Patent Ductus Arteriosus in Pregnancy: Cardio-Obstetrics Management in a Late Presentation

Zhihang Zhang, MD, Aaron Wengrofsky, MD, Diana S. Wolfe, MD, MPH, FACOG, Nicole Sutton, MD, FAAP, FACC, FSCAI, Manoj Gupta, MBBS, MD, Daphne T. Hsu, MD, FAAP, FACC, and Cynthia C. Taub, MD, MBA, *Bronx, New York*

INTRODUCTION

Heart disease is the leading cause of maternal death during pregnancy, and congenital heart disease is dangerous and potentially fatal if not recognized in a pregnant woman. Intracardiac shunts, such as patent ductus arteriosus (PDA), can be particularly hazardous to miss. When unrepaired, they can lead to Eisenmenger syndrome, which carries a 50%-65% mortality rate during pregnancy.[1] Physiologic changes in pregnancy impose further stress on those with congenital heart disease; for example, the decreased systemic vascular resistance that occurs during pregnancy increases right-to-left shunting, leading to reduced pulmonary perfusion and subsequent hypoxemia.[2] This will lead to further decompensation and an increase in maternal and fetal morbidity and mortality.

Here we describe a case of a vaginal delivery in a patient whose PDA was discovered in the third trimester and was noted to have significant left ventricular (LV) dilatation, which placed the patient at increased risk of decompensation and mortality. Due to the coordinated efforts of our Maternal Fetal Medicine (MFM) Cardiology Joint Program, the delivery was successful with no adverse effects to either mother or baby. The PDA was subsequently closed postpartum without complications, and the patient had significant improvement in LV size on routine follow-up. This case highlights the importance of a multidisciplinary team approach to pregnant patients with congenital heart disease.

CASE PRESENTATION

A 24-year-old female at 35 weeks' gestation was referred by the Montefiore Medical Center regional perinatal network to our MFM-Cardiology Joint Program. An astute midwife auscultated a murmur on physical exam. The patient had recently arrived in the United States from Guatemala. Her family was told that she had a murmur at birth, but it was expected to resolve as she grew up. She had no other past medical history and only took prenatal vitamins. She denied any shortness of breath, dyspnea on exertion, orthopnea, chest pain, palpitations, dizziness, or syncope.

Cardiovascular exam was significant for a 4/6 continuous machinery-like murmur radiating from the left sternal border throughout her precordium to the left scapular area and a palpable thrill in the suprasternal notch. Her blood pressure was 100/50 mm Hg with a widened pulse pressure, heart rate 78 bpm, and oxygen saturation 98% on room air. There was no pedal edema, and lungs were clear to auscultation.

Electrocardiogram showed normal sinus rhythm with voltage criteria for LV hypertrophy. Her initial transthoracic echocardiogram (TTE) showed severely dilated LV with low normal LV ejection fraction (LVEF) of 50%. Left ventricular end-diastolic diameter (LVEDD) was 7.2 cm (Figure 1, Video 1). Her left atrium (LA) anteroposterior dimension was measured at 6.1 cm. She had qualitatively normal right ventricle (RV) function.

Further investigation discovered a large PDA measuring 9 mm across the waist (Figure 2, Videos 2, 3, 4). There was no evidence of pulmonary hypertension. The peak systolic gradient across the PDA was 81 mm Hg (Figure 3). The calculated pulmonary artery systolic pressure (PASP) was 20 mm Hg, and Qp/Qs was calculated as 4:1.

The patient was seen at the MFM-Cardiology Joint Program, where both MFM and cardiology specialists completed the assessment together. After multidisciplinary rounds it was determined that there was no need to urgently close her PDA predelivery, since PDA closure often results in worsening LV function. Instead, her volume status was followed clinically at each prenatal visit with MFM. The plan was for labor induction at around 39 weeks given the increased risk of stillbirth in all pregnant patients with congenital heart disease and to avoid development of preeclampsia.[3]

The patient had a successful induced vaginal delivery of a healthy baby boy at 38 weeks 6 days with early epidural anesthesia. Due to the risk of volume overload from the large autotransfusion of blood during the immediate postpartum state, she was transferred to the cardiac intensive care unit immediately postdelivery for close hemodynamic monitoring. She was given intravenous furosemide and remained hemodynamically stable. She was discharged on postpartum day 3. On the postdelivery TTE, the LVEDD was 7.6 cm, the peak systolic gradient across the PDA was 73 mm Hg, and the LVEF remained low normal, not significantly changed from predelivery.

