"Mind Your Arch"—An Unusual Cause of Systemic Hypertension in Pregnancy

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INTRODUCTION

We report a case of a pregnant patient with systemic hypertension who was eventually diagnosed with coarctation of the aorta (CoA), a type of critical congenital heart defect that typically presents with shock and heart failure in neonates and infants within the first few weeks to months of life. However, in a subset of patients, it evolves gradually and may remain undiagnosed for several years due to development of descending aortic collaterals that maintain distal aortic perfusion. These patients may present later in life with uncontrolled systemic hypertension. This case report also highlights the role of multimodality imaging in the diagnosis and management of this congenital heart defect.

CASE PRESENTATION

We report a case of a pregnant 16-year-old G1P0 female who presented at 34 weeks' gestational age with severe uncontrolled hypertension (systolic blood pressure, 171/96 mm Hg) and vaginal bleeding. The patient was diagnosed with hypertension 3 years prior to this clinical presentation, which was poorly controlled mainly due to patient noncompliance with treatment. They had been referred to cardiology multiple times but had missed appointments. Family history was significant for a younger brother with pulmonary hypertension requiring extracorporeal membrane oxygenation as an infant and a maternal cousin's child who passed away from sudden infant death syndrome.

On arrival, upper and lower extremity blood pressures were obtained and noted as follows: right arm, 138/71 mm Hg; left arm, 128/67 mm Hg; left leg, 84/63 mm Hg. The cardiac examination was pertinent for a 1/6 systolic ejection murmur with ejection click at the right upper sternal border and a 2/4 early diastolic murmur at the left lower sternal border. An electrocardiogram was performed that showed sinus tachycardia with possible left atrial enlargement. A transthoracic echocardiogram (TTE) was performed that showed concentric left ventricular hypertrophy with normal systolic function. The aortic valve was bicuspid with fusion of right and left cusps (Video 1). There was mild to moderate aortic regurgitation without any significant stenosis. The ascending aorta was normal in size with absolute dimension of 28 mm. Suprasternal views showed severe narrowing with inability to demonstrate antegrade flow across the aortic isthmus (Figures 1 and 2, Videos 2 and 3). Continuous-wave Doppler through the aortic isthmus showed continuous flow throughout diastole (Figure 3). The peak systolic velocity was 2.0 to 2.5 m/sec, and end-diastolic velocity was 0.5 m/sec. Abdominal aortic spectral Doppler profile showed significant abnormalities including blunted systolic upstroke with continuous flow in diastole suggestive of severe antegrade restriction of blood and presence of distal flow from collaterals (Figure 4).

Cardiovascular magnetic resonance imaging was attempted for cross-sectional imaging of the aortic arch (due to pregnant status); however, the patient was unable to complete the exam due to significant claustrophobia. They remained in the hospital for management of systemic hypertension with intravenous magnesium sulphate, oral nifedipine, and labetalol and also underwent a fetal echocardiogram given their congenital heart diagnosis. The fetal echocardiogram did not show any significant abnormalities. A postnatal TTE was indicated due to the maternal history. Due to difficulty managing blood pressure and gestational age of 34 weeks, a decision was made to proceed with induction of labor. Cardiac anesthesia and cardiology services were consulted for delivery planning. The patient received an arterial line for blood pressure management during labor. Labor was actively managed with oxytocin per local protocol. They underwent assisted second stage of labor with forceps-assisted vaginal delivery with arterial-line monitoring with upper extremity systolic blood pressure goal of <150 mm Hg. The infant was delivered with APGAR scores of 3, 5, and 8 at 1, 5, and 10 minutes, respectively, due to desaturations and need for positive pressure ventilation after birth. A postnatal TTE was obtained on the infant that showed partial fusion of right and left aortic cusps with normal aortic valve function. The infant was discharged home after a 3-week neonatal intensive care unit stay due to the need for respiratory support, prematurity, and feeding issues. The patient required postpartum magnesium sulphate infusions followed by transition to oral antihypertensive medications.

