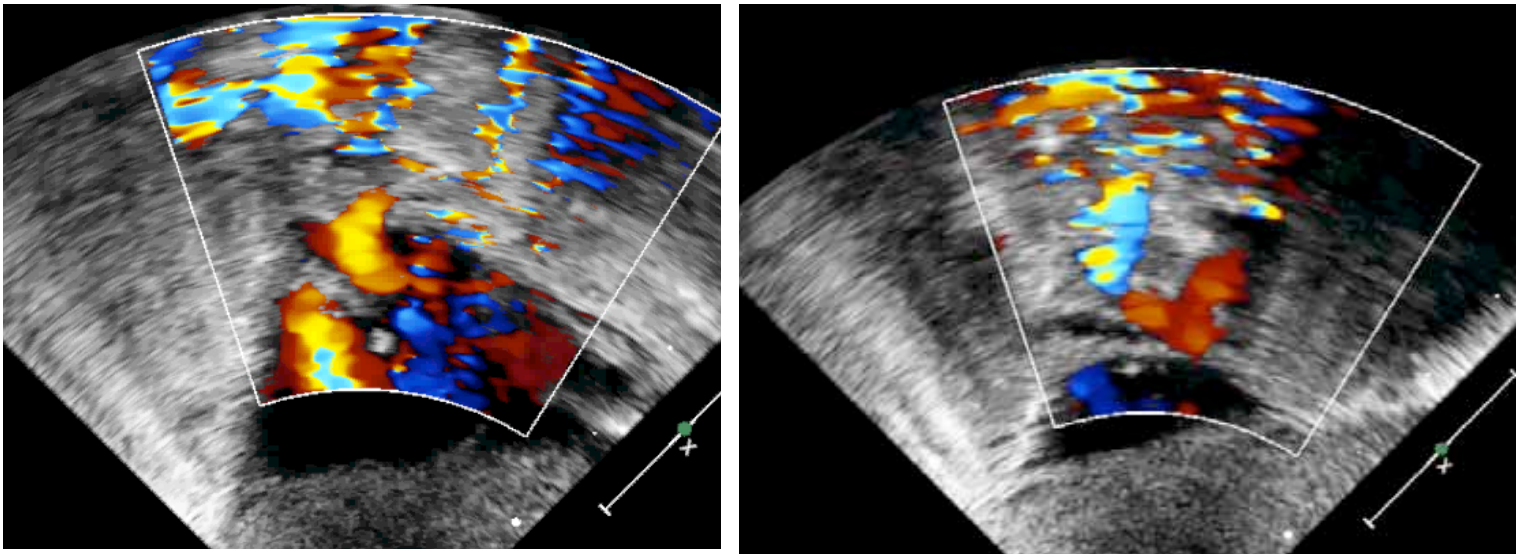

*Figure 1. Two-dimensional transthoracic echocardiographic image with color Doppler from a subcostal coronal view A Christmas tree appearance to the confluence, with upper veins (UV) and B lower veins (LV) entering at different levels.*

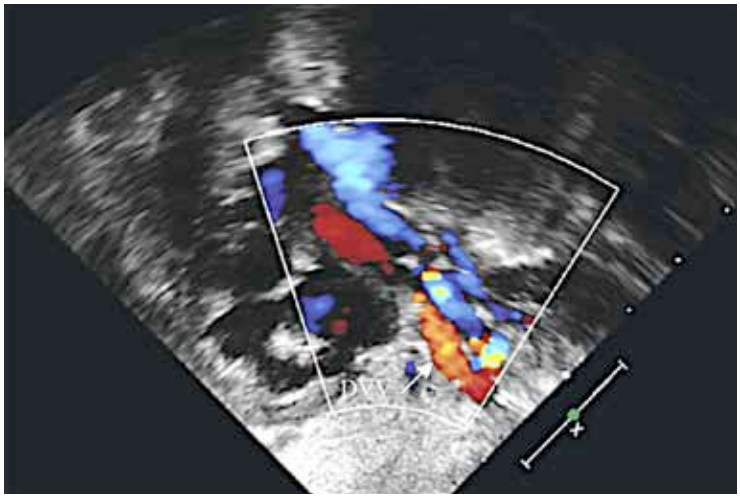


*Figure 2. Two-dimensional transthoracic echocardiographic image with color Doppler from a modified sagittal subcostal view showing red flow towards the transducer in the descending vertical vein (DVV) which then makes a hairpin turn below the diaphragm and ascends back into the thorax.*

(cardiac, 26%), drains directly into the heart usually via the coronary sinus (CS) or directly to the right atrium. Type III (infracardiac, 24%) involves drainage via a descending vein coursing below the diaphragm and joining the inferior vena cava, hepatic veins or, most commonly, the portal system. Type III is usually associated with obstruction of the pulmonary venous return, potentially at the level of the diaphragm or the liver capillary bed. Type IV (mixed, 5%) involves multiple sites of drainage.[4,5] To our knowledge, TAPVC with features of Type I and Type III has not previously been described, and its embryologic nature is unclear. One can hypothesize this curious course occurred potentially due to the persistence of the left cardinal and umbilicovitilline system, however this is only speculative.