Given the large size of her PDA, she ultimately had an outpatient PDA closure with a 12 mm Amplatzer muscular ventricular septal defect (VSD) occluder at 6 weeks postpartum (Figure 4A, B). Her immediate post-PDA closure TTE showed an LVEDD of 6.5 cm with resolution of the PDA (Video 5). Eight months later, her TTE showed an LVEDD of 5.1 cm and mildly decreased LVEF of 45% (Figure 5A, B, Video 6).

From the Department of Cardiology (Z.Z., C.C.T.), Department of Medicine (A.W.), Department of Obstetrics and Gynecology (D.S.W.), and Department of Pediatric Cardiology (N.S., M.G., D.T.H. ,C.C.T), Montefiore Medical Center, Bronx, New York.

Keywords: Unrepaired patent ductus arteriosus, Congenital heart disease, Cardio-obstetrics

Conflicts of Interest: The authors reported no actual or potential conflicts of interest relative to this document.

2468-6441

https://doi.org/10.1016/j.case.2020.12.002

VIDEO HIGHLIGHTS

Video 1: Initial adult TTE parasternal long-axis view.
Video 2: Preclosure TTE with large PDA short-axis view.
Video 3: Initial TTE PDA in suprasternal view off axis.
Video 4: Initial TTE PDA in suprasternal view with color.
Video 5: Post-PDA closure TTE parasternal long-axis view.
Video 6: Post-PDA closure TTE short-axis view.

View the video content online at www.cvcasejournal.com.

DISCUSSION

Congenital heart disease poses a great risk to both mother and baby during pregnancy. In a study with over 26,000 births, maternal mortality was 18-fold higher in those with congenital heart disease compared with those without.[4] Additionally, there have been many cases of pregnant women presenting late in their pregnancy with complications such as preeclampsia, HELLP (hemolysis, elevated liver enzymes, low platelet) syndrome, and decompensated heart failure due to previously undiagnosed PDA.[1] This illustrates the importance of a coordinated effort to both diagnose and manage a patient with an unrepaired PDA early in pregnancy.

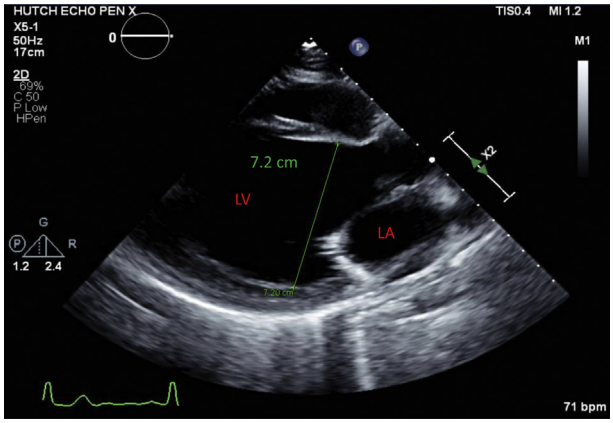

Figure 1 Initial adult TTE LVEDD of 7.2 cm.

Maternal Fetal Medicine and Cardiology at Montefiore Medical Center created a joint outpatient clinic program in 2015 to serve as a multidisciplinary team for optimizing the care of high-risk pregnancies. During a visit, the patient has a same-day TTE and is seen by both a cardiologist and an MFM physician. The team approach was central to the management of this case so that all specialists involved understood the patient's physiology and peripartum risk for heart failure.[5]

This patient had a large PDA with low-normal LVEF along with severely dilated LA and LV, which placed her at high risk for peripartum decompensation. Ideally, her PDA should have been closed prior to attempted pregnancy. However, when she presented at 35 weeks' gestation, the patient was at maximal volume expansion, and the risks of PDA closure outweighed the benefits. If she had presented with such a notably dilated LV in the first or second trimester, she likely would not be able to wait for postdelivery PDA closure. Making this management decision required thoughtful discussion with a multidisciplinary team.

