An Occult Aortocavitary Fistula Presenting as Apparent Tricuspid Valve and Aortic Valve Endocarditis

Thong Nguyen, DO, Mousumi Andersen, MD, Rohesh J. Fernando, MD, FASE, and Karl Richardson, MD, *Winston-Salem, North Carolina*

INTRODUCTION

Infective endocarditis (IE) is a potentially lethal infection with an annual incidence of three to nine cases per 100,000 persons. Patients who are at increased risk include those with prosthetic valves, prior IE, unrepaired congenital heart disease, diabetes mellitus, human immunodeficiency virus infection, and/or intravenous drug abuse.[1] Some of these risk factors can certainly raise the propensity for vegetations to develop on a particular valve. For example, intravenous drug abuse and/or cardiac implanted devices can increase the incidence of right-sided endocarditis, which represents approximately 5% to 10% of all cases of IE.[2] Overall, single-valve involvement accounts for the majority of cases (about 78%), whereas multivalve involvement is much less common (about 17%–19%).[3,4] Complications from the disease can be significant and include valvular destruction, congestive heart failure, stroke and systemic embolizations, conduction abnormalities, and bacteremia.[5] Rarely, perivalvular structures can be involved, leading to abscess formation, valve disruption, and in some cases more serious complications. We present a case in which IE conferred significant morbidity to a patient as a result of an aortocavitary fistula, which is an anomalous connection from the aorta to a cardiac chamber.

CASE PRESENTATION

A 70-year-old woman with a known bicuspid aortic valve and limited cardiology follow-up presented with acute onset shortness of breath following 2 weeks of intermittent fevers and chills. Upon arrival to the emergency department, she was febrile, with a temperature of 102.9°F, and hypoxic, with an oxygen saturation of 91%. A grade III/VI midpeaking crescendo-decrescendo systolic murmur in addition to a grade II/IV diastolic murmur was appreciated in the right upper sternal border. Her laboratory results were notable for leukocytosis (21×10^9/L) and a troponin level of 0.239 ng/mL. Shortly after arrival, the patient experienced respiratory arrest, requiring intubation and 3 mins of cardiopulmonary resuscitation.

Conflicts of interest: The authors report no actual or potential conflicts of relative to this document.

Because of the murmur on examination and new blood cultures revealing bacteremia, transthoracic echocardiography (TTE) was performed. Notable echocardiographic findings included aortic regurgitation, which was seen in the apical five-chamber view (Figure 1, Video 1). The aortic regurgitation was further evaluated with continuous-wave Doppler envelopes, which yielded a pressure half-time of 216 msec (Figure 2). Also, pulsed-wave Doppler envelopes within the left ventricular outflow tract demonstrated an elevated velocity-time integral of 33.1 cm (Figure 3), likely due to increased stroke volume from significant regurgitation. Overall, these findings were consistent with moderate aortic insufficiency. Additionally, elevated continuous Doppler velocities of approximately 3.8 m/sec were measured across the aortic valve, potentially consistent with moderate aortic stenosis (Figure 4).

More immediately concerning, however, was the presence of a mobile echo density that appeared to be associated with the tricuspid valve (Figure 5, Video 2) and an additional smaller mobile echo density that appeared adherent to the heavily calcified, potentially bicuspid aortic valve (Figure 6, Video 3). The presence of these mobile echo densities on both the aortic and tricuspid valves within the clinical context was concerning for an atypical distribution of right- and left-sided endocarditis.

Without an explanation for the respiratory arrest and noting the atypical distribution of infectious vegetations, transesophageal echocardiography (TEE) was urgently performed in the intensive care unit. This confirmed the calcified bicuspid aortic valve (Figure 7, Video 4). The aortic valve echo density seen on TTE was confirmed as a 0.6 × 0.4 cm, irregular, independently mobile mass on the ventricular aspect of the aortic valve adherent to the noncoronary cusp, suspicious for an infectious vegetation. The aortic insufficiency was graded as moderate to severe and was still likely underestimated because of the acute nature of the regurgitation with elevated left ventricular end-diastolic pressures. Additionally, a large, independently mobile 1.8 × 0.8 cm heterogeneous mass was observed in the right atrium superior to the septal leaflet of the tricuspid valve that appeared consistent with a second vegetation. This larger vegetation appeared to intermittently contact the tricuspid valve but did not appear attached to the valve itself (Figure 8, Video 5). There also appeared to be a small aneurysmal section of the noncoronary cusp immediately adjacent to the vegetation near the tricuspid valve. Additional imaging confirmed that the vegetation was superior to the tricuspid valve (Figure 9, Video 6). The tricuspid valve was otherwise normal in function and appearance.

