Aortic Root Replacement in a Patient With Left Ventricular Noncompaction

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We describe the case of a 57-year-old woman with noncompaction of the left ventricle and regurgitant bicuspid aortic valve who presented with progressive congestive heart failure and was successfully treated with aortic root replacement. The long-term outcome for these patients is poor because of progressive left ventricular impairment, increased rates of life-threatening arrhythmias, and intraventricular thrombi. To our knowledge, only 3 patients with noncompaction of the left ventricle have been reported to have undergone aortic valve replacement for severely regurgitant bicuspid aortic valve. Herein, we describe a patient with noncompaction of the left ventricle who underwent successful mechanical aortic root replacement.


Left ventricular noncompaction (LVNC) is a rare cardiomyopathy initially described in the pediatric population [1]. Recently this entity has gained increasing attention, reflected by the growing number of case reports [2–4] and small series of this cardiomyopathy in the adult population. This disease is classified by the American Heart Association as a genetic cardiomyopathy with a sporadic and familial form [5]. Nevertheless, a correlation between genotype and phenotype is not well elucidated with important genetic heterogeneity. This cardiac abnormality is associated with other congenital cardiac defects (pulmonary atresia, ventricular septal defect, aortic stenosis) [1], and the diagnosis is established during childhood. Here, we present a rare case of a 57-year-old woman with an aortic root aneurysm, bicuspid aortic valve, and LVNC.

The patient was a 57-year-old woman with a 3-month history of increasing weakness and dyspnea on exertion. Her medical history included hypertension and paroxysmal atrial fibrillation. On admission she was in sinus rhythm. Dyspnea was classified as class III heart failure according to the New York Heart Association.

The results of her physical examination were unremarkable other than for a diastolic murmur. Chest radiography showed mild cardiomegaly. An electrocardiogram revealed first-degree heart block. Transthoracic echocardiography showed a bicuspid aortic valve type 0 with severe aortic regurgitation and dilated aortic root, the left ventricular end-diastolic diameter was 65 mm, and the left ventricular ejection fraction was 40%, with diffuse left ventricular hypokinesis. Trabeculations were prominent in the apex and the lateral aspect of the left ventricular wall (Fig 1A). The ratio between the noncompacted and compacted layers was 2.5, which met the echocardiographic criteria of isolated LVNC. Cardiac magnetic resonance imaging (MRI) confirmed the diagnosis (Fig 1B). A multislice cardiac-gated computed tomographic scan revealed an aneurysmal aortic root (Fig 2A), with normal coronary arteries and the typical trabecular meshwork and intertrabecular recesses of LVNC. The bicuspid aortic valve is shown in Figure 2B.

Cardiopulmonary bypass was instituted between the ascending aorta and the right atrium. A 27-mm CarboMedics valve conduit (CarboMedics, Austin, TX) was used to replace the bicuspid aortic valve and the aortic root. Weaning from cardiopulmonary bypass was uneventful. The cross-clamp time was 60 minutes, and the cardiopulmonary bypass time was 85 minutes. The postoperative period was unremarkable, and the patient was discharged on the seventh postoperative day. At the last

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Fig 1. (A) Transthoracic echocardiogram demonstrating maximal noncompaction myocardium at the level of the left ventricle apex (white arrows). (LV = left ventricle.) (B) Magnetic resonance image showing the classic aspect of left ventricle noncompaction (white arrow).
follow-up visit 6 months later, she was doing well, and the echocardiogram revealed a good functioning prosthetic valve; the left ventricle ejection fraction increased to 50%.

Comment

Left ventricular noncompaction is a genetic-based abnormality of excessive and prominent trabeculation of the left ventricle, secondary to the arrest in the compaction of the ventricular myocardium during embryogenesis. The American Heart Association classified LVNC as primary genetic cardiomyopathy [5]; by contrast, the European Society of Cardiology does not recognize the genetic basis of this disease and considers it as unclassified cardiomyopathy [6].

The true prevalence of LVNC is still unknown. However, advances in, and the widespread availability of, cardiovascular imaging have led to an increase in the diagnosis of isolated LVNC, and it is no longer considered a rare form of cardiomyopathy. The prevalence of this disease is reported to be between 0.014% and 1.3%; nevertheless, these numbers are from retrospective studies that are exclusively from echocardiography laboratories in tertiary care centers [1].

Adult patients with isolated LVNC presented with symptoms of left ventricular dysfunction; however, the age at onset depends on the extent of noncompacted heart segments. This cardiomyopathy is frequently complicated by life-threatening arrhythmias and thromboembolism [7]. Of note, this cardiac abnormality is often associated with other congenital heart defects in the pediatric population; accordingly, the spectrum of the initial presentation is wide and depends on the gravity of the cardiac abnormality when LVNC is associated with a genetic syndrome [1]. Bicuspid aortic valve is the most common cardiac valvular anomaly, occurring in 1% to 2% of the general population. The present report describes a rare case of a surgical procedure in a patient with LVNC, aneurysmal aortic root, and bicuspid aortic valve. To our knowledge, only one other such case has been previously reported, in which aortic valve replacement was performed in a 61-year-old man with LVNC for severe bicuspid aortic valve regurgitation [2]. The left ventricle ejection fraction of this patient increased from 55% to 60% 1 year after an uneventful operation.

To establish the diagnosis of LVNC, Chin and colleagues [8] were the first to propose echocardiographic criteria. The ratio of noncompacted to compacted thickness >2 at end-systole is the most used diagnostic criteria for LVNC [1]. In challenging cases, new echocardiographic techniques (strain and strain rate, Doppler imaging) may help to distinguish normally trabeculated endocardium from isolated LVNC. Because of the limitation of echocardiographic criteria, many authors advocate the use of an MRI ratio between the noncompacted and compacted layers >2.3 measured at end-diastole [9]. In the present case, this ratio was >2 and >2.3 at echocardiography and MRI, respectively. Areas of noncompaction were predominantly located in the apex and the lateral aspect of the left ventricle.

A remarkable aspect of this case is that the left ventricular dysfunction of the patient was deemed secondary to LVNC associated with long-term aortic valve regurgitation. Undeniably, aortic root dilatation (45 mm) is a marker of a long-lasting associated valvulopathy. Aortic root replacement in a patient with LVNC, even in the setting of left ventricle dysfunction, can be carried out with excellent results. Family screening was recommended as indicated by the guidelines, especially in the presence of two diseases with a genetic background.

References

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