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Anomalous Origin of the Right Coronary Artery from the Pulmonary Artery: Reviewing the Spectrum of Diagnostic and Management Strategies in a Preemie and Septuagenarian

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Introduction

Anomalous origin of a coronary artery from the pulmonary artery is a rare congenital cardiac anomaly with two main subtypes.[1] The more common, anomalous left coronary artery from the pulmonary artery (ALCAPA), receives significant attention due to its high mortality rate in early childhood; however, anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is less well-known.[1] Unlike ALCAPA, which typically presents with congestive heart failure and death within the first year of life, cases of ARCAPA are usually discovered incidentally in asymptomatic patients; however, these patients still carry an increased risk of myocardial ischemia and sudden cardiac death secondary to the coronary steal phenomenon, so the recommended treatment has historically been surgical correction regardless of symptoms.[1-7]

Until the mid-1980's, coronary artery anomalies were primarily diagnosed postmortem, intraoperatively, or by coronary angiography.[4,5,8] Presently, noninvasive imaging such as echocardiography has been increasingly utilized in the diagnosis of ARCAPA, with coronary angiography still considered the "gold standard." However, advances in cardiac computed tomography angiography (CTA) now allow the ability to display higher quality information while avoiding invasive heart catheterization.[6,9,10] In select cases, when myocardial viability is in question, cardiac magnetic resonance imaging (MRI) may be used.[6]

The relative lack of review literature on ARCAPA along with its considerable variation in presentation makes diagnosis and management a challenge. Here, we describe two contrasting cases of ARCAPA in a preemie and septuagenarian which together showcase: the diagnostic approach, the role of complementary imaging, and how conservative management may be preferred over surgical correction in the appropriate clinical setting.

Case Presentations

Case 1: A 36-week premature infant with a prenatal diagnosis of coarctation of the aorta was born with a birth weight of 1.6 kg and required prostaglandin therapy. Postnatal transthoracic echocardiogram (TTE) findings confirmed long-segment coarctation of the aorta **(Figure 1)** and a nearly-closed perimembranous Ventricular Septal Defect. Then, suspicion of ARCAPA was raised **(Figure 2)**. Due to the infant's small size and stable condition, a cardiac CTA was delayed until two months of age; it later confirmed ARCAPA originating from the anterior sinus of the pulmonary artery and a mildly hypoplastic distal arch with coarctation of the aorta **(Figure 3)**. When the patient's weight was deemed appropriate at three months of age, surgical repair of both the anomalous right coronary artery and aortic coarctation was performed. The infant is now 20 months old, asymptomatic, and thriving.

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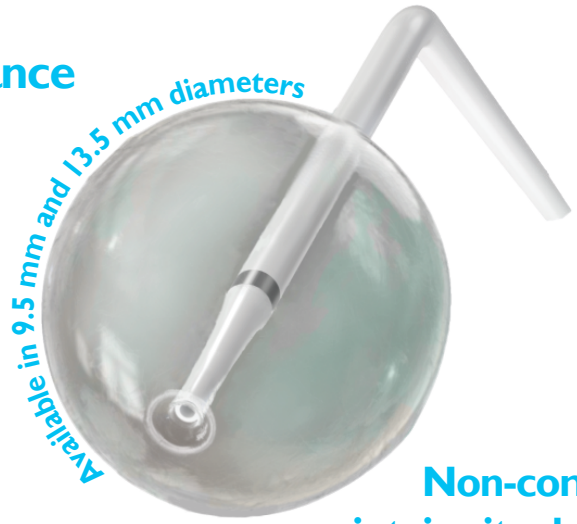

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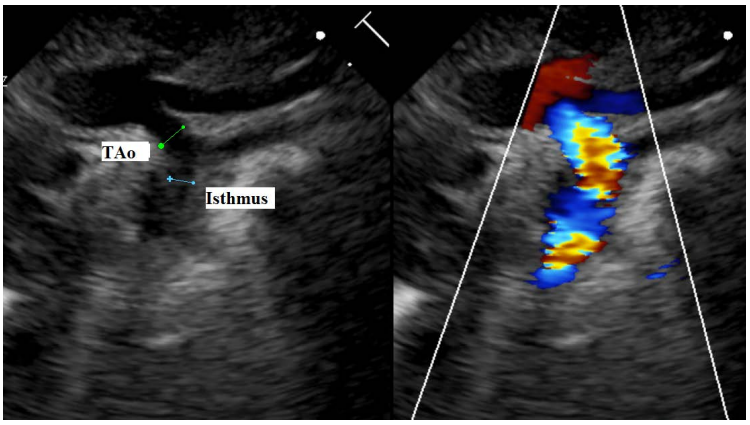

FIGURE 1 *Suprasternal view showing hypoplastic distal transverse arch and isthmus (TAo = Transverse arch)*

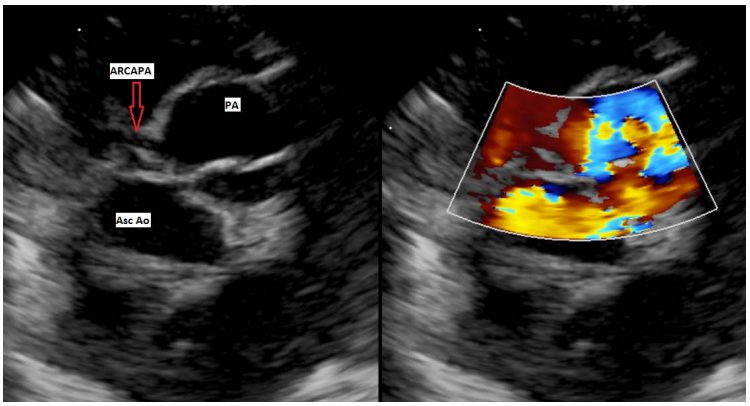

FIGURE 2 *Parasternal short axis view showing anomalous origin of right coronary artery from facing sinus of pulmonary artery (ARCAPA = Anomalous right coronary artery from pulmonary artery; PA = Pulmonary artery; Asc Ao = Ascending aorta)*

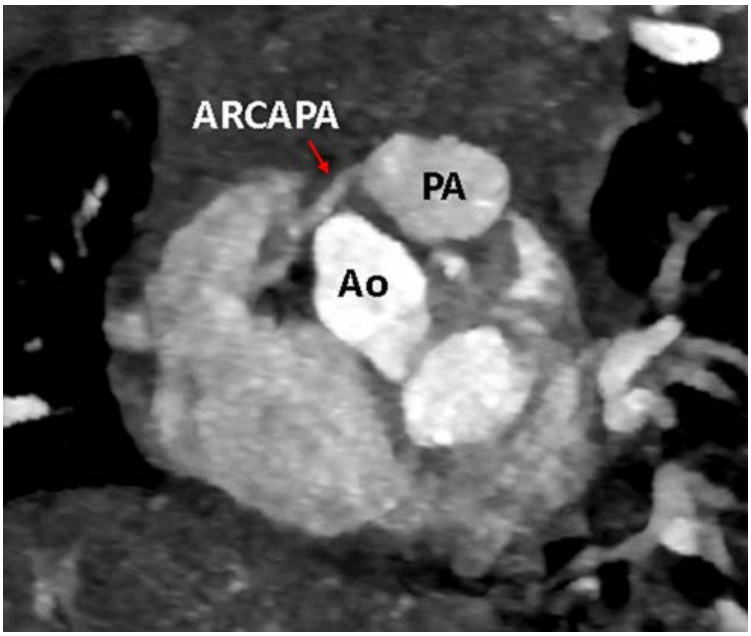

FIGURE 3 *CT Image for **Case 1**: ARCAPA. (ARCAPA = Anomalous origin of the right coronary artery from the pulmonary artery; Ao = Aorta; PA = pulmonary artery)*