The right atrial vegetation was adherent to the aortic root. Further examination revealed a small 1.0 × 0.5 cm aneurysmal section of the noncoronary sinus of the aorta immediately adjacent to the right atrial vegetation with continuous color Doppler flow into the right atrium along the path of the vegetation, suggestive of a fistulous connection between the aortic root and the right atrium (Figures 10 and 11, Video 7). The right ventricle was not dilated, suggesting an acute
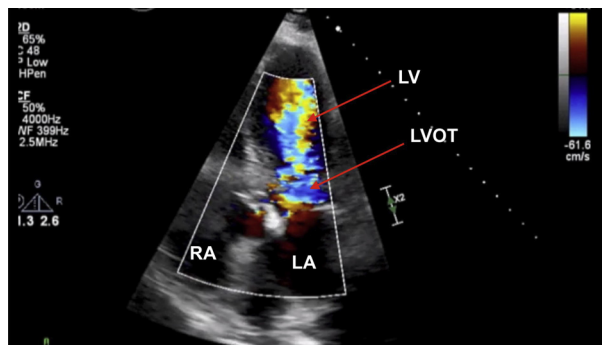
Figure 1 Transthoracic echocardiographic image of apical five-chamber view depicting aortic insufficiency. *LA*, Left atrium; *LV*, left ventricle; *LVOT*, left ventricular outflow tract; *RA*, right atrium.
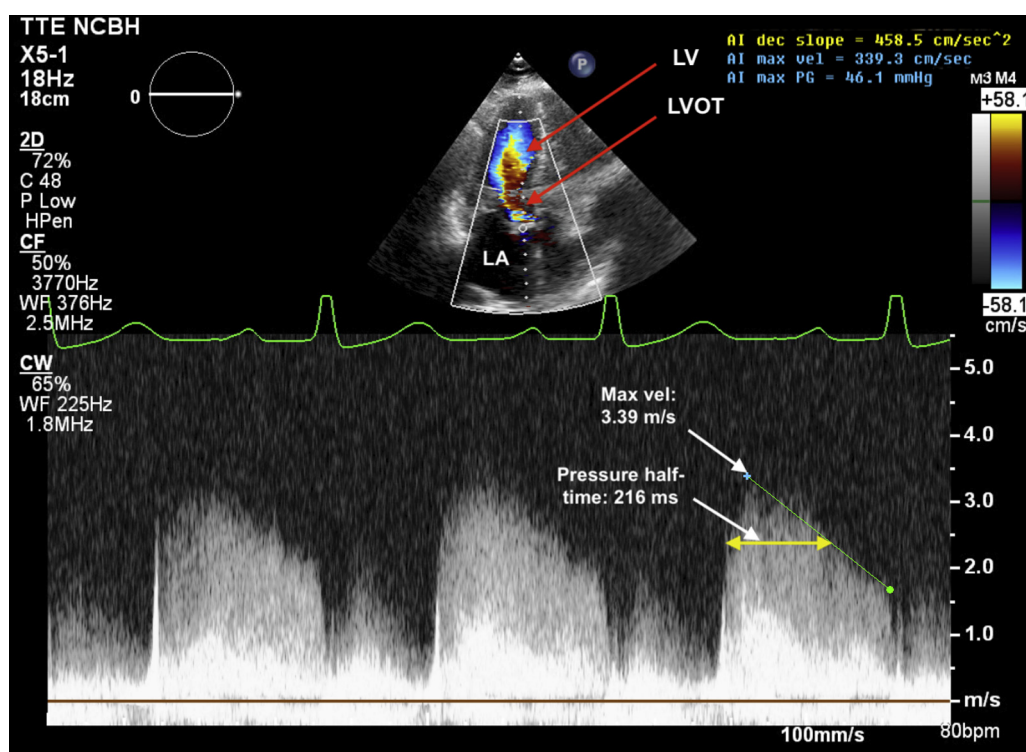

