

AORTIC VALVE CASES

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DISCLOSURES



- None

HANDOUT:

<http://stanford.edu/~hallett>

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CASE 1

- 5 yo male with murmur, FTT

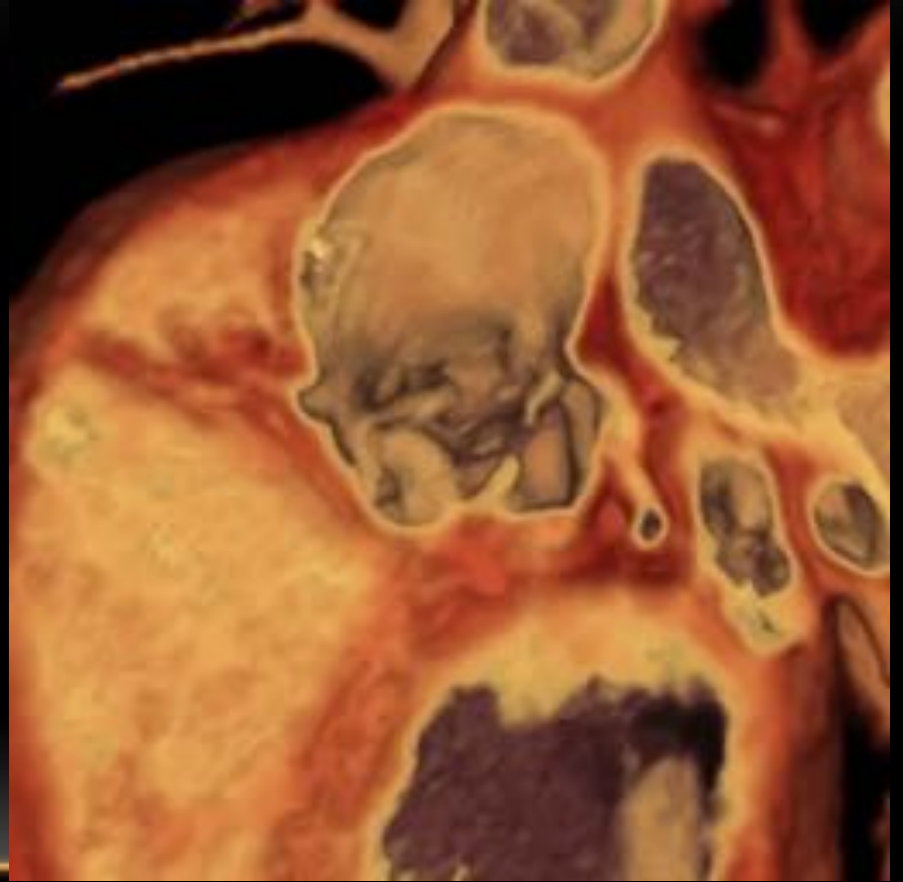
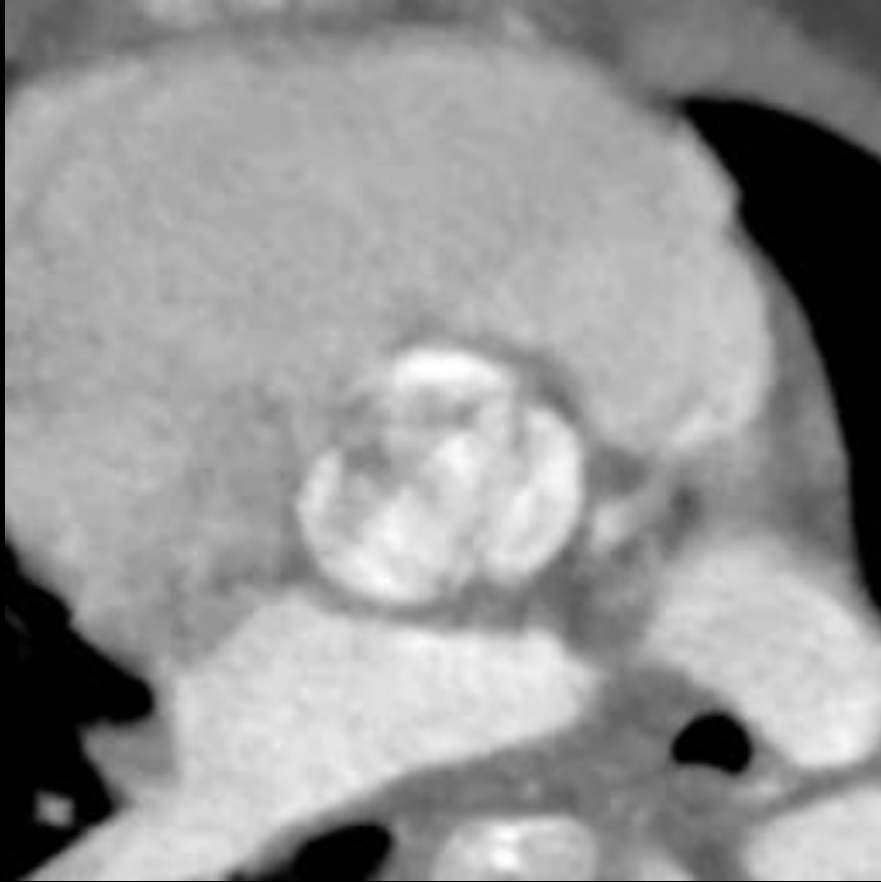
SYSTOLE