Case 2: A 75-year-old female with a history of giant cell arteritis, asthma, hypertension, and hyperlipidemia presented with chronic dyspnea on exertion and no other cardiac symptoms. She was morbidly obese with a blood pressure of 154/92 mmHg and had a normal cardiac and pulmonary examination. TTE was performed at an outside institution and reported a left ventricular ejection fraction of 65% with no significant valvular disease. Considering her age, symptoms, and other risk factors, she underwent coronary CTA—which showed multiple collaterals from the left anterior descending coronary artery to the right coronary artery (RCA). The left main and circumflex coronary arteries were normal. The RCA was dilated and originated from the main pulmonary artery with extensive left-to-right collaterals **(Figure 4)**. There was scattered coronary atherosclerosis without significant stenosis. A positron emission tomography stress test showed a small, mild, reversible basal inferior wall perfusion defect consistent with obstructive coronary artery disease. It otherwise showed normal myocardial perfusion at rest and stress. Given the patient's comorbid conditions, lack of defined ischemic zone, and good collateral formation, she was deemed a poor surgical candidate for coronary reimplantation and appeared to be well-compensated from a cardiovascular standpoint, so continued observation was recommended. She is doing well at her recent visit.

Discussion

First described by Brooks in 1885,[11] ARCAPA is extremely rare, with a reported incidence of 0.002%[5] and only 100–200 cases ever reported.[9,12-14] Because many patients remain asymptomatic, the true incidence might be higher.[5] It is an isolated anomaly in 70% of cases[14] and associated with other congenital cardiac abnormalities 22%–40% of the time, most commonly aortopulmonary window, tetralogy of Fallot, septal defects, and (as seen in our pediatric case) coarctation of the aorta.[1,5,12,14]

ARCAPA is typically well-tolerated in the neonatal period due to the physiologically high pulmonary vascular resistance which promotes antegrade flow from the pulmonary artery into the anomalous RCA; however, as the pulmonary vascular resistance falls over time, there is a reversal of flow from the anomalous RCA into the pulmonary artery leading to the coronary steal phenomenon—a major "hallmark"[1] of this anomaly.[13-16] The resulting left-to-right shunt leads to poor myocardial perfusion in the right coronary distribution followed by extensive collateralization between the two coronary systems in order to preserve adequate ventricular function.[1,14,17]

Ultimately, the pathophysiology of ARCAPA is determined by the direction of coronary blood flow and the quality of myocardial oxygen delivery.[4,7,9] The severity of ischemia and thus, symptoms, is determined by: the degree of collateralization, the presence of stenosis in the RCA, the size of the left-to-right shunt, and the myocardial oxygen demands.[7-9,15,18,19] These factors contribute to the unpredictable presentation of ARCAPA, which is often asymptomatic until adulthood and makes clinical diagnosis challenging.[14,17] When present, the signs and symptoms of ARCAPA are related to myocardial ischemia, including: angina, dyspnea, fatigue, congestive heart failure, myocardial infarction, and sudden cardiac arrest.[3,14,17] The typical ECG findings in anomalous origin of the right coronary artery from the pulmonary artery may be normal, show signs of left ventricular hypertrophy, or indicate ischemia/infarction in inferior leads.[9] While these findings are nonspecific, they may prompt further cardiac workup that ultimately leads to diagnosis as seen in **Case 2**.

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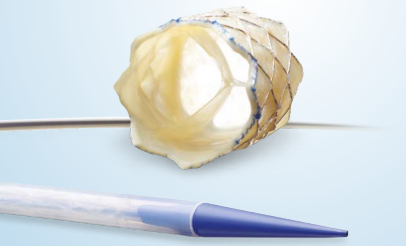

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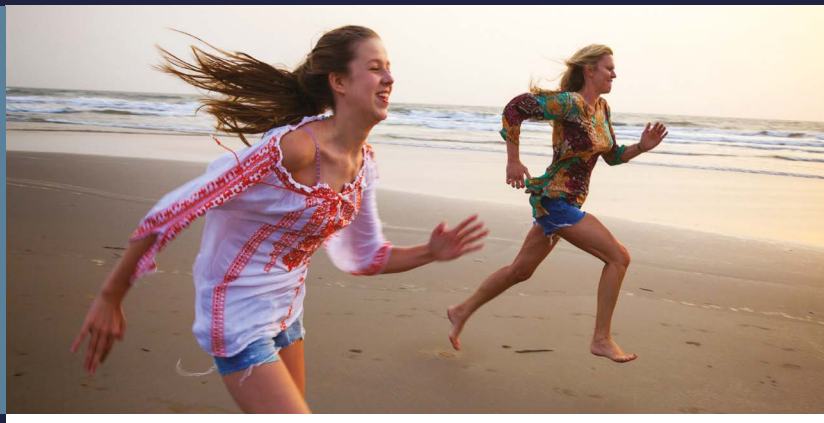

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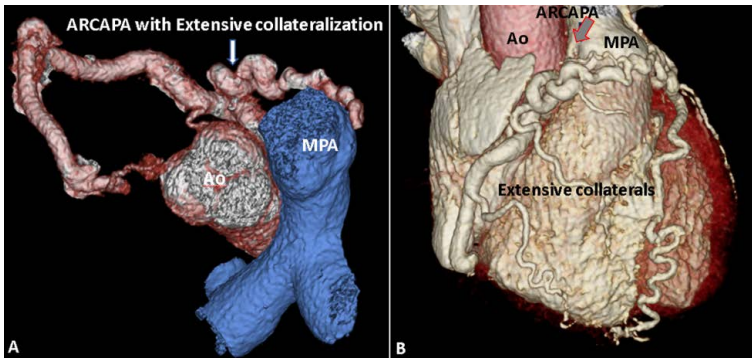

FIGURE 4 *CT Images for **Case 2**: A and **B** showing ARCAPA with extensive collateralization. (ARCAPA = Anomalous origin of the right coronary artery from the pulmonary artery; MPA = main pulmonary artery; Ao = Aorta)*

Although ARCAPA is commonly an incidental finding, it is important to maintain a high index of suspicion. In our first case, fetal ultrasound at 31 weeks identified coarctation of the aorta, but the associated anomalous right coronary artery was only discovered through expert-level imaging acquisition and interpretation on the postnatal TTE. Once suspected on the echocardiogram, the follow-up cardiac CTA was able to further characterize the ARCAPA for timely and optimal surgical planning. This is contrasted by our adult patient, who was diagnosed with ARCAPA in the workup for chronic exertional dyspnea in the setting of multiple cardiovascular risk factors. Both cases highlight the extremely variable clinical presentation of ARCAPA and how the diagnostic evaluation hinges on advanced cardiovascular imaging.

