DIAGNOSTIC DILEMMA

Paul S. Pagel, MD, PHD Section Editor

An Unusual Cause for New Right Bundle-Branch Block

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A 59-YEAR-OLD, 92-kg, 178-cm man with a past medical history of chronic alcohol and tobacco abuse was referred to the authors' institution by his primary care physician because a routine electrocardiogram found a new right bundle-branch block. The patient denied cardiovascular complaints, but he did report occasional dizziness and dyspnea on exertion that he attributed to age and general deconditioning. The physical examination and laboratory analysis were unremarkable with the exception of pulse oximetry values in the low 90s while the

patient was breathing room air. Transthoracic echocardiography showed right atrial and right ventricular dilatation, normal right ventricular systolic function, severe tricuspid regurgitation, and an estimated pulmonary arterial systolic pressure of 65 mmHg. A bubble study was positive for a right-to-left shunt. The patient underwent transesophageal echocardiography (TEE) as part of the diagnostic evaluation, and the following images were obtained (Figs 1–3; Videos 1 and 2). What is the diagnosis?



Fig 1. Midesophageal bicaval transesophageal echocardiography image showing a large atrial septal defect adjacent to the superior vena cava-right atrial junction.

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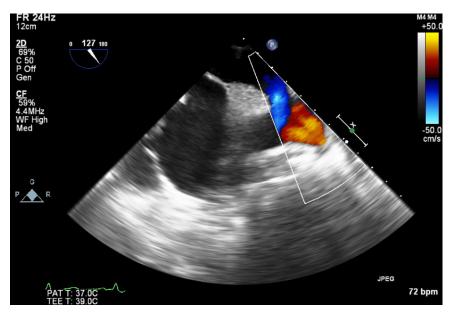


Fig 2. Midesophageal bicaval color Doppler transesophageal echocardiography image showing 2 separate jets of pulmonary venous blood flow (red) entering the distal superior vena cava.

DIAGNOSIS: COMPLEX SINUS VENOSUS ATRIAL SEPTAL
DEFECT WITH PARTIAL ANOMALOUS PULMONARY
VENOUS RETURN OF THE RIGHT SUPERIOR AND MIDDLE
PULMONARY VEINS

A midesophageal bicaval TEE view showed a large defect in the atrial septum adjacent to the superior vena cava-right atrial junction (Fig 1, Video 1). Color Doppler mapping in the midesophageal bicaval plane identified 2 separate jets of pulmonary venous blood flow entering the distal superior vena cava (Fig 2, Video 2) and a large left-to-right shunt through the atrial septal defect (Fig 3, Video 2). These findings were consistent with a complex sinus venosus atrial septal defect

with partial anomalous pulmonary venous return involving at least 2 pulmonary veins. A pulsed-wave Doppler waveform obtained at the level of the atrial septal defect showed systolic pulmonary venous blood flow (positive peak) and pronounced diastolic flow reversal (small and large negative peaks) consistent with the atrial left-to-right shunt (Fig 4). The atrial septal defect (approximately 1.2 cm in largest dimension) also was noted during examination of the superior aspect of the left atrium in a modified upper esophageal TEE imaging plane (Fig 5, Video 3). Marked right atrial and right ventricular dilatation also were observed (Fig 6, Video 4), as previously reported in the transthoracic echocardiography study. Severe



Fig 3. Midesophageal bicaval color Doppler transesophageal echocardiography image showing a large left-to-right shunt through the atrial septal defect.

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