Following delivery, a cardiac computed tomography scan with contrast was obtained for preprocedural planning (Figure 5, Video 4). Cardiac computed tomography showed severe CoA with presence of multiple descending aortic collaterals. The patient was taken to the cardiac catheterization lab with plans to intervene on the aortic arch. In the cath lab, functional atresia of the aortic arch with no flow of contrast across the region of stenosis was noted; however, they were able to pass a wire across the coarctation site. The patient then underwent percutaneous balloon dilation with stent placement with a 14 mm × 3.4 cm premounted covered stent, resulting in a reduction of precoarctation gradient from 58 mm Hg to 10 mm Hg. Postprocedural TTE showed mild flow acceleration across the aortic

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VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, parasternal short-axis view at the level of the aortic valve, demonstrates a bicuspid aortic valve with fusion of right and left aortic cusps.

Video 2: Two-dimensional TTE, suprasternal view, demonstrates severe luminal narrowing of aortic isthmus distal to left subclavian artery origin.

Video 3: Two-dimensional TTE, suprasternal long-axis view with color-flow Doppler, demonstrates aliasing across aortic isthmus and lack of antegrade flow.

Video 4: Computed tomography scan with three-dimensional volume-rendered reconstruction showing the aortic arch. There is severe CoA at the region of the aortic isthmus, distal to the left subclavian artery origin, along with multiple descending aortic collaterals.

Video 5: Poststent two-dimensional TTE, suprasternal long-axis view, demonstrates color Doppler flow across the aortic arch. Peak aortic arch Doppler velocity was 2.5 m/sec with mild aliasing across the aortic isthmus.

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isthmus with peak Doppler velocity of 2.5 m/sec and normal abdominal aortic Doppler profile (Figure 6, Video 5). The patient was discharged home on labetalol 200 mg twice a day for residual systemic hypertension and continues to follow up at regular intervals with the cardiology service.

DISCUSSION

Systemic hypertension in pregnancy can have a broad differential including chronic hypertension, gestational hypertension, and preeclampsia superimposed on chronic hypertension.[1,2] In this case report, we highlight how CoA can be a rare differential of systemic hypertension in a pregnant patient. We also highlight the role of multimodality imaging in this lesion for diagnosis and management.[2-4]

Coarctation of the aorta is a narrowing of the descending aorta and is diagnosed in 7% of patients with congenital heart disease (CHD).[5] This type of heart defect may present as a spectrum ranging from discrete narrowing of the aortic isthmus to long-segment hypoplasia often extending to the transverse aortic arch. Bicuspid aortic valve is the most common associated defect.[5,6] In neonates and infants, it may present as cardiogenic shock following patent ductus arteriosus closure.[6] However, in a subset of patients, it may evolve gradually and present later in life and involve the formation of abnormal collateral vessels between the left subclavian arteries, axillary arteries, and intercostal arteries with retrograde flow into the descending aorta, distal to the site of coarctation.[7] In our patient, this finding was suspected due to the abnormal appearance of the pulsed-wave spectral Doppler profile of the aortic arch and the abdominal aorta, due to antegrade obstruction and abnormal distal perfusion almost exclusively from collateral flow. Older patients with CoA typically present with systemic hypertension (specifically upper extremity hypertension), left ventricular failure, and stroke.[8] This patient's diagnosis was likely delayed due to collateral formation, which maintained distal perfusion but was unmasked during the physiologic alterations that occur in pregnancy.[9] With a World Health Organization pregnancy risk status IV due to risk of cerebral hypertension and fetal hypoperfusion in the setting of underlying unrepaired CoA, our patient required close monitoring of blood pressure with individualized blood pressure goals.[9]

