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Prenatal Detection of Anomalous Right Coronary Artery with an Interarterial Course



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INTRODUCTION

Congenital coronary artery anomalies affect 1% of the general population.¹ These can be classified as abnormalities of origin and course, including such arrangements as an anomalous origin from the opposite sinus and single coronary artery.^{1,2} Although most variants of anomalous aortic origin of the coronary artery (AAOCA) are benign, an important subset may pose a risk for sudden cardiac death, in particular certain variations of those with an interarterial course.¹⁻⁴ The precise prevalence, natural history, and relative risks of many of these lesions are incompletely understood, and the clinical course ranges from entirely benign to fatal. Thus, early identification of patients with these conditions may be clinically useful. We report the first documented case of an anomalous aortic origin of the right coronary artery (AAORCA) from the left main coronary artery with an interarterial course demonstrated by fetal echocardiography (FE) and confirmed by postnatal transthoracic echocardiography (TTE).

CASE PRESENTATION

A 29-year-old woman (gravida 1, para 0) presented to the echocardiography laboratory for evaluation because of a family history of congenital heart disease in a second-degree relative (coarctation requiring subclavian flap repair in infancy). FE was performed at 22 weeks and 4 days' gestation using a 9-MHz curvilinear probe (Vivid E95; GE Healthcare, Little Chalfont, United Kingdom). FE revealed a single intrauterine pregnancy with vertex lie. Fetal size was consistent with established dates. Echocardiography demonstrated a widely patent left aortic arch in the setting of a widely patent ductus arteriosus, normal flow across all four cardiac valves, and a normal-appearing left ventricle and right ventricle. Flow within the right coronary artery was readily identified, and the vessel appeared to arise leftward of the intercoronary commissure, indicating possible AAORCA (Figure 1, Video 1). The infant was born without incident at 38 weeks and 2 days and had planned follow-up in the cardiology clinic at our institution at 2 months of age. Physical examination was normal. Nonsedated TTE was performed and confirmed the diagnosis of an anomalous right coronary artery. The patient had a single ostium

coronary artery originating from the left aortic sinus of Valsalva. The right coronary artery emerged from the common left main coronary just beyond the origin (Figure 2, Video 2). There was an interarterial course, and the vessel appeared to be of consistent caliber throughout. The circumflex and left anterior descending branches had a normal takeoff from the left main coronary artery with normal proximal courses (Figure 3, Video 3). Additional findings included a possible small coronary to main pulmonary artery fistula, normal trileaflet aortic valve, and normal left and right ventricular size and systolic function.

The family was counseled on the diagnosis, prognosis, and treatment options for this condition. No restrictions, precautions, or treatment were recommended for the present time, and a follow-up visit was requested at 3 years of age with electrocardiography and echocardiography. There was a future recommendation for computed tomography imaging and functional testing, including exercise stress testing, at an older age.

DISCUSSION

We present a case of anomalous right coronary artery from the left main coronary artery with an interarterial course discovered incidentally on FE at 22 weeks, 4 days, and confirmed by postnatal TTE. Although evaluation of fetal coronary artery anatomy and flow is not expected during routine FE,⁵ such demonstrations have been described previously. In 2002, Baschat and Gembruch⁶ described coronary artery blood flow velocities during various fetal conditions. They were able to visualize coronary artery blood flow in normal fetuses at a median gestational age of 33 weeks, 6 days. In growth-restricted fetuses, they were able to visualize coronary blood flow at a median age of 28 weeks, 2 days; the detection of coronary blood flow in these fetuses was associated with deteriorating cardiovascular status, often leading to fetal demise. Fetal anemia and ductus arteriosus constriction also allowed the visualization of coronary blood flow, as did the presence of coronary artery fistulae. The coronary fistula in this case was not likely large enough to have affected overall coronary artery size and thus visualization of the coronary arteries. In 2017, Kaji *et al.*⁷ presented the first reported prenatal assessment of coronary artery anatomy in transposition of the great arteries. They imaged coronary artery blood flow on three fetuses with TGA between 28 and 37 weeks' gestation. They found that the prenatal assessment of coronary artery anatomy using color Doppler in transposition of the great arteries was feasible, but the diagnostic accuracy was limited.

In our experience, to locate the coronary artery origins on FE, the fetal lie should be optimal to obtain a parasternal short-axis image with a focus on the aortic valve cusps and sinuses. Meticulous attention should be paid to optimizing spatial and temporal resolution. Using a high-frequency transducer, the depth and sector width should be decreased, followed by zoom (high-definition zoom if available) to increase the image resolution. The color flow settings should be optimized for low-flow imaging. Color flow should be visualized from the

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Keywords: Abnormal coronary artery, Anomalous coronary artery, Interarterial, Fetus, Fetal echocardiography

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VIDEO HIGHLIGHTS

Video 1: FE, short-axis view with two-dimensional and color-compare images, showing the right coronary artery arising from leftward beyond the right-left coronary commissure and coursing between the aorta and pulmonary artery.

Video 2: Transthoracic parasternal short-axis color flow Doppler image showing the right coronary artery origin from the left coronary artery/sinus. Color flow in the wall of the right ventricular outflow tract and into the pulmonary artery likely represent a coronary-to-pulmonary artery fistula discovered incidentally.

Video 3: Transthoracic parasternal short-axis color flow Doppler image further detailing the abnormal origin of the right coronary artery as well as the normal course of the left anterior descending and circumflex coronary arteries.

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

aorta into the proximal coronary artery during diastole to confidently diagnose the vessel origin.

