CONGENITAL CARDIOLOGY TODAY
Timely News & Information for Congenital/Structural Cardiologists & Cardiothoracic Surgeons Worldwide

International Edition
Vol. 22 - Issue 9

September 2024

Table of Contents

A Rare Case of Criss-Cross Heart with Ventriculo-Arterial Discordance with D-Transposed Aorta and Pulmonary Stenosis

Kalyan Munde, MD, DM; Anant Ramkishanrao Munde, MD, DM; Mohan Paliwal, MD, DM

Introduction

Congenital heart defects are present in about eight cases per 1000 newborns at term. Criss-cross heart anomaly is extremely rare, accounting for less than 0.1% of all congenital heart defects, not exceeding 8 per 1,000,000 births.[1] The morphological essence of the criss-cross heart is a rotation of the ventricular mass along its major axis.[1] This conformational change may be associated with any malformation described in cardiac segments, resulting in different relationships and connections between the atria, ventricles and great vessels.[2] Due to the complex structural changes and the rarity of the disease, this anomaly of rotation is often misdiagnosed due to lack of awareness of the medical team, bringing potential harm to appropriate surgical approach.[3] The failure to obtain a characteristic four-chamber view in any plane with transthoracic echocardiography is diagnostic for recognition of the criss-crossed atrioventricular junctions.[4]

Case Report

A 26-year-old male was referred to our clinic with complaints of dyspnea on exertion (NYHA class III) and easy fatigabity since two-years-old. He had underwent bidirectional Glenn shunt operation at the age of three-years-old and was asymptomatic thereafter. Now, his physical examination revealed delayed pulmonary component of second heart sound in the left second intercostal space. A 3/6 systolic murmur was best heard at right lower parasternal area. He had mild cyanosis. A characteristic four-chamber view in any plane with transthoracic echocardiography was not possible. Echocardiography demonstrated situs solitus with levocardia. The base of the heart (atria) remains unchanged in its spatial position, the ventricles appear to have been twisted along their longitudinal axis with atrio-ventricular concordance (right atrium emptying into left sided ventricle with right ventricular morphology and left atrium emptying into right sided ventricle with left ventricular morphology) in subcostal and apical four chamber views. His morphologic right ventricle was situated antero-superior and to the left in parasternal short axis view (PSAX). As well as, transposition of great arteries with dilated right anterior aorta arising from morphologic right ventricle and running parallel to hypoplastic pulmonary artery (arising from morphologic left ventricle) in basal PSAX. Also, patient had large inlet ventricular septal defect (VSD), atrial septal defect (ASD) and valvular and subvalvular pulmonary stenosis (PS) with a maximum pressure gradient of 124 mmHg. The morphologic right ventricle was dilated, hypertrophied and dysfunctional. The patient is currently receiving decongestive therapy for left ventricular (morphologic RV) failure.

SEPTEMBER 2024

International Edition Vol. 22 - Issue 9

TABLE OF CONTENTS

Discussion

This congenital defect was first described by Lev & Rowlatt[4] in 1961, but it was only in 1974 that Anderson et al.[5] first used the term criss-cross heart. The diagnosis of criss-cross heart is based on the intersection of the axes of the ventricular entries. In a normal heart these axes are virtually parallel. This condition is characterized by a spatial change of the ventricular mass that guides each ventricle in a contralateral position in relation to the corresponding atrium. While the base of the heart remains unchanged in its spatial position, the ventricles appear to have been twisted along their longitudinal axis. This promotes a change in hemodynamics characterized by crossing flows through the atrioventricular valves, resulting in the false impression that each atrium is being directed to the contralateral ventricle.[1]

The criss-cross heart may present with concordant or discordant atrioventricular and ventriculo-arterial connections. These connections were demonstrated in 1961 by Lev & Rowlatt[4] through the study of the anatomy of two hearts presenting atria in solitus position communicating with morphologically discordant ventricles in normal position. Another case of criss-cross heart was described by Van Praagh in 1962,[6] in which the morphologically right atrium connected to the morphologically right ventricle on the left side, in a case example with concordant atrioventricular connections.

There are cases of criss-cross heart described in the literature with discordant atrioventricular connections associated with transposition of the great vessels which results in a corrected physiological circulation. Patients with this type of anomaly (which represents 0.05% of Congenital Heart Diseases[7]) may be symptomatic not because of the criss-cross heart, but by the presence of other associated anomalies such as VSD, pulmonary outflow obstruction, tricuspid valve abnormalities.[8] A literature review revealed no cases of this anomaly occurring in isolation. Most patients have ventricular septal defects, transposition of the great arteries, double-outlet right ventricle, hypoplastic right ventricle, pulmonary stenosis, and tricuspid hypoplasia, the latter present in most patients. Other associated defects, although less frequent, are straddling mitral or tricuspid valve, subaortic stenosis, aortic arch obstruction and mitral stenosis.[1,8-10] Anomalies of the coronary circulation may be present and usually related to the ventricular position, and in these cases, magnetic resonance image (MRI) and angiography are useful tools in the diagnosis and approach.[10]