As stated above, heterotaxy patients have a high incidence of TAPVC, most commonly supracardiac. In a review of 72 postmortem cases, Rubino et al described the plethora of variations of TAPVC in the heterotaxy patients.[6] In their series, 58% of patients with heterotaxy had TAPVC, with the largest group of patients having extracardiac connections (42/72), most commonly supracardiac (30/72). Obstruction was found in 55% of cases of TAPVC. The presence of potential obstructed or obstructed TAPVC in heterotaxy patients may require early surgical intervention.[7] Careful examination of pulmonary veins and courses is prudent in patients with heterotaxy.

Our case is unusual in that it appears to have started as a form of infracardiac TAPVC, but then had an altered and reversed course, ascending above the diaphragm to eventually drain into the innominate vein. Its initial course is that of a Type III, but in actuality, it is a Type I. However, given its circuitous course crossing the diaphragm twice, the chance of obstruction is potentially higher than the usual Type I. While no definitive evidence of obstruction was seen by echocardiography, the patient presented clinically with obstructed veins with findings including respiratory distress and edema on CXR. In this patient, two-dimensional echocardiography with color flow Doppler clearly delineated the course of the wandering vertical vein. Other studies have demonstrated the accuracy of echocardiography with color flow Doppler in establishing a diagnosis of total anomalous pulmonary venous connection.[8]

This case demonstrates the importance of tracing the entire course of the vein and not simply its origin or end-destination.

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*Special thanks to Dr. Robert Anderson for reviewing this case and providing insights.*



***Corresponding Author***

*Shazia Bhombal, MD*
*Divisions of Cardiology and Neonatology*
*Children's Hospital Los Angeles*
*4650 Sunset Blvd, MS #34*
*Los Angeles, CA 90027 USA*
*SBhombal@chla.usc.edu*

*Grace C. Kung, MD*
*Division of Cardiology*
*Children's Hospital Los Angeles and Department of Pediatrics*
*Keck School of Medicine*
*University of Southern California Los Angeles*
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|---|---|---|---|---|
| RACHS-1* Category | CM** 1 Year Average | STS*** 1 Year Average | CM 4 Year Average | STS 4 Year Average |
| 1 (least complex) | 0.0 | 0.5 | 0.0 | 0.7 |
| 2 | 0.0 | 1.1 | 1.1 | 1.1 |
| 3 | 2.9 | 3.3 | 3.7 | 3.7 |
| 4 | 5.1 | 7.1 | 8.2 | 7.1 |
| 5 & 6 (most complex) | 15.4 | 14.3 | 20.8 | 16.4 |

*Risk Adjustment for Congenital Heart Surgery classification

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# Wrap-Up LAA Frankfurt 2013

*By Sameer Gafoor, MD*

The *CSI-LAA 2013 - "How to Close the LAA Appendage*" took place November 22-23, 2013 at the Marriott Hotel in Frankfurt, Germany. This amazing conference is unique for its ability to deal with both the theory and practice of LAA closure, all while complementing the conference with numerous live cases. This was done with the help and assistance of world leaders in the field. Over 250 attendees from 31 different countries participated this year in *LAA Frankfurt 2013*, contributing to the success of the meeting by taking part in the vivid discussions.

The conference began with "Transseptal Puncture," taught by Matthew Price, Randall Lee, and Carsten Israel. This was followed by the crucial imaging component - answering both the questions: "Which imaging to use (TEE, angiography, ICE, dynaCT, MRI, or CT) and When?" This topic was presented with excellent pictures from Dr. Jai-Wun Park, Olaf Franzen, and Sergio Berti.

After this came device-specific sessions for: WATCHMAN; Amplatzer Cardiac Plug; Coherex; and the Lariat system. Each session followed a similar format:

- Why was this device designed the way it is?
- How it compares to other devices
- How to select patients for this device with step-by-step, and case examples.

