

A Rare Cause of Right Heart Failure in An Adult: Congenital Gerbode Defect

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ABSTRACT

Gerbode defect is a rare defect describing a left ventricle to right atrium shunt with symptoms dependent on the size and degree of the shunt. It is either a congenital defect detected in infancy, or an acquired case reported in older age. Diagnosis by means of echocardiography and cardiac magnetic resonance imaging is of paramount importance before surgical correction is contemplated to achieve a good prognosis.

INTRODUCTION

Gerbode defect is a rare type of shunt from the left ventricle to the right atrium. It occurs in one of two forms: congenital or acquired. The acquired form is related to previous cardiac interventions and is more commonly reported than the congenital form, which is exclusively reported in infancy. We report a congenital Gerbode defect in a man in his third decade of life. Proper diagnosis is vital to planning surgical correction, which remains the mainstay of treatment.

CASE REPORT

A 37-year-old man of African origin presented with shortness of breath on exertion for the past year that progressively worsened within the last two months. This was associated with paroxysmal nocturnal dyspnea and orthopnea on one pillow and palpitations. He had no prior medical or surgical history. He had a family history of ischemic heart disease affecting his father in the fifth decade of his life. He was a smoker of one pack daily for 15 years. Examination at the time showed signs of right heart failure with a loud pan systolic murmur at the left sternal edge with a thrill. Chest x-ray showed an enlarged heart. He underwent transthoracic echocardiography (TTE), which showed severe functional tricuspid regurgitation (TR) with a 12 mm left ventricle (LV) to the right atrium (RA) defect and severe left to right shunt (Gerbode defect, a mean

gradient was 43 mmHg and peak gradient was 72 mmHg) (Figure 1A and 1B). (Figure 1) Severe right atrial and right ventricular dilatation with a tricuspid annular plane systolic excursion (TAPSE) was 17mm with pulmonary hypertension of 65 mmHg. There were no other anomalies detected on TTE and no regional wall motion abnormalities. There was mild mitral regurgitation with a normal aortic valve. Transesophageal echocardiography (TEE) (Figures 1C and 1D) confirmed the finding (supra-valvular defect) with no additional anomalies. Cardiac catheterization showed normal coronaries with a dominant right system. Ultrasound abdomen showed bilateral renal parenchymal disease (grade 1) with dilated inferior vena cava and hepatic veins and enlarged liver. Cardiac Magnetic resonance imaging (CMR) showed a mildly dilated left ventricle with fair systolic function. There was a 15 mm membranous defect between LV and RA with significant flow across it (Figure 2A). (Figure 2) It was superior to the septal leaflet of the tricuspid valve with severe TR due to failure of coaptation. There was a significantly dilated right ventricle with mild dysfunction and a severely dilated right atrium. The main pulmonary artery was dilated. Liver function tests showed mild elevation of direct Bilirubin and total Bilirubin with normal liver enzymes. The patient underwent surgery through median sternotomy with the aid of cardiopulmonary bypass and cardioplegic cardiac arrest. Ascending aorta and bicaval cannulation with snares were employed. The right side of the heart was severely dilated. An oblique right atriotomy incision was made, and the defect was seen measuring 1.5*2 cm between the left ventricle to the right atrium close to the septal leaflet of the tricuspid valve and above the coronary sinus (Figure 2B). This defect was closed with autologous pericardium using a running polypropylene suture. Then the tricuspid valve was replaced using a size 31 Magna Ease bioprosthetic valve using interrupted Pledgeted Ticron sutures. The atrial septum was visualized and showed no evidence of patent foramen ovale or atrial septal defect. Oblique atriotomy was closed using two running layers of polypropylene sutures. The patient was weaned from cardiopulmonary bypass with inotropic support. He initially was in junctional rhythm but then reverted to sinus rhythm shortly afterward. TEE post separation showed no evidence of any residual shunt (Figure 1D) and well-functioning prosthesis with no para valvular leak. Postoperatively, the patient was extubated same day and then discharged to the ward day 3 after stopping the inotropic support. He made an uncomplicated recovery and was discharged home on day 8, with therapeutic level of warfarin for three months' duration. In the

