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Percutaneous Device Closure of Pseudoaneurysm of the Mitral-Aortic Intervalvular Fibrous Tissue with Konar Multi-Functional Occluder Device – Desperate Closure in a Rare Situation: A Case Report

Nurul Islam, MD & Mirza Mohd Kamran, MD

Background

Mitral-aortic intervalvular fibrous tissue (MAIVF) is an avascular structure and connects the anterior mitral leaflet (AML) and aortic valve. Aneurysm of the tissue is very uncommon and a potentially life-threatening condition. The most common causes of pseudoaneurysm of MAIVF (P-MAIVF) tissue are infective endocarditis or post valve replacement surgical trauma. Here we report an uncommon case of ruptured P-MIVF tissue with intra left atrium (LA) extension and transcatheter closure with a newer device.

Case Summary

Thirty-two-year-old lady admitted with gradual onset shortness of breath. Her primary transthoracic echocardiography (TTE) showed large cystic lesion in between aorta and left atrium (LA). It has a communication from LV cavity through aorto-mitral fibrous area. Morphology of the cyst-like structure was better profiled by doing CT angiogram and subsequently closed percutaneously with a low-profile newer device.

Conclusion

P-MAIVF tissue is rare, but can be a harmful lesion. Majority needs surgical intervention, but transcatheter can be planned as per device availability and size

of the lesion. The newer device which is being used (first time from our best of knowledge) is helpful to track the difficult angulation and orientation with minimal difficulty.

Keywords

Pseudoaneurysm, Mitral-aortic intervalvular fibrous tissue, MFO.

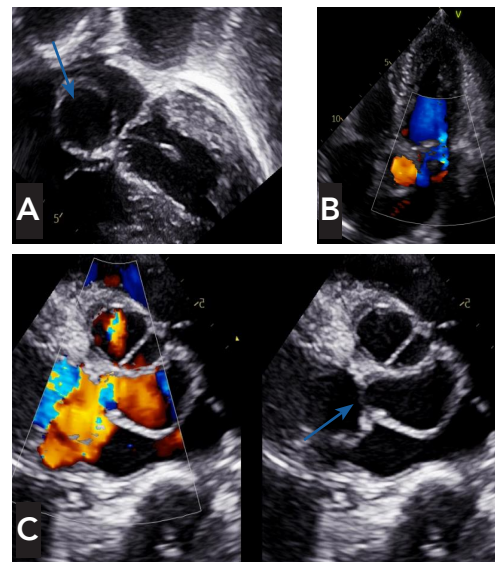


FIGURE 1

A. Sub-costal 4 chambers view showing sac in LA (Blue arrow)
B. Showing significant mitral regurgitation
C. Compressed LA can be well appreciated (blue arrow)

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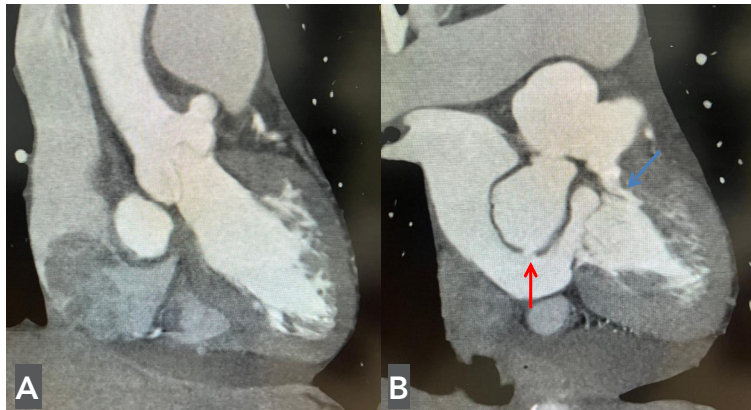


FIGURE 2

A. LVOT view: small aneurysm from LMCA origin
B. Multi-loculated aneurysmal sac arising from LV (blue arrow): Mitral –aortic fibrous tissue- neck diameter: 5.5mm, extending behind, one of the sacs compressing LA & there is a tiny connection (red arrow) in between LA and sac

