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IMAGING VIGNETTE

INTERMEDIATE

CLINICAL VIGNETTE

Double-Chambered Right Ventricle Presenting in Adulthood With Aortic and Pulmonic Valve Endocarditis

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ABSTRACT

A 55-year-old woman admitted with symptoms of right heart failure received a diagnosis of double-chambered right ventricle and a ventricular septal defect with aortic and pulmonic valve endocarditis. This case highlights the use of multimodality imaging and importance of adult congenital heart disease expertise in the diagnosis and treatment of such patients. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;19:101934) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 55-year-old woman with childhood diagnoses of pulmonic stenosis and ventricular septal defect (VSD) presented with several months of progressive dyspnea, weight loss, lower extremity edema, and fatigue. She had not undergone prior cardiac interventions and had been lost to congenital cardiology follow-up since her youth, and no pediatric records were available. She did not describe illicit substance use.

On admission to the adult congenital heart disease (ACHD) service, she had a temperature of 100.3 °F, blood pressure of 139/69 mm Hg, heart rate of 98 beats/min, and a 2 L/min oxygen requirement. The results of her cardiovascular examination were significant for a prominent systolic murmur at the left sternal border and jugular venous distension. She had bibasilar rales and extensive lower extremity edema with petechiae. Oral examination revealed palatal petechiae with poor dentition.

The admission laboratory results were notable for elevated brain natriuretic peptide and creatinine, leukocytosis, anemia, and thrombocytopenia, with low haptoglobin and elevated lactate dehydrogenase, which suggested hemolysis. Blood cultures grew *Streptococcus mutans*.

A chest x-ray showed cardiomegaly with right-sided aortic arch, pulmonary edema, and a small right pleural effusion (**Figure 1A**). A transesophageal echocardiogram (TEE) showed an enlarged, severely hypertrophied right ventricle (RV) with severe subpulmonic stenosis in a pattern of double-chambered right ventricle (DCRV) (**Figures 1B and 1C**, Video 1). There was a small perimembranous VSD, and large vegetations were noted on the aortic and pulmonic valves, with moderate aortic regurgitation (**Figure 1D,** Videos 2 to 4). Cardiac magnetic resonance confirmed the diagnosis of severe subinfundibular stenosis resulting in DCRV with proximal RV hypertrophy (**Figure 1E**, Video 5).

