Unusual Causes of Aortic Regurgitation

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No Disclosures

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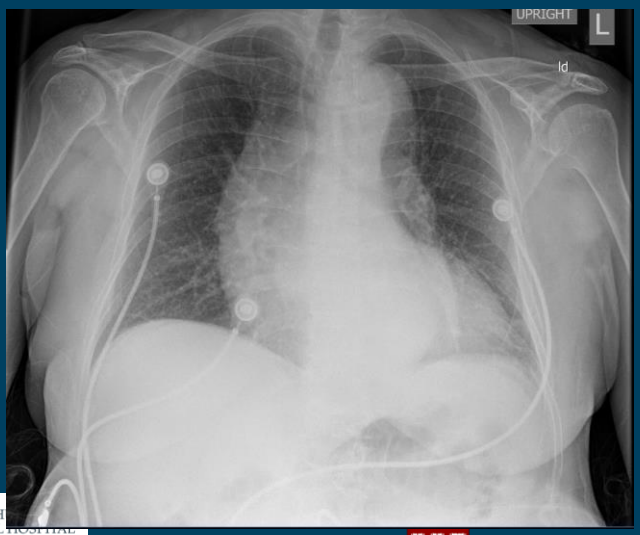
Case 1

- 54 year old female with h/o cerebral aneurysm and vascular malformation requiring surgery as a child and hypertension. FH: Mother also had cerebral aneurysm
- She presents with 3 week h/o persistent dry cough. No fevers, chills or other symptoms.
- CRP elevated 44
- CXR shows:

Case 1



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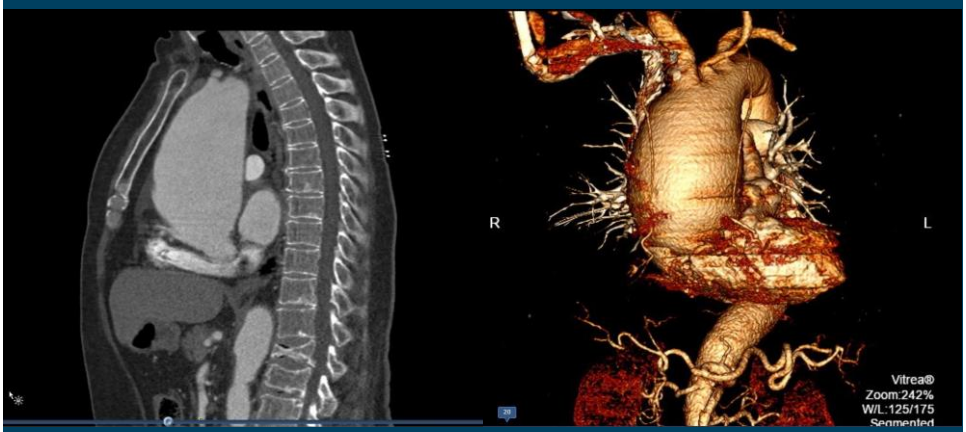
Case 1



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CTA of aorta

- Aortic root: 3.9 cm
- Markedly dilated ascending thoracic aorta at 8.0 cm tapering to 4.6 cm just proximal to the innominate artery, a 3.3 cm distal arch, a dilated proximal descending thoracic aorta at 3.8 cm, and a tortuous descending thoracic aorta tapering to 3.3 cm at the hiatus.
- Of note, there was no significant aortic wall thickening or calcification.