Whether picked up incidentally or through evaluation of signs and symptoms, the diagnosis of ARCAPA can be achieved through several imaging methods. In 1985, the first case of ARCAPA was diagnosed by echocardiography,[20] and since then it has become an excellent initial imaging modality for diagnosing the condition—capable of capturing: the anomalous origin of the RCA from the pulmonary artery **(Figure 2)**, retrograde flow from the right coronary into the pulmonary artery, collateral vessels between right and left coronary systems, and dilation of the left coronary artery.[4,7,9,14] However, echocardiography is limited to two-dimensional images with less spatial resolution and relies on adequate acoustic windows.[9,14]

While coronary angiography is considered the "gold standard" imaging modality to diagnose coronary anomalies, advanced cardiac imaging tools such as cardiac CTA and MRI can provide remarkable three-dimensional reconstructions noninvasively and aid in surgical planning by displaying the coronary anatomy in exceptional detail.[6,9,10] Generally, multi-slice CTA is preferred over MRI due to higher spatial resolution,[6,9,10] which is particularly important in the neonatal population. In sum, these advanced imaging modalities may discover ARCAPA incidentally or be used for further characterization once encountered through other imaging methods. In both **Cases 1 and 2**, the CTA provided the level of detail required for surgical planning; however, our adult patient required additional myocardial perfusion imaging in order to assess the ischemic burden and candidacy for surgery.

Once ARCAPA is identified, surgical correction with reimplantation is generally recommended to reduce the risk of myocardial infarction and sudden cardiac death, even in asymptomatic patients.[1,3-6,9,14] However, selecting the appropriate patient for surgery is dependent on careful evaluation of risks and benefits. Surgery is almost always recommended in pediatric patients such as **Case 1** because the lifelong benefits of a two-coronary system outweigh the relatively low surgical risks in this group.[14] But for the adult patient in **Case 2**, the decision tree is less straightforward. Compensatory adaptations such as the development of collateral vessels over time likely prevented myocardial ischemia, allowing the patient to remain asymptomatic into late adulthood. It also was not entirely clear if the anomalous RCA was contributing to the symptoms in any meaningful way given the lack of well-defined ischemia on myocardial perfusion imaging. In addition, the patient's medical comorbidities—hypertension, hyperlipidemia, and morbid obesity—increased the risk of mortality and postoperative complications associated with open cardiac surgery.[6,21] For all these reasons, the decision was made to pursue conservative treatment, which may be an acceptable option for other adult ARCAPA patients in this situation.

Conclusion

ARCAPA is a very rare congenital cardiac anomaly that leads to chronic myocardial ischemia over time via the coronary steal phenomenon. Patients with this anomaly are typically asymptomatic until adulthood, but the clinical presentation varies widely. Here, we have presented two cases, ARCAPA in a preemie and septuagenarian, to illustrate the diagnostic and management strategies for this condition. Whether encountered incidentally or in the setting of ischemia, noninvasive cardiac imaging with echocardiography and multi-slice CTA is essential to confirm the diagnosis of ARCAPA and assist in surgical planning. In the neonatal population, surgical correction is typically recommended to reduce the risk of sudden cardiac death; however, conservative management should be considered in well-compensated adult patients who are high-risk surgical candidates.

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Conflicts of Interest: None

Ethical Standards: This article does not contain any studies with animals performed by any of the authors. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

References

1. Ganta S, Vanderploeg M, Kavarana M. Repair of anomalous right coronary artery from the pulmonary artery using the modified trapdoor technique. World J Pediatr Congenit Heart Surg 2019; 10(2): 192–196.
2. Askenazi J, Nadas AS. Anomalous left coronary artery originating from the pulmonary artery. Report on 15 cases. Circulation 1975; 51(6): 976–987.
3. Farwati M, Shaker F, Nasser M. Anomalous origin of the right coronary artery from the pulmonary artery in a morbidly obese patient presenting with chest pain. Am J Case Rep 2018: 19: 992–997.
4. Aljohani OA, Sah SP, Murthy RA, Lamberti JJ, Printz BF, Sun HY. Collateral deception: a unique presentation of an anomalous coronary artery. CASE 2018; 2(3): 89–91.

5. Williams IA, Gersony WM, Hellenbrand WE. Anomalous right coronary artery arising from the pulmonary artery: a report of 7 cases and a review of the literature. Am Heart J 2006; 152(5): 1004.e9–17.
6. VanLoozen D, Bykhovsky MR, Kapoor D, Bates WB, Murdison KA, Polimenakos AC. Myocardial ischemia and anomalous origin of the right coronary artery from the pulmonary artery in the adult: management implications and follow-up. World J Pediatr Congenit Heart Surg 2019; 1-3.
7. Saavedra MJ, Mozzi J, Napoli N, Villa A, Barretta J, Marantz P. Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) in an infant with a heart murmur. Case report. Arch Argent Pediatr 2018; 116(6): e789–e792.
8. Kim K, Jo E, Yu J, Kil H. Anomalous right coronary artery from pulmonary artery discovered incidentally in an asymptomatic young infant. Korean J Pediatr 2016; 59(1): S80–83.
9. Wu LP, Zhang YQ, Chen LJ, Liu YQ. Diagnosis of anomalous origin of the right coronary artery from the pulmonary artery by echocardiography. J Med Ultrason 2019; 46: 335–341.
10. Restrepo-Cordoba MA, Arellano-Serrano C, Mingo-Santos S. Right coronary artery with anomalous origin: the role of imaging techniques. JACC Cardiovasc Interv 2016; 9: e137–e139.
11. Brooks HS. Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery: with some remarks upon the effect of this anomaly in producing cirsoid dilatation of the vessels. J Anat Physiol 1885; 20(1): 26–29
12. Chernogrivov AE, Gornostaev AA, Chernogrivov IE et al. Anomalous origin of the right coronary artery from the pulmonary artery: surgical re-implantation into the aorta. Multimed Man Cardiothorac Surg 2015; pii. mmv024.
13. Modi H, Ariyachaipanich A, Dia M. Anomalous origin of right coronary artery from pulmonary artery and severe mitral regurgitation due to myxomatous mitral valve disease: a case report and literature review. J Invasive Cardiol 2010; 22: E49–55.
14. Ugan Atik S, Saltik L, Oztarhan K, Bornaum H. An incidentally detected anomalous origin of the right coronary artery from the pulmonary artery in an infant. Arch Argent Pediatr 2018; 116: e102–e105.
15. Rawala MS, Naqvi STS, Farhan K, Yasin M, Rizvi SB. Anomalous origin of a right coronary artery from pulmonary artery. Case Rep Cardiol 2018; (2583918): 1–3.
16. Zhang Y, Wang Z, Fang M, Jin Y, Wang H. Aortic implantation for anomalous connection of the coronary artery to the pulmonary artery in older children and adults. Thoracic Cardiovasc Surg 2017; 65(1): 18–25.
17. Radke PW, Messmer BJ, Haager PK, Klues HG. Anomalous origin of the right coronary Artery: preoperative and postoperative hemodynamics. Ann Thorac Surg 1998; 66(4): 1444–1449.
18. Mintz GS, Iskandrian AS, Bemis CE, Mundth ED, Owens JS. Myocardial ischemia in anomalous origin of the right coronary artery from the pulmonary trunk. Proof of a coronary steal. Am J Cardiol 1983; 51(3): 610–612.
19. Nakabayashi K, Okada H, Iwanami Y, Sugiura R, Oka T. Anomalous origin of the right coronary artery from the pulmonary artery diagnosed in an adult: a case report. J Cardiol Cases 2014; 10(3): 111–114.
20. Suzuki K, Yokochi K, Yoshioka F, Kato H. Anomalous origin of the right coronary artery from the pulmonary artery: report of a case. J Cardiogr 1985; 15(1): 241–248.
21. Ghanta RK, LaPar DJ, Zhang Q et al. Obesity increases risk-adjusted morbidity, mortality, and cost following cardiac surgery. J Am Heart Assoc 2017; 6(3): 1–8 pii: e003831.

