Response to Letter Regarding Article, “Reversible De Novo Left Ventricular Trabeculations in Pregnant Women: Implications for the Diagnosis of Left Ventricular Noncompaction in Low-Risk Populations”

We are honored by the interest shown by Stöllberger and Finsterer in our article. The aim of our study was to demonstrate that a significant proportion of low-risk individuals were capable of developing de novo left ventricular (LV) trabeculations in response to a physiological increase in cardiac workload. In our study, 25% of women developed de novo trabeculations, and 8% fulfilled criteria for LV noncompaction during pregnancy. The observations highlighted that current diagnostic criteria for LV noncompaction are nonspecific and should not be applied to asymptomatic patients with normal LV function.

One possible explanation for our results is that women who developed the greatest increases in LV size may have shown appearances consistent with trabeculations merely as a result of a stretch effect; however, we did not find any differences in LV size between women with and without trabeculations. Stöllberger and Finsterer raise some pertinent issues that may have affected our results. It is possible that differences in hemoglobin and estrogen/progesterone concentrations occurred in the absence of neuromuscular disease. For example, 8% of nationally ranked British athletes show similar findings, and nobody would doubt that these medalists with extraordinary physical ability are free of neuromuscular disease.

The authors seem obsessed with the association between LV trabeculations and several extremely rare neuromuscular diseases. The bigger picture suggests that in most instances such trabeculations occur in the absence of neuromuscular disease. For example, 8% of individuals with sickle cell disease, arrhythmias, stroke, and neurological disease, but the relatives of the pregnant women with increased LV trabeculations were not systematically investigated to rule out a hereditary cause in relatives of the pregnant women with increased LV trabeculations. One possible explanation for our results is that women who developed de novo LV trabeculations may have led to some women developing trabeculations, but we do not have the data to confirm or refute this theory.

The authors seem obsessed with the association between LV trabeculations and several extremely rare neuromuscular diseases. The bigger picture suggests that in most instances such trabeculations occur in the absence of neuromuscular disease. For example, 8% of nationally ranked British athletes show similar findings, and nobody would doubt that these medalists with extraordinary physical ability are free of neuromuscular disease.

The women recruited to this study did not experience overt neuromuscular disorders, although we concede that we did not conduct electromyograms, nerve conduction tissue studies, or genetic testing. We pursued detailed questioning about a family history of premature cardiac disease, arrhythmias, stroke, and neurological disease, but the relatives of the pregnant women with increased LV trabeculations were not systematically investigated to rule out a hereditary cause in the absence of overt disease in the pregnant woman.

We included women with a completely normal echocardiogram during the first trimester to ascertain whether our hypothesis was correct. It would have been difficult to recruit women before pregnancy for this study because of uncertainties about the time frame to conception. All women, including those with LV trabeculations, had a successful pregnancy. We did not test the babies for LV noncompaction because, as far as we are concerned, we have proven that LV trabeculations can be acquired from increased loading conditions in asymptomatic individuals. We are hopeful that our results will question the specificity of current diagnostic criteria, which have huge limitations, and will prevent overdiagnosis of an otherwise potentially sinister cardiomyopathy. We have greatest concerns about the Jenni criteria, which were poorly reproducible. We did not use the Stöllberger criteria because they have been revised on several occasions but have never been validated.

The aim of our study was to prove that trabeculations could be acquired rather than dispelling the idea that LV noncompaction is indeed a primary myocardial disorder.

Disclosures

None.

References


Response to Letter Regarding Article, "Reversible De Novo Left Ventricular Trabeculations in Pregnant Women: Implications for the Diagnosis of Left Ventricular Noncompaction in Low-Risk Populations"
Sabiha Gati, Michael Papadakis, Nikolaus D. Papamichael, Abbas Zaidi, Nabeel Sheikh, Matthew Reed, Rajan Sharma, Baskaran Thilaganathan and Sanjay Sharma

Circulation. 2015;131:e426
doi: 10.1161/CIRCULATIONAHA.114.014028

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2015 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/131/18/e426

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/