DIASTOLE

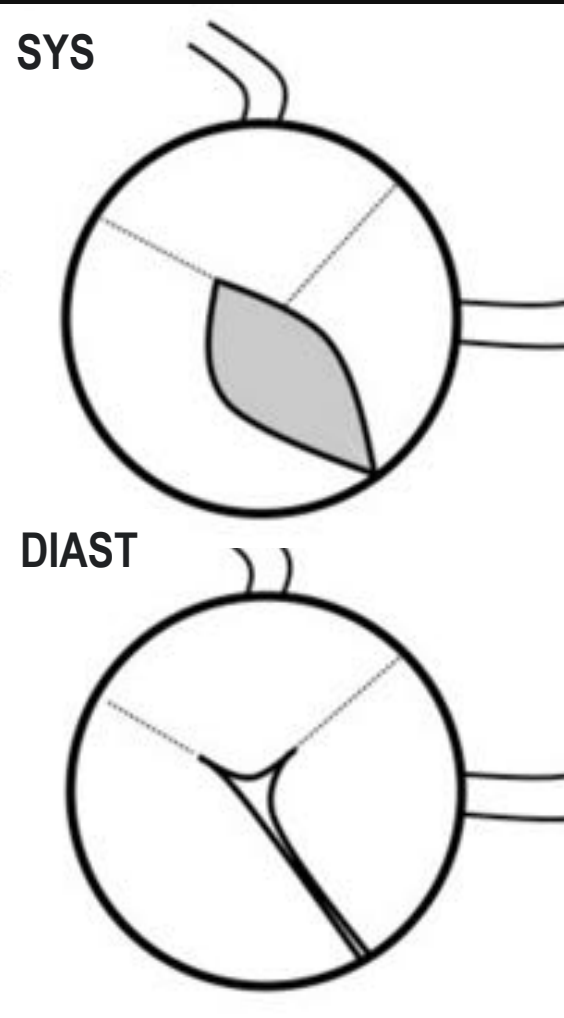


ECG synchronized CTA, HR > 100

CASE 1

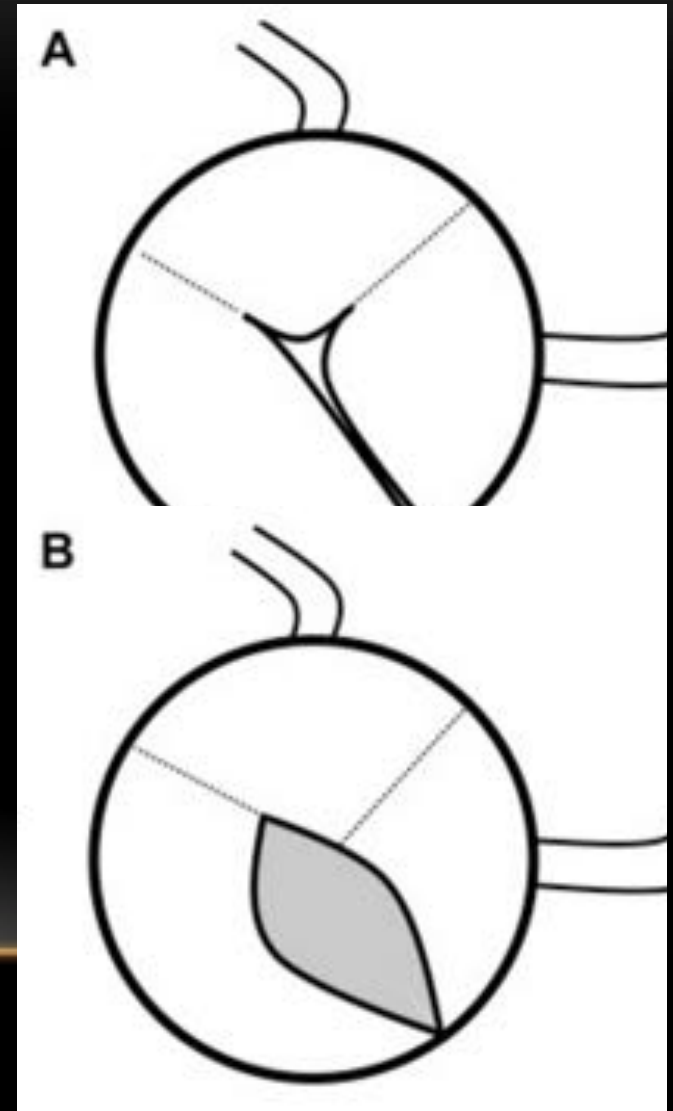
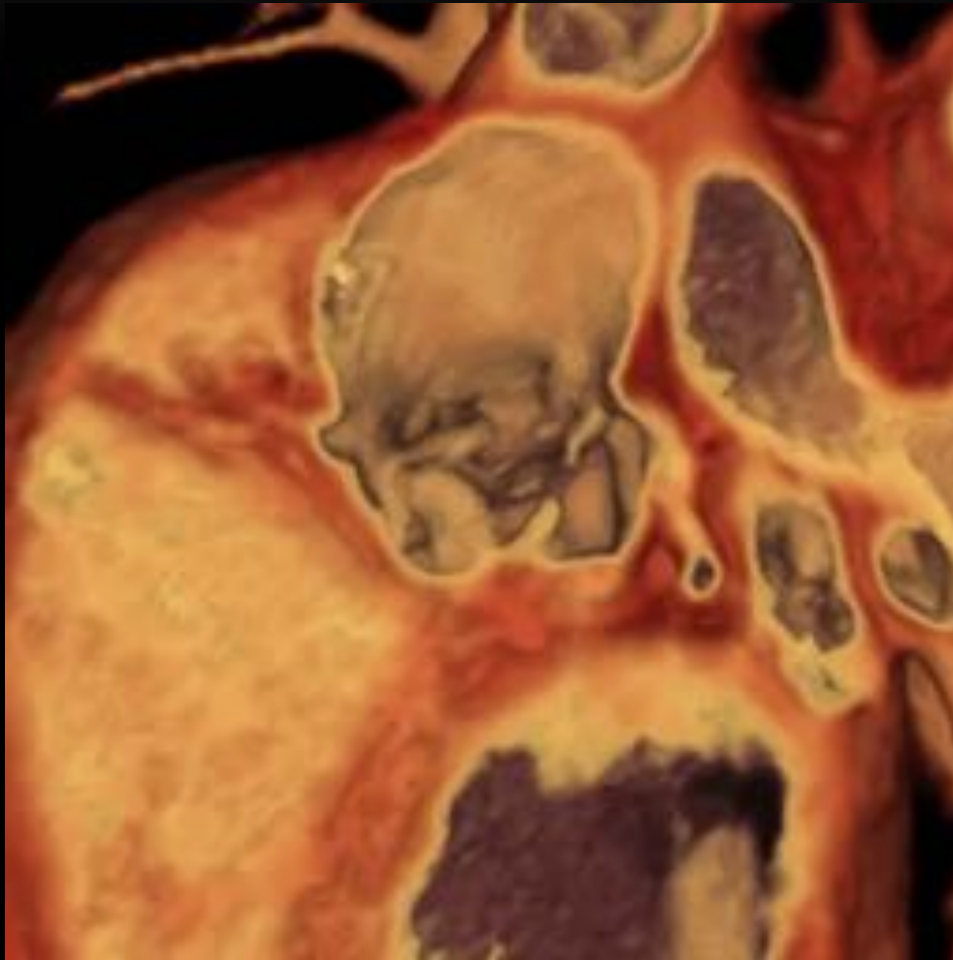


CONGENITAL UNICUSPID AORTIC VALVE



Line drawings: Petit CJ, et al Am J Cardiol 2016; 117(6) 972-9.

CONGENITAL UNICUSPID AORTIC VALVE



CASE 1 QUESTION:
WHICH OF THE FOLLOWING IS TRUE
REGARDING UNICUSPID AORTIC VALVE?

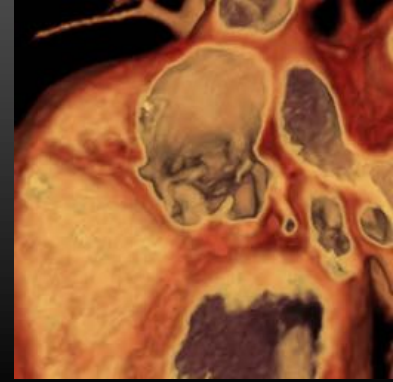
- A. Most cases present in adulthood
- B. UAV have better results from balloon valvuloplasty than bicuspid aortic valve (BAV)
- C. UAV shares some overlap with BAV patients, including risk of aortopathy and coarctation
- D. Have larger valve opening area than BAV

CASE 1 QUESTION: ANSWER

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CONGENITAL “UNICUSPID” AORTIC VALVE

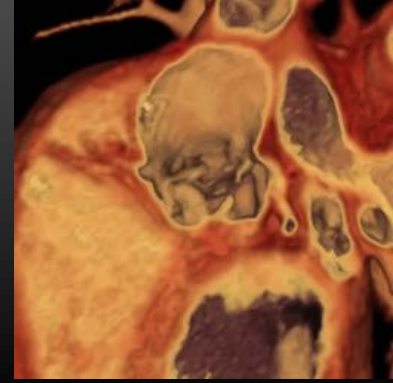


- **Unicuspid unicommissural** (one lateral commissural attachment, slit-like orifice)
- **Unicuspid acomissural** (no lateral attachments, pinhole orifice, neonates)
- Mean age of diagnosis 14 months
- #1 presentation: CHF
- #1 lesion: isolated AS (37%)

