Isolated Endocarditis of Native Pulmonary Valve in a Pediatric Patient: The Unusual within the Unusual

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INTRODUCTION

Right-sided endocarditis encompasses approximately 12% of all cases of infective endocarditis (IE), with the tricuspid valve being the predominant site of infection.[1] Involvement of the pulmonary valve (PV) alone is unusual and often occurs in the presence of congenital heart disease.[2] The occurrence in patients with a native PV is even lower, with an estimate of 0.2% to 1.2%.[3] Literature pertaining to isolated native PV endocarditis is limited to case reports and series primarily in adults with intravenous drug use.[4,5] We report the clinical presentation, diagnosis, and management in a pediatric patient with isolated native PV endocarditis in the absence of predisposing risk factors.

CASE PRESENTATION

A 15-year old previously healthy girl presented to the emergency department with respiratory distress and increasing fatigue in the setting of high-grade fevers over a 3-day period. Her examination revealed mild tachycardia and tachypnea with coarse breath sounds over bilateral lung fields. There was a "new" systolic murmur in the pulmonic area. She had an elevated white blood cell count $(13,000/mm^3)$ and inflammatory markers. Urine drug screen and human immunodeficiency virus test were negative. She had no history of immunodeficiency, underlying congenital heart disease, or intravenous drug use. Chest radiography (Figure 1) revealed bilateral diffuse opacities. Blood cultures were drawn, and she was admitted for intravenous antibiotics and respiratory support for suspected pneumonia. Shortly after admission, she had acute respiratory decompensation requiring endotracheal intubation. Chest computed tomography showed numerous cavitary lesions and bilateral moderate pleural effusions (Figure 2). Her blood culture grew methicillin-sensitive *Staphylococcus aureus*, and she was switched to intravenous oxacillin therapy. Transthoracic echocardiography (TTE) showed mild right ventricular dysfunction and a thickened PV with possible vegetation, albeit with suboptimal visualization of the PV. Spectral Doppler across

VIDEO HIGHLIGHTS

Video 1: Preoperative TTE in parasternal short-axis view demonstrating thickened PV with mild stenosis and moderate regurgitation.

Video 2: Preoperative three-dimensional TEE showing vegetations attached to the PV.

Video 3: Preoperative TEE showing pulmonic valve dysfunction including mild stenosis and moderate regurgitation.

Video 4: Postoperative parasternal short-axis view showing normal right ventricular size and function.

Video 5: Parasternal short-axis view demonstrating good prosthetic PV function with trivial regurgitation.

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the PV showed mild stenosis and moderate regurgitation warranting further evaluation. The heart was structurally normal, without any congenital heart defects. TTE was limited given the patient's body habitus (Figure 3, Video 1). Transesophageal echocardiography (TEE) was performed with a specific effort to assess the pulmonic valve in detail. It revealed a large vegetation protruding into the main pulmonary artery and prolapse of the PV leaflets into the right ventricular outflow tract (Figures 4A-4C, Videos 2 and 3). There was moderate pulmonary regurgitation. One week after admission, the patient continued to have persistent fevers, valve dysfunction, and persistent septic pulmonary emboli despite appropriate antibiotic therapy. It was deemed that surgical removal of the focus of infection would be the best course of action.