Patent ductus arteriosus has an incidence of one in 2,000 births, and the female-to-male ratio is 2:1.[6] In the normal heart, the ductus arteriosus is a vital connection for fetal circulation, connecting the pulmonary artery with the descending aorta, but it usually functionally closes within 72 hours of birth. Small PDAs have a pulmonary flow (Qp) to systemic flow (Qs) ratio (Qp/Qs) < 1.5, moderate PDAs have a Qp/Qs between 1.5 and 2.2, and large PDAs have a Qp/Qs > 2.2.[7] Untreated PDA leads to volume overload with LA and LV dilatation. The amount of flow through the PDA is related to the size of the PDA and the relative pulmonary vascular resistance and systemic vascular resistance.[7]

In this patient's initial TTE, the PASP was calculated by taking the systolic blood pressure and subtracting the PDA gradient, and in the absence of significant RV outflow obstruction, it is equitable to the

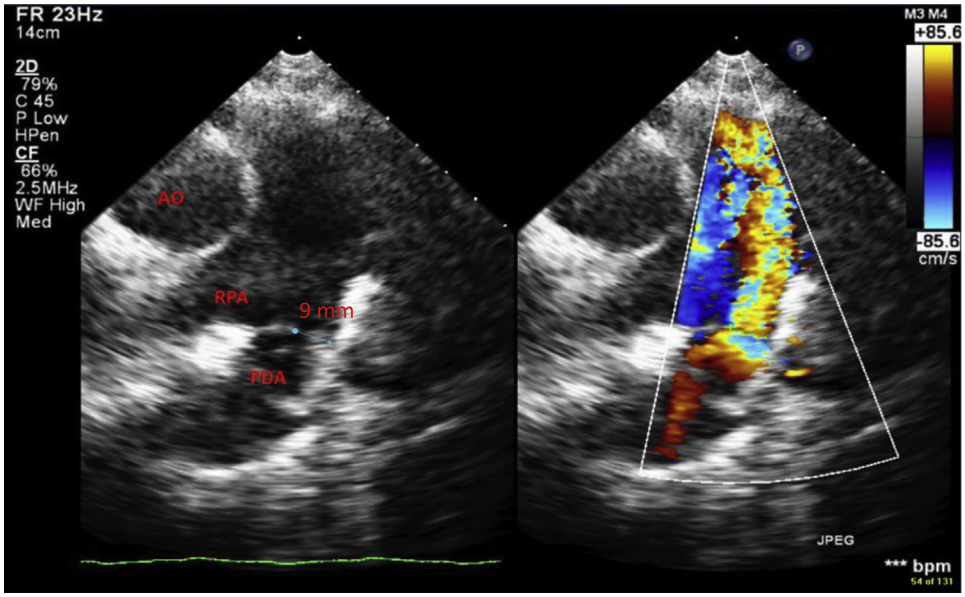

Figure 2 TTE with large PDA of 9 mm.

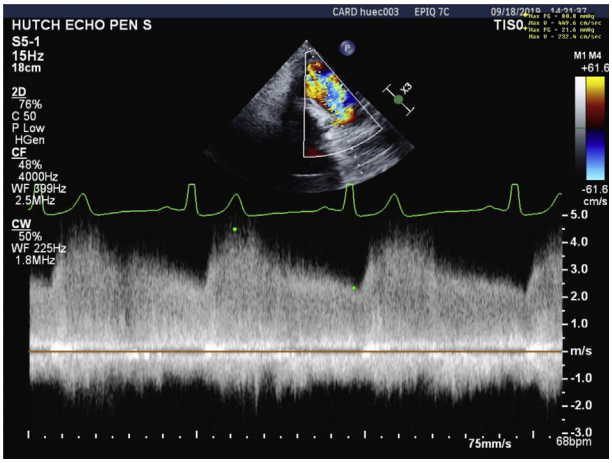

Figure 3 Initial TTE PDA with continuous-wave Doppler peak gradient 80 mm Hg across PDA.

RV systolic pressure. In this patient, the Qp/Qs was 4:1; with a PASP of 20 mm Hg; this is consistent with a large PDA without significant pulmonary hypertension.

Pregnancy increases the circulating maternal blood volume by about 40% for a singleton gestation and maternal cardiac output by 30%-50%, as well as LV mass. During labor, there is an additional rise in blood volume up to 10 L. At the time of delivery, "autotransfusion" of blood from the uterus into the heart occurs with the separation of the placenta, and after delivery, blood from the lower extremity venous system may overload the cardiovascular system.[8] Potential adverse effects of untreated PDAs during pregnancy include LV failure, Eisenmenger syndrome, and infective endocarditis.

