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Table of Contents

Unroofed Coronary Sinus After Device Closure of a Secundum Atrial Septal Defect in a Pediatric Patient

Kevin Delzepich, BS; Lake Ernst, BS; Jackson Beeching, BS; Randy Richardson, MD; Daniel Miga, MD; Deepa Prasad, MD

Introduction

This case highlights a rare congenital cardiac anomaly involving a secundum atrial septal defect (ASD) combined with an unroofed coronary sinus (URCS). URCSs are exceedingly rare and difficult to detect via non-invasive echocardiography, particularly in the presence of another larger anatomical defect. We present a case involving a URCS not detected until follow-up for successful device closure of a secundum ASD closure in a pediatric patient.

Case Presentation

A three-year-old female presented for evaluation following a syncopal episode. Initial evaluation, including echocardiography, revealed a moderate-sized fenestrated ASD with mild right atrial and ventricular enlargement. An ECG demonstrated right ventricular hypertrophy. She was referred for ASD device closure. A Transesophageal Echocardiogram (TEE) revealed a moderate secundum ASD (~8–11 mm) with left-to-right shunting, mild right atrial and ventricular enlargement, and no significant valve abnormalities. The patient then underwent successful transcatheter device closure using an 11mm Amplatzer Septal Occluder device. Post-procedure TEE demonstrated the ASD device in good position with no residual shunting. At her one-month follow-up, TTE confirmed the correct stable ASD device position, but with a possible residual inferior atrial septal defect. During a six-month cardiology follow-up, the patient presented with complaints of irregular breathing

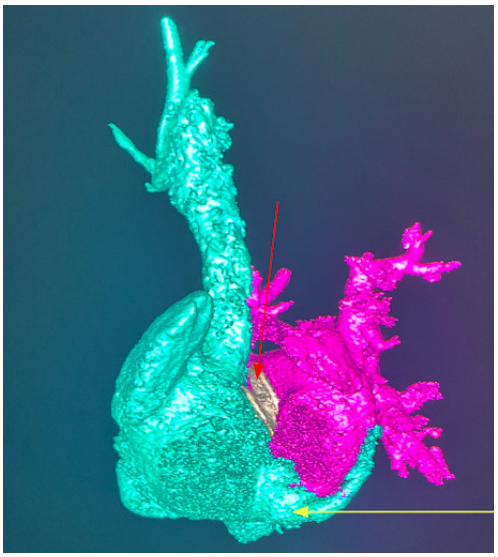

FIGURE 1 3D Blood Volume Image of the Heart

The Coronary sinus is seen directly communicating with the left atrium. The yellow arrow indicates the unroofed coronary sinus. The red arrow indicates the 11mm Amplatzer Septal Occluder device.

AUGUST 2025

International Edition Vol. 23 - Issue 8

TABLE OF CONTENTS

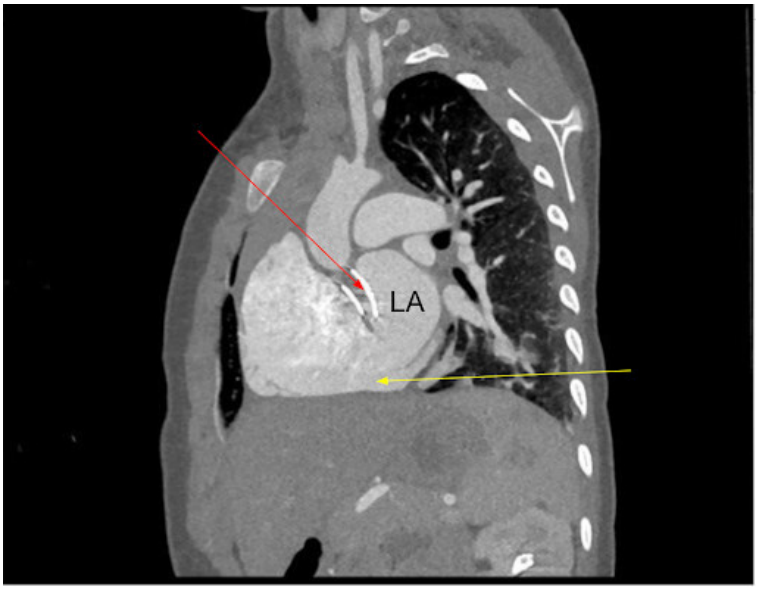

FIGURE 2 Sagittal CT Image with Contrast

The coronary sinus is seen directly communicating with the left atrium. The yellow arrow indicates the unroofed coronary sinus communicating with the left atrium (LA). The red arrow indicates the 11mm Amplatzer Septal Occluder device.

and sleep fits. Her cardiovascular exam was normal with no murmurs. A follow-up TTE demonstrated a persistent moderate-sized inferior defect. A subsequent TEE confirmed a dilated coronary sinus ostium with communication between the left and right atria, indicative of a URCS. Cardiac CT with contrast corroborated these findings, showing significant right atrial and ventricular dilation caused by a URCS. The patient is scheduled for corrective surgery to address the unroofed coronary sinus.

