refused amputation,<sup>3,4</sup> we selected drug therapy and monitored the course. Cases of long-term neglect of an acquired arteriovenous fistula resulting in prominent arteriovenous dilatation and mass formation are very rare.

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# Transesophageal Echocardiography in Congenital Aortic Valvulopathy

La ecocardiografía transesofágica en la valvulopatía aórtica congénita

# To the Editor,

Although new imaging techniques, such as computed tomography and magnetic resonance imaging, can be used for identification and detailed assessment of aortic valve diseases, transesophageal echocardiography (TEE) is still a useful and fundamental technique for diagnosis of such abnormalities.

Congenital aortic valve abnormalities account for 3% to 6% of congenital valve diseases in adults. The most common aortic valve abnormality is bicuspid aortic valve (accounting for 2%), followed

by unicuspid and quadricuspid aortic valves. To illustrate the validity and utility of TEE for the study of aortic valves, we present 2 noteworthy clinical cases.

*Case 1* (Fig. 1). A 30-year-old man who had been diagnosed in infancy with a bicuspid aortic valve was under study due to progressive dyspnea. Transthoracic echocardiography showed extensive stenosis and disruption of the aortic valve. TEE was performed for a more accurate anatomical assessment. The TEE study revealed a unicommisural unicuspid valve (UCV) and extensive calcification and stenosis (Fig. 1A); there was also an aneurysm of the ascending aorta, measuring 50 mm, without associated complications. To complete the study underwent preoperative magnetic resonance that, however, reported that the aortic valve was bicuspid (Fig. 1C) as well as the existence of aortic aneurysm (Fig. 1B) and the absence of birth abnormalities coronary arteries. The patient finally underwent valvular and aortic surgery, and the surgeon confirmed the presence of a UCV (Fig. 1D).



Figure 1. Unicuspid aortic valve.



Figure 2. Quadricuspid aortic valve.

*Case 2* (Fig. 2). A 58-year-old man with a history of chronic ischemic heart disease had undergone 2 prior percutaneous revascularization procedures. In the coronary angiogram, an abnormal origin of the left anterior descending artery and the circumflex artery was detected (Fig. 2A and B), as well as aortic valve sclerosis with moderate regurgitation. Given the deficient transthoracic echocardiography window, TEE was performed. The images obtained revealed a heavily calcified, type G, quadricuspid valve (QCV) with moderate regurgitation (Fig. 2C). To complete the study, cardiac magnetic resonance imaging was performed. This study confirmed the findings of the TEE (Fig. 2D).

UCV is a rare congenital abnormality whose prevalence was 0.02% in series of subjects assessed echocardiographically and 4% to 6% in series of patients who underwent surgery for aortic valve stenosis. UCVs are generally stenotic and require replacement when the patients are in their 20 s. The unicommisural anatomic variant is the least frequent. The positions of the coronary ostia are usually normal, whereas there may be dilatation or aneurysm of the ascending aorta with a predisposition to aorta dissection.<sup>1,2</sup>

On the other hand, the presence of a QCV is even more rare, with a prevalence of 0.008% in autopsy series, 0.013% to 0.043% in echocardiographic series, and 1% in patients who have undergone surgery for aortic valve regurgitation. Seven types or variants have been described; type G (in which the 4 aortic cusps are unequal) is the least common. Congenital aortic valve abnormalities are usually associated with regurgitation and require replacement when patients are in their 40 s or 50 s. A preoperative diagnosis of congenital aortic valve abnormalities is important, given that 10% of the patients also have abnormal origins of coronary arteries; the surgeon should be aware of this to avoid occlusion of the abnormal coronary ostia during attachment of the valve prosthesis.<sup>3,4</sup>

Thus, in aortic valve disease, in addition to determining whether the abnormality is congenital or acquired and assessing the functional impact, it is necessary to define the valvular anatomy. The UCVs may be associated with dilatation of the ascending aorta, with the corresponding risk of complications at this level, and also can be associated with abnormal origins of the coronary arteries, with a potential risk of fatal cardiac events (such as those derived from occlusion of coronary ostia during surgery).

Currently, TEE is an accessible, fast, and relatively noninvasive examination that can determine the type of aortic valve disease and detect abnormalities in the ascending aorta and birth defects in the coronary arteries of most of these patients.

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