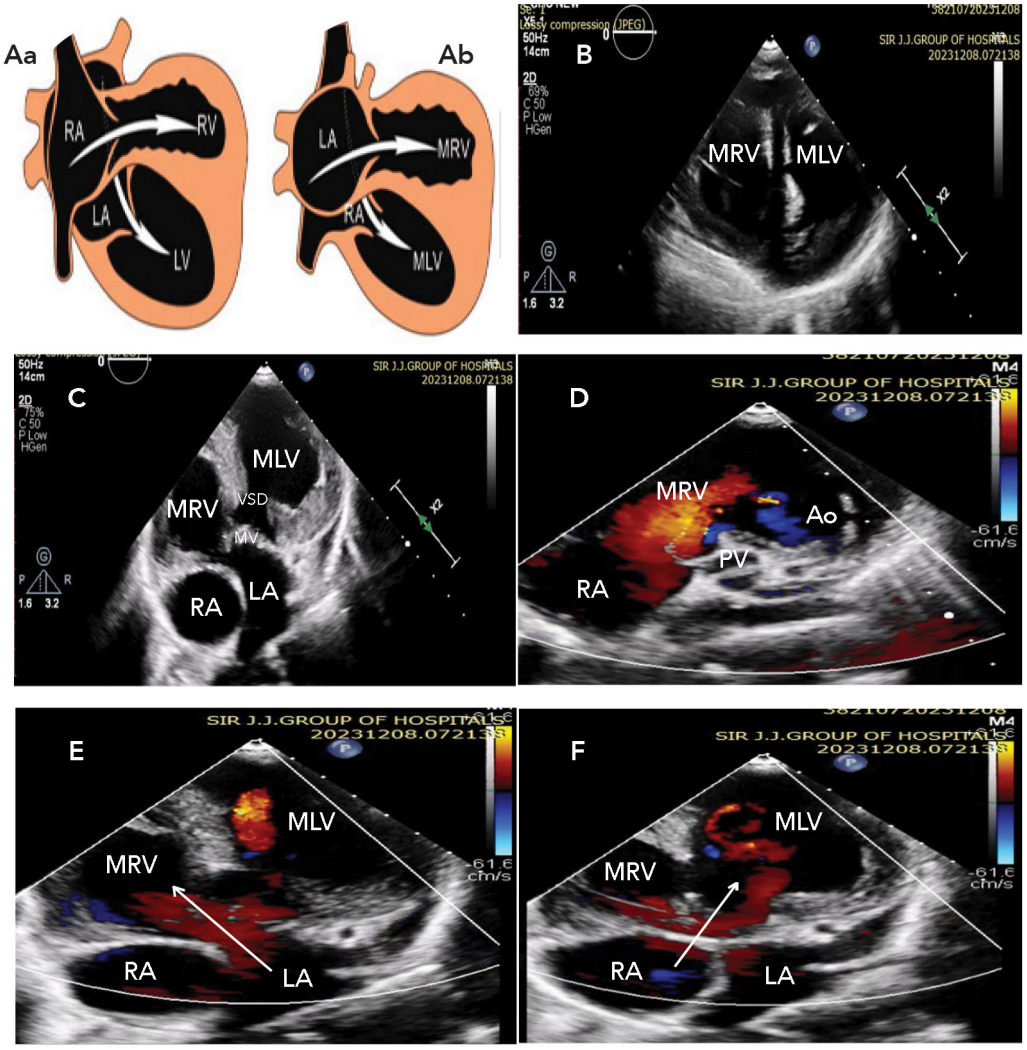


FIGURE 1 Criss-Cross Heart Illustration

Aa) *Schematic diagram of atrioventricular concordance with crisscross morphology.*
Ab) *Schematic diagram atrioventricular discordance with crisscross morphology.*
B) *Parasternal short axis view showing vertical orientation of the inter-ventricular septum and side-by-side ventricular relationship.*
C) *Echocardiography view of atrioventricular discordance with crisscross morphology.*
D) *Right atrioventricular inflow color Doppler image with dilated dextroposed aorta with bicuspid pulmonary valve of hypoplastic pulmonary artery.*
E) *Color Doppler image with arrow showing direction of blood flow from LA to MRV.*
F) *Color Doppler image with arrow showing direction of blood flow from RA to MLV.*
Abbreviations: LA, Left Atrium; RA, Right Atrium; MLV, Morphologic Left Ventricle; MRV, Morphologic Right Ventricle; MV, Mitral Valve; VSD, Ventricular Septal Defect; PV, Pulmonary Valve; Ao, Aorta.

In literature there are some studies linking the Cx43 gene mutation to pathogenesis of the criss-cross heart. Deletion of gene would result in a delay in establishing heart dextroposition, which makes the right ventricle to maintain a craniomedial position, resulting in a 90° rotation of the atrioventricular mass.[11] Reaume et al.,[12] in 1995, reported that animals with deletions in both alleles of Cx43 died shortly after

birth, with no major phenotypic differences compared to controls, except for cyanotic appearance. Necropsy revealed cardiac defects, thereby revealing vital importance of this protein in cardiac development during embryonic development.

The anatomic and physiologic diagnosis of this anomaly can be established by echocardiography, along with other diagnostic methods, such as MRI and cardiac catheterization if necessary.[13]

The transthoracic echocardiography can be used to identify the position and morphology of all cardiac chambers and AV valves, in addition to the connections between chambers and vessels. The subcostal window will determine the location of the heart apex and assess mainly the ventricles characteristics. The trabeculae morphological features, will determine the morphologic ventricle characteristics.[7] The great arteries connections are better visualized in the parasternal window[14,15] Echocardiographic findings include an inability to obtain a characteristic four-chamber view in any plane, displaying the crossing of long axes of atrioventricular valves as seen in the subcostal long axis or coronal plane sweep.[16,17] This anomaly can also be diagnosed prenatally with fetal echocardiography considering these findings.[18]

Studies with cardiac magnetic resonance imaging demonstrated clear visualization of the heart with criss-cross morphology and provided reconstructed three-dimensional images of the heart.[19]

Cardiac catheterization may be necessary only to obtain pressure and oximetry data and to rule out additional septal ventricular defects.[13] Some indications for the invasive study were assessment of pulmonary vascular resistance, angiographic analysis of coronary arteries, and presence of pulmonary valve atresia or pulmonary anomalous veins. [13]