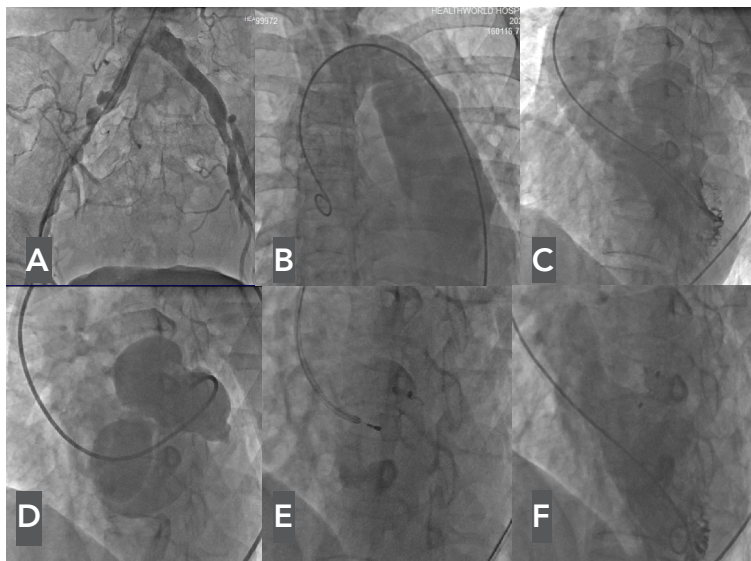


FIGURE 3

Angiographic delineation of lesion and deployment of device
A. Blocked RFA
B. Dilated DTA
C. LV angio 60-20 aneurysmal pouch filling from LV side
D. Multi loculeted aneurysmal cystic lesion super imposing LA
E. MFO device deployment retrogradely
F. Post deployment LV angio

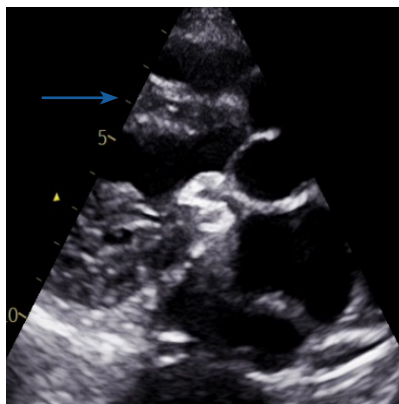


FIGURE 4

Post procedure echo image showing device in good position (blue arrow)

Background

Mitral aortic intervalvular fibrous (MAIVF) tissue connects the anterior mitral leaflet (AML) and aortic valve.¹ This tissue is relatively avascular and susceptible to infection or injury during surgery.² This can result in abscess formation, weakening of the region and pseudoaneurysm formation of MAIVF tissue. In the available literature, this is because of either infective endocarditis or post valve surgery or valve replacement.^{3,4} In this report, we describe a rare incidence of P-MAIVF tissue in a young lady without any pre-existing standard risk factors and successful transcatheter closure with a relatively new device for the first time.

Case Details

A 32-year-old lady was admitted complaining of shortness of breath for the last few months. There was no specific complaint, such as prolonged fever, joint pain, previous hospital admission or prior cardiac surgery. On clinical examination, her body temperature was normal, body weight was only 48kg, BP was 110/68 mm Hg; on auscultation, she had no significant finding, nor on cardiac examination except a soft systolic murmur at the left sternal border. She did not have any risk factors for IE, such as previous cardiac surgery, drug abuse, dental procedures. Her routine blood investigations were normal. TTE showed large cystic lesion within LA, communicating with LV, no signs of IE (Figure 1A, B, C).

The blood was noted flowing during systole from LV to aneurysmal sac. Additional imaging CT angiography was done, which profiled the cyst better. A multi-lobed cyst communicating with LV through MAIVF tissue region. The cyst was found compressing LA and one of the lobes had ruptured into LA, measuring 1.5 to 2mm. The neck of the aneurysm measured 7-9 mm in two different plains (Figure 2A, B).

After thorough evaluation of images from the modalities and detailed discussion with surgical team, it was decided to go for transcatheter closure.

Procedure Details

The defect was profiled in the Cath lab by doing LV angiogram on different angulations. The aneurysm was crossed with exchange length 035 Terumo wire using 5F IMA catheter looking posterior and slightly upward. The wire was secured by making multiple loops inside the sac. The IMA catheter was exchanged with 6F guiding catheter over the stable & secured Terumo wire. The tip of the guiding catheter was kept inside the sac (Figure 3A-F).

In view of the course of the catheter and angulation, we used a relatively low-profile newer device. We used 10x8 mm MFO retrogradely, followed the course of the catheter smoothly, and fitted properly at the site with near occlusion of the aneurysm neck (Figure 4).



There was neither LVOT obstruction nor anterior mitral leaflet mobility restriction or appearance of MR. The procedure was uneventful. The lady was discharged with dual antiplatelets. After eight weeks of discharge, TTE showed no residual flow across the device, reduction of the aneurysmal sac and there was clinical improvement of her symptoms.