Figure 2 Transthoracic echocardiographic image of apical long-axis view depicting aortic insufficiency (AI) continuous-wave Doppler envelopes consistent with moderate aortic insufficiency (*green line*). The pressure half-time (*double-headed yellow arrow*) can be calculated using the difference between the maximal velocity (max vel), 3.39 m/sec (*top white arrow*), and the velocity at half-time, 2.4 m/sec, and the slope of the envelope (4.58 m/sec^2). This value is calculated to be 216 msec, also consistent with moderate aortic insufficiency. Color Doppler flow demonstrating regurgitant jet from the left ventricular outflow tract (LVOT) (*bottom red arrow*) back to the left ventricle (LV) (*upper red arrow*). *LA*, Left atrium; *PG*, pressure gradient.

left-to-right shunt. Cardiothoracic surgery was immediately consulted, and the patient was taken to the operating room, where the presence of the fistula was confirmed (Figure 12). She underwent successful aortic root replacement, right atrial debridement, and repair of the fistula with a pericardial patch. Surgical inspection confirmed that the tricuspid valve was not involved.

A few days postoperatively, the patient was successfully weaned off vasopressor support and mechanical ventilation. The results of her blood cultures grew viridans streptococcus of the mitis/oralis subtype, which was treated with 6 weeks of intravenous ceftriaxone. Her postoperative course was overall uncomplicated, and she returned to the

cardiology clinic a few weeks later reporting enjoyment of cardiac rehabilitation.