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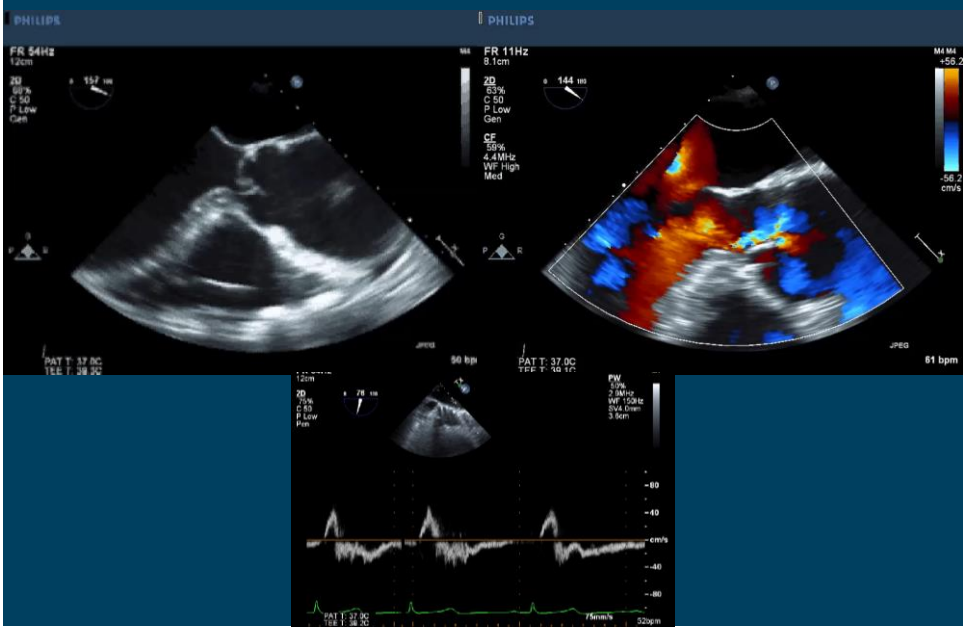
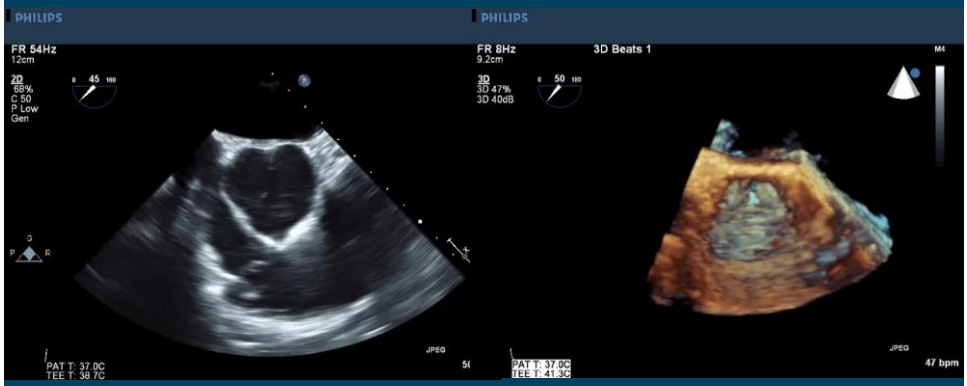
TEE



Tricuspid Aortic Valve; Restricted Motion of left cusp



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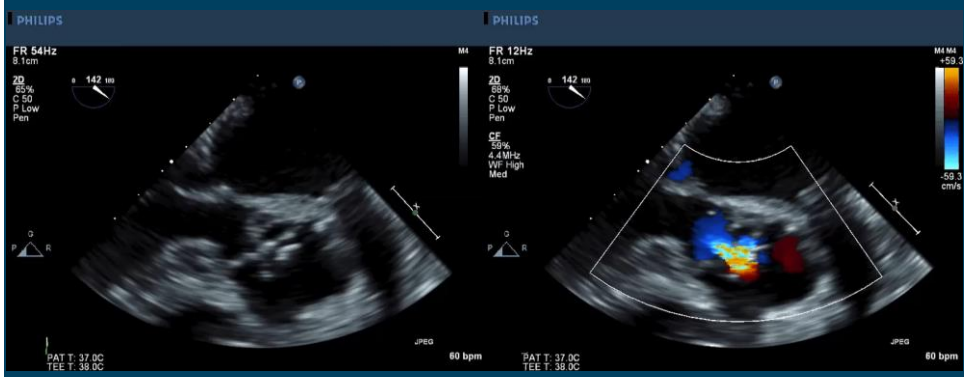
Restricted motion of the left cusp



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Case 1

- Mechanism of AR: Mild aortic root dilation; left aortic cusp was thickened and restricted in motion preventing full closure
- Ascending thoracic aortic aneurysm markedly dilated to 8 cm, which is unusually large for such aneurysms
- The aortic dilatation extended into her proximal aortic arch; the aortic diameter normalized in the distal arch but then enlarged again to 3.8 cm in the proximal descending aortic segment.

- Diffuse thoracic aortic dilatation is unusual and is often seen in the setting of aortitis.
-
- Aortitis typically affects middle-age to late-age women.
- However, usually in chronic aortitis expect to see thickening of the aortic walls or aortic calcification which were not evident by imaging.

- Another possibility is idiopathic diffuse aortopathy
- Given that she and her family have a history of cerebral aneurysms, possible that there is an underlying genetically-mediated connective tissue disorder.