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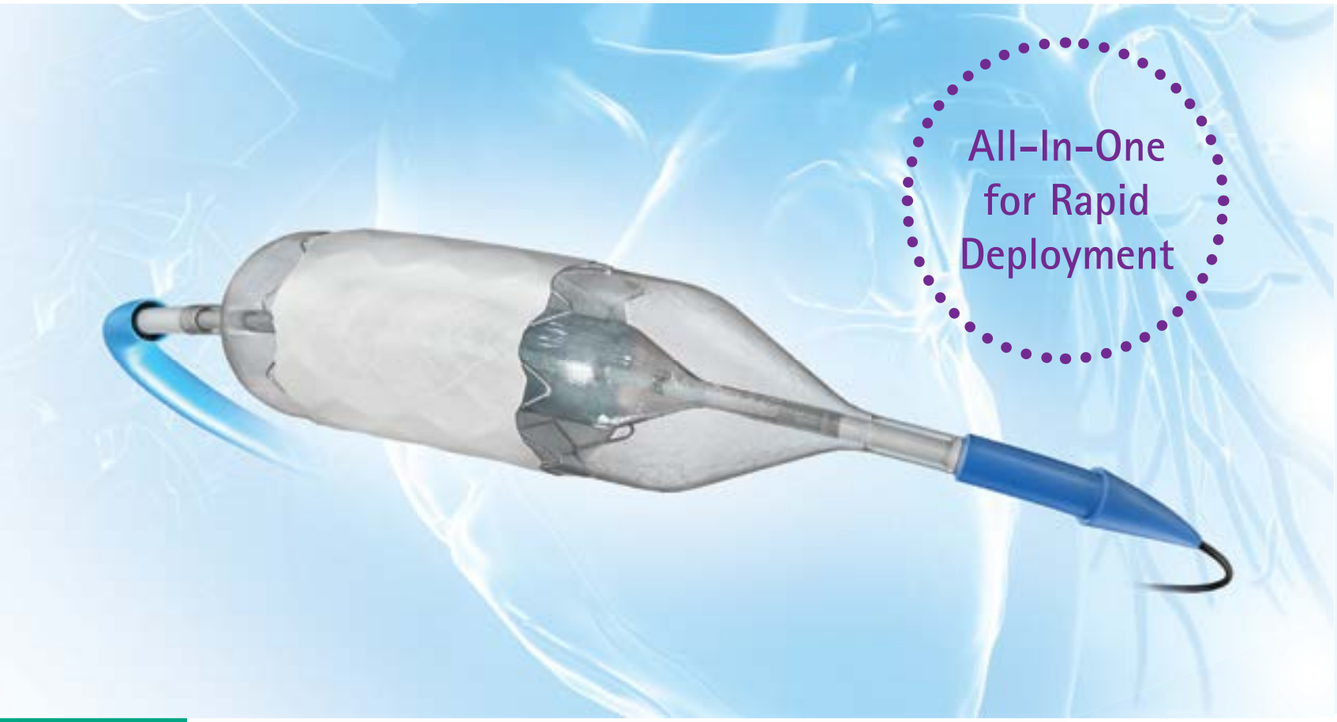

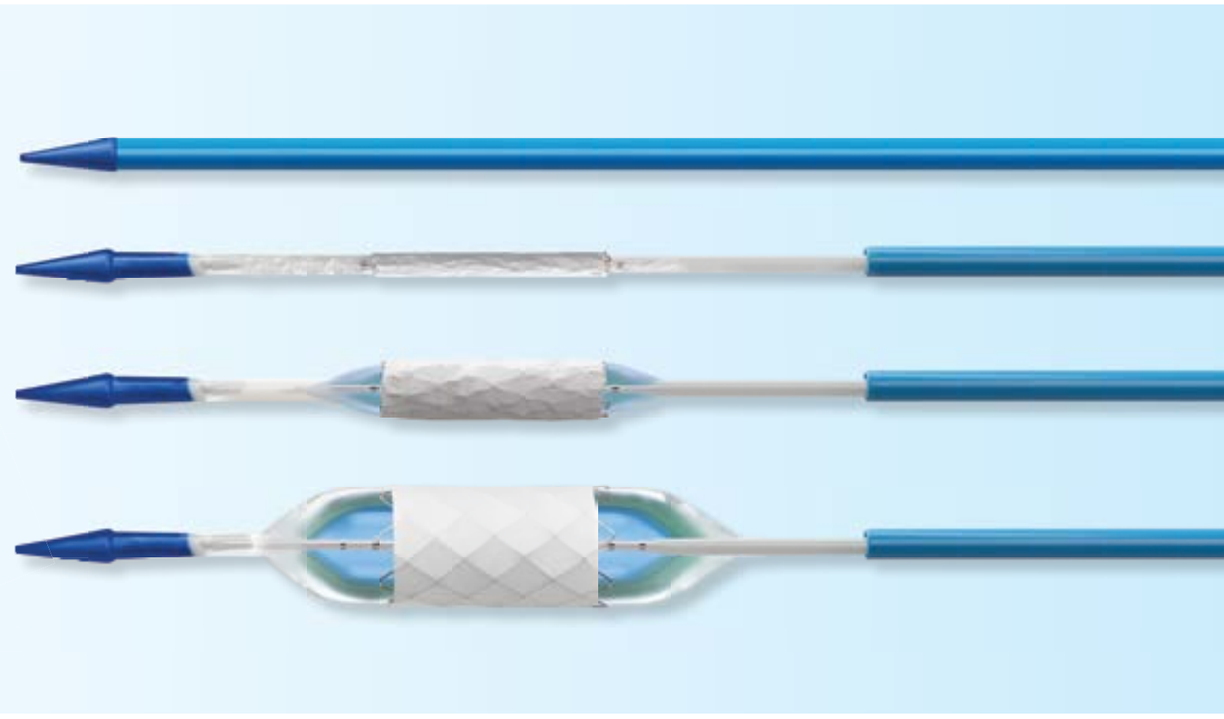

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CV-9107 6/19

Use of 3D Echocardiography in Diagnosis of Double-Orifice Left AV Valve in Neonates

Janelle Buysse, DO & Umang Gupta, MD

Introduction

Double-orifice left atrioventricular valve (DOLAVV) or double-outlet mitral valve (DOMV) is a rare congenital defect that can be seen with other congenital heart defects, most commonly Atrioventricular Septal Defects (AVSD), and can also be found in isolation. The hemodynamic significance of DOMV can vary from a normally functioning valve to a cause of significant stenosis or regurgitation. Regardless of hemodynamic significance, DOMV can pose a challenge to surgeons and complicate surgical repairs.[1] Therefore, the recognition and description of DOMV prior to surgical repair is important in surgical planning and prognostication. We present a case of DOLAVV where a non-sedated, three-dimensional (3D) transthoracic echocardiogram (TTE) was effectively utilized to diagnose the condition in a patient with AVSD when 2D and Doppler imaging could not clarify the diagnosis.

