Review in Translational Cardiology: MicroRNAs and Myocardial Fibrosis in Aortic Valve Stenosis, a Deep Insight on Left Ventricular Remodeling

Addition of Exercise to Dipyridamole Stress Echocardiography in Order to Carry on the Ischemic Cascade: Role in the Diagnosis of Coronary Artery Disease and Prognostic Value

The Diagnostic Challenge of Dipyridamole-atropine Stress Echocardiography in a Patient with Myocardial Bridge

Infected Endocarditis of the Left Main to Right Atrial Coronary Cameral Fistula

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Isolated Double-chambered Right Ventricle with Intact Interventricular Septum

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ABSTRACT

Double-chambered right ventricle (DCRV) is a developmental cardiac anomaly in which anomalous muscle bundles divide the right ventricular (RV) cavity into two chambers. It is usually associated with other congenital cardiac defects, of which ventricular septal defect is the most common association. Isolated DCRV with an intact interventricular septum is very rarely reported. It manifests itself usually in adolescence and adults as the obstruction progresses gradually. It is important to recognize this anomaly as it can progress to severe RV failure if unaddressed. We report a 13-year-old male who presented to us with an isolated DCRV. He successfully underwent a resection of the RV cavity anomalous muscle bundles. We also stress on the utility of transesophageal echocardiography intraoperatively which accurately delineates the location of the muscle bundles and helps in evaluating the adequacy of resection postsurgery.

Key Words: Anomalous, double-chambered, muscle bundle, right ventricle

INTRODUCTION

Double-chambered right ventricle (DCRV) is a rare developmentally acquired heart disease characterized by the division of the right ventricular (RV) cavity into two chambers by anomalous muscle bundles. Typically, DCRV is diagnosed at childhood or adolescence, and most DCRV patients have associated congenital anomalies, such as ventricular septal defect (VSD). Here, we report a case of an isolated DCRV in an adolescent patient with an intact interventricular septum.

CASE REPORT

A 13-year-old boy presented to us with a history of easy fatigability and breathlessness on exertion over 6 months. Physical examination revealed a systolic thrill and an ejection systolic murmur over the left upper parasternal area. Chest X-ray revealed a cardiomegaly with an RV enlargement and two-dimensional transthoracic echocardiography (TTE) findings included a hypertrophied RV and a severe subinfundibular stenosis with a maximum gradient of 100 mmHg across it. The interventricular septum and interatrial septum were intact with no other defects evident on preoperative screening.

Surgical resection was planned. Intraoperative transesophageal echocardiography (TEE) confirmed the preoperative findings [Figures 1, 2a, and 2b].

The maximum gradient across the muscle bundle was calculated to be 65 mmHg which is well above the recommended pressures to be addressed surgically [Figure 3a, and b].

The DCRV bundles were approached trans right atrium (RA) and trans RV under hypothermic cardiopulmonary bypass.

accomplished with aortic and bicaval cannulation. A right ventriculotomy in addition to trans-RA approach was required to completely resect the anomalous muscle bundles. Postresection TEE showed a widely opened up RV cavity with no turbulence or gradient across the mid-cavity and RV outflow tract (RVOT) [Figure 4].

The immediate postoperative period was uneventful with a smooth recovery, and the child was discharged on the 6th postoperative day.

DISCUSSION

DCRV is a developmentally acquired heart disease diagnosed by echocardiography wherein the right ventricle is divided into a proximal high-pressure chamber and a distal low-pressure chamber by anomalous cavity muscle bundles. This condition is almost always associated with other cardiac defects such as VSD, pulmonary valvular stenosis, patent ductus arteriosus, pulmonary atresia, tetralogy of Fallot (TOF), double outlet right ventricle, and rarely transposition of great vessels. The most common among these defects is a VSD which is present in up to 90% of patients with a DCRV. Most of these patients present in early childhood for treatment, but sometimes a patient with isolated DCRV may progress to adulthood without much symptoms till the obstruction becomes more severe. Isolated DCRV with an intact ventricular septum is a very rare occurrence in an adolescent. Several subtypes of divided RV based on the muscle bundles have been described such as anomalous septoparietal band, anomalous apical shelf, hypertrophy of apical trabeculations, anomalous apical shelf with Ebstein's malformation, and sequestration of the outlet portion of the ventricle from a circumferential diaphragm in patients with TOF. Folger described a simple classification of DCRV based on the position of the muscle bundles in the RV cavity. The first group had a low or an oblique partition lower down in the mid-cavity and the second group had a horizontal and a high partition in the subinfundibular portion of RV. Another classification proposed by Galiuto et al. includes a Type 1 DCRV with a mid-cavity anomalous muscle bundle and a Type 2 DCRV with a hypertrophied parietal and septal muscle bands. Isolated DCRV is usually a Type 1 as in our case while Type 2 DCVRs are highly associated with a VSD. Irrespective of the location of the band, the two parts of RV on either side of the bundle possessed a part of the apical ventricular component, with the proximal chamber continuous with the ventricular inlet, and the distal chamber supporting the subpulmonary infundibulum.

