Case Reports

Unicuspid Unicommissural Aortic Valve:

An Extremely Rare Congenital Anomaly

Sukhjeet Singh, DO, JD
Puneet Ghayal, MD
Atish Mathur, MD
Margaret Mysliwiec, MD
Constantinos Lovoulos, MD
Pallavi Solanki, MD, FACC
Marc Klapholz, MD, FACC
James Maher, MD, FACC

Unicuspid aortic valve is a rare congenital malformation that usually presents in the 3rd to 5th decade of life—and usually with severe aortic stenosis or regurgitation. It often requires surgical correction. Diagnosis can be made with 2- or 3-dimensional transthoracic or transesophageal echocardiography, cardiac computed tomography, or cardiac magnetic resonance imaging.

We report the case of a 31-year-old man who presented with dyspnea on exertion due to severe aortic stenosis secondary to a unicuspid unicommissural aortic valve. After aortic valve replacement, this patient experienced complete heart block that required the placement of a permanent pacemaker. ***(Tex Heart Inst J 2015;42(3):273-6)***

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From: *Division of Cardiology, Department of Medicine (Drs. Ghayal, Klapholz, Maher, Mathur, Mysliwiec, Singh and Solanki), and Division of Cardiothoracic Surgery, Department of Surgery (Dr. Lovoulos); Rutgers New Jersey Medical School, Newark, NJ 07103*

Address for reprints:
James Maher, MD, FACC, 185 S. Orange Ave., Newark, NJ 07101

E-mail:
maherja@njms.rutgers.edu

Isolated aortic valve stenosis in adults—with or without aortic regurgitation and without associated mitral stenosis—is almost always secondary to a congenital valvular malformation.[1] These congenital malformations usually present as a bicuspid, quadricuspid, or unicuspid aortic valve. Bicuspid aortic valve is the most common congenital aortic valve anomaly, with an estimated prevalence of 0.9% to 2%.[2] The most common anatomic form of bicuspid aortic valve consists of 2 cusps with a false raphe (between the right and left coronary cusps) and 2 commissures.[3] The prevalence of quadricuspid aortic valve on autopsy is about 0.01%.[4] Quadricuspid aortic valve has been categorized into 7 subtypes ranging from A to H; the 2 most frequent are type A (4 equal cusps) and type B (3 normal cusps and a smaller 4th cusp between the right coronary cusp and the noncoronary cusp).[5]

Unicuspid aortic valve (UAV) is a rare form of aortic valve malformation, the prevalence of which is 0.02% in the adult population.[6] Two forms of UAV, unicuspid acommissural and unicuspid unicommissural, have been described on the basis of the absence (or presence) of a lateral attachment of the commissures to the aorta at the level of the orifice.[7] Unicuspid unicommissural aortic valve usually presents in the 3rd to 5th decade of life,[8] most often as isolated aortic stenosis.[9] Diagnosis can often be made with 2-dimensional (2D) transthoracic echocardiography (TTE) or transesophageal echocardiography (TEE).[10] Definitive treatment is surgical replacement of the valve, performed when significant aortic stenosis or regurgitation develops.[11,12]

After aortic valve surgery, cardiac conduction defects have occurred with a reported prevalence of 5% to 6%; these have been well described.[13,14] Current guidelines recommend the placement of a permanent pacemaker for persistent complete heart block that develops after surgical valve replacement.[15] We present an illustrative case.

Case Report

In August 2012, a 31-year-old man presented with progressive dyspnea on exertion and lower-leg swelling over the course of the previous 2 weeks. He reported no medical or surgical history, was taking no prescription or over-the-counter medication, and reported no history of smoking or illicit-drug use. He reported no family history of cardiac disease or sudden cardiac death. At the time of his presentation, his vital signs were as follows: blood pressure, 100/65 mmHg; heart rate, 101 beats/min; body temperature, 98.5 °F; respiratory rate, 19 breaths/min; and oxygen saturation, 90% on room air. His physical examination was significant for a grade 4/6 systolic ejection murmur heard over the right upper sternal border, and for a diminished S_2.

An electrocardiogram revealed sinus tachycardia with left ventricular hypertrophy, repolarization abnormalities, and left atrial enlargement. A chest radiograph showed an

enlarged cardiomediastinal silhouette (Fig. 1). A TTE displayed a globally hypokinetic left ventricle with an ejection fraction of 0.20 and severe aortic stenosis with a maximum velocity of 4.2 m/s, a mean gradient of 41 mmHg, and a calculated aortic valve area of 0.84 cm^2.