Discussion

ASDs are among the most common congenital heart defects accounting for 10-15% of cases in the United States. Secundum ASDs account for 75% of all ASDs, while unroofed coronary sinuses are exceedingly rare (<1%). In an ASD, there is an abnormal opening in the atrial septum, the wall that separates the right and left atria of the heart. This allows oxygen-rich blood from the left atrium to flow into the right atrium, causing an increase in blood flow to the right side of the heart and the lungs. Over time, this abnormal flow can lead to complications such as pulmonary hypertension, arrhythmias, or heart failure if left untreated.

URCSs are rare congenital defects in which the roof of the coronary sinus, a large vein that drains blood from the myocardium into the right atrium, is absent or poorly developed. This results in an abnormal communication between the coronary sinus and the right atrium. In some cases, URCS can be associated with other congenital defects, including ASD, and can lead to a left-to-right shunt of blood and subsequent right-sided heart dilation. Symptoms of URCS may vary depending on the size and associated defects. Small URCS defects may be asymptomatic, while larger defects can cause issues such as cyanosis, heart failure, or arrhythmias.

A URCS coexisting with an ASD is remarkably rare and complicates diagnosis, as the dominant defect often masks the presence of the secondary anomaly. This case illustrates the diagnostic limitations of less-invasive techniques such as TTE in identifying URCS, especially when there are other dominant heart defects. High-resolution imaging modalities like TEE and cardiac CT are invaluable in diagnosing complex congenital anomalies as they have superior visualization of atrial structures.

Conclusion

This case underscores the importance of high-resolution imaging techniques in diagnosing and managing rare congenital heart defects. While TTE serves as a first-line modality, clinicians should maintain vigilance for less apparent defects, such as URCS, when encountering fenestrated ASDs. The use of TEE and CT, as opposed to TTE, may yield higher fidelity imaging of URCSs, which may improve diagnostic accuracy and management of Coronary Heart Disease.

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Congenital Cardiac Research Collaborative Recognizes Jeffery J. Meadows for Outstanding Leadership

Christopher J. Petit, MD; Andrew C. Glatz, MD MSCE; Athar M. Qureshi, MD; Jeffrey D. Zampi, MD; Nelangi Pinto, MD; Courtney E. McCracken, PhD; Rachel Weber; Bryan H. Goldstein, MD

The Congenital Cardiac Research Collaborative (CCRC) is proud to announce Dr. Jeffery J. Meadows has been recognized as the inaugural recipient of the CCRC Leadership Award.

The CCRC was established in 2013 with the goal of advancing multicenter collaborative clinical research in populations at risk for disappointing outcomes after congenital heart disease (CHD) interventions. Originally comprised of four centers, the CCRC has grown in phases over time to now include 16 centers. The first expansion in 2017 welcomed Dr. Jeffery Meadows and the University of California San Francisco (UCSF) into the CCRC.[1] Immediately upon joining the CCRC, Dr. Meadows brought his iconoclastic style, unique perspectives, and exacting approach to bear upon our organization.

Dr. Meadows' impact on the administrative and organizational structure of the CCRC has been profound. He led initiatives to improve inclusivity and transparency for new members with a focus on early career mentorship. "Jeff has always recognized that the value of our collaborative is measured not by our publications or abstracts, but by the collective energy and enthusiasm of the group and ultimately the friendships that bind us," notes Chris Petit.

Dr. Meadows' contributions to CCRC's science have been impressive. Perhaps the most impactful of the studies he led include evaluations of outcomes in young children with single ventricle CHD who are palliated with either ductal artery stents or systemic to pulmonary surgical shunts.[2] But he also led studies evaluating outcomes following palliative approaches for newborns with pulmonary atresia,[3] and a recently completed study evaluating the late outcomes in children born with symptomatic tetralogy of Fallot.[4]