A preoperative diagnosis of DCRV is of paramount importance. Anatomical relationships may be difficult to define at operation, and there have been instances of unrecognized DCRV in which transventricular channel was inadvertently closed, mistaken for a VSD, and the patients had a fatal outcome. Failure to recognize and address a DCRV in a patient with a VSD could bring back...
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the patient for a redo cardiac surgery at a subsequent later date. Apparently, features of the DCRV that may be evident intraoperatively on opening the pericardium include a dimpling or a grooving of the RV surface and a systolic thrill over the outflow tract of the right ventricle rather than over the pulmonary artery.[10]

Most cases of DCRV are diagnosed and treated during childhood when they present with associated lesions, and very few reports include cases in which the initial presentation for diagnosis and treatment occurs during adulthood. It is difficult to diagnose DCRV with TTE in adults. According to Oliver et al.,[11] a mild right midventricular obstruction shows a fast rate of progression in adolescents and young adults. In one study, of 32 patients, only five were diagnosed by means of TTE (15.6%).[12] It can be difficult to obtain an image owing to the proximity of the RVOT to the transducer and other limitations encountered while studying adults, such as obesity and emphysema. Therefore, it is critical to carry out echocardiography carefully to ascertain DCRV, especially when it is associated with a VSD or with RV hypertrophy on an ECG. TEE appeals to be a more sensitive tool to pick up a DCRV preoperatively and also intraoperatively to address anomalous RV cavity muscle bundles associated with other lesions like a VSD.[13] Cardiac catheterization delineates the coarsely trabeculated inflow being separated by filling defects from a relatively smooth walled distal chamber and defines the pressure gradients across the muscle bundles. Cardiac magnetic resonance imaging may prove to be the most effective noninvasive means of imaging DCRV.[14]

The nature of pathology in a DCRV entails a tendency toward progressive obstruction by the muscle bundles particularly if associated with a VSD, presumably secondary to both muscle hypertrophy and endocardial fibrosis. Patients can develop severe and varied symptoms, such as easy exhaustion, shortness of breath and syncopal attacks and palpitations secondary to transient arrhythmias. Treatment of DCRV cases is surgical which often constitutes a part of the corrective procedure. In general, the surgical procedures consist of the resection of the anomalous muscular bundle and correction of the associated cardiac anomalies. The period for surgical repair usually depends on the associated cardiac anomalies. In the absence of a significant coexisting defect, observation is possible as long as the intracavitary systolic gradient is not >40 mmHg and the obstruction is not progressive.[15] Although traditionally DCRV has been repaired transventricularly,[16] this operative method may depress the RV function and increase the risk of ventricular arrhythmias. Coates et al.[17] suggested that adequate exposure of the mid-cavity muscle bundle could be obtained through a trans-atrial approach. However, because the RVOT may not be seen clearly, an undiagnosed DCRV may be missed. A complete resection of the bundle may require a right ventriculotomy and myectomy.

Figure 3: (a) Two-dimensional transesophageal echocardiography deep transgastric view with a continuous Doppler demonstrating the pressure gradient across the right ventricular cavity muscle bundle. (b) Transesophageal echocardiography transgastric view at 50° with colour Doppler showing the turbulent flow across the double-chambered right ventricle bundle (white arrowhead).

Figure 4: Two-dimensional transesophageal echocardiography mid esophageal right ventricular inflow-outflow view at 60° with colour Doppler revealing an opened up right ventricular cavity.
as in our case. The immediate postoperative period may be complicated by postoperative arrhythmias and a right bundle branch block secondary to a ventriculotomy. The long-term prognosis for patients after the intracardiac repair of DCRV is excellent.

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**REFERENCES**


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