DISCUSSION

An aortocavitary fistula is an uncommon complication of IE. In one large multicenter retrospective study that evaluated 4,681 patients diagnosed with IE on the basis of the modified Duke criteria,[6] only 76 (approximately 1.6%) developed aortocavitary fistulas. The majority were diagnosed using echocardiography, although a few were found during surgery. The origin of fistula formation from the aortic
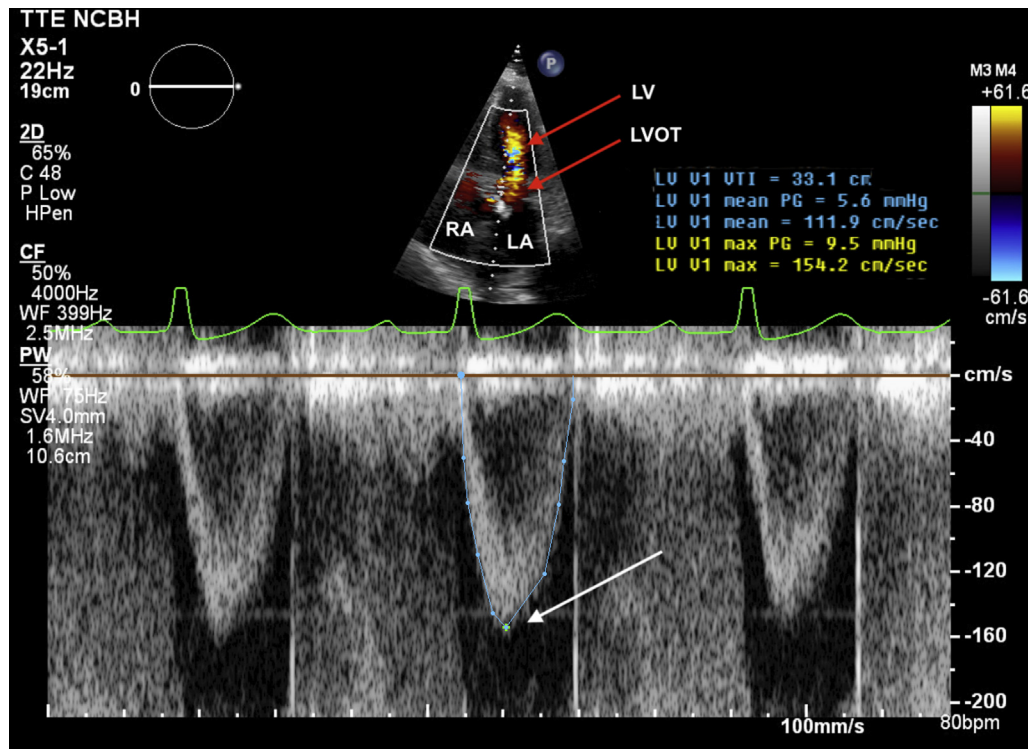
Figure 3 Transthoracic echocardiographic image of apical five-chamber view depicting pulsed-wave Doppler envelopes showing elevated left ventricular outflow tract (LVOT) velocity-time integral (VTI) (33.1 cm), consistent with increased flow from significant aortic insufficiency (*white arrow*). *LA*, Left atrium; *LV*, left ventricle; *PG*, pressure gradient; *RA*, right atrium.
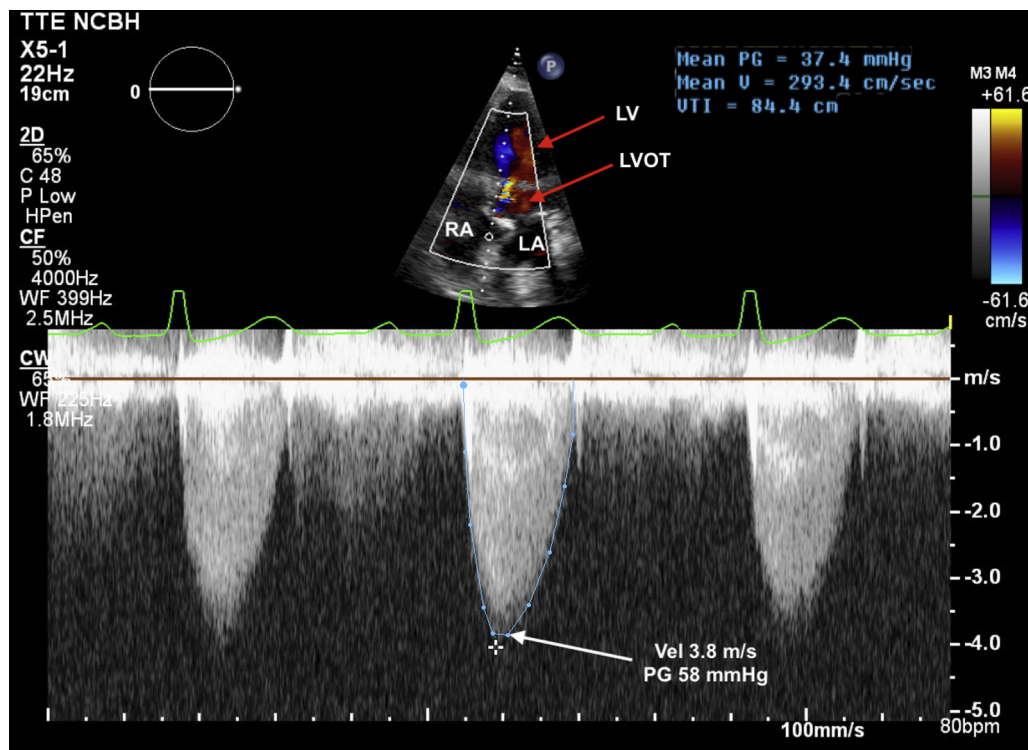

