67F with incidental finding

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Ebstein Anomaly
Atrialization of RV/enlarged RA

Apically displaced leaflet
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Normal attachment of anterior leaflet
CT findings

• Right-chamber enlargement with "atrialized" portion of right ventricle
• Apical displacement of septal/posterior tricuspid valve leaflet
  – Anatomic annulus remains in normal position
  – Functional annulus is displaced apically
• Anterior leaflet has normal level of attachment, but often morphologically abnormal with redundancy, fenestrations, and tethering
• Normal-sized left atrium and ventricle
Ebstein Anomaly

- 3 compartments:
  - Right atrium
  - Atrialized noncontracting inlet portion
  - Functional outlet portion of right ventricle

- Embryology: Insufficient separation of tricuspid valve leaflets and chordae tendineae from right ventricular endocardium
  - Most often sporadic
  - Lithium association

- Associated abnormalities:
  - PFO
  - Secundum atrial septal defect in 90%
Ebstein Anomaly

• Presentation:
  – Sx: asymptomatic, chronic right heart failure, cyanosis (depends on difference between right and left atrial pressures)
  – Age: young to old; avg 14 years
• 0.5% of congenital cardiac defects
  – Most common cause of tricuspid regurg
• M=F
• Prognosis: dependent on hemodynamic significance of tricuspid regurgitation and presence of cyanosis
• Rx: Supportive care when cyanotic, tricuspid valve repair/recon is definitive, arrhythmia treatment
References

1. Stat dx
2. Radiopaedia