Unicuspid Aortic Valve

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Case Report

History of Present Illness:

33 y/o WM with aortic stenosis and reflux gastritis presented on 11/03/09 c/o pre-syncope, weakness, SOB, and chronic chest pain.

Patient states he has had chest pain everyday since he was a child and it has slowly progressed in intensity over the last several years. He has symptoms of weakness, DOE, and pre-syncope with exertion over the most recent 3 years, and he has been curtailing his activity to avoid these symptoms (currently will get symptomatic with climbing a flight of stairs). No orthopnea, no LE edema.
The patient's chest pain occurs daily, up to 5 times per day. Sometimes the pain is sharp, especially when it occurs with exertion, with no radiation, occurs in left chest, lateral to the sternum, lasts 15 minutes, and resolves when he sleeps. Other times the pain is dull, with no associated SOB, and can occur at rest. His last episode of chest pain was 1 day PTA. Patient states the quality and location of pain has not changed since childhood, but the intensity and duration have slowly increased over the last several years.

Past Medical History: Aortic Stenosis and GERD
Surgical History: None
Family History: Father had aortic stenosis and died of MI at age 45; paternal uncles with unknown heart pathology, died from MI at ages 42 and 37
Social History: EtOH - social drinker. No tobacco products. No illicit drug use.
Physical Exam: General: VSS, AF, WDWN, in NAD, healthy appearing, non-cyanotic.
HEENT: Atraumatic head without lesions, normocephalic. Neck supple, without rigidity.
Resp: Clear to auscultation, no rales, no rhonchi, no wheezes, no accessory muscle usage.
Cardio: RRR, no gallops, no rubs, +3/6 systolic murmur with early systolic click, heard best at right upper sternal border, louder toward right periphery, radiates to bilateral carotids. No thrill palpated. No JVD.
Back/Gait: Able to rise from sitting, normal gait.
Extremities: No clubbing, no cyanosis, no edema.
Neuro: CN II-XII intact, motor/sensory/cerebellar function intact bilaterally, gait and speech intact, no focal motor or sensory deficits.
Abdomen: Soft, NTND, normal bowel sounds in all quadrants, no rebound tenderness, no guarding.
Vascular: DP pulses intact bilaterally, radial pulses intact bilaterally, pulses equal bilaterally.
Psych: Judgment intact, oriented to person, place, and time.

Assessment

A trans-esophageal ECHO reveals a unicuspid aortic valve with moderate aortic insufficiency, moderate aortic stenosis, and a mean trans-aortic pressure gradient of 36 mmHg. He was diagnosed unicuspid aortic valve with aortic stenosis.

Overview of Unicuspid Aortic Valve

Unicuspid aortic valve (UAV) is a rare congenital malformation, seen in approximately 0.02% of patients referred for echocardiography, but in as many as 4% to 6% of patients undergoing operations for "pure aortic stenosis." This type of malformation is usually manifest as a valve with a single commissure and a posterior attachment, and often occurs with coexisting ascending aortic dilatation. It presents far more often in males than females, often in the third decade of life when AS becomes clinically significant. These malformed valves tend to fail at an earlier age than normal aortic valves as they tend to suffer from increased pathological change with calcification of the cusp and base, ossification, and ulceration – all contributing to stenosis.

Two Clinical Presentations

One report documented the relationship of pathologic dilatation of the ascending aorta and age at presentation. This separated AS patients into two distinct clinical groups. The relationship was sharp, with a breakpoint at age 47, suggesting two different patterns of this disease. The older patients appeared to have a less aggressive form, with delayed presentation of symptoms and without aortic dilatation. In contrast, the aggressive form of unicuspid malformation was associated with early symptoms and aortic involvement. The latter group may have similar pathologic characteristics to the much more common entity of bicuspid aortic valve and ascending aortic dilatation.

Symptoms

Presenting symptoms are those of aortic stenosis, and may include dyspnea, angina, and dizziness or syncope.

Diagnosis

Clinical symptoms of aortic stenosis correlated with unicuspid valve on TEE or thin slice CT.

Treatment

Treatment involves replacement of the aortic valve when the stenosis is severe enough. However, remodeling of the unicuspid valve to form an operating bicuspid valve has been performed with some success by a cardiothoracic team in Germany.

What Happened to This Patient?