Discussion

Pseudoaneurysm of the MAIVF is a rare but not an uncommon lesion. It mostly happens in an individual suffering from infective endocarditis or who has had a previous heart surgery valve replacement.^{2,3,4} It is usually asymptomatic if it remains uncomplicated. This can have an impact on the cardiovascular system when it causes compression of the left atrium and nearby coronary circulation.^{5,6} It can rupture and jeopardize hemodynamics leading to heart failure or tamponade depending on the site of rupture.^{2,7} In our case the pseudoaneurysm, ruptured into LA. The hemodynamics acted like mitral regurgitation, which was the cause of heart failure.^{7,8} Though development of Pseudoaneurysm of MAIVF is highly related to valve replacement surgery, which may threaten the potential treatment to avascular tissue in MAIVF and cause infective endocarditis. This patient did not have either of these. During angiogram her thoracic aorta showed dilation & tortuous femoral arteries. Her coronary arteries angiogram was normal but there was small (5 mm) aneurysm at origin of LMCA. From these findings, we assumed it might be associated with some collagen vascular disease or not. There are limited reports in the literature regarding transcatheter closure of pseudoaneurysm of MAIVF and those are related to surgical trauma. This was not a simple pseudoaneurysm of MAIVF rather complicated by developing a communication between LA proper and aneurysm, which resulted in volume overload and producing clinical symptoms like heart failure. In terms of etiology, the woman did not have previous conventional risk factors. Though her basic rheumatological work up was within normal limits, angiographic findings strongly indicated possible association

of some immunological disorder which needs to be explored.

Device selection in such complex anatomy is challenging. An ADO-II was unsuitable due to neck size limitation, and a vascular plug or high-profile ADO would have been technically difficult to deliver through the tortuous course. We therefore opted for the Konar-Multifunctional Occluder (Konar-MFO), a relatively new low-profile device with several design advantages:

1. Dual-sided screw hubs enable both antegrade and retrograde deployment without requiring an arteriovenous loop—critical in angulated or difficult-to-track lesions.
2. Soft nitinol mesh and optional PTFE membrane allow it to conform to irregular margins, minimizing the risk of impinging on adjacent valves or conduction pathways.
3. Wider waist size range in a low-profile delivery system permits closure of larger orifices without upsizing the sheath, thus reducing vascular trauma.

The Konar-MFO has shown favorable results in other complex septal defects, including post-infarction ventricular septal rupture⁹ where its versatility and deliverability have been documented in challenging anatomies. In that series, the device demonstrated high procedural success and good short-term outcomes, underscoring its applicability beyond conventional VSD closure to more complex and fragile septal pathologies. To our knowledge, this is the first reported case of transcatheter closure of a ruptured MAIVF pseudoaneurysm using the Konar-MFO device, with an excellent procedural and early clinical outcome.

Conclusion

This report describes the first known use of the Konar-Multifunctional Occluder (Konar-MFO) for transcatheter closure of a ruptured pseudoaneurysm of the mitral-aortic intervalvular fibrous (MAIVF) tissue. The device's low-profile, flexible design

and dual-sided deployment capability facilitated delivery across a tortuous and angulated pathway, ensuring secure defect closure without compromising adjacent valvular structures. This case highlights that, in carefully selected patients, the Konar-MFO can provide a safe and effective alternative to surgery for anatomically challenging pseudoaneurysms traditionally managed by open repair.

Limitations

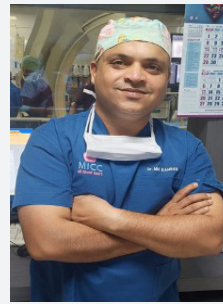
The favorable outcome may not be generalized to all MAIVF pseudoaneurysms, which can vary widely in anatomy and etiology; longer follow-up is required to evaluate the durability of device closure. There is no data available showing any head-to-head comparison with other occluder devices or surgical outcomes. Although angiographic findings suggested a possible connective tissue disorder, no definitive diagnosis was established.

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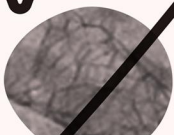

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PDA Stenting: Evolving Strategies, Delaying Surgery, Protecting the At-Risk Brain

Howaida El-Said, MD, PhD; John J. Nigro, MD; Jacqueline Rodriguez

Introduction

The 2025 PDA Stenting Symposium brought together more than 520 clinicians—120 in person and 400+ virtual—from across the globe. Participants represented Europe, Egypt, India, Peru, Mexico, and multiple U.S. centers, making this one of the most internationally collaborative meetings on Congenital Heart Disease. This year's symposium revealed a field undergoing rapid maturation: PDA stenting has evolved beyond a technical alternative to the BT shunt and is now central to neonatal stabilization, neurodevelopmentally informed care, pre-Glenn pulmonary artery growth strategies, and refined surgical timing across single-ventricle pathways.

