Bicuspid Aortic Valve

Válvula aórtica bicúspide

DEFINITION

The aortic valve allows oxygenated blood to flow from the left ventricle to the aorta -the major artery that carries blood to the rest of the arteries in the body.
A normal aortic valve has three flaps, also known as cusps or leaflets. Occasionally, people are born with an aortic valve that has only two leaflets. This is known as bicuspid aortic valve (Figure 1).
The bicuspid aortic valve may occur in isolation in an otherwise healthy heart, or it may occur with other heart defects, such as coarctation (localized narrowing) of the aorta or aortic dilatation.
Bicuspid aortic valve was first described by W. Osler, in 1886.

INCIDENCE

Isolated bicuspid aortic valve is the most common of all congenital cardiac anomalies.
If a bicuspid aortic valve does not open normally, this is called aortic valve stenosis. The aortic valve is generally bicuspid in cases of congenital aortic stenosis, which accounts for 3-6% of all cases of congenital heart disease. It is found more often in men (3:1), and it may lead to complications in the adult patient.
Today, rather than a valve problem, it is considered a genetic disorder involving the development of the heart and the aorta.

DIAGNOSIS

Diagnosis is made by physical examination: cardiac auscultation reveals a typical aortic stenosis murmur which is best heard in the thorax. It can be associated with other murmurs such as aortic regurgitation. Additional tests include patient pulses, ECG, and chest x-ray.
Transthoracic echocardiography is the method of choice to confirm diagnosis, because it determines whether the valve is bicuspid with 92% sensitivity (accuracy to determine the condition) and 96% specificity (to discard it). When there are abundant calcium deposits on the valve or the images have poor quality, transesophageal echocardiography, computed tomography or magnetic resonance imaging can be used to complete the diagnosis.

CLINICAL COURSE

It varies from severe valve disease in infancy –requiring repair and immediate treatment– to asymptomatic involvement in old age.
Aortic valve disease usually occurs between the second and fifth decades of life.
The extent and progression of valve disorders or aortic dilatation vary. During follow-up, age > 30 years and stenosis or mild or severe aortic valve failure are considered risk factors for complications, as established in the Olmsted and Toronto series.

AORTIC STENOSIS: PROGNOSIS

When it occurs in infancy and there are few calcium deposits on the valve, non-surgical valve repair through opening (valvuloplasty) with a balloon performs well and does not require surgery.
In studies on children with gradients > 50 mm Hg, the rate for serious cardiac events is 1.2% per year. Only 1 in 50 children has clinically significant valve disease at adolescence.
In adult patients, progression of the disease is due to calcification of the valve leaflets. It is an active process, initiated by inflammatory mechanisms and fibrosis with calcium and lipoprotein deposits on the edge of the aortic leaflets. Calcification usually occurs after the age of 40.
It is important to evaluate patients' relatives to identify other cases.

RECOMMENDATIONS AND TREATMENT

In most cases, a bicuspid aortic valve does not require any treatment if detected in childhood, and follow-up is made by repeating the ECG every few years. Progression of valve disease is very slow. Only when the stenosis of the valve worsens, a closer follow-up, including echocardiography every 6-12 months, is required.
In adults, echocardiographic follow-up can be performed periodically in mild cases, but if aortic stenosis is severe or symptoms such as chest pain, shortness of breath, syncope or complications (valve infection) occur, surgery is required to replace the valve by a valvular prosthesis, which depending on the patient's age can be biological (made from human or animal tissues) or mechanical (made from inert materials).

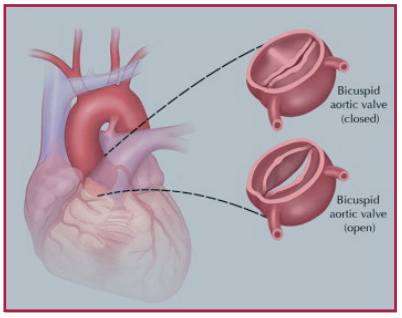

Fig. 1. Image showing the bicuspid aortic valve with two leaflets: closed (diastole) at the top, and open (systole) at the bottom

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INFORMATION YOU MAY FIND IN THE WEB

- ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol 2008;52:e1-e121.
- Patients with Bicuspid Aortic Valve ECOSAC VALVULOPATÍAS: http://www.sac.org.ar/web/es/consejos-cientificos/grandes-temas-/valvulopatias/ Recomendaciones del Comité de la ASE para la cuantificación de cavidades: http://www.ecosiac.org/files/GUIA
- www.webdelcorazon.com/valvula-aortica-bicuspide
- umm.edu/spanishency/articles/valvula-aortica-bicuspide

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