

An acquired Gerbode defect from the left ventricle to the coronary sinus following mitral valve replacement

Richard A Collis¹, Jonathan Afoke¹, Christopher GA McGregor¹

Institution:

1. Institute of Cardiovascular Science, University College London

Corresponding Author:

1. Professor Christopher McGregor, UCL, 5 University Street, London, WC1E 6JF, United Kingdom. E-mail: c.mcgregor@ucl.ac.uk

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Abstract

We report the management of an acquired Gerbode defect, from the left ventricle to the coronary sinus following mechanical mitral valve replacement (MVR). Following a failed percutaneous closure, surgical patch closure of the defect was performed.

Introduction

A shunt from the left ventricle (LV) to the right atrium (RA) was first described by Gerbode in 1958.¹ This defect is now primarily seen in patients after valve surgery.^{2,3,4} We present an unusual case of an acquired Gerbode defect from the LV to the coronary sinus producing cardiac failure following mitral valve replacement (MVR) for the management of left ventricular outflow tract obstruction (LVOTO) in hypertrophic obstructive cardiomyopathy (HOCM).

Patient Profile

The patient was a 73-year-old male who had undergone a right hemicolectomy in 2013 for non-metastatic Duke's A cecal adenocarcinoma followed by chemotherapy and a stroke in 2007.

His symptoms consisted of exertional dyspnea and chest pain. Coronary angiography was unremarkable. Transthoracic echocardiography (TTE) displayed asymmetric left ventricular hypertrophy (18mm), an ejection fraction of 65% and a resting LVOT gradient of 71mmHg (Figure 1). Systolic anterior motion (SAM) of the mitral valve with moderate mitral regurgitation (MR) was noted. Transesophageal echocardiography (TEE) showed a severely regurgitant myxomatous mitral valve with multiple jets of MR. In view of the presence of only mild-moderate septal hypertrophy on TEE and the presence of myxomatous disease of both leaflets, an MVR was considered the most appropriate option in this patient with HOCM.

Surgical Technique

He had a MVR in February, 2014. After median sternotomy and bicaval cannulation, a standard posterior left atriotomy and MVR was performed on cardiopulmonary bypass (130 minutes) with systemic cooling to 33 degrees (°C) and the use of antegrade cold blood cardioplegia. The cardiac tissues were friable, perhaps related to the recent chemotherapy.

The anterior mitral valve leaflet was thickened and calcified. There was bileaflet billowing and an irregular coaptation line with an attenuated posterior leaflet. The mitral valve anterior leaflet was excised entirely. The posterior leaflet was left intact. A 31mm mechanical prosthesis (St Jude, Inc.) was placed using interrupted sutures with an aortic cross clamp time of 105 minutes. After bypass, an intraoperative TEE showed a well seated and functioning prosthesis. The initial post-operative recovery was uneventful with extubation on the day of surgery. Subsequently, the patient deteriorated requiring haemofiltration, inotropic support and reintubation on the sixth postoperative day. TTE (Figure 2) showed a well seated and functioning mitral valve prosthesis with a broad turbulent jet originating inferior to the mitral valve to the free wall of the right atrium. The maximum LVOT gradient was 17mmHg. The patient improved and was extubated after 10 days, but had recurrent pleural effusions despite diuretic therapy. A cardiac magnetic resonance (CMR) scan (Figure 3) showed restrictive (high velocity) flow through the membranous septum into the right atrium along the tricuspid valve with what appeared to be a significant left to right shunt. The O₂ saturations increased from 63% in right atrium to 68% in pulmonary artery on right heart catheterization. Attempted percutaneous closure was carried out via a femoral approach. The defect was identified and a sizer balloon was introduced into the defect with sudden elevation of systemic pressures. At this point it was evident that the communication was between the LV and the coronary sinus which drained into the RA (Figure 4). Closure of the defect was attempted with occluder devices but they were found to be unstable and obstructing the MVR leaflet. At this point it was necessary to proceed with surgical intervention.

The patient had a redo sternotomy at six weeks post initial surgery, with aortic cannulation, and direct cannulation of the superior vena cava and cannulation of the inferior vena cava via the femoral vein. The aorta was cross clamped for 35 minutes using cold blood cardioplegia. A transverse right atriotomy enabled direct visualisation of the defect entering into the coronary sinus 1cm from its orifice in the RA. The defect was 1x1.4cm in size with

well-defined and firm edges. Patch repair was done using an autologous pericardial patch and a continuous 5-0 Prolene suture. After cardiopulmonary bypass (85 minutes), TEE showed the shunt to be completely closed. The patient was extubated on the first postoperative day and discharged home 11 days later. Postoperative TTE showed a small LV cavity, good biventricular function, a well seated mitral prosthesis, with no LVOTO and no left to right shunt (Figure 5).