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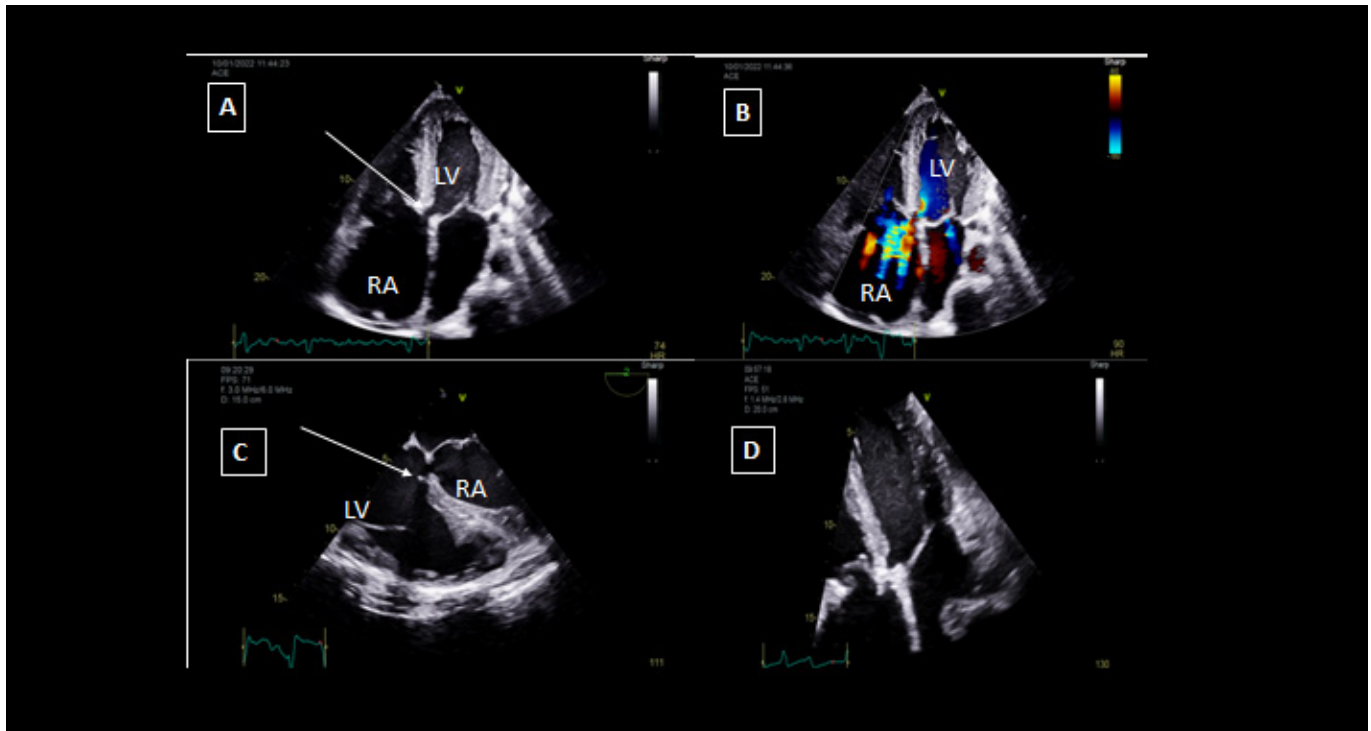


Figure 1. A) 4-chamber view on TTE showing the defect (arrow). B) 4-chamber view on TTE with color Doppler showing flow across the shunt. C) TEE showing the intraventricular septal defect. D) Postoperative TEE showing closure of the shunt with no residual. TTE, transthoracic echocardiography; TEE, transesophageal echocardiography; LV, left ventricle; RA, right atrium

clinic, he was seen three months at follow up. He remained well and was back to all his daily activities and work. TTE at the time showed no residual shunt and a well-functioning tricuspid valve prosthesis with mean gradient of 2 mm HG.

