Hypertrophic Obstructive Cardiomyopathy Presenting with Profound Hypotension* 

Role of Two-Dimensional and Doppler Echocardiography in Diagnosis and Management

Eric Kirschner, M.D.; Marvin Berger, M.D., F.C.C.P.; and Emanuel Goldberg, M.D., F.C.C.P.

Five patients, all women, age ranges 59 to 84 years, with underlying hypertrophic obstructive cardiomyopathy (HOCM) presented with profound hypotension. Initial clinical and hemodynamic evaluation suggested cardiogenic shock in two cases, acute myocardial ischemia in two cases, and hypovolemia in one case. The two patients thought to be in cardiogenic shock were given inotropes without improvement. One patient in whom myocardial ischemia was suspected received nitroglycerin, followed by a marked fall in blood pressure. All five patients remained hypotensive until two-dimensional and Doppler echocardiography were performed, identifying HOCM as the cause of the hypotension, thereby allowing for corrective therapeutic measures.

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CI = cardiac index; HOCM = hypertrophic obstructive cardiomyopathy; LVOT = left ventricular outflow tract; PCWP = pulmonary capillary wedge pressure; SAM = systolic anterior motion of the mitral valve

The two-dimensional and Doppler echocardiographic findings in hypertrophic obstructive cardiomyopathy (HOCM) have been well characterized.1-3 Despite recent reports on the benign course of this disorder,4,5 patients may occasionally present with marked hemodynamic instability. In those cases, a rapid and definitive diagnosis is necessary to guide treatment. This report describes five patients with HOCM who developed profound hypotension, for which the cause remained elusive until two-dimensional and Doppler echocardiography were performed, thereby allowing for corrective therapeutic measures.

CASE REPORTS

CASE 1

An 84-year-old woman with a previous myocardial infarction presented with a one-day history of malaise, weakness, and dizziness. The blood pressure was 70/50 mm Hg, pulse rate was 100 beats/min, and a grade 2/6 systolic ejection murmur was heard at the left parasternal area. An ECG showed normal sinus rhythm with nonspecific ST segment and T wave abnormalities. She was initially treated with intravenous hydration and dopamine. A pulmonary artery catheter revealed hemodynamics suggestive of cardiogenic shock (pulmonary capillary wedge pressure [PCWP] of 21 mm Hg and a cardiac index [CI] of 2.1 L/min/m²). Despite increasing doses of dopamine, the patient remained hypotensive.

An echocardiogram showed a hypertrophic left ventricle with an akinetic apex, a small left ventricular cavity, proximal septal hypertrophy, and systolic anterior motion of the mitral valve (SAM). Continuous-wave Doppler recording across the left ventricular outflow tract (LVOT) demonstrated a late peaking velocity of 6 m/s, corresponding to a peak pressure gradient of 144 mm Hg (Fig 1, left).

Following the study, dopamine therapy was stopped and the patient was treated with intravenous propranolol. Her blood pressure increased to 120/60 mm Hg, the PCWP decreased to 12 mm Hg, and the CI increased to 2.5 L/min/m². A follow-up Doppler study no longer demonstrated a gradient across the LVOT (Fig 1, right).

CASE 2

A 74-year-old woman presented with chest pounding on the seventh postoperative day following repair of a left hip fracture complicated by blood loss. The blood pressure was 80/50 mm Hg, pulse rate was 110 beats/min, and a new harsh grade 4/6 systolic ejection murmur was heard at the left parasternal area. The ECG showed sinus tachycardia with nonspecific ST segment and T wave abnormalities. The hemoglobin level was 8.9 mg/dl.

An echocardiogram showed a hyperdynamic left ventricle, septal hypertrophy, and SAM. Continuous-wave Doppler recording across the LVOT demonstrated increased velocity of 5 m/s, corresponding to a peak pressure gradient of 100 mm Hg.

After the diagnosis of HOCM was made, a pulmonary artery catheter was inserted to guide fluid management. The PCWP was 3 mm Hg and CI was 2.9 L/min/m². Following treatment with intravenous fluids and blood transfusion, the murmur decreased in intensity, and her hemodynamics stabilized (blood pressure, 110/70 mm Hg; PCWP, 12 mm Hg; CI, 4.1 L/min/m²).

