Left ventricular noncompaction (LVNC) is thought to be a rare type of cardiomyopathy likely because of the arrest of myocardial compaction during the fifth to eighth weeks of gestation. The early myocardium consists of deep trabeculations along the endocardium, made of a spongy meshwork of myocardial fibers, that compact to form the capillary network and myocardium of the mature heart, normally progressing from base to apex and from epicardium to myocardium. Many are of the opinion that LVNC is a result of failure to complete this process, though this has been disputed by others who report that LVNC sometimes presents in patients who have had prior normal echocardiograms that did not demonstrate LVNC.¹

To date, the diagnostic criteria of LVNC is largely based on 2-dimensional transthoracic echocardiogram. Typical echocardiographic findings include a 2-layered appearance of the myocardium with a thin outer layer representing the compact myocardium, and a thicker inner layer representing the noncompacted trabeculum, in continuity with the endocardial surface. Three sets of diagnostic criteria have been proposed.

Criteria proposed by Chin et al include: the compact layer to total myocardial thickness ratio is <0.5 in end-diastole in apical or parasternal short axis views. Criteria proposed by Jenni et al include: the ratio of noncompacted to compacted myocardium in parasternal short axis view is >2 at end systole with prominent trabeculations and deep intertrabecular spaces that are perfused by intraventricular blood demonstrated by Doppler imaging, in the absence of other congenital heart disease. Finally, criteria proposed by Stollberger et al include: >3 trabeculations protruding from the left ventricular free wall apically to the papillary muscles, with the intertrabecular spaces perfused from the left ventricular cavity demonstrated by Doppler imaging. However, Kohli et al have called these criteria into question, with a study that showed that only 30% of LVNC met the requirements of all 3 diagnostic approaches.² The clinical utility of cardiac MRI in this setting has yet to be rigorously evaluated. There is some evidence that cardiac MRI provides more detailed morphological data in these patients.³ We present 2 cases of LVNC with atypical left ventricular morphologies in which several imaging modalities were used to assess the appearance of an accessory chamber.

Case 1
A 16-year-old boy treated with a β-blocker for hypertension presented with an episode of chest pressure associated with palpitations shortly after performing in his school’s marching band. The patient denied syncope, dyspnea, or

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exertional symptoms. Pertinent additional past medical history included supraventricular tachycardia that resolved in infancy and an echocardiogram performed in infancy demonstrating mild branch pulmonary stenosis and an atrial septal defect that spontaneously closed. Family history included 2 paternal great-uncles who died suddenly. His examination was normal. A 24-hour Holter monitor was placed, which showed frequent ventricular ectopy and a short run of non-sustained ventricular tachycardia. A follow-up echocardiogram revealed a noncompacted myocardium with mildly depressed systolic function and a left ventricular outpouching forming an accessory chamber (Figure 1 and Clips 1–3 in the online-only Data Supplement). MRI demonstrated a particularly prominent trabeculation forming a muscular band that bisected the left ventricle (Figure 2). Lateral to this band the free wall appeared thin, with dyskinetic movement, giving the appearance of a left ventricular aneurysm. However, unlike aneurysmal tissue, this section of myocardium was viable, without any perfusion deficit or delayed gadolinium enhancement. The patient underwent an electrophysiology study that demonstrated inducible ventricular tachycardia, although not located in the accessory chamber. An implantable cardioverter defibrillator was placed, and β-blockade was continued.

Case 2

A newborn boy initially presented with tachypnea and was diagnosed with an abnormal mitral valve and moderate mitral regurgitation. Over the next 3 years, he developed severe left atrial enlargement, evidence of restrictive physiology, mildly depressed left ventricular systolic function, and an unusual network of trabeculations in the left ventricular apex that formed a hypertensive accessory chamber within the left ventricle (Figure 3 and Clips 4–7 in the online-only Data Supplement). Hemodynamic assessment by catheterization demonstrated a pressure gradient in systole from the chamber to the left ventricle of $120 \text{ mm Hg}$ (Figure 4A and 4B). The main chamber left ventricular end diastolic pressure was $30 \text{ mm Hg}$. Reactive pulmonary hypertension developed and the patient underwent orthotopic heart transplantation at the age of 5 years (Figure 5).

Discussion

LVNC can pose a diagnostic challenge because of differing diagnostic criteria and because of its phenotypic variability, which can overlap with other forms of cardiac pathology. It has been described with congenital heart disease and can be mistaken for hypertrophic or dilated cardiomyopathy, layered mural thrombus, or mycotic invasion of the heart.4 We present 2 cases of LVNC in which accessory chambers formed. Both cases otherwise met each of the 3 proposed sets of diagnostic criteria. In the first case, an outpouching of the inferior wall of the left ventricle developed, resulting in the appearance of an accessory chamber, with viable myocardial tissue. In the second case, a cavity within a relatively large trabecular recess developed, partially occluded by overlaying endocardial fibroelastic tissue. These cases add to the growing body of literature highlighting the variability in the morphological expression and clinical course of LVNC.

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Disclosures

None.

References


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