sectional echocardiograms are able to examine parts of the left ventricle that are inaccessible to accurate diagnosis by the M-mode technique. This advantage is particularly applicable to visualization of the left ventricular apex, a common site of thrombi, which is not reliably examined by conventional echocardiograms but is well visualized with the two-chamber and four-chamber apical views of the two-dimensional echocardiogram; however, with both echocardiographic techniques, accurate diagnosis of a left ventricular mass is critically dependent on proper gain settings on the receiver and on the acoustic mismatch between the mass and blood. The detection of a left ventricular thrombus by current echocardiographic instruments may require the higher acoustic impedance of an organized thrombus, as opposed to that of a recently formed clot.

Our patient is unusual in several respects. Only one other article and recently several abstracts have reported the echocardiographic diagnosis of a left ventricular thrombus occurring as a discrete mass, rather than as vague echocardiographic densities. Contrary to reported experience, the left ventricular thrombus in this patient was well identified by conventional M-mode echocardiograms. Clinically, the repeat retinal arterial emboli seen in this patient have not previously been described in association with left ventricular masses. The extensive calcification of the thrombi in this case is also quite unusual, as is the unexplained absence of coexisting myocardial disease which would predispose to such thrombus formation.

REFERENCES
1 Young RD, Hunter, WC. Primary myxoma of the left ventricle with embolic occlusion of the abdominal aorta and renal arteries. Arch Pathol Lab Med 1947; 43:86-91
3 Gerbode F, Kerth WJ, Hill JD. Surgical management of tumors of the heart. Surgery 1967; 61:94-100
4 Danta G, Williams DO. Multiple emboli from left ventricular myxoma. Br Heart J 1969; 31:799-802
5 Mandel MM, Strimmel WH. Ventricular myxoma associated with cerebral embolism. JAMA 1970; 214:2154-56
13 Reeder GS, Tajik AJ, Seward JB. Detection of left ventricular thrombus with two dimensional echocardiography (abstract). Circulation 1979; 60(suppl 2):II-19
15 Gotttdiener JS, Schooley RT, Maron BJ, Fauzi AS. Cardiac abnormalities in the hypereosinophilic syndrome (abstract). Circulation 1979; 60(suppl 2):II-19

Midystolic Closure of the Aortic Valve in Primary Pulmonary Hypertension*

Yoshinori L. Doi, M.D.; Richard L. Bishop, M.D.; Tetsuro Sugiura, M.D.; and David H. Spodick, M.D., F.C.C.P.

Echocardiographic examination in a patient with primary pulmonary hypertension demonstrated midys-
When individuals, especially at a young age, present with dyspnea, chest pain, and/or syncope, the differential diagnosis may include primary pulmonary hypertension and hypertrophic cardiomyopathy.\(^1,2\) Often these entities can be separated on examination and on 

**Case Report**

A 48-year-old woman was admitted with a six-week history of progressive dyspnea on exertion associated with a dry, nonproductive cough. She had lost 4 kg over the prior six months and had had a recent episode of ventricular bigeminy. Based on an examination and echocardiogram done elsewhere, hypertrophic cardiomyopathy had been diagnosed. On examination, she was mildly dyspneic at rest. The blood pressure was 114/70 mm Hg, and there was a palpable right ventricular heave. The first heart sound was normal and the second physiologically split with a prominent pulmonary component. There was a grade 3/6 pansystolic murmur loudest at the lower left sternal border that radiated to the axilla. Borderline cardiomegaly was present on chest x-ray film, the lungs appearing clear. The ECG (Fig 1) revealed normal sinus rhythm; QRS axis of +30°; peaked P wave in lead 2, and slight ST-T changes in the inferolateral leads. There was no evidence of right or left ventricular hypertrophy. Echocardiograms (Fig 2 and 3) revealed enlarged right ventricle (30 mm). The septum was not thick (10 mm), and its motion was normal except for slightly increased amplitude. The left ventricle was small. The anterior mitral leaflet contacted the septum during diastole and had a reduced diastolic closure rate. Motion of the posterior mitral valve leaflet was normal. Left atrial size was within normal range (36 mm). The pulmonic valve had an “a” wave, and the aortic valve showed midystolic closure.

Propranolol (20 mg four times daily) was administered for the treatment of frequent ventricular bigeminy with a presumptive diagnosis of hypertrophic cardiomyopathy, but her blood pressure fell, requiring dopamine therapy. Swan-Ganz catheterization revealed the following: central venous pressure, 16 mm Hg; right ventricular pressure, 85/3 mm Hg; pulmonary artery pressure, 85/15 mm Hg (mean: 48 mm Hg); and a normal pulmonary capillary wedge pressure of 11 mm Hg. There was no oxygen step-up. After the resolution of her hypotensive episode, phentolamine (50 mg three times daily) was given, but a week later, she was found dead in bed. Postmortem examination reconfirmed the clinical

---

**Figure 1.** Electrocardiogram revealing normal sinus rhythm.

**Figure 2.** Echocardiogram demonstrating enlarged right ventricle (RV) and small left ventricle (LV). Left ventricular end-systolic dimension was 20 mm and end-diastolic dimension 38 mm. Hyperdynamic septum (IVS) and posterior wall (PW) as well as reduced diastolic closure rate of mitral valve (MV) are present. Left atrial (LA) size is normal. AO indicates aorta.
diagnosis of primary pulmonary hypertension. The heart weighed 330 gm with a normal left ventricular shape and contour. The right ventricle was hypertrophied, wall thickness being 9 mm. There were atheromatous plaques in the pulmonary arteries but no emboli or thrombi. Microscopic examination was normal except for hypertrophy of the right ventricular myocardium. There were no lesions suggesting hypertrophic cardiomyopathy.

