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A Five Chambered Heart

George Hanna

HCA Healthcare, George.Hanna@hcahealthcare.com

Javad Savoj

HCA Healthcare, javad.savoj@hcahealthcare.com

Syed Iftikhar

HCA Healthcare, Syed.Iftikhar@hcahealthcare.com

Scott Kubomoto

HCA Healthcare, scott.kubomoto@hcahealthcare.com

Patrick Hu

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A five chambered heart

George Hanna, Javad Savoj, Syed Iftikhar, Scott Kubomoto and Patrick Hu
Internal Medicine Residency – Riverside Community Hospital / UC Riverside School of Medicine

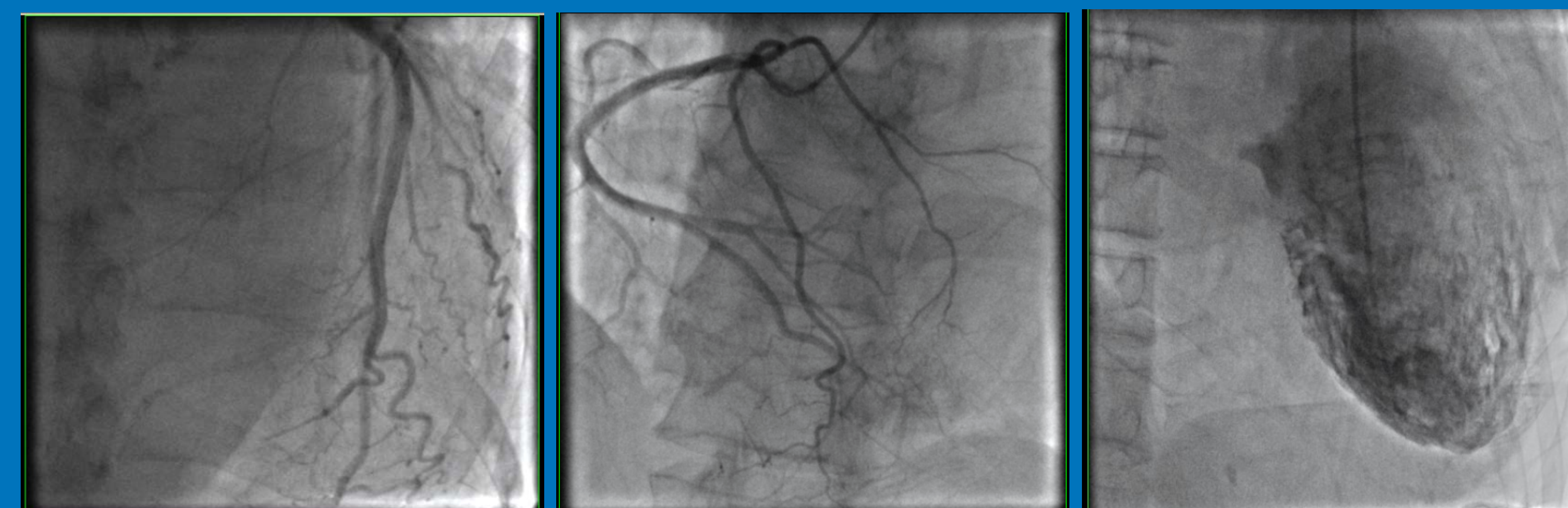
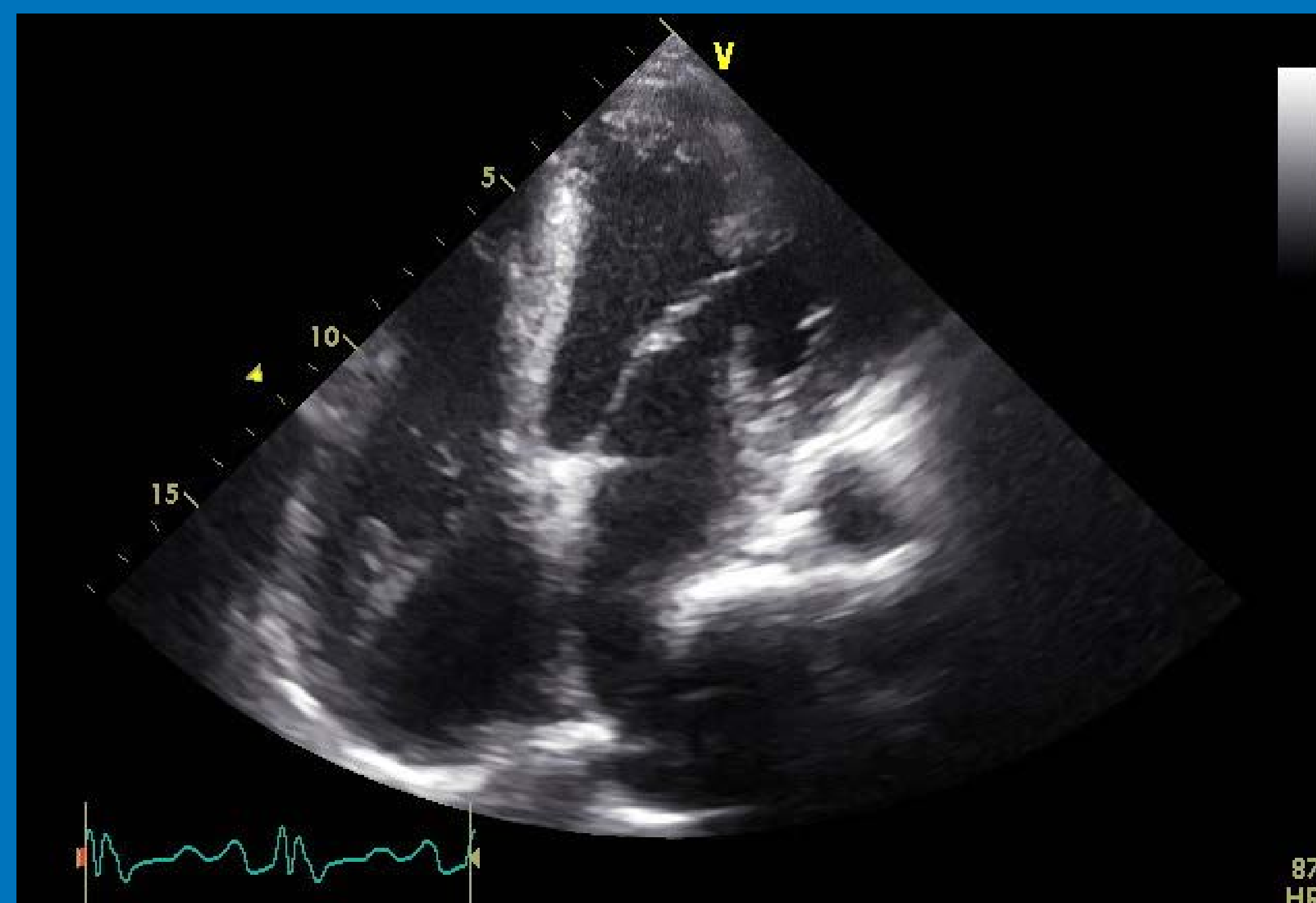
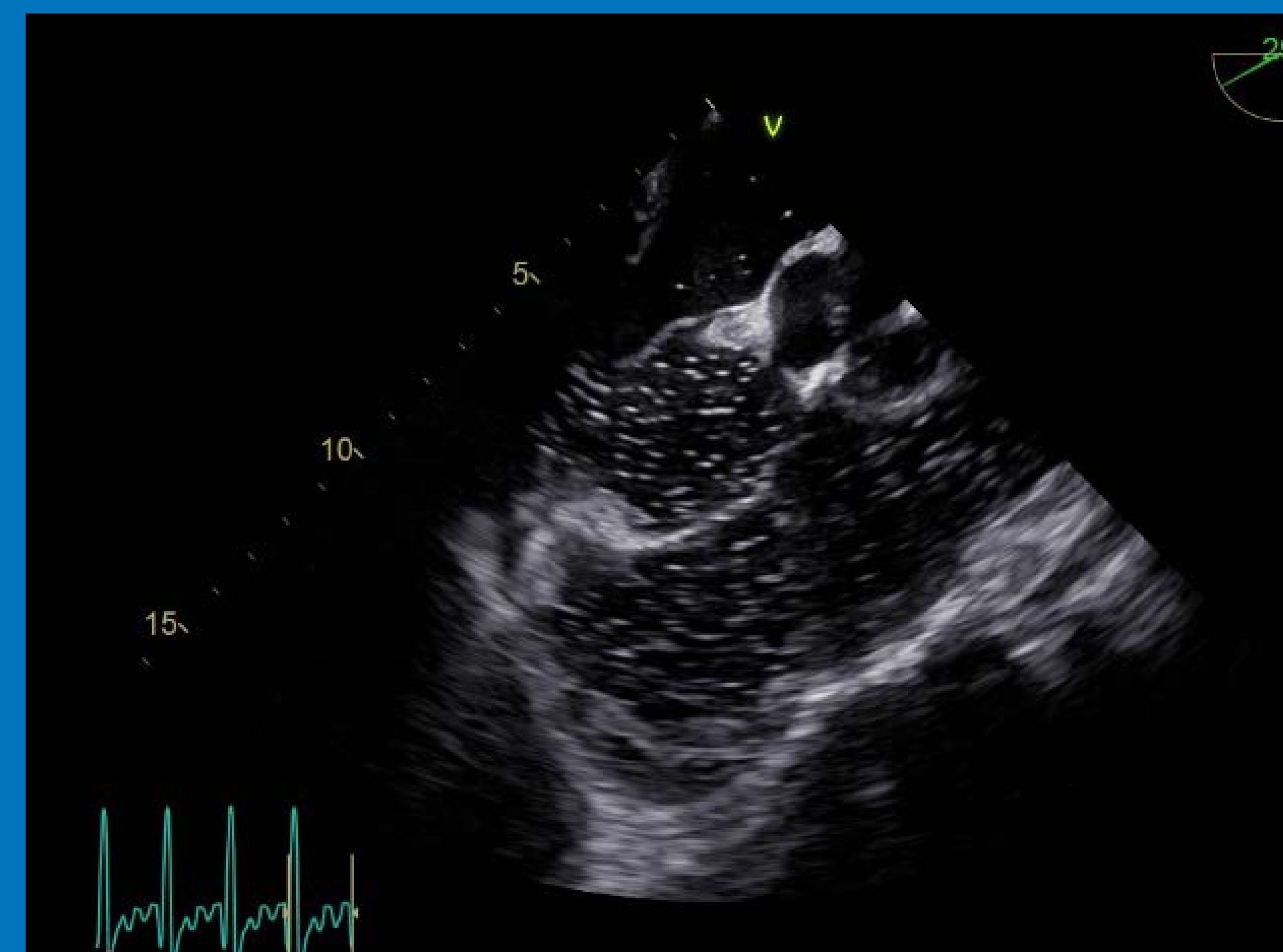
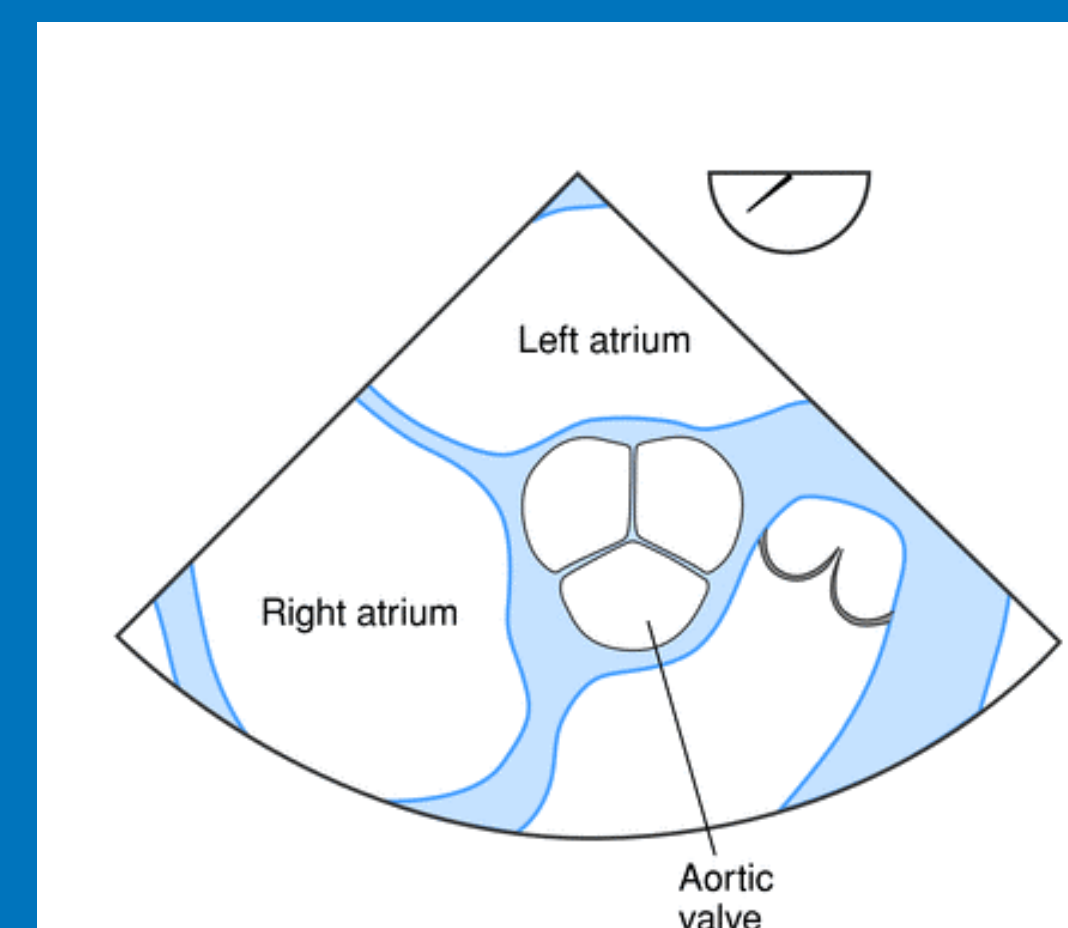
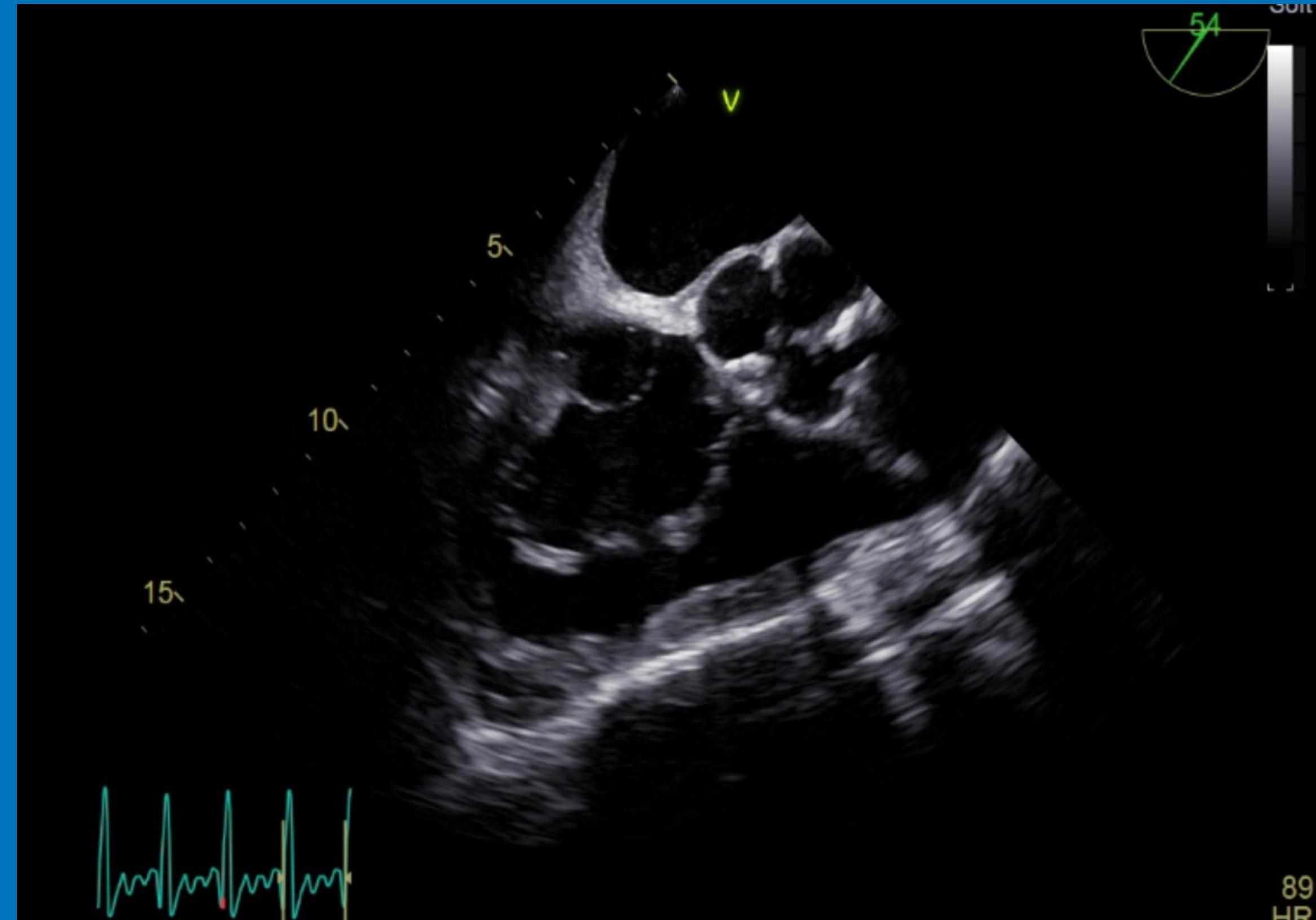
Introduction