Case Presentation

A 12-month-old male presented to a local emergency department for evaluation of fever and respiratory distress. He was found to be influenza A-positive and was transferred to a pediatric unit in a community hospital for bronchiolitis treatment where a chest X-ray was obtained which showed cardiomegaly. An electrocardiogram (ECG) and transthoracic echocardiogram (TTE) were obtained. ECG showed biatrial enlargement, biventricular hypertrophy, right-axis deviation, and ST segment changes. TTE showed large Atrial Septal Defect (ASD), right-atrium (RA) and left-atrium (LA) dilation, right ventricle (RV) dilation, and concerns for pulmonary hypertension. No concerns for DOLAVV were raised. He was transferred to our pediatric intensive care unit. On arrival, further history revealed poor weight gain since around six months of age despite taking appropriate volumes of formula for age. Family denied any diaphoresis or fatigue with feeds, but reported noticing his "heart racing" through his chest that was more prominent during illnesses and had seemed to get more noticeable over the few months prior to admission. Physical examination on arrival revealed an active precordium with visible point of maximal impulse and regular rate and rhythm with normal S1 and S2. There was a III/VI systolic ejection murmur appreciated best at the left-upper sternal border. Repeat TTE at our institution revealed intermediate type AVSD with a single annulus and two separate atrioventricular (AV) valves with a large primum ASD and a small-inlet Ventricular

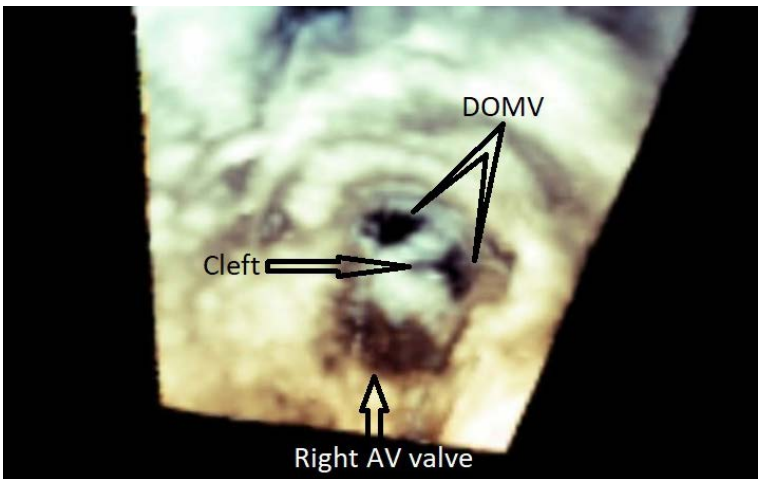


FIGURE 1 *TTE left AV valve with double orifice and cleft reconstructed from full volume 3D data set acquisition seen from ventricular side. DOMV = Double orifice mitral valve*

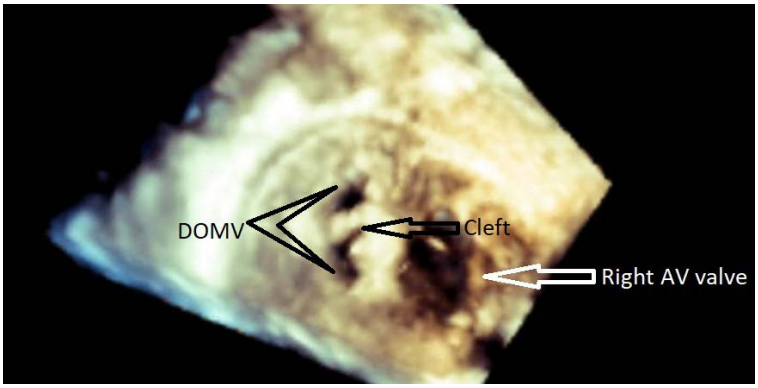


FIGURE 2 *TTE surgical view of DOMV and cleft reconstructed from full volume 3D data set acquisition. DOMV = Double orifice mitral valve*

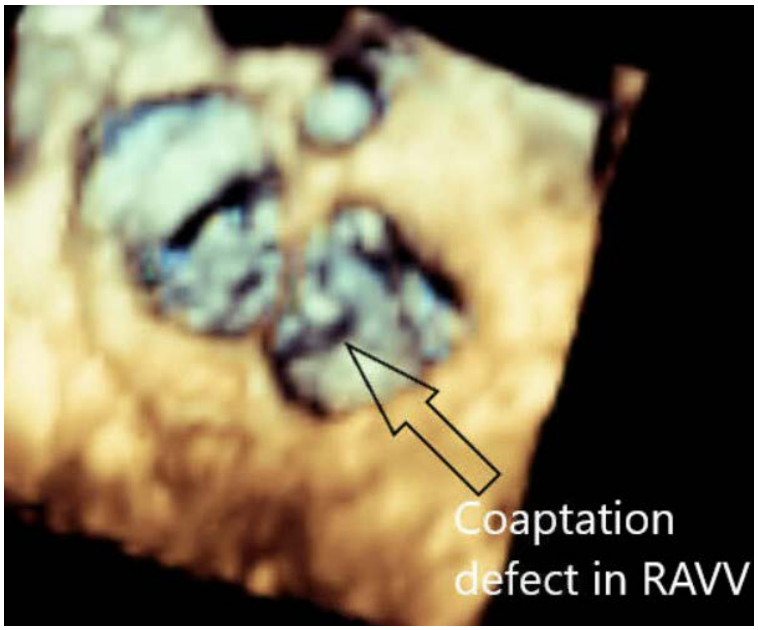
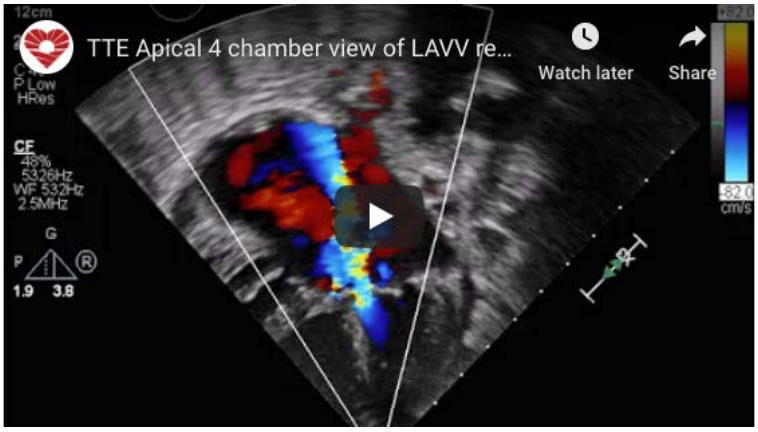


