

A Hole and One



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INTRODUCTION

Atrial septal defects (ASDs), after bicuspid aortic valve, are the most common congenital heart defect (CHD) in the adult population. Clinical presentations differ greatly as most are asymptomatic, but others can develop significant pulmonary hypertension and subsequent right heart failure.¹ They may be associated with other CHDs such as ventricular septal defects or anomalous pulmonary venous drainage. Anomalous aortic origins of coronary arteries are relatively rare congenital defects, and although they can be associated with some more complex CHDs such as transposition of great arteries or tetralogy of Fallot, they are often seen in isolation. We present a case of a patient with a secundum ASD who was incidentally found to have left main artery origin from the right sinus of Valsalva.

CASE PRESENTATION

An asymptomatic 65-year-old woman with no known previous history of cardiac or medical illness was referred for cardiology consultation and an echocardiographic examination for an incidental finding of cardiomegaly identified on chest radiograph.

Physical examination revealed a left upper extremity polydactyly with an extra thumb and no other dysmorphic features. There was no digital clubbing or cyanosis. Heart rate was 72 beats per minute and regular with blood pressure of 120/80 mm Hg in both arms. Jugular venous pulse was not distended, and there was no hepatojugular reflux. Precordial examination revealed a normal apical impulse and presence of a right ventricular lift indicating a possible right ventricular enlargement. Auscultation revealed normal first heart sound and a fixed-split second heart sound. There was a grade II/VI systolic ejection murmur best heard at the left upper sternal border. No extra heart sounds were heard. Peripheral pulses were all normal in volume, and there was no peripheral edema.

A 12-lead electrocardiogram (Figure 1) was significant for an incomplete right bundle branch block and a rightward axis. Transthoracic echocardiography (Figure 2, Videos 1 and 2) showed enlarged right-sided chambers (right atrial volume index of 30 mL/

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VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiogram from the right ventricle–focused view showing dilated right heart chambers.

Video 2: Transthoracic echocardiogram from the modified apical four-chamber view showing left-to-right shunting across the atrial septum.

Video 3: Transesophageal echocardiogram from the mid-esophageal window with and without color Doppler showing presence of a secundum ASD with evidence of left-to-right shunting between the left and right atria.

Video 4: Coronary angiogram from the left anterior oblique plane showing the origin of the LAD artery from the right.

Video 5: CT coronary angiogram with volume rendered display showing the LAD and the right coronary artery arising from the same sinus.

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

m²) with a preserved right ventricular systolic function (tricuspid annular plane systolic excursion of 1.9 cm and fractional area change of 42%) and a right ventricular systolic pressure within normal limits at 28 mm Hg, indicating absence of significant pulmonary hypertension. This study also showed the presence of left-to-right shunting across a moderate-size secundum ASD. Subsequent transesophageal echocardiogram (Figure 3, Video 3) confirmed the presence of a secundum ASD measuring 18 mm in diameter with an aortic rim of 10 mm, posterior rim of 15 mm, superior vena cava rim of 20 mm, and inferior vena cava rim of 22 mm. The right heart chambers were dilated with no other abnormalities seen. Based on the echocardiographic findings, the ASD was deemed suitable for percutaneous device closure.² Given the patient's age, a diagnostic coronary angiography was performed to assess for hemodynamically significant coronary artery disease (Figure 4, Video 4). Surprisingly, this showed a single coronary artery arising from the right sinus with no obstructive lesions. To better assess the course of the anomalous left main coronary artery (ALMCA) and to rule out a malignant course, a computed tomography (CT) angiogram with three-dimensional reconstruction was performed (Figures 5 and 6, Video 5). The images revealed a right dominant system with a single coronary artery originating from the right coronary sinus with a nonmalignant course of the left anterior descending (LAD) artery.

As there was no evidence of significant coronary artery disease, with a benign course of the single anomalous left coronary artery, the patient was referred for and underwent a successful transcatheter ASD closure with the use of a 22 mm Amplatzer septal occluder. On follow-up, the patient remains asymptomatic 12 months postclosure, with echocardiogram demonstrating a well-seated interatrial occlusion device with no significant residual shunt and the return of the dilated right-sided chambers to normal range.

