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Cases: Adult Congenital Heart Disease

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October 10, 2017; ASE Echo Florida

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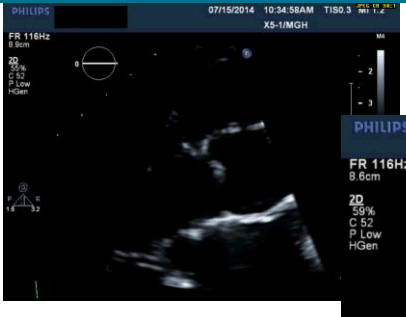

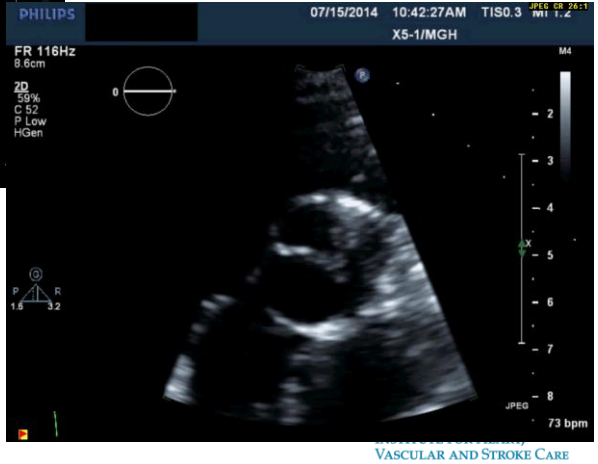

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The following is a true statement:

- A. The open commissure in a unicuspid valve is most often between the right and left cusps
- B. The open commissure in a unicuspid valve is always between the left and non cusps
- C. AVA planimetry can be accurate with unicuspid valves

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3

Case: LM, angina evaluation

- 62F history of hypertension, prior breast cancer
- Murmur all her life
- Generally very well, daily exercise, lives in FL but visiting in Boston
  - Chest heaviness with inclines
- Recent 6 months: exertional chest heaviness, relieved with rest
- No dyspnea or cough, no dysphagia

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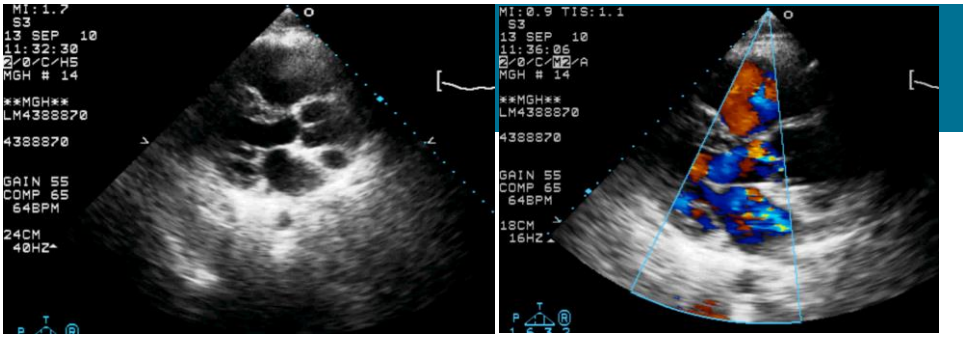