Singh S, et al. Texas Heart J. 2015; 42: 273-276

Mookadam F, et al. J Heart Valve Dis 2010; 19(6)678-83

CONGENITAL “UNICUSPID” AORTIC VALVE

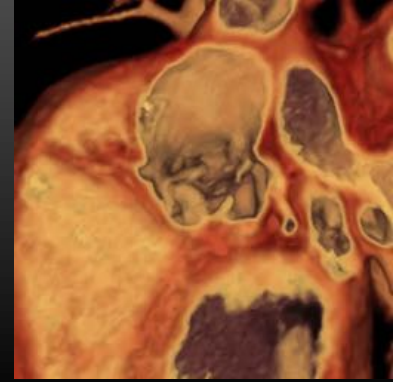


- Associated anomalies:
 - Coarctation (37%)
 - VSD (12%)
 - PDA (5%)
 - Aortic Aneurysm (5%)
 - Coronary artery anomalies

Singh S, et al. Texas Heart J. 2015; 42: 273-276

Mookadam F, et al. J Heart Valve Dis 2010; 19(6)678-83

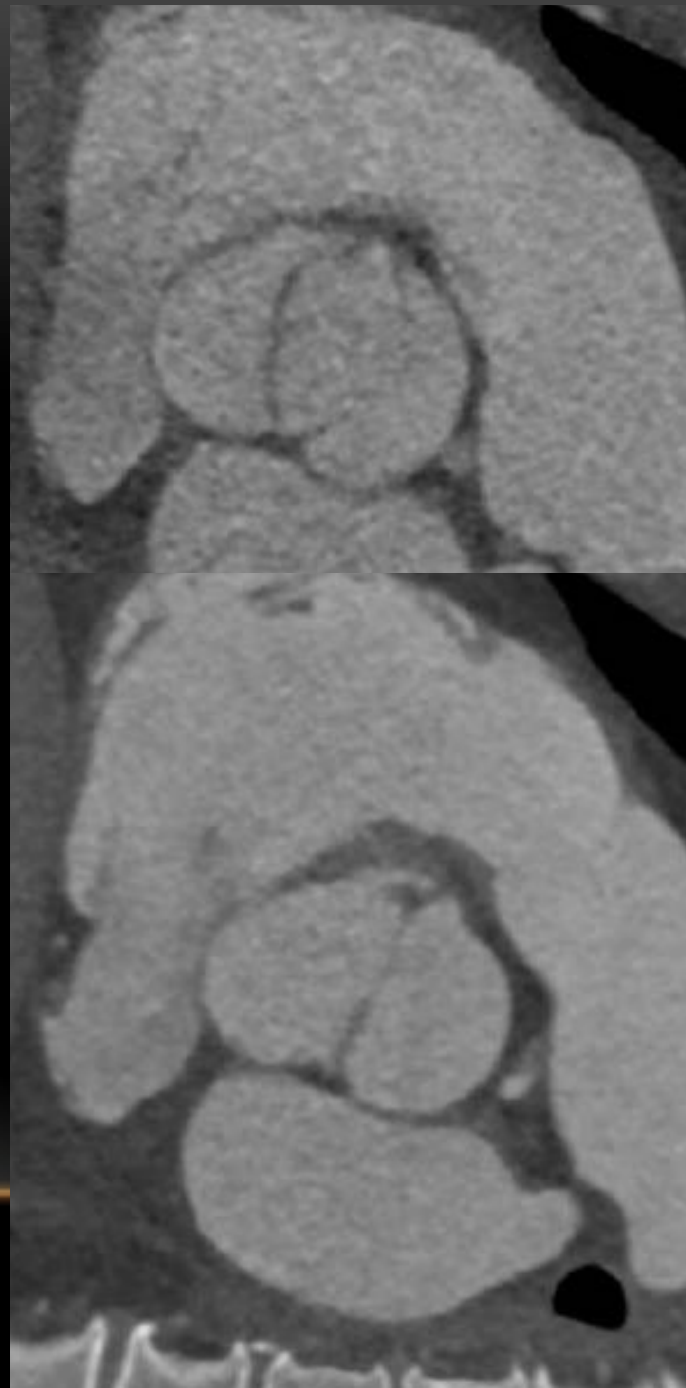
CONGENITAL “UNICUSPID” AORTIC VALVE



- Unicuspid (or functionally unicuspid) valves^{1,2}:
 - Have lower associated LVEF than BAV
 - Lower opening area (AVOA)
 - Greater fusion length
 - Higher incidence of leaflet tears and resultant aortic insufficiency after balloon v-plasty

1. Gao, K, et al. J Invasive Cardiol. 2016; 28(9):381-8.
2. Petit CJ, et al Am J Cardiol 2016; 117(6) 972-9.

CASE 2



BICUSPID AORTIC VALVE

SAM QUESTION:

Bicuspid aortic valve (BAV) is best characterized as:

- A. An isolated defect causing aortic valve abnormalities
- B. A systemic disorder with aortic tissue showing decreased levels of matrix metalloproteinases
- C. A systemic disorder that shares some histopathologic findings with Marfan syndrome
- D. A systemic disorder with sporadic inheritance

SAM QUESTION: ANSWER

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- B. A systemic disorder with aortic tissue showing decreased levels of matrix metalloproteinases
- C. A systemic disorder that shares some histopathologic findings with Marfan syndrome**
- D. A systemic disorder with sporadic inheritance

BICUSPID AORTIC VALVE

- 1-2% of the population
 - Decreased Fibrillin-1 levels (M)
 - Increased matrix metalloproteinases (M)
 - NOTCH1 and ACTA2 genes
 - Some inherited - autosomal dominant
-

