

Transient Midventricular Ballooning Syndrome

A New Variant

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We describe a new variant of transient left ventricular (LV) ballooning in North American Caucasian patients in which only the midventricle is affected. The patients described in this case series initially presented with emotional or physical stress and had similarities to transient apical ballooning syndrome; however, this variant is unique in that the transient ballooning involves the midventricle with hypercontractility of the apical and basal segments. The presentation, clinical features, and transient nature of the reported cases in this series are similar to transient LV apical ballooning and suggest a shared pathophysiologic etiology. Sparing of the apical segment with involvement of midventricle only supports etiologies not related to an epicardial coronary artery distribution. Although the pathophysiologic mechanism of the transient ventricular ballooning syndromes and other cases of catecholamine-associated transient ventricular dysfunction are not well understood, the emergence of this new variant raises further questions in the understanding of the “brain-heart” relationship. (J Am Coll Cardiol 2006;48:579–83) © 2006 by the American College of Cardiology Foundation

We report a new variant of transient left ventricular (LV) ballooning in 4 patients. Each case involves a female patient who presented with symptoms of acute coronary syndrome, a normal coronary angiogram, a left ventriculogram demonstrating midventricular dilation and akinesis with a hypercontractile apex and base, and a follow-up echocardiogram demonstrating resolution of the wall motion abnormality. These cases have similarities to apical ballooning syndrome; however, this variant is unique in that the transient ballooning involves the midventricle with sparing of the apical and basal segments.

CASE 1

A 69-year-old Caucasian woman with a history of hypertension but no clinical history of coronary artery disease presented to the emergency department (ED) with substernal chest pain. She was 1-day postoperative from shoulder arthroscopy with subacromial decompression and had been experiencing significant postprocedure discomfort. The electrocardiogram had minimal nonspecific T-wave changes when compared with a preprocedure electrocardiogram. Cardiac enzymes, including a troponin T, were mildly elevated. Cardiac catheterization was performed and showed angiographically normal epicardial coronary arteries; however, the left ventriculogram demonstrated midventricular dilation and akinesis with a hypercontractile apex and base (Figs. 1A and 1B, Video 1 [please see the

Appendix]). A transthoracic echocardiogram performed 3 days later revealed a normal LV size and function with no regional wall motion abnormalities (Video 2 [please see the Appendix]).

CASE 2

A 49-year-old Caucasian woman with no cardiac history or risk factors for atherosclerosis presented to the ED with acute substernal chest pain. Her symptoms began during a vigorous exercise session. The patient also noted being under a tremendous amount of work-related stress during this time. There was no history of illicit drug use, and our pertinent review of systems was negative. Her blood pressure (BP) at admission was 132/94 mm Hg. The initial electrocardiogram was normal. Troponin T level was mildly elevated, with a normal creatine kinase and MB fraction. A transthoracic echocardiogram showed dilation and akinesis of the midventricle with a hypercontractile apex and base (ejection fraction [EF] 40%) (Figs. 2A and 2B, Video 3 [please see the Appendix]). The epicardial coronary arteries were angiographically normal; however, the left ventriculogram demonstrated midventricular dilatation and akinesis with a hypercontractile apex and base (Figs. 3A and 3B, Video 4 [please see the Appendix]). She was initiated on angiotensin-converting enzyme (ACE) inhibitor and beta-blocker medication; however, she did not tolerate either medication because of symptomatic hypotension and bradycardia. A transthoracic echocardiogram 3.5 months later showed normal LV size and function (EF 60%) and no regional wall motion abnormalities. The patient has remained asymptomatic in 30 months of follow-up.