Intraoperative findings were significant for a destroyed PV with multiple vegetations. After debridement and removal of vegetations, a 23-mm homograft was placed in the pulmonic position. Pleural washout and drainage were performed. The patient's postsurgical TEE showed good homograft function and normal right ventricular function (Figure 4D). Pathology specimen of the excised PV confirmed endocarditis with gram-positive cocci (Figure 5). Her immediate postoperative course was significant for hemodynamic instability, likely from a systemic inflammatory response. She was maintained on inotropes and broad spectrum intravenous antibiotics for the first postoperative week. Given dependency on mechanical ventilation, a tracheostomy was performed. She was discharged to a rehabilitation facility and completed a 6-week course of intravenous antibiotics. She was successfully decannulated 3 months later with improved computed tomographic findings (Figure 6). At her most recent follow-up 1 year later, she had no evidence of recurrent infection and had resumed all baseline physical activities. Echocardiography showed normal PV function (Figure 7, Videos 4 and 5).
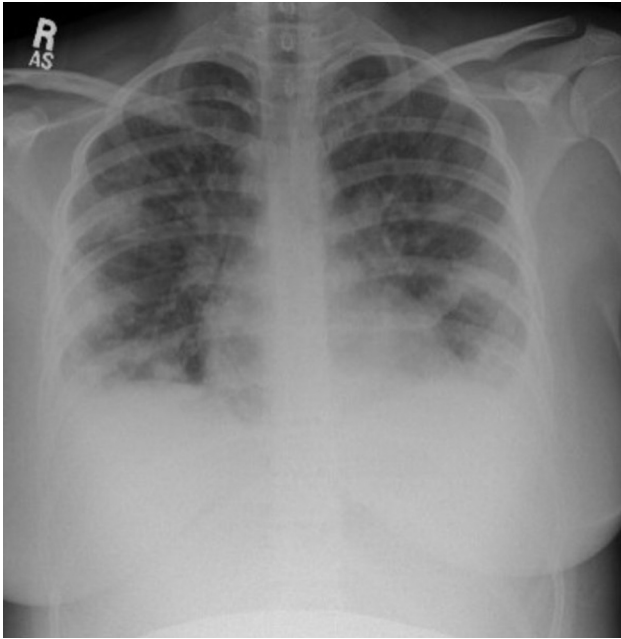
Figure 1 Anteroposterior chest radiograph demonstrating multiple bilateral hazy opacities as well as pleural effusions.

DISCUSSION

We present the case of a healthy young female patient with PV endocarditis complicated by recurrent septic pulmonic emboli and respiratory compromise. TEE was key in the diagnosis and delineation of the vegetation on the pulmonic valve. Despite a thorough workup and history, we were unable to identify any risk factors for her condition. Although her initial course was protracted, she eventually had a favorable outcome using a combination of surgical and medical management.

IE of the right-sided valves, particularly of the PV, is rare. A recent review article in 2016 identified a total of 70 cases of isolated PV endocarditis in literature, with the majority being adults (average age, 44 ± 18 years).[6] In a large international cohort of adult patients with native valve endocarditis, 0.2% to 1.2% of the population had isolated PV endocarditis.[3] In the pediatric population, native PV endocarditis is further uncommon and typically reported in the presence of risk factors.

