High-Velocity Jet in the Right Atrium: What Is the Diagnosis?

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66-year-old man with a history of hypertension and hyperlipidemia presented to the emergency department with palpitations and was found to have paroxysmal supraventricular tachycardia. Auscultation of the chest revealed a high-pitched holosystolic murmur. A transthoracic echocardiogram showed a disturbed color flow signal in the right atrium (RA) not originating from the tricuspid valve (TV), with peak velocities of approximately 5 m/s (Fig. 1; Supplemental Digital Content 1, Supplemental Video 1, http://links.lww.com/AA/B283). Written informed consent was obtained from the patient for publication of this report and accompanying images.

A transesophageal echocardiogram (TEE) was performed to clarify the diagnosis. A modified midesophageal 4-chamber view with Doppler interrogation of the RA showed a turbulent jet that traversed the RA in a medialto-lateral trajectory and was directed toward the free wall. The jet was systolic in timing but had a peak velocity of approximately 5 m/s, which is extremely unusual for a tricuspid regurgitation (TR) jet (Supplemental Digital Content 2, Supplemental Video 2, http://links.lww.com/AA/B284). In the midesophageal aortic short-axis view, advancing and withdrawing the probe slightly on color Doppler demonstrated no turbulence above the level of the aortic annulus, as would be seen with a ruptured sinus of Valsalva aneurysm, and no evidence of extracardiac flow as seen with an aorta-RA tunnel, but instead localized the origin of the jet to the level just below the aortic valve, consistent with a small perimembranous left ventricular (LV)-to-RA communication (Supplemental Digital Content 3, Supplemental Video 3, http://links.lww.com/AA/B285). A deep transgastric view with color Doppler interrogation of the interventricular septum confirmed this finding (Fig. 2). Examination of the TV did not reveal any gross anatomic defects, and no other significant valvular pathology was observed. The right heart chambers were dilated, and LV and right ventricular systolic functions were normal.

The patient was taken to the operating room, where an LV-to-RA shunt was confirmed and repaired successfully under cardiopulmonary bypass. A cleft was noted on the septal leaflet of the TV and also was repaired. This abnormality was not detected in the prebypass TEE examination but could be identified upon retrospective review of the images (Fig. 3). The postbypass examination revealed no residual shunt.

DISCUSSION

The finding of a high-velocity jet in the RA can be a diagnostic challenge, and several etiologies need to be considered (Table 1). When performing a TEE examination, careful visualization of the jet direction, timing during the cardiac cycle, velocity, and spectral Doppler morphology are essential to provide an accurate diagnosis.

TR is the most common cause of a high-velocity jet in the RA. It is most frequently caused by dilation of the tricuspid annulus and right ventricle (RV) (functional TR) in patients with pulmonary hypertension, RV ischemia or infarction, constrictive pericarditis, or pulmonic stenosis. Spectral Doppler interrogation of the jet is commonly done to estimate pulmonary artery systolic pressure. The TR jet is typically central and always systolic, with a velocity that is typically 2 to 2.5 m/s and is related to the pressure gradient between the RV and the RA (and not to severity of TR). As RA pressures increase over time, the RV-RA gradient and consequently the velocity of the TR jet will decrease. If a jet originating from a left-sided (systemic pressure) chamber such as the LV or the aorta is interpreted as a TR jet, the patient may be incorrectly diagnosed with pulmonary hypertension.¹

Sinus of Valsalva aneurysms are rare, accounting for 0.14% cardiac surgical procedures.² Aneurysms arise most frequently from the right sinus, and these are also most likely to rupture, typically into the RV. Those from the noncoronary sinus typically project and fistulize into the RA.³ Echocardiographic examination in these patients shows aneurysmal dilation of one or more of the sinuses. If a fistula is formed, it may be seen as a "windsock" protrusion into the RA, with a high-velocity systolic and diastolic turbulent jet on color Doppler that is directed laterally toward the RA free wall. M-mode Doppler is invaluable when assessing the timing of the jet.

