Cases in Adult Congenital Heart Disease

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No Disclosures
“I Have Palpitations”
18 Year old Man

- Palpitations
- “abnormal” ecg and cxr
Normal Delamination of the TV from the RV Myocardium
Failure of Delamination From the Myocardium

Spectrum with Infinite Variability
Failed Delamination results in ...

- adherence of leaflets to underlying RV myocardium
- displacement of the anular hinge points
Displacement Apically AND Toward the Right Ventricular Outflow Tract
Echocardiographic Diagnosis

- Apical displacement of the septal leaflet of the tricuspid valve > 8mm/m²
- Right sided chamber enlargement with “atrialized” RV
- Tricuspid valve regurgitation – often appears laminar
- Elongated, tethered anterior TV leaflet
Ebstein Anomaly Associated Lesions

- Secundum ASD
- RV outflow tract obstruction
- LV non-compaction
- Accessory pathways
Ebstein Anomaly
Indications for Operation

- symptoms, ↓ exercise tolerance, cyanosis
- progressive RV dilatation
- *before* significant RV dysfunction
- onset, progression of atrial arrhythmias
- ? earlier operation if TV repair is likely
- prior to LV dysfunction
“I Have a Headache”
36 Year Old Man

- Undergoing evaluation in neuro for headache
- Found to be hypertensive
Is This Coarctation?

A. Yes
B. No
C. Not Sure
Is This Coarctation?

A. Yes
B. No
C. Not Sure
Abdominal Aorta Doppler
Imaging of Coarctation of the Aorta

- Abdominal aorta Doppler
- Suprasternal notch imaging
- Parasternal short axis - ?BAV
- Parasternal long axis – ascending aortic dimension
Discrete Coarctation
Coarctation Caveats

- Doppler gradient through the coarctation may be low 2° collaterals
- Abdominal Doppler pattern is critical
- Continuous flow in the thoracic aorta is helpful
- Don’t forget association to BAV
“Abnormal ECG on Life Insurance Exam”
31 year old man

Told as a child he had a “hole in his heart” – no intervention
EKG
Diagnosis?

A. Non-compaction cardiomyopathy
B. L-TGA
C. D-TGA
D. Ebstein anomaly
Diagnosis?

A. Non-compaction cardiomyopathy
B. L-TGA
C. D-TGA
D. Ebstein anomaly
Congenitally Corrected Transposition
Complete Transposition (D-TGA)

Congenitally Corrected Transposition (L-TGA)
Conus present in the “LVOT”
Left A-V valve displaced apically
Side-by-side semi-lunar valves
Lesions Associated with ccTGA

- Ventricular Septal Defect (70%)
- Subpulmonary ventricular outflow tract obstruction (40%)
- Tricuspid valve dysplasia/Ebstein malformation (90%)
- Situs Inversus
- Dextrocardia
“Second Opinion”
38 Year Old Woman

- Present for second opinion re: treatment of pulmonary hypertension
- Significantly limited
- Marked cyanosis
Past Medical History

- Evaluated at 3 months of age for pneumonia
- Diagnosed with VSD, PDA, coarctation
- PA banding, PDA ligation and coarctation repair performed
Past Medical History

- 6 years: diagnosed with Eisenmenger syndrome
- Treated with frequent phlebotomy
- Placed on Coumadin in adulthood
- Placed on the heart/lung transplant list 5 years (elsewhere)
- No birth control being used
Current Exam

- Significant cyanosis
- Conjunctival injection
- 2+ RV impulse, normal LV impulse
- 3/6 systolic crescendo-decrescendo murmur left upper sternal border
- No diastolic murmur
PWD velocity = 0.5 m/s
CWD velocity = 4.1 m/s
Non-imaging Doppler velocity $= 4.5 \text{ m/s}$
Does This Patient Have Eisenmenger’s Syndrome?

A. Yes
B. No
Does This Patient Have Eisenmenger’s Syndrome?

A. Yes
B. No
Non-imaging Doppler velocity = 4.5 m/s
Cath

- Tight PA band in appropriate location without distortion of the pulmonary valve
- Distal PA pressure 35/11
- Band gradient: 80 mmHg
- Pulmonary blood flow < 1 L/min/m2
- No residual coarctation
- No PDA
Outcome

- Successful PA debanding and VSD closure
- Transient post-op reperfusion lung injury
- Returned for 6 month follow-up: room air sat 95%. Normal 6 minute walk. RVSP: 51 mmHg
- Discontinued disability and began a new job
Teaching Points

- A VSD with a bidirectional shunt ≠ Eisenmenger syndrome – look for obstruction to RV outflow causing RV hypertension
- Patients with Eisenmenger VSD do not have loud systolic ejection murmurs
- Review cath reports carefully with your interventionalist – communication between the care team is essential