Clinical Image

Double-chambered Right Ventricle after Ventricular Septal Defect Operation Presenting as Syncope

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Abstract

Double-chambered right ventricle (DCRV) is a cardiac disease of the right ventricular outflow tract obstruction characterized by anomalous muscle bundles that divide the right ventricle into two chambers. It may be also develop over time as an acquired lesion in patients with an abnormally short distance between the moderator band and the pulmonary valve. This report highlights the case of a man with double-chambered right ventricle after ventricular septal defect operation, who presented with syncope.

Key words: Double-chambered right ventricle, Ventricular septal defect, Echocardiography, Syncope



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A 32-year-old man was referred to our hospital for evaluation of chest pain on exertion and recurrent syncope. He has undergone ventricular septal defect operation 20 years ago. On physical examination, a prominent systolic ejection murmur was heard on the left parasternal border. Electrocardiogram showed right ventricular hypertrophy, and chest X-ray revealed no cardiomegaly (Fig. 1). Transthoracic echocardiogram showed normal left ventricular function with a protruded muscle band in enlarged right ventricular wall on parasternal short-axis view (Fig. 2A, 2B). Color Doppler showed a turbulent mixed color flow through the stenotic lesion in the right ventricle (Fig. 2C). Continuous-wave Doppler revealed with a flow acceleration of 4.0 m/s (peak pressure gradient of 74 mm Hg) (Fig. 2D). The patient underwent treadmill test. During recovery phase, frequent ventricular premature contraction presented and occurred syncope (Fig. 3). We recommended further evaluation and

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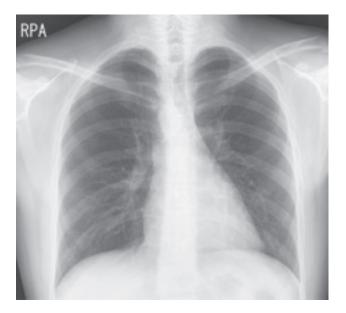


Figure 1: Chest X-ray shows suspected right sided aorta with post median sternotomy.

management. However, the patient refused it. DCRV is a rare cardiac disease of the right ventricular outflow tract obstruction characterized by anomalous muscle bundles.

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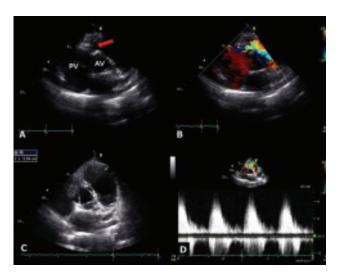


Figure 2: Transthoracic echocardiography shows significant narrowing of the right ventricular cavity (arrow) by abnormal muscle band (A), mixed turbulent flow through the stenotic lesion within the right ventricular cavity on color Doppler imaging (B),post-stenotic dilatation of pulmonary artery (C), midcavitary pressure gradient of 74 mmHg by continuous wave Doppler (D). AV; aortic valve, PV; pulmonary valve.

DCRV is usually diagnosed in childhood or adolescence, with most reported cases in patients less than 20 years old.¹ DCRV is a rarely found as an isolated anomaly. Perimembranous ventricular septal defect (VSD) is the most common coexisting congenital defect.² Other coexisting anomaly including subaortic stenosis, pulmonary valve stenosis, double outlet right ventricle, TOF, anomalous pulmonary venous drainage, complete or corrected transposition of the great arteries, pulmonary atresia with intact ventricular septum and Ebstein anomaly. Hemodynamic state and cardiac morphologic structure changes associated with the unusaual evolution of acquired infundibular stenosis in patient after operative closure of the ventricular septal defect.³ Clinical presentation is depending on the degree of obstruction. The patients may be asymptomatic or present various symptoms, such as exertional dyspnea, syncope, dizziness. Echocardiography is considered a good method for diagnosis of DCRV in adult. ² The management of the DCRV is surgical correction.

Conflict of Interest: None

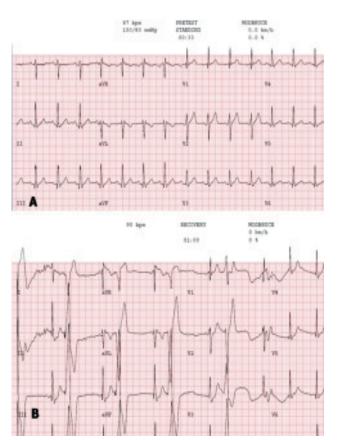


Figure 3: Exercise stress test reveals right ventricular hypertrophy in baseline electrocardiogram (A), frequent premature ventricular contractions occurred in the exercise recovery period within 1 minute after exercise (B).

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