Imaging goals for postnatal studies should include further characterizing the anatomic features of AAOCA that have a bearing on management. Criteria for identifying an intramural course have been described for TTE,^{8,9} computed tomographic angiography,¹⁰ and cardiac magnetic resonance angiography.¹¹

Recommendations for management of AAOCA remain an evolving topic. In the case of AAOCA involving the left coronary artery, the recommendations are clearer given the higher risk for sudden cardiac death, particularly in the case of an interarterial or intramural course.¹² However, because the risk for sudden cardiac death in patients with AAORCA is lower, management is more controversial, particularly in the asymptomatic population. For asymptomatic patients with AAORCA, a recent American Heart Association/American College of Cardiology scientific statement

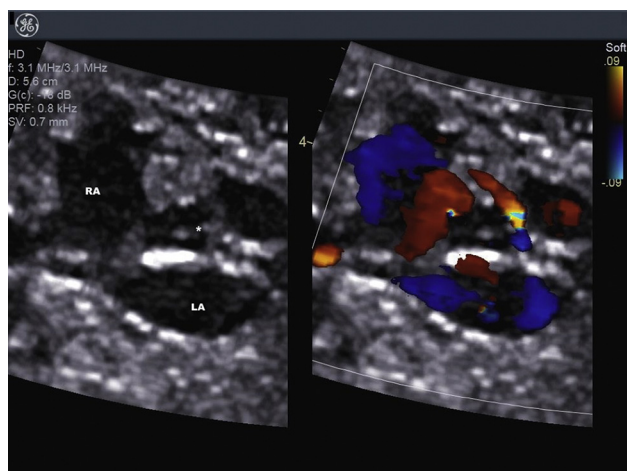


Figure 1 FE, short-axis view with color Doppler comparison, showing color flow in the right and left coronary arteries. The flow into the right coronary artery originates from leftward beyond the right-left coronary commissure (asterisk).

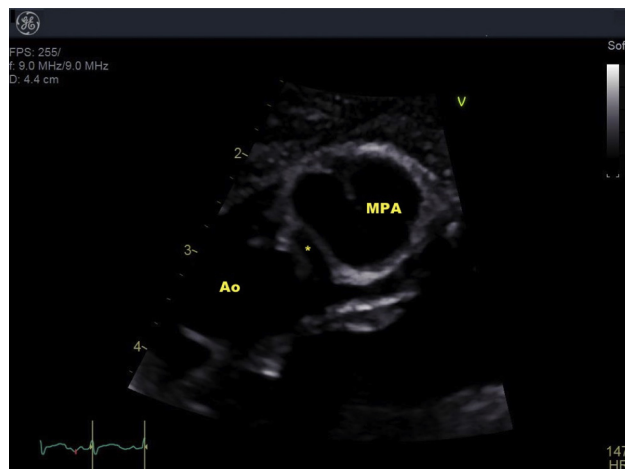


Figure 2 Transthoracic short-axis two-dimensional image showing the right coronary artery (asterisk) originating from the base of the left main coronary artery with an interarterial course.

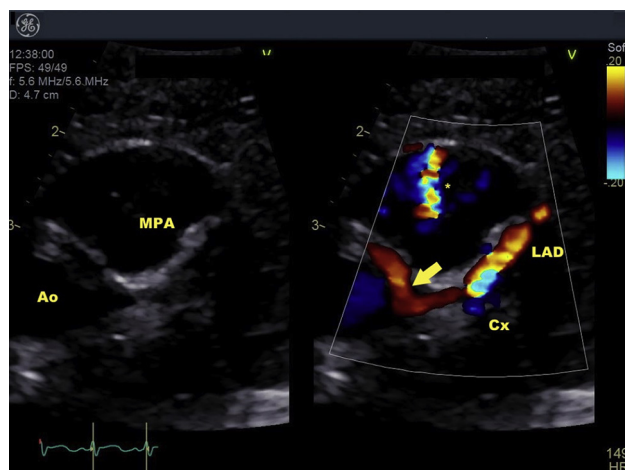


Figure 3 Transthoracic short-axis color Doppler comparison image, showing the right coronary artery originating from the base of the left main coronary artery (arrow). The left main coronary artery branches normally into the left anterior descending (LAD) and circumflex (Cx) coronary arteries. Also shown is a turbulent jet of flow into the pulmonary artery (asterisk), likely representing a coronary-to-pulmonary fistula.

offers the potential to return to competitive sports after a complete evaluation.¹³ However, if a patient with AAORCA is symptomatic or has inducible ischemia, activity restriction and surgery are recommended.¹² Early identification, diagnosis, and risk stratification¹⁴ may benefit these patients, as the understanding of the prognosis improves over time.

CONCLUSION

We present a case of anomalous right coronary artery with an interarterial course identified on FE and confirmed on postnatal TTE. Imaging of the coronary artery origins and proximal course is feasible with FE, allowing the diagnosis of rare and subtle abnormalities.

Ongoing studies are needed to evaluate the diagnostic accuracy of this imaging modality. With advancements in technology, prenatal recognition of these rare congenital coronary artery abnormalities will occur with more frequency. Collaborations such as the Congenital Heart Surgeons' Society Registry of AAOCA,¹³ the registry of proximal anomalous connections of coronary arteries of the French Society of Cardiology, and the Sudden Death in the Young Registry,¹⁵ will further reduce knowledge gaps, strengthen consensus guidelines, and aid in counseling patients and their families from the fetus through adulthood.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.10.001>.

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