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Clinical Course

- Ascending aortic replacement using a 28-mm graft and a zone 2 aortic arch replacement with reimplantation of innominate and left carotid artery.
- There was tethering of the aortic valve due to the aneurysm and restricted left aortic leaflet causing incomplete aortic valve closure and severe aortic insufficiency, aortic valve repair with resuspension of leaflets and plication attempted but with significant residual AR; aortic valve replacement with a 23-mm St. Jude mechanical tilting disc valve

Biopsies of aorta obtained Pathology showed:

- FINAL PATHOLOGIC DIAGNOSIS:
  ASCENDING AORTA, RESECTION:
  ACTIVE AORTITIS WITH A GRANULOMATOUS / GIANT CELL PATTERN OF INFLAMMATION (SEE NOTE).

  Note: There is extensive destruction of the medial elastic lamellae with adventitial, medial and intimal fibrosis. Within the media of the aortic wall there are foci of granulomatous inflammatory infiltrate, as well as an associated lymphoplasmacytic infiltrate primarily in the adventitia. By immunohistochemistry for IgG, IgG4, IgA, IgD, IgM, IgE, and CD138, most of the plasma cells express IgG and well under half of the plasma cells stain for IgG4 which is non-specific. Immunohistochemistry for CD68 and CD163 highlights the granulomatous component. The lymphocytes are both CD3+ T-cells and CD20+ B-cells, the latter of which are primarily in aggregates in the adventitia. The CD3+ cells are mixture of CD4+ and CD8+ cells with scattered FoxP3+ cells and rare Granyzyme-B+ cells. Trichrome and elastic stains and immunohistochemical stain for myeloperoxidase are also examined. The histologic findings can be seen in clinically isolated aortitis with a granulomatous / giant cell pattern of inflammation as well as systemic giant cell arteritis.

Case 2

- Patient is 33 yr old female who presents to establish cardiology care after moving to area.

- She had normal active childhood, She developed palpitations in her early 20s and was diagnosed with ruptured sinus of Valsalva aneurysm and underwent surgical repair. Intraoperatively, she developed complete heart block and had insertion of epicardial pacemaker leads

- She presents to establish cardiology care as she has recently moved to area
- She feels well, exercises regularly and denies functional limitations
- She is interested in having children

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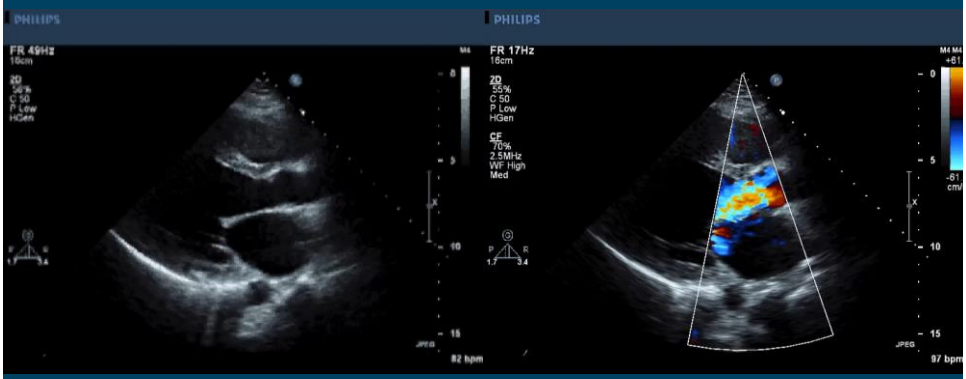
Case 2



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Case 2

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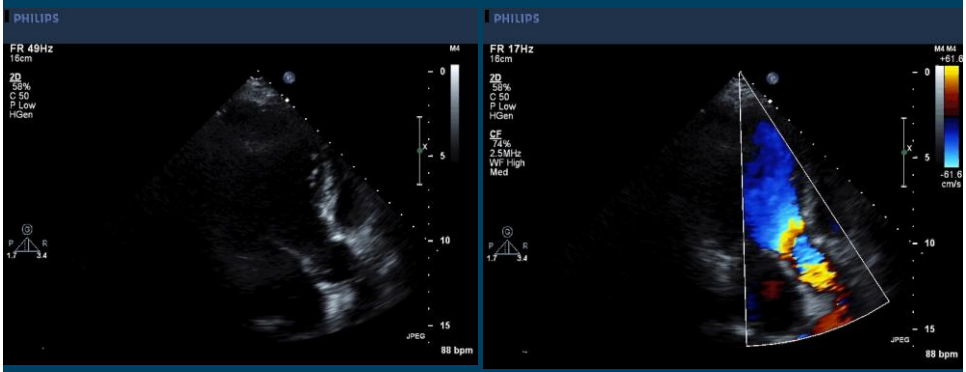

