Case Reports

Murmur Associated with Diastolic Paradoxical Jet Flow

in a 43-Year-Old Man with Hypertrophic Cardiomyopathy

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A diastolic paradoxical jet flow, often seen in patients with hypertrophic cardiomyopathy, is a unique flow from the apex toward the base of the left ventricle during isovolumic relaxation. To date, this phenomenon appears to have been noninvasively detected only on echocardiograms. We report the case of a 43-year-old man with hypertrophic cardiomyopathy and a diastolic paradoxical jet flow, in whom cardiac auscultation revealed a soft S_4, a systolic ejection murmur, and a low-pitched early diastolic murmur immediately after S_2 at the apex. On comparing his echocardiographic findings with those on phonocardiograms and apexcardiograms, we confirmed that the unusual murmur coincided with the diastolic jet flow. To our knowledge, this is the first case in which heart murmurs associated with a diastolic paradoxical jet flow have been clearly described. Because these flows can increase the risk of adverse outcomes, detecting any associated murmurs by methods other than echocardiography is worthwhile, even in the era of advanced imaging techniques. ***(Tex Heart Inst J 2018;45(2):102-5)***

Key words: *Blood flow velocity; cardiomyopathy, hypertrophic/diagnostic imaging/physiopathology; echocardiography; heart sounds; heart ventricles/physiopathology; isometric contraction; phonocardiography*

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A diastolic paradoxical jet flow is a unique flow directed from the apex toward the base of the left ventricle during isovolumic relaxation. Such flows are often observed in patients with hypertrophic cardiomyopathy (HCM), and they are associated with an increased risk of adverse outcomes, such as ventricular arrhythmia or systemic embolism.[1-4] This phenomenon has been noninvasively detected only on echocardiograms. We report the case of a patient with HCM whose paradoxical jet flow was identified on phonocardiograms and apexcardiograms.

Case Report

An asymptomatic 43-year-old man with HCM presented at Matsushita Memorial Hospital for further testing after abnormal electrocardiographic results. Cardiac auscultation revealed a soft S_4; a systolic ejection murmur; and a low-pitched, early diastolic murmur immediately after S_2 at the apex. The other results of the physical examination were normal. An electrocardiogram indicated left ventricular hypertrophy, and a chest radiograph showed nothing notable.

A phonocardiogram was performed to clarify the components of the patient's complex heart sounds. It was recorded by placing microphones at the patient's upper left sternal border and near the apex, and an apexcardiogram was performed simultaneously by placing a transducer at the true apex, as described previously.[5] The tracings revealed a soft heart sound before S_1, a high-pitched ejection murmur during midsystole, and another soft heart sound after S_2 (Fig. 1A). The 2 additional heart sounds were thought to be an S_4 and an S_3 because they respectively coincided with an A wave and a rapid filling wave on the apexcardiogram. A phonocardiogram, obtained at the true apex after removal of the transducer for the apexcardiogram, revealed a low-pitched murmur immediately after the pulmonary component of S_2 (Fig. 1B). The change in wave shape during the last part of the early diastolic murmur—an increase in amplitude and duration approximately 200 ms after the onset of S_2—provided further evidence of an S_3.

Echocardiograms showed myocardial hypertrophy of the ventricular septum, the anterior wall of the left ventricle, and the apex of both ventricles, with a maximum

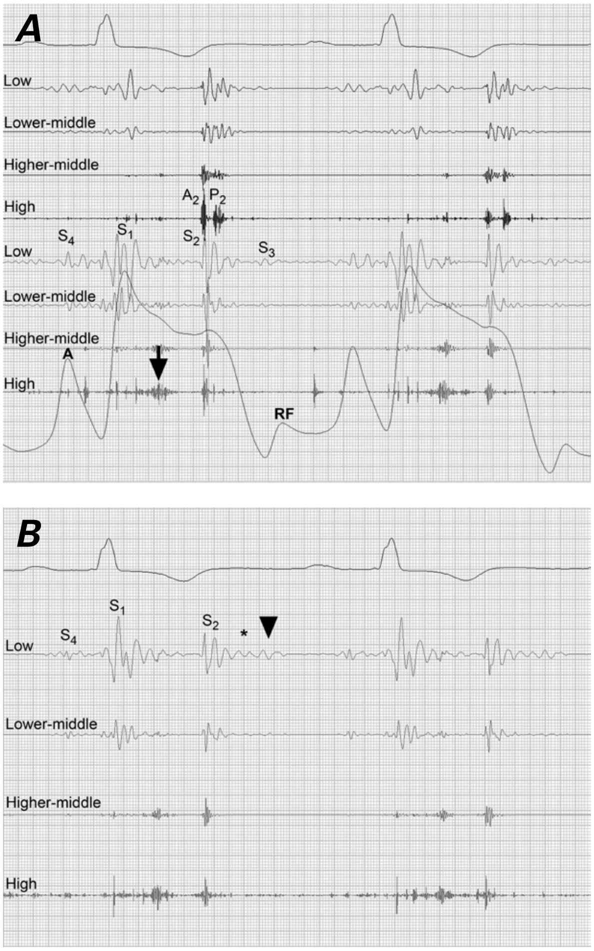


Fig. 1 ***A****) Phonocardiogram obtained at the upper left sternal border and near the apex shows an S_4 before S_1; a high-pitched midsystolic ejection murmur (arrow); and an S_3 approximately 200 ms after the onset of S_2. Apexcardiogram simultaneously recorded at the true apex shows a prominent A wave (A) coinciding with S_4 and a rapid filling wave (RF) coinciding with S_3.* ***B****) Phonocardiogram obtained at the true apex shows a low-pitched, early diastolic murmur immediately after S_2 (asterisk). The last part of the murmur, which shows a change in wave shape occurring about 200 ms after S_2 onset, suggested an S_3 (arrowhead).*

