Bilateral Ductus Arteriosus and Discontinuity of the Pulmonary Branches and Pulmonary Atresia: An Unusual Anatomy Diagnosed by Echocardiography

Tamika K. Rozema, MD, Ravi Ashwath, MD, and Christopher S. Snyder, MD, *Cleveland, Ohio; and Iowa City, Iowa*

INTRODUCTION

This case report describes a neonate that was found to have complex intracardiac anatomy in addition to discontinuous pulmonary arteries that were supplied by bilateral patent ducti. Initial treatment was bilateral transcatheter stenting of the ducti to allow for growth prior to complete repair. To our knowledge, this case is the first reported bilateral ducti transcatheter stenting via the femoral approach with expected results and demonstrates the vital role echocardiograms play in the diagnosis and management of these patients.

CASE PRESENTATION

A 22-year-old woman was referred for fetal echocardiography at 26 weeks' gestational age; imaging demonstrated double-outlet right ventricle (DORV), malposed great arteries, large subpulmonic ventricular septal defect with pulmonary atresia, and poorly visualized branch pulmonary arteries because of poor acoustic windows. The patient was born at 38 weeks' gestational age after an otherwise uncomplicated pregnancy and delivery. The patient was stable on room air with normal vital signs, including peripheral capillary oxygen saturation of 96% and a single S2. Initial arterial blood gas on room air demonstrated a pH of 7.32, partial pressure of carbon dioxide of 31 mm Hg, partial pressure of oxygen of 51 mm Hg, and lactate of 1.2 mmol/L. Prostaglandin E (PGE) infusion was started immediately after birth.

Transthoracic echocardiography confirmed the diagnosis of DORV with malposed great arteries, large ventricular septal defect, and pulmonary atresia with discontinuous pulmonary arteries supplied by bilateral patent ducti (Figures 1 and 2, Video 1). DORV was diagnosed on the basis of >50% aortic override and lack of aortic-to-mitral fibrous continuity. The left pulmonary artery arose from a reverse-angle patent ductus arteriosus that originated from the undersurface of the left-sided transverse aortic arch (Video 2). The right pulmonary artery arose from the ductus arteriosus that originated from the base of the right innominate artery (Video 3). The point of origin of the vessels from the undersurface of the aorta was more suggestive of patent ducti instead of aortopulmonary (AP) collaterals. Transthoracic echocardiography was vital in determining this unusual anatomy, understanding the critical nature of the lesion, and supporting the need for PGE to maintain pulmonary artery blood flow. Subcostal coronal sweeps helped in delineating the aortic arch, pulmonary artery, ductal connections, and their relationships. High parasternal short-axis imaging was also helpful in delineating the discontinuous pulmonary arteries and ductal connections. Color Doppler showing continuous flow in the duct connecting the aorta and pulmonary arteries was helpful in strengthening the two-dimensional diagnosis. Computed tomographic angiography (CTA) was performed primarily to confirm the diagnosis and also to delineate coronary anatomy for completion and further management planning (Figures 3 and 4).

After a thorough review of all of the patient's data and discussion with our team and the family, the decision was made to palliate the neonate by bilateral ductal stenting using a femoral approach. Three Integrity coronary artery stents (Medtronic, Minneapolis, MN) were placed in the right patent ductus arteriosus, and two Integrity coronary artery stents were placed in the left patent ductus arteriosus (Figure 5). The systemic oxygen saturations off PGE after catheterization remained unchanged (90% range). The patient tolerated the procedure without complication but was not discharged home until 6 days later because of feeding issues.

DISCUSSION

Bilateral patent ducti are an uncommon abnormality usually associated with pulmonary atresia and discontinuous branch pulmonary arteries and can be mistaken for AP collaterals if full-color and spectral Doppler assessment is not performed.[1] They are commonly associated with heterotaxy syndrome. This case is unique in that the patient had pulmonary atresia with discontinuous pulmonary arteries in the setting of DORV with malposed great arteries. Two previously reported case series from the Hospital for Sick Children in Toronto over 37 years described 38 cases of bilateral patent ducti.[2,3] The cases presented included none in isolation, associated with congenital heart disease including various forms of pulmonary atresia, aortic arch anomalies including aortic atresia or interrupted aortic arch, and isolation of one subclavian artery.[2,3] This wide range of presentation illustrates and underscores the range seen in pediatric cardiology and the importance of understanding the embryology of the paired primitive aortic arch system.

From the The Congenital Heart Collaborative, Pediatric Cardiology Division, Rainbow Babies and Children's Hospital, Cleveland, Ohio (T.K.R., C.S.S.); and the Pediatric Cardiology Division, University of Iowa Stead Family Children's Hospital, Iowa City, Iowa (R.A.).

