Cases in Adult Congenital Heart Disease

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➢ No Disclosures
“I Have Palpitations”

18 Year old Man

- Palpitations
- “abnormal” ecg and cxr
ECHO
Ebstein Anomaly
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

Significant Coarctation  Normal
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
“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 m/s
Does This Patient Have Eisenmenger’s Syndrome?

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

What Next?

A. Cath
B. MRI
C. Sildenafil
D. Bosentan
E. Flolan
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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