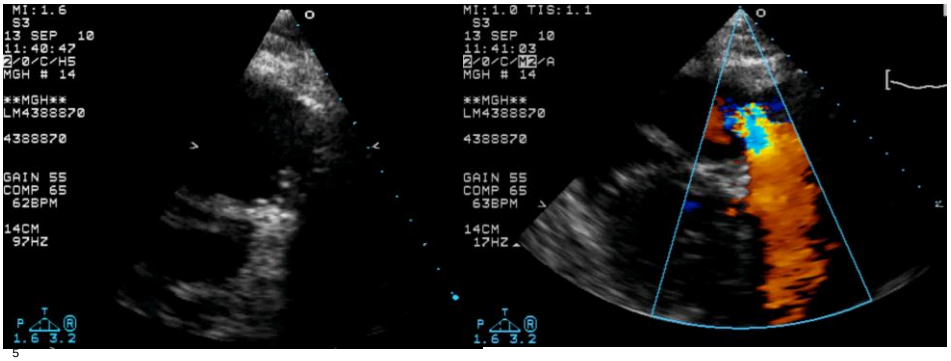

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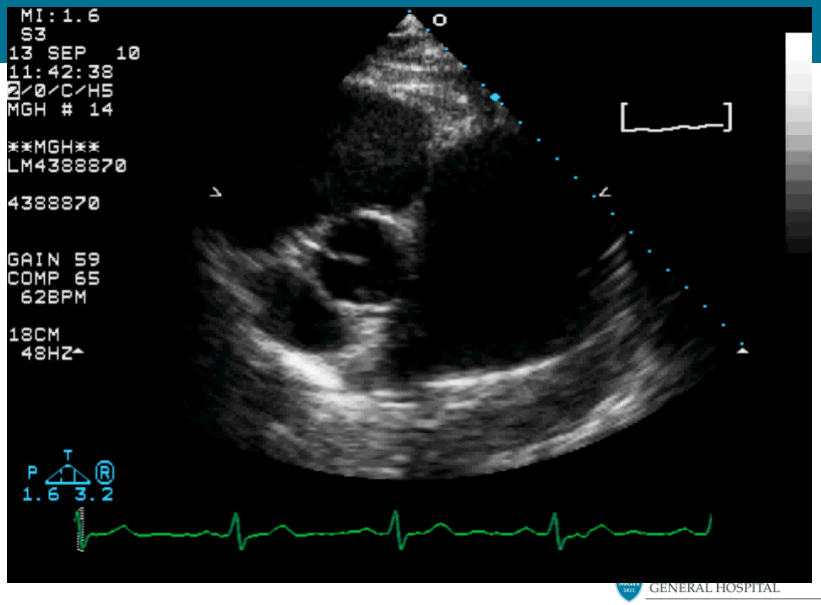

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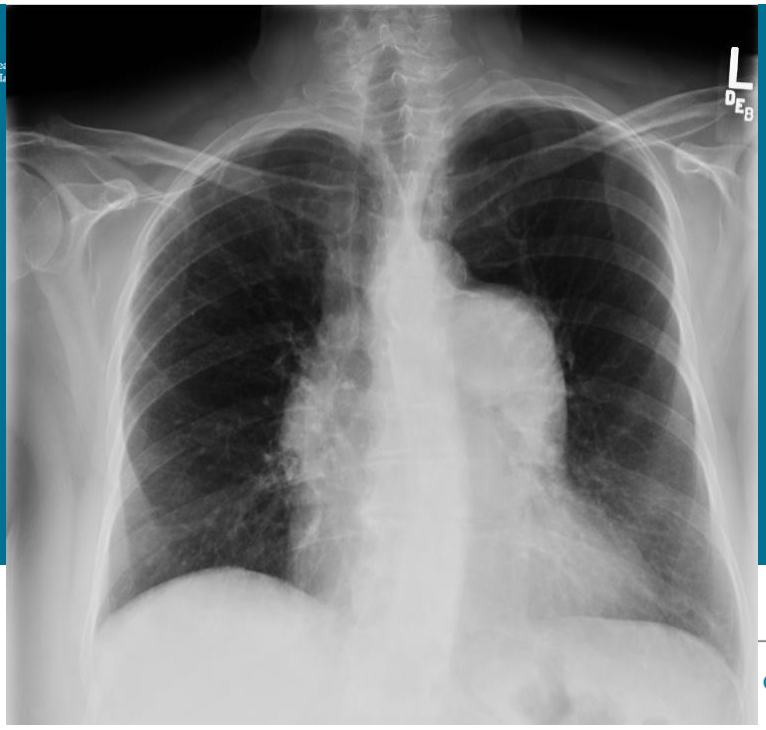
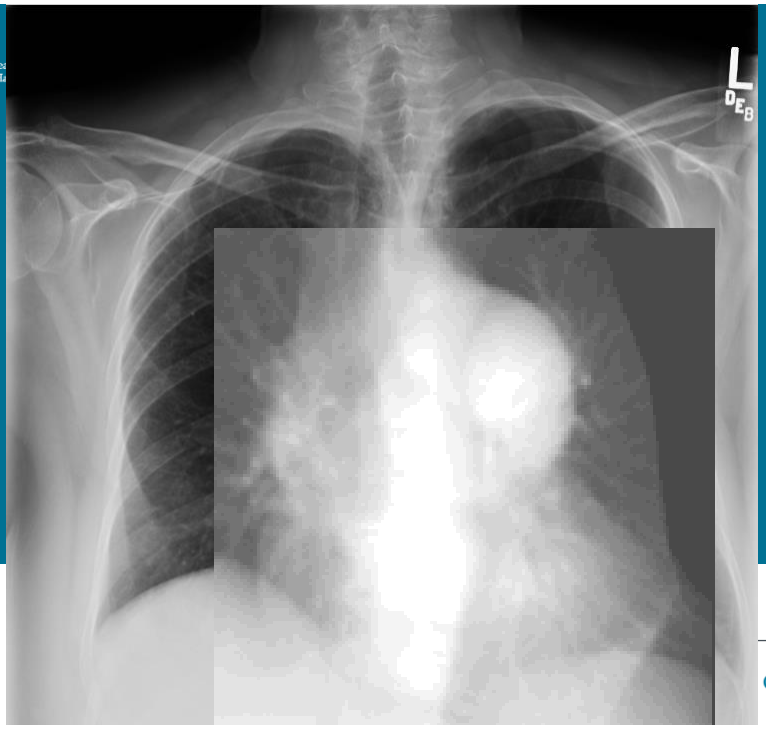
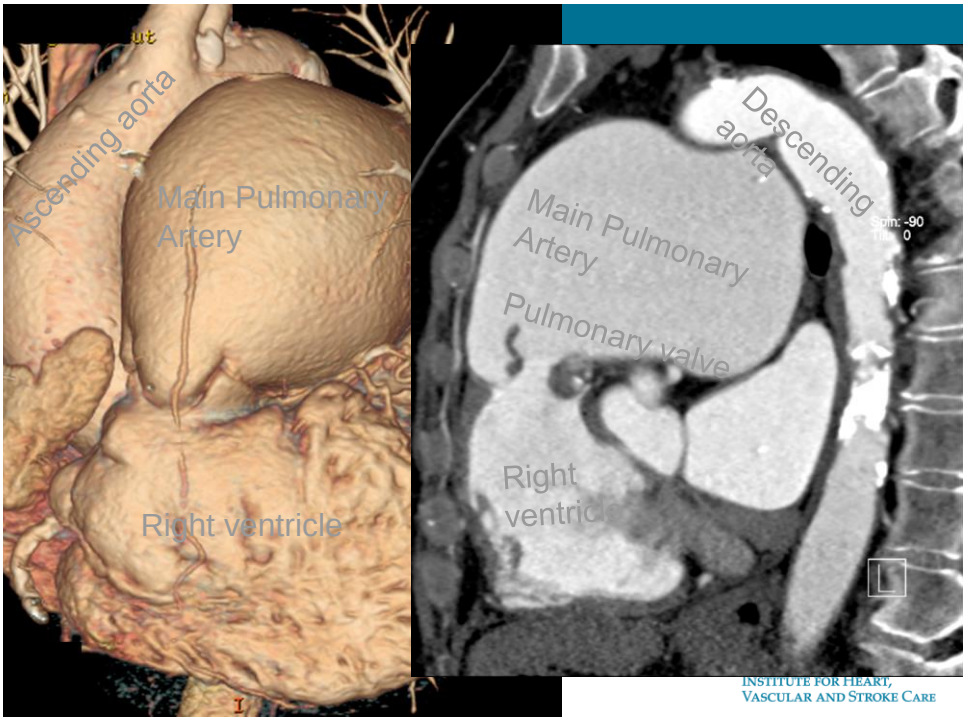


