

# CONGENITAL CARDIOLOGY TODAY

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## Subpulmonary Obstruction Due to Aneurismal Ventricular Septum in a Patient with Congenitally Corrected Transposition of the Great Arteries and Dextrocardia

By Tharakanatha R. Yarrabolu, MD; Mohinder  
K. Thapar, MD; P. Syamasundar Rao, MD

### Introduction

Congenitally Corrected Transposition of the Great Arteries is usually associated with multiple cardiac defects. Some defects are complex and others are simple, such as atrial or ventricular septal defects. Membranous ventricular septal defects tend to close spontaneously; such closures are usually due to plastering down of one of the leaflets of the tricuspid valve, commonly referred to as formation of aneurysm of the membranous ventricular septum. Rarely, the aneurismal tissue occluding the ventricular septal defect may prolapse into the outflow tract of the morphologic left ventricle and cause significant obstruction to the pulmonary outflow tract, requiring surgical therapy. We report a case of severe subpulmonary stenosis due to an aneurysm of the membranous ventricular septum in a patient with Congenitally Corrected Transposition of Great Arteries and Dextrocardia.

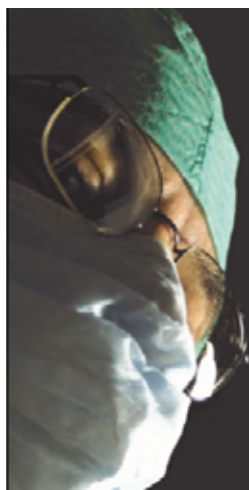
### Case Report

A three-year-old asymptomatic male child was referred to us for evaluation of dextrocardia. On cardiovascular examination, the apical impulse

*“We report a case of severe subpulmonary stenosis due to an aneurysm of the membranous ventricular septum in a patient with Congenitally Corrected Transposition of Great Arteries and Dextrocardia.”*

was felt in the right midclavicular line at the 5<sup>th</sup> intercostal space with: right precordial heave, a normal first sound at the right apex, a single second heart sound at right upper sternal border and a grade III/IV ejection systolic murmur best heard at the right upper sternal border. Liver edge was palpable in the left upper quadrant of the abdomen. There were no clinical signs of congestive heart failure.

Chest x-ray (Figure 1) revealed dextrocardia, left-sided liver and left-to-right reversal of bronchi, indicating situs inversus totalis. The electrocardiogram showed negative P waves



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in lead I suggestive of atrial inversion and Q waves in all chest leads (Figure 2).

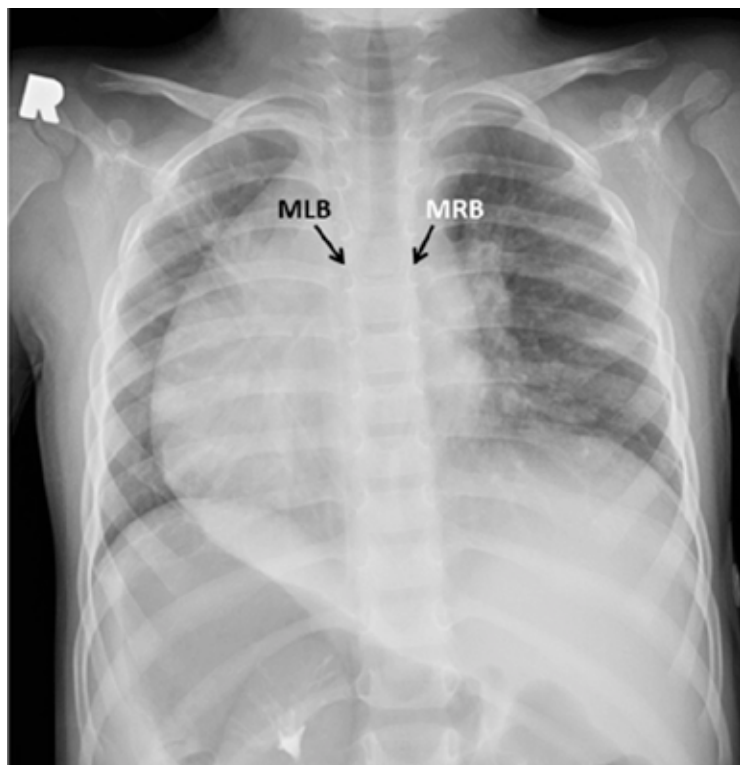


Figure 1: Chest X-ray in posterior-anterior view showing dextrocardia, reversal of right (MRB) and left (MLB) main stem bronchi and left sided liver.



Figure 2: Electrocardiogram showing negative P waves in lead I suggestive of atrial inversion and Q waves in all chest leads.

Echocardiogram revealed dextrocardia with atrial situs inversus, atrio-ventricular discordance, D-loop of ventricles, and ventriculo-arterial discordance. The features are consistent with corrected transposition physiology. There was a moderate-sized perimembranous ventricular septal defect with left-to-right ventricular shunting (Figure 3). There was aneurismal tissue beneath the pulmonary valve (Figure 4) causing

severe sub pulmonary stenosis with a peak Doppler flow velocity in excess of 5.0 m/s with a peak instantaneous gradient of 110 mmHg and a mean of 59 mmHg (Figure 5).

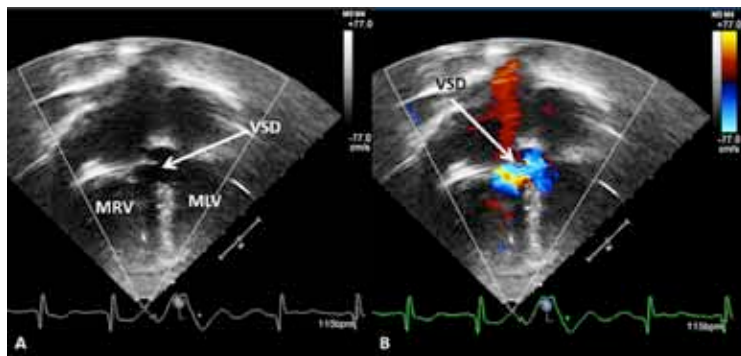


Figure 3: Two-dimensional (A) and color flow mapping (B) images of apical four-chamber echocardiographic views obtained from the right chest demonstrating moderate to large ventricular septal defect (VSD). MLV, morphological left ventricle; MRV, morphological right ventricle.

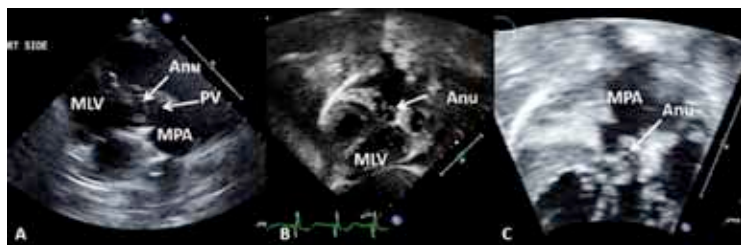


Figure 4: (A) Selected video frame from a long axis two-dimensional echocardiographic view of the morphological left ventricle (MLV) showing aneurismal tissue (Anu) protruding into the pulmonary outflow tract, causing obstruction. The Anu is located beneath the pulmonary valve (PV). Dilated main pulmonary artery (MPA) is also seen. (B.) The Anu is also demonstrated in the subcostal four chamber view. (C) Magnified view of B, again demonstrating the Anu and dilated MPA.