Congenitally corrected transposition of the great arteries in criss-cross morphology was reported previously.[3,20-23] Our patient had a d-transposed aorta (right anterior aorta), described as "S,L,D" according to the Van Pragh notation system, which is a rare finding in ccTGA. To the best of our knowledge very few cases of criss-cross heart with corrected transposition with d-transposed aorta (right anterior aorta) have been reported to date. In a study by Allwork et al.,[24] of the 32 patients with ccTGA, two had a right-sided aorta. Symons et al.[3] reported a case with a criss-cross heart presenting with ccTGA and a d-transposed aorta. In another study carried out by Fang et al.,[17] of 10 patients, all had abnormal ventriculo-arterial connections including transposed in five and double-outlet right ventricle in the remaining patients. Fang et al.[17] reported the incidence of hypoplastic right ventricular and pulmonary stenosis in 30% and 60% of the patients, respectively. However, it is unclear that whether these are secondary or primary in the pathogenesis of a criss-cross heart. Straddling atrioventricular valve and double-inlet ventricle were also reported in the literature with a criss-cross heart.[17] Since it is almost always with other complex cardiac defects, patients usually present early in life with cyanosis, murmur or with heart failure symptoms, as seen in our patient for which he underwent Glenn shunt surgery. The clinical outcome of a criss-cross heart predominantly depends on the underlying hemodynamic abnormalities. Surgical treatment varies from palliative correction to definitive anatomic correction. In the majority of the patients with a criss-cross heart, a two-ventricle repair may not be possible and these patients are staged toward a Fontan-type operation.

Conclusion

A criss-cross heart is a rare and complex anomaly which should be kept in mind to recognize during echocardiography. Inability to obtain a characteristic four-chamber view in any echocardiographic plane is diagnostic for this anomaly in most cases.

References

1. Sanders, S. Crisscross Heart. Orphanet encyclopedia, Feb 2003. Disponível em <http://www.orpha.net/data/patho/Pro/ en/CrissCrossHeart-FRenPro8602.pdf> Acesso em 29 de fevereiro de 2012.
2. Cavellucci PMSM, Suaide CE, Ferreira LDC, Tandrafilov MD, Pimentel BDPA, Lerner M, et al. Entrecruzamento das vias de entrada ventriculares (criss-cross heart) e transposição ana-tomicamente corrigida das grandes artérias: relato de caso e revisão de literatura. Rev Bras Ecocardiogr Imagem Cardiovasc. 2012;25(4):292-7.
3. Symons JC, Shinebourne EA, Joseph MC, Lincoln C, Ho Y, Anderson RH. Criss-cross heart with congenitally corrected transposition: report of a case with d-transposed aorta and ventricu-lar preexcitation. Eur J Cardiol. 1977;5(6):493-505.
4. Lev M, Rowlatt UF. The pathologic anatomy of mixed levocardia. A review of thirteen cases of atrial or ventricular inversion with or without corrected transposition. Am J Cardiol 1961;8:216-63.
5. Anderson RH, Shinebourne EA, Gerlis LM. Criss-cross atrioventricular relationships pro-ducing paradoxical atrioventricular concordance or discordance. Their significance to nomen-clatura of congenital heart disease. Circulation. 1974;50(1):176-80.
6. VanPraagh R, VanPraagh S, Vlad P, Keith JD. Anatomic types of congenital dextrocardia: diagnostic and embryologic implications. Am J Cardiol. 1964;13:510-31.
7. Wallis GA, Debich-Spicer D, Anderson RH. Congenitally corrected transposition. Orphanet J Rare Dis. 2011;6:22.
8. Croti UA, Mattos SS, Pinto Jr VC, Aiello VD. Cirurgia cardiovascular pediátrica. 1ª ed. São Paulo:Roca;2008.
9. Gatzoulis MA, Webb G, Broberg C, Uemura H. Cases in adult congenital heart disease. Complex congenital heart conditions. 2010;1:278-80.
10. Angelini P, Lopez A, Lufschanowski R, Nemeth MA, Flamm SD. Coronary arteries in crisscross heart. Tex Heart Inst J. 2003;30(3):208-13.
11. Ya J, Erdtsieck-Ernste EBHW, Boer PAJ, Van Kempen MJA, Jongsma H, Gros D, et al. Heart defects in connexin 43-deficient mice. Circulation Research 1998, 82:360-6.
12. Reaume AG, Sousa PA, Kulkarni S, Langille BL, Zhu D,