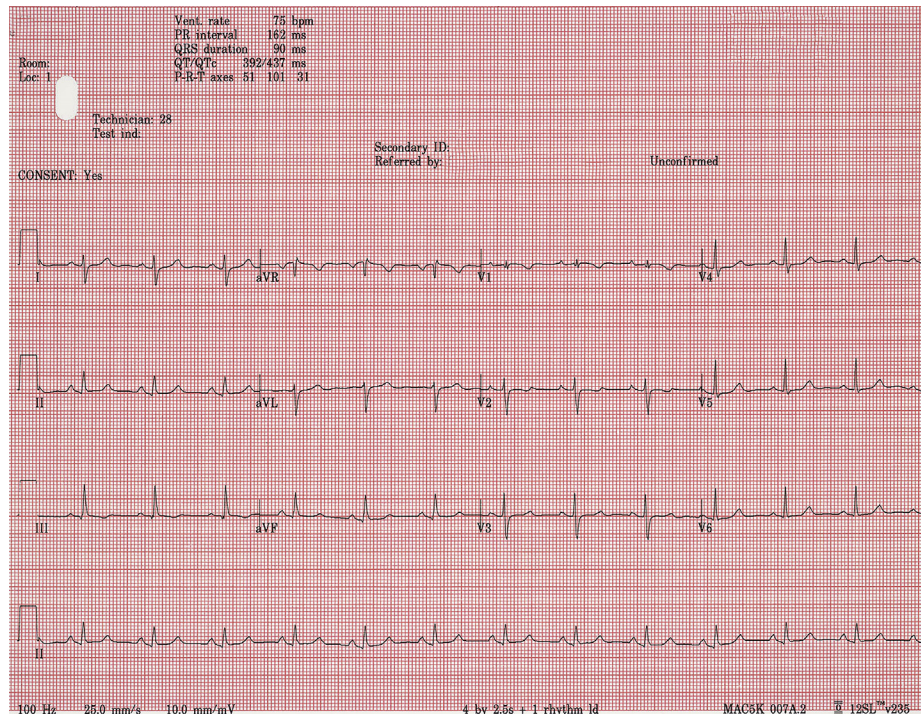


Figure 1 Twelve-lead electrocardiogram showing sinus rhythm with an incomplete right bundle branch block and a rightward axis.

DISCUSSION

Atrial septal defects are the second most common CHD after bicuspid aortic valve, seen in the adult population with an incidence of one in 1,500 live births, with the majority being of the ostium secundum type.³ Most patients are asymptomatic, but some develop pulmonary

hypertension with increased left-to-right shunt and eventually right-sided heart failure due to the resulting volume overload.⁴ Other potential complications of untreated ASDs include paradoxical embolization, atrial arrhythmias due to atrial enlargement, and progressive pulmonary vascular disease, which can lead to shunt reversal and Eisenmenger physiology. The aforementioned complications are associated with