Cor triatriatum dexter is a rare congenital heart disease resulting from persistence of the right valve of the sinus venosus. The membrane divides the right atrium into an upper and a lower chamber.

Case description

A 70-year-old Asian man with past medical history of hypertension, cataracts and a known heart murmur since childhood presents with worsening dyspnea on exertion and severe fatigue. Denies any history of chest pain, palpitations, diabetes, dyslipidemia or smoking. No family history of heart disease.

Vitals were stable. BMI was 17.2. He had a 4/6 systolic murmur with a palpable thrill best heard at the left lower sternal border and a right ventricular heave. Trans-thoracic echocardiography was significant for an EF of 55-65% with moderate/severe mitral and tricuspid regurgitation. Trans-esophageal echocardiography revealed normal left ventricular size and systolic function with evidence of a septum within the right atrium diagnosed as Cor triatriatum dexter. There was an intra-atrial septal aneurysm with a small patent foramen ovale and a right-to-left shunt. Left heart catheterization was significant for a vertically rotated heart and large caliber coronary vessels with no stenosis. The patient was followed up in the cardiology clinic for continued evaluation of his valvular disease and cor triatriatum, which appear to be the main causes of his symptoms.



Discussion

Cor triatriatum dexter has an incidence of approximately 0.1% of congenital heart malformations. During fetal life, the right valve of the sinus venosus divides the right atrium into two and serves to direct the oxygenated venous return from the inferior vena cava across the foramen ovale to the left side of the heart. Normally, the valve regresses at 12 weeks gestation. Persistence results in separation between the smooth and trabeculated portions of the right atrium, forming cor triatriatum dexter. The upper chamber receives the venous blood from both vena cavae and the lower chamber is in contact with the tricuspid valve. Clinical manifestations and age of presentation depend on the degree of septation. Mild disease is asymptomatic and is an incidental finding. More severe septation can cause right-sided heart failure due to obstruction of the tricuspid valve, the right ventricular outflow tract, or the inferior vena cava. It can occur in isolation or associated with other malformations such as pulmonary artery stenosis/atresia, tricuspid valve abnormalities, ASD, or Ebstein anomaly. Cyanosis is a rare presentation. The PFO in our patient was small and would not have resulted in significant right-to-left shunting.

MRI has higher detection rates (95%) when compared with echocardiography (38%) and cardiac angiography (69%). Asymptomatic patients are generally not treated unless they are undergoing cardiac surgery for other reasons. Treatment for symptomatic patients is percutaneous catheter disruption of the membrane which is preferred to open heart surgery.

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