Davies TC, et al. Cardiac mal-formation in neonatal mice lacking connexin43. Science. 1995;267(5205):1831-4.
13. Robinson PJ, Kumpeng V, Macartney FJ. Cross sectional echocardiographic and angiocar-diographic correlation in criss cross hearts. Br Heart J. 1985;54(1):61-7.
14. Carminati M, Valsecchi O, Borghi A, Balduzzi A, Bande A, Crupi G, et al. Cross-sectional echocardiography study of criss-cross heart and superoinferior ventricles. Am J Cardiol. 1987:59(1):114-8.
15. Attie F, Zabal C, Buendia A. Cardiologia pediátrica diagnóstico y tratamiento. México. 1993;1:353-62.
16. Yang YL, Wang XF, Cheng TO, Xie MX, Lü Q, He L, et al. Echocardiographic characteristics of the criss-cross heart. Int J Cardiol 2010;140:133-7.
17. Fang F, Li ZA, Yang Y, Zheng CH, Lam YY. Deciphering the mysteries of crisscross heart by transthoracic echocardiography. Echocardiography 2011;28:104-8.
18. Ayabakan C, Binnetoğlu K, Sarısoy Ö, Tokel K. Crisscross heart with tricuspid atresia diagnosed in utero. Congenit Heart Dis 2013;8:153-6.
19. Ming Z, Yumin Z. Magnetic resonance evaluation of crisscross heart. Pediatr Cardiol 2008;29:359-65.
20. Taksande AM. Echocardiographic recognition of a crisscross heart with double outlet right ventricle. Images Paediatr Cardiol 2013;15:3-7.
21. Yadav S, Haranahalli PE, Shukla M, Varma CM. Criss-cross heart with atrial inversion and juxtaposed atrial appendages: an echocardiographic study. Turk Kardiyol Dern Ars 2013;41:564.
22. Muneer PK, Kalathingathodika S, Chakanalil GS, Sony MM. Crisscross heart with dextrocardia and intact interventricular septum. Ann Pediatr Cardiol 2014;7:70-1.
23. Martínez Quintana E, Rodríguez González F, Agredo Muñoz J. Criss cross heart in a congenitally corrected transposition of the great arteries. Int J Cardiol 2008;130:81-2.
24. Allwork SP, Bentall HH, Becker AE, Cameron H, Gerlis LM, Wilkinson JL, et al. Congenitally corrected transposition of the great arteries: morphologic study of 32 cases. Am J Cardiol 1976;38:910-23.

KALYAN MUNDE, MD, DM

Professor, Department Head, Cardiology
Grant Government Medical College &
Sir J.J. Group of Hospitals
Mumbai, Maharashtra, India
kalyanmunde@yahoo.com

ANANT RAMKISHANRAO MUNDE, MD, DM

Assistant Professor, Cardiology Department
Grant Government Medical College &
Sir J.J. Group of Hospitals,
Mumbai, Maharashtra, India
anantmunde09@gmail.com

MOHAN PALIWAL, MD, DM

Corresponding Author
Senior Resident, Cardiology Department
Grant Government Medical College &
Sir J.J. Group of Hospitals
Mumbai, Maharashtra, India
drmohanpaliwal@gmail.com

Matters of The Heart and Mind: Bad Decisions Make Good Experience, or is it… Bad Experience Leads to Good Decisions?

Neil Wilson, MBBS, DCH, FRCPCH, FSCAI

Who would have thought Mark Twain was an interventional cardiologist? I think we have all learned from his take on decision and experience theory. I adjust his quotes only slightly when I say at meetings with nightmare type sessions that 'Bad experience leads to good decisions…or is it…bad decisions lead to good experience'? That 'I will not do that again' ia a feeling you occasionally get in the catheter lab. Certainly same old same old decisions can breed familiarity and indolent thinking, though please note I am not advocating change just for the sake of it. But here is a funny story. Talking of same old, same old decisions…

Two mornings ago, I tried to brush my teeth with shaving cream. I did not actually get to the brushing stage. I absent mindedly had put shaving cream on my toothbrush but recognised the folly before plunging the brush into my mouth. Now why on earth would I do that? The tube of toothpaste and the tube of shaving cream sit upright on the basin. They are different colours. The toothpaste tube is white (of course) and the toothpaste is white. On the other hand, the shaving cream tube is morphologically identical except blue. The shaving cream itself is white. Obviously, I have dispensed and used the said tubes, toothbrush, and razor thousands of times and until two days ago had never tried to shave using toothpaste or brush my teeth using shaving cream. So, all those thousands of appropriate actions and then two days ago folly. Perhaps a demonstration that if it is possible to make a bad decision without real provocation or consequence then sure enough when the chips are down, we can do the same. What on Earth is the old timer going on about?

'That device is going nowhere' I said confidently about fifteen years ago as the very competent senior registrar had positioned an Amplatz duct device in a slightly 'kinked' medium sized duct in a two year old child. With the device still attached to the cable an aortogram showed that there was a puff of residual flow over the superior aspect of the device. "Give it a tug," I said. Tug performed. "Looks OK to me." "Shall I release?" says the senior registrar. "You make the decision and I'll support whichever decision you make," I said, supremely confident that all would be well. Well, he did release…upon which the device dislodged and did a triumphant cartwheel briefly into the main pulmonary artery and then flew into the mid-right pulmonary artery. Of course, this being a busy day and only the first case as a warmer upper of trickier cases ahead, the device settled with the attachment screw pointing distally. Sure, we got it out eventually and even used and repositioned the same device but that's not the point. Nobody, particularly me, felt anything other than an idiot. I should have paid a bit more attention to the kink in the duct, the residual shunt, I should have perhaps used an AVP II, should have done an echo. Should not have been impatient wanting to get on with the next case. You know, the could have/would have/should have feelings. That release was a bad decision, but led to a good experience, not least of all --the experience of retrieving duct devices from the RPA when the screw is pointing in the wrong direction! Turning the device over so the screw is accessible and then snaring the tip of the screw for retrieval through the sheath. But the real experience is the point above about the decision to release with residual over the superior margin of the device. And there is more…

But hey, we have all had devices embolise. Nightmare case sessions at conferences almost always have a presentation with a device of some sort doing a walk about in the circulation. In the days of the popular use of Gianturco coils for occluding ducts, it was relatively common that multiple coils were required. They were

sometimes positioned separately, releasing one before inserting another, often begging the question how much of a residual puff of flow on an angiogram you would tolerate before deciding another coil was required. Some operators chose to position coils simultaneously with separate delivery cables before release, creating a 'nest.' Not a bad idea.