Although intravenous drug use is the most common risk factor for right-sided IE, the presence of chronic vascular access and pacemakers,[7] immunosuppressed states such as diabetes, cancer, dialysis, and human immunodeficiency virus are also identifiable risk factors in patients with native valve endocarditis.[3] PV endocarditis can occur with preexisting pulmonary stenosis, intracardiac shunts, and surgical PV replacement.[2]
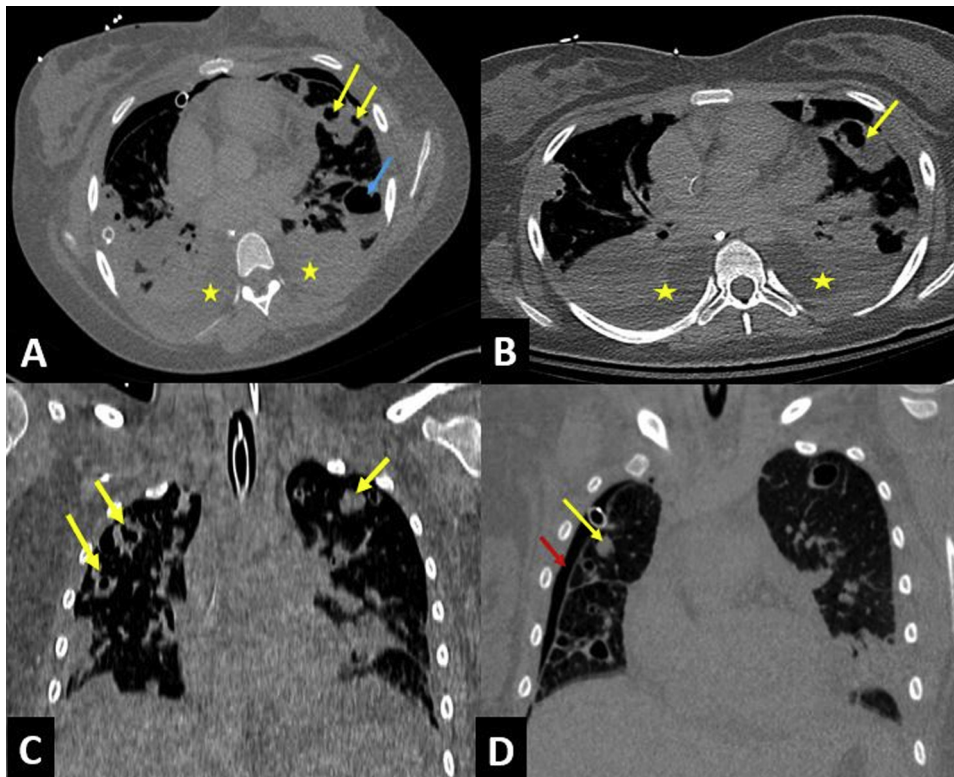

