Plugging the Hole: Diagnosis and Management of Post–Myocardial Infarction Ventricular Septal Defect

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INTRODUCTION

Mechanical complications after myocardial infarction (MI) pose a significant management dilemma. Ventricular septal defect (VSD) usually occurs 3 to 5 days following the infarction, with advanced age and female gender being risk factors. The rupture usually occurs at the margin of the necrotic tissue and healthy myocardium. In cases of anterior wall MI involving the left anterior descending coronary artery, the VSD may be located in the mid or apex of the ventricular septum. In cases of right coronary artery or left circumflex coronary artery MI, the VSD is most commonly located in the basal part of the septum, but it can also be found in the mid or less often close to the apex of the septum.

Patients may present with dyspnea, hypotension, or shock, and on physical examination a harsh systolic murmur is present, which may be accompanied by a thrill. Echocardiography, both transthoracic and transesophageal, can demonstrate the defect, its location and size, and its hemodynamic significance. Although current guidelines recommend urgent surgery, surgical mortality is very high.

We present a case of a large, mid-to-apical post-MI VSD, which was initially managed with venoarterial (VA) extracorporeal membrane oxygenation (ECMO) and percutaneous closure, followed by salvage open surgery. The role of echocardiography is highlighted throughout the patient's course.

CASE PRESENTATION

A 68-year-old woman with a medical history of hypertension, hyperlipidemia, and prediabetes presented to a community hospital with vomiting and jaw pain for 36 hours. At baseline, she was active and swam 1 mile three times a week. On presentation she was afebrile and had normal vital signs. Electrocardiography showed inferior-posterior ST-segment elevations with q waves, consistent with a late-presenting MI (Figure 1). The first troponin level was significantly elevated at 19.72 ng/mL. She was transferred to another hospital for cardiac catheterization, which revealed a thrombotic occlusion of the right coronary artery. This was treated with aspiration thrombectomy and a drug-eluting stent. She had residual mid left anterior descending (80%) and mid circumflex (80%) disease, which was not revascularized (Video 1).

In the coronary care unit, the patient reported ongoing chest and jaw pain. Her troponin peaked at 92.39 ng/mL after intervention. Twelve hours after intervention, she became hypotensive, requiring vasopressor support. Repeat electrocardiography showed the expected evolution of her inferior-posterior MI, and her troponin level trended downward.

Transthoracic echocardiography (TTE) demonstrated a VSD along the midportion of the septum, closer to the apex, with left-to-right shunting (Figures 2A and 2B, Video 2). Left ventricular (LV) systolic function was mildly reduced with inferior-posterior akinesis, and right ventricular (RV) systolic function was moderately reduced. Right heart catheterization confirmed significant left-to-right shunting (Qp/Qs = 3).

An intra-aortic balloon pump was placed as a temporizing measure, and the patient was maintained on inotropic support. Because of the VSD location, RV infarction, and the friability of the myocardium after infarction, it was thought that immediate open surgical repair would carry an extremely high mortality risk. The patient was placed on peripheral VA ECMO as a bridge to repair.

After VA ECMO was initiated, the patient was weaned off vasopressors. However, repeated attempts to wean ECMO support were unsuccessful. Five days after ECMO initiation, percutaneous VSD closure was attempted, as the patient was believed to be extremely high risk for open repair.

By transesophageal echocardiography (TEE), the VSD measured 26 mm at its largest diameter, with an area of 4.2 cm^2 on three-dimensional reconstruction. Because of the defect size, a 30-mm atrial septal defect occluder was used, as the largest VSD occluder available was too small. The VSD was crossed with a glide wire from a catheter in the left ventricle and advanced across the defect through the right ventricle and out the pulmonary artery, where it was snared and externalized out the femoral vein. A delivery sheath was then advanced over the glide wire across the VSD from the femoral vein, allowing a 30-mm Amplatzer atrial septal defect occluder to be deployed (Figure 3, Video 3). After deployment of the occluder, there was only minimal residual shunt by echocardiography. Because of the location of the VSD in the mid-to-apical septum, there was no interference with the mitral, aortic, or tricuspid valves (Figure 4, Video 4). The mixed venous saturation improved from 90% before closure to 71% after closure.