Thirty years ago I had an entertaining case of embolization using two 5mm coils to close a duct. First coil in fine, released, angiographic residual flow which was predictable, (we were anticipating using two or even three coils), so a second coil was positioned and released. Hey, presto! In a heartbeat, one coil embolised the RPA and the other to the LPA. Bad experience. Retrieval uncomplicated, but certainly extended fluoroscopy time. Better decision next time?

There is even more… at about the same time, I gained good experience which led me to learn that I would do all I could to ensure there was complete occlusion of the duct on angiography when using coils to close a duct. This bad experience I am about to describe which, subsequently led to me making good decisions in the future involved an adult lady of 63 from a small town in a remote area of Northern, Scotland, an area of outstanding natural beauty you would pay money to go and vacation there. Did I mention there is a selection of outstanding whisky distilleries in the same area? To add insult to injury, the patient was a doctor, family practitioner. Now you know this is going to be a story... I can not quite remember how the duct was diagnosed. Certainly, she had no symptoms that you could attribute to a small duct. I seem to remember she had a mild anaemia (clue). Down to Glasgow she comes for an interventional closure. I think this was the first duct in an older person (60+) I had closed. I did note heavy calcification in the wall of the duct, but morphologically, it funneled nicely symmetrically and was a slam dunk for a 5mm coil delivered from the arterial side. There was a puff of residual flow on repeat angio. "That's definitely going to close completely in the next hour or two!" I confidently announced to the lab. The next morning, Echo showed a tiny jet of residual flow. Well, it probably just needs another day or two I conclude. The patient goes home back to the North.

A few days later I get a lovely thank you letter from her. A big 'Thank You' to the team. Very kind. A few weeks later, I get another letter, this time, from her cardiologist in Inverness. My patient has (have you guessed yet?) a haemolytic anaemia. Haemoglobin 7.6 g/dl. Of course, calcium, stainless steel, high velocity narrow jet. I should have thought about that but, (but there is always an excuse) this was the fist calcified duct I'd come across for interventional closure. It was a bad decision to leave even that tiny residual flow, smashing red cells into the calcium & stainless steel interface. Idiot Wilson (yet again).

So, this is now an unforgettable bad experience leading to a new good decision not to leave any residual flow with coils and calcium in juxtaposition. Thankfully, the second coil did the trick for complete closure. Home. Two more letters. A second 'Thank You' and a few weeks later an anxiety relieving letter from the cardiologist… Haemoglobin up to 11.4 g/dl.

Back to Mark Twain… is it a bad decision leading to good experience or is it a bad experience leading to good decisions? You choose. Good luck with your own decisions. You make them and I will support whichever decisions you make.

NEIL WILSON, MBBS, DCH, FRCPCH, FSCAI

Formerly Professor of Pediatrics
University of Colorado School of Medicine
Formerly Director Cardiac Catheter Laboratory Children's Hospital Colorado
Colorado, USA
Neilwilson1955@icloud.com

The SickKids Advanced Cardiology Education (ACE) Program — September 6th, 2024

Alyssa Gumapac, BHSc.

The SickKids Advanced Cardiology Education (ACE) Program, https://cvent.me/Mg2aaw, offers a unique opportunity for healthcare professionals worldwide to benefit from the groundbreaking advancements at the Labatt Family Heart Centre at the Hospital for Sick Children (SickKids).

Over the past nine years, the SickKids ACE Program has attracted healthcare professionals from six continents. This initiative fosters global collaboration through live webinars, real patient case discussions, and peer-to-peer debates on ethical issues in pediatric cardiology, creating a worldwide community dedicated to improving pediatric heart health outcomes.

The 33-week online curriculum, https://cvent.me/eDnZyL?RefId=Sessions, is delivered by over 100 esteemed experts from SickKids to healthcare professionals globally, ranging from early career to novice physicians, nurse practitioners, nurses, respiratory therapists and more. This diverse audience enriches the learning experience for everyone involved. Here's what some participants have shared about their experiences:

Meet Dr. Megha Unadkat MD, MMED MsC

In 2022, Dr. Megha Unadkat was completing her pediatric cardiology fellowship at the Jakaya Kikwete Cardiac Institute, the only cardiac center in East Africa.

Despite the challenges faced by local setups like Jakaya Kikwete, ranging from limited human and financial resources, Dr. Unadkat valued the exposure to the latest innovations in pediatric cardiology that complemented her fellowship:

"There's more to cardiology than just performing echocardiograms and administering anti-heart failure medication. [The ACE Program] integrated seamlessly with my fellowship. Even though my fellowship faces challenges due to limited resources, [the program] introduced me to international advancements, the latest drug trials, and advanced heart failure management. I gained insights from both perspectives, which made learning about pediatric cardiology ideal. I was surprised by how advanced pediatric cardiology is at SickKids and other parts of the world compared to here—such as available drugs and advanced equipment like VADs, ECMOs, and heart transplants, which I've never seen here."

Dr. Unadkat also appreciated the flexibility of the online format. Despite the 7-hour time difference from Tanzania, she leveraged online resources to understand complex concepts and build a global network:

"These are challenging concepts, but the course's repeated reinforcement and access to recordings and slides made them easier to grasp over time. Even though the course was online, I made friends, participated in group presentations with people from around the world, and even found a mentor—I had never experienced that before."

While the ACE Program equips healthcare professionals with technical, collaborative, and cognitive skills through live webinars, Dr. Unadkat highlights how it also broadened her focus to include affective skills. She now feels more confident in managing children with heart disease and communicating with patients and families:

"A case that particularly stood out to me was on 'Long QT Syndrome.' It was fascinating because I hadn't encountered such a patient before. Initially, I struggled to understand it, but the presentation made it clear. Now, if I encounter such a patient, I feel equipped to read the ECG, diagnose, and manage the condition effectively! It can be challenging in East Africa, as the only cardiology site, with many patients and occasional lapses in communication with parents. The course emphasized the importance of compassionate care, patient counseling, and genetic counseling, which significantly impacts patient outcomes. Now that I know more, I want more for my patients and want to save more lives."