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Abbreviations and Acronyms

ACE = angiotensin-converting enzyme
LV = left ventricular

CASE 3

A 37-year-old Caucasian woman with no cardiac history presented to the ED with substernal chest pain. The symptoms began during a public speaking presentation for which she had experienced significant anxiety. There was no history of illicit drug use, and a pertinent review of systems was negative. Her BP was 110/63 mm Hg. Serial electrocardiograms demonstrated new inverted T waves in the precordial leads without ST-segment deviation. Cardiac biomarkers were significant for a mild elevation in troponin T. The patient underwent cardiac catheterization for unremitting substernal chest discomfort. The epicardial coronary

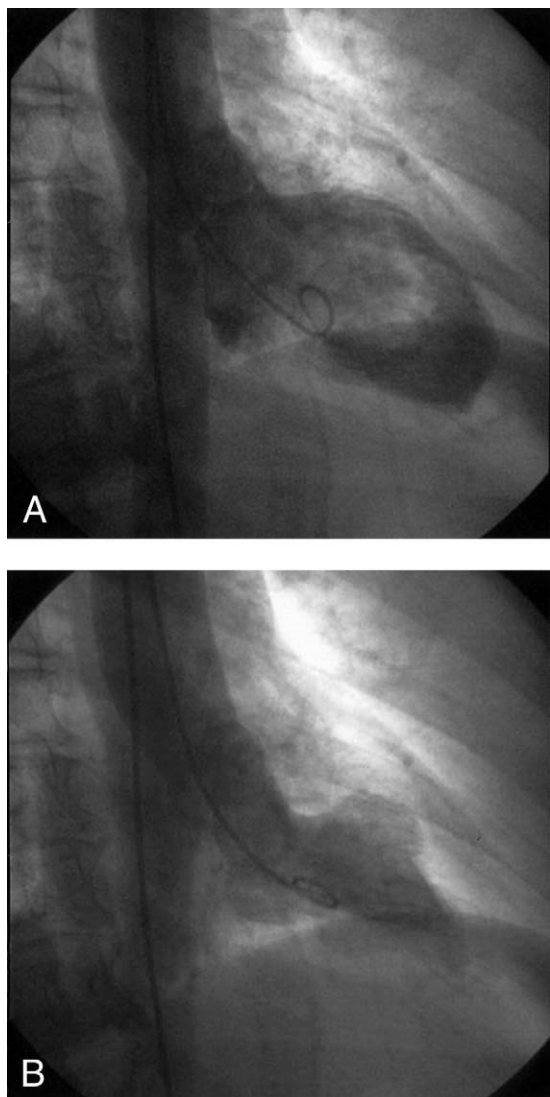


Figure 1. Left ventriculogram in right anterior oblique view of Case 1 in diastole (A) and systole (B). Note the hypercontractility of the basal and apical segments and ballooning of the midventricular segments.

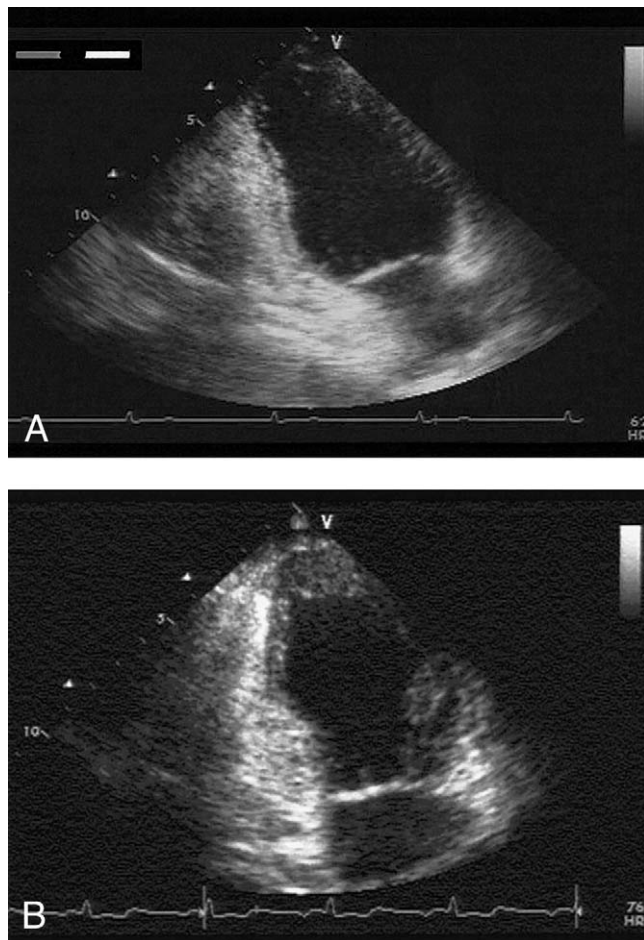


Figure 2. Echocardiographic still frame of the left ventricle of Case 2. The apical 2-chamber view of the left ventricle is shown in diastole (A) and systole (B). In systole, note the ballooning of the midventricle only.

