

Diagnosis of congenital unicuspid aortic valve by 64-slice cardiac computed tomography

Wende N. Gibbs, MD, Baron L. Hamman, MD, William C. Roberts, MD, and Jeffrey M. Schussler, MD

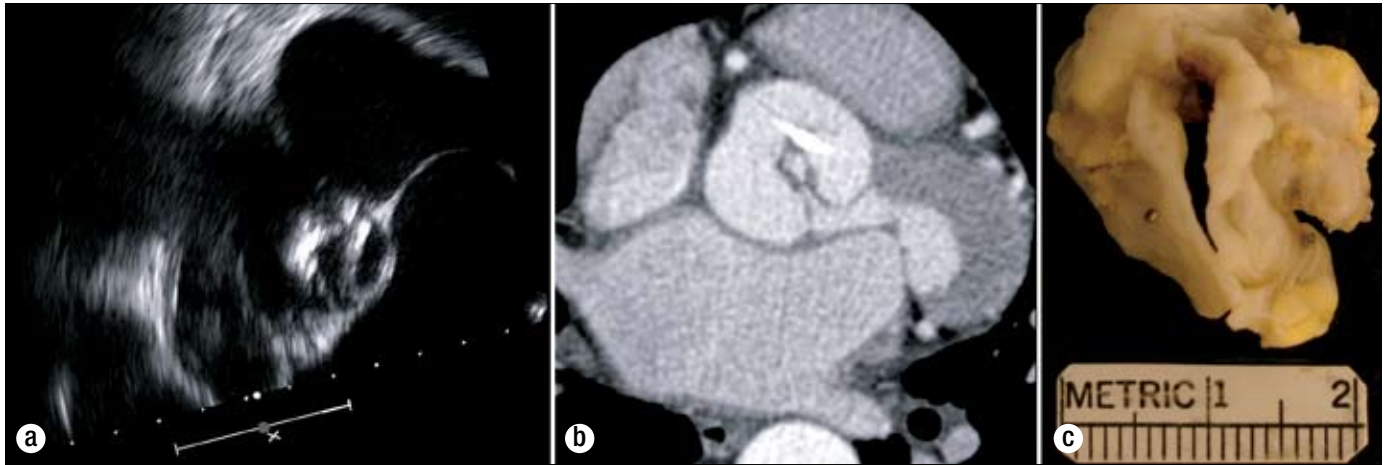


Figure. Unicuspid aortic valve evaluated with (a) transesophageal echocardiography, (b) computed tomography, and (c) gross pathology.

A 28-year-old man presented to our hospital with 3 days of intermittent, escalating dyspnea and chest tightness. He reported that he often experienced dyspnea when smoking, chewing tobacco, or exerting himself beyond the level of normal daily activity. His discomfort was partially relieved by his albuterol inhaler, which he used ≥ 10 times per day. As a child, he was told that he had a precordial murmur, but he had not sought medical attention. Five years prior to this evaluation, he was having similar symptoms and was treated with antibiotics. He was told at that time that he had mild asthma.

On examination, the patient was mildly dyspneic, with a harsh systolic murmur at the right upper sternal border. Transthoracic echocardiogram revealed left ventricular thickening with normal systolic and diastolic function and a left ventricular ejection fraction of 65%. A peak gradient of >4 meters per second was noted, with a calculated aortic valve area of <0.9 cm². The morphology of the valve was not clearly seen, but a unicuspid valve was suspected.

Transesophageal echocardiogram demonstrated a heavily calcified unicuspid aortic valve with reduced cuspid excursion and moderate to severe aortic regurgitation (Figure 1a). Preoperative 64-slice computed tomographic coronary angiography (Lightspeed VCT, GE Healthcare) confirmed the valve morphology and demonstrated no significant coronary narrowing (Figure 1b).

At operation, the valve was found to be unicuspid with one true unfused commissure. The free edge traversed the cusps without contact with the aortic wall (Figure 1c). The patient received a St. Jude medical mechanical prosthesis, and his postoperative course was uncomplicated.

The estimated incidence of unicuspid aortic valve is 0.02% (1–3). During development, the aortic valve is formed from three tubercles, which each develop a cusp and sinus of Valsalva. Fusion of the cusps results in a unicuspid valve. Unicommissural unicuspid valves, as in our case, have one lateral attachment and an eccentric orifice. Acommissural unicuspid valves have no lateral attachment to the aorta.

1. Novaro GM, Mishra M, Griffin BP. Incidence and echocardiographic features of congenital unicuspid aortic valve in an adult population. *J Heart Valve Dis* 2003;12(6):674–678.
2. Roberts WC, Ko JM. Frequency by decades of unicuspid, bicuspid, and tricuspid aortic valves in adults having isolated aortic valve replacement for aortic stenosis, with or without associated aortic regurgitation. *Circulation* 2005;111(7):920–925.
3. Roberts WC, Ko JM. Clinical and morphologic features of the congenitally unicuspid aocommissural stenotic and regurgitant aortic valve. *Cardiology* 2007;108(2):79–81.

From the Department of Internal Medicine (Gibbs), Department of Thoracic Surgery (Hamman), Department of Pathology (Roberts), and Division of Cardiology (Schussler), Baylor University Medical Center, Dallas, Texas.

Corresponding author: Jeffrey M. Schussler, MD, 621 North Hall Street, Suite 500, Dallas, TX 75226 (e-mail: Jeffrey.Schussler@heartplace.com).