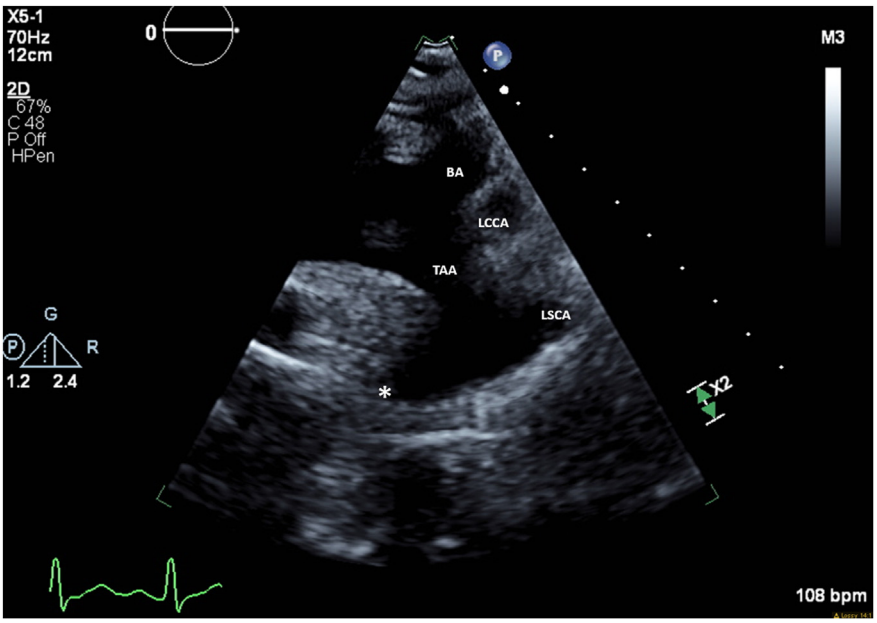

Figure 1 Two-dimensional TTE, suprasternal long-axis view of the aortic arch, demonstrates severe luminal narrowing (*) in the aortic isthmus distal to the origin of the left subclavian artery. *BA*, Brachiocephalic artery; *LCCA*, left common carotid artery; *LSCA*, left subclavian artery; *TAA*, transverse aortic arch.

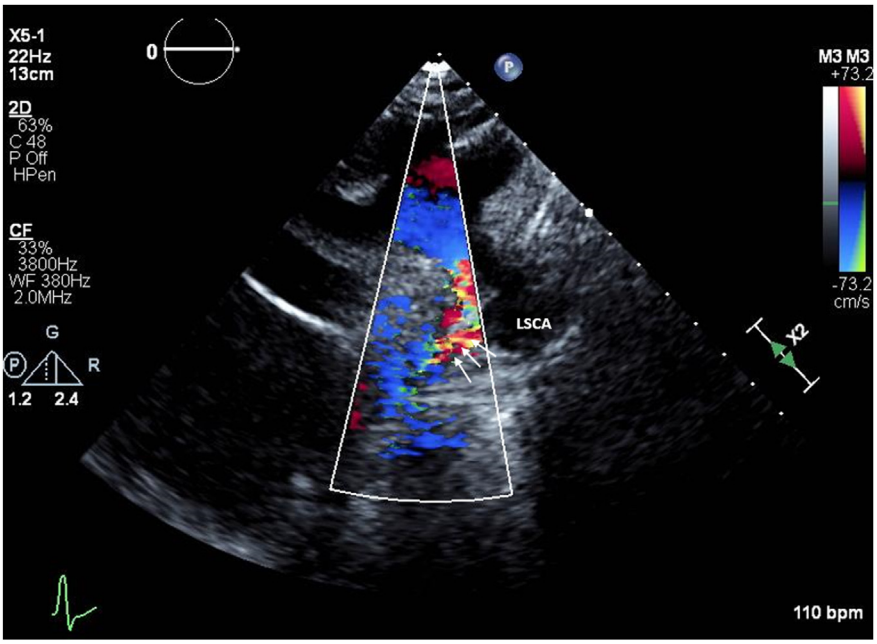

Figure 2 Two-dimensional TTE, suprasternal long-axis view with color-flow Doppler, demonstrates aliasing (*arrows*) and lack of antegrade flow across aortic isthmus. *LSCA*, Left subclavian artery.

Coarcation of the aorta is also considered a vascular disease with the highest risk of vascular complications in CHD.[10] There is a 3.2% risk of stroke or transient ischemic attack and a 5.1% risk of myocardial infarction in these patients.[8,11] Even in the absence of anatomical narrowing, the aorta in these patients can have abnormal compliance and vascular stiffness, which predisposes them to systemic hypertension.[12] Despite successful placement of the covered stent without any residual gradient, our patient continued to have significant systemic hypertension that needed aggressive treatment with antihypertensives. Additionally, 10% of patients can experience cerebral aneurysms, termed Berry aneurysms, and require neurovascular imaging for screening.[13]