Care



Care


Ascending aorta
Main Pulmonary Artery
Right ventricle
Descending aorta
Main Pulmonary Artery
Pulmonary valve
Right ventricle
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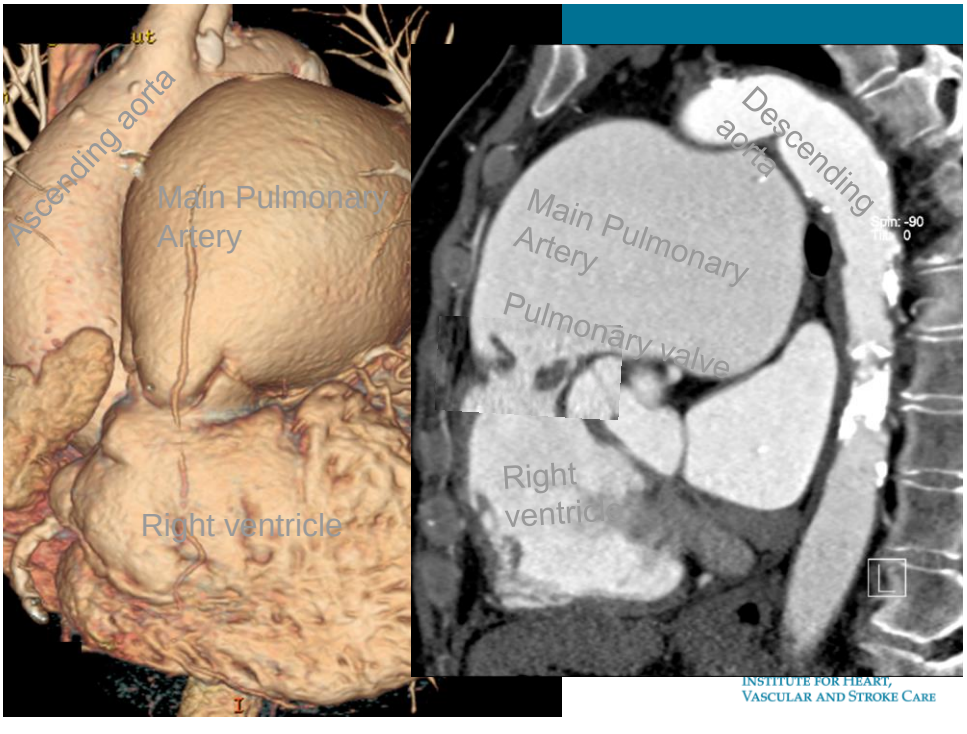
Ascending aorta
Main Pulmonary Artery
Right ventricle
Descending aorta
Main Pulmonary Artery
Pulmonary valve
Right ventricle
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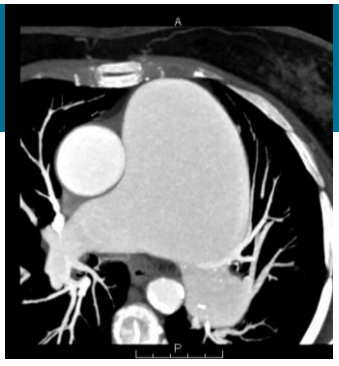