From the opening sunset reception with Dr. Sanjay Sinha's Gen-X band to the closing call for a multi-institutional PDA stent registry, the meeting highlighted multidisciplinary integration—fetal cardiology, neonatology, ICU care, interventional cardiology, cardiac surgery, pulmonology, imaging, computational modeling, and long-term ACHD perspectives.

Keynote Address

From Urgency to Strategy: Reimagining the Surgical Timeline — Drs. Gil Wernovsky, John Moore and Marc Gewillig

Dr. Gil Wernovsky reframed neonatal congenital heart disease (CHD) care as a complex developmental journey. He emphasized that many high-risk infants have a "35-week-equivalent brain," impaired placental function, and vulnerability to ischemia-reperfusion injury. PDA stenting provides a stabilized physiology that may avoid early circulatory and neurological insults. He argued that the "tipping point" toward improved neurodevelopment hinges on: 1). integrated teams, 2). sophisticated imaging, 3). consistent ICU practices, and 4). family mental-health-informed care structures.

PDA Stenting from 10,000 Feet: First Principles Matter

Dr. John Moore emphasized that while PDA stenting offers clear physiologic advantages over surgical shunts, the field still lacks the long-term evidence base needed to guide practice. He stressed the urgency of developing **purpose-built neonatal ductal stents**—devices with true flexibility, conformability, and controlled flaring rather than repurposed adult coronary platforms. He underscored the importance of **transparent family counseling**, detailed failsafe planning for spasm or obstruction, and consistent postoperative monitoring. Above all, Dr. Moore called for **rigorous multicenter data**

collection, standardized definitions, and comparative studies so that PDA stenting can advance from promising practice to evidence-driven standard of care.

The Goal Is Not Just Survival to the Next Stage, But Quality of the Final Repair

Dr. Marc Gewillig reframed Fontan care as an engineering problem: the TCPC functions like a **dam**, creating chronic upstream venous congestion and downstream low cardiac output unless the pulmonary vascular bed has exceptionally low-resistance. Because **neonatal physiology is the only true window for PA catch-up growth**, he urged abandoning age-based Glenn timing and instead aggressively optimizing pre-Glenn anatomy through shunt revision, PDA stent redilation, or PA augmentation so that by Glenn the PAs are already "final size." He emphasized that the Fontan ventricle is chronically **underfilled and stiff**, that excessive collaterals reflect inadequate early PA flow, and that **exercise—not medication—is the only physiologic way to push through the Fontan bottleneck**. His central message: long-term Fontan success is determined almost entirely by **early, intentional engineering of PA size, symmetry, and ventricular readiness**.





How Do You Decide on Stent Length? Roundtable Approaches

Dr. Frank Ing emphasized angiography-based measurement after wire placement, noting that AP and lateral views help anticipate straightening. He routinely overestimates length slightly, keeps multiple stent options available, and stresses having a clear rescue plan for spasm, thrombosis, or geometry change.

Dr. Omar Deyaa favored echo-guided length estimation, reserving CT for complex ducts. Traditional 'add 1–2 mm' rules fail in tortuous PDAs, and he advises shorter stents with planned overlaps to avoid PA jailing. Surgical operability outweighs the ideal of a single-stent repair.

Dr. Shakeel Qureshi advocated blending 3D CT with curvature indices. With severely tortuous ducts, straightening is incomplete; operators should calculate a straightened length (L1) and curved length (L2) and choose a stent size between them. Dual-stent constructs are often appropriate.

Dr. Alwi Mazeni stressed that the PDA must conform to the stent, not vice versa. Straighten the duct with two wires before measuring, and expect to deploy two stents in ducts with sharp proximal angles. Avoiding overextension into branch PAs remains paramount.

Dr. Gunter Kerst described RAO 30° plus lateral imaging for aortic ducts, frequently

needing telescoping self-expanding stents. Ducts are often longer than assumed.

Dr. David Balzer integrates CT, angiography, and microcatheter measurements. His multicenter review shows angiography underestimates duct length by ~3 mm, and PDAs shorten ~26% after stenting. He prepares two to three plausible lengths and confirms alignment before deployment.

Across speakers, consensus emerged: duct length estimation remains imprecise, CT improves but does not replace intraprocedural assessment, and slightly long is better than slightly short. Multi-stent strategies are common, safe, and physiologically appropriate.

