Disclosures

• I have no relevant financial disclosures or conflicts of interest.
Outline of Today’s Talk

• Introduction of Simple Congenital Heart Disease
• Specific Cardiac Lesions:
  • ASD
  • PAPVR
  • VSD
  • AVSD
  • PDA
  • Coarctation of Aorta

Introduction

• Incidence – 0.5 to 0.8% of live births per year in the United States.

• Acyanotic ~ 70% (Simple shunts – ASD, VSD, PDA; Complex obstruction)
• Cyanotic ~30%

• Over 1 to 1.3 million adults with CHD in US; The population of adult CHD patients is growing at a rate of 5% per year.
Clinical Presentation

- Size and location of defect
- Left-to-right shunt (volume overload)
- Pulmonary hypertension
- Associated syndromes
Atrial Septal Defects - Types

- Patent foramen ovale (PFO).
- Ostium secundum atrial septal defect (ASD).
- Ostium primum ASD.
- Sinus venosus ASD.
- Unroofed Coronary sinus (CS) ASD.

Echo evaluation of ASD

- Location of the defect
- Right heart dilatation
- Estimation of PA pressure
- Tricuspid Regurgitation
- Other associated lesions
Secundum Atrial Septal Defect

1. Defect in the septum primum.
2. Most common type of ASD - 70% of ASDs are secundum defects
Secundum ASD – Parasternal short axis view

4 Chamber – off axis view
Adult patient with large ASD – 4 chamber view
Sinus Venosus Defect

- Two types
  - Superior (more common compared to Inferior SVASD) defect located superior and posterior near SVC
  - Inferior defect located posterior and inferior near IVC

- Associated with anomalous pulmonary vein return
  - Right upper pulmonary vein

- Best imaged by TEE; subcostal- bical view used in TTE.
Superior Sinus Venosus Defect

15-year-old with right heart dilatation
Superior Sinus Venosus Defect - PAPVR

Vertical Vein – Left sided pulmonary veins
Right sided PAPVR

Over-riding IVC to inter-atrial septum
ASD - Presentation

• Asymptomatic
• Increased RV impulse
• Fixed split S2 (65%)
• Systolic ejection murmur LUSB
• Diastolic rumble LLSB – relative increased flow across TV
• ECG – 1° AVB, RBBB, RVH

Atrial Septal Defect

2 year old  22 year old
Management

• Adequate Septal Rims – Cardiac Cath Lab – Device Closure

• Not amenable to Cath Closure – Surgical Closure

Ostium Primum Atrial Septal Defect

• Located in the most anterior and inferior aspects of the atrial septum.

• Deficient atrioventricular (AV) septation.
  • Associated with Trisomy 21.
  • Usually seen in association with AV septal defects with a cleft in the anterior leaflet of the mitral valve, although primum defects can occur in isolation.
  • Both AV valves (AVVs) appear at the same level.
  • The tricuspid valve (TV) will not be apically displaced.
Ventricular Septal Defects
Ventricular Septal Defects

Most common isolated congenital cardiac defect.
Most common defect – 25% of CHD
VSDs are frequently associated with more complex cardiac malformations such as:
  - Conotruncal malformations including
  - Double-outlet RV.
  - Truncus arteriosus.
  - Tetralogy of Fallot (TOF).
  - Interrupted aortic arch.
  - Complete AVC defects.
  - Coarctation of the aorta (Ao).

VSD – Clinical Features

• VSD shunt usually – left to right
• Timing and inset of symptoms – depend on the size of the VSD
• Large VSD-pulmonary edema- tachypnea- FTT
• Unoperated large defects – pulmonary vascular disease – Shunt reversal and cyanosis (Eisenmenger’s complex)
Types of Ventricular Septal Defects

- VSDs may overlap two or more classifications depending on the size and shape of the defect.
  - Perimembranous – 70%
  - Muscular – 20%
  - Inlet – AV Septal Defects 5%
  - Supracristal (subpulmonary) 5%

Common Types of VSD

Perimembranous

Muscular
Perimembranous VSD

Muscular VSD
Inlet VSD

Clinical Features

• Small VSD – asymptomatic.
• Moderate to large
  • Delayed growth
  • Decreased exercise tolerance
  • Increased pulmonary infections
  • CHF
  • Long standing – PHTN – Cyanosis
  • Supracristal VSD – complication – Aortic Insufficiency
VSD Confounding Factors

• VSD – tricuspid regurgitation contamination.
• High right sided pressures - reduced – left to right shunt.

VSD - Management

• Anti-congestive therapy – Diuretics
• Indications for Closure:
  • Surgical repair Qp/Qs >1.5
  • Aortic valve prolapse/progressive aortic insufficiency
  • RV outflow tract obstruction
• Catheter-based occlusion – mostly for muscular VSDs
30-year-old with multiple large VSD

30-year-old with multiple large VSD – 4 Ch view
Patent Ductus Arteriosus

- A patent ductus arteriosus is a persistent patency of a fetal connection (6th left branchial arch) between the descending aorta (6th left branchial arch) just distal to the origin of the left subclavian artery and PA.

