Case Report

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Repair of isolated double-chambered right ventricle

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ABSTRACT

The finding of a double-chambered right ventricle (DCRV) is exceptionally rare as an isolated anomaly. It is a congenital cardiac anomaly in which the right ventricle is separated into two chambers, a proximal high-pressure chamber and a distal low-pressure chamber, by anomalous muscles or fibrous tissues in the right ventricular cavity. We report the case of a 6-year-old infant who was admitted for growth retardation. The patient was diagnosed with an isolated DCRV without any other associated congenital anomalies. The patient underwent a successful cardiac surgical procedure of enlargement repair; he was discharged in good clinical condition with a normal cardiac function.

Keywords: Enlargement repair, isolated double-chambered right ventricle

INTRODUCTION

Double-chambered right ventricle (DCRV), causing right ventricular outflow tract obstruction, is an uncommon congenital heart disease. Diagnosis can be complicated when it presents with unusual symptoms. The majority of patients have associated congenital anomalies, most commonly a ventricular septal defect or fixed subaorticstenosis. Anisolated DCRV is extremely rare. Here, we report a case of an isolated DCRV that presented with growth retardation in a young infant. The patient underwent a successful cardiac surgical procedure of enlargement repair.

CASE REPORT

A 6-year-old infant was admitted for growth retardation without any cardiac symptoms. On physical examination, a systolic ejection murmur at the left parasternal border was noted, an electrocardiogram (ECG) showed the right ventricular hypertrophy with right axis deviation suggestive of right ventricular overload. The transthoracic echocardiography (TTE) showed a turbulent doppler color flow jet through a stenotic mid-right ventricle and continuous wave (CW) doppler revealed a mid-cavitary pressure gradient of 100 mmHg. There was also amoderatedilatation of pulmonary artery with normal infundibular contraction. In addition, the CT imaging revealed an anomalous muscle bundle. The diagnosis of an isolated DCRVwas confirmed. The infant was taken to the operating room at the age of 6 years, where he was given a general anesthesia and intubated. The repair was done on cardiopulmonary bypass with bicavalcannulation. Through a vertical right ventriculotomy, the muscular obstruction in the roof of the chamber leaves a small circular orifice, covered with fibrous tissue. Resection, initially starting anterior and to the left of the opening, will start to open up the connection with the distal chamber, taking care not to divide the papillary muscles of the tricuspid valve. Alternatively the vertical incision was closed with a patch of pericardium [Figure 1]. Trans-esophageal echocardiography in the operating room shows a good expansion of the RV without residual gradient. Postoperative course was uneventful.

DISCUSSION

Surgical correction of DCRV results in excellent functional and hemodynamic long-term results, with complete relief of the right ventricular obstruction. The presence of a DCRVshould be considered in anomalies with high or persistent right ventricular outflow tract obstruction.[1,2] Two basic morphologic patterns based on angiographic findings have been identified: A high-position abnormal muscle bundle with a horizontal orientation and a low-position muscle bundle with an oblique orientation across the apical component of the right ventricle.[3] In DCRV the infundibular

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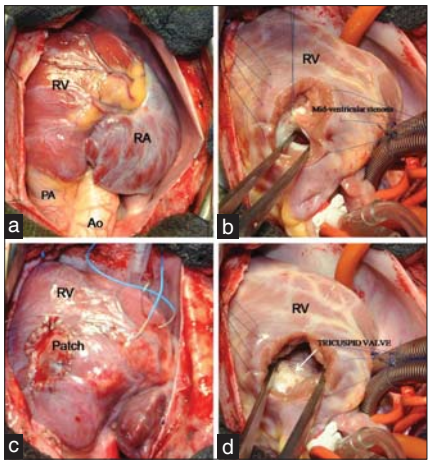


Figure 1: Before ventriculotomy (a). Vertical right ventriculotomy (b). The muscular obstruction in the roof of the chamber leaves a small circular orifice, covered with fibrous tissue (b). After resection (c). The vertical incision was closed with a patch of pericardium (d) LV = Left ventricle, RV = right ventricle, PA = pulmonary artery

septum is not involved.[4] It is morphologically distinct from tetralogy of Fallot in which the infundibulum is hypoplastic due to anterior and superior displacement of the crista supraventricularis.The patients presenting with isolated DCRV may have had a ventricular septal defect (VSD) present at younger age that spontaneously closed from mechanisms such as adherence of tricuspid valve tissue, fibromuscular proliferation adjacent to the anomalous muscle bundles, or hypertrophy of the anomalous muscle bundles and/or ventricular septum.[2] The majority of patients with DCRV have coexisting cardiac lesions: Most commonly it is associated with a membranous type VSD, pulmonary valve stenosis, subaortic stenosis, atrial septal defect, double-outlet right ventricle, and Ebstein's anomaly of the tricuspid valve.[5] It has been considered that the pressure gradient is one of the indications of surgical therapy.[6] A discrepancy between the gradient measured by CW Doppler and the gradient measured by right ventricular catheterization in the case of DCRV is possible.[7] There is often a branch for the right coronary artery thatcourses over the underlying obstruction which can often be relieved via a right atrial approach. The ventricular cavity is explored via the tricuspid valve, entering the proximal chamber or through a transverse right ventriculotomy, immediately above the site of the obstruction and of the coronary branch (if present). In appearance of the muscular obstruction, the small circular orifice between DCRV is very similar to a small ventricular septal defect. Gentle probing of the opening will reveal connection with the distal chamber. Secondary infundibular hypertrophy is a common cause of residual gradient between the right ventricle and the pulmonary artery. It is prudent to close theventriculotomyafter infundibular resection, through apatchof enlargement.

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