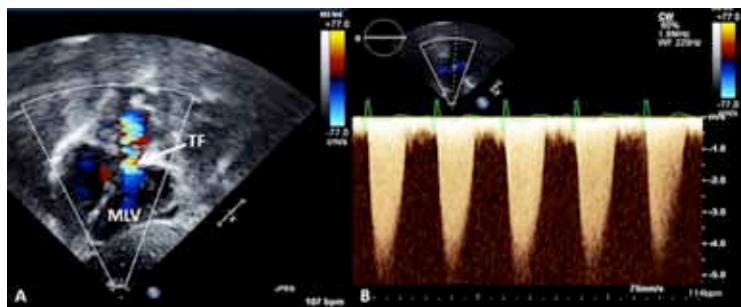


Figure 5: (A) Selected video frame from a subcostal four chamber view (similar to figure 4B) with color flow mapping demonstrating turbulent flow (TF) in the pulmonary outflow tract. (B) Continuous wave Doppler recording across the pulmonary outflow tract shows peak Doppler velocity in excess of 5 m/sec suggesting severe obstruction. See the text for the calculated gradients. MLV, morphological left ventricle.

Cardiac catheterization and selective cineangiography confirmed the diagnosis of dextrocardia and atrial situs inversus. The left-sided



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morphologic right atrium was connected to the left-sided morphologic left ventricle which gave rise to the pulmonary artery (Figure 6A). The right-sided morphologic left atrium was connected to the right sided morphologic right ventricle which gave rise to the aorta (Figure 6B). The aortic valve is anterior (not shown), superior (Figure 6) and to the right (Figure 6) of pulmonary valve (D-loop). These data confirmed echocardiographic findings of corrected transposition physiology. The systolic pressure in the left-sided, morphologic left (pulmonary) ventricle was at systemic level, but pulmonary artery pressure was normal. There was 57 mmHg peak-to-peak systolic pressure gradient across the left ventricular (pulmonary) out flow tract. Right-sided morphologic right ventricular (systemic) and aortic pressures were normal. Selective morphologic left ventricular angiography revealed an aneurysm protruding into the subpulmonary region causing severe outflow tract obstruction (Figures 7 & 8). There was a moderate-sized ventricular septal defect with left-to-right ventricular shunting (Figure 7 A).

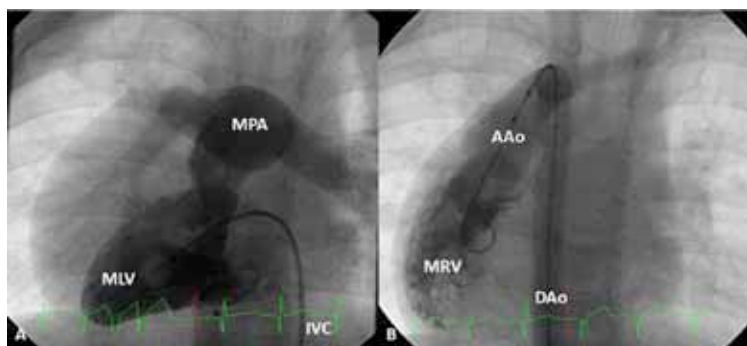


Figure 6: (A) Selected cineangiographic frame from a left-sided ventriculogram in a postero-anterior view showing a finely trabeculated, left-sided, morphological left ventricle (MLV) which gives rise to a dilated main pulmonary artery (MPA). Also note that the inferior vena cava (IVC) is on the left of the spine which is connected the left-sided right atrium (not shown). (B) Right-sided ventricular angiogram, also in antero-posterior view, showing a coarsely trabeculated, right-sided, morphologic right ventricle (MRV) which gives rise to the aorta (AAo). Note that the descending aorta (DAo) is on the right of the spine. The aortic valve is anterior (not shown), superior and to the right of pulmonary valve. These data would indicate D-loop of the ventricle, D-transposition of the great arteries in a subject with dextrocardia, a scenario indicative of corrected transposition physiology.

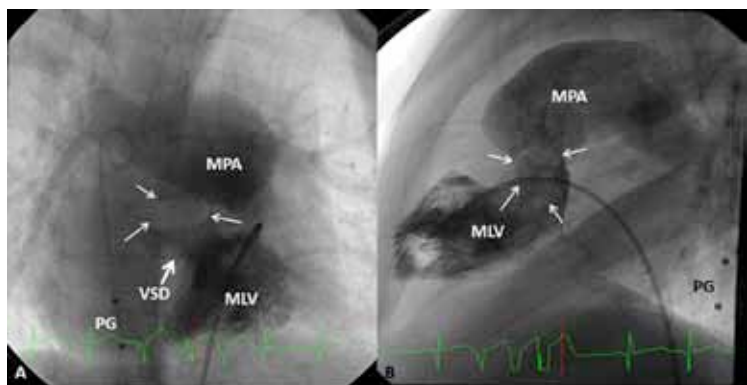


Figure 7: Selected cineangiographic frames from morphological left ventricular (MLV) angiogram in 60° left anterior oblique (A) and lateral (B) views showing pulmonary outflow tract obstruction from aneurysmal tissue (thin arrows), ventricular septal defect (VSD) (thick arrow) and post-stenotic dilation of the main pulmonary artery (MPA). PG, pigtail catheter in the descending aorta.

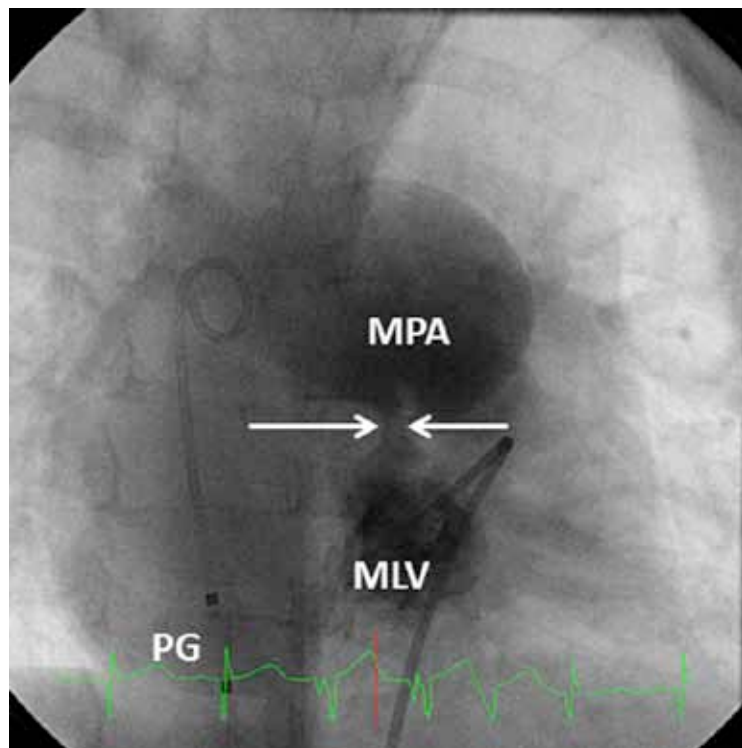


Figure 8: Selected cineangiographic frame from morphological left ventricular (MLV) angiogram in 60° left anterior oblique view showing severe narrowing of the right ventricular outflow tract (double arrows) due obstruction from aneurysmal tissue (not shown) and post-stenotic dilation of the main pulmonary artery (MPA). PG, pigtail catheter in the descending aorta.