During the procedure, dense echocardiographic contrast was noted in the aortic root and ascending aorta, with minimal opening of the aortic valve (AV) during systole and aortic regurgitation was present throughout the cardiac cycle (Figure 5, Video 5). These findings were thought to be a result of blood stasis from poor LV output due to significant left-to-right shunting and increased afterload due to retrograde aortic flow from VA ECMO.

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VIDEO HIGHLIGHTS

Video 1: Coronary angiography showing acute thrombotic occlusion of the proximal right coronary artery, treated with a drug-eluting stent with restoration of flow. There is residual left anterior descending coronary artery and left circumflex coronary artery disease.

Video 2: The first half of the video shows subcostal views obtained by TTE with and without color, demonstrating a VSD along the mid-to-apical portion of the septum with left-to-right shunting. The second half of the video shows transgastric short-axis views on TEE at the level of the left ventricle with and without color, again demonstrating a large VSD. LV systolic function appears preserved, while RV systolic function appears moderately reduced.

Video 3: Transgastric short-axis views obtained by TEE, during echocardiographic guidance of percutaneous closure of the VSD with an Amplatzer atrial septal defect occluder. A well-seated closure device is demonstrated between the left and right ventricles by both two- and three-dimensional imaging, and no significant residual shunt is shown by color Doppler.

Video 4: Midesophageal long-axis view by TEE showing the atrial septal defect occluder in the mid-to-apical septum, with no interaction with the tricuspid valve, mitral valve, or AV.

Video 5: Midesophageal long-axis view by TEE, with the patient on 2 L ECMO support. Dense echocardiographic contrast is demonstrated in the aortic root and ascending aorta, with minimal opening of the AV, because of poor LV output. There is continuous aortic regurgitation throughout the cardiac cycle by color Doppler.

Video 6: Midesophageal long-axis view by TEE with weaning of ECMO support the day after the closure device was placed. The video shows clearance of the echocardiographic contrast in the aorta and improved opening of the AV, due to a decrease in afterload and increase in LV output.

Video 7: Combined transgastric short-axis view by TEE and apical four-chamber view by TTE demonstrating migration of the closure device toward the right ventricle. Color flow demonstrates increased shunt across the device. A small pericardial effusion is also present (TTE).

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Because of concerns of blood stasis and possible thrombus, anticoagulation was intensified, and VA ECMO was maintained at 4.5 L/min. TEE the following day showed that with weaning of ECMO support, the AV opened, allowing forward flow and clearing the echocardiographic contrast (Figure 6, Video 6). There was also slightly increased flow across the occluder, though the mixed venous saturation remained stable. Over the next few days, the mixed venous saturation began to rise again. Repeat echocardiography showed partial migration of the LV disk of the occluder into the right ventricle, with significant shunting across the VSD (Figure 7, Video 7).

After 16 days on ECMO support, the patient continued to have minimal improvement, and the Amplatzer occluder was failing. Salvage open VSD repair was then performed with a bovine pericardial patch. The Amplatzer occluder was removed, and after successful patching, there was no residual shunt on TEE. However, the patient developed significant postoperative bleeding due to significant coagulopathy, followed by multisystem organ failure. Despite massive transfusions, the patient passed away on postoperative day 2.

DISCUSSION

In the current era of widely available catheter-based reperfusion therapies for MI, the incidence of mechanical complications has significantly decreased. However, when post-MI mechanical complications occur, the mortality remains as high as 30 years ago.[1] In the case of VSD, patients with RV involvement have worse outcomes.[2] Our case highlights the challenges of managing these extremely ill patients and offers insights into the critical role of echocardiography in their management.