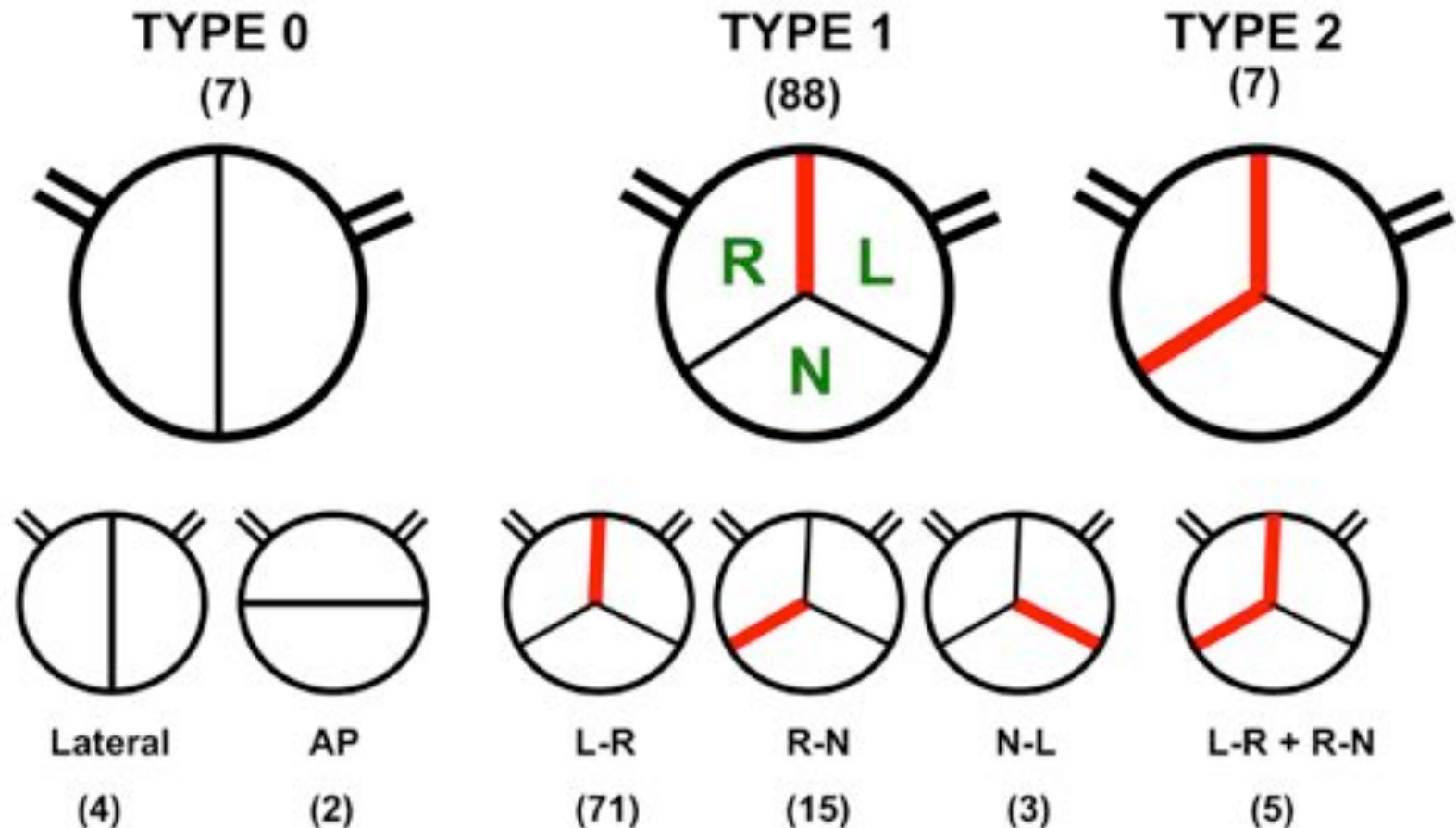
BICUSPID AORTIC VALVE

- Aortic Dilatation / Aneurysms
- Aortic Dissection
- Aortic Coarctation
- Family members of BAV patients can show aortic dilatation even though they have trileaflet valves

BAV: CLASSIFICATION

- Sievers and Schmidtke – 2007
- Classification based on 304 surgical BAV specimens
- Raphe: conjoint or fused portions of two adjacent leaflets that become a malformed commissure

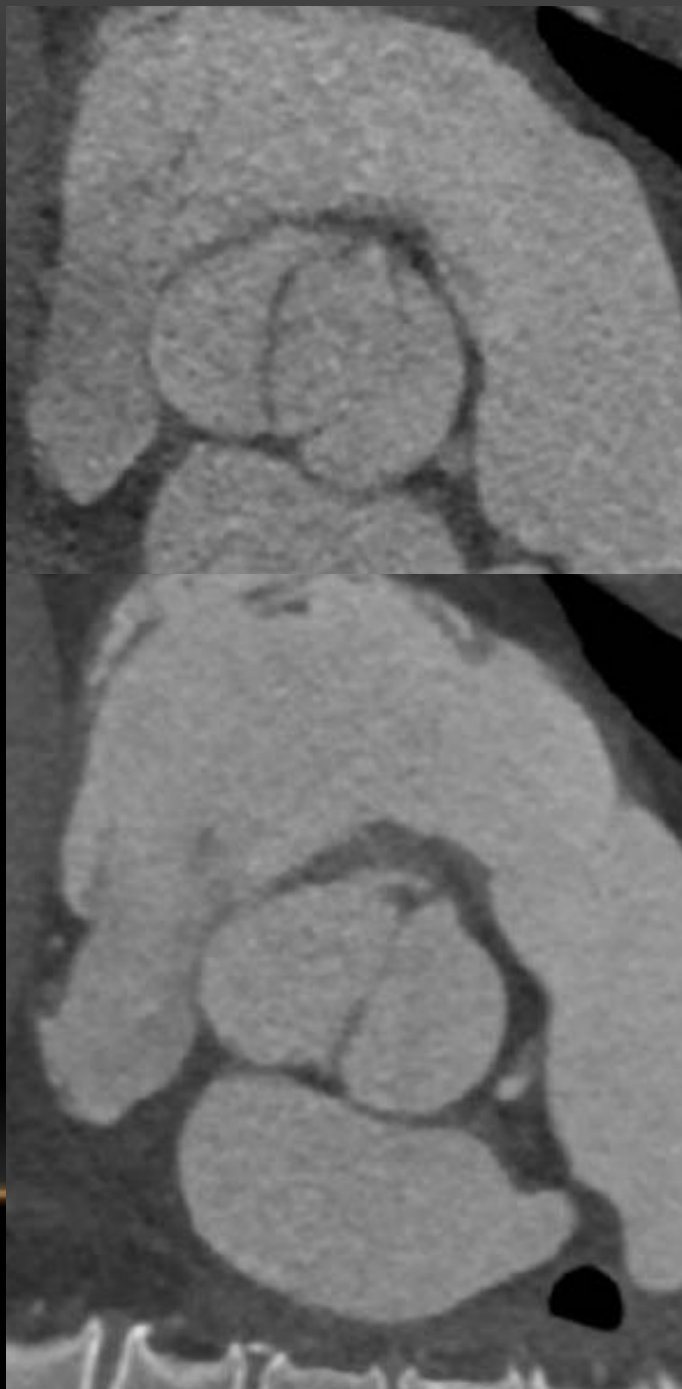
SIEVERS CLASSIFICATION OF BAV



S = Stenosis Dominant
 B = Balanced stenosis / insufficiency

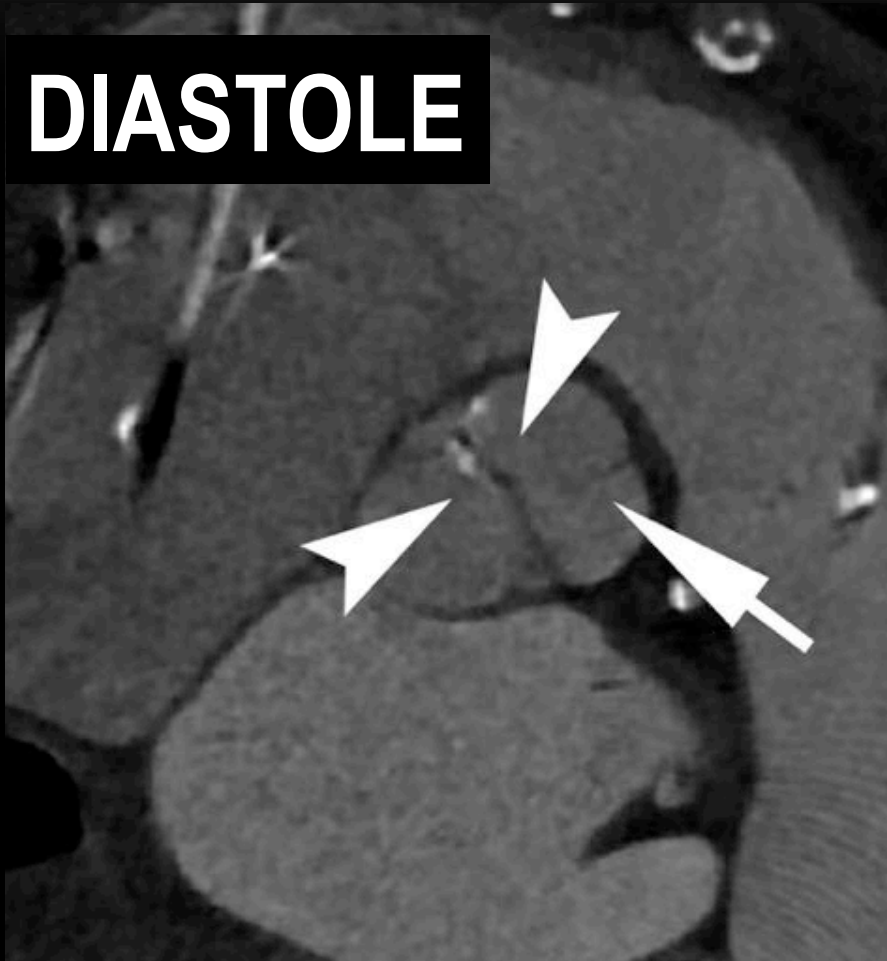
I = Insufficiency Dominant
 No = Normal Function