It was recommended that the patient undergo surgical resection of the aneurysmal tissue along with closure of the ventricular septal defect. Intraoperative transesophageal echocardiographic findings are consistent with those of the transthoracic echocardiographic and angiographic data. Intraoperative findings were a moderate-sized ventricular septal defect with tricuspid valve tissue prolapsing through the defect forming an aneurysm protruding into the left-sided, morphologic left (pulmonary) ventricular outflow tract. The patient underwent resection of the aneurysmal tissue and closure of the ventricular septal defect with a Dacron patch. He developed complete heart block after the surgery, for which he received a permanent pacemaker at the same time. Currently, he is followed in the pediatric cardiology clinic, and at the last visit eighteen months after surgery, he was asymptomatic, had no residual ventricular septal defect but has mild residual pulmonary outflow tract obstruction with a Doppler peak instantaneous gradient of 30 mmHg and a mean of 16 mmHg.

## Discussion

Congenital Corrected Transposition of the Great Arteries was originally described by von Rokitsansky in 1875.<sup>1</sup> Several case series have been reported since 1950 documenting associated lesions and hemodynamic abnormalities. The incidence of this lesion is 1 in 33,000 live births which is approximately 0.05% of congenital cardiac malformations.<sup>2-4</sup> The majority are seen with situs solitus and only 5% are associated with situs inversus,<sup>5</sup> similar to our case.

The hallmark of the lesion is the so called “double discordance:” atrio-ventricular and ventriculo-arterial discordance. Because of this double discordance, the circulatory physiology is normal; systemic venous return goes into the lungs and the pulmonary venous return to the body.<sup>6,7</sup> The most common anatomical arrangement is levocardia with situs solitus, I-loop of the ventricles and anterior aorta which is located to the left of the pulmonary artery {S,L,L}. The less common form is



Dextrocardia with situs inversus, d-loop of the ventricles and anterior aorta which is rightward {I,D,D}, as in our case.

The most common lesions are: Ebstein's malformation of the morphologic tricuspid valve, ventricular septal defect, morphologic left ventricular (pulmonary) outflow tract obstruction and complete heart block.<sup>8-10</sup> Extensive study of the atrioventricular conduction system in corrected transposition with situs solitus patients by several investigators<sup>11-16</sup> determined that it is abnormally positioned, coursing in the anterior aspect of the subpulmonary tissue and along the anterior rim of the ventricular septal defect. In patients with small or atretic pulmonary trunk and normal septal alignment, the conduction system may consist of dual atrioventricular nodes with sling-like arrangement of conduction tissue.<sup>17</sup> The location of the atrioventricular conduction system is more variable in patients with situs inversus with corrected transposition physiology; Wilkinson et al<sup>18</sup> reported that the atrioventricular conduction system is usually situated in the posterior and inferior margin of the ventricular septal defect (anterior node may be present, but does not connect to the ventricular myocardium), in contrast to the superior and anterior location found in corrected transposition of the great arteries in situs solitus. These observations were similar to those reported by Dick and associates<sup>19</sup> in intracardiac electrophysiological study during surgery and by Thiene et al<sup>20</sup> in post-mortem hearts. These abnormal courses of the conduction system are of great surgical importance, especially in the presence of a ventricular septal defect or subpulmonary obstruction, since the conduction system is vulnerable to injury during surgical repair of the ventricular septal defect, with the potential for producing iatrogenic heart block, as in our case. Careful review of the location of the conduction system in a given situation (levocardia vs. dextrocardia) and, if necessary, intraoperative electrophysiological definition of the conduction system<sup>19</sup> may be necessary to prevent such a complication. This lesion is also prone to develop re-entry tachyarrhythmia and varying degrees of heart block.<sup>15</sup>

In this review we will focus on morphologic left ventricular (pulmonary) outflow tract obstruction. Issues related to pulmonary outflow tract obstruction in corrected transposition have been addressed in both early and more recent reports.<sup>8-10,17,21-27</sup> According to these authors the pulmonary outflow tract obstruction is due to several causes, and the most common causes are pulmonary valve stenosis or atresia and subvalvar pulmonary stenosis related to muscular malalignment and/or hypertrophy. Other less common causes are: fibrous tags or accessory valve tissue,<sup>28</sup> aneurysm of the membranous system,<sup>6,29-34</sup> tuberculoma<sup>34</sup> and intracardiac blood cyst.<sup>35</sup> Pulmonary outflow obstruction due to a prolapsing aneurysm of the membranous ventricular septum is rare,<sup>6,29-34</sup> including a detailed clinical, trans-thoracic and trans-esophageal echocardiographic, catheterization and angiographic and surgical description of this entity in 1996 in a situs solitus patient by the senior author.<sup>6</sup>

Aneurysms of membranous ventricular septum are commonly seen in association with perimembranous ventricular septal defects in patients with normally related great arteries<sup>36-38</sup> and constitute one of the most common mechanisms by which the ventricular septal defects close spontaneously. Although popularly called "ventricular septal aneurysm," it may not be a true aneurysm, nor derived from the ventricular septum. The origin of the aneurysm is difficult to ascertain even in pathologic studies; these studies suggest that this pouch (aneurysm) is derived either from redundant tricuspid valve tissue or

either from membranous septum itself.<sup>38-40</sup> Even though these closures with aneurysms are beneficial in many patients with ventricular septal defect, sometimes the aneurysms can cause obstruction of the pulmonary outflow tract. In patients with normally related great arteries, the aneurysm rarely causes outflow tract obstruction due to interposition of the conal septum and crista supraventricularis between the aneurysm and pulmonary valve. By contrast, in patients with Transposition of the Great Arteries who have higher right ventricular pressure, the aneurysm protrudes into the left ventricular outflow tract and causes pulmonary outflow tract obstruction.<sup>41</sup> Similarly, in patients with corrected transposition, in absence of conal septum and crista supraventricularis in the morphologic left ventricle the aneurysm is closer to the pulmonary valve leading to pulmonary outflow tract obstruction even when the aneurysm is small.

According to the previous reports<sup>6,29-34</sup> the age of presentation of this condition varies from 5 to 54 years, with a median age of 8 years; most patients were male. Our patient is 3-years-old and is younger than any patient reported so far in the literature. The presentation may be a cardiac murmur in an otherwise asymptomatic patient or symptomatic with chest pain, exertional dyspnea or easy fatigability. Precordial heave and a single loud second heart sound are present. A loud ejection systolic murmur of pulmonary stenosis at the right upper sternal border and a holosystolic murmur of ventricular septal defect at the right lower sternal border may be heard in patients with dextrocardia. If there is Ebstein's anomaly, a holosystolic murmur due to regurgitation of morphologic tricuspid valve is heard at the apex with radiation to the axilla. Cyanosis and clinical signs of heart failure are uncommon.