arteries were angiographically normal. The left ventriculogram showed a hypercontractile base and apex with a dilated and akinetic midventricle (Figs. 4A and 4B, Video 5 [please see the Appendix]). A transthoracic echocardiogram obtained 1 year later showed normal LV size and function. The EF was 62%, with no regional wall motion abnormalities. The patient was initiated on ACE inhibitor and beta-blocker therapy. The patient has had infrequent episodes of atypical chest pain in 2 years of follow-up, none similar to the initial presentation.

CASE 4

A 55-year-old Caucasian woman with hyperlipidemia and a family history of premature coronary artery disease had significant anxiety related to her responsibilities in organizing a social event. She developed a “tingling” sensation and then experienced an out-of-hospital cardiac arrest requiring bystander cardiopulmonary resuscitation and defibrillation. She was transported to the nearest ED, where an electrocardiogram was notable for ST-segment elevation in the inferior leads and cardiac enzymes, including troponin T, were elevated. Her BP was 122/68 mm Hg. Thrombolytics were administered, and the patient was emergently trans-

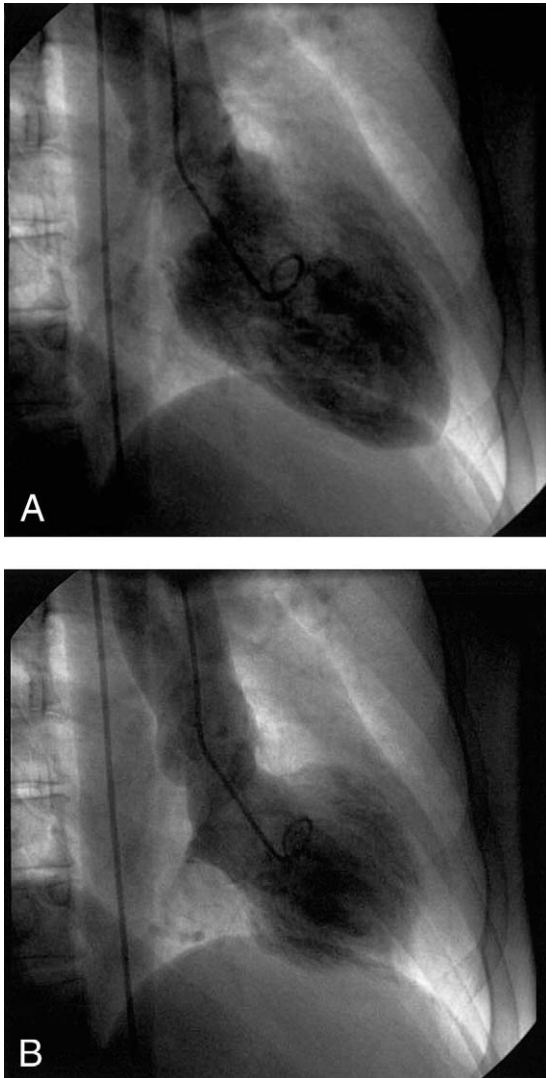


Figure 3. Left ventriculogram in right anterior oblique view of Case 2 in diastole (A) and systole (B). Note the hypercontractility of the basal and apical segments and ballooning of the midventricular segments.

ferred. An echocardiogram was performed on admission, which demonstrated regional wall motion abnormalities and an estimated LV EF of 35%. Emergency coronary angiography revealed normal epicardial coronary arteries. The left ventriculogram demonstrated a hypercontractile base and apex with midventricular akinesia (Figs. 5A and 5B, Video 6 [please see Appendix]). She was managed with conservative medical therapy using beta-blockers and ACE inhibitors. A transthoracic echocardiogram obtained 4 weeks later showed normal LV size with recovery of function (EF 57%) and marked improvement of the midventricular wall motion abnormalities. The patient has not experienced any subsequent cardiac events after more than 4 months of follow-up.

