Cases: Adult Congenital Heart Disease

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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

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
Pulmonic Stenosis:

- Where is the obstruction?
  - Supravalvular (branch)
  - Subvalvular
  - Valvular \( \rightarrow \) associated with pulm arteriopathy and PA aneurysms

- RVSP **does not** equal PASP

- Intervention if valvular PS:
  - Peak gradient > 60mmHg
  - Peak > 50mmHg if symptoms
  - Absence of significant PR
Case: 50F with dyspnea

- Progressive fatigue and dyspnea at work
- Difficulty lifting
- Co-workers noted her chest would heave
Diagnosis?
A. Giant RA syndrome
B. Ebsteins anomaly
C. Dysplastic tricuspid valve syndrome

Normal delimitation (separation) of the RV from the RV myocardium
Failed TV delamination: Ebstein Anomaly

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
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
Thank You
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