Electrocardiogram shows negative P waves in lead I suggestive of atrial inversion and abnormal Q waves in the chest leads. Chest x-ray confirms the dextrocardia, left-to-right reversal of the bronchi and left sided liver, suggestive of situs inversus (Figure 1). Echocardiogram confirms the diagnosis of atrial inversion, atrio-ventricular discordance, ventricular inversion, and ventriculo-arterial discordance and specific relationship of the great vessels.<sup>42,43</sup> Echocardiographic studies are also useful in demonstrating aneurysmal pouch protrusion into the morphologic left ventricular (pulmonary) outflow tract (Figure 4) and ventricular septal defect (Figure 3). Doppler interrogation quantifies the gradient across the obstruction (Figure 5). Cardiac catheterization is useful as an additional diagnostic tool to accurately measure the peak-to-peak systolic pressure gradient across the pulmonary outflow tract. Angiography clearly demonstrates the ventricular morphology (Figure 6), location and size of the aneurysm (Figure 7) and of the associated lesions. In the current era cardiac magnetic resonance imaging may be used as an alternative to angiography.<sup>44</sup> Three-dimensional echocardiogram<sup>45</sup> is another helpful diagnostic tool. Transesophageal echocardiogram is useful during surgery to reconfirm the diagnosis and to evaluate the extent of relief after surgery.<sup>6,33,46</sup>

Treatment of choice is surgical resection of the obstruction and closure of ventricular septal defect,<sup>6,29-34</sup> although some investigators<sup>32</sup> used valved conduit to bypass the obstruction. Complete heart block is a common complication for patients with corrected transposition leading some investigators<sup>47,48</sup> to advocate intra-operative electrophysiological identification of the conduction system. de Leval et al<sup>49</sup> described a technique of placing the sutures on the morphologically right side of the septum without opening the systemic ventricle to close the ventricular septal defect and reported no significant arrhythmias



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related to closure of ventricular septal defect. Various other surgical strategies, particularly double-switch,<sup>50-53</sup> have also been described as ways to restore the morphologic left ventricle to pump against systemic circulation; some physicians<sup>54,55</sup> consider double-switch preferable to conventional repair.

## Summary

Congenitally corrected Transposition of Great Arteries is usually associated with multiple cardiac defects. Morphologic left-ventricular outflow (pulmonary) tract obstruction due to aneurysm of the membranous ventricular septum in patients with corrected transposition and ventricular septal defect is rare, but was reported in the past. This is even more uncommon in patients with dextrocardia, prompting us to document this case. Absence of the conus with resultant proximity of the aneurysm to the subpulmonary region and higher pressures in the left-sided morphologic right ventricle lead to obstruction of outflow tract in corrected transposition. Echocardiogram with Doppler interrogation and cardiac catheterization with selective cineangiography are the diagnostic tests of choice. Surgical resection of the aneurysm with patch closure of ventricular septal defect, avoiding injury to the conduction system, is recommended.

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### Letters to the Editor

*Congenital Cardiology Today* welcomes and encourages Letters to the Editor. If you have comments or topics you would like to address, please send an email to: [LTE@CCT.bz](mailto:LTE@CCT.bz), and let us know if you would like your comment published or not.



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# Case Report: Scimitar Syndrome with Anomalous Left Coronary Arising from the Pulmonary Artery

By Tabitha G. Moe, MD; Ashish B. Shah, MD, MBA

## Case Report

A two-month-old female was seen by her primary care physician for a routine well-child check and thought to have a murmur. She was referred to a cardiologist, who diagnosed the murmur as a gallop, and ordered an echocardiogram. The echocardiogram demonstrated Scimitar Syndrome with: right pulmonary veins anomalously draining to the inferior vena cava, partial anomalous pulmonary venous return, atrial septal defect, and a dilated right ventricle with decreased right ventricular function. An electrocardiogram showed normal sinus rhythm with extreme rightward axis (Image 1). Referred for cardiac computed tomography and found to have 2 aortopulmonary collaterals, in addition to PAPVR (Image 2). the patient was then referred for cardiac catheterization for coiling of AP collaterals prior to operative repair of partial anomalous venous return. Cardiac catheterization was performed following lung perfusion imaging with a 75-25 perfusion mismatch left lung vs. right, and demonstrated anomalous left coronary arising from the left posterior pulmonary sinus (ALCAPA) (Images 3 and 4). Pulmonary hypertension with systemic right ventricular pressures secondary to her Scimitar Syndrome are thought to have maintained adequate coronary flow to avoid myocardial ischemia. At 2 ½ months she underwent operative repair with reimplantation of the left coronary using trap door technique, pulmonary artery reconstruction with a pericardial patch, partial Atrial Septal Defect (ASD) closure, and patent

ductus arteriosus ligation. After her initial operative repair the right ventricular pressures decreased to less than half systemic. At 5 months she underwent repair of Scimitar Syndrome with sutureless technique and anastomosis of scimitar vein to left atrium with completion of ASD closure, and open dilation of RPA stenosis. At age four she required right pulmonary artery balloon dilatation for right

pulmonary artery stenosis, but otherwise has done quite well.

## Discussion

Scimitar Syndrome and ALCAPA are both exceedingly rare congenital abnormalities. After an extensive review of the literature, there are three previously reported cases, and we report

***“The echocardiogram demonstrated Scimitar Syndrome with: right pulmonary veins anomalously draining to the inferior vena cava, partial anomalous pulmonary venous return, atrial septal defect, and a dilated right ventricle with decreased right ventricular function.”***