Today, Dr. Megha Unadkat is a Pediatric Cardiologist at the Jakaya Kikwete Cardiac Institute. She remains passionate about bridging the gap in Tanzania's cardiology services and improving health outcomes. The ACE Program team had the privilege of meeting Dr. Unadkat in person at the 8th World Congress of Pediatric Cardiology and Cardiac Surgery 2023, where she presented posters on "Age of Diagnosis and Timing of Intervention in Children with Truncus Arteriosus at the Jakaya Kikwete Cardiac Institute, Dar Es Salaam, Tanzania" and "Incidental Diagnosis of Large Aortopulmonary Window Post Patent Ductus Arteriosus Ligation: A Case Report." Recently, she was involved in the first-ever replacement of a stenotic pulmonary valve using the Melody valve (bovine jugular vein) in

Tanzania, East, and Central Africa—a significant milestone for the Jakaya Kikwete Cardiac Institute.

Meet Annette Klingmann, NP

Annette completed the ACE Program in 2021 while working as a Clinical Nurse Specialist in the Anesthesia and Intensive Care Unit at the University of Heidelberg, where she has been employed since 1998.

Annually, her unit treats 4,000 to 5,000 patients in the outpatient clinic and 800 to 900 in the inpatient unit, performing over 2,000 surgeries, including 250 on infants. Despite her extensive experience in pediatric cardiology, Annette found the ACE Program to be an invaluable source of new perspectives:

"The program was appealing due to its broad range of topics—from general heart disease in children to cyanotic lesions. Germany has many small centers and fewer cases compared to Toronto, so I learned a lot. I also discovered differences in therapies compared to Europe. For instance, Ventricle Assist Devices (VADs) are used here only in children aged 6 and older, whereas I learned about their use in infants. The program also covered ECG interpretation, heart murmurs, and heart sounds, areas usually handled by physicians rather than nurses in Germany. Learning about ECMO, pacemaker therapy, and the pathology series was incredibly interesting!"

Annette also found the case presentations and collaboration with international colleagues enriching:

"I presented a case from our hospital—a Single Ventricular patient—and learned a lot from the class. Collaborating with colleagues from the Emirates and England provided valuable insights into different approaches. I returned to work and discussed pharmacological differences with my senior doctor and came away with new ideas!"

Like Dr. Unadkhat, Annette appreciated the online format, interactivity, and ability to rewatch lectures:

"There were numerous interactive activities, polls, and breakout groups, and the communication from Carrie and Cecilia was excellent. I always tell my colleagues about the program. The topics are well-chosen, the speakers are outstanding, and I look forward to Fridays. Although the time shift sometimes made the last few lectures tiring, I could listen to the recordings later when I had time."

Annette, a seasoned nurse, acknowledges the nuances of caring for children with heart disease and the need for improved interprofessional collaboration and patient communication in her center:

"It's interesting to see engagement from the interdisciplinary team. When congenital heart disease is diagnosed, there's extensive instruction for parents and assistance from social workers. Here, parents often only speak to physicians, and the process isn't as organized as in Canada. The course provided great input on how we can improve. Nurses are with parents most of the time, and I can apply these lessons to enhance communication in my ward."

Despite being on opposite sides of the world, both Dr. Unadkat and Annette highly recommend the ACE Program:

"If you have any interest in cardiology, go for it! I know it's intense and time-consuming, but it's worth it. It's not only about learning cardiology but also engaging with people, learning about the latest drug trials, devices, and interventions. Even if you don't apply everything now, it will be valuable in the future." — Dr. Megha Unadkat

"You must take the program! I always tell my colleagues, especially physicians. I've been working a long time in my ward and suggest changes, but it's more impactful when physicians hear things from other physicians." — Annette Klingman, NP

You can join many like Dr. Unadkat and Annette this fall. It's not too late to register! Semester 1 begins on **September 6, 2024**. Registration will remain open passed the Semester 1 start date.

Visit our website to register or for more information:
https://cvent.me/dknaYG

For inquiries, email: ace.program@sickkids.ca

Quotes may have been edited for brevity and grammatical correctness.

First-Ever Successful Use of Modified Double-Decker Technique for Scimitar Syndrome in a Child

Scimitar Syndrome, a rare congenital heart disease, involves an anomalous pulmonary venous return where the right pulmonary veins return to the inferior vena cava instead of the left atrium. It is mainly diagnosed in infants, with an estimated prevalence of 1–3 per 100,000 births. Delayed treatment can lead to pulmonary hypertension, right heart failure, respiratory failure, heart arrhythmia, and growth disorders.