On 2D TTE, the aortic valve appeared to be unicuspid (Fig. 2). Transesophageal echocardiography confirmed a stenotic, unicuspid, unicommissural aortic valve without substantial aortic dilation (Fig. 3). Because of the symptomatic, severe aortic stenosis, the cardiothoracic surgery staff was consulted. The patient was taken to the operating room for aortic valve replacement (AVR). After cardiopulmonary bypass was achieved, the ascending aorta was opened. Inspection of the aortic valve revealed a heavily calcified unicuspid valve. In the area where the right coronary cusp is usually located, there was no well-defined annulus—only the intraventricular septum was present. Moreover, the aortic annulus, at the level of the noncoronary cusp, was poorly defined, and the aortic valve appeared to be a folding web of tissue, with calcification that coursed into the anterior mitral valve leaflet.

Given the absence of a solid annulus in the area of the interventricular septum, the friable appearance of the coronary ostia, and the insignificant dilation of the aortic annulus and the aortic root, the Bentall procedure was deemed inappropriate. The UAV was replaced with a 23-mm valve (St. Jude Medical, Inc.; St. Paul, Minn), positioned in a supra-angular manner with 15 Ethibond 2-0 pledgeted sutures (Ethicon LLC, a Johnson & Johnson company; Somerville, NJ), thereby sparing the coronary ostia. After the aortic valve was inserted, it was tested and was observed to be working properly.

Postoperatively, the patient developed persistent complete heart block, which ultimately necessitated the placement of a permanent pacemaker. After his discharge from the hospital in stable condition, the patient was followed up in the medicine and cardiology clinics. As of December 2014, the patient remained asymptomatic and was taking his oral antiplatelet and vitamin K antagonist appropriately.

Discussion

Unicuspid aortic valve is an extremely rare congenital malformation, first reported by Edwards in 1958.[16] The annual incidence of UAV has been estimated at 0.02% in the adult population. However, in those patients who undergo surgery for isolated aortic stenosis, it is encountered at a rate of 4% to 5%.[1] During embryogenesis, a normal aortic valve develops with valve cusps, sinuses, and commissures. A normal trileaflet aortic valve consists of 3 cusps with 3 associated commissures, which develop from embryonic tubercules of the aortic trunk. Unicuspid aortic valve develops due to failure of the 3 aortic cusps to separate before birth.[1]

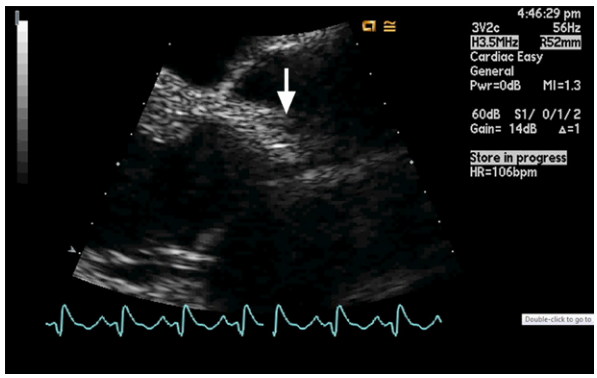

Fig. 2 *Transthoracic echocardiogram (parasternal long-axis view) shows a unicuspid aortic valve (arrow). Seen in systole, the valve has a single leaflet.*

Supplemental motion image is available for Figure 2.

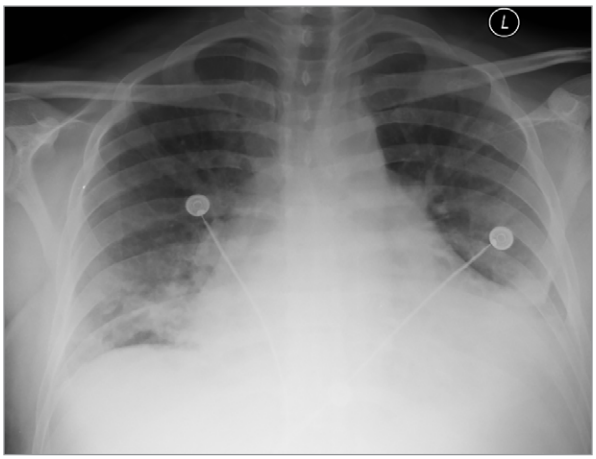

Fig. 1 *Chest radiograph (anteroposterior view) shows a large cardiac silhouette.*