Figure 4 Transthoracic echocardiographic image of apical five-chamber view depicting moderately elevated aortic valve continuous-wave Doppler velocities (*white arrow*) at about 3.8 m/sec, potentially consistent with moderate aortic stenosis. *LA*, Left atrium; *LV*, left ventricle; *LVOT*, left ventricular outflow tract; *PG*, pressure gradient; *RA*, right atrium; *V*, velocity; *Vel*, velocity; *VTI*, velocity-time integral.
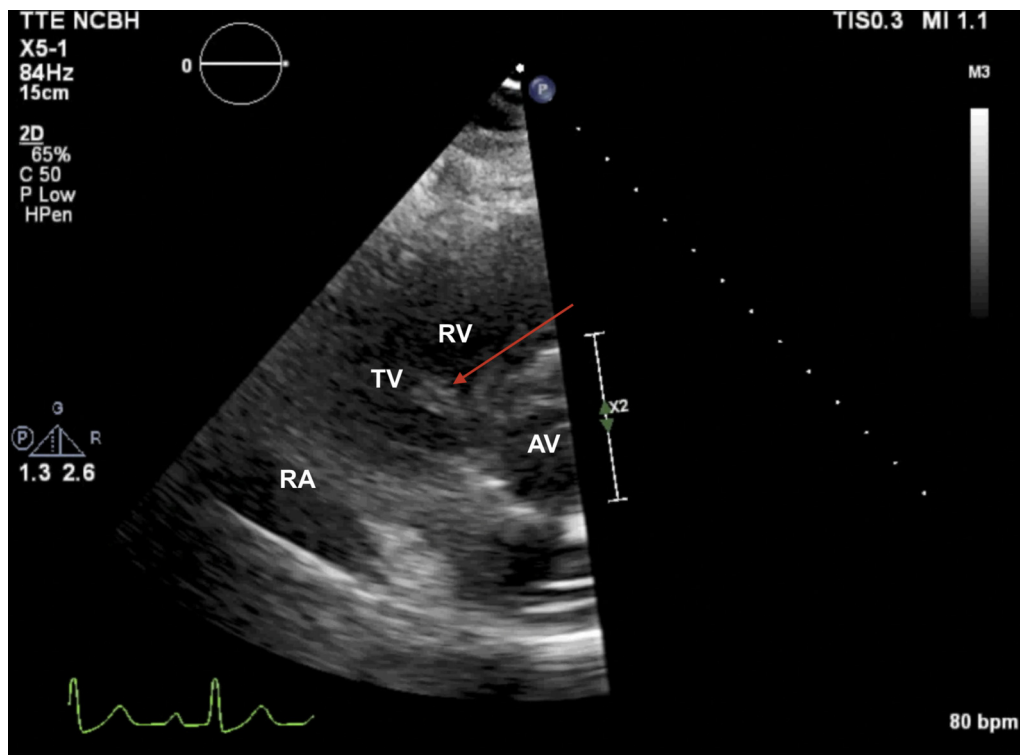
Figure 5 Transthoracic echocardiographic image of parasternal short-axis view depicting mobile echo density that appears to be associated with the tricuspid valve suggesting vegetation (*red arrow*). *AV*, Aortic valve; *RA*, right atrium; *RV*, right ventricle; *TV*, tricuspid valve.
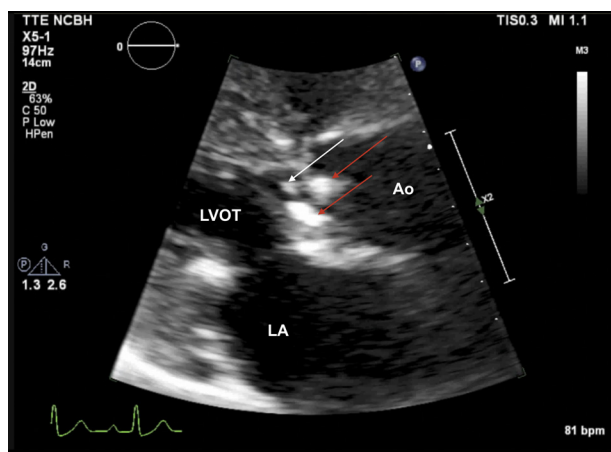

Figure 6 Transthoracic echocardiographic image of parasternal long-axis view depicting heavily calcified aortic valve (*red arrows*) and a small mobile echo density on the LV aspect of aortic valve (*white arrow*). *Ao*, Aorta; *LA*, left atrium; *LVOT*, left ventricular outflow tract.
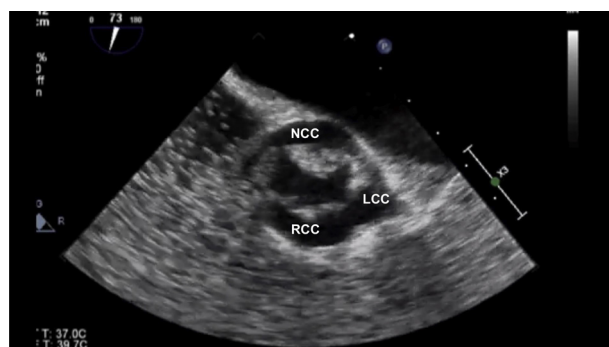

Figure 7 Transesophageal echocardiographic midesophageal aortic valve short-axis view depicting a bicuspid aortic valve. *LCC*, Left coronary cusp; *NCC*, noncoronary cusp; *RCC*, right coronary cusp.

sinuses in this series was right coronary sinus (37%), left coronary sinus (38%), and noncoronary sinus of Valsalva (25%). In terms of fistula development from the noncoronary sinus, there appeared to be an equal occurrence of tract development to all four chambers of the heart.