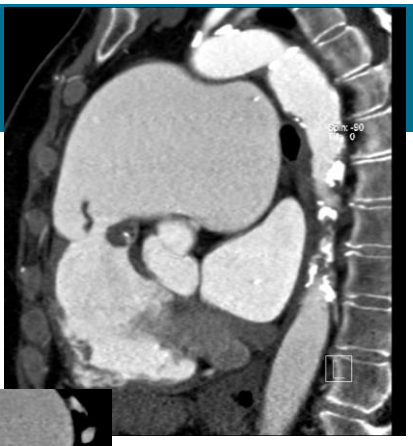

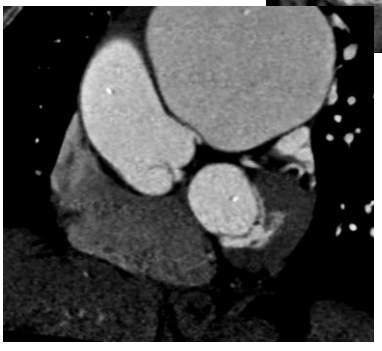

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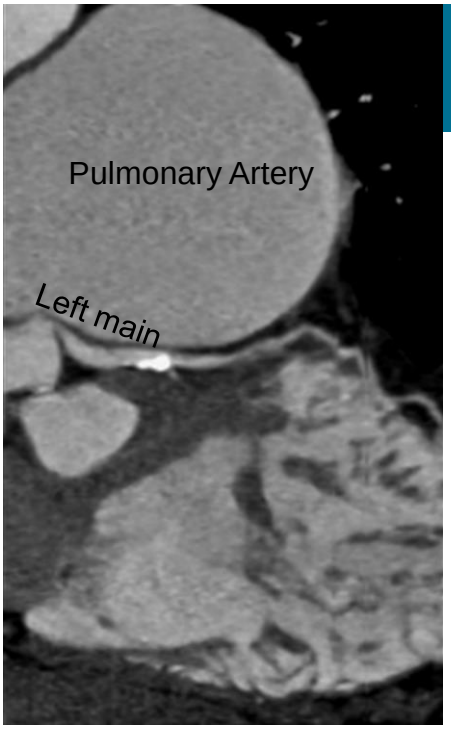

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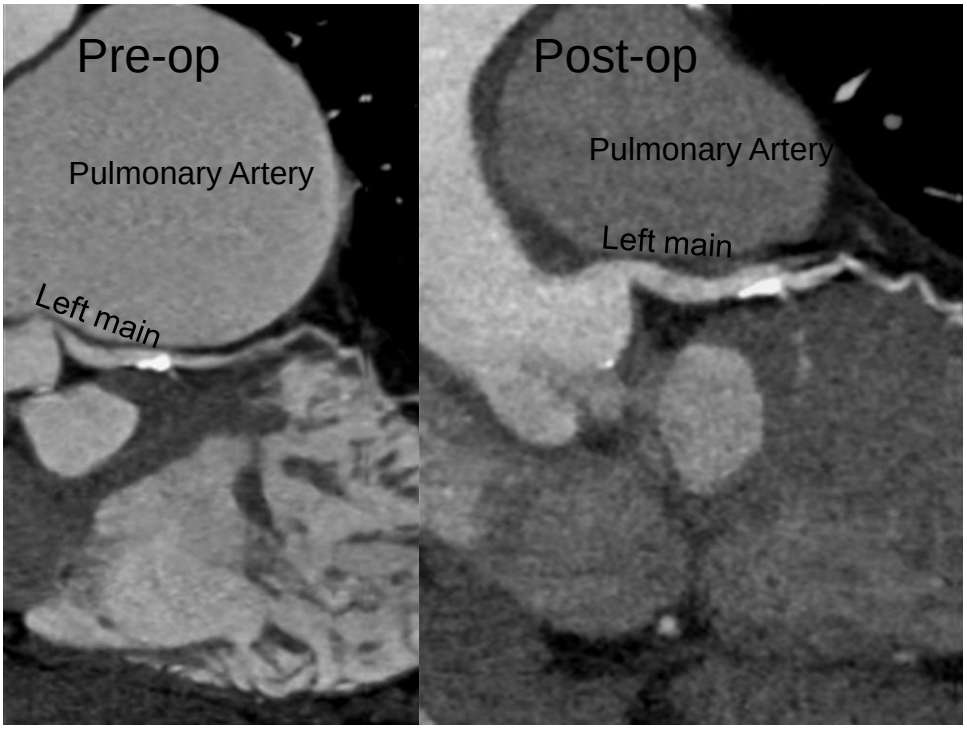
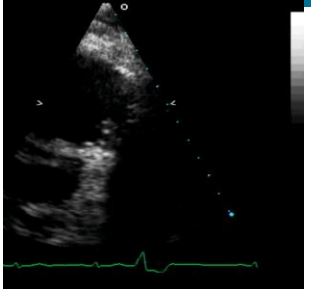


Pulmonic Stenosis:



- Where is the obstruction?
  - Supravalvular (branch)
  - Subvalvular
  - Valvular → <u>associated with pulm arteriopathy and PA aneursyms</u>
- RVSP **<u>does not</u>** equal PASP
- Intervention if valvular PS:
  - Peak gradient > 60mmHg
  - Peak > 50mmHg if symptoms
  - Absence of significant PR

