Mechanical Hemodynamic Support as a Bridge to Recovery in Severe Takotsubo Cardiomyopathy with Marked Left Ventricular Outflow Tract Obstruction and Cardiogenic Shock

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Introduction

Takotsubo Cardiomyopathy (TC) is an acute syndrome of transient left ventricular (LV) dysfunction, presenting with chest symptoms, electrocardiographic changes, and cardiac biomarker elevations mimicking acute coronary syndrome. In the typical variant, this condition results in regional dysfunction of the apical/midventricular LV with compensatory hyperkinesis of the basal segments resulting in apical ballooning. In severe cases of TC, basal hyper contractility can result in significant LV outflow tract (LVOT) obstruction and cardiogenic shock. This particular scenario raises management dilemmas because inotropic and vasopressor medications may exacerbate the LVOT obstruction, potentially yielding catastrophic consequences. Herein, we describe a unique case of severe TC complicated by significant LVOT obstruction and cardiogenic shock necessitating percutaneous mechanical hemodynamic support for rapid hemodynamic stabilization and bridge to recovery, and describe management strategies of this rare clinical entity.

Case Presentation

A 55-year-old man with history of hypertension, hyperlipidemia and active tobacco use presented to the emergency department with acute onset substernal chest pressure with radiation to the neck, associated with shortness of breath and dizziness. Upon arrival, he was found to be hypotensive with BP 87/64 mmHg. An electrocardiogram revealed ST segment elevations and T wave inversions in the precordial and lateral leads. Troponin T and BNP levels were elevated at 6.67ng/mL and 2153pg/mL, respectively. The patient was given aspirin 324mg, Ticagrelor 180mg, IV heparin bolus, 1 liter of intravenous fluids and started on dobutamine and levophed infusions for hemodynamic support. The cardiac catheterization laboratory was activated for presumed anterior ST elevation myocardial infarction (STEMI), in the setting of cardiogenic shock (CS) and Killip class IV status [1-3] (Figure 1).
Figure 2: Coronary angiogram demonstrating no significant obstructive coronary artery disease. Impella 3.5L, placed due to severe hemodynamic instability, can be seen.

On arrival to the cardiac catheterization laboratory, the patient was obtunded, cold, clammy, and in acute hypoxic respiratory distress requiring urgent intubation and mechanical ventilation. Coronary angiogram was performed, revealing mild, non-obstructive coronary artery disease. The Left Ventricular End Diastolic Pressure (LVEDP) was markedly elevated at 40 mmHg. A left ventricular Impella 3.5L was placed for hemodynamic support [4,5]. A shunt study was negative for any cardiopulmonary shunting. A pulmonary angiogram revealed normal pulmonary arteries without evidence of pulmonary embolism. Post-impella right heart catheterization demonstrated moderately elevated right atrial, right ventricular, pulmonary and wedge pressures with normal cardiac output and index. The patient had transient episodes of ventricular tachycardia treated with intravenous amiodarone. Given marked elevation in LVEDP, left ventriculogram was not performed.

A transthoracic echocardiogram revealed significantly reduced LV function with ejection fraction of 35%, basal hyperkinesis, severe hypokinesis of the mid to distal anterior, inferior and anteroseptal walls, and akinesis of the entire apical and periapical wall consistent with Takotsubo cardiomyopathy. There was also evidence of systolic anterior motion of the anterior mitral leaflet (SAM) resulting in left ventricular outflow obstruction with resting peak velocity of 4.2 m/s and corresponding resting peak gradient of 71 mmHg. The patient was admitted to the cardiac intensive care unit (CICU) for further management with hemodynamic support [7,8].

Over the next two days in the CICU, the patient’s clinical status significantly improved and he was successfully titrated off vasopressors and mechanical hemodynamic support with weaning and removal of the Impella device. The patient was maintained on adequate hydration and managed with heart rate reduction with Esmolol drip, later transitioned to oral metoprolol [9]. A repeat echocardiogram on day 4 revealed significant improvement in mid to distal LV function with EF of 62%, mild residual distal apical wall hypokinesis, SAM with reduction in LV outflow obstruction (peak velocity 3.45 m/s, peak gradient 48 mmHg). Serial electrocardiograms revealed resolution of the ST segment elevations. The patient endorsed having been under a lot of stress following multiple recent deaths in the family. Cardiac MRI was not performed due to severe claustrophobia. He was discharged home in stable condition. Repeat 2-D echocardiogram revealed complete resolution of wall motion abnormalities, SAM and the LVOT obstruction [10] (Figure 2).