DISCUSSION

We report the first series of 4 cases of transient midventricular ballooning syndrome in Western literature. Our literature search revealed only 2 similar cases of transient midventricular ballooning in the Japanese literature (1,2).

This new variant of LV ballooning has unique features on echocardiography and left ventriculography that distinguish it from LV apical ballooning in that the apex and base are hypercontractile whereas the midventricle segments show akinesia and ballooning. The distribution of regional wall motion abnormalities does not follow epicardial coronary vascularity and, much like LV apical ballooning, the ventricular dysfunction is transient.

There has been much speculation regarding the etiology of transient LV apical ballooning. Although initially recognized in the Japanese population and described as “takotsubo-type cardiomyopathy” (3,4) (named for the similarities in appearance of the left ventriculogram during systole to the “short neck round-flask” appearance of the Japanese fishing pot used for trapping octopus) (5,6), this syndrome has subsequently been well-described in Europe and the U.S. (7–10). Currently the term “transient left ventricular apical ballooning” is commonly used to describe this syndrome. A recent review has been published outlining

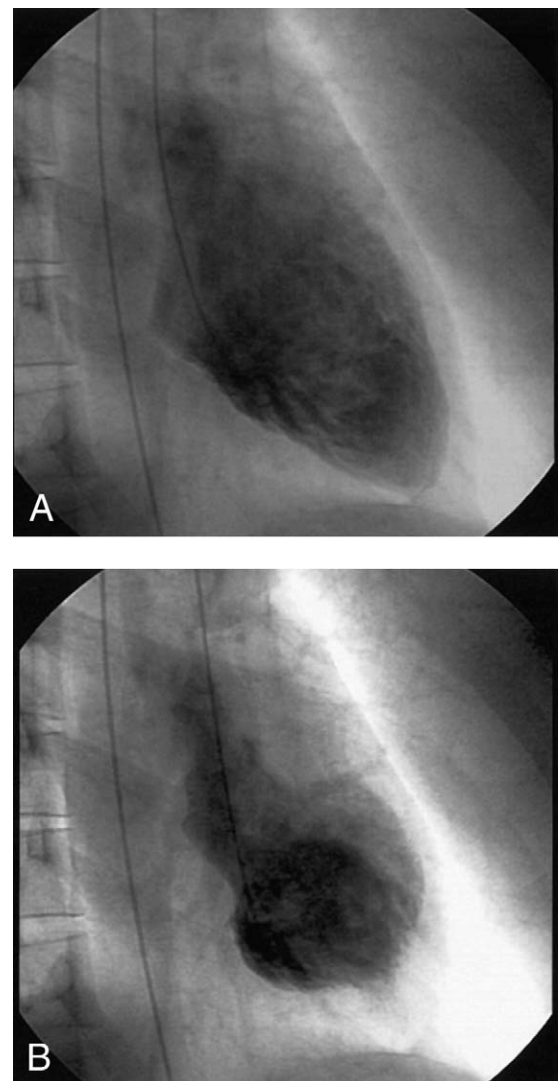


Figure 4. Left ventriculogram in right anterior oblique view of Case 3 in diastole (A) and systole (B). Note the hypercontractility of the basal and apical segments and ballooning of the midsegments.