DISCUSSION

Gerbode defect accounts for 0.08% of intra cardiac shunts and <1 % of all congenital heart defects [Saker 2017]. It is defined as abnormal shunt between the left ventricle to right atrium and has been classified as congenital or acquired [Saker 2017; Yuan 2015]. It involves the atrioventricular portion of the interventricular septum [Kelle 2009] and the shunt typically in the membranous part of the septum. It is classified according to its position in relation to the tricuspid valve as supra valvular (one-third of cases) or infra valvular (two-thirds of cases) [Saker 2017; Yuan 2015]. The supra valvular defects (as in our case) are in the atrioventricular membranous septum immediately superior to the septal leaflet of the tricuspid valve and anterior to the coronary sinus. The shunting occurs from the LV to RA, due to large pressure gradient leading to dilated right side of the heart with pulmonary hypertension, and also can lead to enlargement of the left heart chambers, due to increased blood volumes [Saker 2017]. Congenital Gerbode defect usually is diagnosed in infancy [Kelle 2009]. However, it also can occur as an acquired defect in the setting of endocarditis or right

coronary artery infarction or iatrogenic (previous valve surgery or previous percutaneous intervention, such as atrioventricular node ablation) and blunt cardiac trauma [Uslu 2007; Venkatesh 1996]. In our case, the patient was a 37-year-old man when the diagnosis was made.

Symptoms vary from asymptomatic (in small shunts) to severe, including heart failure, endocarditis, and death [Yuan 2015]. Right heart failure symptoms (as in our case) occur with large shunts. Diagnostic modalities include echocardiography [Dzwonczyk 1995; Acar 2011] and cardiac magnetic resonance imaging (CMR). Treatment of Gerbode defect depends on the severity of symptoms. Chronic asymptomatic small defects can be managed conservatively although endocarditis may occur [Yuan 2015]. Surgical correction remains the mainstay of treatment and entails the use of a patch to close the defect from the right atrial side to prevent atrioventricular nodal block [Saker 2017; Yuan 2015; Kelle 2009]. This is performed in combination with tricuspid valve repair with annuloplasty or replacement. Transcatheter closure techniques have been used in high-risk surgical candidates [Saker 2017; Dangol 2013], due to previous cardiac surgical procedures or advanced age, or multiple comorbidities.

CONCLUSION

Gerbode defect is a rare defect causing LV to RA shunt with symptoms dependent on the degree of the shunt. It is usually

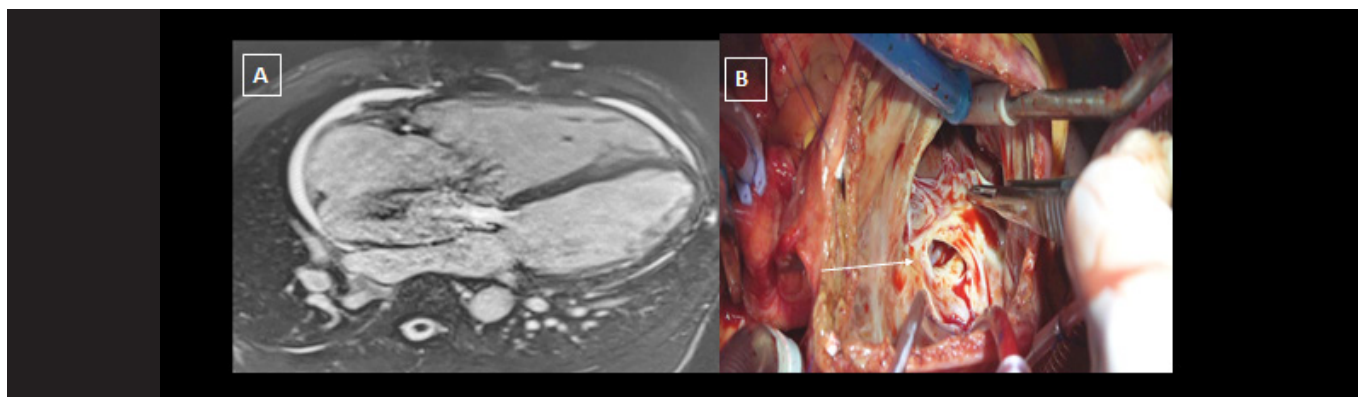


Figure 2. A) 4-chamber view SSFP cine MRI showing the defect between the left ventricle and right atrium. B) Operative view through right atrium showing the defect (arrow) between the left ventricle and right atrium. The defect was superior to the coronary sinus cannula and abutting the tricuspid valve. SSFP, steady-state free precession; MRI, magnetic resonance image

a congenital defect detected in infancy with acquired cases reported in older age. Diagnosing this condition by means of echocardiography and CMR is of paramount importance before surgical correction is contemplated to achieve good prognosis.

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