

Use of transillumination echocardiography in the assessment and diagnosis of bicuspid aortic valve

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Abstract

Bicuspid aortic valve is the most common congenital heart defect. Transthoracic echocardiogram is the initial tool to assess and diagnose this condition, however, transesophageal echocardiogram with 3D modalities, including transillumination have a better anatomical and functional evaluation of the valve, allowing to classify the bicuspid aortic valve according to the position of the raphe and assess the main vessels for complications or exclude other cardiovascular diseases.

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Abstract.

Bicuspid aortic valve is the most common congenital heart defect. Transthoracic echocardiogram is the initial tool to assess and diagnose this condition, however, transesophageal echocardiogram with 3D modalities,

including transillumination have a better anatomical and functional evaluation of the valve, allowing to classify the bicuspid aortic valve according to the position of the raphe and assess the main vessels for complications or exclude other cardiovascular diseases.

A 56-year-old male with a 10-year history of hypertension presented to a routine primary care consult. Physical examination revealed a grade II/VI diastolic murmur in the aortic focus. As such, the patient was referred to a cardiologist where a transthoracic echocardiogram (TTE) was performed. (Panel A; Video S1) Despite a suboptimal valve acoustic window, it revealed mild aortic insufficiency and suspicion of bicuspid aortic valve (BAV). The left ventricular ejection fraction was 63% with a pulmonary artery systolic pressure of 35 mmHg and grade I diastolic dysfunction.

Transesophageal echocardiogram (TEE) was performed for optimal valve visualization. (Panel B; Video S2) This study revealed moderate aortic insufficiency with eccentric flow. Transillumination echocardiography confirmed the presence of BAV with a raphe between the right and left coronary cusps. (Panel C; Video S3) The patient is currently asymptomatic.

BAV is considered the most common congenital heart defect, with a prevalence of 1-2% in general population, predominantly in men. [1] It can present isolated or associated with other congenital defects. [2] According to the Sievers classification, BAV is categorized according to the orientation of the valve orifice and the presence or absence of a raphe. The subtypes include type 0 (without a raphe), type 1 (one raphe) and type 2 (two raphes). The fusion of two cusps creates a raphe, which can be located in different positions: between the left-right coronary cusps (L-R), right-noncoronary cusps (R-N) and noncoronary-left cusps (N-L). [3] After comparison with a corresponding anatomopathological specimen, this BAV was classified as Sievers 1 L-R. (Panel D)

TTE is the standard and initial imaging tool for diagnosing BAV. Nevertheless, limitations, such as suboptimal acoustic windows or restricted evaluation of the ascending aorta, have prompted the consideration of other alternatives for appropriate valvular assessment. [1] An alternative is 3D TEE, offering real higher spatial resolution and diagnostic accuracy, also guiding management decisions for valvular replacement with evaluation of the outflow tract size. [4] Previous studies have shown that 2D TEE compared to 2D TTE has a higher accuracy in identifying the valvular phenotype, (90.1% vs 47.4%) and those unable to be identified with this method, were successfully identified by 3D TEE. [3]

Transillumination 3D TEE increases the depth perception and border delineation, as such it can be an alternative to accurately detect aortic valve disease as BAV and assess the main vessels for complications as bicuspid aortopathy or exclude other cardiovascular diseases. [5]

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Figure legends

Figure 1. Bicuspid aortic valve. A) 2D transthoracic echocardiogram with suboptimal valve acoustic

window, no raphe was observed. **B)** 2D transesophageal echocardiogram showing a raphe between the left and right coronary cusp. **C)** 3D transillumination transesophageal echocardiography confirming the presence of the raphe between the left and right coronary cusp. **D)** Anatomopathological specimen corresponding with this patient, Sievers type 1 L-R.

1: non-coronary cusp, 2: right coronary cusp, 3: left coronary cusp, LA: left atrium, PA: pulmonary artery, RA: right atrium.

Video legends

Video S1: 2D transthoracic echocardiogram with color flow. Demonstrates suspicion of bicuspid aortic valve, however no raphe is observed.

Video S2: 2D transesophageal echocardiogram at 53°. Presence of bicuspid aortic valve with a raphe between the right and left coronary cusps.

Video S3: 3D transillumination transesophageal echocardiogram. Cristal mode allowing a better visualization of the bicuspid aortic valve with a raphe between the right and left coronary cusps.