CCRC Steering Committee recognizes Dr. Jeffery Meadows (from left): Abhay Divekar, Dana Boucek, Nelangi Pinto, Shawn Batlivala, Allen Ligon, Jeffrey Zampi, Christopher Petit, Shiraz Maskatia, Ryan Callahan, Athar Qureshi, Brian Morray, Michael O'Byrne, Bryan Goldstein, Jeffery Meadows, Allison Meadows, Ravi Birla, Tacy Downing, George Nicholson, Abby Waldon, Dominic Zanaboni, Jeannette Wong-Siegel, Mark Law, Yun Zhang, Andrew Glatz, Rachel Weber, Andrea Les

This year, the CCRC unanimously voted to recognize Dr. Meadows for his outstanding leadership and contributions to our group. We presented him with the CCRC Leadership Award this May during our biannual CCRC Steering Committee Meeting in Houston, Texas – hosted by Dr. Athar Qureshi at Texas Children's Hospital. Unbeknownst to Jeff, the organizers of the meeting arranged to have his wife, Dr. Allison Meadows, fly in for the ceremony. The award presentation (pictured below) was a special moment for all of us as we celebrated all that Dr. Meadows has accomplished within the CCRC over the last eight years.

In ensuing years, this award will heretofore be named the "Jeffery J. Meadows' CCRC Leadership Award" and given to a member of,

Founding members of the CCRC (from left to right): Andrew Glatz, Athar Qureshi, Christopher Petit, and Bryan Goldstein (far right), along with Jeffery Meadows (holding award) and Allison Meadows

CCRC President Christopher Petit presenting Dr. Meadows with the Leadership Award

or contributor to, the CCRC who embodies the spirit of what Dr. Meadows brought to the CCRC. On behalf of all of the investigators and administrative staff of the CCRC, we wish Dr. Jeff Meadows heartfelt congratulations on this award and enduring thanks for all his contributions and being a role model for us all!

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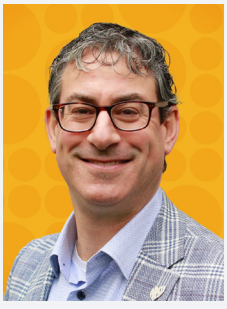

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Driscoll Children's Hospital Unveils $100 Million Heart Center Expansion

Driscoll Children's Hospital Heart Center marked a historic milestone today with the ribbon-cutting of its newly expanded facility, a $100 million investment that significantly enhances pediatric heart care for children across South Texas. The expansion adds over 50,000 square feet of dedicated cardiac care space, including two state-of-the-art operating rooms, two advanced cardiac catheterization labs, and a 35-bed Cardiac Intensive Care Unit (CICU).

"This expansion is more than just adding space, it's about ensuring that every child receives the best possible care with the latest advancements in pediatric cardiology and surgery," said Karen Long, Vice President of Clinical Operations at Driscoll. "We are making an unparalleled commitment to the children and families of South Texas, ensuring they have access to world-class heart care without leaving home."

The investment comes as Driscoll Heart Center continues to achieve some of the best surgical outcomes in North America. Over the past four years, the center has performed more than 600 congenital heart surgeries with a 98.5% survival rate, surpassing national benchmarks and placing Driscoll among the top programs in the country according to Society of Thoracic Surgeons data.

"Pediatric heart surgery demands precision, expertise, and a commitment to progress. At Driscoll, every case is part of a continuum—each advancement builds on the last, driving better outcomes and greater possibilities for our patients. This expansion strengthens that mission, equipping us to take on more complex cases and push the boundaries of what we can achieve in pediatric heart care," said Dr. Stephen Langley, Medical Director of the Heart Center and Chief of Pediatric Cardiac Surgery.

Driscoll's Heart Center treats a high percentage of complex congenital heart conditions and provides expert multidisciplinary care from a team of more than 40 specialists.

"Every child we care for is helped by a hundred hands—every person in our program plays a role in that child's outcome," said Dr. Daniel McKenna, Medical Director of Pediatric Cardiology. "This expansion ensures that our entire team has the best resources to provide the highest level of care."

As South Texas grows in population, Driscoll Heart Center will continue to expand services to more children with the long-term goal of performing more than 500 pediatric heart surgeries each year. The Heart Center is increasing surgical capacity, recruiting additional cardiologists and specialized staff to strengthen regional access to care. A newly opened cardiology clinic in El Paso extends services to West Texas, while the upcoming launch of South Texas's first Mobile ECMO Program will provide lifesaving extracorporeal membrane oxygenation (ECMO) support for critically ill cardiac patients during transport.

