Commonly Encountered Congenital Heart Disease in the Adult

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No Disclosures
Atrial Septal Defect

- Second most common congenital defect recognized in adulthood
- Symptoms progressive
- Physical exam findings subtle

Atrial Septal Defects

- Secundum
- Primum
- Sinus Venosus
- Unroofed Coronary Sinus
Atrial Septal Defect

- **Echo Diagnosis and Evaluation**
  - Location of Defect
  - Right sided chamber size and function
  - Estimation of PA pressure
  - Tricuspid Regurgitation
  - Other Lesions
  - Repair Options

Secundum Atrial Septal Defect

Image Courtesy of Dr. Bill Edwards
Parasternal Short Axis Imaging
Subcostal Imaging
Primum Atrial Septal Defect

Image Courtesy of Dr. Bill Edwards

Apical 4 Chamber Imaging
Apical 4 Chamber Imaging

Valvular Abnormalities Associated with Primum ASD

Cleft Mitral Valve

Double Orifice Mitral Valve
LVOT Elongation, Narrowing, Anomalous Chords

Sinus Venosus Atrial Septal Defect

Image Courtesy of Dr. Bill Edwards
Subcostal Imaging

TEE Imaging
Anomalous Right Pulmonary Vein

Coronary Sinus Atrial Septal Defect
Coronary Sinus Atrial Septal Defect
Partial Anomalous Pulmonary Venous Return/Connection

Variants of Partial Anomalous Pulmonary Venous Connection

- Right pulmonary venous anomalies are most common
- Left pulmonary venous anomalies only comprise 4% of PAPVC
- Scimitar syndrome 3% of PAPVC
- Connections to the CS exceedingly rare
- Bilateral PAPVC occurs, but rare
PAPVC Physiology
- Left to right shunt
- Right chamber volume overload and dilatation
- Single anomalous veins – low risk of hemodynamic compromise
- Less than 50% shunt – rare to have symptoms in childhood

ECHO Evaluation of PAPVC
- Type of connection
- Associated anomalies
- Right chamber size
- Right ventricular function
- Pulmonary artery pressure
Vertical Vein
Right Pulmonary Vein to SVC
Scimitar Syndrome

MRA
Suprasternal Notch Coronal View ("Crab")

Ventricular Septal Defects
Echo Evaluation of VSDs

- Location
- Size
- Involvement of other structures
  - Left ventricular and left atrial size
- Estimated right ventricular systolic pressure
- Associated anomalies

Post- MI VSD
VSD Caveats

- The VSD jet may contaminate the TR signal
- Patients with high RV pressures may not have much color flow
Pulmonary Hypertension?

TR estimates RV pressure

Outflow Obstructions:
1. PS
2. Double chamber RV

ERRORS:
Contamination from VSD jet

Pulmonary Hypertension

MPA

Branch
PS

Pulmonary vascular disease

Pulmonary vein stenosis

LA

LA HTN
PAP = QP x PVR

PAP: Pulmonary artery pressure
QP: Pulmonary blood flow
PVR: Pulmonary vascular resistance

Double Chambered RV
Indications for Closure

- Large VSD (left heart enlargement, QP/QS > 1.5) without irreversible pulmonary vascular disease
- Aortic valve prolapse with progressive regurgitation
- RV outflow tract obstruction
- Recurrent endocarditis

Atrial Septal Defect vs. Ventricular Septal Defect

**ASD**
- Increased pulmonary blood flow
- Primarily volume load
- Low incidence of pulmonary hypertension in adulthood

**VSD**
- Increased pulmonary blood flow
- Primarily pressure load
- High incidence of pulmonary hypertension in adulthood
Patent Ductus Arteriosus

- Left Heart Enlargement
- Pulmonary hypertension common if the PDA is large – may not see a shunt on echo (equal pressures)
QP/QS

Ebstein Anomaly
Normal Delamination of the TV from the RV Myocardium

Failed Delamination results in …

- adherence of leaflets to underlying RV myocardium
- displacement of the anular hinge points
Failure of Delamination From the Myocardium

Spectrum with Infinite Variability

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

Apical Displacement: 20 mm
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
Coarctation of the Aorta

Imaging of Coarctation of the Aorta

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

Significant Coarctation

Normal

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
Congenitally Corrected Transposition

Image Courtesy of Dr. Bill Edwards

Complete Transposition (D-TGA)  Congenitally Corrected Transposition (L-TGA)

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Conus present in the “LVOT”

Left A-V valve displaced apically
Lesions Associated with ccTGA

- Ventricular Septal Defect (70%)
- Subpulmonary ventricular outflow tract obstruction (40%)
- Tricuspid valve dysplasia/Ebstein malformation (90%)
- Situs Inversus
- Dextrocardia
Sequelae of L-TGA

- Systemic (RV) failure
- Systemic AV valve regurgitation
- Complete heart block
- SVT
- Sudden cardiac death

Systemic AV Valve Regurgitation

- Surgical intervention is needed prior to significant decline in systolic function
Conclusion

• There are congenital heart defects that present for the first time in adulthood, and they are not all “simple” lesions
• Right heart enlargement: ASD, PAPVR, TR, PR
• Left heart enlargement: VSD, PDA, AI, MR