Clinical Medicine Insights: Circulatory, Respiratory and Pulmonary Medicine

SHORT REPORT

A Rare Case of Double-Chambered Right Ventricle Associated with Ventricular Septal Defect and Congenital Absence of the Pulmonary Valve

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Abstract: Double-chambered right ventricle (DCRV) is a rare congenital heart disorder involving 2 different right ventricle (RV) pressure compartments that is often associated with ventricular septal defect (VSD). Usually, the obstruction is caused by an anomalous muscle bundle crossing the RV from the interventricular septum to the RV free wall. We are reporting a case of double-chambered right ventricle associated with ventricular septal defect and congenital absence of the pulmonary valve, a rare form of congenital infundibular pulmonary stenosis. In addition to ventricular septal defect, our patient had congenital absence of the pulmonary valve, which is very unusual and has never been reported to our knowledge.

Keywords: cardiology, magnetic resonance imaging, echocardiography, doubled-chambered right ventricle, ventricular septal defect

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Case Presentation

A 45-year-old female presented to the cardiology office complaining of persistent dyspnea on exertion. She also reported intermittent chest pain along with bilateral lower extremity edema. Symptoms had been present for past 2 years and were becoming progressively worse. Her past medical history was consistent with hypothyroidism. There was no evidence of deep vein thrombosis on lower extremity duplex. A transthoracic echocardiogram showed a normal left ventricular function and a small ventricular septal defect (VSD) (Video 1). Color Doppler analysis using transesophageal echocardiogram revealed turbulence in the proximal portion of right ventricle with a high velocity jet (Image 1A, Video 2). An abnormal muscle bundle was also evident separating the proximal from the distal infundibular chamber of right ventricle (Image 1B and C, Video 2). Measurement of the pressure gradient beteween the proximal and distal chamber was not done since the ultrasound beam could not be aligned with the jet. Magnetic resonance imaging (MRI) confirmed the finding of membranous ventricular septal defect without significant shunting and increased trabecular markings of the subinfundibular region of the right ventricle consistent with double-chambered right ventricle (DCRV) (Image 1D). The pulmonary valve was not seen on echocardiogram or MRI.

Discussion

DCRV is a rare form of congenital infundibular pulmonary stenosis. It is characterized by aberrant hypertrophied muscle bands that divide the right ventricle into 2 cavities with a pressure gradient across. Several subtypes of divided RV have been discribed.[1] Associated defects are present in approximately 80% to 90% of patients. VSD that involves the membranous septum is the most common associated congenital abnormality. Other frequent associated lesions include pulmonary valve stenosis and discrete subaortic stenosis. Vogel et al described 36 patients with membranous VSD and double-chambered right ventricle, 88% of who had echocardiographic evidence of subaortic stenosis, with evidence of progressive left ventricular outflow tract obstruction.[2]

Congenital absence of the pulmonary valve is uncommon and usually associated with VSD and

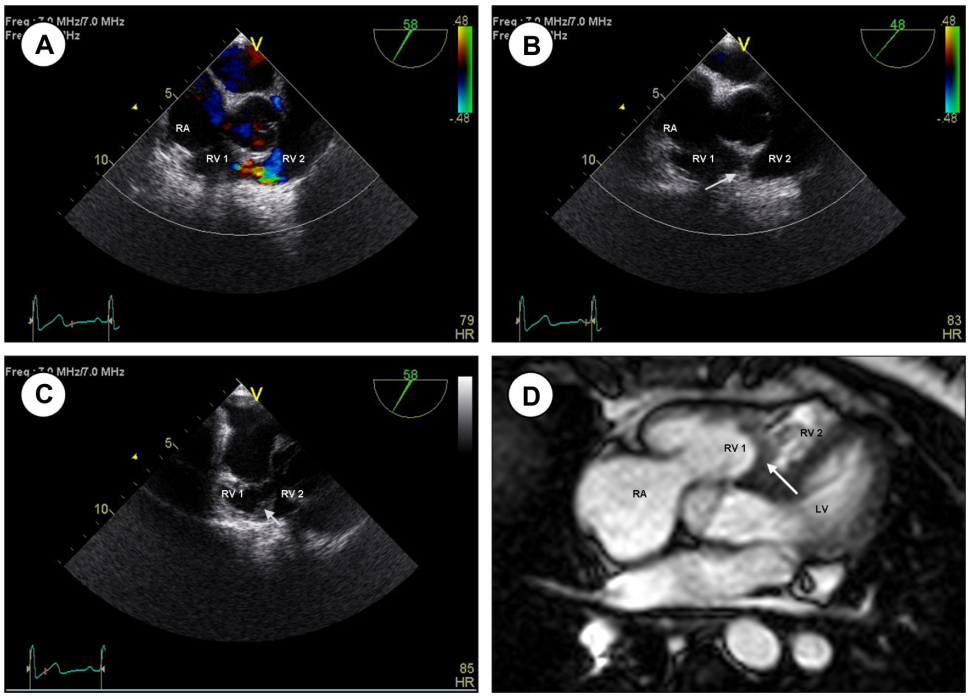

Figure 1. (A) A transesophageal echocardiogram with color doppler (short axis midesophageal view) showing turbulence in the proximal portion of right ventricle with a high velocity jet. (**B**) In the same view, the muscle bands are clearly illustrated (white arrow) and appear to divide the right ventricle into a proximal and distal infundibular chamber. (**C**) A modified transesophageal view also showing the muscle bands (white arrow). (**D**) MRI image (horizontal long axis view) showing increased trabecular markings and anomalous muscle bundles (arrow) dividing the right ventricle into a proximal and a distal chamber.

obstructive subvalvular pulmonary ring but not typically with DCRV.

Author Contributions

Analyzed the data: GK. Wrote the first draft of the manuscript: MM, MA. Contributed to the writing of the manuscript: TB, MT. Agree with manuscript results and conclusions: WH. Jointly developed the structure and arguments for the paper: TB, GK, NAR. Made critical revisions and approved final version: TB. All authors reviewed and approved of the final manuscript.

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Competing Interests

Author(s) disclose no potential conflicts of interest.

Disclosures and Ethics

As a requirement of publication the authors have provided signed confirmation of their compliance with ethical and legal obligations including but not limited to compliance with ICMJE authorship and competing interests guidelines, that the article is neither under consideration for publication nor published elsewhere, of their compliance with legal and ethical guidelines concerning human and animal research participants (if applicable), and that permission has been obtained for reproduction of any copyrighted material. This article was subject to blind, independent, expert peer review. The reviewers reported no competing interests.

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