Congenital Heart Disease: An Introduction

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Epidemiology

- CHD occurs in 8/1000 live births
- The most common birth defect
- About 1/4 of infants with CHD will require surgery in the 1st year of life

1 mil children
300K severe
1.4 mil ACHD

Categories

• I. Left to right shunts
• II. Left heart obstructive lesions
• III. Cyanotic lesions
• IV. Others
I. Left to Right Shunts

*Left to Right Shunting Lesions:*

- Ventricular Septal Defects
- Atrial Septal Defects
- Atrioventricular Septal Defects
- Patent Ductus Arteriosus
- Aorto-Pulmonary Window
Ventricular Septal Defects
VSD - Physiology
VSD - Presentation

Small vs Large Defects
Echo is diagnostic
VSD - Management
Atrial Septal Defects

- 2nd most common type of CHD
- classified by location in septum
Atrial Septal Defects

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Atrial Septal Defects

- 2nd most common type of CHD
- classified by location in septum
  - sinus venosus
  - anomalous return of R pulmonary veins
ASD - Physiology
ASD - Management

- elective closure before school age
- earlier for primum, sinus venosus
- cath vs. surgery for secundum ASD
II. Obstructive Lesions

*Left Heart Obstruction:*

- Mitral Stenosis
- Subaortic Stenosis
- Aortic Stenosis
- Coarctation of the Aorta
- Hypoplastic Left Heart Syndrome
Coarctation of the Aorta
Coarctation - Physiology
Coarctation - Presentation

- Discrepant pulse
- Cardiogenic Shock (infant)
- Hypertension (older child/adult)
- Murmur over the back
Coarctation - Diagnosis

Clinical: with your fingers and your ears

Confirm with the echo
Echo image of a coarct

AA = ascending aorta
TA = transverse aorta
* = isthmus, most common site of coarctation
Coarctation: Management

- Infant: PGE until surgery
  - end to end anastomosis

- Child/Adult:
  - balloon angioplasty
  - stent placement
  - surgery
III. Cyanotic Heart Disease

5 T’s of Cyanotic Heart Disease:

- Tetralogy of Fallot (TOF)
- Transposition of the Great Arteries (TGA)
- Tricuspid Atresia
- Truncus Arteriosus
- Total Anomalous Pulmonary Venous Return (TAPVR)
Tetralogy of Fallot (TOF)
TOF - Physiology

- Degree of cyanosis results from amount of pulmonary blood flow
- Murmur reflects pulmonary blood flow
  - no murmur = no flow
- Spectrum of disease, from pink to severe cyanosis and PGE dependent
TOF - Presentation

Clinical Manifestations:

• Murmur
• Progressive cyanosis
• Single S2, loud murmur
• Cyanotic “Tet” spell
TOF - Management

Spell treatment:
- increase pulmonary blood flow
- alter the ratio of PVR/SVR

Surgical
- Blalock - Taussig Shunt
- Rastelli Repair
- Complete repair
Surgical repair of tetralogy of Fallot: (A) preoperatively; (B) resection of the obstructive infundibular tissue; (C) patch closure of the ventricular septal defect; and, when enlargement of the annulus is necessary, an annular patch from the right ventricular outflow tract to the main pulmonary artery.
Transposition of the Great Arteries
TGA - Anatomy

- VentriculoArterial Discordance
- VSDs
- Coronary Artery Anomalies
- Dynamic LVOT obstruction
TGA - Physiology

- Parallel Circulations
- Shunting
  - Ductus Arteriosus
- Mixing
  - Atrial Septum
  - Ventricular Septum
TGA - Clinical Manifestations

- Cyanosis
- Tachypnea
- If no VSD, no murmur
- If not recognized: moribund
CXR: “egg on string”
TGA - Management

- PGE
- Balloon Atrial Septostomy
- Surgical: Arterial Switch Operation
Epidemiology

- CHD occurs in 8/1000 live births
- The most common birth defect
- >90% of patients with severe disease survive to adulthood

CHD Population Growth

Triumph: survival to adulthood is the
Challenges

ACHD Population
Do we have resources to care for this population?
Do we have the knowledge?
Do we have the quality metrics?

Individual Concerns
Heart failure
Arrhythmias
Cyanosis
Unforeseen complications
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