This case report also highlights the role of multimodality imaging in this congenital lesion.[14,15] Although echocardiography was the initial imaging modality and showed significant abnormalities that pointed toward the diagnosis, cross-sectional imaging with three-dimensional reconstruction demonstrated the additional features such as near interruption of the aortic arch, along with distribution of the

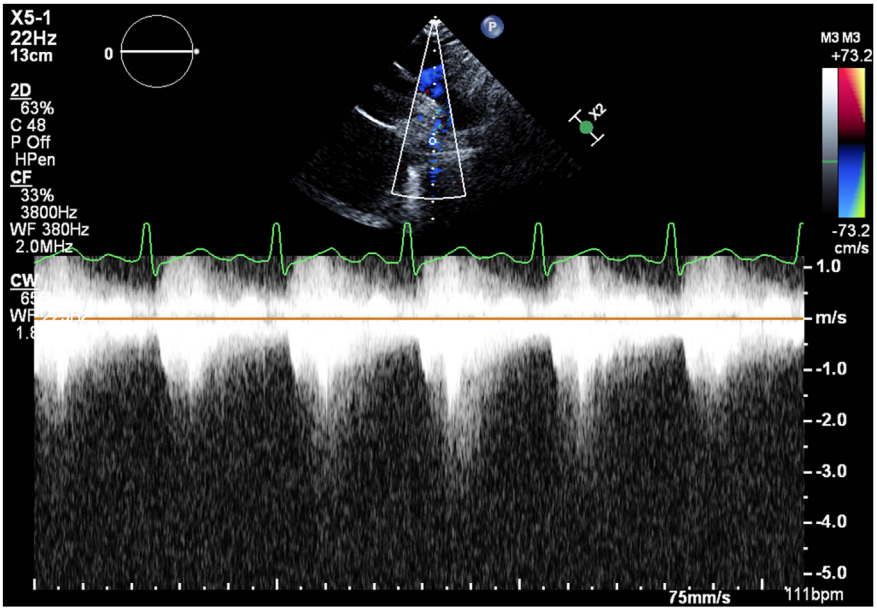

Figure 3 Two-dimensional TTE, continuous-wave Doppler across descending aorta, demonstrates abnormal forward flow in diastole and a peak Doppler velocity of 2.5 m/sec.

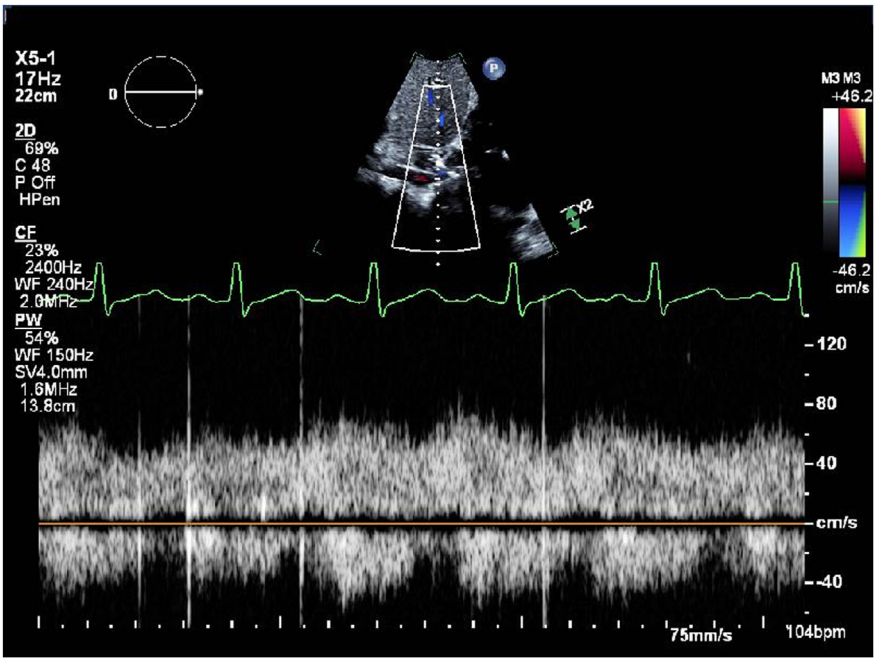

Figure 4 Two-dimensional TTE, abdominal aortic pulsed-waved spectral Doppler profile, demonstrates significantly abnormal Doppler pattern with blunted profile and forward flow in diastole.