**DISCUSSION**

Midsystolic closure of the aortic valve has not previously been reported in primary pulmonary hypertension. It may be present in hypertrophic cardiomyopathy, in discrete subaortic stenosis, and in ruptured aneurysm of the right coronary sinus of Valsalva. It was reported in a patient with mitral regurgitation and in patients with ventricular septal defects. In our patient, the interval from the opening of the aortic valve to the point of midsystolic closure was 0.12 second, making discrete subaortic stenosis unlikely. Normal left atrial size indicated that significant mitral regurgitation was unlikely. Ventricular septal defects and ruptured aneurysm of the right coronary sinus of Valsalva were excluded on clinical grounds. Given the patient's history and clinical findings, however, the diagnosis of hypertrophic cardiomyopathy was difficult to exclude, and the echocardiogram could easily be misinterpreted.

Midsystolic closure of the aortic valve is most commonly seen in hypertrophic cardiomyopathy, either obstructive or nonobstructive. These conditions are generally associated with a hyperdynamic heart, brisk early systolic ejection, and at times, a narrowed outflow tract, which result in a high initial bloodflow velocity across the aortic valve, followed by some reduction in flow velocity as ejection of the small stroke volume is nearly completed. A similar hemodynamic state has been observed in normal humans and dogs with adrenergic stimulation or with volume depletion. It is possible that a low cardiac output imposed on a normal left ventricle may produce a similar situation in primary pulmonary hypertension. This may be especially true in a patient with right ventricular pressure overload and hypertrophy, which can result in hyperdynamic septum as well as distortion of left ventricular geometry. It is of interest that a patient with dyspnea, syncope, and chest pain recently reported also had midsystolic closure of the aortic valve as well as other echocardiographic features of hypertrophic cardiomyopathy but at autopsy was found to have pulmonary embolism.

The echocardiogram of this patient (Fig 2) showed almost all features of primary pulmonary hypertension except for normal septal thickness and an "a" wave on the pulmonic valve echo. Normal septal thickness, confirmed at autopsy, correlated well with lack of evidence of ventricular hypertrophy on her ECG. However, the presence of a pulmonic valve "a" wave is somewhat unusual among patients with pulmonary hypertension. The explanation may lie in low pulmonary arterial diastolic pressure. If the pulmonary arterial diastolic pressure is low, relatively close to the right ventricular diastolic pressure, a rise in the right ventricular end-diastolic pressure following right atrial systole may result in relatively normal motion of the pulmonic valve.

To date, attention has been paid to reduced diastolic closure rate of the anterior mitral leaflet in primary pulmonary hypertension, and consequently, the differentiation from mitral stenosis has been emphasized. However, many features, including a small left ventricle, thickened septum, and reduced diastolic closure rate of the anterior mitral leaflet, are also features of hyper-
trophic cardiomyopathy. Although an abnormally thickened septum was not seen in our patient, Goodman et al. noted increased septal thickness of 13 mm or greater and increased septal to posterior wall ratio in all of nine patients with primary pulmonary hypertension. In addition, when midsystolic closure of the aortic valve is demonstrated as in our patient, differentiation from hypertrophic cardiomyopathy may be extremely difficult, particularly from those cases without systolic anterior motion of the mitral valve. A possible clue in differential diagnosis may be dilated right ventricle and abnormal pulmonary valve motion in primary pulmonary hypertension. Another difficulty may be raised in differentiating ruptured aneurysm of the right coronary sinus of Valsalva into the right ventricle, though the differentiation is usually possible on clinical grounds. In ruptured aneurysm of the right coronary sinus of Valsalva, the right coronary cusp shows midsystolic closure, but other cusps appear to show normal movement, and this could be a clue for echocardiographic differentiation.

In summary, midsystolic closure of the aortic valve in a patient with primary pulmonary hypertension extends the differential diagnosis of this disorder to hypertrophic cardiomyopathy as well as other diseases. We hope that this documentation of midsystolic closure of the aortic valve in primary pulmonary hypertension will provoke thought concerning its etiology as well as drawing attention to the differential diagnosis from hypertrophic cardiomyopathy.

ACKNOWLEDGMENT: We gratefully acknowledge DeLores Paladino, R.N., and Kathleen Moreau, L.F.N., for technical assistance.

REFERENCES