Congenital Heart Defects

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Congenital Heart Defects

- Incidence:
  - 8-12 of 1000 live births (~ 1%)

- Heart Defects:
  - Acyanotic defects
  - Cyanotic defects
  - Obstructive defects
Normal Cardiac Anatomy & Physiology

- Four chambers and four valves
- Blue on right, pink on left
- Circulation in series
- PA/RV pressures = $\frac{1}{4}$ systemic pressure
- No mixing allowed!
Fetal Circulation
Acyanotic Heart Defects

**Volume Overload (left-to-right lesions)**

- Atrial Septal Defect (ASD)
- Ventricular Septal Defect (VSD)
- Patent Ductus Arteriosus (PDA)
- Atrioventricular Septal Defect (AVSD)
Pulmonary Vascular Changes

- Pulmonary vascular resistance abruptly decreases at birth, but takes another 4 to 8 weeks to reach adult level in humans.
- Pulmonary over-circulation will therefore gradually increase as pulmonary resistance decreases.
Atrial Septal Defect (ASD)

- Communication in the septal wall between the left and right atria
  - Sinus Venosus
  - Secundum
  - Primum
ASD.. Signs & Symptoms

* Usually asymptomatic
* Usually slender
ASD.. Treatment & Prognosis

- **Smaller defects**
  - Observation
- **Larger defects**
  - Medical management
  - Surgery vs. device closure – at 3-5 years of age

- Surgical results-excellent
- Follow up is infrequent
- May be d/c from follow up 3-5 yrs post-op
Amplatzer Septal Occluder Device
Ventricular Septal Defect (VSD)

- An Opening in the septal wall between the Left and right ventricles
  - Size
  - Location
    - Perimembranous
    - Muscular
    - Inlet
    - Outlet
VENTRICULAR SEPTAL DEFECTS
(Viewed from Right Ventricle)

1. Posterior A-V Canal Type
2. Perimembranous
3. Muscular
4. Apical Muscular
5. Multiple Anterior Muscular
6. Supracristal
7. Midcristal

Parietal Band of Crista Supraventricularis
Papillary Muscle of Conus
Septal Leaflet of Tricuspid Valve
Anterior Leaflet of Tricuspid Valve
VSD.. Signs & Symptoms

- May be asymptomatic
- Congestive Heart Failure
  - Tachypnea
  - Poor weight gain
  - Sweating
- Simple colds can lead to infections or pneumonia
VSD.. Treatment & Prognosis

- **Small defects**
  - Observation
- **Large defects**
  - Medical therapy
  - Surgery
    - Symptoms
    - Pulmonary hypertension
  - 12 months

- Surgical results-good
- Heart block, RBBB
- Cardiology follow up every 2-3 yrs
Patent Ductus Arteriosus (PDA)

- Communication between the pulmonary artery and aorta
PDA.. Signs & Symptoms

- Usually asymptomatic (unless premature infant)
- Large defect
  * May lead to Congestive Heart Failure or Pulmonary Hypertension
PDA. Treatment & Prognosis

- **Small Ductus**
  * Coil at > 1 yr
- **Large Ductus**
  * Surgery

- No residual heart disease
- D/C 1-2 yrs post op
- Coil follow up yearly
Atrioventricular Septal Defect (Endocardial Cushion Defect)

- Primum ASD
- Inlet VSD
- Common AV valve
- Down Syndrome

[Diagram of Atrioventricular Septal Defect]
AVSD.. Signs & Symptoms

- Failure to thrive
- Respiratory Infections
- Congestive Heart Failure
AVSD.. Treatment & Prognosis

- Medical Management
  * Congestive Heart Failure
- Surgical
  * Complete Repair at 6 months
- Heart block
- Residual pulmonary Hypertension
Cyanotic Heart Defects (The 5 Ts)

- **T**etralogy of Fallot (TOF)
- **T**ransposition of the Great Arteries (TGA)
- **T**runcus Arteriosus
- **T**otal Anomalous Pulmonary Venous Return (TAPVR)
- **T**ricuspid Atresia (and other single ventricle physiology)
Transposition of the Great Arteries (D-TGA)

- Aorta arises from the Right ventricle
- Pulmonary Artery arises from the left ventricle
- Communication between systemic and pulmonary circulation is crucial for survival
  
  * VSD, PDA, PFO
TGA.. Signs & Symptoms

- Cyanosis
- Tachypnea
TGA.. Treatment & Prognosis

- Medical (mixing)
  * Prostaglandin Infusion (PGE1)
  * Balloon Atrial Septostomy
- Surgical
  * Arterial Switch Procedure

- Surgical results
  * Coronary arteries
  * Supravalvar PS

- Cardiology follow up every 1-2 yrs
  * Cardiac Catheterization 1 yr post op
Balloon Atrial Septostomy (Rashkind Procedure)

- Balloon-tipped catheter is inserted through the atrial septal defect (ASD).
- Once the balloon is inflated, the catheter is pulled back through to widen the ASD.
- An opening in the septum allows oxygen rich and oxygen poor blood to mix to improve circulation.

