Commonly Encountered Congenital Heart Disease in Adults

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No Disclosures
A 24 year old woman presents with dyspnea on exertion. On echocardiogram is found to have right heart dilatation with normal estimated RV systolic pressure. You should assess for which shunt lesions?

A. Unroofed coronary sinus  
B. Ventricular septal defect  
C. Patent ductus arteriosus  
D. None of the above  
E. Both A and B
A 30 year old man is known to have an unrestrictive membranous ventricular septal defect. On echocardiogram you would expect to find which of the following?

A. Systolic velocity across the VSD > 4 m/s
B. Pulmonary valve cusp prolapse into the defect
C. Laminar color Doppler flow across the defect
D. Left ventricular dilatation
E. AV valves at the same level – no apical displacement of the tricuspid valve
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
Apical 4 Chamber Imaging
Apical 4 Chamber Imaging
Parasternal Short Axis Imaging
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

No tape cassette detected in VCR. Check VCR

HR = 79 bpm
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")
Patent Ductus Arteriosus

- Left Heart Enlargement
- Pulmonary hypertension common if the PDA is large – may not see a shunt on echo (equal pressures)
Ventricular Septal Defects
Ventricular Septal Anatomy

- Membranous
- Muscular
  - Inlet: Separates ventricular inflow
  - Trabecular
  - Outlet: Separates outflow tracts
Ventricular Septal Defects

- Membranous (80%)
- Muscular (trabecular septum)
- Inlet
- Outlet
  - Infundibular
  - Supracristal/Subarterial (5%)
- Post-MI
Echo Evaluation of VSDs

- Location
- Size
- Involvement of other structures
- *Left* ventricular and *left* atrial size
- Estimated right ventricular systolic pressure
- Associated anomalies
Location by Echocardiogram
Parasternal Long Axis

Trabecular
Outlet

RV AO LV LA
Parasternal Short Axis

Diagram showing the membranous, outlet, and trabecular regions of the heart.
Apical

Inlet

Membranous

Trabecular

Trabecular
Outlet/Infundibular Septal Defect
Supracristal VSD
Don’t Get Confused!
Perimembranous Defect
Trabecular (Muscular) Defect
Inlet VSD
VSD Size

- Small (restrictive): Defect size <1/3 aortic root; velocity > 4 m/s
- Moderate: Defect size ½ aortic root; velocity 3 m/s
- Large (non-restrictive): right and left ventricular systolic pressure near equal
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

Errors:
- Contamination from VSD jet

Outflow Obstructions:
1. PS
2. Double chamber RV
PAP = QP x PVR

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

- ASD and PAPVC = Right heart enlargement
- VSD and PDA = Left heart enlargement
- Patients with pulmonary HTN may not have significant shunt visible
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