Reversible Left Ventricular Trabeculations in Pregnancy
Is This Sufficient to Make the Diagnosis of Left Ventricular Noncompaction?

Sharon C. Reimold, MD

Left ventricular noncompaction cardiomyopathy (LVNC) is a disorder characterized by significant ventricular trabeculations on cardiovascular imaging. The most common echocardiographic criteria used to define the disorder are the Jenni et al.¹ and Chin et al.² criteria. These criteria were developed after the observation that some patients, primarily children, had cardiomyopathies characterized by significant trabeculations associated with recesses. These criteria differ in that the Jenni et al. criteria focus on comparing the noncompacted and compacted myocardium at end systole, whereas the Chin et al. criteria compare the noncompacted and total myocardial thickness at end diastole. Diagnosis of noncompaction based on imaging studies is variable but in 1 large review was estimated to involve 0.24% of the general population.³ Trabeculations fulfilling the criteria for LVNC have been seen in a variety of individuals, including patients with sickle cell anemia, athletes, pregnant women, and patients with dilated cardiomyopathy.⁴⁻⁵ In a retrospective evaluation of patients with systolic heart failure, the number of patients satisfying the criteria for LVNC far exceeds (23.6%) that reported in the general population.⁶

Why are these trabeculations seen? Pregnancy is associated with increased preload and pregnancy are associated with increased preload, Gati and colleagues⁷ believe that increased preload may be the common pathway mediating the development of trabeculations. They hypothesized that women would develop trabeculations during pregnancy and that these trabeculations would resolve in the postpartum period. These investigators studied 102 women with morphologically normal-appearing hearts by echocardiography in the first trimester of pregnancy. These women were followed up throughout pregnancy with additional 2-dimensional echocardiograms obtained during the third trimester and postpartum. A quarter of the women (26 of 102) developed new trabeculations late in pregnancy. These trabeculations resolved in the majority of these women early postpartum; however, in 7 of the 26 women, the trabeculations persisted. None of these women had clinical evidence of cardiac dysfunction or had evidence of systolic or diastolic dysfunction on echocardiography.

It is important to assess how likely it is for these echocardiographic assessments to be accurate. The diagnosis of increased trabeculations was based on serial echocardiographic images rather than cardiac magnetic resonance imaging given the pregnancies. Echocardiographic images may be technically more difficult for serial evaluation than cardiac magnetic resonance imaging, but the frequency of the observation makes it likely that the development of increased trabeculations in pregnancy is a real finding. The investigators have extensive experience in assessing cardiac morphology, and they demonstrate reasonable interobserver and intraobserver variability, increasing the validity of their results. The majority of the women had resolution of trabeculations after delivery, so they satisfied the criteria for LVNC only during pregnancy. These patients are unlikely to have underlying LVNC because the trabeculations were not a persistent finding and their hearts otherwise appear structurally normal.

As explained in this issue of Circulation, because these disorders and pregnancy are associated with increased preload, it is important to assess how likely it is for these echocardiographic assessments to be accurate. The diagnosis of increased trabeculations was based on serial echocardiographic images rather than cardiac magnetic resonance imaging given the pregnancies. Echocardiographic images may be technically more difficult for serial evaluation than cardiac magnetic resonance imaging, but the frequency of the observation makes it likely that the development of increased trabeculations in pregnancy is a real finding. The investigators have extensive experience in assessing cardiac morphology, and they demonstrate reasonable interobserver and intraobserver variability, increasing the validity of their results. The majority of the women had resolution of trabeculations after delivery, so they satisfied the criteria for LVNC only during pregnancy. These patients are unlikely to have underlying LVNC because the trabeculations were not a persistent finding and their hearts otherwise appear structurally normal.

Why are these trabeculations seen? Pregnancy is associated with an increase in chamber sizes and volumes.⁸ Increased chamber volumes occur in response to the hormonal changes associated with pregnancy. It is possible that increases in left ventricular volumes are associated with localized changes in stress on the myocardium, which cause muscle bundles to become apparent on echocardiograms. When chamber preload decreases after delivery, these small muscle bundles may no longer protrude into the left ventricular chamber. The explanation for the failure of trabecular regression in a small subset of women postpartum is not clear. In pressure overload of the left ventricle, we know that effective treatment of hypertension or critical aortic stenosis is associated with a change in left ventricular morphology and a decrease in left ventricular mass.⁹⁻¹⁰ The degree of regression of left ventricular mass is not uniform in these populations. It is possible that there is variability in the regression of trabeculations in pregnancy similar to the variation in the reduction of left ventricular mass in effectively treated pressure-overload states.

LVNC is more common in individuals with neuromuscular and congenital cardiac disorders. A variety of genes have been associated with the development of LVNC. These include mutations in, CSX, DMPK, dystrophin, mitochondrial DNA, G4.5, and α-dystrobrevin.¹¹⁻¹³ Individuals with a variety of neuromuscular disorders, including Becker muscular dystrophy, Friedreich ataxia, myotonic dystrophy, mitochondriopathy, and myoadenylate deaminase deficiency, may have echocardiographic phenotypes consistent with LVNC.¹⁴⁻¹⁵ In patients with morphologically normal-appearing hearts except for trabeculations, it is unknown
whether genetic variation plays a role in the appearance of trabeculations. Gati and colleagues also noted that trabeculations were seen more commonly in black women. Trabeculations have been reported more commonly in black individuals with heart failure. This supports the concept that there may be genetic variation in the response of the myocardium to volume and pressure overload. Whether this variation results in different long-term clinical outcomes is not known.

This article highlights the need to revise the current criteria for LVNC. These criteria were initially developed to categorize individuals with a poorly defined cardiomyopathy. LVNC is most typically associated with left ventricular systolic dysfunction, arrhythmias, and cardioembolic events resulting from thrombi. With the improved delineation of left ventricular endocardium by echocardiographic and cardiac magnetic resonance imaging methods, left ventricular trabeculations are identified in a variety of disorders, increasing the frequency of diagnosing LVNC. Most of these patients do not exhibit the common clinical presentations of patients with LVNC. In addition, there is poor correlation between the various cardiovascular imaging criteria (the Chin et al and Jenni et al criteria used in this article and the Stölzblegger criteria). Thus, the current criteria are likely significantly overly sensitive for the diagnosis of LVNC.

In addition, the observations of Gati et al highlight the ability of the left ventricle to undergo morphological changes in response to changes in preload. There is very little understanding of the mechanism of these changes. Future directions not only should focus on redefining the criteria of LVNC but also should determine the mechanism for the development of trabeculations in otherwise normal hearts. In addition, it will be important to determine whether there is a relationship between trabeculations and long-term clinical outcomes.

Disclosures

None.

References


Key Words: Editorials ■ cardiomyopathy, dilated with left ventricular noncompaction ■ pregnancy
Reversible Left Ventricular Trabeculations in Pregnancy: Is This Sufficient to Make the Diagnosis of Left Ventricular Noncompaction?
Sharon C. Reimold

Circulation. 2014;130:453-454; originally published online July 8, 2014;
doi: 10.1161/CIRCULATIONAHA.114.011481
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2014 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/130/6/453

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org/subscriptions/