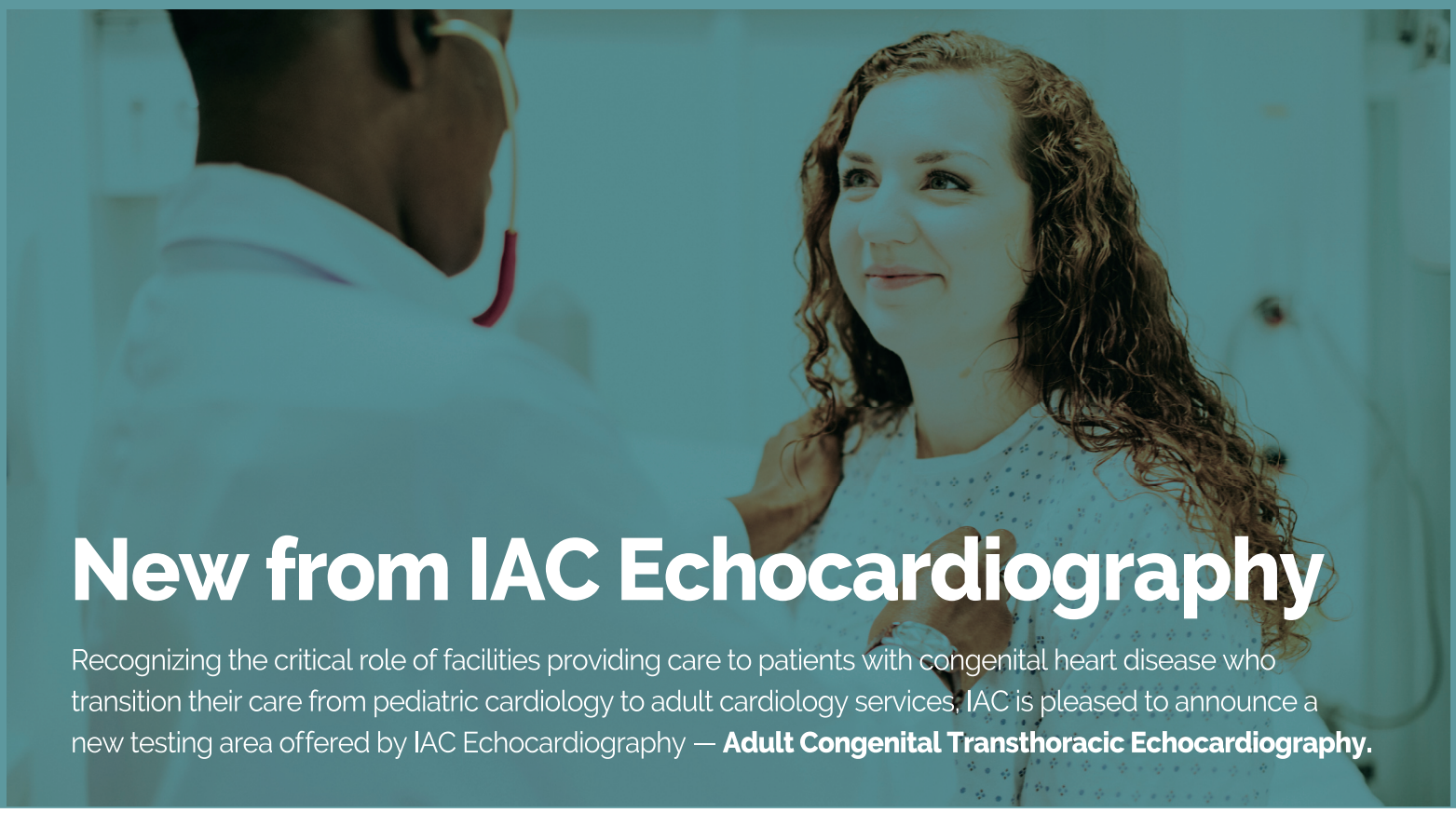
This syndrome is characterized by anomalous pulmonary venous drainage to the inferior vena cava, and the usual surgical repair involves re-implanting the right pulmonary veins (scimitar vein) to the left atrium or creating an intra-atrial tunnel to redirect the scimitar vein to the left atrium. However, these methods have a critical problem of postoperative pulmonary vein obstruction. If this occurs, it can lead to severe pulmonary venous congestion and subsequent hemoptysis. In such patients, the success rate of re-intervention for pulmonary venous obstruction is very low.

We have just performed an unprecedented surgical procedure on a two-year-old child diagnosed with scimitar syndrome. The procedure was the world's first successful application of the "Double-decker Technique" used to repair another type of partial anomalous pulmonary venous return. This new procedure, a modified double-decker technique for scimitar syndrome, uses no artificial material and reconstructs two blood flow pathways (right pulmonary vein and inferior vena cava) using only the patient's atrial wall. This novel surgery was conducted by a pediatric cardiac surgery team led by Senior Professor Genichi Sakaguchi from the Department of Cardiovascular Surgery, Kindai University Hospital, Affiliate Professor Shinichiro Oda from the Department of Cardiovascular Surgery, Kindai University Hospital, and Associate Professor Satoshi Asada from the Department of Cardiovascular Surgery, Kindai University Hospital, University Hospital in Osakasayama, Japan.

The patient was referred to our hospital for suspected congenital heart disease from another public hospital after developing a fever. The patient was diagnosed with scimitar syndrome by cardiac echocardiogram and contrast-enhanced computed tomography. After the surgery, there were no problems with the reconstructed blood flow pathways in the right pulmonary veins and inferior vena cava. The patient was discharged from our hospital 10 days after the surgery without any postoperative complications.

The advantage of this new technique is that the new blood flow pathway of the inferior vena cava is created outside (on the pulmonary venous pathway). This technique can create wide pathways separately and reduces the risk of obstruction. The conventional intra-atrial tunneling divides the inferior vena cava into two pathways for the right pulmonary vein and the inferior vena cava. That is why the conventional technique is likely to create narrower pathways and develop obstructions. In addition, the surgical site is expected to grow following the patient's somatic growth because these pathways were reconstructed by the pedicled atrial wall without any artificial material.

The success of this surgery can make this a common surgical technique for scimitar syndrome, and surgical outcomes for this rare disease are expected to be improved in the future.