Once an aortocavitary fistula is diagnosed, the characteristics of the fistulous connection between the aorta and one of the other chambers of the heart should be evaluated to determine the potential for hemodynamic instability. First, the size should be considered, because a larger fistula allows a bigger shunt, which could increase the potential for volume overload.[6] In addition, the site of the fistula could also play a role, as the relative pressures of the two chambers and blood vessels will influence the direction and magnitude of blood flow. Finally, the presence of a fistula should prompt the echocardiographer to look for pathology that may be associated with the fistula, such as abscess, tis-
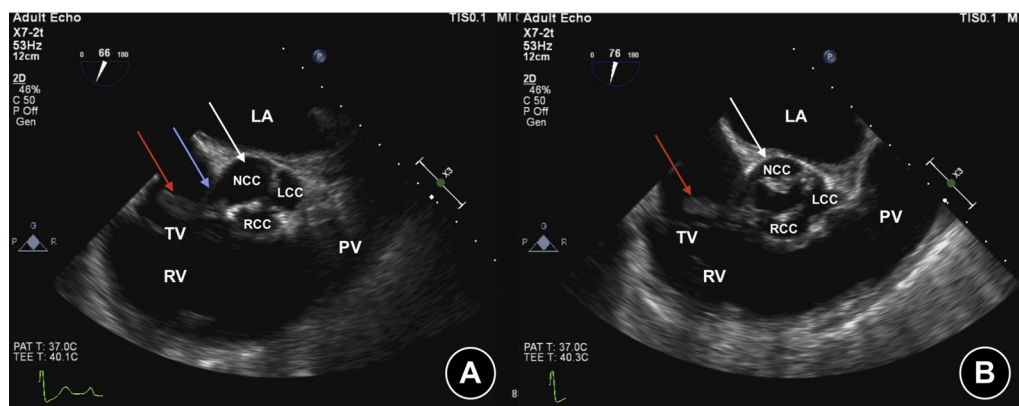
Figure 8 Transesophageal echocardiographic images of midesophageal aortic valve short-axis view with left image **(A)** depicting closed bicuspid valve (*white arrow*) and right image **(B)** depicting open bicuspid valve (*white arrow*). Large mobile vegetation is clearly superior and not attached to tricuspid valve (TV) (*red arrows*). In the *left image* **(A)**, there is a small aneurysmal section of the noncoronary cusp (NCC) immediately adjacent to the right atrial vegetation (*blue arrow*). *LA*, Left atrium; *LCC*, left coronary cusp; *PV*, pulmonary valve; *RCC*, right coronary cusp; *RV*, right ventricle.
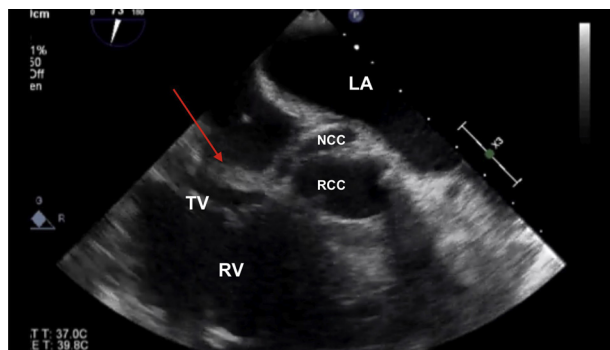

Figure 9 Additional transesophageal echocardiographic midesophageal aortic valve short-axis view depicting vegetation superior to tricuspid valve (TV) leaflet (*red arrow*). *LA*, Left atrium; *NCC*, noncoronary cusp; *RCC*, right coronary cusp; *RV*, right ventricle.