Figure 2 Chest computed tomography without contrast. **(A, B)** Axial views demonstrating multiple solid and cavitary pulmonary lesions, some with air fluid levels (*blue arrow*) consistent with diffuse septic emboli (*yellow arrows*), bilateral pleural effusions (*stars*). **(C, D)** Coronal views with multiple solid and cystic lesions (*yellow arrows*) and small right pneumothorax (*red arrow*).
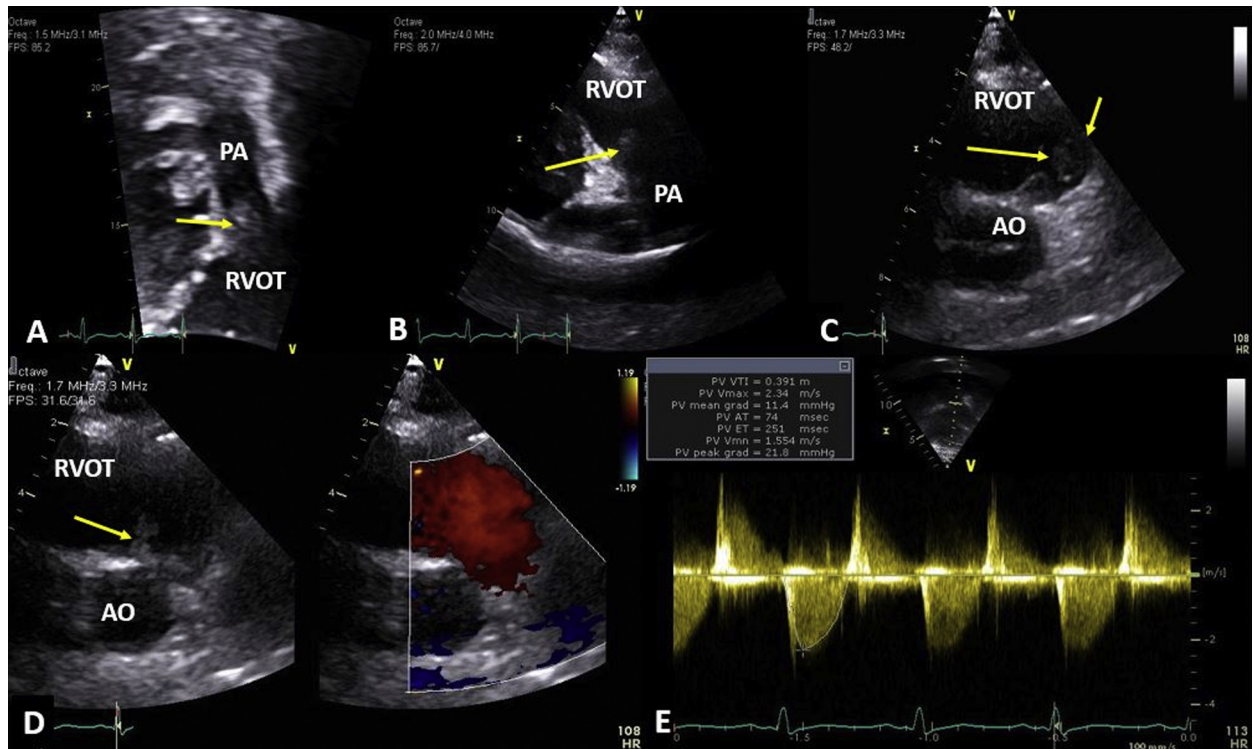
Figure 3 TTE. Subcostal view **(A)**, parasternal long-axis **(B)** and short-axis **(C)** views demonstrating thickened PV (*yellow arrow*) and possible vegetation. **(D)** Parasternal long-axis color-compare image with thickened PV. **(E)** Spectral Doppler image showing mild pulmonic stenosis (peak gradient 22 mm Hg) and moderate regurgitation. *AO*, Aorta; *PA*, pulmonary artery; *RVOT*, right ventricular outflow tract.
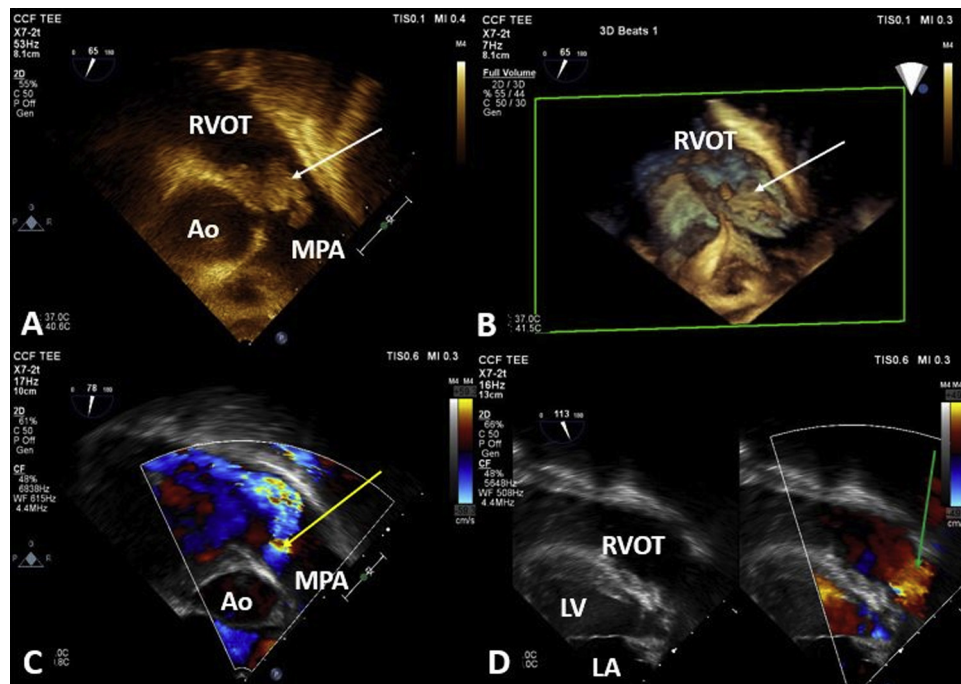