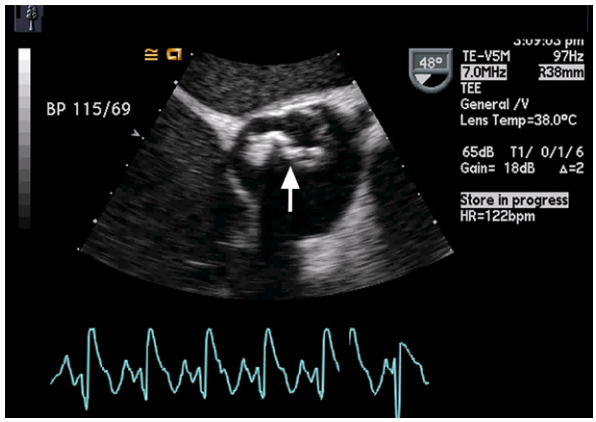

Fig. 3 *Transesophageal echocardiogram (parasternal short-axis view) shows the leaflet tips (arrow) of the unicuspid aortic valve.*

Supplemental motion image is available for Figure 3.

The unicuspid acommissural valve has no commissures or lateral attachments to the aorta at the level of the orifice and appears as a pinhole on imaging.[8] As a result, severe aortic stenosis develops at an early age, and patients can present at birth or infancy in need of surgical correction.[17,18] The unicommissural UAV has one lateral commissural attachment to the aorta at the level of the orifice and, in its morphology, appears as a slit-shaped structure.[8] Because the orifice in unicommissural UAV is larger than that in acommissural UAV, these patients typically remain asymptomatic until the 3rd to 5th decade of life,[1,19] as did our patient.

Unicuspid aortic valve is predominantly found in males (male-to-female ratio, 4:1). Patients with UAV develop symptomatic aortic stenosis 20 to 30 years earlier than do patients with a normal trileaflet aortic valve. Unicuspid aortic valve also has a bimodal presentation: a less aggressive form in older patients and a more aggressive form in younger individuals.[6] The diagnosis of UAV can be made with use of 2D or 3D TTE or TEE,[19] cardiac computed tomography,[20] or cardiac magnetic resonance imaging.[21] Unicuspid aortic valve is best imaged by means of echocardiography during systole, because the absence of cusp separation during aortic valve opening reveals the eccentric "teardrop" opening in a unicommissural UAV.[6]

Abnormalities associated with UAV include aortic aneurysm,[22] aortic regurgitation, aortic dissection, coarctation of the aorta, patent ductus arteriosus,[9] and aortic dilation. Current guidelines from the American College of Cardiology (ACC), the American Heart Association (AHA), and the Society of Thoracic Surgeons propose aortic root replacement as a Class IC recommendation when the maximum aortic diameter exceeds 5 cm. These guidelines also recommend that patients undergoing aortic valve surgery for valvular conditions undergo aortic root replacement as well, when the maximum aortic diameter exceeds 4.5 cm.[11,23]

The most common of the many valvular conditions associated with UAV is isolated aortic stenosis. Symptoms of aortic stenosis, regardless of commissural type, usually arise when the transvalvular mean gradient exceeds 40 mmHg, the aortic jet velocity is greater than 4 m/s, and the valve area is less than 1 cm^2. Current ACC/AHA guidelines on valve replacement recommend AVR for symptomatic severe aortic stenosis, a Class IB recommendation. Aortic valve replacement with a mechanical device was performed on our patient in response to hemodynamic abnormalities consistent with severe aortic stenosis and to his symptoms of exertional dyspnea.

Conduction abnormalities are a common and well-known sequela of isolated AVR. In one study of 261 patients, the necessity of a permanent pacemaker for complete heart block after isolated AVR was observed in 5% to 6% of patients.[24] When postoperative complete heart block is not expected to resolve spontaneously, the ACC/AHA guidelines set forth a Class IC recommendation for permanent pacemaker placement.[15]

In conclusion, unicuspid unicommissural aortic valve is a rare congenital disorder that often leads to severe aortic stenosis. Unicuspid aortic valve is an important clinical entity that should be in the differential diagnosis of younger patients who present with symptoms of heart failure and with a systolic murmur that suggests aortic stenosis. Patients usually present in the 3rd to 5th decade of life and often must undergo corrective valve replacement. As in our patient, complete heart block can develop as a sequela of aortic valve surgery, requiring the placement of a permanent pacemaker to avert sudden cardiac death.

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