

# Can Left Ventricular Noncompaction Be Acquired, and Can It Disappear?

Paolo Angelini, MD

**T**he report by Papadopoulos and colleagues<sup>1</sup> is interesting because it reopens an unfinished discussion about left ventricular noncompaction (LVNC): can de novo LVNC appear after birth and then disappear?

Authors typically refer to LVNC as a rare cardiomyopathy; however, this conclusion is premature for 2 reasons. First, echocardiography, by means of which LVNC was initially described, is not well suited for viewing soft-bordered apical anomalies, such as LVNC in adult patients. Second, more accurate imaging methods, such as cardiac magnetic resonance (CMR), have revealed a 6% to 20% prevalence of LVNC in large populations.<sup>2,3</sup>

Pediatric cardiologists in hospital-based cardiomyopathy clinics were the first to become aware of LVNC. In this setting, LVNC is typically observed in the context of complex congenital heart malformations, dilated cardiomyopathy, and congestive heart failure, implying a preliminary entry bias. Subsequently, a prevalence of 0.1% to 0.3% was reported in hospital-based echocardiography laboratories.<sup>4-6</sup> Later, during large-population echocardiographic screenings (in studies of sudden cardiac death in the young), LVNC's prevalence ranged from 0 to 0.1%.<sup>7,8</sup> More recently, investigators have used CMR—a method more accurate than echocardiography for detecting LVNC<sup>9</sup>—and found a much higher prevalence of LVNC in large general populations.<sup>2,3</sup> The Multi-Ethnic Study of Atherosclerosis,<sup>2</sup> an epidemiologic multiyear project involving adults, included a study of LVNC in 1,000 individuals; the investigators reported a 6% prevalence of LVNC (using the criterion of “more than one myocardial segment involved”). Screen to Prevent,<sup>3</sup> a study of 5,130 subjects 10 to 19 years of age, reported a surprising prevalence of 19.9%, with strict use of Petersen and colleagues' criteria in CMR diagnosis.<sup>9</sup> In Screen to Prevent, most LVNC carriers had no symptoms or abnormal left ventricular (LV) function: only 3 of 982 individuals had dilated cardiomyopathy (0.3%).

By itself, LVNC does not imply dilated cardiomyopathy; the latter is an exception in young patients. Coexisting congenital cardiomyopathy is rare and probably arises from additional genetic mutations to LVNC. However, more studies are needed to evaluate long-term changes, especially in children.

## What Is the Nature of Papadopoulos and Colleagues' Case?

Papadopoulos and colleagues' case<sup>1</sup> was clearly one of LVNC and was accurately evaluated by means of CMR at the patient's clinical presentation and 2-year follow-up. In contrast, the echocardiographic findings suggested “appearing and disappearing” LVNC associated with transient dilated cardiomyopathy. Initially, the authors considered dilated cardiomyopathy possibly intrinsic to LVNC. Indeed, the new onset of diffuse, severe cardiomyopathy (LV ejection fraction, 0.15–0.25) in a middle-aged, previously asymptomatic patient suggested either reversible myocarditis or takotsubo cardiomyopathy (the most typical transient cardiomyopathy that has segmental distribution and rapidly evolving features). A final diagnosis was not available. Severe cardiomyopathy was not a clinical issue after 2 years, when the patient's LV ejection fraction had improved to 0.60; however, LVNC remained.

Reports<sup>9-14</sup> of seemingly reversible LVNC in pregnant women and athletes<sup>10-14</sup> have evoked claims that LVNC is inducible in adults and is curable when predisposing

**From:** Department of Cardiology, Texas Heart Institute, Houston, Texas 77030

**Address for reprints:**  
Paolo Angelini, MD,  
Texas Heart Institute,  
6624 Fannin St., Suite 2780,  
Houston, TX 77030

**E-mail:** pangellini@leachmancardiology.com

© 2017 by the Texas Heart® Institute, Houston

conditions (such as pregnancy or strenuous exercise) disappear. Some authors have conjectured that a latent cardiomyopathy is implied in LVNC. Most likely, milder forms of LVNC go undetected by imaging methods of inferior precision, yielding the presumption that LVNC appears during the pregnancy or strenuous physical training and disappears afterwards. However, this occurs only because LVNC becomes more visible on echocardiography in the presence of an acquired and transient LV dilation. Conversely, CMR would reveal LVNC's features at any point. One lesson from the present case<sup>1</sup> is that more accurate imaging is needed to detect LVNC, especially when echocardiographic windows are poor. The authors proved this well.