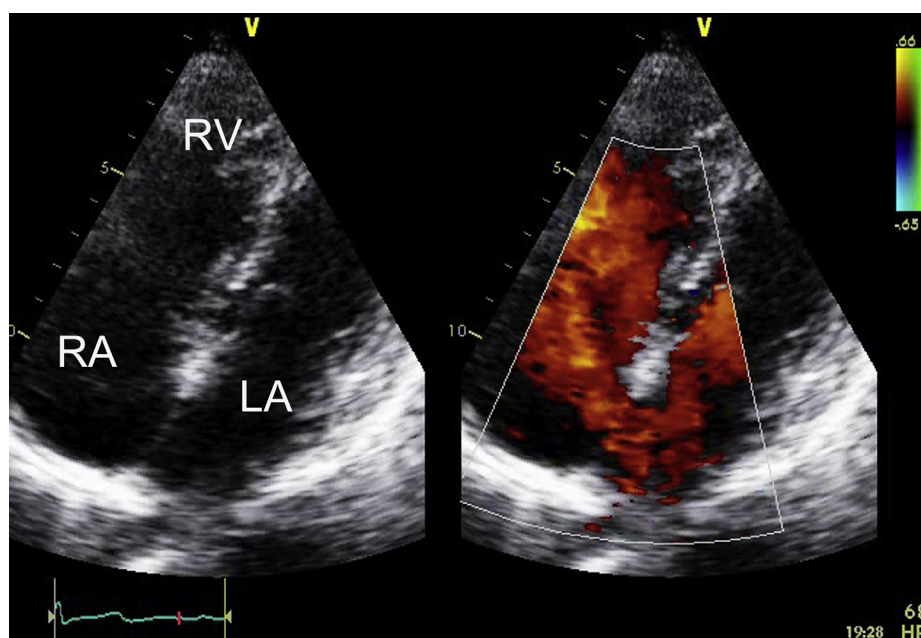


Figure 2 Trans thoracic echocardiogram from a modified apical window showing dilated right atrium (RA) and right ventricle (RV) and evidence of left-to-right shunting between the left atrium (LA) and the RA.

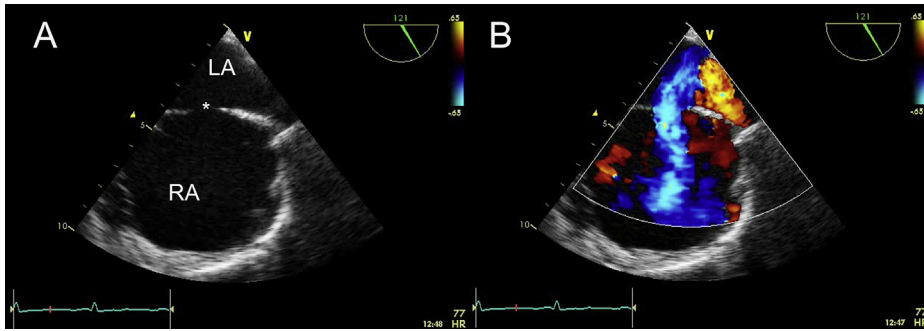


Figure 3 Transesophageal echocardiogram from the midesophageal window without (A) and with (B) color Doppler showing presence of a secundum ASD (*) with evidence of left-to-right shunting between the left atrium (LA) and the right atrium (RA).

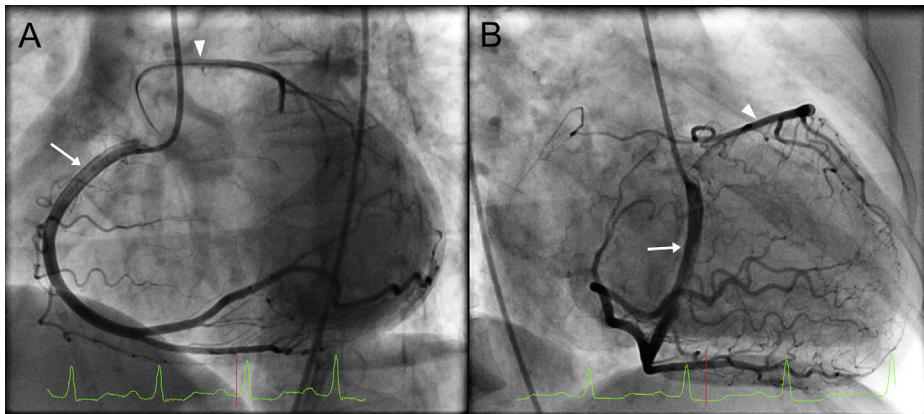


Figure 4 Coronary angiogram with the injection of the right ostium from left anterior oblique (A) and right anterior oblique (B) planes showing the LAD artery (arrowhead) and the right coronary artery (arrow) originating from the same sinus.

