Reversible Left Ventricular Trabeculations in Pregnancy:

Is This Sufficient to Make the Diagnosis of Left Ventricular Noncompaction?

Running title: Reimold; Pregnancy and Trabeculations

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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 and Chin 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 criteria focus on comparing the noncompacted to compacted myocardium at end-systole whereas the Chin criteria compare the noncompacted to total myocardial thickness at end-diastole. Diagnosis of noncompaction based on imaging studies is variable but in one large review was estimated to involve 0.24% of the general population.

Trabeculations fulfilling the criteria for LVNC have been seen in individuals with a variety of different disorders including sickle cell anemia, athletes, pregnancy, and dilated cardiomyopathy. In a retrospective evaluation of patients with systolic heart failure, patients fulfilling criteria for LVNC far exceed (23.6%) that reported in the general population.

In this issue of *Circulation*, since these disorders and pregnancy are associated with increased preload, Gati and colleagues 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 throughout pregnancy with additional 2-dimensional echocardiograms obtained during the third trimester and post-partum. A quarter of the women (n=26/102) 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 (CMR) given the pregnancies. Echocardiographic images may be technically more difficult for serial evaluation than CMR 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 inter- and intra-observer variability, increasing the validity of their results. The majority of the women had resolution of trabeculations after delivery, so they satisfied criteria for LVNC only during pregnancy. These patients are unlikely to have underlying LVNC since 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. 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 causes 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. An 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 are associated with a change in left ventricular morphology and a decrease in left ventricular mass. The degree of regression of left ventricular mass is not uniform in these
populations. It is possible that there is variability in regression of trabeculations in pregnancy similar to variation in 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 has been associated with the development of LVNC. These include mutations in CSX, DMPK, Dystrophin, mitochondrial DNA, G4.5, and alpha-dystrobrevin. 12, 13 Individuals with a variety of neuromuscular disorders including Becker’s muscular dystrophy, Friedreich’s 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. 8 Trabeculations have also been reported more commonly in black individuals with heart failure. 7 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 manuscript 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 due to thrombi. 16 With the improved delineation of left ventricular endocardium by echocardiographic and CMR 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 (Chin and Jenni
used in this manuscript and Stollberger)\textsuperscript{7,17,18} 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 morphologic changes in response to changes in preload. There is very little understanding of the mechanism of these changes. Future directions should not only focus on redefining the criteria of LVNC but should also determine the mechanism for 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.

**Conflict of Interest Disclosures:** None.

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