Innovation Mix: Novel Interventions and Evolving Pathways

Dr. Sherine Abdelsalam presented a collateral-access strategy for high-risk RVOT perforation. Mapping ductal override patterns and using contrast-timing to identify dependent lung segments improved wire safety and reduced perforation complications. Dr. Marjan Hesari summarized Rady Children's stent strategy for obstructed TAPVR in high-risk neonates, using vertical vein plus ductus venosus stenting to create a V-shaped egress when needed. She demonstrated that this strategy decreases early obstruction and improves surgical survival compared with historical data.

Dr. Matthew Brigger highlighted airway vulnerability in CHD infants. Left bronchial compression from trachea-bronchomalacia, and fixed airway lesions frequently limit cardiopulmonary stability. He reviewed metallic vs biodegradable airway stents and emphasized early multidisciplinary aerodigestive care.

Dr. Shyam Sathanandam demonstrated MVP-based flow restriction for oversized PDAs. Fenestration diameter governs Qp:Qs, and strategic 'flaring' stabilizes device position. This is particularly useful when ductal stents cause excessive pulmonary blood flow.

Dr. Howaida El-Said presented a physiologic method for rebuilding the pulmonary artery tree in PA/IVS + MAPCA physiology. RVOT stenting delivers antegrade flow into native PAs, sometimes identifying small MAPCA-to-PA connections that can be balloon-dilated to incorporate territory without the need for early unifocalization.

Dr. Diego Porras addressed the ASD-closure debate in PA/IVS. Closure improves oxygenation and RV preload but may cause decompensation if RV compliance remains poor. Test occlusion and individualized physiology—not fixed rules—should guide decision making.

Engineering and Device Innovation

Dr. El-Said highlighted the need for neonatal-specific self-expanding ductal stents. Current adult coronary platforms are too rigid,



causing ductal spasm and poor conformability. Soft, self-expanding stents may reduce trauma, improve curvature matching, and lower branch PA jailing risk.

Dr. Henri Justino presented a polymer-based pediatric pulmonary valve designed for infants as small as 14–18 mm, expandable to adult sizes. This calcification-resistant leaflet technology—supported by extensive NIH funding—may redefine RV–PA conduit management.

Fontan Strategy Sessions —d’Udekem, Al Halees

Dr. Yves d’Udekem highlighted lifespan-oriented approach to Fontan care, emphasizing that optimal Fontan timing (typically **two to seven** years) depends on early PA growth rather than age alone, and that many problems seen in “late Fontans” originate from suboptimal upstream decisions. He underscored the need for appropriately sized conduits—adult survivors ultimately require **22–26 mm**, making planned upsizing in later childhood essential. Ventricular filling physiology, including insights from inspiratory IVC collapse, may revive selective use of the lateral tunnel. About **11%** of Fontan patients can achieve **≥80% predicted VO₂**, with early structured exercise as the strongest modifiable factor. He supports routine fenestration closure when feasible, as raising saturations from mid-80s to low-90s significantly improves well-being. Emerging tools—4D-flow MRI, venous viscoelasticity metrics, and computational models—hold promise, but remain underutilized clinically. He emphasized early intervention for lymphatic failure (including catheter-based decompression) and early repair of even moderate AV valve regurgitation to protect long-term ventricular function. Overall, his philosophy centers on **proactive, physiology-driven, preventive care across the entire single-ventricle lifespan**.

Dr. Al Halees reviewed modern Fontan preparation: low PVR is fundamental; PA growth occurs almost exclusively before Glenn; and Fontan completion between 18 months and four years prevents deterioration. His percutaneous Fontan completion program achieves extubation in the cath lab and minimal effusions.

Critical Physiologic Care — ICU, Airway, and Interstage

Dr. Gewillig emphasized tailoring prostaglandin (PGE) use to ductal anatomy. Straight, short ducts benefit from **allowing natural constriction**—avoid early PGE and stop it two-to-three hours before cath to improve stent grip. Tortuous ducts, however, should receive **early PGE** to maintain curvature and avoid collapse during intervention. For short ducts supplying a single lung, **aggressive PGE and early stenting** are essential. His key message: **match PGE strategy to duct morphology**, as mild constriction can significantly improve stent stability.

Dr. Ghanayem emphasized that PDA stent patients require surgical-shunt-level vigilance: two-site NIRS, structured handoffs, and early detection of physiologic decline. Early saturation drop or poor weight gain predicts interstage risk.

Dr. Aparna Rao reviewed airway vulnerabilities, including tracheobronchomalacia, vascular compression, fixed airway lesions, and parenchymal disease. The heart cannot thrive without matching pulmonary function; early pulmonology involvement is essential.

