

When the Atrial Septal Defect is No Longer Silent: A Case of a Large, Symptomatic Adult Congenital Heart Defect

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Clinical Presentation

A 49-year-old woman with diabetes mellitus and hyperlipidemia presented with atypical chest pain, palpitations, orthopnea, and reduced exercise tolerance for a week. Physical exam was significant for a 3/6 holosystolic murmur heard loudest in tricuspid area with splitting of S1 in the pulmonic area and louder with inspiration. Her initial vitals and laboratory studies were unremarkable except for a stable normocytic anemia. Electrocardiogram (EKG) showed a normal sinus rhythm with incomplete right bundle branch block (RBBB). Computed tomographic angiogram of the chest showed signs of right-sided heart failure with a large atrial septal defect (ASD) but no aortic dissection. Further imaging studies revealed a large ASD not amenable to percutaneous closure.

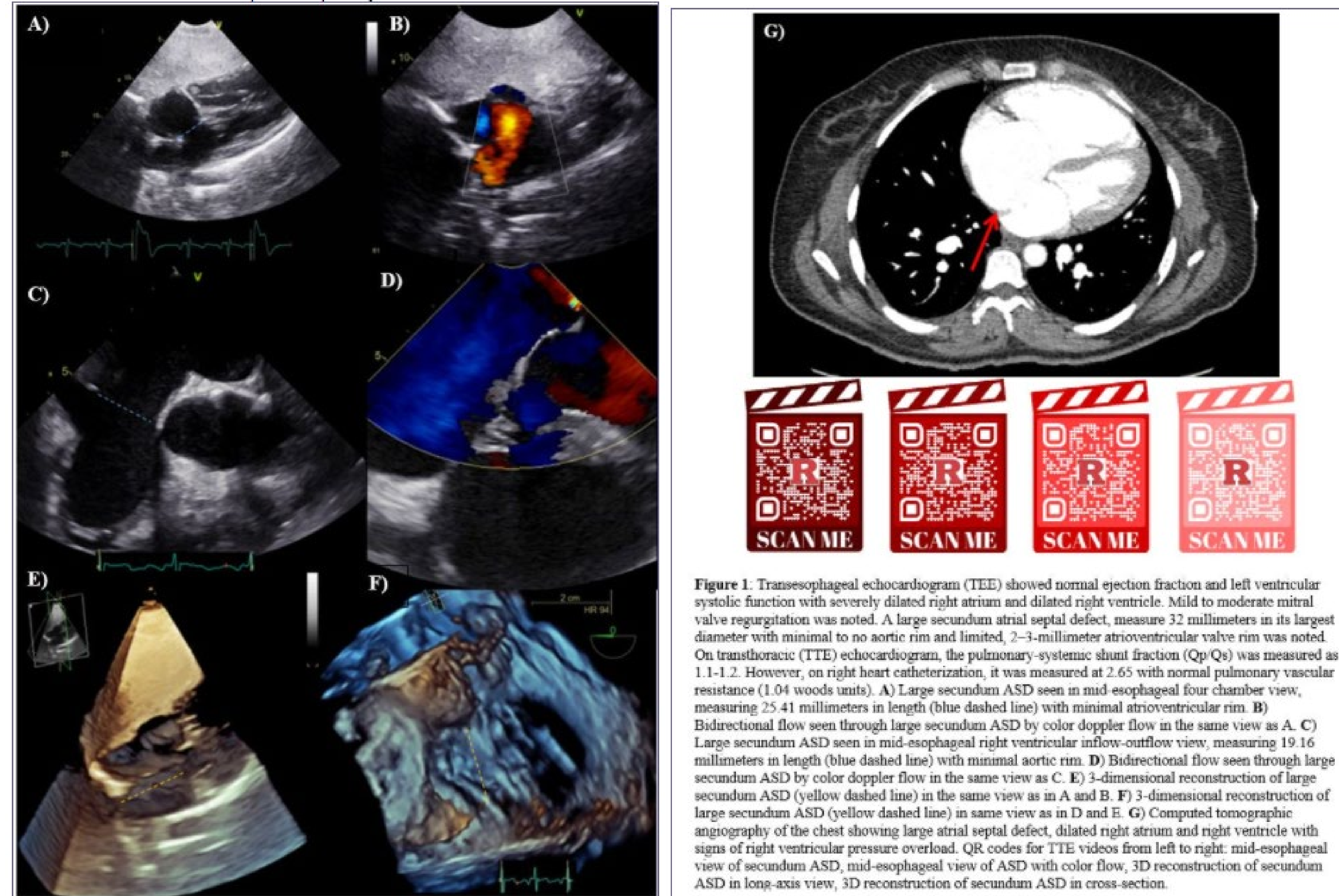


Figure 1: Transesophageal echocardiogram (TEE) showed normal ejection fraction and left ventricular systolic function with severely dilated right atrium and dilated right ventricle. Mild to moderate mitral valve regurgitation was noted. A large secundum atrial septal defect, measure 32 millimeters in its largest diameter with minimal to no aortic rim and limited, 2–3-millimeter atrioventricular valve rim was noted. On transthoracic (TTE) echocardiogram, the pulmonary-systemic shunt fraction (Qp/Qs) was measured as 1.1-1.2. However, on right heart catheterization, it was measured at 2.65 with normal pulmonary vascular resistance (1.04 woods units). A) Large secundum ASD seen in mid-esophageal four chamber view, measuring 25.41 millimeters in length (blue dashed line) with minimal atrioventricular rim. B) Bidirectional flow seen through large secundum ASD by color doppler flow in the same view as A. C) Large secundum ASD seen in mid-esophageal right ventricular inflow-outflow view, measuring 19.16 millimeters in length (blue dashed line) with minimal aortic rim. D) Bidirectional flow seen through large secundum ASD by color doppler flow in the same view as C. E) 3-dimensional reconstruction of large secundum ASD (yellow dashed line) in the same view as in A and B. F) 3-dimensional reconstruction of large secundum ASD (yellow dashed line) in same view as in D and E. G) Computed tomographic angiography of the chest showing large atrial septal defect, dilated right atrium and right ventricle with signs of right ventricular pressure overload. QR codes for TTE videos from left to right: mid-esophageal view of secundum ASD, mid-esophageal view of ASD with color flow, 3D reconstruction of secundum ASD in long-axis view, 3D reconstruction of secundum ASD in cross-section.

Imaging Findings

See Figure 1 for full details.

Role of Imaging in Patient Care

TTE and TEE were crucial in identifying and characterizing the ASD. With minimal atrioventricular and aortic rim, percutaneous closure most likely would have failed. As a result, cardiothoracic surgery was involved early and the patient underwent secundum ASD repair with a Synovius bovine pericardium patch with complete resolution of symptoms. She was discharged on metoprolol tartrate, magnesium oxide, aspirin, atorvastatin, and iron sulfate.

Discussion

ASDs can result from malformation of the ostium primum, ostium secundum, sinus venosus, or the coronary sinus. Though most patients with these lesions are asymptomatic in early adulthood, pathogenesis can occur when increased pulmonary blood flow leads to remodeling of the right ventricle, pulmonary hypertension, and ultimately, shunt reversal. ASDs that lack a viable rim necessitate open closure. Thus, early detection of these lesions can preclude development of devastating cardiovascular morbidity and mortality and vastly improve quality of life.