Echocardiography is the test of choice in patients with hemodynamic instability and cardiogenic shock after MI because of its widespread availability, detailed assessment, and rapid interpretability.[3] Mechanical complications such as postinfarction VSD should be suspected in all patients who develop hemodynamic instability after successful coronary revascularization and prompt urgent echocardiographic evaluation, as fast recognition is essential to guide further management.[4] TTE can establish the anatomic location of the VSD, calculate the shunt fraction, and evaluate biventricular and valvular function, which can aid in identifying patients who are at extremely high risk for poor outcomes. Although two-dimensional TTE is a valuable tool for initial diagnosis, the use of TEE allows a more comprehensive assessment of the morphology and size of the shunt and can evaluate eventual postsurgical residual shunt. However, two-dimensional TEE often underestimates the size of the defect because of the nonparallel orientation of the ultrasound beam to the long axis of the cardiac defect. This limitation can be avoided with the use of three-dimensional TEE, which can reconstruct the full extent of the defect in three dimensions. Three-dimensional TTE or TEE can further elucidate size, location, and morphology of the VSD and may assist in the determination of the appropriateness of a percutaneous approach and in guiding the procedure, such as was used in this case.[5]

Because of the low incidence and high mortality of postinfarction VSD, guidelines are based largely on expert consensus and data from small observational studies. Current American College of Cardiology/American Heart Association guidelines recommend emergent surgical repair in all cases of post-MI VSD, though they acknowledge high operative mortality rates up to 87%.[6] Percutaneous closure is mentioned as a possible, though unstudied, option in appropriate patients.

There are several anatomic considerations in selecting patients for a percutaneous defect closure, and echocardiography can be extremely helpful in the evaluation and decision-making process. Most authors suggest that a defect <15 mm is optimal for percutaneous closure, because of the device sizes that are available, as well as the size of the septum. Extremely apical defects are especially challenging, as they frequently lack an adequate tissue "rim" to secure the device, and alternative closure methods such as exclusion of the right ventricle may be needed.[7] Conversely, the location of the tricuspid valve apparatus (especially the septal leaflet) can make closure of basal defects more challenging. Serpiginous defects are not only difficult to

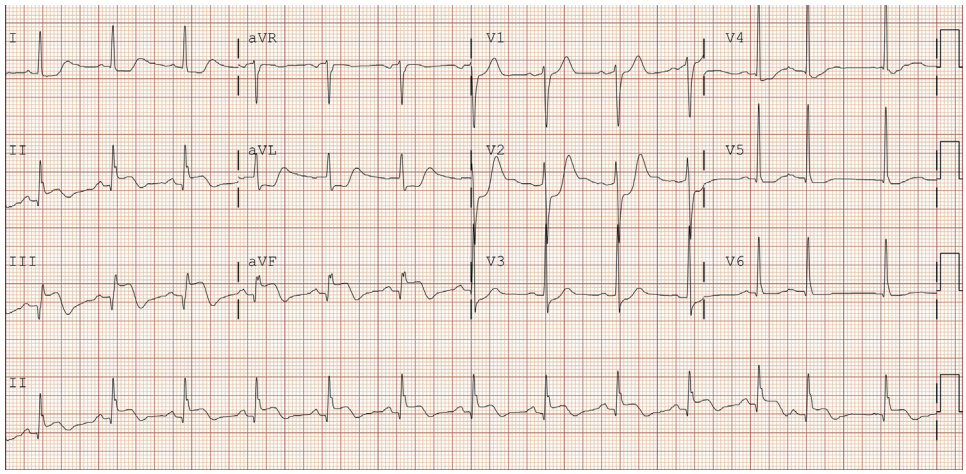

Figure 1 Electrocardiogram upon patient's presentation, demonstrating an inferior-posterior MI.

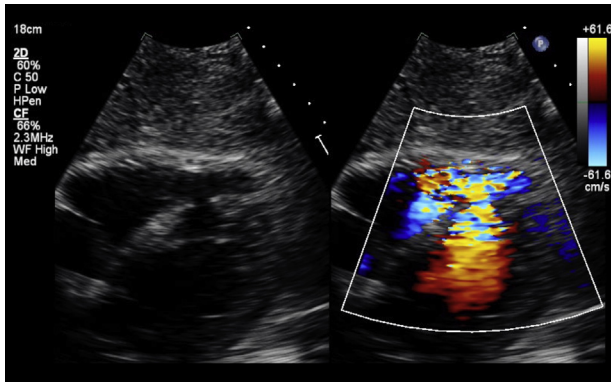

Figure 2 Subcostal view on TTE, without and with color Doppler (B), demonstrating a large VSD along the mid-to-apical portion of the septum, with left-to-right shunt.