FIGURE 3 *Coaptation defect as the mechanism for right AV valve regurgitation reconstructed from 3D full volume data set as seen from atrial side. RAVV = Right AV valve*

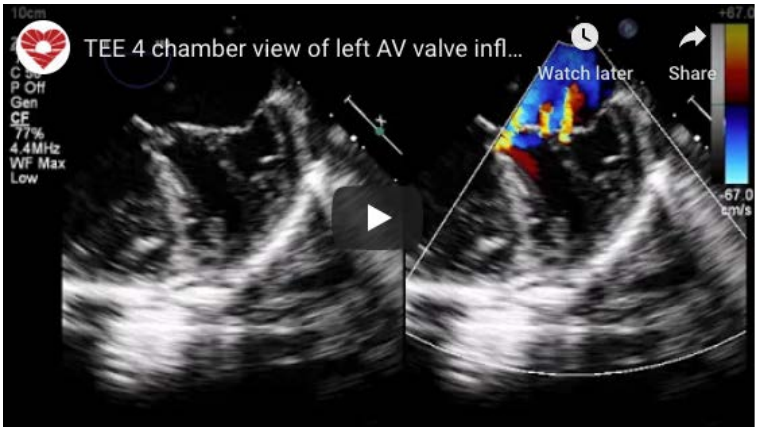
Septal Defect (VSD). There was mild-to-moderate right-sided AV valve regurgitation. The Left AV valve appeared to have cleft with left ventricle (LV) to RA shunt with no significant left AV valve insufficiency. There was some septal/left ventricular outflow tract (LVOT) attachment from left AV valve without any LVOT obstruction. There was dilation of RA, RV and main and branch pulmonary arteries. ECG showed sinus tachycardia with left axis deviation, right-atrial enlargement and biventricular hypertrophy. He was admitted for four days for influenza A bronchiolitis-related respiratory failure. He was started on scheduled furosemide and discharged home with close outpatient cardiology follow-up with anticipated surgical repair when he recovered from his viral illness.



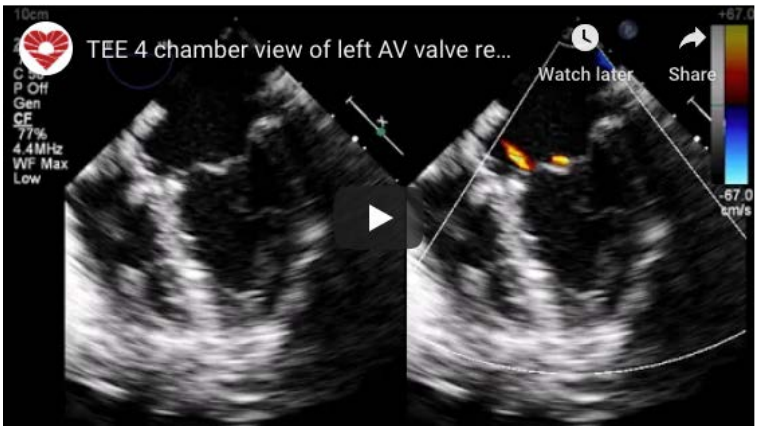
VIDEO 1 *TTE Apical 4 chamber view of LAVV regurgitation with 2 separate jets of regurgitation.*
Video: https://congenitalcardiologytoday.com/ed-resources/

VIDEO 2 *TTE Subcostal image of left AV valve en face.*
Video: https://congenitalcardiologytoday.com/ed-resources/

About six weeks after initial presentation, he underwent cardiac catheterization which showed Qp:Qs of 3.02 and normal pulmonary vascular resistance (indexed pulmonary vascular resistance of 1.48 Woods units x m2 on room air). While recuperating in the ICU after the procedure he underwent TTE with 3D imaging (using real time 3D imaging, single-beat full volume acquisition and zoom imaging with Phillips Epiq 7™ machine and post-processed on Phillips QLab™) in an attempt to better define the AV valves which confirmed the transitional AVSD with two separate AV annuli and valves. The right AV valve was dysplastic with poor cooptation resulting in moderate-to-severe insufficiency. The Left AV valve was found to have two separate orifices with a cleft in the more anteriorly located orifice. This had not been previously identified on 2D imaging and the valve was noted to be mild to moderately regurgitant (**Video 1-2, Figures 1-3**). He was scheduled for surgical repair the next week.



VIDEO 3 *TEE 4 chamber view of left AV valve inflow demonstrating 2 inflow jets.* **Video: https://congenitalcardiologytoday.com/ed-resources/**



VIDEO 4 *TEE 4 chamber view of left AV valve regurgitation demonstrating 2 regurgitant jets.*
Video: https://congenitalcardiologytoday.com/ed-resources/

Preoperative TEE showed findings consistent with previous TTE **(Video 3-4)**. The direct surgical inspection of the intracardiac anatomy revealed the presence of a partial AVSD with a primum defect and no clearly visible ventricular component. There was severe dysplasia of the right atrioventricular valve. The right atrioventricular valve demonstrated an area of deficiency of the superior bridging leaflet which was significant and associated with the presence of severe right-atrioventricular valve regurgitation. Inspection of the left atrioventricular valve confirmed the presence of a DOLAVV in which the smaller orifice was the one associated with the cleft. The area of the cleft resulted in lack of support of the edges and was associated with the left-atrioventricular valve regurgitation. The larger orifice was eccentric and located posterior inferiorly. The diameter of the larger opening was approximately 10 to 12 mm **(Figure 4)**. It was somewhat small, considering the expected diameter for a child of this weight and body surface area (BSA) is approximately 14 mm. Hence, partial closure of the cleft was performed due to concerns that completely closing the small orifice could have resulted in severe left-AV valve stenosis. The right AV valve was reconstructed and the ASD was closed with a pericardial patch. Postoperative TEE revealed adequate biventricular systolic function with mild-to-moderate right-AV valve regurgitation and mild left-AV valve regurgitation. Post-operative recovery was uneventful, and he was discharged home after five days on furosemide and enalapril. At subsequent outpatient follow-up

appointments, he has done well with no clinical concerns, improved weight gain and has been weaned off all his cardiac medications.

Discussion

DOLAVV is a rare congenital heart defect characterized by two distinct valvular orifices, each with their own chordal support and papillary muscles.[2] DOMV has been reported with variable frequency from 0.05% to 1% of all patients with congenital heart defects.[3] DOLAVV is most commonly seen associated with AVSD, as seen in our patient, though typically not described with intermediate-type AV septal defects. It can also be found with obstructive left-sided defects, cyanotic heart disease, and in association with LV non-compaction.[3] Retrospective reviews of patients who underwent repair for AVSD have reported the prevalence of DOLAVV in this patient population as 4%-6.7%.[1,3] The largest review of 44 patients with AVSD and DOLAVV demonstrated a predominance of partial AVSD among patients with DOLAVV (64% of patients) with complete AVSD accounting for 34% of the patients and only one patient with intermediate-type AVSD (2%).[1] Our patient falls into this latter, rarer, category of DOLAVV associated with intermediate-type AVSD.

