Adult Congenital Heart Disease: What All Clinicians Should Know
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DISCLOSURES

- I have no disclosures relevant to today’s talk
Why should clinicians know/care about congenital heart disease?

Classification of Congenital Heart Disease

- Right versus Left-sided lesions
  - According to side of heart affected
- Cyanotic versus Acyanotic lesions
  - May be difficult to use these classifications as severity of disease may affect degree of cyanosis
- Simple/Moderate/Complex
Atrial Septal Defects (ASD)

- 5 Types
  - Secundum
  - Primum
  - Sinus venosus
  - Coronary sinus
  - Patent foramen ovale
- Right sided lesions—increased blood flow across defect results in right atrial and ventricular enlargement
Types of Repair

- Catheter Closure
  - Must have rim of tissue around defect for device to attach
- Surgical Closure
  - Sinus venosus ASDs
  - Primum ASDs
  - Large secundum ASDs with inadequate tissue ring
    - Relationship of aortic valve to defect
Septal Occluder Devices

Potential Complications

- Device migration
- Erosion
  - Aorta
  - Atrium
- Atrial arrhythmias
Ventricular Septal Defects (VSD)

- **Inlet**
  - Separates mitral and tricuspid valves
- **Muscular (trabecular)**
  - Attachments of tricuspid leaflets outward to apex and upward to the crista supraventricularis
- **Outlet**
  - Crista to pulmonic valve
  - Smooth walled structure
- **Membranous**
  - Fibrous portion of septum just below aortic valve
  - Small

Physiologic Effects of VSDs

- Shunt flow is dependent on size of defect and relative pulmonary and systemic vascular resistance
  - Blood flows towards lowest resistance
  - SVR>PVR $\rightarrow$ left-to-right shunting
  - PVR>SVR $\rightarrow$ right-to-left shunting $\rightarrow$ CYANOSIS
- Shunt flow is from left ventricle to right ventricular outflow tract $\rightarrow$ little effect on right atrial/ventricular size.
- With large defects $\rightarrow$ increased pulmonary blood flow $\rightarrow$ increased pulmonary vascular resistance $\rightarrow$ RVH
Estimating Pulmonary Artery Pressures

- Simplified Bernoulli Equation = 4 x Vm²
  - Assumes no intracardiac shunting
  - Right ventricular pressure
- In VSDs, the right ventricular pressure is the left ventricular pressure minus the gradient across the defect
  - Systemic systolic blood pressure-VSD gradient (4 x Vm²) where Vm is the velocity across the defect
- In other situations of intracardiac shunting, should only report the right ventricular pressure

Atrioventricular Canal Defects (Endocardial Cushion Defects)

- Spectrum of defects
  - Partial AV canal—primum ASD + cleft mitral valve
  - Complete AV canal defect with one common (5 leaflet) AV valve and both atrial and ventricular septal defects
- Atrioventricular valves are at same level
- Will always have “cleft” in mitral valve
Atrio-ventricular canal defect
Tetralogy of Fallot

- Ventricular septal defect
- Varying degrees of pulmonic stenosis
  - Valvar or subvalvar
  - Branch pulmonary stenosis
- Overriding aorta
- Right ventricular hypertrophy
- Pentalogy of Fallot: TOF + ASD

Long-term Complications

- Right ventricular enlargement/failure
- Ventricular arrhythmias
- Residual intracardiac shunting
- Branch pulmonary artery stenosis
- Aortic dilatation
Coarctation of the Aorta

- Due to narrowing in distal aortic arch/proximal descending aorta near insertion site of ductus.
- May be discrete lesion or involve a longer segment or diffuse arch hypoplasia.
- Several types of surgical repair
  - End-to-end anastomosis
  - Tube graft
  - Subclavian flap repair
  - Patch aortoplasty
  - Interposition ("jump") graft

Types of Repair

A. B. C. D. E.
Imaging

- **Echo**
  - Subcostal imaging of the descending aorta is imperative
  - Suprasternal notch views are also very important (provide best angle of interrogation of the coarctation)
  - Pulse and continuous wave doppler
- **MRI**
- **CT**
- **Angiography**
  - Done only if intervention being performed
Aortic Valve Disease

- Bicuspid aortic valves are the most common congenital heart disease occurring in ~2% of the population
- Frequently associated with an aortopathy regardless of degree of valvular disease
- Screening of first degree relatives
Complex Congenital Heart Disease

- Examples: Single ventricle (Fontan), Transposition of the great arteries (D- or L-), Truncus arteriosus
- Ideally should be performed at facility with congenital experience (sonographer and cardiologist)
- Single ventricle physiology
  - Glenn: Anastomosis of SVC to pulmonary artery
  - Fontan (various types): Anastomosis of IVC flow to pulmonary artery

Bidirectional Glenn
Fontan Operation

EXTRACARDIAC

LATERAL TUNNEL

D-Transposition of Great Arteries

KEY
SVC – Superior Vena Cava
IVC – Inferior Vena Cava
RA – Right Atrium
RV – Right Ventricle
MPA – Main Pulmonary Artery
Aoa – Aorta
LA – Left Atrium
LV – Left Ventricle
Mustard or Senning Operation

- "Atrial Switch"
  - Baffling systemic venous return to mitral valve → morphologic LV → PA
  - Pulmonary venous return then flows around baffle to tricuspid valve → morphologic RV → aorta
  - Mustard: Baffle created from pericardium.
  - Senning: Baffle created from atrial tissue.

“Arterial Switch”

- LeCompte maneuver
L-TGA (Congenitally Corrected or Ventricular Inversion)

Key:
- RA - Right Atrium
- RV - Right Ventricle
- PA - Pulmonary Artery
- LA - Left Atrium
- LV - Left Ventricle
- Ao - Aorta
- DAO - Descending Aorta

THANK YOU