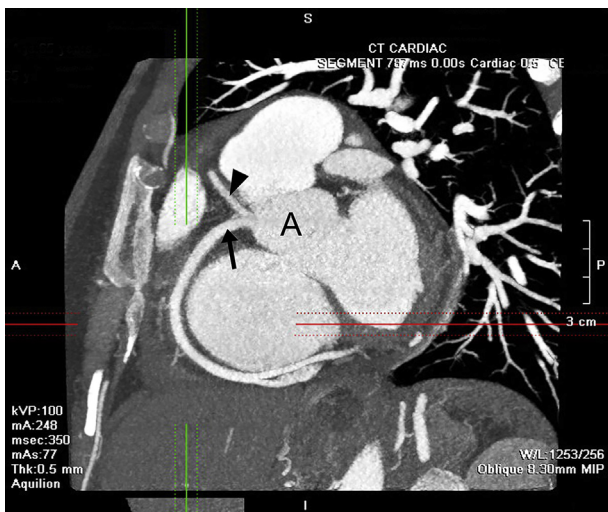


Figure 5 CT coronary angiogram showing the LAD artery (arrowhead) and the right coronary artery (arrow) arising from the aorta (A).

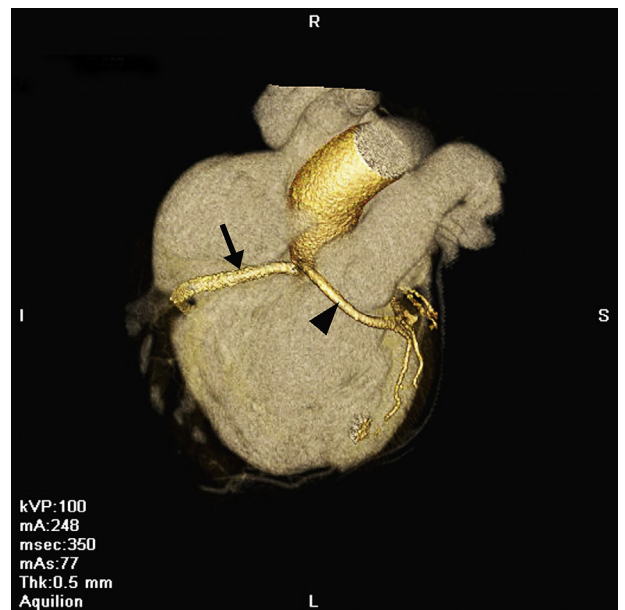


Figure 6 CT coronary angiogram with volume rendered display showing the LAD (arrowhead) and the right coronary artery (arrow) arising from the same sinus.

significant morbidity and mortality.⁴ Current guidelines recommend closure of ASDs when symptoms are present and with evidence of right heart volume overload even in the absence of symptoms.⁵

On the other hand, anomalous origin of the coronary artery from the opposite sinus is rare by comparison and occurs in approximately 1% of live births, with the ALMCA variant representing 0.15%, while right coronary artery origin from the left sinus of Valsalva has an incidence of 0.92%.⁶ The presence of a single coronary artery is even less common, with an incidence of 0.12%.⁷ The clinical presentation differs greatly as the vast majority of cases are asymptomatic, but those whose variant includes the left coronary artery coursing in between the pulmonary artery and aorta (preaortic or posterior course) have a higher incidence of sudden cardiac death due to coronary compression as the aorta and pulmonary artery enlarge during strenuous physical activity.⁸ Surgery is usually indicated only in the preaortic course due to the increased risk. The combination of the two congenital lesions of ASD and a single coronary artery is rare, with only a single reported case of a 23-year-old man with exertional dyspnea who was found to have both a secundum ASD and ALMCA, which, interestingly, ran between the aorta and pulmonary artery (preaortic), necessitating surgery for both the ASD and coronary artery anomaly.⁹ In the case we are presenting, given the lower risk of sudden cardiac death associated with patient's coronary artery anomaly, we elected to proceed with device closure of the ASD and to not intervene on the anomalous coronary artery.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2021.08.003>.

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