Congenital Heart Disease Cases
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No Disclosures
CASE 1

63 year old Woman

- Healthy throughout life except:
  - Rheumatic fever at age 7
  - Palpitations for several years
- 2 months ago noticed increasing shortness of breath
Recent Hospitalization

- Anasarca - responded to diuresis
- Atrial fibrillation – treated with rate control and anti-coagulation

Physical Examination

- Neck: JVP is moderately elevated with prominent V wave
- Heart:
  - 2+ RV; Normal LV impulse
  - Irreg RR; soft \( S_1 \); single \( S_2 \); \( S_3 \) present
  - III/VI systolic murmur
  - I/IV diastolic murmur at L sternal border
- Abdomen: Liver is pulsatile 4cm below costal margin
- Ext: Trace edema
Medications

- Aspirin 81 mg QD
- Furosemide 40 mg QD
- Lanoxin 250 mcg tablet QD
- Metoclopramide 10 mg TID
- Metoprolol Tartrate 50 mg BID
- Potassium Chloride 20 mEq QD
- Warfarin 3 mg QD

Exercise Stress Test

- Exercise Time: 3.8 minutes
- FAC: 48%
- Peak VO₂ = 13.4 mL/kg/min (59%)
ECG

Chest X-Ray
RVSP = 29 mmHg
Diagnosis?
A. Ebstein anomaly
B. Membranous VSD
C. Gerbode defect
D. Coronary artery fistula
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Echo Results

- Enlarged left coronary system
  - Circumflex to RV fistula
- Flail Tricuspid valve leaflet with severe TR
- Severe RV enlargement with mildly decreased function; RVSP = 29 mmHg
- Severe RA enlargement
- LV EF = 58%
Catheterization
What Next?

A. Catheter intervention  
B. Surgical consultation  
C. More testing
Surgical Intervention

- Repair of coronary artery fistula with 2 layer primary closure in the RV
- 31-mm Epic porcine tricuspid valve replacement
- PFO closure
- Radiofrequency maze procedure
- Uncomplicated hospital course
Case 2

26 Year old Woman

• Diagnosed with tetralogy of Fallot in the neonatal period
• Operative repair at age 2
• Sporadic follow up after age 12
• Presented with progressive decline in stamina
• Elsewhere underwent mitral valve replacement with a tissue prosthesis for “mitral valve prolapse” and regurgitation
• Presents 4 weeks after surgery with intractable pleural effusions, fatigue and high grade AV block
Echocardiogram
Diagnosis?

A. Constriction  
B. Severe tricuspid valve regurgitation  
C. Severe pulmonary valve regurgitation  
D. Pulmonary hypertension
Diagnosis?
A. Constriction
B. Severe tricuspid valve regurgitation
C. Severe pulmonary valve regurgitation
D. Pulmonary hypertension

ECHO Report
• Severe RV enlargement, moderate-severe decrease in function, RVSP 39 mmHg.
• Severe (free) pulmonary regurgitation
• LV EF 30% - 35%
• Abnormal hepatic vein Doppler related to junction rhythm
• Normal mitral tissue prosthesis

Patient medically optimized then referred for PVR
Post-Operative Hemodynamic Concerns

- Pulmonary valve regurgitation
- Residual/recurrent RVOT obstruction
- Residual VSD
- Tricuspid valve regurgitation
- Aortic root enlargement +/- aortic valve regurgitation

Consequences of Pulmonary Valve Regurgitation

- Exercise intolerance
- Right ventricular dilatation
- Right ventricular dysfunction
- Increased risk of ventricular tachycardia
- Increased risk of atrial arrhythmia
- Left ventricular dysfunction
Evolution of PVR Timing

Pulmonary Valve Not Important

Pulmonary valve should be replaced for right heart failure symptoms

Pulmonary valve should be replaced to prevent right ventricular dysfunction
Case 3

24 year old

- Denies complaints
- Echo obtained after murmur was heard on exam for assessment to be kidney donor
24 year Old: Wants to be kidney donor
Diagnosis?

A. Parachute mitral valve
B. Cleft mitral valve
C. Supravalvular mitral ring
D. Rheumatic mitral stenosis
Diagnosis?
A. Parachute mitral valve
B. Cleft mitral valve
C. Supravalvular mitral ring
D. Rheumatic mitral stenosis

Parachute Mitral Valve
• Abnormal compaction of ventricular trabecular myocardium and abnormal delamination of the trabecular ridge
• Unifocal attachment of the mitral valve cordae to a single/fused papillary muscle
• Papillary muscle usually centrally placed
• Often associated with other left heart abnormalities
• Mitral stenosis most common hemodynamic consequence
Mitral Ring

- Supramitral ring: fibrous membrane just above the mitral annulus. Does not adhere to the leaflets. Subvalvular apparatus normal
- Intramitral ring: thin membrane within the funnel created by the valve leaflets. Always combined with an abnormal subvalvular apparatus
- Results in stenosis

Supramitral Ring
Cor Triatriatum

- Failure of proper embryologic development of the common pulmonary vein
- Mitral valve structure usually normal
- Fibromuscular membrane *proximal* to the left atrial appendage that divides the atrium into two parts
Supramitral Ring

Cor Triatriatum
Mitral Arcade

- Caused by an arrest in the developmental stage of the mitral valve before attenuation and lengthening of the chordae
- Chords are thickened, very short, or absent
- Fibrous bridge may join the two papillary muscles
- Results in both stenosis and regurgitation