Then the conference circled back to the basic question, "When and why do we close the LAA?" Jennifer Franke discussed the balance of stroke and bleeding risk, followed by Martin Bergmann with antithrombotic strategies. New arenas included: Stefan Bertog's talk about LAA closure in valvular afib and Randall Lee's talk about LAA closure before and after Atrial fibrillation ablation. To round out the session came the debate between Drs. Jai-Wun Park and Dr. Tom de Potter, on "Whether LAA closure is indicated only if anticoagulants are contraindicated, or in all patients."

The first day led us into the second day of talks, which started with "Building a Program." This covered building a referral network (Dr. Richard Folkeringa), picking a device for an LAA, and the new topic of residual leaks closure (Dr. Simon Lam).

This was followed by evidence for LAA closure by device (Dr. Krzysztof Bartus for Lariat; Dr. Matthew Price for WATCHMAN; Dr. Apostolos Tzikas for ACP/Amulet; Dr. Dhjanunjaya Lakkireddy for Lariat; and Dr. Vivek Reddy for Coherex Wavecrest). We learned both randomized and registry data for each device, allowing the consolidation of data to determine risk of stroke, bleeding, thrombus, and residual leak.

As one knows, the technology is ever changing and growing. Next the conference members discussed design considerations and experiences with the Amplatzer Amulet device (Dr. Apostolos Tzikas); Lifetech LAA Closure device (Dr. YY Lam); the Atricure device (Dr. Mokracek); and other LAA closure devices in early stages (Dr. Simon Lam).

As is usual for the CSI conferences, the last session was on challenging cases, discussed by Drs. Brendan Gunalingam, Kevin Walsh, and Neil Wilson.

In addition, eight live cases of LAA closure were shown, including the techniques and tips using a step-by-step approach. TEE and ICE were both used for these cases, showing the advantages and challenges of each approach. Cases were performed using a variety of devices, including: WATCHMAN, ACP, Coherex, and Lifetech. Materialise 3D models were also used prior to the certain selected cases, helping to determine procedural challenges and also best fit for device selection.

This amazing conference happens yearly in Frankfurt. Due to the popular demand, we are replicating this conference with one specifically devoted to the physicians in Asia-Pacific. Our conference entitled, *LAA Asia Pacific* (www.csi-laa-asia-pacific.org) will take place April 18-19, 2014 in Shanghai, China. We look forward to more exciting developments and discussion and invite you to be part of this conference as we discuss and grow in this exciting field.

**CCT**

*Sameer Gafoor, MD*
*Advanced Cerebrovascular, Structural Heart, and Peripheral Vascular Interventions Fellow*
*Cardiovascular Center Frankfurt*
*Frankfurt Germany*

*s.gafoor@cvcfrankfurt.com*

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**Dr. Ziyad M. Hijazi Appointed Clinical Chief for Pediatrics at Sidra Medical and Research Center, and Chair of Pediatrics at Weill Cornell Medical College in Qatar**

Sidra Medical and Research Center. Doha, Qatar announced in November 2013 that Dr. Ziyad M. Hijazi has been appointed Clinical Chief for Pediatrics. In this role, the pioneering and highly experienced clinician will drive the strategic direction of the Department of Pediatrics and integrate research and education priorities into a program of excellent clinical service delivery. He will also hold the position of Chair of Pediatrics at Weill Cornell Medical College – Qatar, Sidra's partner for medical education.

"Sidra will be a beacon of excellence for pediatric care in Qatar and the Middle East so this is a key appointment for us," said Sidra's Chief Medical Officer, Dr. Edward S. Ogata. "Dr. Hijazi's wide-ranging clinical and academic expertise in both pediatrics and cardiology, and his passion for research of the highest quality – evident from his exceptional record of publications – perfectly complement our vision for Sidra. I am particularly pleased that he will be here when Sidra hosts the *Excellence in Pediatrics Conference* in December 2013, the first time that this major conference has ever been held here in the Middle East."

Dr. Hijazi graduated from the University of Jordan School of Medicine before attending Yale University School of Medicine where he completed a Master of Public Health degree, a residency in Pediatrics and a fellowship in Pediatric Cardiology. He spent eight years on the faculty at Tufts University School of Medicine and another eight years at the University of Chicago. In 2007 he was appointed Professor, Pediatrics and Internal Medicine, Director, Rush Center for Congenital and Structural Heart Disease, and Section Chief, Pediatric Cardiology at Rush University Medical Center.