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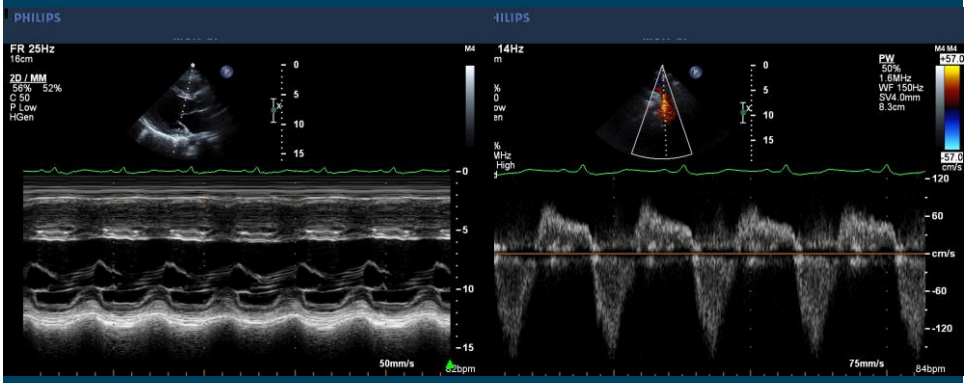
Premature closure of MV

Holodiastolic flow reversal in aorta



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What is mechanism of AR?

- Bicuspid with prolapse
- Aortic root dilation
- Discrete sub aortic membrane
- Leaflet restriction

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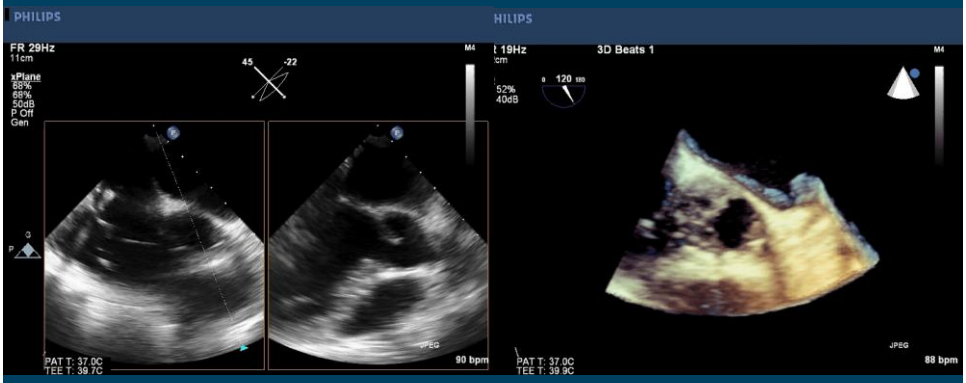
TEE: Restricted motion of the left aortic cusp



Summary of echo findings

- Severe AR due to restricted motion of the left aortic cusp; likely related to prior patch repair of ruptured sinus of Valsalva aneurysm with distortion of the aortic cusp
- LVEF 60%, no WMA. LVIDd 54 mm, LVIDs 38 mm. LA 41 mm. RVSP 32 mmHg normal ascending aortic and aortic sinus dimensions

What to do?

- Refer for aortic valve replacement before consideration of pregnancy
- Advise against pregnancy, defer AVR
- Defer AVR, proceed to pregnancy

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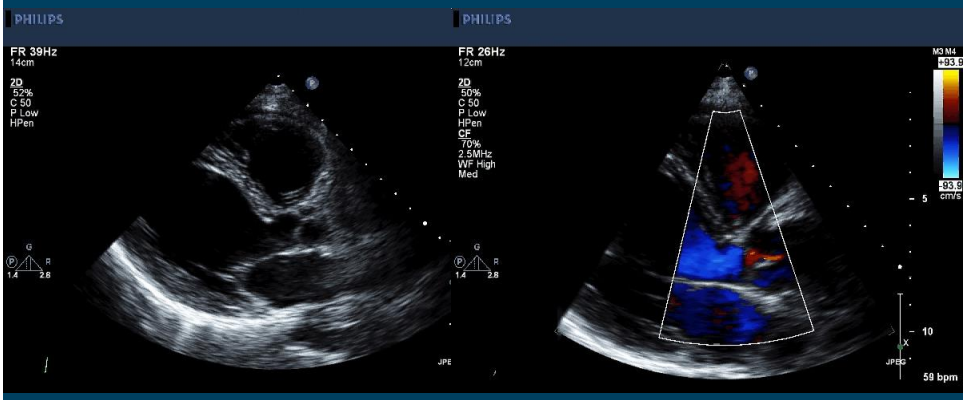
Recommendation

- Defer AVR, proceed to pregnancy
  - Patients with severe AR have acceptable pregnancy risk if asymptomatic with preserved LV function
  - In contrast, patients with severe AR with associated symptoms (stage D) or left ventricular systolic dysfunction or severe left ventricular dilation (stage C2), or pulmonary hypertension may develop heart failure during pregnancy due to the volume load.