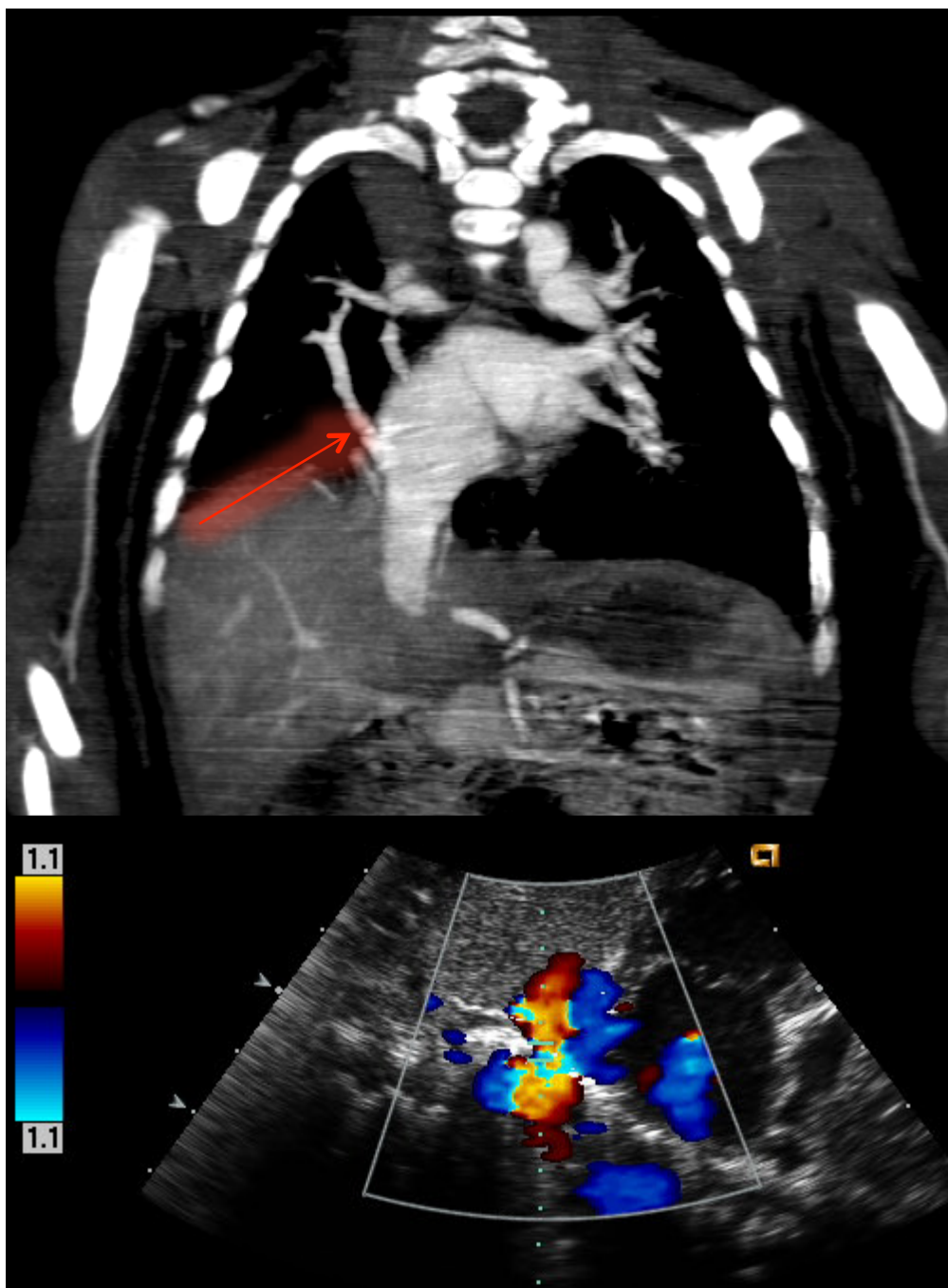


Image 1. Computed Tomography of the chest with angiography demonstrates the partial anomalous venous return of the right pulmonary vein to the inferior vena cava. Right pulmonary vein flow on transthoracic echo.

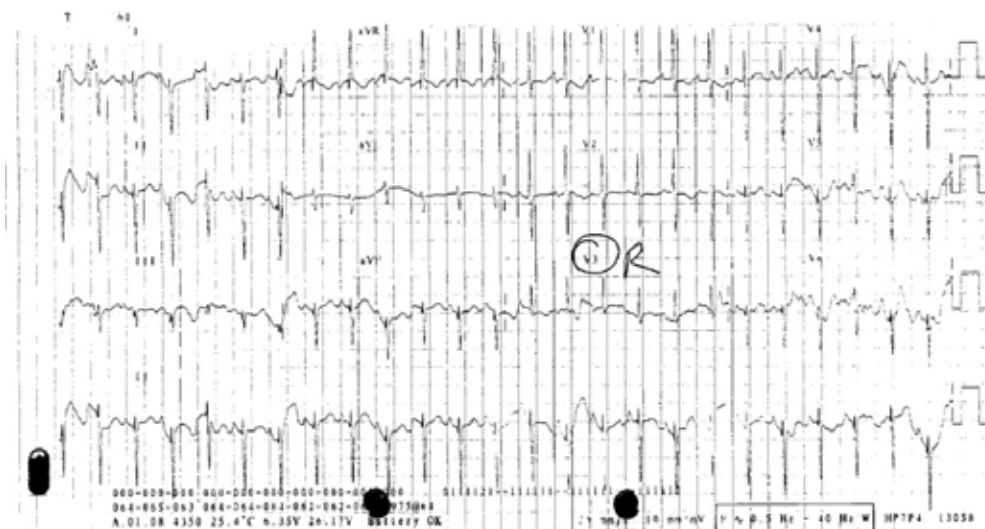


Image 2. Initial EKG shows extreme rightward axis.

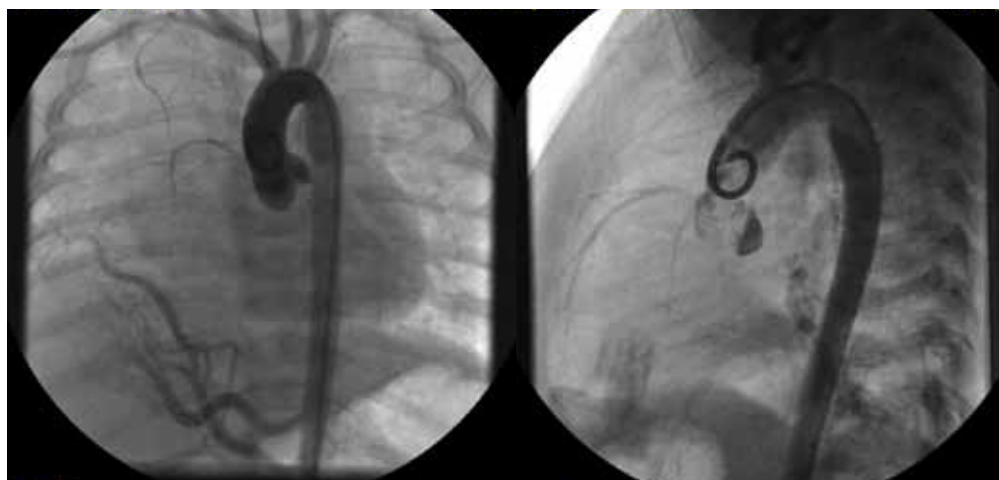


Image 3. Aortic Angiography demonstrating normal origin and course of the right coronary artery, with distinct absence of contrast within the distribution of the left coronary system.

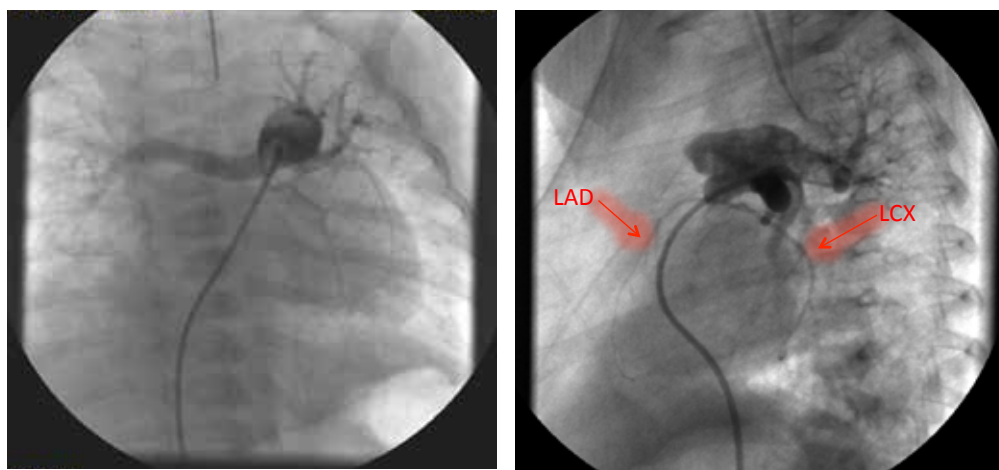


Image 4. Pulmonary Artery angiography demonstrating the abnormal origin of the left main coronary artery from the posterior main pulmonary artery. The left anterior descending (LAD) artery, and septal perforators can be seen coursing in the anterior interventricular groove, and the left circumflex (LCX) artery is well-visualized as it courses through the expected atrioventricular groove.