Dr. Hijazi specializes in treating congenital and structural heart disease in both children and adults. He is a pioneer in the non-surgical repair of congenital and structural heart defects and a Past President of the Society for Cardiovascular Angiography and Interventions (SCAI).

"It's an honor to join Sidra and its prestigious leadership team. It's also a personal pleasure for me to come back to the region where my love for medicine began. I hope to bring back what I have learned and contribute a small part to improving the lives of women and children in Qatar and the region. I'm optimistic that my work with transcatheter septal defect closure in children, which will be the focus of my presentation at the Excellence in Paediatrics Conference, is helping find alternatives to the invasive open heart surgery which is especially traumatic for pediatric patients and I look forward to bringing these techniques to Sidra," said Dr. Hijazi.

Hosted by Sidra, the *5th Annual Excellence in Pediatrics Conference* took place in Doha on December 4-7, 2013, at the Qatar National Convention Centre. The conference brought together international academics, scientists and clinicians to share cutting edge research and best practice with the aim of improving care for pediatric patients. It was expected to be one of the largest international pediatric conferences of the year and was attended by more than 3,000 healthcare professionals from 98 countries.

Sidra Medical and Research Center is a groundbreaking hospital, research and education institution, based in Doha, Qatar, that will focus on the health and wellbeing of women and children regionally and globally. Sidra will be a fully digital facility, incorporating the most advanced information technology applications in clinical, research and business functions. Sidra will initially have around 400 beds with infrastructure to enable expansion to 550 beds in a subsequent phase.

Sidra represents the vision of Her Highness Sheikha Moza bint Nasser, who serves as its Chairperson. The high-tech facility will not only provide world-class patient care but will also help to build Qatar's scientific expertise and resources. Sidra will be funded by a US$7.9

billion endowment from Qatar Foundation, one of the largest endowments of its kind in the world.

Sidra is part of a dynamic research and education environment in Qatar that includes leading international institutions, including Sidra's academic partner Weill Cornell Medical College in Qatar. Through strong partnerships with leading institutions around the world, Sidra is creating an intellectual ecosystem to help advance scientific discovery through investment in medical research.

Sidra will have a unique working structure with inter-professional collaboration at the heart, providing the best holistic care for patients and an unparalleled learning environment for its medical professionals. Sidra will combine the best in design, technology, operations and practices from medical centers around the world to offer its employees an environment that is at once familiar and extraordinary.

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**BJC Healthcare Selects Digisonics Cardiovascular Information System for Pediatric Cath**

BJC Healthcare (www.bjc.org) in St. Louis, MO, will implement the Digisonics Cardiovascular Information System (CVIS) for their pediatric cath studies. The Digisonics CVIS will provide the facility with a comprehensive clinical database, high-powered PACS and structured pediatric reporting in one complete system.

The Digisonics CVIS will provide BJC Healthcare with functionality designed specifically for the pediatric cardiology workflow including Mullins and Mayer congenital heart diagrams which can quickly be modified using graphical drawing tools and drop down labels. An added HemoLink interface between the Digisonics cardiovascular information system and the hospital's hemodynamics system will autopopulate the demographics, hemodynamic measurements, medications and other data. Hemodynamic measurements including pressures and flow measurements will be autopopulated directly onto the congenital heart diagram for fast and easy review. The Digisonics cardiovascular information system will also be integrated directly with Cedaron for IMPACT Registry data submission.

HL7 interfaces between the hospital's EMR and the Digisonics cardiovascular information system will facilitate a fully electronic workflow. A DigiLink add-on to the HL7 Results Out interface provides their clinicians with access to PDFs of the finalized pediatric cath reports within their EMR. DICOM Modality Worklist will automate transfer of patient demographics to the facility's imaging modalities, creating a list of scheduled patient studies and significantly reducing manual data entry time. Legacy images will be migrated and forwarded to BJC Healthcare's vendor neutral archive. Access of archived images will be via Digisonics DICOM Query & Retrieve with Prefetch module.

For further information, www.digisonics.com.


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Mailing Address: PO Box 444, Manzanita, OR 97130 USA
Tel: +1.301.279.2005; Fax: +1.240.465.0692
Editorial and Subscription Offices: 16 Cove Rd, Ste. 200, Westerly, RI 02891 USA
www.CongenitalCardiologyToday.com

**Publishing Management:**
Tony Carlson, Founder, President & Sr. Editor - *TCarlsonmd@gmail.com*; Richard Koulbanis, Group. Publisher & Editor-in-Chief - *RichardK@CCT.bz*;
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