SIEVERS 0, LATERAL

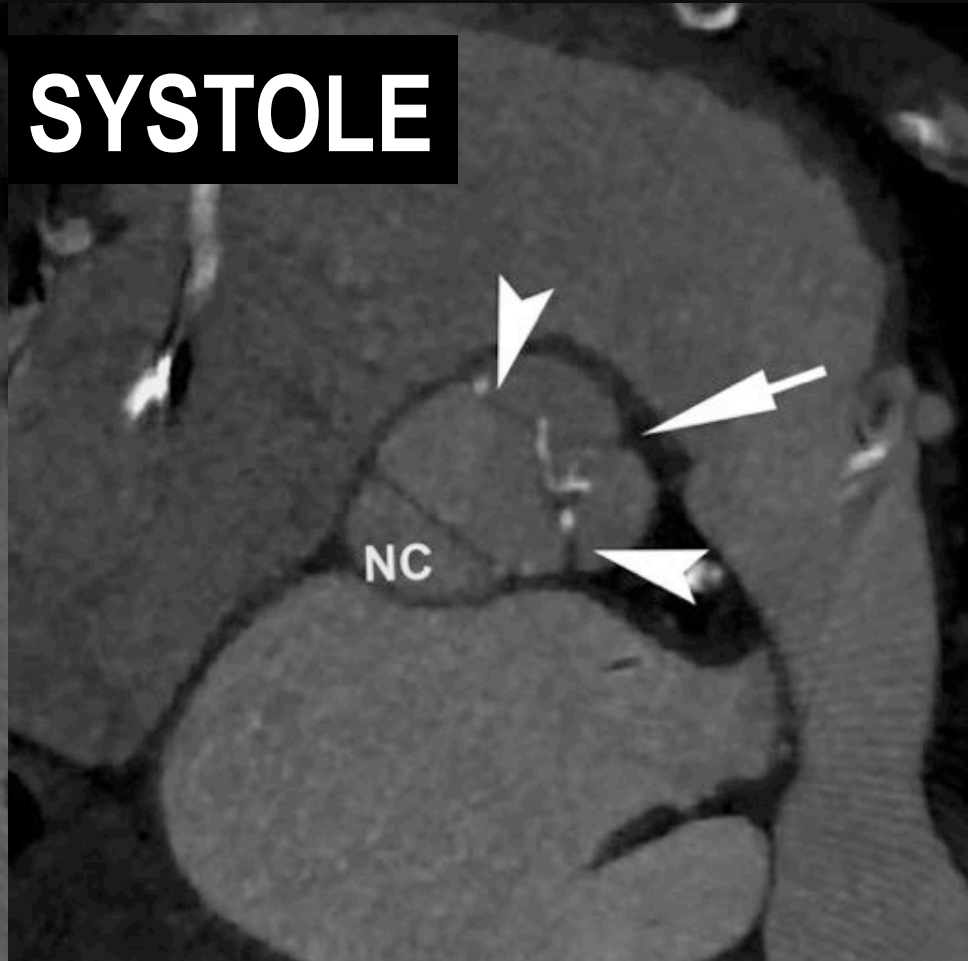


SIEVERS 1, L-R

DIASTOLE



SYSTOLE

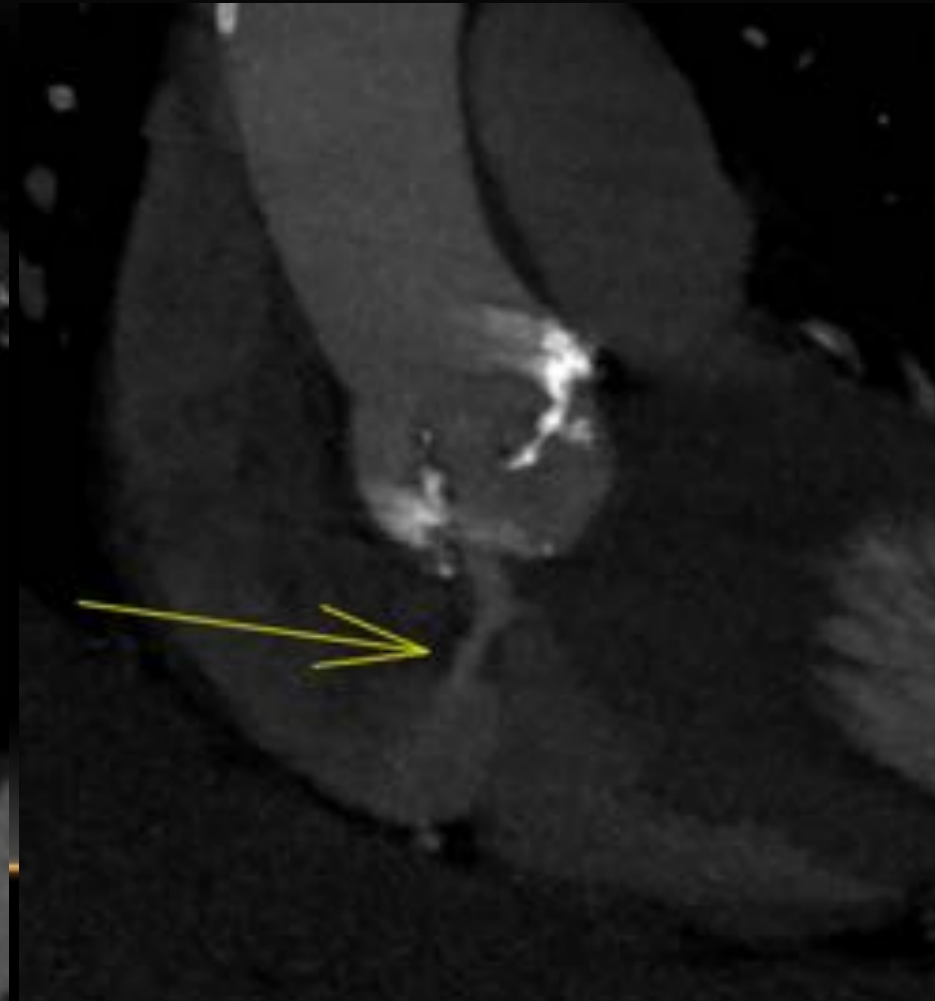
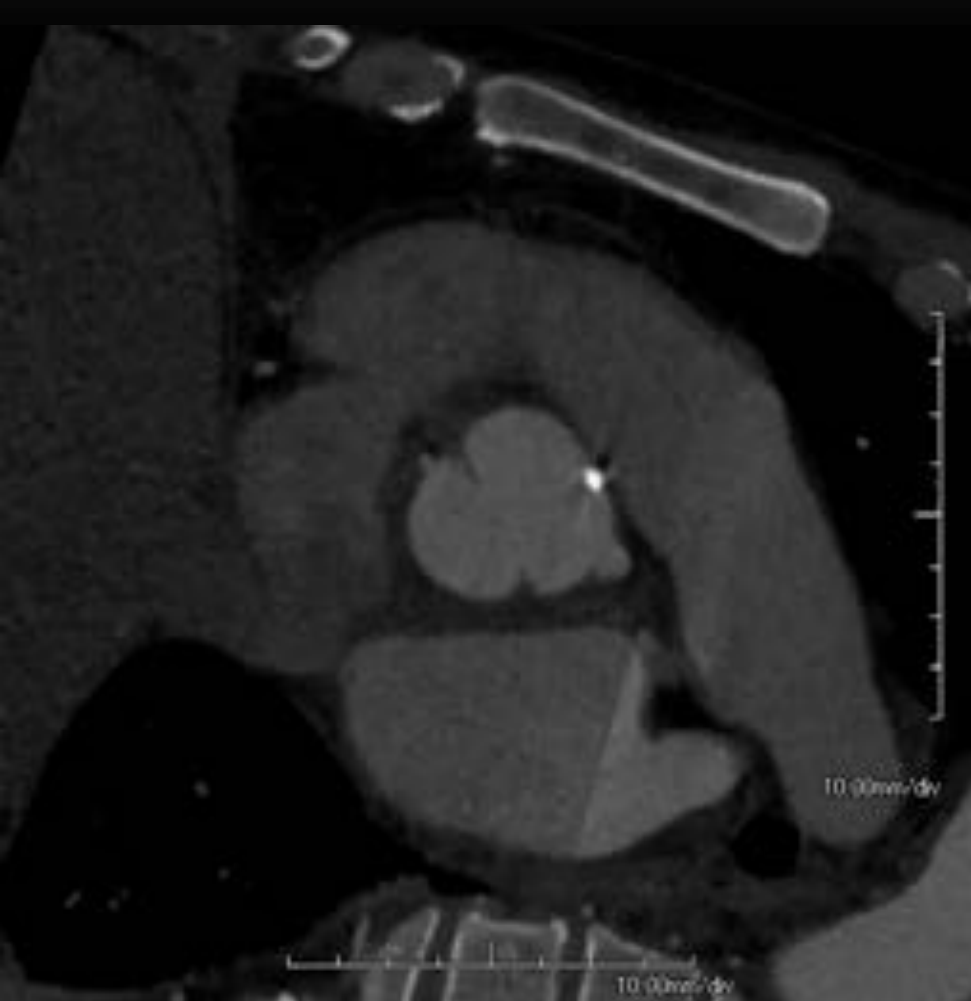


