An Early Systolic Sound Associated With Midventricular Obstruction in a Patient With Hypertrophic Cardiomyopathy

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A 57-year-old woman was admitted for examination because of chest discomfort. Transthoracic echocardiography was performed and she was diagnosed as having hypertrophic cardiomyopathy. An echocardiogram also revealed that she had midventricular obstruction with a pressure gradient of 125 mmHg determined by Doppler echocardiography. A phonocardiogram showed an early systolic sound and the beginning of the sound coincided with the time of septal-posterior wall contact. In addition, the timing also corresponded to the sudden obstruction of blood flow in the region of the midventricular narrowing. Furthermore, this sound markedly decreased with the reduction in pressure gradient caused by cibenzoline treatment. Thus, it was concluded that the early systolic sound was associated with midventricular obstruction and produced by a rapid deceleration of the interventricular flow caused by midventricular obstruction. (Jpn Circ J 1998; 62: 385–388)

Key Words: Early systolic sound; Hypertrophic cardiomyopathy; Midventricular obstruction

The occurrence of systolic sounds, also called pseudoejection sounds or early systolic sounds, has been reported in idiopathic hypertrophic subaortic stenosis (IHSS). Braunwald et al. noted this sound in 7 of 64 patients with IHSS, an incidence of 11%. Sze and Shah reported that this sound was associated with systolic anterior motion (SAM), that is the sound coincided with the sudden halting of SAM of the anterior mitral leaflet.

To our knowledge, however, there has been no report of a patient with midventricular obstruction who has an early systolic sound. We report on such a patient with midventricular obstruction.

Case Report

A 57-year-old woman was admitted to our hospital because of worsening chest discomfort on effort on 17 June 1996. She was diagnosed as having hypertrophic cardiomyopathy (HCM) in 1993 and had been treated with the calcium antagonist nifedipine. Her past history and family history were unremarkable.

Physical examination on admission revealed the following. Her pulse was irregular at 88 beats/min and her blood pressure was 134/84 mmHg. On auscultation, a Levine II/VI systolic murmur that was maximal at the apex was also heard. Breath sounds were clear and no peripheral edema was observed. A neurologic examination was unremarkable.

An electrocardiogram on admission revealed atrial fibrillation and left ventricular hypertrophy with ST-segment depression and inverted T waves (Fig 1). On the day of admission, atrial fibrillation returned to sinus rhythm after treatment with diltiazem. A chest radiograph showed cardiac enlargement, indicating a cardiothoracic ratio of 60%. Congestion of the lung field was not observed.

A transthoracic echocardiogram, recorded on the day that atrial fibrillation returned to sinus rhythm, showed asymmetric septal hypertrophy with a small left ventricular cavity. Mid-ventricular hypertrophy at the papillary muscle level was especially prominent on the 4-chamber view (Fig 2). An M-mode echocardiogram showed no SAM of the anterior mitral leaflet, and an early systolic sound was also recorded on phonocardiogram (PCG) (Fig 3A). An M-mode echocardiogram at the papillary muscle level showed septal-posterior wall contact at systole (Fig 3B). At the same time, the beginning of the early systolic sound was concordant with the time of septal-posterior wall contact, as shown in Fig 3B.

To investigate blood flow in the left ventricle, Doppler echocardiography was performed. As shown in Fig 4A, the waveform at the region of midventricular narrowing was biphased, and lasted until the end of isovolumic relaxation or the beginning of a rapid filling phase beyond the second heart sound as a result of the diastolic paradoxical jet flow. The systolic peak flow velocity at the same portion was on average 5.6 m/sec and the calculated pressure gradient was 125 mmHg.

Cardiac catheterization was performed. Left ventriculography revealed marked hypertrophy at the interventricular septal site and basal free wall (arrow marks), and the left ventricular cavity was shaped like a sandglass, indicating midventricular obstruction (Fig 5). The press...
sure measurements in the apical and basal chambers could not be confirmed because of catheter-induced ventricular arrhythmia. However, the pressure gradient at the left ventricular outflow tract was absent.

**Effect of Cibenzoline on Midventricular Obstruction**

These findings confirmed the diagnosis of HCM with midventricular obstruction. To reduce the midventricular obstruction, 300 mg per day of cibenzoline, a class Ia antiarrhythmic agent, was administered. After treatment with cibenzoline, Doppler trans-midventricular flow velocity decreased to an average of 4.3 m/sec and thus the pressure gradient decreased to 75 mmHg. As shown in Fig 4B, the diastolic paradoxical jet flow in the region of midventricular narrowing disappeared and the waveform showed a single-peaked pattern. In addition, the patient's chest symptoms improved significantly and the early systolic sound almost disappeared.

**Discussion**

Clinically, 2 types of hypertrophic obstructive cardiomyopathy have been confirmed, so-called IHSS and HCM with midventricular obstruction. HCM with midventricular obstruction is rare, and its pathophysiology and prognosis remain unknown. In some patients with IHSS, the existence of a pseudo-ejection sound or an early systolic sound has been confirmed.

The characteristics of the early systolic sound in our patient were as follows: the sound was heard maximally at the left sternal margin of the IVth intercostal space, and not so high pitched. Its appearance coincided with the beginning of septal-posterior wall contact and the beginning of the cessation of midventricular blood flow. Thus, the genesis of this sound seems to be related to a rapid deceleration in blood flow associated with septal-posterior wall contact, and the vibration made by the structures of the left ventricle, including chordae and papillary muscles seems to be the main source of this sound. As observed in Fig 4, this early systolic sound was delayed more than 150 msec from the beginning of ejection flow. This is very different from the characteristics of aortic ejection sounds.

The class Ia antiarrhythmic drug disopyramide has been shown to decrease the pressure gradient in the majority of patients with hypertrophic obstructive cardiomyopathy and to improve left ventricular diastolic filling.
However, disadvantages of this drug are the side-effects caused by its strong anticholinergic action. Recently, we reported that cibenzoline, which is a class Ia antiarrhythmic agent with only weak anticholinergic effects, has the same effect as disopyramide, i.e., it can attenuate left ventricular pressure gradient and relieve symptoms in IHSS.

In our study, cibenzoline also reduced the pressure gradient and symptoms in a patient with midventricular obstruction. At the same time, the early systolic sound decreased in intensity. This finding also supports the hypothesis that the early systolic sound in our patient is clearly related to the high pressure gradient caused by midventricular obstruction. The main action of cibenzoline (reducing the midventricular pressure gradient) may be caused by the ability of cibenzoline to reduce the myocardial contractility.
References