Figure 10 Transesophageal echocardiographic midesophageal aortic valve short-axis view with color Doppler flow demonstrating fistulous connection from noncoronary sinus of the aortic valve to right atrium (*white arrow*) adjacent to the tricuspid valve (TV) vegetation. Tricuspid regurgitation (*red arrow*) is also seen. *LA*, Left atrium; *NCC*, noncoronary cusp; *PV*, pulmonary valve; *RV*, right ventricle.

sue destruction, and infectious vegetations that could compromise valvular function. This can help clarify the etiology of the fistula as infectious or noninfectious. This information along with the size and location may inform the urgency of surgical repair.

Regarding our patient's case, determining the etiology of this patient's acute respiratory failure was initially challenging. Her findings on TTE of moderate aortic insufficiency and moderate aortic stenosis were both likely chronic in nature and would not have likely explained the acute decompensation. The noted endocarditis, although immediately concerning, did not clearly explain acute hypoxic respiratory failure. It was the atypical distribution of right- and left-sided endocarditis and relatively unexplained hemodynamic compromise that prompted urgent further investigation with a more invasive imaging modality. Careful inspection on TEE revealed continuous color Doppler flow from the aorta to the right atrium, consistent with an aortocavitary fistula. The left-to-right continuous flow was entirely obscured on TTE by turbulent color Doppler signals produced by concomitant aortic stenosis, aortic regurgitation, and tricuspid regurgitation. The normal-sized right ventricle suggested an acute manifestation of the left-to-right shunt. This shunt was therefore presumed to be an acute complication of her endocarditis, likely originating on the
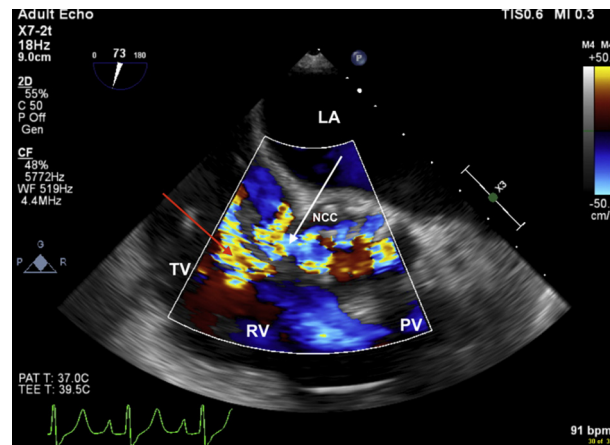
diseased bicuspid valve, leading to her hemodynamic collapse. Prompt surgical intervention was sought, and the patient eventually made a remarkable recovery.