CASE 3

- 50 M w hx BAV, cerebral abscess, new heart failure

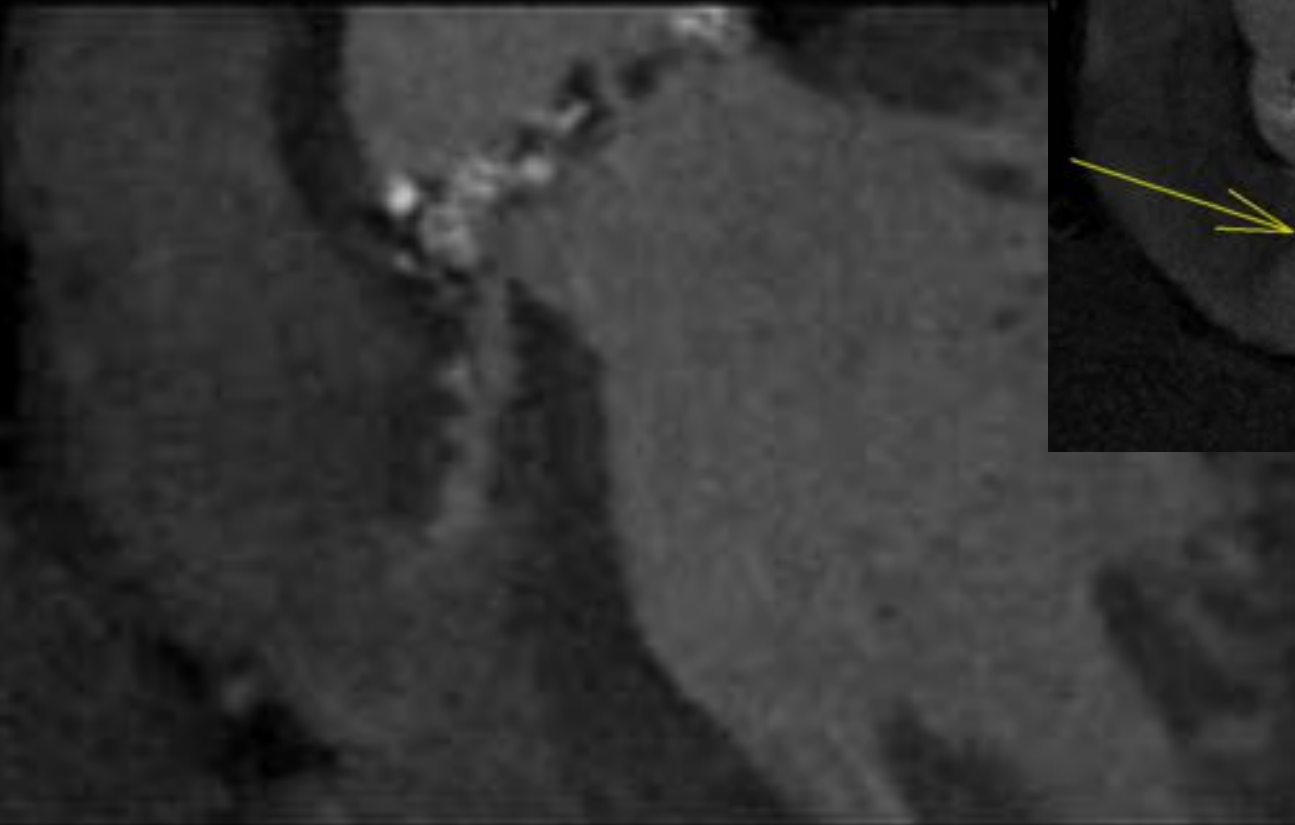
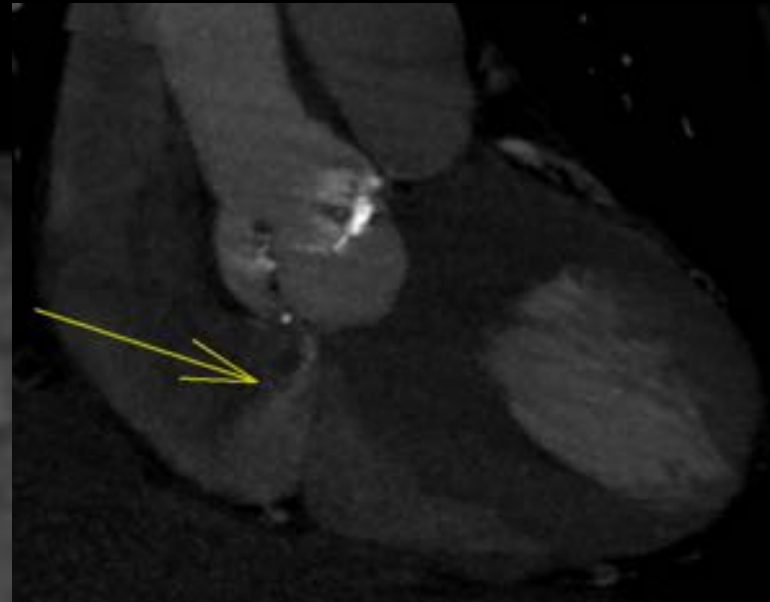
CASE 3

- 50 M w BAV, cerebral abscess



INFECTIOUS ENDOCARDITIS (IE) WITH SUBVALVULAR ABSCESS AND AORTO-RIGHT ATRIAL FISTULA

- HACEK endocarditis



CASE 3 QUESTION

Regarding HACEK infectious endocarditis (IE), which is true?