Dr. David King described the emerging role of remote interstage monitoring: daily saturations, weights, feeding logs, and automated alerts embedded in parent-friendly dashboards. Human oversight remains essential to avoid alarm fatigue.

Should We Have a PDA Stent Registry?

A multidisciplinary panel (El-Said, Nigro, Alshawabkeh, Zablah, Kobayashi) discussed the need for a PDA stent registry. Clinical practice has outpaced evidence; operators need multicenter data to refine reintervention timing, PA growth expectations, and surveillance strategies. Key data elements should include: duct morphology, access strategy, stent type and dimensions, PA symmetry, complications, reinterventions, and interstage outcomes. Barriers include IRB variability, data standardization, and cost. Solutions include modular datasets, automated extraction, and alignment with established CHD networks. Consensus: a PDA stent registry is essential, and a working group should form immediately.

Concluding Remarks

The symposium highlighted a field in rapid maturation. PDA stenting has become a central stabilizing strategy for ductal-dependent circulations, touching every stage from fetal diagnosis to adult congenital care. Precision planning, neonatal-specific devices, advanced imaging, computational modeling, airway and lymphatic science, and structured interstage monitoring are moving the field toward physiologic, sustainable palliation. The next leap will require multicenter registry data, engineering collaboration, and unified standards of care.

Acknowledgment

We extend our deepest gratitude to Jackie Rodriguez, whose exceptional leadership and meticulous organization made this meeting possible. Her dedication ensured a seamless and productive symposium from start to finish. We are also sincerely thankful to Daniel Roberson, Mark Abcede, Alexis Duran, and Danica Peterson for their invaluable support behind the scenes. Their commitment, coordination, and hard work were essential to the success of this event. We also gratefully acknowledge Rady Children’s Hospital–San Diego for its unwavering support, and Dr. John Nigro for his leadership, guidance, and continued commitment to advancing congenital heart care.



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Letter to the Editor

Dear Mrs. Baldwin,

It was a year ago that your editors censored my article (without asking or informing me) about the sad state of the supply chain of essential medical equipment and supplies to the Soler Pediatric Medical Center in Havana Cuba. The deficiency of supplies is only due to the cruel US Embargo and designation of Cuba as a state sponsor of terrorism. This cruel embargo that results in the deaths of many Cuban children has been condemned by a UN vote of 165 to seven.

You rationalized your censorship of my article based on a supposed journal policy of "not publishing political views."

I read the excellent article by Dr. Andrii Maksymenko in the November 2025 issue. I liked the article and understood why Dr. Maksymenko at the bottom of page 2, wrote that "A separate priority of the Center's work has become assistance to the defenders of Ukraine." This statement is political. There are two political sides on the issue of the Ukraine war. One side says that Ukraine is a tool of the US and that Ukrainian children are dying due to mistaken policies of the West. The other political side says that the entire conflict is the fault of Vladimir Putin and the Russians. The journal had no problem, in this instance, taking political sides.

This discrepancy between your censorship of my article about the plight of Cuban children and Andrii's article about the plight of Ukrainian children shows a clear bias on the part of the journal.

In the interest of objectivity, I ask that you republish my article in its original form or at least publish an erratum regarding the censored paragraph.

I call your attention to the fact that the late, great William Roberts, editor, and founder of the American Journal of Cardiology, took a principled stand regarding the cruel US embargo of Cuba in Roberts' articles about this subject. Congenital Cardiology should do the same.

Regards,

Robert Detrano MD, PhD
Retired University of California School of Medicine
China California Heart Watch

Dear Dr. Detrano,

Thank you for the letter regarding your article, "My Visit to Havana," published in the January 2025 issue of Congenital Cardiology Today (CCT). As a technical medical newsletter, CCT's focus is on articles of interest to the pediatric and congenital cardiology community of practitioners in North America and worldwide. CCT's mission is to provide reliable and timely information on congenital and structural heart disease. As part of that mission, CCT publishes occasional articles about practice and practice conditions in places and environments unfamiliar to most of our readership. Moreover, one of our editorial policies is to exclude statements which we consider overtly political. We acknowledge that judgement is required. We exercise that judgement rarely, by slightly editing manuscripts to remove obvious political content.

We deemed the following statements included in your original manuscript to be too political for publication:

"The American blockade is unhelpful and cruel and is materially harming the people of Cuba. The United States should institute full diplomatic and economic relations with Cuba. Travel to this beautiful, extremely safe, and friendly country should be open to US travelers without restrictions. Everyone will benefit from this."