This case encourages clinicians to maintain a high level of suspicion for complications that arise from IE in the setting of known risk factors such as a congenital cardiac malformation. Bicuspid aortic valve is the most common cardiac congenital malformation, and approximately 12% of these patients eventually develop IE, though variable numbers have been reported.[7-9] A retrospective analysis of a cohort of patients with bicuspid aortic valves observed a significantly higher rate of periannular complications compared with patients with tricuspid aortic valves.[10] Although practitioners may focus on intravenous drug abuse and prosthetic valves as common risk factors for IE, vigilance should also be maintained in febrile patients with congenital cardiac abnormalities.
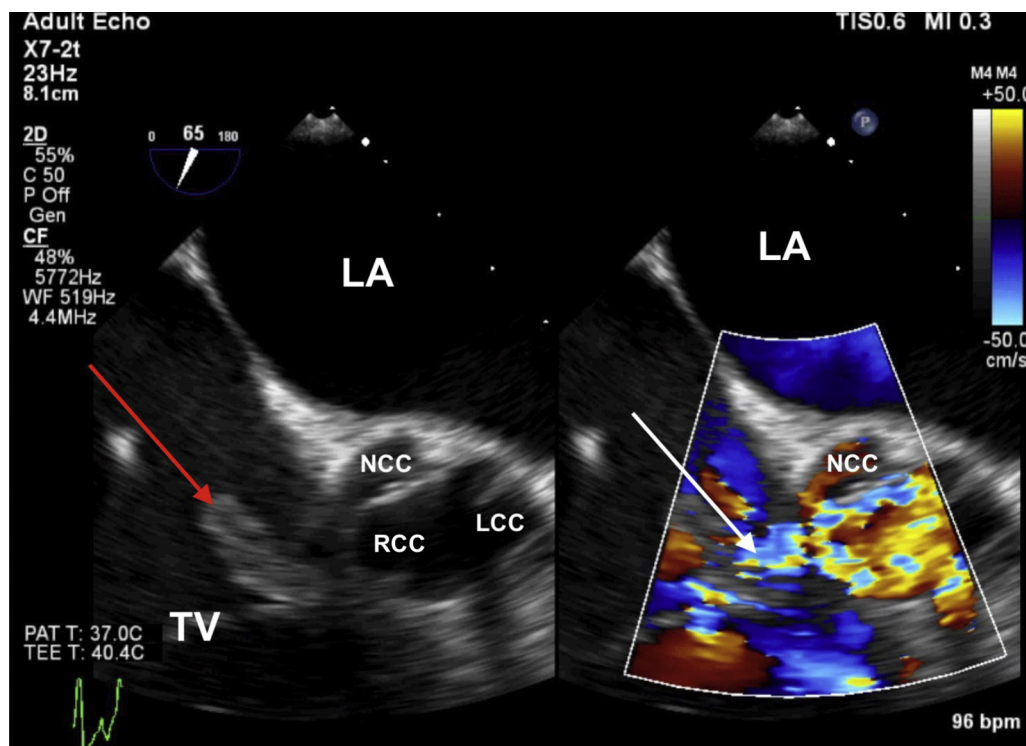
Figure 11 Transesophageal echocardiographic midesophageal aortic valve short-axis view using color compare mode. The *left image* highlights the tricuspid vegetation (*red arrow*), while the *right image* depicts the abnormal fistulous connection (*white arrow*) using color Doppler. *LA*, Left atrium; *LCC*, left coronary cusp; *NCC*, noncoronary cusp; *RCC*, right coronary cusp; *TV*, tricuspid valve.
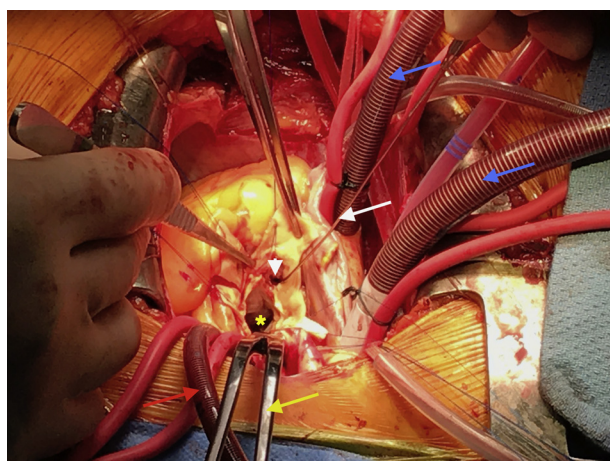

Figure 12 Intraoperative image taken from the head of the surgical table after the native aortic valve was excised (*yellow star*). The image depicts an instrumental probe (*white arrow*) with the tip (*white arrowhead*) placed into the fistula from the noncoronary sinus of Valsalva to the right atrium. The superior vena cava cannula (*lower blue arrow*), inferior vena cava cannula (*upper blue arrow*), aortic cannula (*red arrow*), and aortic cross clamp (*yellow arrow*) are also seen.

Last, the superiority of TEE compared with TTE to detect periannular complications is observed in this report, as well as being demonstrated in previous studies.[11-13] In patients with concerning findings consistent with complications from IE (new murmur, volume overload, pulmonary edema, persistent fevers despite broad antibiotic therapy), early TEE may be indicated to evaluate for these complications before hemodynamic compromise. Diagnoses of these complications (fistula formation, severe valvular abnormalities, or heart failure) are usually indications for emergent surgery.[14,15]

CONCLUSION

Complications from IE require a high level of suspicion and warrant early TEE in an effort to expedite surgery. This case represents an excellent example of the incorporation of clinical suspicion, physical examination findings, and imaging to make a prompt clinical diagnosis and urgent referral to surgery.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.09.003.

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