Keywords: Discontinuous branch pulmonary arteries, Pulmonary atresia, Bilateral patent ducti

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

Copyright 2017 by the American Society of Echocardiography. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

2468-6441

https://doi.org/10.1016/j.case.2017.09.007

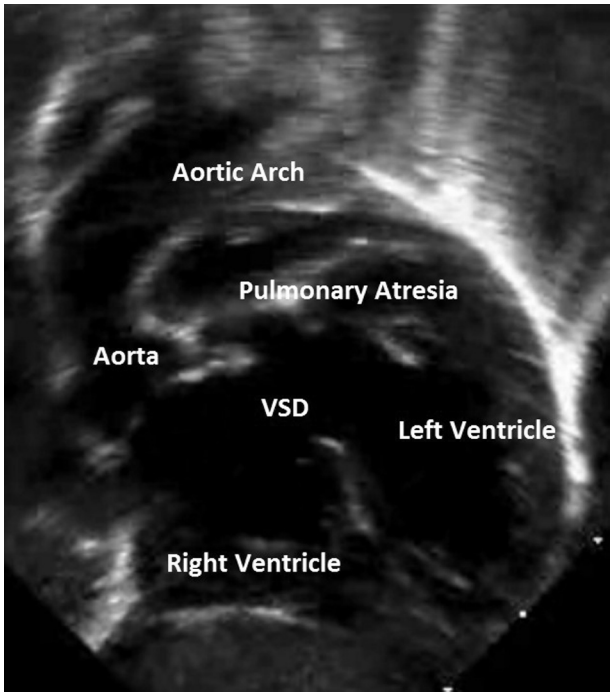

Figure 1 Intracardiac anatomy, DORV, malposed great arteries, and large vetricular septal defect (VSD) with pulmonary atresia.

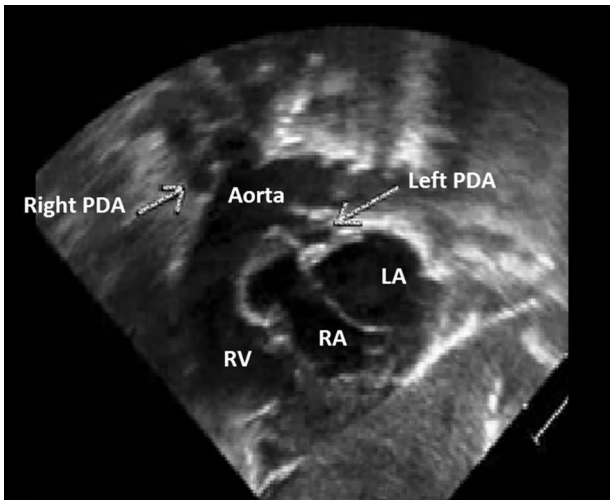

Figure 2 Bilateral patent ductus arteriosus (PDA) arising from separate origins from the undersurface of the aortic arch. *LA*, Left atrium; *RA*, right atrium; *RV*, right ventricle.

Fetal diagnosis of this condition can be quite challenging because of multiple levels of interconnecting aspects. Echocardiographic windows may be limited because of maternal factors, lack of cooperation from the fetus, and limitations in assessing small structures. Discontinuous pulmonary arteries were considered in the diagnosis of this case with supply from either bilateral ducti or AP collaterals. One way to assess if the vessels are ductal or collateral tissue is to stop PGE infusion and assess for changes in the vessels. If there is narrowing of the vessels that then responds to PGE, this is more suggestive of ductal tissue and not AP collaterals.

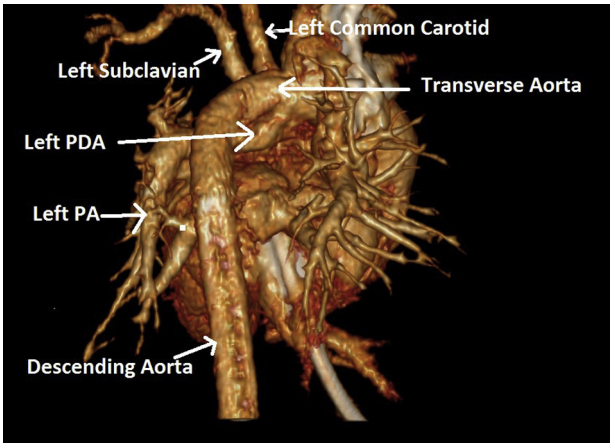

Figure 3 Three-dimensional reconstruction of the aortic arch, focusing on the left patent ductus arteriosus (PDA) and left pulmonary artery (PA).

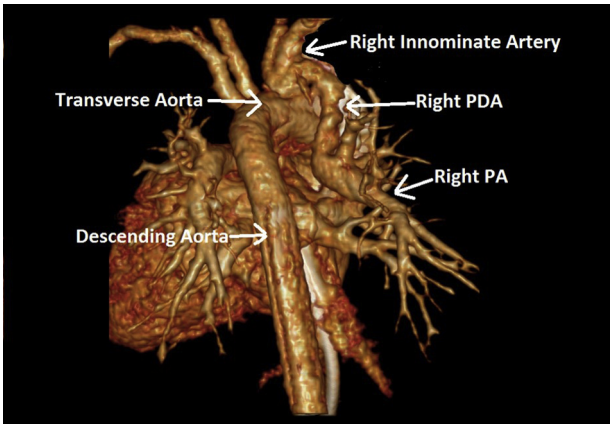

Figure 4 Three-dimensional reconstruction of the aortic arch, focusing on the right patent ductus arteriosus (PDA) with right pulmonary artery (PA).