- A. HACEK are gram positive bacteria that are part of normal skin flora
- B. HACEK organisms cause a majority of infectious endocarditis (IE) cases
- C. HACEK IE patients tend to be younger than other IE patients
- D. Outcomes are poorer than other IE patients

CASE 3 QUESTION - ANSWER

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- D. Outcomes are poorer than other IE patients

Chambers ST et al. HACEK Infective Endocarditis: Characteristics and Outcomes from a Large, Multi-National Cohort

Abbate A, editor. PLoS ONE. 2013;8:e63181.

HACEK ENDOCARDITIS

- **H**aemophilus spp.
 - **A**ggregatibacter spp.
 - **C**ardiobacterium hominus
 - **E**ikenella corodens
 - **K**ingella spp.
-

HACEK ENDOCARDITIS

- Heterogenous group of gram negative fastidious bacteria
- Frequently colonize oropharyngeal
- Cause < 5% of IE cases
- Better outcomes
 - 11% 1-year mortality (vs 39%)

AORTO-RIGHT ATRIAL FISTULA^{1,2}

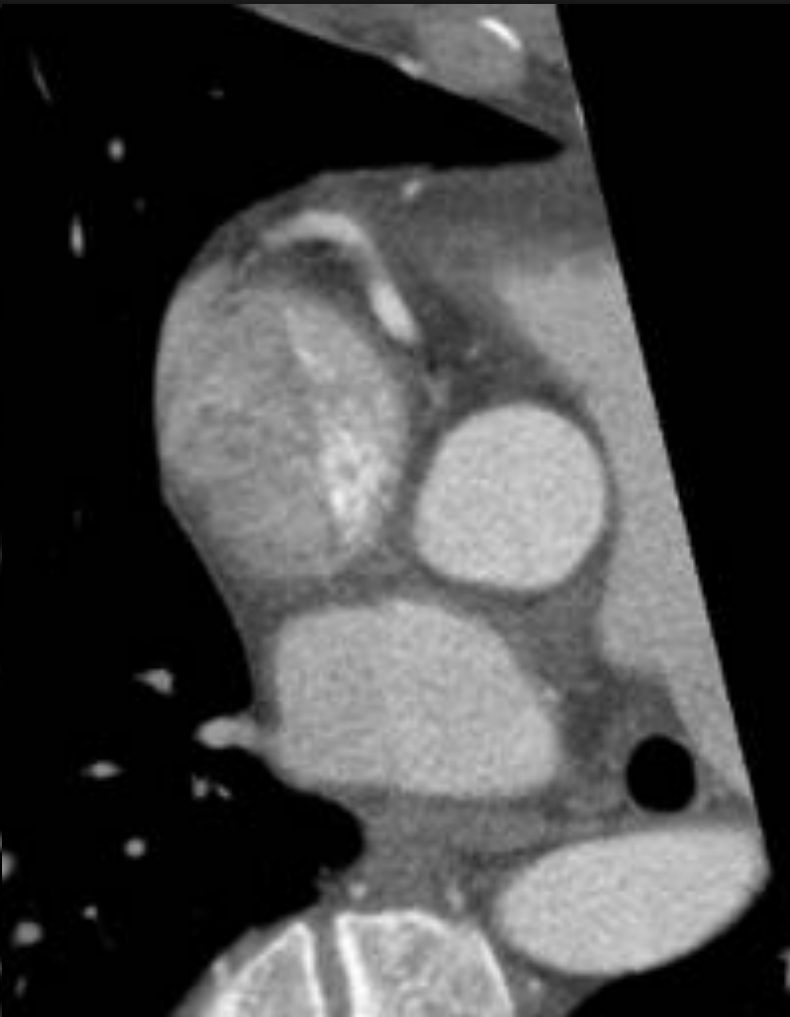
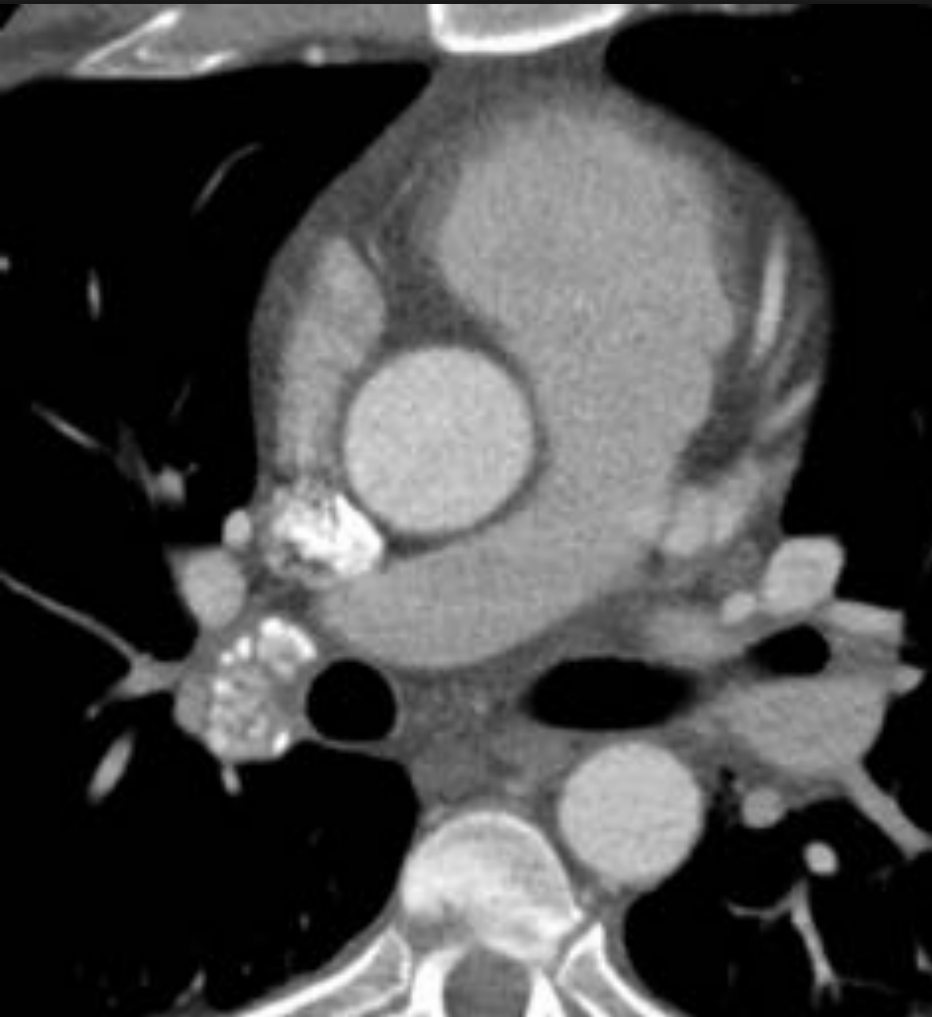
- Congenital
- Aneurysm / Dissection
- Infective endocarditis
 - Aorto-cavernous fistulae in only 1-2% IE pts
 - Staphylococcus: worse outcomes