A_2 = aortic component of S_2; P_2 = pulmonary component of S_2

wall thickness of 23 mm (Fig. 2A). The peak early mitral inflow velocity (E) was 0.6 m/s, and the E-wave deceleration time was 140 ms; the ratio of E to the peak early septal mitral annular velocity (E/e′) was 19.4. At end-systole, a midventricular obstruction and an apical chamber were observed (Fig. 2B); Doppler images revealed the coincident diastolic paradoxical jet flow from the apex to the base of the left ventricle (Figs. 2C and D). The peak velocity was 3.1 m/s, and it did not change during the Valsalva maneuver. We found trivial mitral regurgitation but no valvular or congenital heart disease. A color-flow Doppler M-mode image and a phonocardiogram obtained at the apex suggested that the early diastolic murmur was a combination of a paradoxical flow murmur and an S_3 (Fig. 2E).

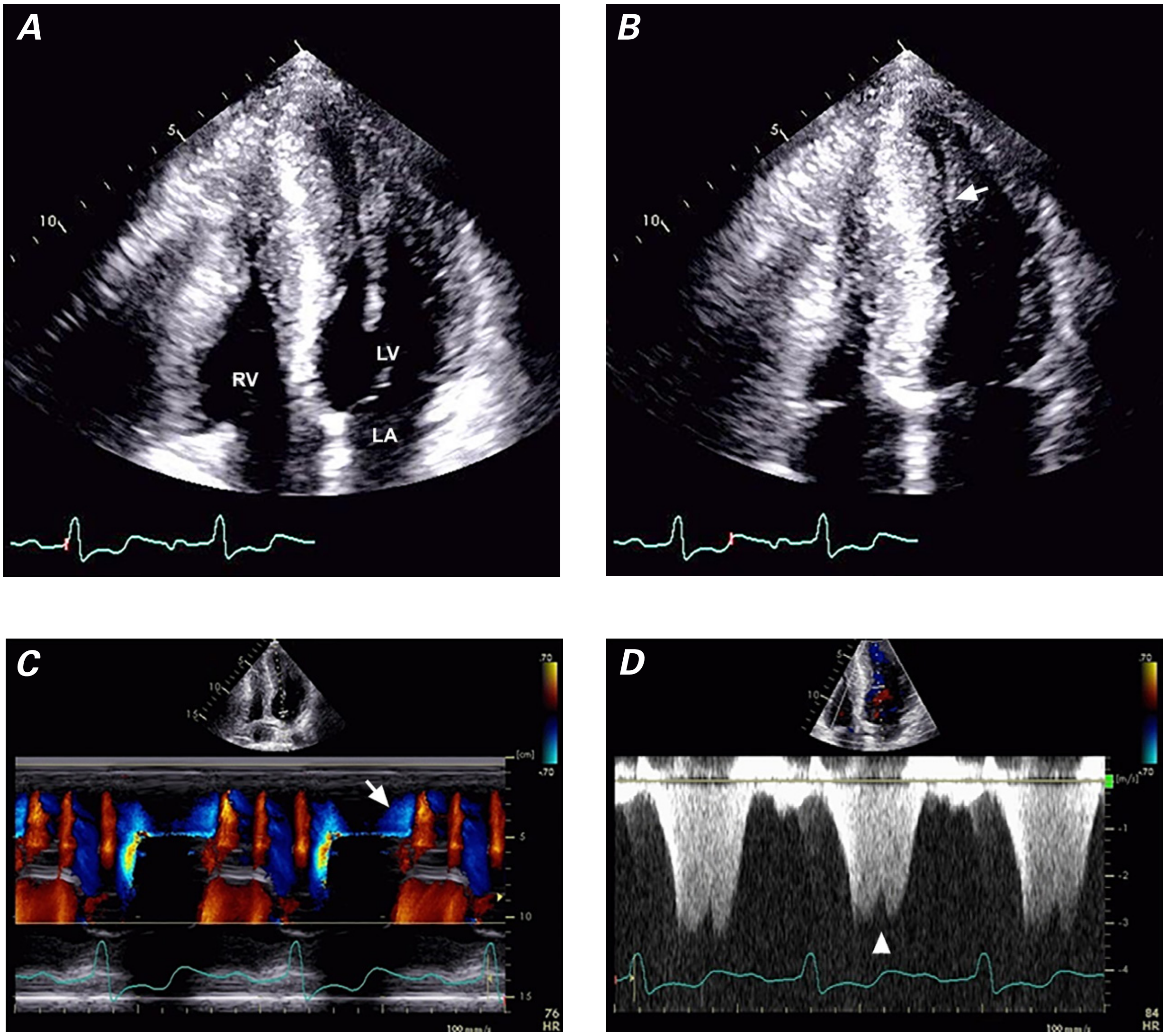
The patient declined medical therapy, including β-blockers. His unique early diastolic murmur waxed and waned during his 1-year follow-up visit. He was still asymptomatic at that time but was lost to follow-up thereafter.