Clinical spectrum of presentation can be quite varied for DOLAVV. Most have a functionally-normal valve with up to 25% having significant mitral stenosis or regurgitation.[4] However, among patients with AVSD, the presence of moderate-to-severe regurgitation has been reported as high as 80%.[1] DOLAVV has clinical significance beyond valve function; it has been reported to enhance severity of heart failure when found with a VSD and may amplify pulmonary hypertension.[5] Presence of DOLAVV complicates surgical repairs and requires careful consideration of surgical approach.[1,3] Despite the clinical significance of DOLAVV, identification remains challenging.

Due to the rarity of DOLAVV, there are no studies assessing accuracy of echocardiographic diagnosis. One case series of four patients with DOLAVV 2-dimensional (2D) and Doppler echocardiography failed to detect DOLAVV in 50% of the cases.[5] There are few case reports that describe the challenges of diagnosing DOMV and describe the use of 3D Transesophageal echocardiography as superior to 2D and Doppler.[6] Unfortunately, this is only possible in adults and currently there is no capability of performing 3D TEE-imaging in pediatric patients. Further 3D TTE-imaging in pediatric patients have long been hamstrung by requirement of children to remain still and their inability to breath hold. However, with advancement of 3D TTE-imaging technology, new opportunities have provided utilization of this modality in younger, unsedated children using real time 3D imaging, as well as single-beat acquisitions as we were able to demonstrate in this case.

Conclusion

DOLAVV is a rare entity found in isolation or with other congenital heart defects, most commonly partial AVSD. We present a rare case of DOLAVV associated with intermediate-type AVSD. Diagnosis of DOLAVV remains a challenge, especially in smaller children with other congenital heart diseases. The advancements in 3D TTE technology and our own skill sets in obtaining and processing images has provided additional tools that enable us to diagnose this condition as we have demonstrated in this case. While 3D imaging still has limitations, nevertheless, this should be attempted in targeted fashion with all children, especially if there are questions regarding the atrioventricular valves. Any additional information gathered would allow better surgical planning and prognostication.

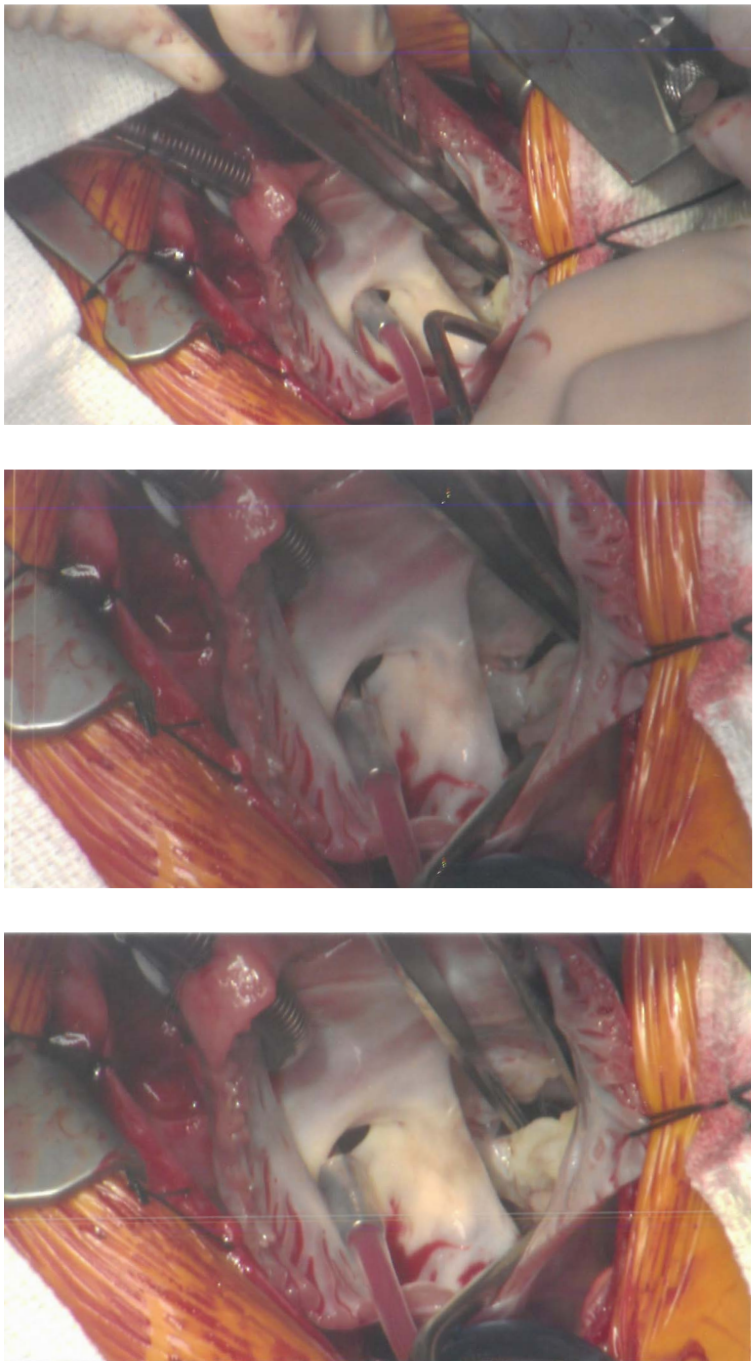


FIGURE 4 *Surgical images of DOLAVV*

Keywords
Double-orifice mitral valve, transitional AV canal

Declarations

Funding: No funding received
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References

1. Sharma V, Burkhart HM, Schaff HV, Cabalka AK, Grogan MA, Dearani JA. Double-orifice left atrioventricular valve in patients with atrioventricular septal defects: surgical strategies and outcome. Ann Thorac Surg. 2012;93(6):2017-2020; discussion 2020-1. doi:10.1016/j.athoracsur.2012.02.055.
2. Nielsen JC, Panesar LE. Mitral Valve and Left Atrial Anomalies. Lai WW, Mertens LL, Cohen MS, Geva T, eds. Echocardiogr Pediatr Congenit Heart Dis Fetus Adult. Published online 2016:244.
3. Pontailler M, Haidar M, Méot M, et al. Double orifice and atrioventricular septal defect: dealing with the zone of apposition. Eur J Cardiothorac Surg. 2019;56(3):541-548. doi:10.1093/ejcts/ezz085.
4. Bhattacharyya S, West C, Chinasamy D, Senior R, Li W. Utility of three-dimensional echocardiography for assessment of double-orifice mitral valve. Eur Heart J Cardiovasc Imaging. 2012;13(8):672. doi:10.1093/ehjci/jes061.
5. Ito-Akabori S, Nakagawa M, Okamoto N, et al. Clinical characteristics and diagnosis of double-orifice left atrioventricular valve associated with other congenital heart disease. Heart Vessels. 2005;20(6):286-289. doi:10.1007/s00380-004-0818-0.
6. Kim IC, Cho YK, Kim H, Park NH, Kim KB. Three-dimensional echocardiographic reconstruction of double-orifice mitral valve and mitral leaflet prolapse. Circulation. 2014;130(10):e87-8. doi:10.1161/CIRCULATIONAHA.114.011373.

