Gerbode Defect Formation Two Decades after Tetrology of Fallot Repair

Abstract

Although most intracardiac defects are congenital, a small fraction may be acquired during life. The Gerbode defect is an abnormal anatomical connection between the left ventricle and the right atrium. We describe herein a patient who initially underwent repair of tetralogy of Fallot (TOF). Years after TOF repair, he developed severe dyspnea. Extensive evaluation revealed that he had developed a Gerbode defect. Very few cases of acquired Gerbode defect have been previously reported. Management options are predominantly surgical interventions.

Keywords: Gerbode defect, tetralogy of Fallot, ventricular septal defect

Introduction

“The lesion consists of a high ventricular septal defect (VSD) associated with a defect of the septal leaflet of the tricuspid valve which allows left ventricular blood to enter the right atrium.”

— Dr. Frank Gerbode (presenting at the American Surgical Association, New York, April 1958)

The Gerbode defect is an abnormal anatomical communication between the left ventricle (LV) and the right atrium (RA). Gerbode et al. published a case series of five patients with this surgically correctable defect.[1] Gerbode defects comprise of <1% of all congenital defects and around 0.08% of all intracardiac shunts.[2]

Patient Presentation

A 71-year-old man presented with complaints of shortness of breath and bilateral leg swelling. The shortness of breath started 1 week previously and had been progressively getting worse, accompanied by orthopnea. He also noticed the development of swelling over both feet. He did not describe any chest pain, palpitations, cough, fever, or chills. Medical history included essential hypertension, atrial fibrillation, and Tetralogy of Fallot (TOF) surgically corrected 27 years previously. Medications included digoxin and warfarin for atrial fibrillation.

Physical examination revealed distended neck veins and bilateral pedal edema. Palpation of the precordium revealed a thrill over the left sternal accompanied by pulsatile hepatomegaly. Cardiac auscultation revealed a nonradiating holosystolic grade IV blowing murmur at the left sternal border. Dynamic auscultation resulted in a decrease in murmur intensity with inspiration.

Routine blood chemistry determinations were within normal limits. Chest roentgenogram demonstrated increased vascular markings and trace bilateral pleural effusions. Sternotomy wires were observed along with moderate cardiomegaly. A 12-lead electrocardiogram revealed marked right-axis deviation. Atrial fibrillation with a right bundle branch block was also evident.

Computed tomography angiography of the chest demonstrated marked RA dilatation (calculated volume 340 mL), right ventricular (RV) hypertrophy (calculated volume 211 mL), and narrowing of RV outflow tract. There was a 4–5 mm defect between the distal left ventricular outflow tract (below the right coronary cusp) and the right ventricle. The pulmonic valve was deformed with mild narrowing of the RV outflow tract. No evidence of anomalous pulmonary venous return was noted. The great veins, superior vena cava (SVC), inferior vena cava (IVC), and the coronary sinus were dilated.

Transthoracic echocardiogram revealed an ejection fraction of 51%–55% by visual 

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estimation. RV pressure–volume overload was present with abnormal (paradoxic) septal motion consistent with postoperative status. LV filling pressures were normal. There was malalignment of the VSD with a repair patch noted. A ventriculoatrial septal defect with a left to right shunt was found [Figure 1].

A small residual restrictive VSD with a left to right shunt was present too (VSD velocity 5.0 m/s). Severe pulmonic valve regurgitation was noticed. Mild subpulmonic and pulmonic valvular stenosis was present with a peak gradient of about 25 mmHg. The estimated RV systolic pressure was moderately elevated at 56.0 mmHg, with a normal pulmonary artery pressure of 31 mmHg. The IVC was dilated, with respiratory size variation less than 50% [Videos 1 and 2].

**Discussion**

The Gerbode defect is an abnormal anatomical communication between the LV and the RA.[1] Gerbode defects comprise of <1% of all congenital defects and approximately 0.08% of all intracardiac shunts.[2]

The direct type of Gerbode defect involves the membranous septum above the tricuspid valve between the LV and the RA; the indirect type has a VSD with concomitant tricuspid regurgitation. Thus, the blood is shunted indirectly from LV to RV through the VSD, and then from RV to RA through the defective tricuspid valve. The direct form of the defect is less frequent than the indirect.

A large systolic pressure gradient exists between the LV and the RA which creates a high-velocity systolic flow from LV to RA. A characteristic finding is a high Doppler gradient between the respective chambers. This results in blood shunting between the two chambers. Increased blood flow to the RA results in high flow to the RV, which subsequently dilates. RV hypertrophy and raised RA pressure suggest the presence of pulmonary artery hypertension. In cases where the shunt is not closed in a timely manner, the left heart starts to enlarge as well. This causes a decline in global cardiac function.

As a result of technological advances in the past decade, the number of invasive procedures on the heart has remarkably increased. This has resulted in an increased incidence of acquired Gerbode defects (AGD), although AGDs are rare and almost always occur from invasive procedures performed near the membranous AV septum. AGDs are classified into iatrogenic and noniatrogenic; 68% occur in men.[3] Acquired iatrogenic Gerbode defects are most commonly caused by previous cardiac surgery or percutaneous cardiac interventions. Noniatrogenic causes include blunt cardiac trauma, endocarditis, and myocardial infarction in the right coronary artery perfusion domain.

Physical examination may reveal signs of right heart failure. A prominent auscultatory finding in Gerbode defect is a harsh holosystolic murmur, unchanged with respiration. A thrill may also be palpated along the left sternal border. It is often challenging to distinguish the holosystolic murmurs of a Gerbode defect from that of a VSD. Vogelpoel et al. stipulate that VSD murmurs have a higher pitch and an intensity that decreases during inspiration.[4]

In our patient, a meticulous physical examination pointed toward the diagnosis of right heart failure. Since there are myriad causes of right heart failure, a transesophageal echocardiogram (TEE) is essential to make the diagnosis of Gerbode defect. A two-dimensional TEE is the gold standard diagnostic study for an LV–RA shunt. Differential diagnosis includes ruptured sinus of Valsalva. Cardiac catheterization can be used to confirm the diagnosis. An increase in oxygen saturation from SVC to RA is typically found. Left ventriculography demonstrates opacification of a dilated RA prior to the RV. Symptom severity is the main factor in guiding treatment. It depends on the magnitude of the shunt, the volume of blood flow, comorbidities, and preexisting cardiac defects. Asymptomatic or longstanding defects can be managed conservatively. Defects causing severe dyspnea need emergent surgical repair.

**Clinical Outcome**

Surgical correction of the Gerbode defect was recommended for this patient. He is currently being evaluated for corrective heart surgery. In our case, the Gerbode defect was most likely iatrogenic due to TOF repair.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.
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Conflicts of interest

There are no conflicts of interest.

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