Discussion

Patients with obstructive HCM often have both an S_4 and a systolic ejection murmur. Given our patient's short E-wave deceleration time and high E/e′, it is likely that he also had an S_3,[6] which is sometimes audible in patients with HCM.[7] The most striking finding in our case, however, was the low-pitched murmur between the pulmonary component of S_2 and S_3. Differential diagnoses for early diastolic murmurs include valvular heart disease, congenital heart disease, and arteriovenous fistula; in our patient, however, clinical and echocardiographic findings supporting these diagnoses were lacking. On detailed analysis of the phonocardiographic, apexcardiographic, and echocardiographic findings, we concluded that our patient's unusual murmur during early diastole was related to his diastolic paradoxical jet flow. To our knowledge, this is the first case in which heart murmurs associated with a diastolic paradoxical jet flow—or "paradoxical flow murmur"—has been clearly described.

The mechanism of a paradoxical jet flow is an increased pressure gradient between the high-pressure apical chamber and the low-pressure basal left ventricle during early diastole.[3] It is conceivable that flow through a narrowed midventricle could produce a high-pitched murmur, given the characteristics of heart murmurs: the greater the gradient, the higher the pitch of the murmur.[8] However, in our patient, a high-pitched ejection murmur was recorded during midsystole because of the midventricular obstruction, whereas the paradoxical flow murmur was low-pitched, even though the peak blood flow velocities of these 2 blood flows were almost identical. Although the mechanisms underlying our patient's condition are unknown, it is possible that the volume of the paradoxical jet flow was not large enough to produce high-energy turbulence. Another possibility is that the presence of an opposite-direction signal (that is, early mitral inflow) during early diastole and after isovolumic relaxation prevented the acceleration of flow.

A paradoxical flow murmur may not be easy to detect because it could occur in a limited area of the thoracic wall, as was the case in our patient. These paradoxical flow murmurs might also appear and disappear due to various factors, such as hemodynamics, heart rate, preload, and afterload.[9] In fact, our patient's murmur was not always audible during his follow-up visits. However,



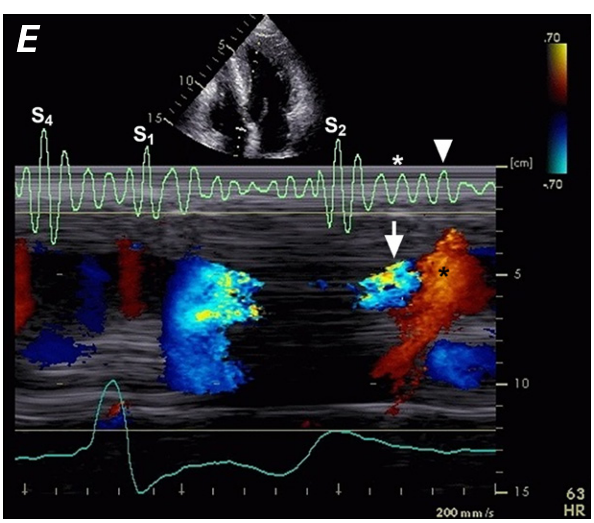


Fig. 2 *Transthoracic echocardiograms (apical 4-chamber views) show* ***A****) myocardial hypertrophy of the ventricular septum, the anterior wall of the left ventricle, and the apex of both ventricles at end-diastole, and* ***B****) midventricular obstruction with an apical chamber at end-systole (arrow).* ***C****) Color-Doppler M-mode image shows the near-absence of midventricular flow during late systole and the diastolic paradoxical jet flow from the apex toward the base of the left ventricle (arrow).* ***D****) Continuous-wave Doppler-mode image shows bimodal flows toward the base of the left ventricle with a notch between end-systole and early-diastole (arrowhead).* ***E****) Color-Doppler M-mode image and a phonocardiogram obtained at the apex show the first part of a diastolic murmur after* S_2 *(white asterisk) coinciding with the diastolic paradoxical jet flow (arrow) and the last part of the murmur (arrowhead) coinciding with the mitral E wave (black asterisk). The phonocardiogram is not identical to that shown in Figure 1 because it was recorded by using a simple phonocardiograph connected to the echocardiograph. The probe used to obtain the corresponding echocardiogram was not placed at the true apex because a microphone for phonocardiography was placed in that location.*

LA = left atrium; LV = left ventricle; RV = right ventricle the association between diastolic paradoxical jet flows and an increased risk of adverse outcomes has been reported,[3,4] so detecting paradoxical flow murmurs on auscultation is worthwhile, even in the era of advanced imaging techniques.

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