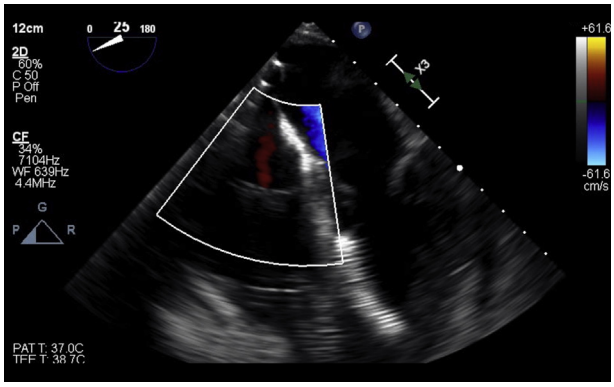

Figure 4 Midesophageal long-axis view by TEE showing the atrial septal defect occluder in the mid-to-apical septum with no interaction with the tricuspid valve, mitral valve, or AV.

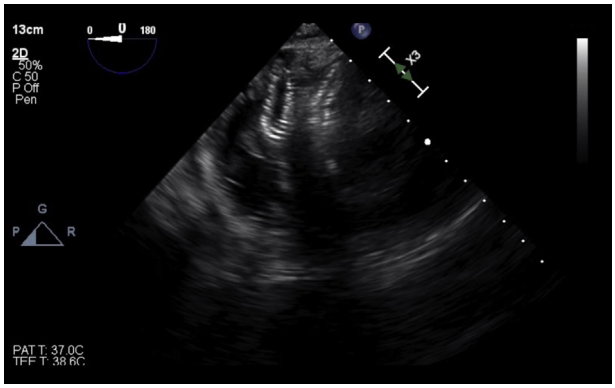

Figure 3 Transgastric view, short-axis window at the level of the left ventricle by TEE, demonstrating a well-seated closure device between the left and right ventricles.

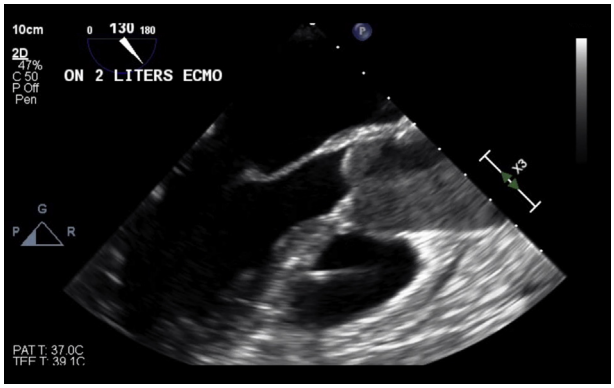

Figure 5 Midesophageal long-axis window by TEE, demonstrating dense echocardiographic contrast in the aortic root and ascending aorta, on 2 L ECMO support.

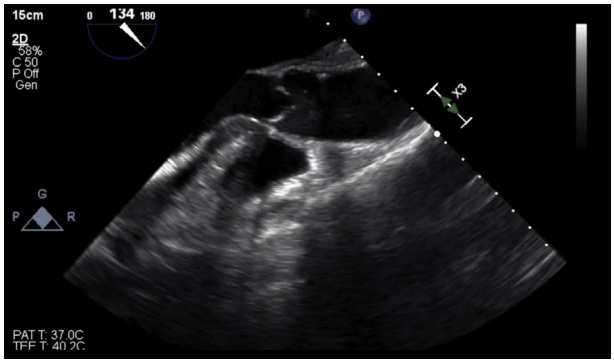

Figure 6 Midesophageal long-axis window by TEE, 1 day after device placement and with ECMO support wean, demonstrating clearing of the echocardiographic contrast from the ascending aorta.