descending aortic collaterals.[14] It also provided high-resolution images of the arch anatomy so accurate measurements could be made for preprocedural planning in terms of stent size and placement. Cardiovascular magnetic resonance imaging provides additional data such as collateral flow burden, but its use was limited in this case due to patient claustrophobia.[14]

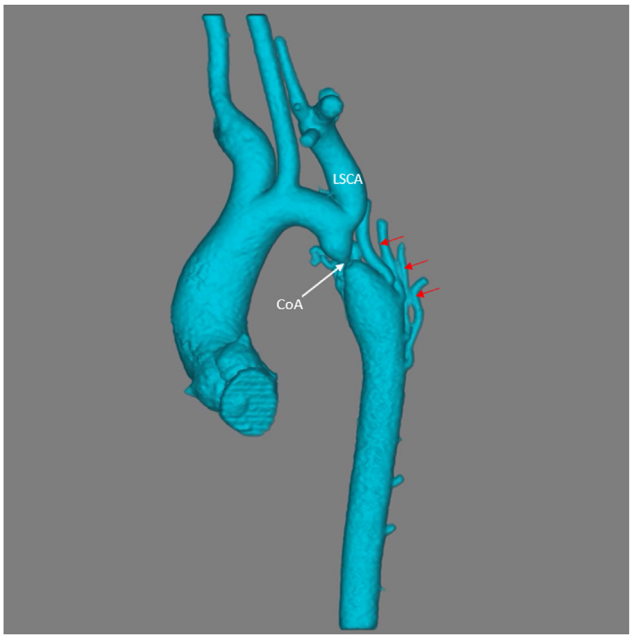

Figure 5 Computed tomography scan with three-dimensional volume-rendered reconstruction showing the aortic arch. There is severe CoA at the region of the aortic isthmus, distal to the left subclavian artery (LSCA) origin, along with multiple descending aortic collaterals (*red arrows*).

CONCLUSION

We report a case of aortic coarctation that was diagnosed in a young pregnant patient. Aortic coarctation is a type of CHD that should be ruled out in a young patient with severe systemic hypertension. Presentation in older patients may be delayed by the presence of collateral vessels, which bypass the coarctation segment to supply the descending aorta. This case also highlights the role of multimodality imaging in the diagnosis and management of this congenital heart defect.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

CONSENT STATEMENT

The authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

FUNDING STATEMENT

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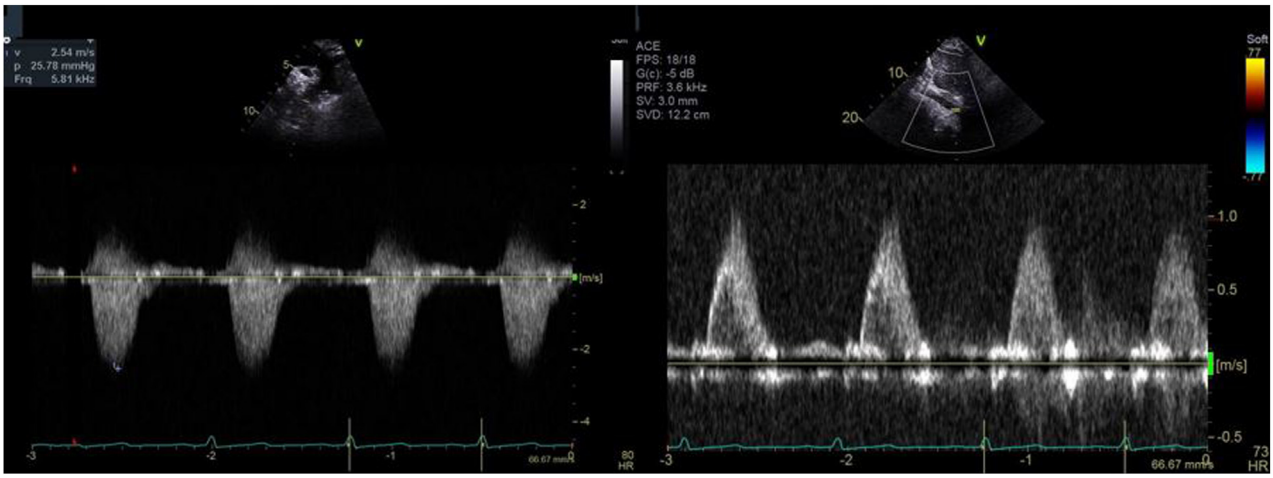

Figure 6 Two-dimensional TTE, aortic arch continuous-wave Doppler, demonstrates peak aortic arch Doppler velocity of 2.5 m/sec (*left*). Two-dimensional TTE, abdominal aortic pulsed-wave Doppler, demonstrates normal pulsatile flow with absence of forward flow in diastole after intervention (*right*).

