Unicuspid Aortic Valve
Amber Bazler, MD and Christian Killer, MD
Homayoun Ataei, MD - Faculty Advisor
LSUHSC-Shreveport – Department of Family Medicine
Family Medicine Residency – Shreveport, LA

Case Report

History of Present Illness:
33 y/o WM with aortic stenosis and reflux gastritis presented on 11/3/39 /c/o pre-synecope, weakness, DOE, 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-synecope 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 sits down. Other times the pain is dull, with no associated DOE, 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: EIOH - social drinker. No tobacco products. No illicit drug use.
Physical Exam: General: VSS, AF, WW, in NAD, healthy appearing, non-cyanotic.
Cardio: RRR, no gallops, no rubs, +3/6 systolic murmur with early systolic click, heard best at right upper sternal border, louder toward right peripheral, 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.
Neuromus: CN I-VII 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 38 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 manifested as a valve with a single commissure and a posterior attachment, and often occurs with coexisting ascending aortic dilation. It presents far more often in males, 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 cusps and base, ossification, and ulceration – all contributing to stenosis.

Two Clinical Presentations
One report documented the relationship of pathologic dilation 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 dilation. 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. Other times the pain is dull, with no associated DOE, and can occur at rest.

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, including 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.

References
5. Circulation 2005;111:920-925; originally published online Feb 14, 2005;