Hypertensive Hypertrophic Obstructive Cardiomyopathy Crisis Resolved with Transvenous Pacing Guided by Bedside Echocardiography

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Background

Hypertensive hypertrophic obstructive cardiomyopathy (HHoCM) is phenotypically similar to hypertrophic obstructive cardiomyopathy (HoCM) and usually presents in elderly patients with longstanding, poorly controlled hypertension. Cardiogenic shock due to HHoCM is clinically challenging because vasopressors, inotropes, and acute afterload reduction can worsen LVOT obstruction.

Presentation

An 81 year old female with longstanding poorly controlled hypertension was admitted with hemorrhagic lacunar intraparenchymal hematoma. For target systolic blood pressure of 100-140 mmHg, the patient was treated with concomitant oral hydralazine, lisinopril, hydrochlorothiazide, verapamil, intravenous hydralazine, and labetalol, and a nicardipine drip. After initial achievement of blood pressure control, she became hypotensive and hypoxic. Vasopressor requirement escalated rapidly, and hypotension was refractory to metoprolol, nicardipine, phenylephrine, and vasopresin. The patient developed an acute kidney injury (AKI) as a result of shock and required noninvasive positive pressure ventilation for pulmonary edema.

Echocardiogram revealed moderate to severe asymmetric septal hypertrophy (1.7-2.0 cm) with moderate concordant left ventricular hypertrophy, LVOT outflow tract obstruction with peak LVOT gradient of 102 mmHg, left ventricular ejection fraction (LVEF) of 80%, systolic anterior motion of mitral valve (SAM) with severe MR, and calcification of the mitral annulus and leaflet. (Figure 1) A diagnosis of hypertrophic hypertensive cardiomyopathy of the elderly (HHoCME) was established.

She then developed bradyarrhythmia due to junctional escape rhythm with heart rate (HR) of 45 beats/min (bpm) refractory to atropine and epinephrine.

Intervention

A right internal jugular transvenous pacing wire was inserted into the right ventricle (RV). At bedside with simultaneous echocardiography, HR was set at 55 bpm and increased sequentially to 80 bpm with LVOT peak velocity and velocity time integral (VTI) measurement at each HR. (Figure 2) At the optimal setting of 75 bpm, mitral SAM was absent, LVOT peak gradient was 36 mmHg, LVEF 65%, mitral regurgitation was moderate in severity, and cardiac output was 4.9 L/min with low beat to beat variability, further confirmed by noninvasive continuous cardiac output monitoring.

Within an hour, norepinephrine was weaned off, and no vasopressors or inotropes were required after 12 hours. Diuresis was tolerated and AKI resolved. After cessation of vasopressors and addition of low dose metoprolol, optimized LVOT gradient was further reduced to 15 mmHg.

Discussion

This patient developed HHoCM crisis due to aggressive blood pressure control, via afterload reduction, initiated to limit intracerebral hemorrhage. Acute preload or afterload reduction increases the severity of LVOT obstruction and compromises hemodynamics. This was further exacerbated by initiation of pharmacologic agents with positive inotropic effect. We successfully resolved our patient’s HHoCM crisis with transvenous RV pacing guided by echocardiography. By measuring LVOT diameter and LVOT VTI at each HR, we calculated stroke volume and cardiac output at HR from 55 to 80 bpm. The degree of mitral regurgitation was reduced from severe to moderate with reduction of dynamic LVOT obstruction. (Figure 3) Systolic anterior motion of the mitral valve worsens LVOT obstruction and results in subsequent dynamic mitral regurgitation. Slower heart rates allow for increased filling of the left ventricle and therefore decrease the cardiac outflow obstruction, however with more time in systole for flow across the mitral valve, mitral regurgitation worsens. Therefore, balancing mitral regurgitation and outflow tract obstruction with heart rate control can optimize forward flow.

First line therapy for symptomatic LVOT obstruction due to mitral SAM is pharmacological with beta blockers or non-dihydropyridine calcium channel blockers, and for treatment of hypotension related to LVOT obstruction pure alpha agonists are recommended as positive inotropes can increase dynamic obstruction and thus LVOT gradient. Theoretically, RV pacing results in asynchonous septal movement, reducing LVOT obstruction, SAM and dynamic mitral regurgitation. Echocardiographically guided rate optimization allows for real time balancing between LV filling time and absolute heart rate in order to maximize cardiac output.

Conclusions

Permanent pacemaker implantation with synchronous atrioventricular (AV) pacing and AV delay of 75-100 ms has been shown to reduce LVOT and intraventricular gradient. However, its use for symptomatic LVOT gradient relief is controversial due to lack of consensus on long term benefit. In the acute setting, as presented in this patient, RV pacing could provide a significant, and rather immediate reduction of dynamic LVOT obstruction and restore hemodynamic stability.

References