This patient was evaluated by cardiology and cardiothoracic surgery during his hospitalization in November 2009. He underwent a series of tests, included echocardiogram, left and right heart catheterization, and pharmacologic stress testing. Based on the degree of his stenosis, the decision was made to delay valve replacement until his stenosis was severe enough to meet criteria for replacement. His disease will be monitored with scheduled echocardiogram every 6 months until that time.

Figure 1. Images from the patient's nuclear stress test reveal moderately enlarged left ventricle with EF 51%. Normal pharmacologic rest and stress images.

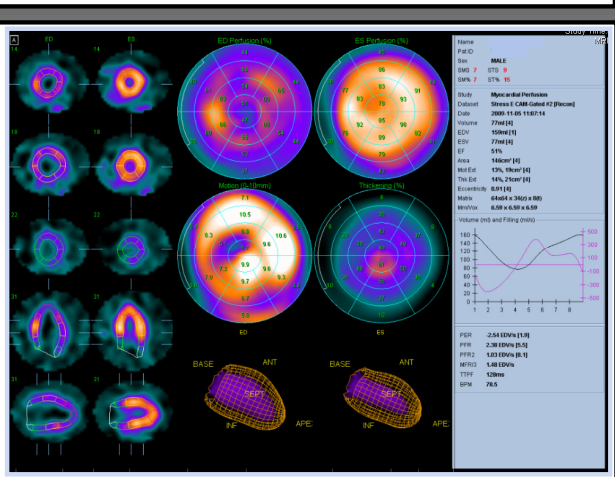
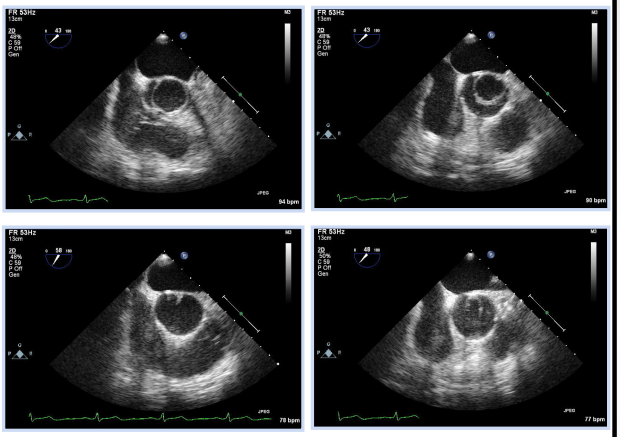


Figure 2. The patient's echocardiogram is displayed in various stages from systole (top left) to diastole (bottom left) in clockwise fashion. Note the eccentric orifice in systole, and the single lateral commissural attachment to the aorta (posterior) in diastole. Besides revealing a unicommissrual unicuspid valve, the study also revealed moderate aortic insufficiency and moderate aortic stenosis. Mean trans-aortic pressure gradient was 36 mm Hg.



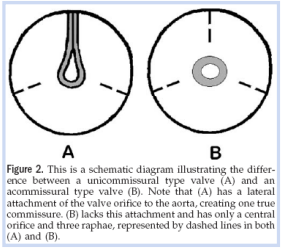


Figure 2. This is a schematic diagram illustrating the difference between a unicommissural type valve (A) and an acommissural type valve (B). Note that (A) has a lateral attachment of the valve orifice to the aorta, creating one true commissure. (B) lacks this attachment and has only a central orifice and three raphae, represented by dashed lines in both (A) and (B).

Figure 4. Image from Sneicinski et al., IARS, p 789

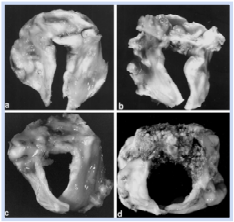


Figure 4. Examples of unicuspid unicommissural aortic valves.
Image from Roberts et al., Circulation 2005 p 921

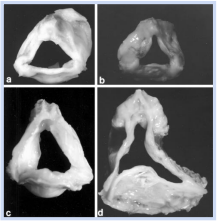


Figure 5. Examples of unicuspid acommissural aortic valves.
Image from Roberts et al., Circulation 2005 p 922

Summary

Unicuspid aortic valve is a rare congenital malformation which caries significant morbidity for affected individuals. As was the case for our patient, it is often diagnosed via echocardiogram, but the clinical presentation is that of aortic stenosis, except at a much younger age than when aortic stenosis usually appears. The importance of a complete cardiologic evaluation is emphasized by the fact that these patients will require eventual surgical address and close monitoring until that time.

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