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Case: 50F with dyspnea

- Progressive fatigue and dyspnea at work
- Difficulty lifting
- Co-workers noted her chest would heave

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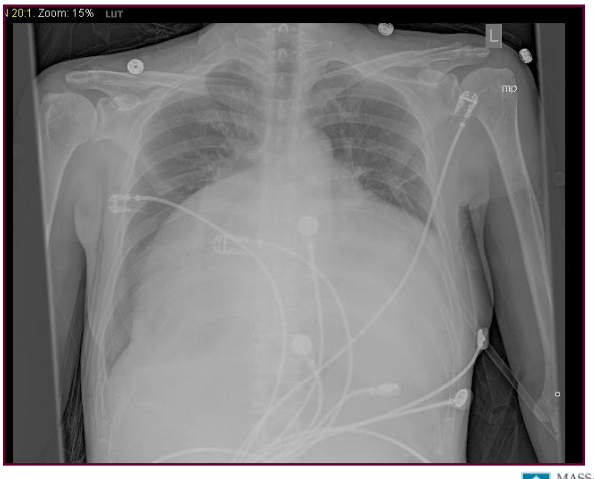
50F with dyspnea



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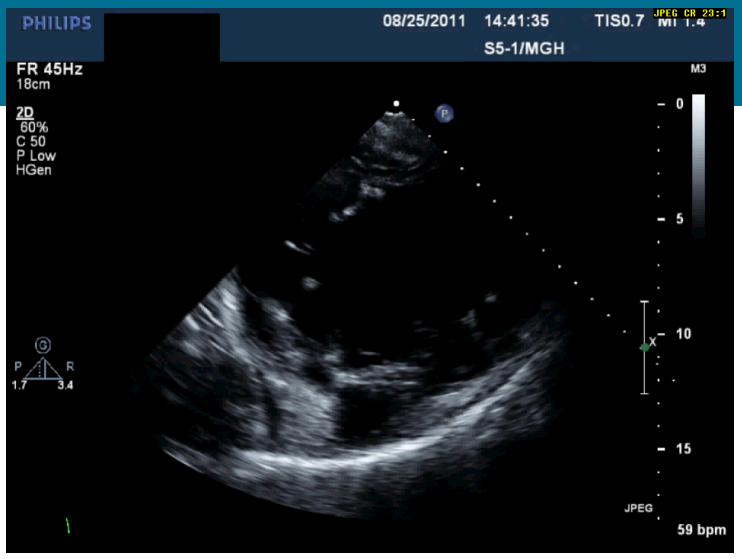

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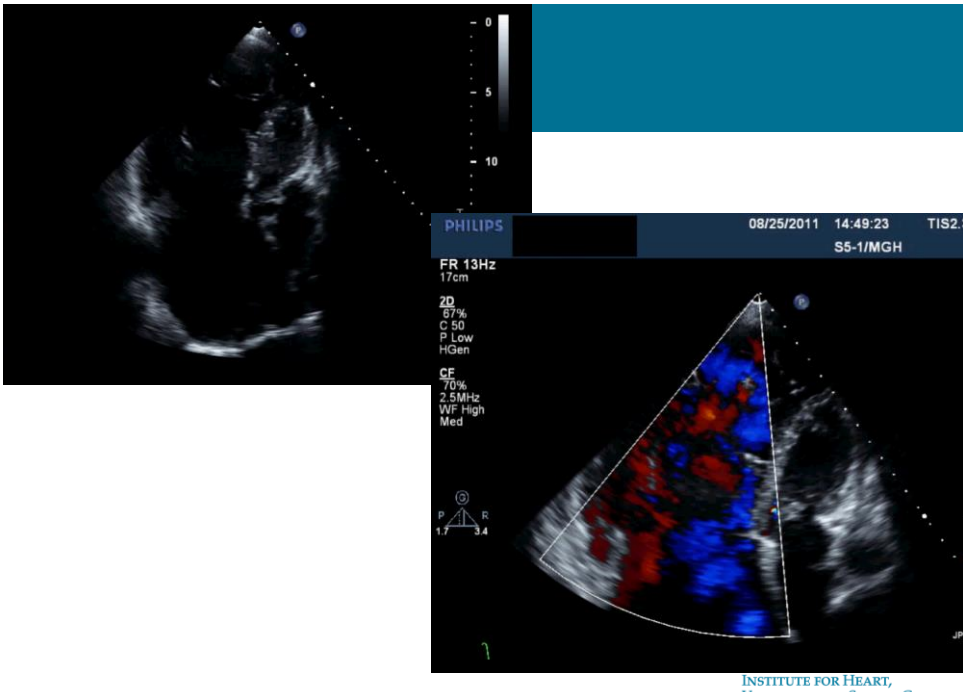

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Diagnosis?