DISCLOSURE STATEMENT

The authors report no conflicts of interest.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2023.12.016.

REFERENCES

1. Kametas NA, Nzelu D, Nicolaides KH. Chronic hypertension and superimposed preeclampsia: screening and diagnosis. Am J Obstet Gynecol 2022;226:S1182-95.
2. Ramlakhan KP, Malhame I, Marelli A, Rutz T, Goland S, Johnson MR, et al. Hypertensive disorders of pregnancy in women with structural heart disease: data from the ESC EORP Registry of pregnancy and cardiac disease (ROPAC). Eur Heart J 2022;43:3749-61.
3. Wei L, Hu S, Gong X, Ahemaiti Y, Zhao T. Diagnosis of covert coarctation of the aorta in adolescents. Front Pediatr 2023;11:1101607.
4. Lip GYH, Singh SP, Beevers DG. Aortic coarctation diagnosed after hypertension in pregnancy. Am J Obstet Gynecol 1998;179:814-5.
5. Talner CN. Report of the New England regional infant cardiac program. Pediatrics 1980;65:376e461.
6. O'Brien P, Marshall AC. Coarctation of the aorta. Circulation 2015;131:e363-5.
7. Nance JW, Ringel RE, Fishman EK. Coarctation of the aorta in adolescents and adults: a review of clinical features and CT imaging. J Cardiovasc Comput Tomogr 2016;10:1-12.
8. Tanous D, Benson LN, Horlick EM. Coarctation of the aorta: evaluation and management. Curr Opin Cardiol 2009;24:509-15.
9. Lindley KJ, Bairey Merz CN, Asgar AW, et al. Management of women with congenital or inherited cardiovascular disease from pre-conception through pregnancy and postpartum: JACC focus seminar 2/5. J Am Coll Cardiol 2021;77:1778-98.
10. Roeleveld PP, Zwijsen EG. Treatment strategies for paradoxical hypertension following surgical correction of coarctation of the aorta in children. World J Pediatr Congenit Heart Surg 2017;8:321-31.
11. Verheugt CL, Uiterwaal CS, Grobbee DE, Mulder BJ. Long-term prognosis of congenital heart defects: a systematic review. Int J Cardiol 2008;131:25-32.
12. Chen SSM, Donald AE, Storry C, Halcox JP, Bonhoeffer P, Deanfield JE. Impact of aortic stenting on peripheral vascular function and daytime systolic blood pressure in adult coarctation. Heart 2008;94:919-24.
13. 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: executive summary: a report of the American college of cardiology/American heart association task force on clinical practice guidelines. J Am Coll Cardiol 2019;73:1494-563. Erratum in: J Am Coll Cardiol. 2019 May 14;73(18):2361.
14. Fratz S, Chung T, Greil GF, Samyn MM, Taylor AM, Buechel ERV, et al. Guidelines and protocols for cardiovascular magnetic resonance in children and adults with congenital heart disease: SCMR expert consensus group on congenital heart disease. J Cardiovasc Magn Reson 2013;15:51.
15. Writing Group, Sachdeva R, Valente AM, Armstrong AK, Cook SC, Han KB, et al. ACC/AHA/ASE/HRS/ISACHD/SCAI/SCCT/SCMR/SOPE 2020 appropriate use criteria for multimodality imaging during the follow-up care of patients with congenital heart disease: a report of the American College of Cardiology Solution Set Oversight Committee and Appropriate Use Criteria Task Force, American Heart Association, American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, Society for Cardiovascular Magnetic Resonance, and Society of Pediatric Echocardiography. J Am Soc Echocardiogr 2020;33:e1-48.