CASE 3

A 73-year-old woman was transferred to our institution following a barium enema during which she developed chest pain, dyspnea, and diaphoresis. Her blood pressure was 84/50 mm Hg, pulse rate was 90 beats/min, and a grade 3/6 systolic murmur was heard at the left parasternal area. Lung examination revealed bibasilar rales. The ECG showed normal sinus rhythm, a right bundle branch block, and evidence suggesting an acute myocardial infarction (Fig 2A).

An echocardiogram showed a hypertrophic left ventricle with a dysskinetic apex, proximal septal hypertrophy, and SAM. Continuous-wave Doppler recording across the LVOT demonstrated a peak

*From the Department of Medicine, Division of Cardiology, Beth Israel Medical Center, New York, and the Department of Medicine, Mount Sinai School of Medicine of the City University of New York.

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Reprint requests: Dr. Kirschner, Division of Cardiology, Beth Israel Medical Center, 1st Avenue at 16th Street, New York 10003.
velocity of 4 m/s, corresponding to a peak pressure gradient of 64 mm Hg.

With intravenous fluids, the hypotension resolved and the systolic murmur decreased in intensity. There was no elevation of cardiac isoenzymes and there was resolution of the ischemic ECG changes (Fig 2B). A repeated echocardiogram and Doppler study showed improvement in the apical wall motion abnormality and no resting gradient across the LVOT. Cardiac catheterization revealed normal coronary arteries with no evidence of intramyocardial systolic compression, and a hyperdynamic left ventricle with a small area of apical dyskinesis. There was a provokable LVOT gradient of 130 mm Hg with amyl nitrate.

**CASE 4**

A 59-year-old woman with a history of hypertension presented with chest pain for several hours. Her blood pressure was 130/60 mm Hg, pulse rate was 90 beats/min, and a grade 2/6 systolic ejection murmur was heard at the left parasternal area. The ECG showed normal sinus rhythm with left ventricular hypertrophy. Following treatment with sublingual nitroglycerin, she became hypotensive to 70/50 mm Hg.

An echocardiogram showed a hyperdynamic left ventricle with concentric left ventricular hypertrophy, mitral annular calcification, and SAM. Continuous-wave Doppler recording across the LVOT demonstrated a peak velocity of 6 m/s, corresponding to a peak pressure gradient of 144 mm Hg.

Her symptoms and hypotension resolved with intravenous fluids and β-blocker therapy.

**CASE 5**

A 74-year-old woman with known HOCM presented with chest pain, nausea, and vomiting. She was found to be in atrial fibrillation with a rapid ventricular response that slowed and converted to normal sinus rhythm after treatment with digoxin and verapamil. She subsequently developed respiratory distress and was hypotensive to 80/50 mm Hg. A grade 2/6 systolic murmur was heard at the left parasternal area. Lung examination revealed bilateral rales. The ECG showed normal sinus rhythm and left ventricular hypertrophy. She was placed on mechanical ventilatory support, and a pulmonary artery catheter revealed a PCWP of 45 mm Hg and CI of 2.5 L/min/m². She was believed to be in cardiogenic shock, but despite inotropic support, she remained hypotensive.

An echocardiogram showed a hyperdynamic left ventricle with an akinetic apex, septal hypertrophy, mitral regurgitation, and SAM. Continuous-wave Doppler recording across the LVOT demonstrated a peak velocity of 5 m/s, corresponding to a peak pressure gradient of 100 mm Hg (Fig 3, left).

The inotropes were tapered and she was treated with intravenous

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**FIGURE 1.** Left. Continuous-wave Doppler recording across the left ventricular outflow tract (LVOT) showing a peak velocity of 6 m/s and a gradient of 144 mm Hg. Mitral regurgitation (MR) is also present. Right. After discontinuation of dopamine therapy and treatment with propranolol, the peak velocity has decreased to 1.2 m/s and an outflow tract gradient can no longer be detected.