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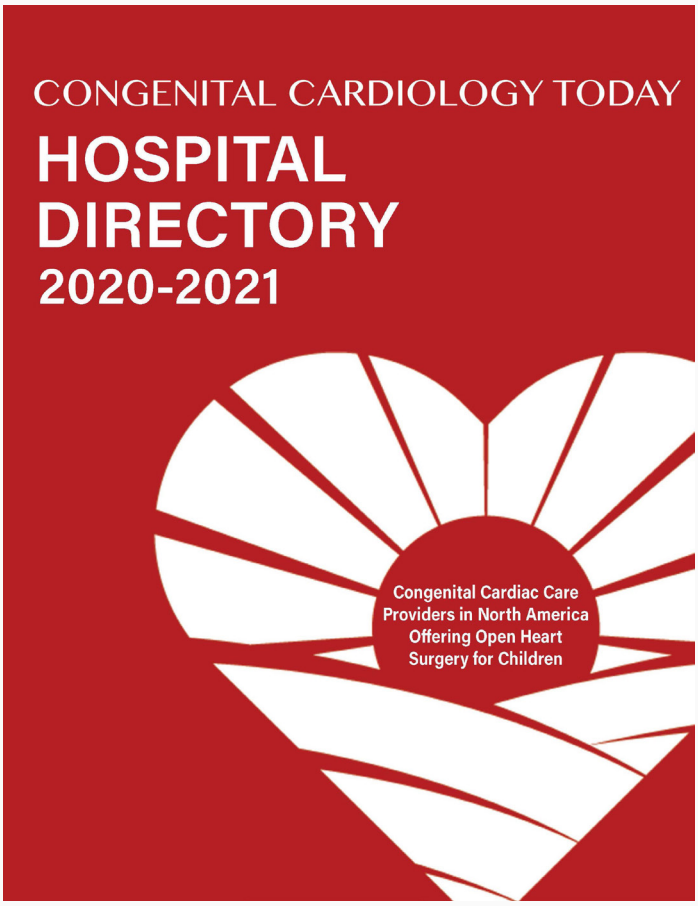

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Pediatric Cardiac Critical Care Collaborative Credited with Saving Lives and Preventing Cardiac Arrests in Children with Critical Heart Disease

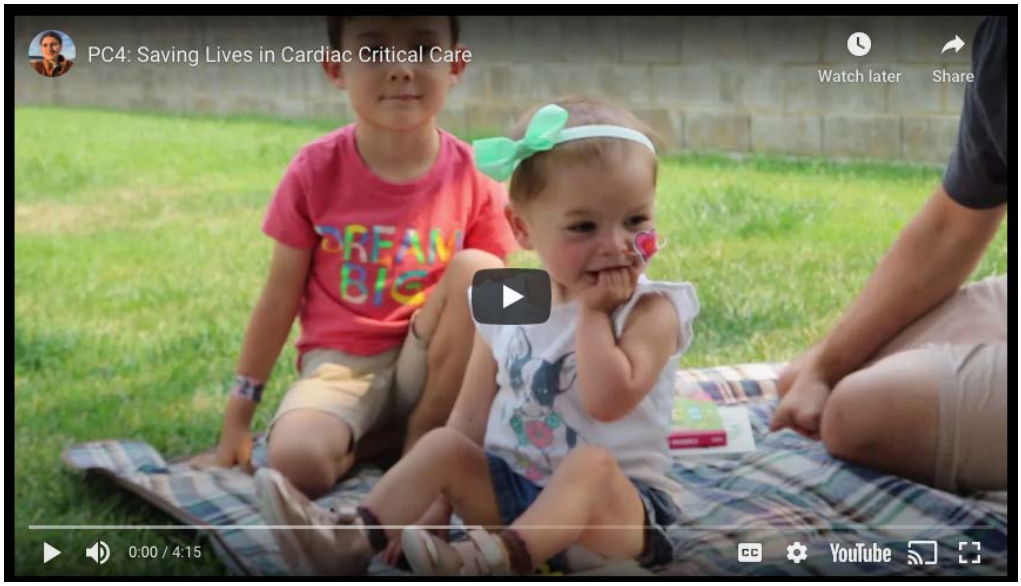
Saving over 160 lives and reducing the incidence of cardiac arrest by nearly half, PC4's fundraising initiative launched to continue life-saving mission

PRNewswire/ -- The Pediatric Cardiac Critical Care Consortium (PC4) announced today remarkable progress in improving outcomes in children with critical heart disease. At the 16th Annual International Meeting of the Pediatric Cardiac Intensive Care Society, both physicians and hospital administrators applauded PC4 for the impact of the collaborative on preventing cardiac arrest and saving children's lives.

The average person is often unaware of the significance of Congenital Heart Disease. Congenital heart disease impacts 1 in 100 children born each year, with most of them needing open-heart surgery and a stay in the intensive care unit to survive. One in 12 newborns suffers a cardiac arrest after open heart surgery and only half survive.

Speaking this week at the Pediatric Cardiac Intensive Care Society meeting, Kay Stewart-Huey, Vice President of the Children's Heart Center at Children's Healthcare of Atlanta, praised PC4. "Through participation in the PC4 Cardiac Arrest Prevention national collaborative we were able to reduce our incidence of cardiac arrest. PC4 enables hospitals to improve the quality of their clinical care by facilitating comparison of many outcomes with other high performing centers and sharing best practices with one another."

PC4 started in 2013, just five hospitals with a grant from the National Institutes of Health (NIH) and an overarching goal to improve the experience, care and outcomes for these vulnerable children with heart disease. PC4 has now grown to over 60 intensive care units across North America. PC4 hospitals believe in collaborative learning as a way to improve the care of patients and families fighting critical cardiovascular disease. This includes sharing best practices, promoting teamwork, working together on innovative projects, and communicating important findings across hospitals. Applying these practices across intensive care units, PC4 has recently reported saving over 160 lives and reducing the incidence of cardiac arrest by nearly half.



Samantha and Julian are two young cardiac patients cared for at PC4 hospitals. This video shows their stories and highlights how the Pediatric Cardiac Critical Care Consortium works towards the goal of improving outcomes of patients with congenital heart disease across the country.
Video: https://youtu.be/YEkNI4OqGfI

Launched with funding from the University of Michigan, the organization has made considerable contributions to the medical field, including over 30 publications in the past five years. For the first time, PC4 is reaching out to the community to help give hope to families of children with heart disease.

Dr. Sarah Tabbutt, the Executive Director of PC4, and a cardiac intensive care doctor at the University of California San Francisco Benioff Children's Hospital, says the organization has made so much progress, but will need generosity to help discover more ways to improve outcomes in these fragile children. We have created such a robust infrastructure and collaboration within PC4, that we really can answer questions which were previously un-answerable. PC4 is already underway on several important initiatives directed at shortening the time in the hospital, avoiding unexpected additional procedures, equalizing care across race/ethnicity, and reducing pain and anxiety after open-heart surgery. "The PC4 community is passionate about finding ways not only to improve care, but to give a brighter future to these children and their families." PC4 has a fundraising site where you can see heartwarming stories in addition to details of upcoming projects that donations will help fund.

PC4 fundraising site:
https://give.communityfunded.com/o/michigan-medicine/i/pc4/s/pc4

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