Congenital Heart Disease: An Introduction



Robert Elder, MD



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



Gilboa et al. Circ. 2016



- 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

VSD - CXR





VSD - Management



Atrial Septal Defects

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







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- 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

ΓA

*

AA

AA = ascending aorta TA = transverse aorta * = isthmus, most common site of coarctation +80.2

-80.2 cm/s

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



TOF - "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



Gilboa et al. Circ. 2016

CHD Population Growth



Marelli A et al. Lifetime Prevalence of Congenital Heart Disease in the General Population From 2000 to 2010 . Circ. 2019.

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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