Correspondence

Mitral Regurgitation 43 Years after Commissurotomy by Dr. Denton A. Cooley

To the Editor:

In 2010, a 77-year-old woman presented at our hospital with shortness of breath and congestive heart failure. In 1967—43 years earlier—she had been a patient of Dr. Denton A. Cooley, who had performed commissurotomy for her mitral valve disease. The replacement valve was made of bovine pericardial tissue. In 2007, the patient had developed chronic atrial fibrillation, which was controlled with digoxin. She had no other significant comorbidities. Upon her presentation in 2010, her electrocardiogram showed atrial fibrillation, and her echocardiogram showed severe mitral regurgitation and a left ventricular ejection fraction of 0.65.

The patient underwent mitral valve replacement at our hospital in March 2010. After median sternotomy and bicaval cannulation, cardiopulmonary bypass was started. The aorta was cross-clamped, myocardial protection was achieved with use of topical cooling, and cardioplegic solution was administered antegrade and retrograde. The preoperative transesophageal echocardiogram had suggested severe mitral sclerosis and insufficiency, and these were confirmed upon direct inspection. The left atrium was opened, and the mitral valve was noted to be extremely sclerotic—typical of a rheumatic-type valve. The valve was excised and was replaced with a 29-mm bovine pericardial tissue valve. A CryoMaze procedure was performed to eliminate the atrial fibrillation. The atrium was closed with use of 4-0 polypropylene suture. The patient was weaned from cardiopulmonary bypass without difficulty, and the chest was closed in standard fashion. An immediate postoperative transesophageal echocardiogram revealed no mitral insufficiency. The patient had a prolonged postoperative course because of her congestive heart failure but was discharged from the hospital in sinus rhythm and with her warfarin therapy discontinued. In 2013, an echocardiogram showed only trace amounts of mitral regurgitation. The patient subsequently developed bradycardia and sick sinus syndrome that necessitated pacemaker placement in 2014. As of May 2015, she was alive and doing well.

Our patient's original mitral valve surgery was performed by Dr. Cooley during the infancy of cardiac surgery. Our patient's long life after that procedure is remarkable, and it is impressive even in the era of percutaneous balloon valvotomy.

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Lamin A/C Mutations Do Not Cause Left Ventricular Hypertrabeculation/ Noncompaction

To the Editor:

We read the article by Parent and colleagues[1] about a family in which 4 members carried the lamin A/C (LMNA) mutation n.644R>C and 3 members presented with left ventricular hypertrabeculation/noncompaction (LVHT). We have the following comments and concerns.

The authors describe the LMNA mutation as causative of LVHT. Although LVHT is frequently associated with mutations in various genes that encode for structural proteins, transport proteins, or enzymes in myocardial or muscle cells, none of these mutations has ever been proved to "cause" LVHT. One argument against a causal relation between the many mutated genes and LVHT is that a large number of different genes are associated with LVHT.[2] Another argument is that a mutation is often present in various family members, but only one member presents with LVHT. Furthermore, a mutation might be associated with different myocardial manifestations in the same family, such as LVHT, hypertrophic cardiomyopathy, dilated cardiomyopathy, arrhythmias, or sudden cardiac death.

Lamin A/C mutations not only cause cardiac disease, but also Emery-Dreifuss muscular dystrophy, limb-girdle muscular dystrophy, Dunnigan-type familial partial lipodystrophy, mandibuloacral dysplasia, Hutchinson-Gilford progeria syndrome, restrictive dermopathy, and autosomal recessive Charcot-Marie-Tooth disease type 2.[3] Did the authors' patient develop muscular manifestations? Was he seen by a neurologist to investigate

whether muscle, peripheral nerve, or other noncardiac tissues were clinically or subclinically involved?

The patient was reported to have experienced primary syncope. Were neurologic causes of syncope considered? Were cerebral magnetic resonance imaging (MRI), electroencephalography, and carotid ultrasound carried out? Because LVHT might be associated with stroke or embolism, it is essential to exclude ischemic stroke on cerebral MRI. What were the results of long-term blood pressure monitoring? Because systolic function was normal, there is no sufficient explanation for syncope.

De novo development of LVHT (acquired LVHT) is not unusual. It has been reported in patients with neuromuscular disorders (NMDs) and recently in pregnant women and professional athletes.[4]

Lamin A/C mutations have been reported not only in association with the authors' patient,[1] but also in other patients with LVHT.[3,5,6]

Overall, this interesting case could profit from further investigations for neurologic disease, cardioembolism, and neurologic causes of syncope. The detection of a mutated gene does not necessarily imply causality of LVHT.

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