Complementary imaging such as CTA and magnetic resonance angiography is recommended to fully delineate both the intracardiac and extracardiac anatomy.[1] However, as this case illustrates, with excellent echocardiographic technique, understanding of complex anatomy, and patience, one can often define the cardiac anatomy with echocardiography alone, especially in the neonatal period. However, in certain cases of DORV with malposed great arteries, defining the coronary artery anatomy may be challenging by echocardiography alone, and CTA may provide more benefit in these rare cases to help avoid invasive angiography before repair.

The repair of the underlying anatomy with bilateral patent ducti ranges from palliation with transcatheter ductal stenting to complete repair. This is the first reported case of successful bilateral ductal stenting via the femoral approach. A previously published case reported stenting of bilateral ducti; the left duct was approached from the femoral artery and the right duct via the right carotid artery, as the angle of the right ducti from the aortic arch was thought to be extremely difficult for stenting via the femoral vessels.[4] This case report demonstrates that echocardiography was vital to obtain all the information needed to diagnose and further guide the

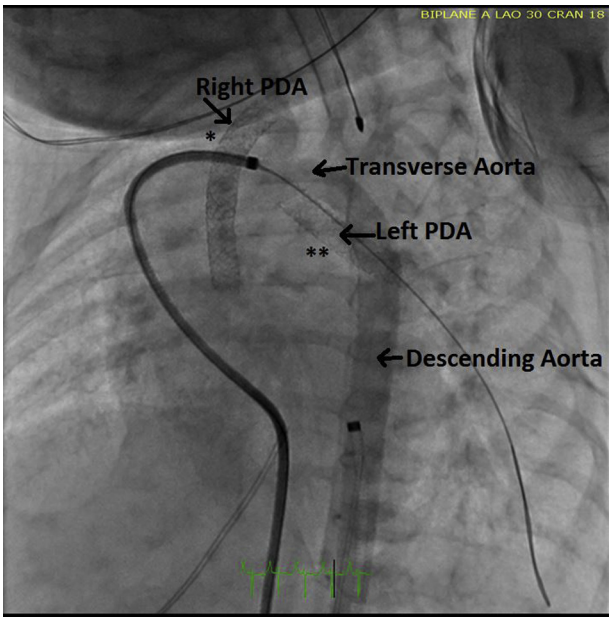

Figure 5 Angiogram showing the coronary artery stents placed in the bilateral patent ductus arteriosus (PDA): right PDA (*asterisk*) and left PDA (*double asterisk*).

interventionalist in planning the procedure. In a number of other cases, this problem was approached through a wide range of surgical interventions, including placing an AP shunt to complete repair. The approach generally elected in these complex patients is based largely on patient and institutional preference.[2,5-7]

CONCLUSION

This case underscores the wide range of anatomy seen in pediatric cardiology and the importance of excellent echocardiographic images as the initial imaging modality in many complex cases. Under certain circumstances, complementary imaging modalities, such as CTA and magnetic resonance angiography, may be helpful to confirm the anatomy when it is not clearly defined by echocardiography. Also, this case illustrates the feasibility of initial transcatheter palliation as an alternative to allow patient weight gain and improved successful repair in cases of complex congenital heart disease.

ACKNOWLEDGMENTS

We thank Jennifer Staley for edits of this report.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found online at https://doi.org/10.1016/j.case.2017.09.007.

REFERENCES

1. Moreira VW. Persistent bilateral patent ductus arteriosus and confluent pulmonary arteries—a rare congenital cardiac malformation. SOMATOM Sessions Online. Available at: https://health.siemens.com/ct_applications/somatomsessions/index.php/persistent-bilateral-patent-ductus-arteriosus-and-confluent-pulmonary-arteries-a-rare-congenital-cardiac-malformation/. Accessed October 11, 2017.
2. Peirone A. Echocardiographic evaluation, management and outcomes of bilateral arterial ducts and complex congenital heart disease: 16 years' experience. Cardiol Young 2002;12:272-7.
3. Freedom RM. Bilateral ductus arterious (or remnant): an analysis of 27 patients. Am J Cardiol 1984;53:884-91.
4. Baspinar O, Sahin DA. Bilateral ductal stenting for discontinuity of the pulmonary artery via the femoral and carotid arteries in an infant. Case Rep Cardiol 2015;2015:619653.
5. Ismail TF. Pulmonary atresia with double ductus arteriosus. J Cardiovasc Comput Tomogr 2015;9:463-5.
6. McElhinney DB. Bilateral branch pulmonary artery obstruction due to kinking at insertion sites of bilateral ductus arteriosus. Ann Thorac Surg 1997;64:537-9.
7. Ugurlucan M. Bilateral PDA in a patient with VSD and pulmonary atresia. J Card Surg 2011;26:107-10.