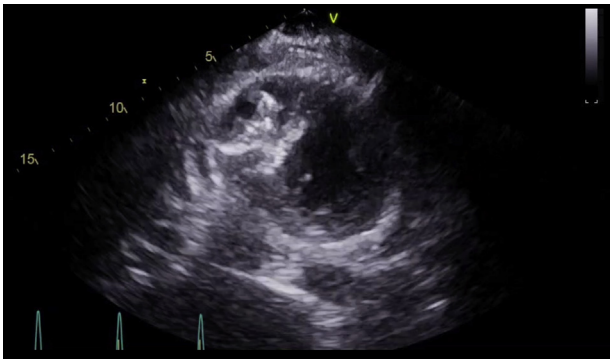

Figure 7 Transgastric short-axis window at the level of the left ventricle by TEE, demonstrating migration of the closure device toward the right ventricle.

cross with a wire but can also be complicated by significant leakage around the device and are associated with an increased risk for device embolization. Thus, attempting percutaneous closure of post-MI VSD requires a thorough understanding of the defect's size, shape, and borders, usually requiring characterization by both surface and TEE.

Early surgery is often complicated by the friability of the necrotic myocardium around the VSD, which makes it difficult to adequately anchor patch repairs and increases the risk for recurrent VSD and subsequent mortality.[7] VA ECMO has been reported as a bridge to percutaneous closure and surgical repair.[8,9] Percutaneous VSD closure may be optimal in smaller VSDs (≤15 mm), though larger defects, such as the one in this case, have been successfully repaired.[10] Some operators prefer to advance the occluder from the venous side via the internal jugular vein, as there may be less tension on the friable VSD borders through this method.

To our knowledge, this is the first reported case in which all three techniques were used. Additionally, though percutaneous closure was not intended as a bridge to surgery, it provided time for myocardial remodeling, improving the technical success of operative repair.

VA ECMO, as opposed to percutaneous LV assist devices such as Impella, was chosen in this case because of the significant RV failure, which would have been exacerbated by placement of an LV assist device. However, the increased afterload caused by the retrograde aortic flow from the ECMO femoral artery cannula resulted in minimal opening of the AV during systole (Video 4). This resulted in stasis of blood flow at the aortic root, appearing as spontaneous echocardiographic contrast in the aortic root.

Aortic root thrombi have been reported in patients on VA ECMO because of LV nonejection and stasis in the root.[11] Patients with VSDs on VA ECMO are likely at greater risk for aortic root thrombus because of pressure gradients favoring increased flow through the VSD rather than the AV, with potentially devastating embolic complications, including stroke. This complication can be recognized promptly by echocardiography. The presence of spontaneous echocardiographic contrast in the setting of VA ECMO has been associated with an increased risk for intracardiac thrombus and stroke in one observational study of 98 patients on VA ECMO, though not necessarily in the aortic root.[12] Patients with spontaneous echocardiographic contrast had a lower ejection fraction and a lower pulsatility index. In the setting of VSD and spontaneous echocardiographic contrast in the aortic root, the most important factor in preventing stasis and thrombus formation in the aortic root is forward flow through the AV.

Importantly, the presence of spontaneous echocardiographic contrast in the aortic root does not equal the presence of thrombus. There are no existing societal guidelines on how to manage these patients with spontaneous aortic root echocardiographic contrast on VA ECMO, and our management is based on evidence from case series and expert opinion. Anticoagulation should be optimized and intensified, and the likelihood of true thrombus on the valve and aortic root should be assessed through echocardiography and clinical factors, such as poor pulsatility or insufficient anticoagulation. If the likelihood of thrombus is low, such as in our case, pulsatility should be restored by lowering the ECMO flow. This will increase flow through the AV and dissipate the spontaneous echocardiographic contrast within a few beats (Video 5). If thrombus is identified on the valve apparatus or in the root, afterload should be increased to maintain persistent AV closure, and the patient should be sent for surgical embolectomy when feasible to avoid systemic embolism. In appropriate patients, Impella along with VA ECMO ("EcPella") can be considered early on in the clinical course to avoid stasis and thrombosis in the ascending aorta.

CONCLUSION

VSD is a rare but often fatal complication after MI. Urgent surgical repair is recommended but may be complicated by recurrent VSD and high mortality rates. Combined VA ECMO and percutaneous closure, especially in patients with concurrent RV involvement, represents a possible strategy in select patients at very high operative risk but presents its own difficulties and complications, such as aortic root stasis. Echocardiography plays a key role in the diagnosis and management of these patients.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.04.001.

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