Quadricuspid Aortic Valve

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Abstract
Quadricuspid aortic valve is a rare congenital heart defect usually detected by echocardiography or at the time of aortic valve surgery. Preoperative recognition of this abnormality has clinical significance because it is frequently associated with other congenital cardiac abnormalities and anomalies of coronary origin. We report a case of quadricuspid aortic valve, which was detected preoperatively by transthoracic echocardiography and was associated with severe aortic regurgitation.

Case Presentation
A 56 year old lady presented to us with a 1-month history of palpitation. On physical examination, her blood pressure was 140/70 mmHg and a diastolic murmur of grade 4/6 was heard at the lower left sternal border. The pulses were bounding and her ECG showed sinus rhythm with high QRS voltage and ST-T configuration consistent with left ventricular hypertrophy. Chest X-ray revealed cardiomegaly.

Trans thoracic echocardiography displayed a Quadricuspid aortic valve with two equal, larger cusps and two unequal smaller cusps associated with severe aortic regurgitation (central jet) (Figures 1, 2). There were no associated structural defects. The left ventricle was mildly dilated and hypertrophied with mild systolic dysfunction. Coronary arteriogram performed subsequently revealed normal origin of the coronaries with no luminal stenosis. This patient successfully underwent aortic valve replacement and was discharged in a stable condition.

Discussion
Quadricuspid aortic valve (QAV) is a rare congenital cardiac disorder, with a reported incidence between 0.008% and 0.033% at autopsy and ~ 1% in those posted for aortic valve surgery.1,2 Balington for the first time reported a case of QAV in 1862 and about 200 cases have been reported since then.

As per the reported literature, most of the cases of QAV are diagnosed by echocardiography but some are picked up only at the time of surgery.3 The diagnosis may thus be missed on transthoracic echocardiography.4 In our case, transthoracic echocardiography was able to detect the anomaly preoperatively and there was no need to perform transesophageal echocardiography.

The mechanism of formation of a QAV is not well understood. Aberrant fusion of the aorticopulmonary septum or abnormal mesenchymal proliferation in the common trunk may lead to abnormal cusp formation.

Anatomical classification of QAV according to the size of each individual cusp has been described by Hurwitz and Roberts5 as-Type A: Three equal-sized cusps and one smaller (most common type), Type B: Four equal-sized cusps (second most common type), Type C: Two larger and two smaller cusps, Type D: One large cusp, 2 intermediate-sized cusps, and 1 smaller cusp, Type E: Three equal-sized cusps and one larger cusp, Type F: Two equal and two unequal smaller cusps and Type G: Four unequal cusps (least common type).

The mean age of diagnosis is around 50 years with a slight male preponderance3 and the most common valvular
abnormality in QAV is aortic regurgitation. QAV stenosis is extremely rare (<1%). QAV has been associated with other congenital cardiac abnormalities such as variations in coronary anatomy, ventricular or atrial septal defects, aneurysm of sinus of valsalva, patent ductus arteriosus etc.

When there is an indication for surgical intervention, aortic valve replacement is the most acceptable procedure. It is very important for the surgeons to know the presence of QAV since there is a possibility of abnormally placed coronary ostia (specially the left) which may be damaged during aortic valve replacement.

References