Transposition of Great Arteries

- Balloon catheter
- Atrial septum
Tetralogy of Fallot (TOF)

- VSD
- Pulmonary Stenosis
- Aortic Override
- Right Ventricular Hypertrophy
TOF.. Signs & Symptoms

- Cyanosis
- Clubbing
- Exertional dyspnea
- Hypercyanotic Spells (Tet Spell)
- Squatting

Children with Tetralogy of Fallot exhibit bluish skin during episodes of crying or feeding.

“Tet spell”
TOF.. Treatment & Prognosis

- Medical Management of Tet Spells
  - * Acute
    * Knee chest position, oxygen, morphine
  - * Prevention
    * Propranolol
- Surgical
  * Complete repair 3-6 mo
- Ventricular arrhythmias/
  * Pulmonary insufficiency
- Cardiology follow up every 1-2 yrs
Total Anomalous Pulmonary Venous Return

- Pulmonary veins drain into the RA
  * Supracardiac
  * Cardiac
  * Infracardiac
  * Mixed
Pulmonary Atresia/Intact Ventricular Septum

- Atretic pulmonary valve
- Small Right Ventricle
Signs & Symptoms

- Severe Cyanosis
- Tachypnea
Treatment & Prognosis

- Medical
  - PGE 1 Infusion
  - Cardiac Catheterization (BAS)
- Surgical
  - Staged Repairs
    - Bi-directional Glenn
    - FONTAN/TCPC
- Followed closely
Tricuspid Atresia

- Atretic Tricuspid valve
- RV & PA are usually hypoplastic
  - * Associated defects
    - * ASD
    - * VSD
    - * PDA
    - * PS
Signs & Symptoms

- Severe Cyanosis
- Poor Feeding
- Tachypnea
Treatment & Prognosis

- Increases PBF
- Medical
  * PGE 1 Infusion
  * BAS
- Surgical
  * BT Shunt
  * Glenn
- Fontan/TCPC
- Cardiac Catheterization
- Followed yearly
  * Atrial Arrhythmia’s
  * Liver Congestion
  * Protein-losing enteropathy
Truncus Arteriosus

- A single arterial blood vessel leaves the heart and feeds both the pulmonary and systemic circulations
- Often present with mild cyanosis and loud murmur
- Saturations first climb, then fall as severe CHF develops
- High risk of sudden death if not treated
- DiGeorge Association
Obstructive Heart Defects
(Pressure Overload Defects)

- Pulmonary Stenosis
- Aortic Stenosis
- Coarctation of the Aorta
- Interrupted Aortic Arch
- Hypoplastic Left Heart Syndrome (HLHS)
Pulmonary Stenosis (PS)

- Thick, domed valve
- Post stenotic dilatation
- Thickened Right Ventricle
PS.. Signs & Symptoms

- **Mild**
  - *Asymptomatic*

- **Moderate**
  - *Exertional dyspnea*
  - *Fatigue*

- **Severe**
  - *Congestive Heart Failure*

- **Newborn Critical Stenosis**
  - *Cyanotic, tachypnea*
PS.. Treatment & Prognosis

- Balloon Valvuloplasty
- Surgical Valvotomy
- Follow up infrequently
  * Echocardiogram
Aortic Stenosis (AS)

- Thickened Aortic Valve
- May be bicuspid
- Post Stenotic Dilatation
- Thick Left Ventricle
AS.. Signs & Symptoms

- Mild to Moderate
  * Asymptomatic
- Severe
  * Exertional chest pain
  * Syncope

**Newborn Critical AS**

* Congestive Heart Failure/low cardiac output
AS.. Treatment & Prognosis

- Critically Ill
  - Aggressive CHF therapy
    - PGE 1, Oxygen,
    - Inotropes, Diuretics
  - Balloon Valvuloplasty
- Surgery
  - Valvotomy
  - Valve Replacement
  - Ross Procedure
- Cardiology follow up is frequent
Coarctation of the Aorta (COA)

- Narrowing in the descending Aorta
COA.. Sings, Symptoms, Treatment, & Prognosis

- Symptomatic Infants
  * Medical CHF management
    * Diuretics, PGE 1
    * Inotropes, O2
    * Surgery
- Asymptomatic Children (HTN)
- Cardiology follow up is yearly
- Recurrence - balloon angioplasty
Hypoplastic Left Heart Syndrome (HLHS)

- Most Serious of all defects
- Staged surgical repair
  * Significant morbidity/mortality
THANK YOU