I also exclude the biological possibility of de novo LVNC and its later disappearance because the structural changes in LVNC (intertrabecular recesses with endothelial lining, and trabecular structures with peculiar myocardial vascularization<sup>15</sup>) are not reversible after birth. The compacted myocardium in LVNC is approximately 30% thinner than normal.<sup>2,3,15</sup> Accordingly, although transient or irreversible dilative evolution can develop during the life of a patient with LVNC, in all probability this congenital anomaly will never disappear.

## References

1. Papadopoulos K, Petrou PM, Michaelides D. Left ventricular noncompaction cardiomyopathy presenting with heart failure in a 35-year-old man. *Tex Heart Inst J* 2017;44(4):260-3.
2. Kawel N, Nacif M, Arai AE, Gomes AS, Hundley WG, Johnson WC, et al. Trabeculated (noncompacted) and compact myocardium in adults: the multi-ethnic study of atherosclerosis. *Circ Cardiovasc Imaging* 2012;5(3):357-66.
3. Angelini P, Uribe C, Masso A, Lopez A, Ali S, Willerson JT. Results of MRI-based screening study of 5130 candidates for sports participation [abstract]. Available from: <http://abstracts.online.com/pp8/#!/4223/presentation/38112> [cited 2017 Aug 2].
4. Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium. A study of eight cases. *Circulation* 1990;82(2):507-13.
5. Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol* 2000;36(2):493-500.
6. Pignatelli RH, McMahon CJ, Dreyer WJ, Denfield SW, Price J, Belmont JW, et al. Clinical characterization of left ventricular noncompaction in children: a relatively common form of cardiomyopathy. *Circulation* 2003;108(21):2672-8.
7. Zeltser I, Cannon B, Silvana L, Fenrich A, George J, Schleifer J, et al. Lessons learned from preparticipation cardiovascular screening in a state funded program. *Am J Cardiol* 2012;110(6):902-8.
8. Grazioli G, Merino B, Montserrat S, Vidal B, Azqueta M, Pare C, et al. Usefulness of echocardiography in preparticipation screening of competitive athletes. *Rev Esp Cardiol (Engl Ed)* 2014;67(9):701-5.
9. Petersen SE, Selvanayagam JB, Wiesmann F, Robson MD, Francis JM, Anderson RH, et al. Left ventricular non-compaction: insights from cardiovascular magnetic resonance imaging. *J Am Coll Cardiol* 2005;46(1):101-5.
10. Gati S, Papadakis M, Papamichael ND, Zaidi A, Sheikh N, Reed M, et al. Reversible de novo left ventricular trabeculations in pregnant women: implications for the diagnosis of left ventricular noncompaction in low-risk populations. *Circulation* 2014;130(6):475-83.
11. Coris EE, Moran BK, De Cuba R, Farrar T, Curtis AB. Left ventricular non-compaction in athletes: to play or not to play. *Sports Med* 2016;46(9):1249-59.
12. Finsterer J, Stollberger C. Regression of left ventricular hypertrabeculation/noncompaction. *J Cardiol* 2016;68(4):357-8.
13. Gati S, Chandra N, Bennett RL, Reed M, Kervio G, Panoulas VF, et al. Increased left ventricular trabeculation in highly trained athletes: do we need more stringent criteria for the diagnosis of left ventricular non-compaction in athletes? *Heart* 2013;99(6):401-8.
14. D'Ascenzi F, Pelliccia A, Bonifazi M, Mondillo S. Left ventricular non-compaction and hypertrabeculation in the athlete: distinguishing between pathology and physiology. *Int J Cardiol* 2015;190:122-3.
15. Burke A, Mont E, Kutys R, Virmani R. Left ventricular non-compaction: a pathological study of 14 cases. *Hum Pathol* 2005;36(4):403-11.