Thus, we removed these statements from the published article.

Regarding your concerns about editorial consistency, as reflected in an article by Dr. Maksymenko about pediatric heart care in Ukraine during wartime (November 2025); we judged this article to be a report about practices and conditions at the Ukrainian Children's Cardiac Center during war. The statement: "A separate priority of the Center's work has become assistance to the defenders of Ukraine," in our view reports a practice of the Center and is not overtly political. Therefore, we did not exclude it from the published article.

We acknowledge that we were not explicit in our communications with you about removal of the statements cited earlier from the published version of your article. We apologize for this omission. Your letter has provided a learning experience for us, and we will improve communications about editing with authors going forward.

Thank you again for your letter and for being a reader of Congenital Cardiology Today. We hope your concerns have been adequately addressed.

Sincerely,

Kate Baldwin	Tony Carlson	John Moore, MD, MPH
Publisher &	Founder &	Co-Founder &
Editor-in-Chief	Senior Editor	Medical Editor

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Play-based LEGO® MRI Scanner Set Reduces Anxiety and Sedation for Children in Hospitals Worldwide

Over one million children around the world have used the LEGO Group's MRI Scanner set, helping transform a potentially stressful medical procedure through a playful, hands-on learning experience.

The set, which features a scanner, patient bed, waiting room, scanning room, staff minifigures and medical accessories, was designed by the LEGO Group and the LEGO Foundation to help children understand MRI procedures by learning through play. MRI scans are frequently used in pediatric care because they do not use radiation. But for many children, the experience can be overwhelming, requiring them to lie still in a large, noisy machine for extended periods. As a result, sedation or general anesthesia is often used.

New research conducted by the LEGO Group revealed that 96% of healthcare professionals globally who used the LEGO MRI Scanner set say the model helps alleviate children's anxiety, and 46% report that it has reduced the need for sedation or anesthesia during MRI procedures.

Since 2023, more than 10,000 LEGO MRI Scanner sets have been donated to hospitals and health professionals around the world as part of the LEGO Group's commitment to unlocking the power of play for children, especially those who may need it most.

From Scan to Smiles

Through role play and storytelling, the LEGO MRI Scanner allows hospital staff to offer a more child-centered, playful approach when they prepare a child for a scan. The research also showed that 95% of healthcare professionals using the set say it improves the family's hospital experience, while 94% find it fun and engaging for children.

The LEGO Group has gathered testimonials from children, parents and healthcare professionals around the world, including five-year-old Ivy from Edinburgh, Scotland. At the age of two, Ivy began having prolonged seizures. After being put to sleep for her first MRI scan, her family and hospital team wanted to try her second scan at age four awake. Ivy was referred to the procedural anxiety team at the Royal Hospital for Children and Young People in Edinburgh, UK, where she was first introduced to the LEGO MRI Scanner model.



Speaking about their recent experience, Ivy's mum Rachel said: "On the day of her second scan Ivy did really well and was even a bit excited! If we hadn't played with the LEGO MRI model beforehand, I think she would have had a full meltdown and would no doubt have needed general anesthetic. Nobody wants their child to be put to sleep if you can avoid it."

"Ivy is a visual learner and loves LEGO bricks, so seeing and explaining the procedure through play was a game changer. It really helped her to understand what to expect, took away any nerves and made things go plain sailing. Coming into hospital can be a stressful experience, but playing with this model made our whole family feel more relaxed, calm and prepared."



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Partial Heart Transplant Shows Promise in Congenital Heart Disease

Patients showed appropriate valve growth, but long-term immunosuppression data is needed

Tara Haelle, Contributing Writer, MedPage Today

Key Takeaways

- Partial heart transplants for congenital heart disease using donor semilunar valves showed valve growth matching patient development in infants.
- Transplanted valves functioned well over time, with no major complications from immunosuppression reported.
- While promising, this approach requires further study on long-term outcomes and equitable valve access.

Partial heart transplants using donor semilunar valves in patients with Congenital Heart Disease appeared to be a safe and feasible procedure resulting in appropriate tissue growth, according to a case series.

Among 19 patients undergoing transplant, nine in the initial cohort had functioning valves at a median follow-up of 26 weeks, with the annular diameter of aortic valves growing from a median 7 mm to 14 mm and that of pulmonary valves growing from a median 9 mm to 17 mm ($P=0.004$ for increase in pulmonary valve), according to Joseph W. Turek, MD, PhD, of Duke University in Durham, North Carolina, and colleagues.