CASE 4

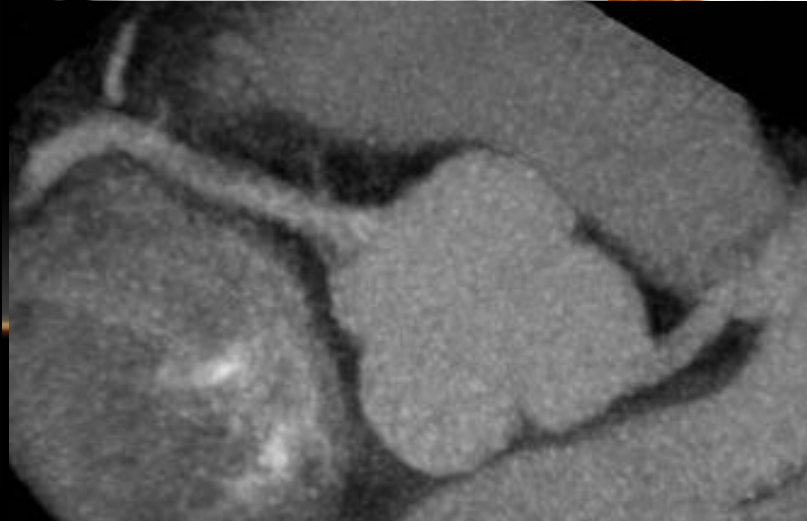
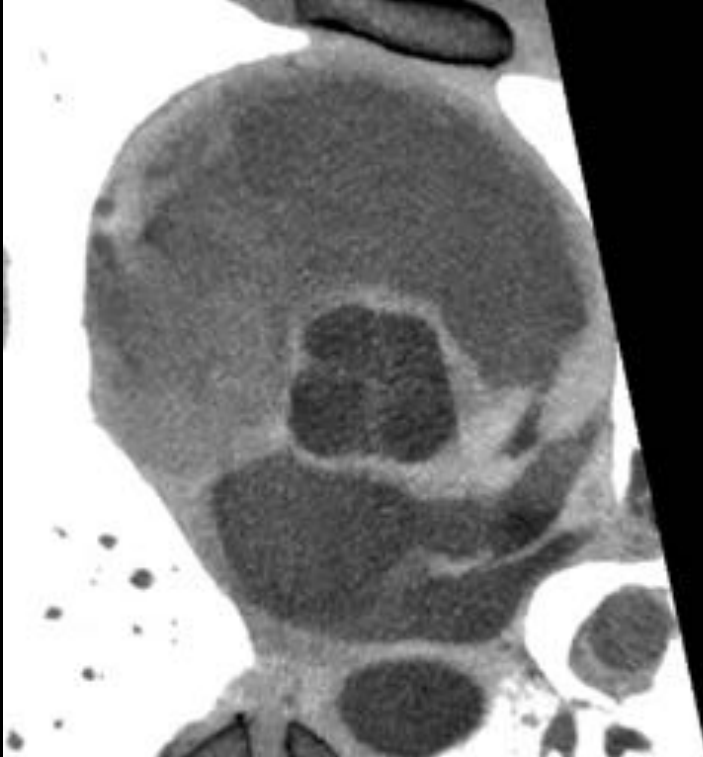
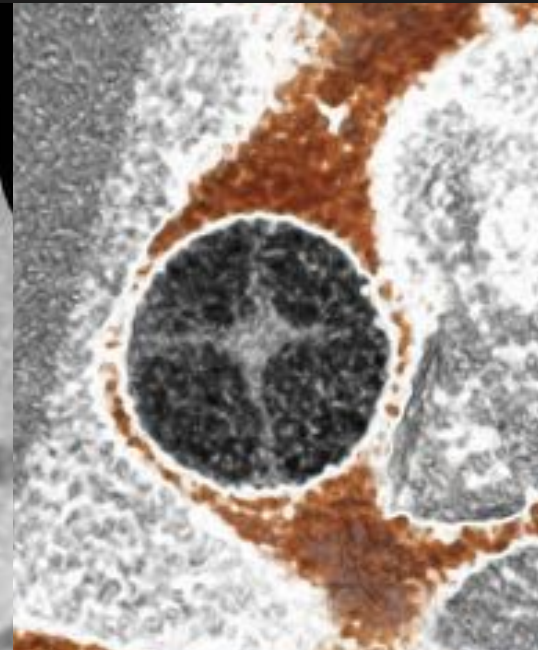
CASE 4

- 32 yr old male
- Atypical CP, equivocal stress echo
- Cath: “No vessel coming off R sinus”, concern for anomalous coronary artery

CASE FOUR



QUADRICUSPID AORTIC VALVE



QUESTION

The most common complication of Quadricuspid Aortic Valve (QAV) is:

- A. Valvular aortic stenosis
- B. Aortic regurgitation
- C. Atrial fibrillation
- D. Left ventricular hypertrophy

QUESTION: ANSWER

The most common complication of Quadricuspid Aortic Valve (QAV) is:

A. Valvular aortic stenosis

B. Aortic regurgitation

C. Atrial fibrillation

D. Left ventricular hypertrophy

QUADRICUSPID AORTIC VALVE (QAV)

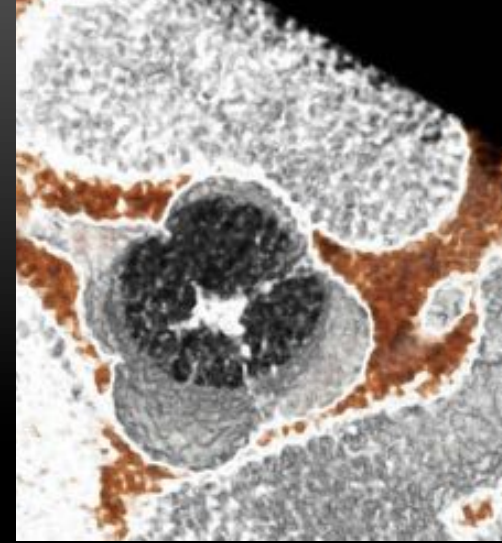
- Rare, 1/6000 aortic valve surgery patients¹
- M = F, avg. age at Dx ~ 50
- Classification by size of cusps²
 - Most common: 3 same size + 1 smaller cusp (type B)
- Echo: “X”-shaped SAX view
- CT/MR: confirmatory; perform planimetry and/or flow measurement³

1. Douglas, H., Moore, M., & Purvis, J. (2012) *Heart* 2012; 98(24), 1838-1838.

2. Hurwitz L, et al. *Am J Cardiol* 1973; 31(5) 623-626.

3. Khan SK, Tamin SS, Araoz PA. *J Comput Assist Tomogr.* 35 (5): 637-41.

CLASSIFICATION OF QAV¹



A	4 equal sized cusps
B	3 equal + 1 smaller (most common)
C	2 equal + 2 equal smaller
D	1 large + 2 intermediate + 1 smaller
E	3 equal + 1 larger
F	2 equal large + 2 smaller unequal sizes
G	4 unequal sized cusps

1. Hurwitz L, et al. *Am J Cardiol* 1973; 31(5) 623-626.

QUADRICUSPID AORTIC VALVE (QAV)

- **Usually isolated**, but can be associated with:
 - Single or Anomalous Coronary Arteries
 - Displacement of coronary ostia (from aortic cusp)
 - HCM / Subaortic Stenosis
 - PDA, VSD
 - Endocarditis

1. Jagganath AD, et al. *Echocardiography* 2011; 28(9), 1035-1040.

2. Zhu J, et al. *J Cardiothor Surg* 2013; 8(1) 87.

3. Tutarel O. *J. Heart Valve Dis.* 2004;13 (4): 534-7.

QUADRICUSPID AORTIC VALVE (QAV)

- Complications:
 - **Aortic Insufficiency (#1)**
 - Up to 75% at time of Dx
 - LVH
 - Conduction problems (BBB)
- TX: Reconstruction and/or Surgical valve replacement



1. Jagganath AD, et al. *Echocardiography* 2011; 28(9), 1035-1040.
2. Zhu J, et al. *J Cardiothor Surg* 2013; 8(1) 87.
3. Tutarel O. *J. Heart Valve Dis.* 2004;13 (4): 534-7.

CONCLUSION

- Whether 1, 2, 3, or 4 cusps, aortic valve pathology can be well assessed by ECG synchronized CTA

THANKS FOR YOUR ATTENTION!!

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