The patient continues to have good symptomatic and functional improvement two years postoperatively.

Discussion

Acquired Gerbode defects can occur due to septal injury or weakening of the tissue following infection, necrosis or trauma. Although rare, the majority of acquired Gerbode defects are iatrogenic in nature following aortic and mitral valve replacement surgery. Less commonly cases of defects are reported in infective endocarditis, post myocardial infarction or trauma.² Patients may present acutely with heart failure or cardiogenic shock depending on the shunt size. Three types of Gerbode defects have been described based on the location of the shunt to the tricuspid valve (TV) anatomically. A type 1 defect, the most prevalent, is located above the TV in the region of the membranous ventricular septum. A type 2 defect is below the TV with type 3, a mixture of type 1 and 2 at the level of the TV itself.^{2,3} Awareness of this complication aids in timely diagnosis, as imaging can be difficult to interpret in postoperative patients in the presence of tricuspid regurgitation. The value of percutaneous closure of such defects is, as yet, uncertain and has been associated with significant procedural complication rates.² As these patients can be high risk for surgery this was thought to provide a less invasive alternative to surgical repair. Certain closure devices however can interfere with prosthetic valve leaflet function as seen in this case.⁴ Surgical correction is more commonly used in non-iatrogenic acquired cases such as infective endocarditis, post

MI and trauma.² In this case, the recent chemotherapy for adenocarcinoma may have contributed to the friability of tissues encountered at surgery and predisposed to the development of the defect. The attempt at balloon inflation may also have increased the shunt requiring urgent surgical reintervention. This case illustrates a new variant of an acquired Gerbode defect that can be treated effectively by surgical re-intervention with direct visualisation of the defect and patch repair.

References

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Figure Legend

Figure 1: Preoperative parasternal long axis TTE image illustrating mild asymmetric septal hypertrophy with an angulated septum and a thickened calcified anterior mitral valve leaflet with SAM.

Figure 2: Apical TTE image illustrating a basal septal defect LV to RA. The arrow points to color flow across the defect from LV to RA.

Figure 3: CMR 4-Chamber view. The arrow points to a basal interventricular septal defect below the level of the mitral valve.

Figure 4: Fluoroscopy image of inflated sizer balloon across the defect. The arrow points to contrast injection into the coronary sinus from the RA.

Figure 5: Apical postoperative TTE view with absence of left to right flow.

Figures



Figure 1

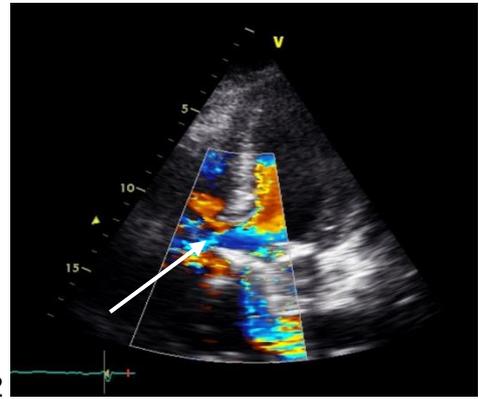


Figure 2



Figure 3

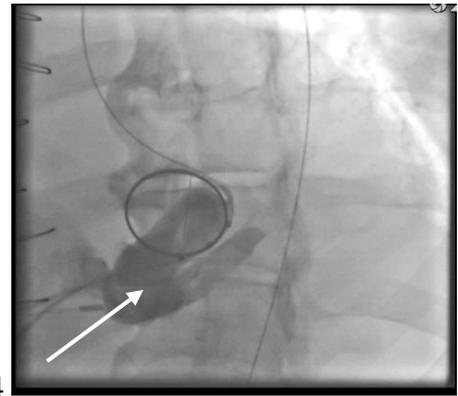


Figure 4

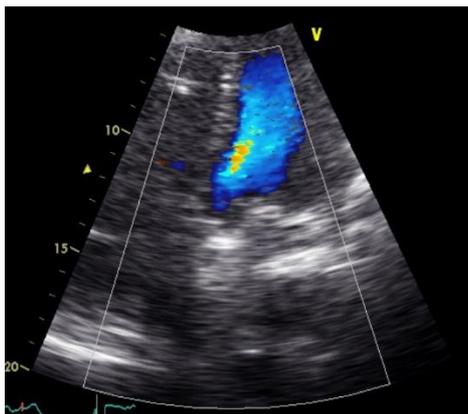


Figure 5