Figure 4 Preoperative TEE. **(A, B)** Two-dimensional (2D) and three-dimensional images showing large lobulated mass attached to the PV (*white arrow*). **(C)** Color Doppler image demonstrating moderate pulmonary regurgitation (*yellow arrow*). **(D)** Postoperative 2D and color-compare transesophageal echocardiographic images showing normal pulmonary homograft function without regurgitation (*green arrow*). *AO*, Aorta; *LA*, left atrium; *LV*, left ventricle; *MPA*, main pulmonary artery; *RVOT*, right ventricular outflow tract.
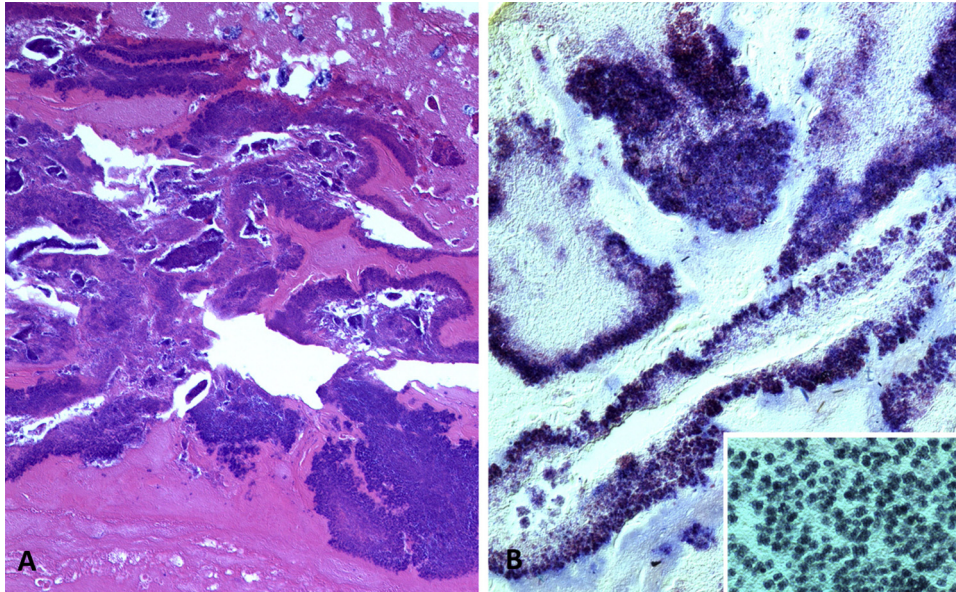
Figure 5 Photomicrograph of excised pulmonic valve. **(A)** Histologic section of the vegetation shows abundant colonies of bacterial microorganisms that appear basophilic in a background of fibrin (hematoxylin and eosin, 100× magnification). **(B)** The microorganisms are stained gram positive (200× magnification). *Inset* shows Grocott methenamine silver stain that demonstrates the morphology of bacteria as cocci (600× magnification).
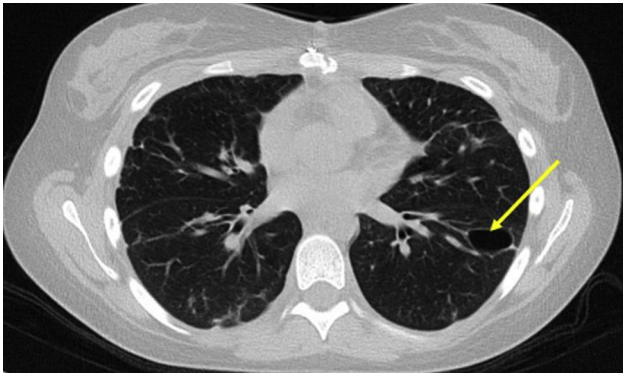

Figure 6 Axial view on chest computed tomography without contrast showing resolution of previously seen lung lesions with scattered areas of parenchymal scarring and a residual cyst (*yellow arrow*) in the left lower lobe.