New from IAC Echocardiography

Recognizing the critical role of facilities providing care to patients with congenital heart disease who transition their care from pediatric cardiology to adult cardiology services, IAC is pleased to announce a new testing area offered by IAC Echocardiography — **Adult Congenital Transthoracic Echocardiography.**

IAC provides a unique approach to accreditation and is leading the field with innovative, customized solutions for your facility. IAC provides quality improvement-focused solutions such as the IAC QI Self-Assessment Tool, to help facilities optimize processes and improve patient safety and outcomes.

In addition to the testing area for Adult Congenital Transthoracic, **IAC offers accreditation specific to Echocardiography in the following areas:**

Adult Transthoracic
Adult Transesophageal
Adult Stress
Adult Congenital Transthoracic *(New!)*
Pediatric Transthoracic
Pediatric Transesophageal
Fetal
Perioperative Transesophageal *(New!)*

ACCREDITED
IAC
INTERSOCIETAL ACCREDITATION COMMISSION
FACILITY

Now Available! IAC Heart and Vascular Center of Quality Designation

The IAC Center of Quality designation recognizes health care organizations that demonstrate an ongoing, multi-disciplinary integration of diagnostic imaging and intervention-based procedures. Their commitment to the patients they serve throughout the continuum of care is exemplary. Learn more at **intersocietal.org/centerofquality**.

IAC
Improving health care through accreditation®
intersocietal.org | 800.838.2110

Contact IAC today or visit **intersocietal.org/echo**.

Children's Colorado Accepted into the Pediatric Heart Network

Children's Hospital Colorado (Children's Colorado) announced its acceptance into the Pediatric Heart Network (PHN), a collective of leading hospitals working to improve outcomes and quality of life for children – and more recently adults – with heart disease. The hospital's Heart Institute will become one of nine clinical research centers across North America selected to be a part of this national network.

"We are thrilled to be accepted as a new core site for the Pediatric Heart Network. As one of only eight stand-alone pediatric cardiology research centers in the country, we are uniquely positioned to ensure that children and young adults with heart disease in the Midwest and Rocky Mountain regions have equitable access to these important research studies," shares Shelley Miyamoto, MD, Professor and Jack Cooper Millisor Chair of Pediatric Cardiology and Co-Director of the Heart Institute.

The goal of the PHN is to bring cutting-edge treatments, trials and studies to patients through the collaboration of the nine participating hospitals. Over the next seven years, the selected congenital heart centers will work together to conduct multisite research in pediatric and congenital heart disease projects, bringing even more leading and innovative trials, studies and treatments to patients treated at these sites.

Funded by the National Heart, Lung and Blood Institute of the National Institutes of Health (NIH), the PHN will provide $2.4 million over the term of the award to Children's Colorado, which was selected as a core site in the Gateway to the West consortium with Washington University School of Medicine in St. Louis. As such, Children's Colorado pediatric leaders will participate in all PHN-led clinical studies and serve on the executive committee and all other PHN committees.

The network aims to improve health outcomes in children with heart disease – particularly congenital heart disease –and more recently, in adults, by unifying an at-times fragmented congenital heart disease research community. Over the past 22 years, the multicenter collaborative effort has supported 25 large studies, including 10 clinical trials, adding treatments and improving care for pediatric heart disease patients.

The application process, to be a part of the PHN, is a competitive and rigorous endeavor that spanned six months. The application process began in January of 2023. Awardees had to demonstrate a strong institutional commitment to pediatric cardiovascular research and education, the infrastructure to participate and lead multicenter clinical trials, and a broad range of expertise among study team members. In addition, each site had to develop and pitch a study proposal, which is currently being considered for the next phase of clinical studies.

"We are confident that our exceptional outcomes and team members were key to securing this award," Miyamoto said. "We are thrilled to be able to have even more collaboration and information from other PHN hospitals and to be able to offer even more valuable resources and options to our patients."

Leading the new core site alongside Children's Colorado's Dr. Miyamoto is Emily Bucholz, MD, PhD, Assistant Professor of Pediatrics and a Fetal Cardiologist, in collaboration with Washington University's Andrew C. Glatz, MD, the Louis Larrick Ward Professor of Pediatrics, and Jennifer N. Silva, MD, a Professor of Pediatrics and of Biomedical Engineering. The two centers will work together as the Gateway to the West consortium, with regular virtual and in-person meetings, as well as a shared plan to enhance diversity in research participants, and a shared mentorship model to train the next generation of pediatric cardiology clinical investigators.

SEPTEMBER

04TH-07TH

PICS 2024
San Diego, California, USA
https://www.picsymposium.com/

05TH-07TH

PEDS Cardio AI Conference
Austin, Texas, USA
https://cvent.utexas.edu/event/13c4218a-9814-431e-b116-b3661d7adea6/summary

06TH-08TH

Annual PICS Fellows & Early Career Course
San Diego, California, USA
https://register.rcsreg.com/r2/pics2024/fellow/top.html

OCTOBER

04TH-06TH

CSI ASIA-PACIFIC 2024
Bangkok, Thailand
https://www.csi-congress.org/asia-pacific

04TH-05TH

Tips and Tricks in Congenital & Structural Interventions
Milan, Italy
https://www.victoryproject.it/Pdf/654_TIPS_AND_TRICKS_PROGRAM.pdf

NOVEMBER

07TH-10TH

Great Wall International Congress of Cardiology 2024
Beijing, China
http://www.gw-icc.com/en

CONGENITAL CARDIOLOGY TODAY

CORPORATE OFFICE
PO Box 52316
Sarasota, FL 34232 USA

CORPORATE TEAM

EDITORIAL BOARD

OFFICIAL NEWS & INFORMATION PARTNER OF

PICS Society
Pediatric and Congenital Interventional Cardiovascular Society

ISSN 1554-7787 print. ISSN 1554-0499 electronic.
Published monthly.