a fourth.<sup>1,2</sup> Of the previously reported cases there are details for one, who died.<sup>3</sup> The incidence of Scimitar syndrome is 1/100,000 live

births, accounting for 0.1% of congenital cardiac disease.<sup>4</sup> The incidence of ALCAPA is estimated at 1/300,000 live births, accounting

for 0.3%<sup>5</sup>. The combined incidence is estimated at 3/1,000,000. There are no statistics available in previously published literature. Scimitar Syndrome is named for the radiographic appearance of the right lung border. Infantile Scimitar Syndrome is associated with pulmonary hypertension, often diagnosed as persistent pulmonary hypertension of the newborn. Pulmonary pressures have been recorded as systemic, and even suprasystemic. The ALCAPA depends upon pulmonary blood flow for perfusion of the coronary artery territory. Therefore, systemic or suprasystemic pulmonary pressures or persistent pulmonary hypertension of the newborn is palliative for the ALCAPA patient. Normal hemodynamic transition from fetal to adult circulation requires closure of the ductus. In the setting of ductal closure, right coronary artery and collaterals form to perfuse the left coronary myocardial distribution. This subsequently results in right coronary artery dilatation and aneurysm formation, and ultimately, reversal of flow through the left coronary ostia into the main pulmonary artery.<sup>6</sup> Natural history of the disease leads to an additional phase of coronary steal where myocardial perfusion of left coronary artery territory from the right coronary artery has progressive shunting.<sup>7</sup> Scimitar Syndrome and the resultant pulmonary hypertension in our patient, was protective, and the ALCAPA was found incidentally at the time of cardiac catheterization. However, careful review of office echocardiography does not demonstrate the origin of the left coronary artery from the left coronary sinus raising suspicion of ALCAPA.

The authors have no disclosures.

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***“Scimitar Syndrome and the resultant pulmonary hypertension in our patient, was protective, and the ALCAPA was found incidentally at the time of cardiac catheterization. However, careful review of office echocardiography does not demonstrate the origin of the left coronary artery from the left coronary sinus raising suspicion of ALCAPA (Image 5).”***



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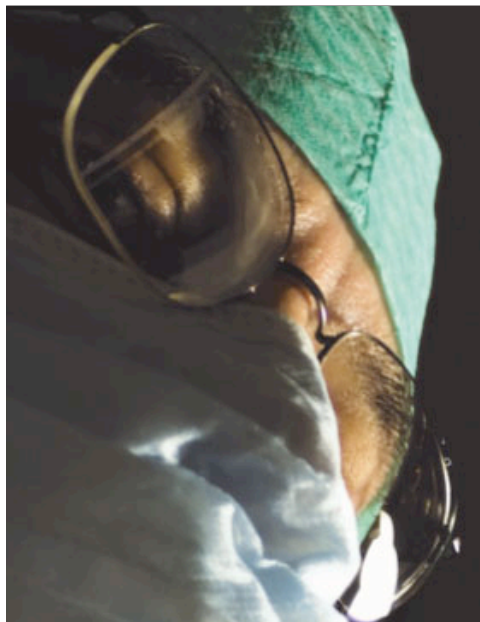
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# Synopsis of the Fifth Phoenix Fetal Cardiology Symposium, April 23-27, 2014

By Christopher L. Lindblade, MD

*The Fifth Phoenix Fetal Cardiology Symposium* that occurred April 23-27, 2014, was the largest symposium in the history of this conference. Nearly 250 participants traveled to Arizona from North America, as well as from several countries around the world including: Kuwait, Brazil, Japan, Italy, Australia, Chile, Netherlands, Egypt, and Great Britain. There was nearly equal representation from fetal cardiologists, maternal-fetal-medicine specialists, and sonographers. Several neonatologists, radiologists, fellows, residents, and nurses found great educational value in the Symposium as well. The prestigious faculty was comprised of leading experts in the field of fetal cardiology from highly respected fetal cardiology programs throughout North America.

Drs. Norman Silverman, Julia Solomon, and Christopher Lindblade co-directed the Symposium this year bringing new elements to this recurring conference. Fellows and attending physicians presented poster abstracts on their current research and observations. Dr. Stefano Faiola, maternal-fetal-medicine specialist from Milan, Italy, won the abstract competition with his work on Biochemical and Ultrasound Markers of Fetal Cardiac Dysfunction in Twin-to-Twin Transfusion Syndrome. Another session gave four selected physicians the opportunity to share with the Symposium attendees their most "nightmarish" fetal cardiac case. Dr. Emily Lawrence, fetal cardiologist from Texas Children's Hospital, took first place in this competition showing a case of ectopia cordis and presenting a brief review of her institution's experience with this rare condition.

The faculty delivered state of the art lectures focused on specific cardiac disease including other obstetrical conditions such as Twin-to-Twin Transfusion Syndrome. A conference highlight was Dr. Mary Donofrio's remarks about the American Heart Association Scientific Statement on Diagnosis and Treatment of Fetal Cardiac Disease minutes after its online publication. Symposium attendees commented that nearly every fetal cardiologist who contributed to the AHA Scientific Statement were guest faculty at the Symposium.

The Ritz-Carlton was a beautiful venue for the four and half day conference in Arizona. Dr. Norman Silverman presented an informative and entertaining dinner lecture on The History of Fetal Echocardiography on the first evening of the Symposium. On Friday night, over 30 attendees took advantage of an Arizona Diamondback baseball game with several experiencing America's favorite pastime for the very first time. Sunday morning workshops gave



Nearly 250 attendees heard over fifty lectures on fetal cardiac disease using with their electronic syllabus to follow along during the Symposium. Dr. Julia Solomon impressed the audience with her 3D/4D images and their clinical application.



Symposium faculty was comprised of leading experts in fetal cardiology from all over North America. An open question forum, moderated by Dr. Norman Silverman, gave the audience the opportunity to ask the faculty thought provoking questions sparking dialogue and discussion for all to enjoy.



Attendees interacted with Dr. Anita Moon-Grady and other Symposium faculty during hands-on scanning workshops focused on assessment of fetal cardiac anatomy, function, Doppler, and rhythm. A 4D volume manipulation workshop gave attendees the opportunity to work on GE laptops with assistance from faculty and GE clinical applications specialists.

participants the opportunity to put their hands on the ultrasound probe as well as perform post-processing on 3D/4D fetal echocardiographic imaging with faculty at their side.

*The Fifth Phoenix Fetal Cardiology Symposium* was a dynamic and rewarding educational experience for those who attended. Attendees and faculty alike took advantage of the

opportunity to collaborate with one another for future projects. Several left the Symposium asking when is next year's conference. The organizing committee is pleased to announce that *The Sixth Phoenix Fetal Cardiology Symposium* is scheduled for April 15-19, 2015. Go to [www.fetalcardio.com](http://www.fetalcardio.com) to check for more details as they become available. We hope to see you and many new faces there!

CCT



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# MD1World: Connecting the Global Medical World

By John-Charles Loo, MD

Two years ago, my friend and neighbor in Southern California, Brian Tran, asked for my help. A baby, named Tra My in Vietnam, had a heart problem; her mother was desperately seeking advice wherever she could get it. I'm certainly no more expert than any other pediatric cardiologist, but she sent me the echo images via DropBox, and I wrote down my findings for her. DILV, transposition, Hypoplastic aortic valve and arch, and coarctation. Not that unusual really, except the child was already 7 weeks old. And to make it more complicated, she was hospitalized with presumed pneumonia. The good news was that she was still in the hospital. At home, she would most certainly die.