Abnormal valvar morphologies such as bicuspid or unicuspid PVs may cause predisposition to isolated pulmonary endocarditis.[8] Valve morphology could not be determined intraoperatively, as it was destroyed. Given the absence of a known preexisting murmur in this patient, it can be postulated that the valve was likely normal, although this cannot be known with certainty.

The clinical presentation of these patients can often masquerade as community-acquired pneumonia with fever, elevated inflammatory markers, and respiratory symptoms. The presence of blood cultures growing S aureus and the clinical finding of a new murmur prompted echocardiography, which helped in the diagnosis. Although TTE can be used to diagnose PV endocarditis in the majority of cases (88%), it can be limited by lack of acoustic windows.[9] TEE can be used as an additive diagnostic modality, with successful visualization of pulmonic vegetations.[6,7] This was similar to our experience, in which although a

strong suspicion for PV involvement was raised by TTE, TEE was key in solidifying the diagnosis. Recent studies have used electrocardiographically gated contrast computed tomography to simultaneously assess both cardiac involvement, such as pulmonary vegetations and right-sided dysfunction, and pulmonary involvement, such as septic emboli and pleural effusion.[10]

Medical management involves prolonged intravenous antibiotic therapy, particularly if the microorganism is susceptible, as in the large majority of cases.[3,5] Typically right-sided endocarditis is better tolerated than left-sided endocarditis and is managed conservatively. However, one reason for early surgical consideration is suggested in the presence of S aureus infection given the higher risk for valve destruction and embolic manifestations.[11,12] Indications for surgery in right-sided IE include symptomatic severe valve dysfunction, large vegetations, evidence of persistent infection manifested by persistent bacteremia or fevers lasting >5 to 7 days after the initiation of appropriate antimicrobial therapy, and evidence of septic pulmonary embolism.[13] Valve-sparing surgery when possible should be performed, and if replacement is necessary, a bioprosthetic valve is used. Overall, the outcome of right-sided IE is largely favorable.

CONCLUSION

In this report, we present an unusual case of isolated native PV endocarditis in an otherwise healthy pediatric patient with no identifiable risk factors. Although rare in the pediatric population, a high index of suspicion for this diagnosis should be maintained in a patient presenting with pneumonia-like symptoms and recurrent positive blood cultures. Although transthoracic imaging can typically visualize the PV well, it is important to use adjunctive imaging modalities such as TEE and computed tomography to visualize the vegetations and to assess the cardiopulmonary sequelae of PV endocarditis.
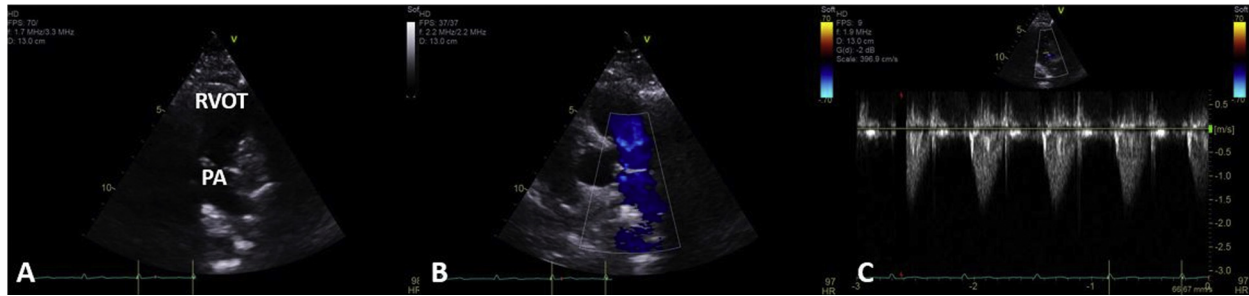
Figure 7 Postoperative TTE. **(A, B)** Parasternal short-axis view demonstrating normal prosthetic PV function. **(C)** Spectral Doppler across the right ventricular outflow tract (RVOT) showing no stenosis or regurgitation. *PA*, Pulmonary artery.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2019.10.004.

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