In women with unrepaired PDAs, vaginal delivery is preferred over cesarian section, due to the increase in mortality seen with the latter.[9] In the peripartum setting, supplemental oxygen should be considered, which works as a pulmonary vasodilator to decrease blood flow across the right-to-left shunt. Anticoagulation is controversial in these patients; patients with unrepaired PDA and resulting pulmonary hypertension are at risk for pulmonary vascular thrombosis but are also at risk of pulmonary hemorrhage, in addition to blood loss during delivery.[2]

Patients with moderate or large PDAs with left-to-right shunting, signs of left-sided overload (LA dilatation and/or LV enlargement), or pulmonary arterial hypertension have a grade 1B recommendation to close the PDA.[6] Any PDA with previous endocarditis has a grade 1C indication for intervention.[6] For intervention type, there is a grade 1B recommendation to perform percutaneous PDA closure

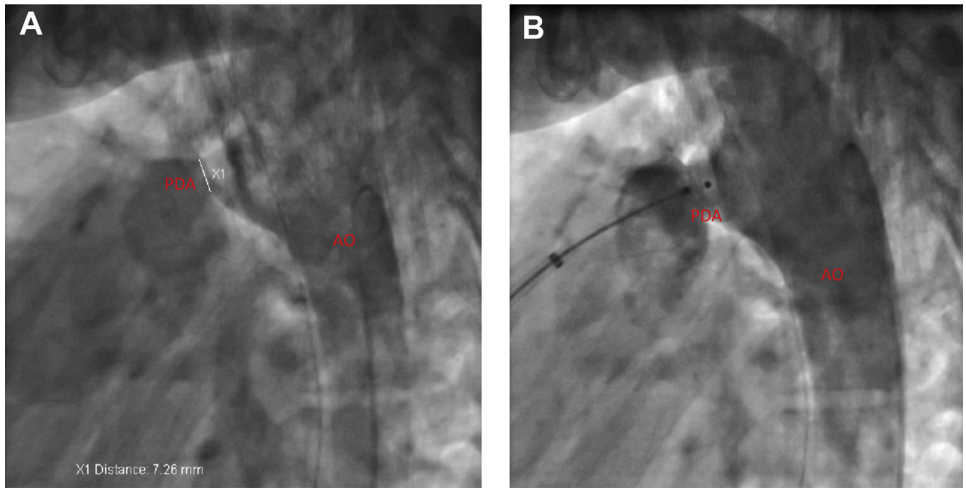

Figure 4 **(A)** PDA visualized on cardiac catheterization; **(B)** PDA with Amplatzer VSD occlusion device.

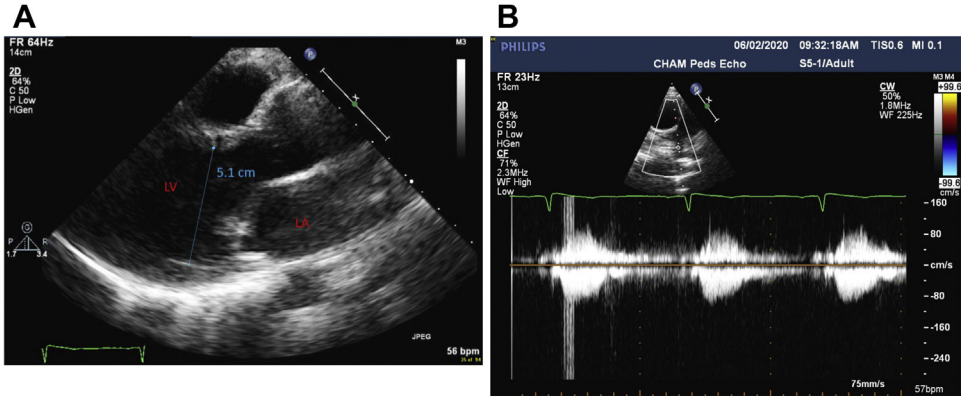

Figure 5 **(A)** Post-PDA closure TTE LVEDD of 5.1 cm. **(B)** Follow-up TTE closure of PDA continuous-wave Doppler.

rather than surgical ligation in adolescents and adults. Postclosure, prophylactic antibiotics for endocarditis are recommended for 6 months.