What is mechanism of AR?



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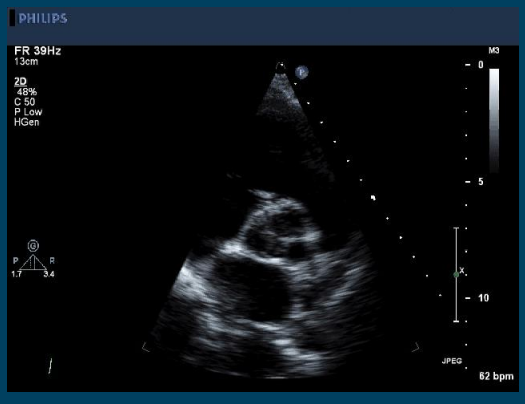
SAX of Ao valve



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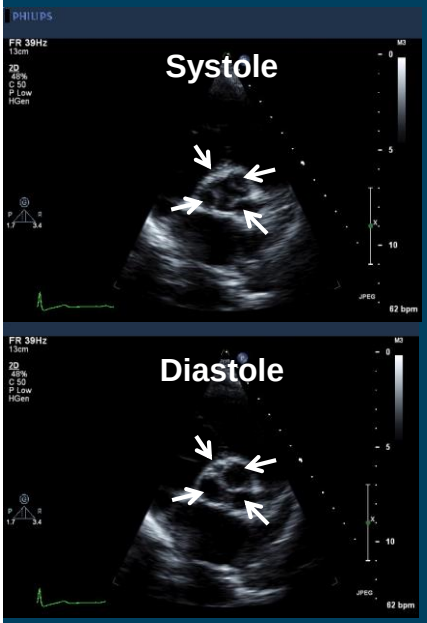
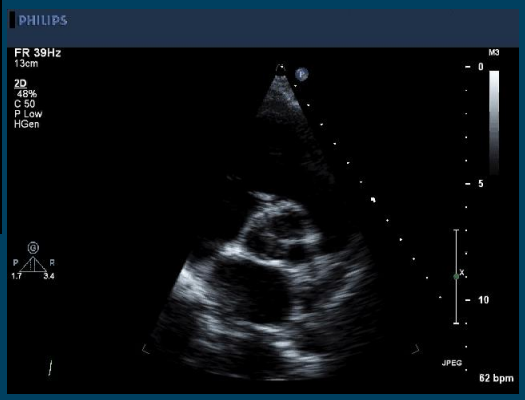
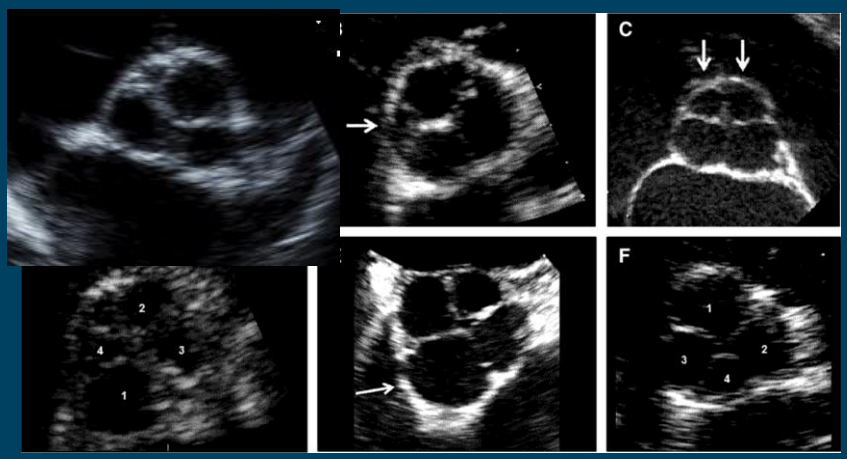
How many cusps?





Quadricuspid aortic valve classification in accordance with anatomic variations.