A. Giant RA syndrome

B. Ebsteins anomaly

C. Dysplastic tricuspid valve syndrome

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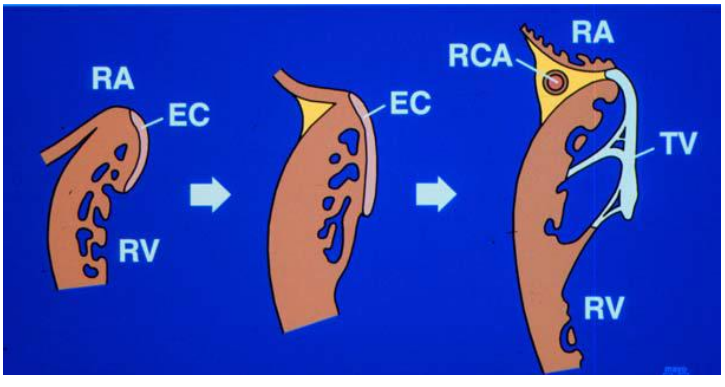
Normal delimination (separation) of the RV from the RV myocardium



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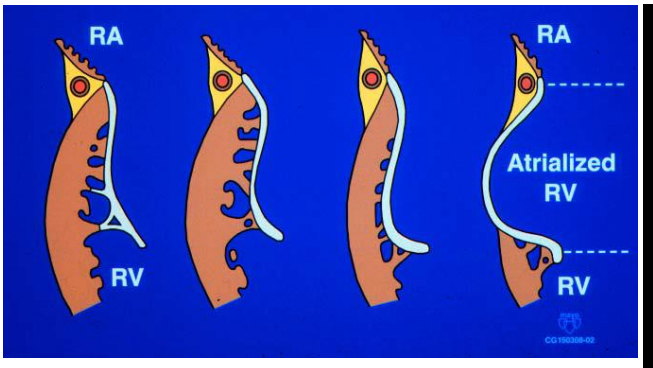
Failed TV delamination: Ebstein Anomaly



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Case II:

- 78 year old female with a murmur as a child
  - Ebstein's anomaly diagnosed after 3rd pregnancy
  - Intermittent atrial arrhythmias
    - managed with digoxin, no prior ablation
  - Sat 95% on RA, normal HCT, euvolemic