- PDA occurs in 5-10% congenital heart defects.
Clinical Features

- Continuous murmur, bounding pulses, wide pulse pressure.
- Small PDA usually asymptomatic.
- Large PDA – causes CHF in infants.
- Tachypnea, tachycardia, bounding peripheral pulses.
- Exertional dyspnea in older children.
- Endarteritis

- Echo: LA, LV dilatation

PDA - high left parasternal view
Management:

• Medical: Indomethacin, Ibuprofen, Acetaminophen.

• Cath: PDA Coils, Amplatzer PDA device.

• Surgical ligation and division.
Atrioventricular Septal Defect
Atrioventricular Septal Defect

- Complete endocardial Cusson Defect occurs in 2% of Congenital Heart Defects.
- ECD- 70% have Down’s Syndrome.

Common AV Valve

A. Normal AV Valve
B. Partial ECD with Cleft in Left AV Valve
C. Complete ECD
Clinical Features

- Clinical Presentation varies based on the size of the VSD (inlet) and AV valve regurgitation.
- Heart Failure usually sets in in 1-2 months.
- Many patients with AV canal defect develop pulmonary hypertension.

Partial AV Canal
Atrioventricular Septal Defect

- AVSDs represent about 4% to 5% of congenital heart defects.

- Deficient atrioventricular (AV) septation.
  - Associated with Trisomy 21.
  - Usually seen a cleft in the anterior leaflet of the mitral valve, although primum defects can occur in isolation.
  - Both AV valves (AVVs) appear at the same level.
  - The tricuspid valve (TV) will not be apically displaced.

Types of AV Septal Defects

1. Complete AV Septal Defect
2. Partial AV Septal Defect
3. Transitional AV Septal Defect
Complete AV Canal

A complete AVSD has a common AV junction,
- a primum ASD,
- an IVSD, and
- a common AVV.

The primum ASD is anterior and inferior to the fossa ovalis, adjacent to the AVVs.

The AVV consists of five leaflets: superior and inferior bridging leaflets, a left mural leaflet, a right mural leaflet, and a right anterosuperior leaflet.

Partial AV Septal Defect

A partial AVSD usually has:
- a primum ASD and two separate AVVs
- cleft in the anterior leaflet of the left-AVV.

The cleft in the left-sided AVV usually results in some degree of regurgitation of the valve.
Primum Atrial Septal Defect

• Located in the most anterior and inferior aspects of the atrial septum.

Presentation in Adults with repaired AVSD

• Left atrioventricular valve regurgitation and stenosis
• Left ventricular outflow tract (LVOT) obstruction attributable to the abnormal shape of the LVOT
• Arrhythmias – late onset complete heart block.

Left atrioventricular valve regurgitation is the most common reason for later surgical reintervention.
Management

• Medical for CHF and Pulmonary Hypertension

• Surgical:
  • Usually performed at 2-4 months of age.
  • Single patch or 2 patch technique.

• Complications:
  • Residual VSD, AV Valve regurgitation or Stenosis.
  • Pulmonary Hypertension.

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Coarctation of the Aorta
Coarctation

**Definition:**
- Discrete narrowing or long segment stenosis
- Usually in descending aorta opposite ductus arteriosus

**Prevalence:**
- 8 to 10% of congenital heart disease
- 1.5:1 Male predominance

**Etiology:**
- Multifactorial: Genetic and environmental

**Clinical Features:**
- Infants: Poor feeding, dyspnea, poor weight gain, severe cases circulatory shock.
- Peripheral pulses may be weak and thready.
- Can develop CHF by 3 months of age.
- Upper extremity Hypertension
Coarctation of the Aorta

Contrast enhanced MRA
Coarctation

**Types:**
1) Preductal - Infantile Type
2) Juxtaductal - Most common type
3) Postductal - Adult Type
4) Long Segment or Tubular hypoplasia
   - Associated with complex anomalies
5) Pseudocoarctation
   - Non-obstructive kinking of the aorta
Coarctation

Associated Cardiac Anomalies:
• VSD - 44%
  • More common with isthmus hypoplasia
• Bicuspid Aortic Valve - 42%
  • More common with isolated juxtaductal coarctation
• Valvular Aortic Stenosis - 8%
• Subaortic Stenosis - 8%
• Mitral Valve abnormalities - 4%
• Patent Ductus Arteriosus

Aortic Arch 3D Recon
Coarctation

• **Clinical presentation**
  • Asymptomatic murmur
  • Hypertension
  • Congestive heart failure and shock

• Symptoms depend on severity of stenosis and associated cardiac defects (PDA, VSD, AS, etc..)
Coarctation-Treatment

- Untreated infants presenting with CHF in the first year of life
  - High mortality rate
- Operative Treatment:
  - Resection with end-to-end anastomosis
  - Prosthetic patch aortoplasty
  - Left subclavian flap aortoplasty
- Catheterization Treatment:
  - Percutaneous balloon angioplasty +/- stent

Coarctation

Resection with end-to-end anastomosis
Coarctation

Post-Operative Complications:

- Residual obstruction
  - Gradient less than 20 mm Hg is normal
  - More common if surgical repair < 3 yrs of age
- Aortic Aneurysm
  - Diameter greater than 1.5 times the descending aorta
- Aortic Dissection
- Hypertension
- Endocarditis
Questions?

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Thank you

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