The mother feverishly got her hands on every report she could and interpreted them for me. She sent emails to over a hundred people and organizations, driven to help her baby live. There were multiple challenges due to location, resources, not to mention my inability to speak Vietnamese. And unlike the typical medical mission trip, we were 8,000 miles apart.

Feeling a bit lost in an unfamiliar world and language, I decided to look for help. I had friends in organizations like *Samaritans Purse* and *For Hearts and Souls* who travel the world offering pediatric cardiac care. Someone must know more than me about how to best help. Then I came across an article written in *Congenital Cardiology Today*. It was written by an American who had been instrumental in building a pediatric cardiac program in South Vietnam. He carefully described the successes of developing the program, outcomes, limitations, and the huge wait list for children in need of heart surgery there. Dr. Culbertson just so happened to live up the coast from me in Northern California. His work and article was about the very hospital our DILV baby was at. I found his email address, and we went to work. We were in contact with groups throughout Vietnam, America, Europe, Canada, Australia, India, Thailand, even the U.S. Navy! We explored all the options available. Through an amazing series of divine coincidences, at nearly 3 months of age, Tra My became Vietnam's first Norwood. Post-op management was long and challenging, as expected. At the end of the 3 month journey, we looked back at the long email trail, countless phone conversations, and the beautiful photo of a baby and mother who fought hard and overcame one of the most complex medical conditions known.

Being that it was 2012, we wondered how we might do this better. How do we more efficiently align with colleagues across the world in order to improve the lives of children and families wherever needed. And how do we draw on the

knowledge, wisdom, and resources of the vast community of pediatric cardiologists and surgeons everywhere?

Out of these questions, an answer started to take shape. Just over 1 year later, we launched our website called *MD1World1.com* ([www.MD1World.com](http://www.MD1World.com)). We are dedicated and committed to bringing the pediatric cardiac community together, in an effort to support each other and colleagues worldwide, and ultimately enhance the care delivered to these children. It is the same reason deep in our own hearts that we decided to pursue this field to begin with. And we have already seen it work through baby Maya and others.

The *MD1World.com* platform is simple; it is specifically designed for pediatric and congenital cardiologists, and surgeons. After logging in, the dashboard highlights the most active and popular discussions, polls, and cases. Your activity enables the site to know your interests and can make better use of your valuable time. "Discussions" are helpful for asking questions and offering wisdom. You can also follow educational discussions and share rare entities and images. Important journal updates and discussions with the author in an online journal club is an exciting feature. "Polls" allow you to survey the entire community, and with very fast turnaround, you can receive hundreds of responses on controversial or challenging scenarios. You can virtually generate your own mini study data by responses from your trusted colleagues. The "Cases" section is where colleagues around the world post complex cases and invite you to help manage real patients in a virtual ICU telemedicine format. Engaging in any of these activities can literally take less than 15 seconds or as much time as you have to share. And there is so much more to come.

Physicians have always been in this noble and honorable profession that gives us the privilege of caring for human life. As pediatric cardiologists and surgeons, that tension is all the more tangible. The story and life of this precious baby girl in Vietnam touches our hearts in a deep and powerful way. The potential impact of *MD1World* to help transform the lives of these children and families is tremendous. No doubt that the "on the ground mission" trips with face-to-face education and support must continue. However, we believe that the collaborative community that includes you at: *MD1World.com* will be an invaluable asset to your practice, your soul, and that of colleagues and children around the world.

We look forward to you joining us today as we help to save lives around the world.

CCT

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## MD1World Founder Profiles

### Brian Tran, Founder

Mr. Brian Tran is a Vietnamese-American entrepreneur whose business efforts in Southeast Asia led him to develop MD1World. It was at his door manufacturing plant in Vietnam that an employee requested a leave of absence to care for her baby who was diagnosed with a rare heart defect and was given no chance of survival.

Mr. Tran enlisted the help of Drs. Casey Culbertson and John Charles (JC) Loo, both pediatric cardiologists, who worked together to collaborate with Vietnamese doctors and together extend the baby's life. Through this experience, Mr. Tran, and Drs. Culbertson and Loo saw both a need and an opportunity to bring advanced medical collaboration to developing nations while giving physicians a way to network and share with one another.

### Dr. Casey Culbertson, Co-Founder

Dr. Culbertson is a published pediatric cardiologist / cardiac intensivist with 26 years of experience in his field. He also has a passion for international medical philanthropy. He is the Founder of and is presently the Cardiac Advisor for the Pediatric Cardiac Open Heart Surgery Programs at National Hospital for Pediatrics in Hanoi, and 3 hospitals in Ho Chi Minh City, Vietnam. It was for these efforts that he was awarded the Global Citizen Award by the United Nations Association USA - East Bay Chapter.

Dr. Culbertson's public service includes serving as a Team Leader for Project Vietnam and a Pediatric Cardiology Advisor for the East Meets West Foundation. He is a member of the AAP and Sub-section of Pediatric Cardiology, the ACC, the Western Society of Pediatric Cardiology, and the Pediatric Cardiac ICU Society.

### Dr. John-Charles Loo, Co-Founder

Dr. Loo is a pediatric cardiologist with 13 years of experience in his field. He has long been interested in bringing advanced medical care to developing nations, helping patients in Mexico and Vietnam in addition to his practice in the U.S. Dr. Loo is a member of the Southern California Permanente Medical Group and has won several awards for his work at University of California at San Diego Department of Pediatrics, Miller Children's Hospital and Univ. of California at Irvine Dept. of Pediatrics.

Dr. Loo is a member of the American Board of Pediatric Cardiology, the American Board of Pediatrics, and the Medical Board of California.

# Medical News, Products & Information

By Tony Carlson, Senior Editor, CCT

## CFI Medical unveils Arm-IR™, a Stable and Safe Positioning Solution for Critical Upper Arm Placement.



CFI Medical, a leading proponent of improving clinical efficacy, has announced the release of Arm-IR™.

When positioning the arms on an anesthetized patient, prolonged placement above the head during the procedure may cause peripheral nerve injury. To address this risk, Arm-IR safely and comfortably stabilizes the upper arm during biplane cardiac catheterization to protect patients—pediatric as well as adult—from brachial plexus injury.

“Conventional wisdom is to position the arms above the head with the patient supine and keep the angles of flexion/extension in accordance to AORN guidelines to minimize injury to the brachial plexus nerve,” said Michael Czop, President, CFI Medical. “To date, this arm position has been achieved with rolled up blankets and tape. These are often difficult to keep in place and, when the blankets fall off the table, so do the patient’s arms.”

An effective alternative to using rolled up blankets or towels, adjusting Arm-IR to safely position the patient is simply a matter of tightening and loosening a single knob per arm and positioning the straps on the arm pads. The single knob provides quick and easy re-positioning, as one operator hand is occupied stabilizing the arm during re-positioning.

“The Arm-IR positioning device is another example of CFI Medical’s ability to realize a clinician concept into a beneficial clinical tool for patient and caregiver,” said Czop.

Arm-IR is designed with a variety of custom-fit clamps for compatibility with a majority of current-generation original equipment manufacturer catheterization tables, providing easy attachment to the head of the table. By utilizing standardized positioning, Arm-IR helps save facilities time, reduce costs and mitigate risk for better patient outcomes.

For more information about Arm-IR, visit [www.cfimedical.com/arm-ir](http://www.cfimedical.com/arm-ir).