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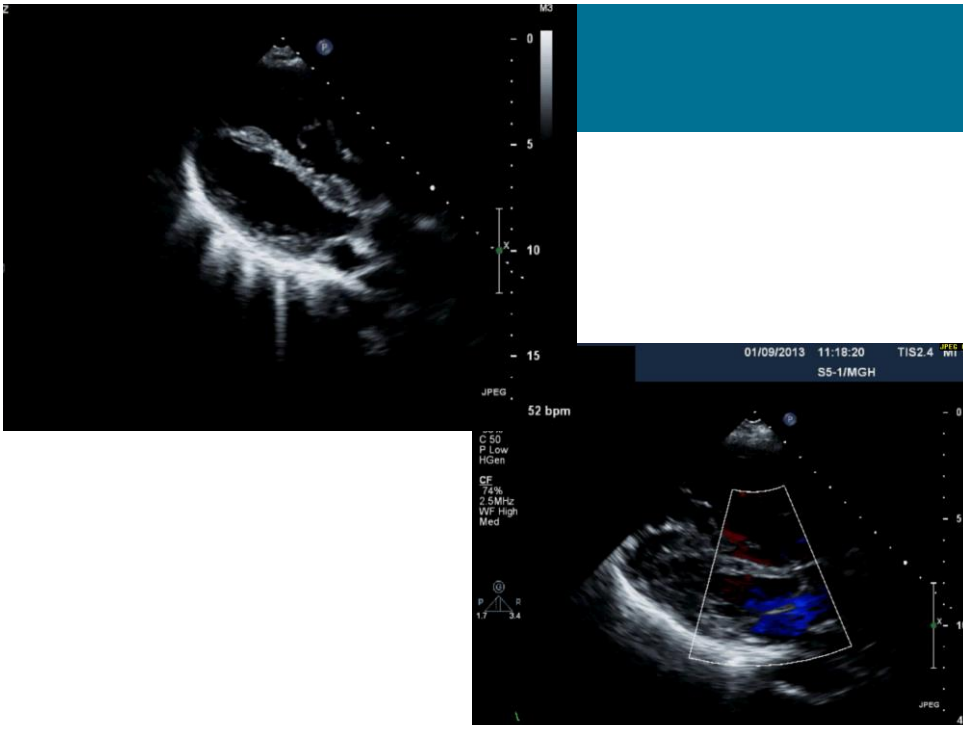
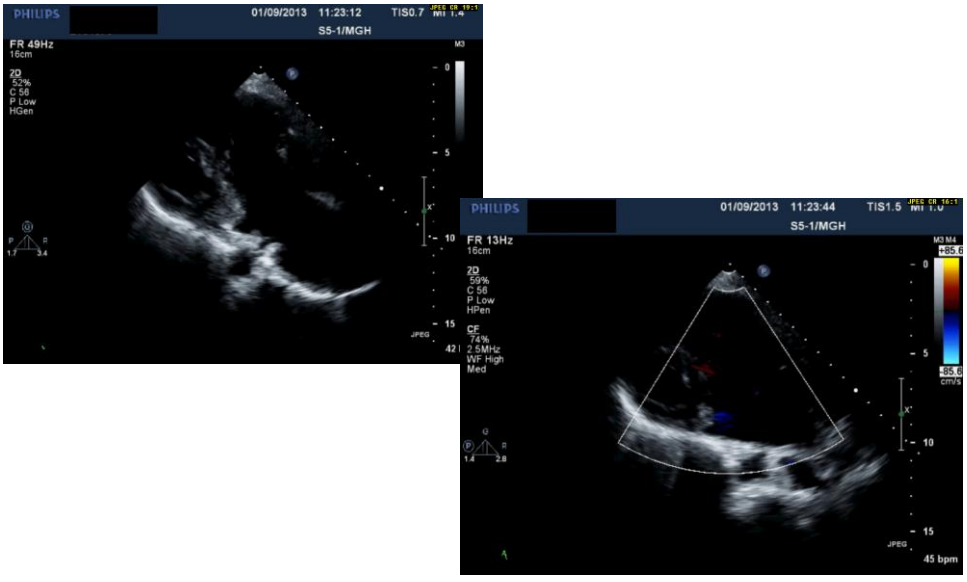
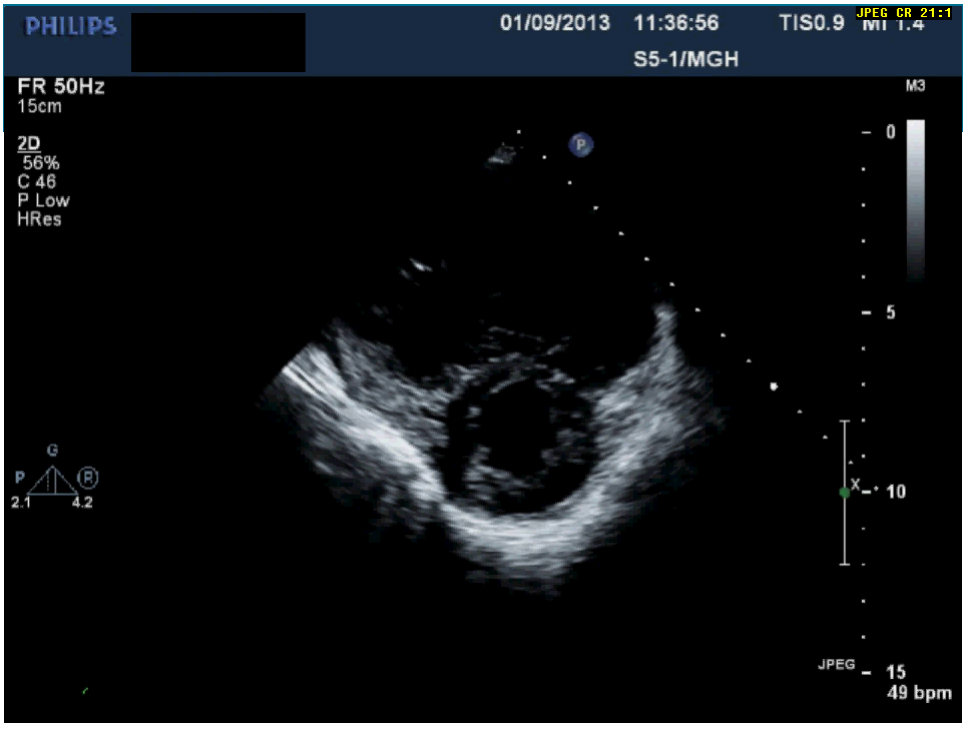
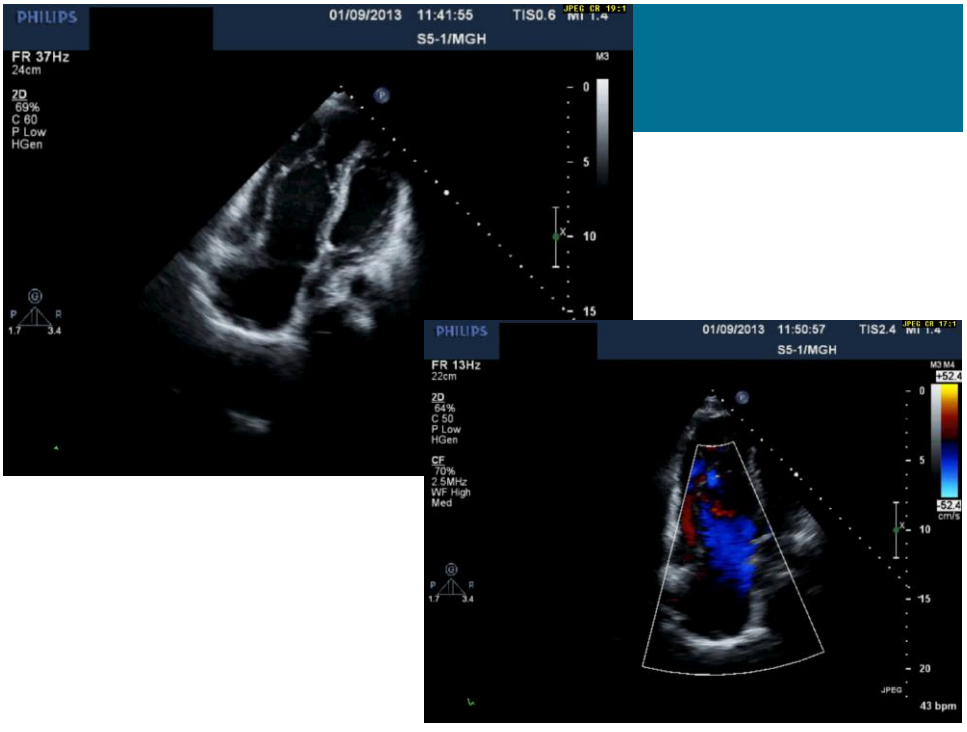
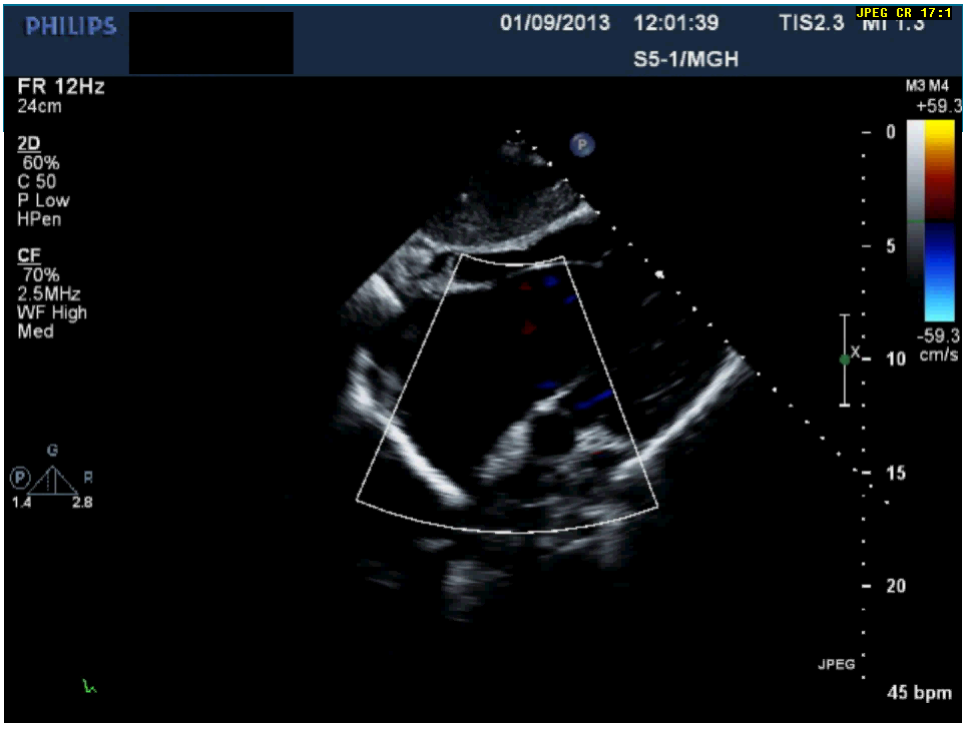