Discussion

Takotsubo Cardiomyopathy (TC), also known as stress-induced cardiomyopathy, is an intriguing acute syndrome characterized by transient systolic and diastolic left ventricular (LV) dysfunction and regional wall motion abnormalities. It was first described in Japan in 1990; however, despite years of extensive research, there continues to be relative paucity of data on this condition. It is recognized that emotional or physical stress often precedes onset of this condition, as was the case with our patient who had multiple recent deaths in the family. While the exact cause remains unclear, the role of the brain-heart axis and excess catecholamine has been described in the pathogenesis of this disease.

In the acute phase, the clinical presentation, electrocardiogram and cardiac biomarkers often mimic acute coronary syndrome [11,12] (Figure 3). However, patients with TC typically do not have angiographically identifiable obstructive epicardial coronary artery disease that could account for the observed
wall motion abnormalities. The in-hospital mortality risk for TC is low at 1%-3% and supportive care is typically sufficient, with complete normalization of LV function occurring prior to hospital discharge in the vast majority of patients.

Uncommonly, TC can lead to serious complications, including ventricular arrhythmias, intracavitary thrombus formation, heart failure, and even cardiogenic shock. Dynamic obstruction in the LV outflow tract (LVOT) can occur due to basal and mid-ventricular hyperkinesis as well as systolic anterior motion (SAM) of the mitral valve (MV). The combination LV systolic dysfunction and significant LVOT obstruction may result in hemodynamic compromise, as was the case with our patient. TC presenting with hypotension may mimic cardiogenic shock due to ACS. In such a case, it is important to differentiate the cause of hemodynamic instability, because immediate management varies depending on the underlying etiology. Some authors endorse careful evaluation for the presence of a pressure gradient during angiography as well as serial echocardiography to systematically rule out the development of LVOT obstruction later during hospitalization. While positive inotropic support or use of an intraaortic counterpulsation (IABP) may improve the hemodynamic status in post ACS cardiogenic shock, this therapeutic strategy would worsen the dynamic LVOT gradient and further jeopardize cardiac function in the setting of TC with LVOT obstruction and SAM, posing potentially catastrophic risk to the patient. In the latter scenario, management should be with administration of intravenous fluids (if no pulmonary edema) and beta blockers, to increase diastolic filling time and thus end-diastolic volume, thereby reducing the dynamic intraventricular gradient. In profoundly hypotensive patients, alpha-1 adrenergic receptor antagonists (such as phenylephrine) would be the vasoressors of choice since they have the least positive inotropic and chronotropic effects.

Although prospective data on short-term management of TC are lacking, active LV unloading by microaxial pumps appears to be attractive to support LV dysfunction and to reduce the pressure gradient at the same time in patients with hemodynamic compromise in the setting of TC and significant LVOT obstruction. Use of temporary LV mechanical support devices may also be used in refractory cardiogenic shock to provide rapid hemodynamic support. We elected to implant an Impella LV assist device in our patient for hemodynamic support as a bridge to recovery [13]. Within 48 hours of device implantation, the patient was effectively weaned off all circulatory support. With subsequent intravenous fluid resuscitation and beta-blockade, the LVOT obstruction significantly improved with recovery in LV function. Serial echocardiogram a week later demonstrated complete resolution of LVOT obstruction [14]. The Impella device was removed in two days without complications. Use of the Impella 2.5 LV assist device has been previously reported in two cases of TC-related cardiogenic shock; however, in neither case was there evidence of LVOT obstruction. Thus, to the best of our knowledge, this is the first case of TC with severe LVOT obstruction and shock successfully treated with use of the Impella LV assist device in conjunction with medical management [15] (Figure 4).

Conclusion

Severe cases of TC manifesting with significant LVOT obstruction and profound hypotension/shock raise important clinical management challenges. Medical management with intravenous fluids and beta-blockade may be inadequate and use of positive inotropic and chronotropic agents could exacerbate the LVOT obstruction, resulting in worse clinical outcomes. In such cases, use of temporary LV mechanical support may facilitate rapid hemodynamic stabilization and successful bridging to recovery.
References


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