**FIGURE 2.** A. The ECG shows ST segment elevations in leads 1, 2, 3, aVF, and V₃-V₆. B. On follow-up ECG, the ST segment elevations are no longer present.
propranolol resulting in an improvement in her hemodynamic status (blood pressure, 139/60 mm Hg; PCWP, 96 mm Hg; CI, 3.0 L/min/m²). A follow-up Doppler recording revealed a decrease in the peak velocity across the LVOT to 2.3 m/s, corresponding to a peak gradient of 21 mm Hg (Fig. 3 right). Despite the transient hemodynamic improvement, she died several days later of overwhelming sepsis.

**Discussion**

In all of our patients, the presence on two-dimensional echocardiogram of a hyperdynamic, nondilated, hypertrophic left ventricle with SAM, and an increased late peaking velocity across the LVOT by Doppler recording, allowed for the rapid recognition of HOCM as the cause of hypotension. In such cases, early recognition of HOCM is important as the clinical presentation may mimic a variety of acute conditions, where conventional therapy may exacerbate the underlying disorder.

Two patients had hemodynamics suggestive of cardiogenic shock unresponsive to inotropic therapy. After HOCM was recognized by echocardiography, they “paradoxically” responded to negative inotropic therapy. Similar results were noted by Topol et al in a group of elderly patients with hypertrophic cardiomyopathy presenting with congestive heart failure. By decreasing myocardial contractility and increasing ventricular filling, negative inotropes will decrease the outflow tract obstruction, increase stroke volume, and improve hemodynamics.

Acute myocardial ischemia was suspected in two patients. One patient had ECG changes suggestive of an acute myocardial infarction, yet no significant lesions were detected on coronary angiography. The precipitating factor appeared to be the barium enema. Although the cardiovascular complications of a barium enema are rare, dehydration from the bowel preparation and increased sympathetic tone may have increased the LVOT obstruction and myocardial oxygen consumption, resulting in the severe hypotension and ischemia. This would suggest that patients with HOCM undergoing a barium enema may be at increased risk for cardiovascular complications.

In addition to the ECG changes, this patient showed improvement in a segmental wall motion abnormality, suggesting reversible myocardial ischemia. Myocardial ischemia can occur in hypertrophic cardiomyopathy despite normal epicardial coronary arteries and can result in permanent left ventricle scarring and dysfunction. Reversible myocardial ischemia in hypertrophic cardiomyopathy has been demonstrated with atrial pacing and exercise thallium-201 perfusion studies. Proposed mechanisms include inadequate capillary density, narrowed small intramural coronary arteries, or systolic compression of large intramyocardial coronary arteries.

Chest pain, which is a common complaint among patients with hypertrophic cardiomyopathy, may result in the inadvertent administration of nitrate therapy. By decreasing preload, nitrates will increase the outflow tract obstruction and may result in profound hypotension as in one of our patients.

It is interesting to note that all our patients were women, and that four of five were elderly (73 to 84 years). These findings are in agreement with previous reports on hypertrophic cardiomyopathy in the elderly that have shown a female predominance ranging from 64 to 94 percent. The reasons for this are unknown.

Although hypertrophic cardiomyopathy is in general a benign disorder, a subset of patients, particularly
those with dynamic outflow tract obstruction, may under certain conditions (i.e., hypovolemia, increased sympathetic tone, vasodilator therapy) develop severe hypotension for which the cause is not suspected at bedside. As demonstrated, invasive hemodynamic monitoring may not be helpful and may even be misleading. Early two-dimensional and Doppler echocardiography can be of significant value in establishing the diagnosis of HOCM as well as differentiating it from other cardiovascular causes of acute hypotension. Immediate volume replacement is standard therapy in patients who develop hypotension secondary to nitrate therapy or dehydration. Our findings suggest that in the profoundly hypotensive patient with a systolic murmur, ECG evidence of left ventricular hypertrophy, or a history of hypertension, further evaluation with two-dimensional and Doppler echocardiography is warranted to help guide management.

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