## LUMEDX and Scientific Software Solutions Announce Strategic Partnership

LUMEDX Corporation, a leading provider of cloud-based cardiovascular imaging and information systems (CVIS), and Scientific Software Solutions, Inc., (SSS), maker of PedCath congenital catheterization (cath) reporting software, announced today that they are partnering to create an innovative solution for congenital cardiac data and image management.

The partnership brings together Scientific Software Solutions’ expertise in pediatric cath software and LUMEDX’s unique suite of cardiology data analytics, integration and registry tools, and clinical workflow applications to form a Comprehensive Congenital Cardiac Information System (3CIS). 3CIS will be the first of its kind to serve congenital cardiologists and surgeons, delivering fully integrated congenital cardiac patient records and advanced analytics.

“Collaborating with Scientific Software Solutions enables us to draw on both companies’ strengths to deliver a solution that provides unparalleled efficiency and value,” said LUMEDX VP of Strategic Products, Praveen Lobo. “Today there is no integrated solution for comprehensive congenital cardiac information management. Patient data is silo’d in disparate systems. Physicians have to log into multiple workstations to access patient history, review procedural data, annotate images and Mullins diagrams, and complete reports. Our partnership with SSS will allow us to offer an integrated solution with complete, up-to-date patient records, all with a single sign-on.”

“For the last twenty-seven years, we have focused PedCath development efforts on the unique requirements of congenital cath docs,” said Jack Wilson, CEO of Scientific Software Solutions. “There remained a great unmet need for a Comprehensive Congenital Cardiac Information System. Partnering with LUMEDX means that we will be able leverage LUMEDX HealthView technology to avoid duplicate data entry while improving accuracy. 3CIS will make it possible to easily access longitudinal congenital cardiac patient records—the cath report, NCDR and STS registry data, Echo reports, labs, more—so that clinicians can make fully informed treatment decisions more quickly. Participation in registries will be seamless.”

PedCath is the most widely used software for storing and reporting congenital and structural cardiac cath information. It combines the features of a patient database with customized drawing routines to provide a complete solution for recording pediatric catheterizations. LUMEDX will integrate PedCath with the full suite of LUMEDX HealthView CVIS tools and services, making detailed patient information available to physicians from within LUMEDX HealthView applications.

The LUMEDX-PedCath 3CIS will:

- Offer fully integrated patient information—data, images, study context, labs, meds—with single sign-on for longitudinal pediatric cardiology patient records
- Enable meaningful pediatric analytics and research initiatives by capturing discrete data



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- Embed PedCath modifiable Mullins Diagram into LUMEDX structured reports
- Support seamless enterprise-EHR integration with ADT and Results Reporting interfaces
- Make patient images immediately available in reports for comparison and review
- Support ACC NCDR® IMPACT Registry® and STS reporting
- Improve operational efficiency and enhance data integrity

This partnership will also enable LUMEDX and Scientific Software Solutions to provide seamless support and services to their clients at hospitals around the globe.

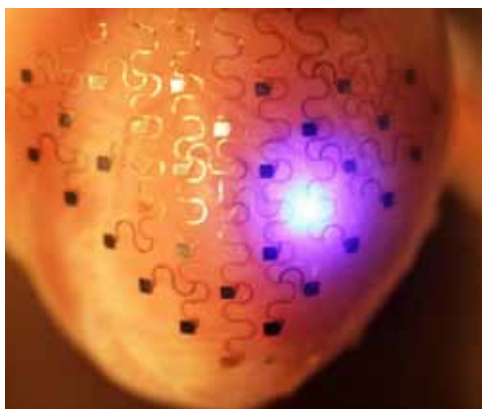
"We at LUMEDX have been expanding our pediatric cardiology offerings for several years, especially in the non-invasive space. PedCath complements our current offerings and is the preferred solution for many of our customers with congenital cath labs," said Lobo.

For more information, [www.lumedx.com](http://www.lumedx.com)

### 3-D Heart Sock Could Replace Pacemaker - Stretchable Silicone Device Can Monitor Vital Signs, Could Help Doctors Pinpoint Heart Problems

The "heart sock" is made of a thin silicone membrane embedded with flexible sensors that can monitor vital signs.

An international research team that includes a University of Alberta engineering professor has designed a 3-D silicone "heart sock" that could eventually replace the venerable pacemaker.



The heart sock is made of a thin silicone membrane embedded with flexible sensors that can monitor vital signs.

Hyun-Joong Chung, Professor of Chemical and Mechanical Engineering at the University of Alberta, John Rogers, Professor of Engineering and Chemistry at the University of Illinois, and Rogers' research group were co-authors of two recent articles published in *Nature Communications* and in *Advanced Healthcare Materials* on the development of the heart sleeve, which is designed to monitor vital signs.

"My role specifically involved developing the first stretchable multiplexing chemical sensor, namely a pH sensor with multichannel mapping ability," said Chung. "The pH sensor array was embedded in the heart sock format, enabling real-time observation of the heart's chemical activities."

The researchers embedded 68 tiny sensors into a sheet of silicone that they fit around a 3-D printed replica of a rabbit heart. The circuits were laid out in a curved, S-shaped design that allows them to stretch and bend without breaking.

The heart sock physically resembles the shape of the pericardium, the naturally occurring membrane surrounding the heart. The sensors in the soft, flexible membrane track vital signs such as temperature, mechanical strain and pH. The device is designed to maintain a stable fit to the heart tissue, while exerting minimal force on the contracting and relaxing heart muscle.

The heart sock could be used to identify critical regions that indicate the origin of conditions such as arrhythmias, ischemia or heart failure — information that could guide therapeutic interventions.

The finished design will feature electrodes capable of regulating heartbeat, like a pacemaker, and it could counteract heart attacks. Although human trials may be "a ways down the road," doctors and researchers recognize the significant potential of the technology.

The team is now looking at ways to dissolve the implant in the body once it is no longer needed and finding the optimal way to power the electrodes embedded in the device. They are also looking at opportunities to use the device to monitor other organs.

"I am currently pursuing various polymeric material systems that are stretchable and can be installed into living organs," Chung said.

"One can simply envision cell scaffolds or surgical adhesives from such an approach."

Chung notes that many of the key technologies from this research could also be adapted to industrial uses, such as wear-resistant coatings for drills.

"The next step will be to develop a novel processing pathway to fabricate non-conventional electronic devices," he said.

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**Melody® Transcatheter Pulmonary Valve  
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**Indications:** The Melody TPV is indicated for use in a dysfunctional Right Ventricular outflow Tract (RVOT) conduit ( $\geq 16$  mm in diameter when originally implanted) that is either regurgitant ( $\geq$  moderate) or stenotic (mean RVOT gradient  $\geq 35$  mm Hg)

**Contraindications:** None known.

**Warnings/Precautions/Side Effects:**

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

Potential procedural complications that may result from implantation of the Melody device include: 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, and pain at the catheterization site.

Potential device-related adverse events that may occur following device implantation include: stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, and hemolysis.

For additional information, please refer to the Instructions for Use provided with the product or call Medtronic at 1-800-328-2518 and/or consult Medtronic's website at [www.medtronic.com](http://www.medtronic.com).

**Humanitarian Device.** Authorized by Federal law (USA) for use in patients with a regurgitant or stenotic Right Ventricular Outflow Tract (RVOT) conduit ( $\geq 16$  mm in diameter when originally implanted). The effectiveness of this system for this use has not been demonstrated.

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