01/09/2013 11:18:20 TIS2.4
S5-1/MGH
52 bpm
JPEG


PHILIPS
01/09/2013 11:23:12 TIS0.7
S5-1/MGH
FR 49Hz
16cm
01/09/2013 11:23:44 TIS1.5
S5-1/MGH
FR 13Hz
16cm
45 bpm
JPEG

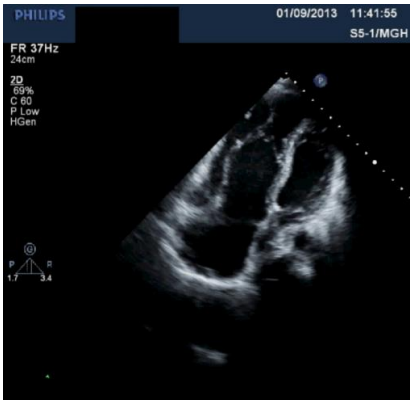

PHILIPS
01/09/2013 11:36:56 TIS0.9
S5-1/MGH
FR 50Hz
15cm
2D
56%
C 46
P Low
HRes
M3
0
5
10
15
JPEG
49 bpm


PHILIPS
01/09/2013 11:41:55 TIS0.6
S5-1/MGH
FR 37Hz
24cm
2D
69%
C 60
P Low
HGen
M3
PHILIPS
01/09/2013 11:50:57 TIS2.4
S5-1/MGH
FR 13Hz
22cm
2D
64%
C 50
P Low
HGen
CF
70%
2.5MHz
WF High
Med
M3 M4
+52.4
-52.4
cm/s
JPEG
43 bpm



Ebstein Anomaly Key Points:

- Degree of TR, RV function, desaturation will determine symptoms
- Wide spectrum of anatomic variation
- 50% with ASD/PFO
  - Desaturation
  - Paradoxical embolization
- Arrhythmia: WPW



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Thank You

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