Driscoll Children's Hospital

Driscoll Heart Center Expansion Highlights:

- 53,333 sq. ft. expansion – The Heart Center's new Pavilion space on the third and fourth floor adds over 50,000 square feet of dedicated cardiac care space at Driscoll Children's Hospital in Corpus Christi.
- $100 million investment – The largest single investment in pediatric heart care in Driscoll's history.
- Two cardiac operating rooms – Designed for the most advanced pediatric heart surgeries.
- Two cardiac catheterization labs – Expanding capabilities for minimally invasive and hybrid procedures.
- 35-bed Cardiac Intensive Care Unit (CICU)

CHLA Team Is First in World to Investigate Cardiac Xenotransplant as a 'Bridge' for Babies

The Pioneering Preclinical Research Could Pave the Way for Xenotransplant to Support Critically-Ill Babies Who Have Few Options While Waiting for a New Heart

Katie Sweeney

A team led by Children's Hospital Los Angeles has become the first in the world to demonstrate the possibility of using genetically engineered pig hearts as a potential "bridge" to heart transplant for critically ill babies.

The groundbreaking preclinical research could lead to a lifesaving alternative for supporting critically-ill babies waiting for a new heart—particularly those with single-ventricle heart disease, most of whom currently die while waiting for a transplant.

John David Cleveland, MD, a congenital heart surgeon in the Heart Institute at Children's Hospital Los Angeles, led the study and presented the results April 28 in a plenary session at the International Society for Heart and Lung Transplantation Annual Meeting in Boston. He also presented on this research May 4 at the American Association for Thoracic Surgery meeting in Seattle.

"The goal is to be able to use pig hearts instead of machines to support these babies until a suitable human heart can be found," Dr. Cleveland says. "We still need to do more research, but our studies show this is a viable model."

Xenotransplant has long been explored as a potential solution to the severe shortage of donor organs. Although ventricular assist devices (VADs)—mechanical pumps that help the heart circulate blood to the body—can support many patients during the long wait for a heart, infants with single-ventricle heart disease have a critical need for new options.

Currently, only about 30% of those babies will survive three months on a VAD, due to high rates of strokes, bleeding, and infections—meaning that few live long enough to receive a new heart.

"For those with single-ventricle disease, it's difficult to balance their circulations with a VAD—to find a way to give enough blood to the body and enough blood to the lungs," Dr. Cleveland explains. "A machine cannot constantly adapt to the physiology like an actual heart."

Xenotransplant could offer these babies a much better chance at surviving the time it takes to match with a suitable heart for transplant. "Not only could the outcomes be better, but a pig heart could potentially allow babies to go home while they wait for a human heart, as opposed to being connected to a VAD in the hospital," he adds.

A First-Ever 'Bridge'

The researchers began their studies five years ago, supported by an R33 Catalyze grant from the National Institutes of Health. So far, they have transplanted genetically modified pig hearts into

14 young, size-matched baboons at research facilities outside of California.

To date, eight of the 14 baboons have survived for several months with a pig heart. One of the baboons has so far lived for 620 days—nearly 21 months—the longest a nonhuman primate has ever lived with a pig heart. In addition, the team early on successfully reversed an episode of acute rejection in that baboon, the first time that's been done for a xenograft. That work was recently published in Xenotransplantation.

Importantly, the team also has become the first in the world to replace a functioning xenograft with a same-species heart. That was done in two of the baboons, with one living to 105 days.

"We have shown that replacing a xenograft with an allograft is feasible, which no one has done before," Dr. Cleveland says. "Based on those learnings, we are working to further modify and improve the protocol."

He notes that science has come a long way since 1984, when "Baby Fae" received a baboon heart, becoming the first infant to receive a cardiac xenotransplant. She died 21 days later after her body rejected the heart.

One key advance is the ability to genetically modify organs to make them more humanlike. Immunosuppression technology is also far superior now, increasing the ability of the immune system to accept a xenograft.

The CHLA team is using an investigational medication called tegoprubart, an anti-CD40L antibody developed by Eledon Pharmaceuticals that aims to retrain the immune system to better accept a foreign organ as "self." The researchers' protocol also reduces the amount of time that the donor pig heart is out of circulation, which can help reduce the chances of rejection.

Through a partnership with eGenesis, a biotech company that specializes in xenotransplant, the researchers have also been using hearts from Yucatan miniature swine, a pig that grows more slowly than large white pigs.

Dr. Cleveland also says that xenotransplant may have a greater chance of success in babies than in older adults. In earlier studies, the team found that children younger than 18 do not have significant antibodies to pigs, while most adults do.

"The immature pediatric immune system is much more capable of accepting something completely foreign than the adult system is," he says. "We still have more research to do. But our hope is that we can eventually offer these babies a much better chance to live."

FDA Approval for Cordis' PALMAZ MULLINS XD Pulmonary Stent™

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