A Five Chambered Heart

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Vitals were stable. BMI was 17.2. He had a 4/6 systolic murmur 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

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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.

References