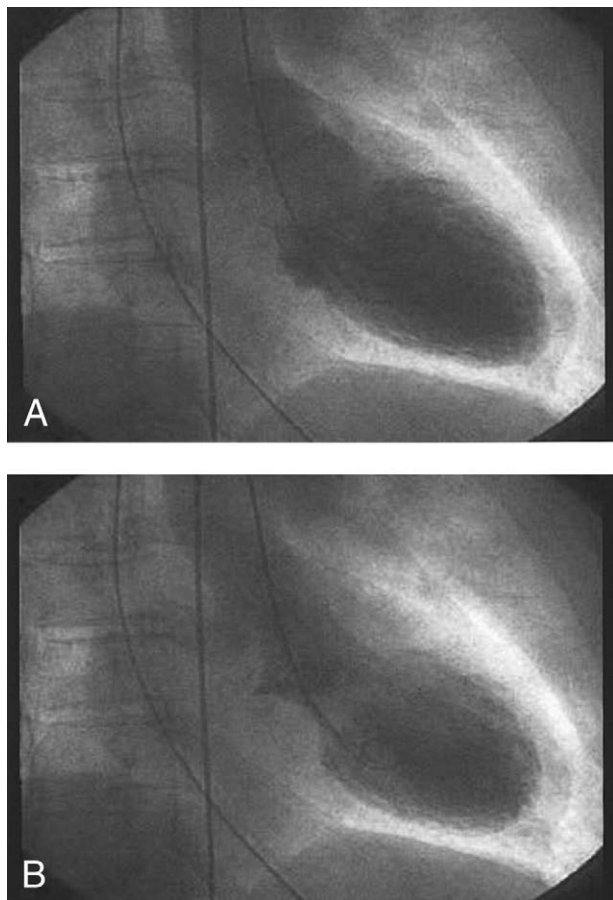


Figure 5. Left ventriculogram in right anterior oblique view of Case 4 in diastole (A) and systole (B). Note the hypercontractility of the basal and apical segments and ballooning of the midsegments.

clinical features, findings, and potential pathophysiologic mechanisms (11). Multivessel epicardial spasm, coronary microvascular dysfunction or spasm, acute coronary syndrome with reperfusion, impaired fatty acid metabolism, myocarditis, transient obstruction to LV outflow, and catecholamine-mediated myocardial dysfunction all have been proposed as potential mechanisms (5,12–18).

The presentation, clinical features, and transient nature of the reported cases in our series are similar to transient LV apical ballooning and suggest a shared pathophysiologic etiology. As previously described with LV apical ballooning (5,7,8,10) and also observed in this series of patients, there is a strong preponderance of women. The seemingly increased susceptibility of women to stress-related LV dysfunction and potential gender-related differences in response to catecholamines is not well understood (10). Why only the midventricle is involved is an intriguing observation without a clear explanation. We can only speculate that the differences in wall motion abnormalities between apical and midventricular ballooning in our patients may reflect a temporal variation in resolution in which the apical ballooning has recovered by the time of angiographic diagnosis. However, our patients underwent coronary angiography and left ventriculography within 24 h (and 3 of the 4 at <12 h) of symptom onset, making it unlikely for such rapid apical

recovery to not only normalize but to demonstrate hypercontractility. Transient midventricular ballooning syndrome is a clinical entity that is probably a variant of apical ballooning syndrome. The prognostic implications of midventricular ballooning as compared with apical ballooning are unknown; however, 2 of these patients have been followed for longer than 2 years, with no recurrence or demonstrable morbidity.

The transient myocardial “stunning” observed with LV ballooning, the myocardial dysfunction seen with catecholamine excess related to pheochromocytoma, and the neurally mediated “neurogenic stunned myocardium” are suggestive of a common mechanism. Given the typical clinical scenario of an associated emotional or physical stressor in these patients, many researchers believe that the complex interaction between catecholamines and the adrenergic innervation of the heart is largely responsible. Catecholamines can have a toxic effect on cardiac myocytes (19,20) and animal models have suggested that activation of alpha- and beta-adrenoreceptors are the primary trigger of emotional stress-induced cardiac changes (12,13,21). Perhaps the variations in regional wall motion observed in transient LV apical ballooning, transient midventricular ballooning, and other entities involving excess catecholamines (pheochromocytoma and neurocardiogenic injury) relate more to differences in the anatomic location of cardiac adrenergic receptors, the degree of excess sympathetic activity involved, or differing susceptibilities to such sympathetic stimulation from individual to individual. Although the pathophysiologic mechanism of the transient ventricular ballooning syndromes and other cases of catecholamine-associated transient ventricular dysfunction are not well understood, future research will likely enhance our understanding of this complex “brain-heart” relationship.

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APPENDIX

For accompanying videos, please see the online version of this article.