To ensure annular diameter growth was not simply dilation, leaflet length was assessed. Aortic valve leaflets grew from a median 0.5 mm to 1 mm and pulmonary valve leaflets grew from a median 0.49 mm to 0.675 mm ($P=0.004$ for pulmonary leaflet growth), the research team reported in JAMA.

One patient needed reoperation, though not due to the implanted valve, and there were no major complications related to immunosuppression.

"Partial heart transplant has the potential to provide growing and living tissue for valve replacement, addressing a critical limitation of current technologies," the authors wrote. "However, it is crucial to recognize that partial heart transplant is not a panacea but, rather, a promising step forward that requires further refinement."

Treatment of heart valve disease in infants is challenging, primarily because current valve

replacements cannot grow with them as they get older. Cryopreserved valved homograft implants from deceased donors are currently the accepted standard for those who are not candidates for valvuloplasty; however, these often succumb to calcification and fibrosis, which requires children to undergo multiple, increasingly risky subsequent operations over time, often with high rates of poor outcomes, the authors noted.

Partial heart transplant -- implanting freshly obtained donor tissue and using standard immunosuppression to maintain its viability -- is potentially a viable way to advance surgical management of the disease, as it allows for adaptive growth, much like a full heart transplant, though it comes with its own challenges and is not universally applicable.

"While partial heart transplant offers an alternative to conventional valve replacements, particularly for growing patients, it will never eliminate the need for future interventions by nature of the underlying congenital heart disease," the researchers wrote.

"This study highlights the promise of using viable heart valves for repair of congenital valvular disease in children to decrease the need for reoperation and reintervention if valve growth and function are maintained in the longer term," Kevin P. Daly, MD, of Boston Children's Hospital and Harvard Medical School, wrote in an accompanying editorial.

But he also highlighted its challenges, including equitable allocation of valves, as well as better understanding the long-term consequences of immunosuppression in these patients.

"Major adverse events related to immunosuppression include: serious infection, acute and chronic kidney damage, posttransplant lymphoproliferative disease, teratogenicity of mycophenolate mofetil, and other drug-related adverse events." Many of these develop over time, he said, so their risks in these patients are not clear.

"In the case of partial heart transplant in children, it would be reasonable to hypothesize that immunological injury would result in sufficient damage to prevent continued vascular growth, but that has

not yet been demonstrated," Daly wrote. "Understanding the long-term consequences of immunological injury, and the degree to which such injury can be tolerated, will be key to properly balancing the benefits and burdens of immunosuppression."

The authors described a case series of 19 patients with Congenital Heart Disease who underwent partial heart transplants using donor hearts' semilunar valves at a single U.S. pediatric cardiac surgery and transplant center between April 2022 and December 2024. The valve implantation was done in the same manner as cryopreserved homograft valve replacement.

Most of the participants were young infants, with a median patient age of 97 days at time of transplant, but the case series also included seven pediatric patients over one-year-old and one 34-year-old patient; ten patients were male. Most patients had truncus arteriosus or tetralogy of Fallot; others had critical aortic stenosis, severe aortic insufficiency, right ventricular outflow tract (RVOT) obstruction, and biventricular outflow tract obstruction.

Patients were screened for cytomegalovirus and Epstein-Barr virus prior to surgery and received cytomegalovirus prophylaxis. Immunosuppression for most patients involved mycophenolate mofetil, started immediately after surgery and given every 12 hours for one year, and tacrolimus, started two days after surgery with trough levels of 6-10 ng/mL for the first year and 4-8 ng/mL after that.

Seven patients received RVOT replacement with a living pulmonary valve, seven received RVOT replacement with a living aortic valve, three received both semilunar valves, and two received a living aortic valve in the left ventricular outflow tract (LVOT). Median operation time was 294 minutes, and median cardiopulmonary bypass time was 145 minutes. Patients spent a median nine days in intensive care and a median 13.5 days in the hospital.

Growth was analyzed in the first nine patients, who were all under four months old at the time of their transplant. "All transplanted valves demonstrated growth over the interval time frame, tracking along appropriate z scores," including in one patient who had to stop



receiving immunosuppression due to an infection after an unrelated procedure, the authors reported.

Aside from the patient with infection from an unrelated procedure, one had a group B Streptococcus respiratory infection that was successfully treated with antibiotics. Two had some evidence of kidney injury that required reducing the dose.

No patients developed significant stenosis or regurgitation of the neo-aortic or neopulmonary valves at the time of discharge, though two patients later developed mild regurgitation of the transplanted aortic valve in the LVOT position and two others showed mild regurgitation of the transplanted pulmonary valve in the RVOT position.

Disclosures

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