Strategic selection for PDA closure is driven by duct size at its narrowest point, ampulla size and shape, and patient age and weight. It is generally possible to occlude most ducts <5 mm with coils, whereas ducts >8 mm are better occluded with a variety of different devices.[10] The Amplatzer ductal occluder was approved by the Food and Drug Administration in 2003 for PDA occlusion in individuals over the age of 6 and has the capability of closing PDAs of up to 11-12 mm in diameter. The Amplatzer VSD muscular occluder, used off-label in this case, is approved for congenital VSD closure and has a diameter from 4 to 18 mm but has a long history of being used off-label to close large PDAs due to its larger maximum diameter.[11,12]

A retrospective analysis of 447 patients with PDA and normal LVEF showed that 22.7% of patients had LV dysfunction after closure. The larger the PDA, the higher the risk of LV dysfunction, with 91% of patients having LV dysfunction when PDA sizes are >9 mm, but only 2.8% of patients with PDA of <3 mm had LV dysfunction.[13] Other rare and severe complications following PDA closure include infective endocarditis, encroachment of the left pulmonary artery, and narrowing of the descending aorta. A delayed PDA closure strategy at 6 weeks postpartum served our patient well.

CONCLUSION

We present a rare case of a large, unrepaired PDA discovered late in pregnancy, which poses management challenges. This case demonstrates that with a multidisciplinary team of interventional cardiologists, maternal fetal medicine obstetricians, congenital heart disease specialists, cardiac imaging specialists, and obstetric-cardiac anesthesiologists, safe vaginal delivery can be achieved with delayed PDA occlusion postpartum.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2020.12.002.

REFERENCES

1. Vashisht A, Katakam N, Kausar S, Patel N, Stratton J. Postnatal diagnosis of maternal congenital heart disease: missed opportunities. BMJ Case Rep 2015. https://doi.org/10.1136/bcr-2015-209938.
2. Yuan SM. Eisenmenger syndrome in pregnancy. Brazil J Cardiovasc Surg 2016;31:325-9.
3. Kloster S, Tolstrup JS, Olsen MS, Johnsen SP, Søndergaard L, Nielsen DG, et al. Neonatal risk in children of women with congenital heart disease: a cohort study with focus on socioeconomic status. J Am Heart Assoc 2019;8:e013491.
4. Karamlou T, Diggs BS, McCrindle BW, Welke KF. A growing problem: maternal death and peripartum complications are higher in women with grown-up congenital heart disease. Ann Thoracic Surg 2011;92:2193-9.
5. Wolfe DS, Hameed AB, Taub CC, Zaidi AN, Bortnick AE. Addressing maternal mortality: the pregnant cardiac patient. Am J Obstet Gynecol 2019;220:167.e1-8.
6. Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Circulation 2019;139:e698-800.
7. Sommer RJ, Hijazi ZM, Rhodes JF Jr. Pathophysiology of congenital heart disease in the adult. Part I: shunt lesions. Circulation 2008;117:1090-9.
8. Sanghavi M, Rutherford JD. Cardiovascular physiology of pregnancy. Circulation 2014;130:1003-8.
9. Brickmer EM. Cardiovascular management in pregnancy: congenital heart disease. Circulation 2014;130:273-82.
10. Atiq M, Aslam N, Kazmi KA. Transcatheter closure of small-to-large patent ductus arteriosus with different devices: queries and challenges. J Invasive Cardiol 2007;19:295-8.
11. Fernando R, Koranne K, Loyalka P, Kar B, Gregoric I. Patent ductus arteriosus closure using an Amplatzer(™) ventricular septal defect closure device. Experi Clin Cardiol 2013;18:e50-4.
12. Kanabar K, Bootla D, Kaur N, Pruthvi CR, Krishnappa D, Santosh K, et al. Outcomes of transcatheter closure of patent ductus arteriosus with the off-label use of large occluders (≥16 mm). Indian Heart J 2020;72:107-12.
13. Kiran VS, Tiwari A. Prediction of left ventricular dysfunction after device closure of patent ductus arteriosus: proposal for a new functional classification. Eurointervention 2018;13:e2124-9.