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

ACHD = adult congenital heart disease

DCRV = double-chambered right ventricle

RV = right ventricle

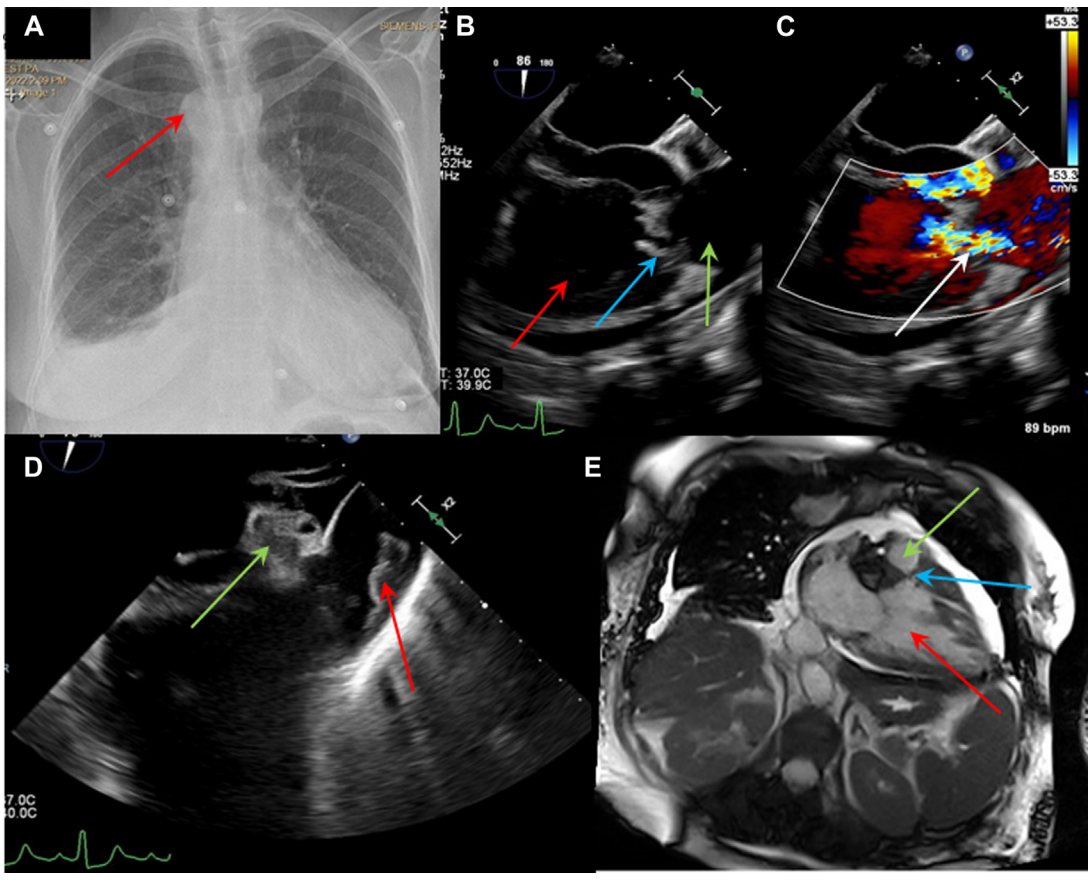
RVOT = right ventricular outflow tract

TEE = transesophageal echocardiogram

VSD = ventricular septal defect

Our patient was treated with intravenous ceftriaxone, and clearance of bacteremia occurred within 48 hours. After an ACHD team discussion, she was scheduled to undergo aortic and pulmonary valve replacements with resection of the RV subinfundibular obstruction and VSD closure. Owing to the need of congenital cardiothoracic surgery expertise, she required transfer to the local children's hospital with a plan for surgery the day after transfer. Unfortunately, on arrival there, the result of her test for COVID-19 was positive, which led to further delay in surgical date, which was ultimately scheduled 2 weeks after diagnosis. However, just before surgery she experienced a sudden onset of headache, chest pain, and cardiogenic shock followed by cardiac arrest. She was resuscitated and underwent cannulation for extracorporeal membrane oxygenation. A repeated echocardiogram showed severely reduced left ventricular function with wall motion abnormalities and severe mitral regurgitation concerning for a coronary event. She unfortunately expired rapidly.

FIGURE 1 Multimodality Imaging of Double-Chambered Right Ventricle and Endocarditis



(A) Chest x-ray showing cardiomegaly with right-sided aortic arch **(red arrow)**, pulmonary edema and small right pleural effusion. **(B, C)** Color transesophageal echocardiogram comparison images show proximal right ventricle high-pressure chamber with hypertrophy **(red arrow)** and subvalvular muscular obstruction **(blue arrow)** with color Doppler of high gradient **(white arrow)** and distal low-pressure chamber **(green arrow)**. **(D)** Transesophageal echocardiogram shows aortic valve vegetation **(green arrow)** and pulmonic valve vegetation extending into pulmonary artery **(red arrow)**. **(E)** Cardiac magnetic resonance shows hypertrophied proximal right ventricle chamber **(red arrow)** with subinfundibular obstruction **(blue arrow)** and distal low-pressure chamber **(green arrow)**.

DISCUSSION

Double-chambered right ventricle is a rare congenital cardiac defect that often develops in adulthood.[1] Multimodality imaging using ACHD expertise is frequently necessary to clearly define anatomy and hemodynamics. Endocarditis in DCRV is rare and requires prompt diagnosis and surgical intervention. Our unusual case in a patient with both left- and right-sided involvement was likely related to shunting from the VSD. The treatment of these complex patients at a center with ACHD expertise is essential to ensure timely, appropriate diagnosis and treatment.

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KEY WORDS congenital heart defect, endocarditis, imaging, ventricular septal defect

APPENDIX For supplemental videos, please see the online version of this paper.