LV-to-RA communications, also referred to as Gerbode defects, are <1% of congenital cardiac defects but may also be acquired.⁴ Normally, the TV annulus is slightly (5–10 mm) apically displaced compared with the mitral valve annulus. Consequently, a small membranous septum separates the LV from the RA (atrioventricular segment). Type 1 (direct) Gerbode defects are a direct LV-to-RA communication through the atrioventricular septum and typically are acquired. Type 2 (indirect) Gerbode defects are most frequently of congenital origin and consist of a combination of a perimembranous ventricular septal defect associated with a defect in the septal or anterior leaflet of the TV.⁵ In both

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Figure 1. A, Apical 4-chamber transthoracic echocardiographic view. Color Doppler interrogation shows a high-velocity jet at the level of the tricuspid valve. B, Continuous-wave Doppler interrogation of the jet demonstrates a peak velocity of approximately 5 m/s. RV = right ventricle; RA = right atrium; LV = left ventricle; LA = left atrium.



Figure 2. Deep transgastric transesophageal echocardiographic zoomed in "color compare" view demonstrating the presence of a high-velocity jet originating from the left ventricle (LV; on the left ventricular outflow tract [LVOT] side of the aortic valve). The receiving chamber is the right atrium (RA). Involvement of the tricuspid valve could not be determined from this view. RV = right ventricle.



Figure 3. Modified midesophageal 4-chamber view with focus on the tricuspid valve in early systole. A, The asterisk (*) indicates the septal leaflet defect. B, Color Doppler interrogation of the tricuspid valve area shows a high-velocity jet originating in the LV and entering the RA toward the RA free wall. RV = right ventricle; RA = right atrium; LV = left ventricle; LA = left atrium.

cases, the jet is directed toward the RA-free wall, occurs during systole, and has a high velocity (approximately 5 m/s). Given the morphology and the lack of a history of cardiac surgery, infective endocarditis, or trauma, we speculate that the origin of our patient's defect was congenital.

A high-velocity jet in the RA also may occur when a perimembranous ventricular septal defect orifice extends below the septal TV leaflet, allowing the jet to force the anterior leaflet away from the septal leaflet and resulting in significant TR.⁶ Other infrequent etiologies for a high-velocity RA jet include a ventricular septal defect with aneurysmal transformation, aorta-to-RA tunnel, coronary artery fistula into the RA, and caval obstruction (Table 1).⁷ Although the types of atrial septal defect typically demonstrate low-velocity flow, the combination of a small atrial septal defect with high left atrial pressure could increase the shunt velocity.

In conclusion, the presence of a high-velocity jet in the RA can be a diagnostic challenge. Knowledge of its potential causes, including infrequent congenital and acquired cardiac defects, and careful visualization of the jet direction, timing, velocity, and spectral Doppler morphology by means of TEE are essential to provide an accurate diagnosis and to direct appropriate management.

DISCLOSURES

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Clinician's Key Teaching Points

By Roman M. Sniecinski, MD, Martin M. Stechert, MD, and Martin J. London, MD

- Left ventricular-to-right atrial (LV-RA) shunts, also known as "Gerbode defects," can be distinguished by 2 types: type I defects are direct communications of the LV to the RA through the superior aspect of the atrioventricular septum. They usually are acquired from endocarditis, trauma, or ischemic heart disease. Type 2 defects are mostly congenital, and they are described as a combination of a perimembranous ventricular septal defect with a defect in one of the tricuspid valve leaflets, thereby creating an indirect LV-RA communication via the right ventricle.
- Gerbode defects may be suspected when marked RA enlargement is seen on a transesophageal echocardiogram that is not otherwise explained. The hallmark is a high-velocity jet (typically 5 m/s or more, depending on LV-RA pressure gradient) noted on Doppler within the RA during systole. This can be confused with the commonly observed systolic jet of tricuspid regurgitation (TR), although in the absence of pulmonary hypertension, the TR jet is significantly lower velocity than 5 m/s.
- In this case of a high-velocity color flow Doppler jet within the RA, spectral Doppler examination confirmed the absence of a diastolic component, thus excluding possible diagnoses such as a ruptured sinus of Valsalva aneurysm or a coronary artery fistula, both of which have Doppler signals extending throughout systole and diastole. Meticulous examination using modified midesophageal and transgastric imaging planes revealed a direct LV-RA communication.
- Color flow Doppler jets within the RA can originate from a variety of sources, not all are a result of TR. Key components for identifying the etiology include determination of the pressure gradient, as well as timing of the jet, that is, during systole, diastole, or throughout the cardiac cycle.