Michael Y.C. Tsang et al. Circulation. 2016;133:312-319

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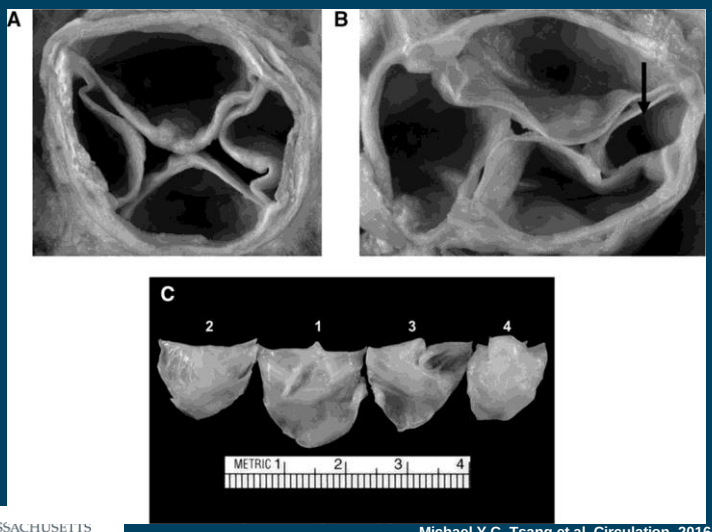
Pathological specimens of various subtypes of quadricuspid aortic valve.



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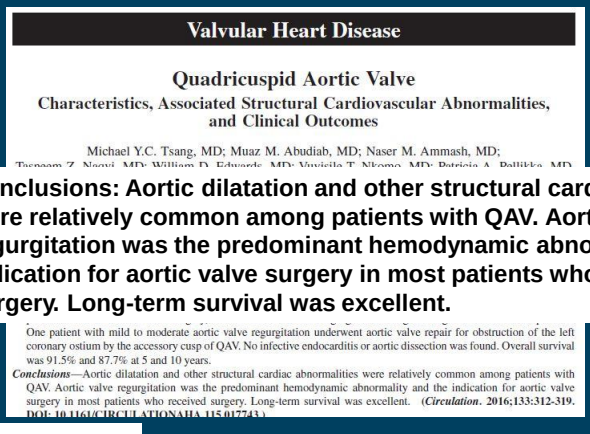

Valvular Heart Disease

Quadricuspid Aortic Valve

Characteristics, Associated Structural Cardiovascular Abnormalities, and Clinical Outcomes

Michael Y.C. Tsang, MD; Muaz M. Abudiab, MD; Naser M. Ammash, MD;

One patient with mild to moderate aortic valve regurgitation underwent aortic valve repair for obstruction of the left coronary ostium by the accessory cusp of QAV. No infective endocarditis or aortic dissection was found. Overall survival was 91.5% and 87.7% at 5 and 10 years.

Conclusions—Aortic dilatation and other structural cardiac abnormalities were relatively common among patients with QAV. Aortic valve regurgitation was the predominant hemodynamic abnormality and the indication for aortic valve surgery in most patients who received surgery. Long-term survival was excellent. (***Circulation***. 2016;133:312-319.

Conclusions: Aortic dilatation and other structural cardiac abnormalities were relatively common among patients with QAV. Aortic valve regurgitation was the predominant hemodynamic abnormality and the indication for aortic valve surgery in most patients who received surgery. Long-term survival was excellent.

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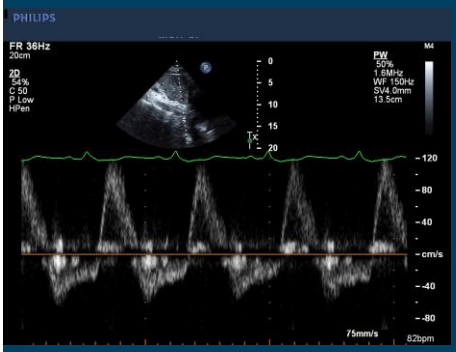
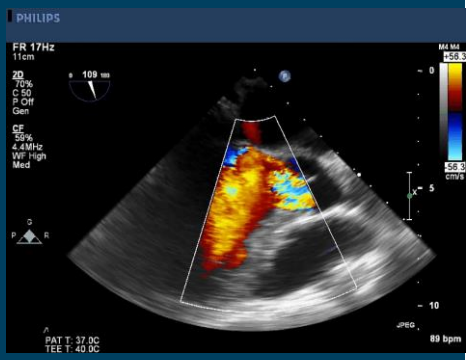
Thank you





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