Congenital Heart Disease

- Cyanotic
  - Diagnosing cyanosis
  - Pulse Oximetry screening
  - Transitional Circulation
  - Specific lesions
- Acyanotic
  - Shunt lesion
  - Obstructive lesions
  - Specific lesions
Diagnosing Cyanosis

- Central vs Acrocyanosis
  - Central = Trunk and lips
  - Acrocyanosis= Hands feet and skin of the face (perioral)
Diagnosing Cyanosis

- Hyperoxia test
  - Used to differentiate cyanotic heart disease from other causes of cyanosis
  - Obtain ABG to measure pO2
  - Place in 100% oxygen for >10 minutes
  - Measure pO2 (not saturation) on oxygen
  - pO2 <50 or unchanged - cardiac likely
  - pO2 50-150 - cardiac or pulmonary possible
  - pO2 >150 - cardiac etiology unlikely
Diagnosing Cyanosis

- Pulse oximetry screening

[Diagram showing decision tree for diagnosing cyanosis]
Transitional Circulation

- PDA closes (hours to days)
- Ductus venosus closes (minutes to days)
- PFO closes (minutes to months)
- ↓PVR- slowly drops to normal by 2 months.
A full term male newborn has failed initial pulse oximetry screening due to a saturation of 88%. The baby is in no distress and has a normal physical exam except for a loud second heart sound and a weight that is LGA. Echocardiography is not available. The pO2 is 36 mmHg at baseline and 64 mmHg on 100% inspired FiO2, after 4 hours, he is found to have a saturation of 90% in his left foot and 63% in his left arm. Which of the following is indicated prior to transfer?

1. PGE1 infusion
2. Surfactant
3. Intubation and ventilation with 100% FiO2
4. Phenylephrine
Cyanotic CHD - Specific Lesions

- The five “T”s of Cyanotic Congenital heart disease
  - Transposition of the Great Vessels
  - Tetralogy of Fallot
  - Truncus Arteriosus
  - Tricuspid atresia
  - Total Anomalous Pulmonary Venous return
  - Pulmonary atresia
  - DORV, HLHS..... Other single ventricle
Others

- Pulmonic stenosis/atresia
- Ebstein’s anomaly (SEVERE)
- Other Single ventricles
  - Double inlet left ventricle
  - Hypoplastic left heart syndrom
  - Double outlet right ventricle
    - DOVR can act as anything
    - TOF-like DORV
    - Single Ventricle – DORV
    - VSD - DORV
Transposition of the Great Arteries

- Aorta arises from pulmonary ventricle, pulmonary artery from systemic ventricle
- “blue baby” (cyanosis)
- 5% of CHD
- Palliation by Balloon Atrial Septostomy
- NEED TO KNOW: SATs low above, high below, Balloon septostomy to improve UE SATs, big baby with normal to incr pulmonary blood flow
Transposition of the Great Vessels

- **Presentation**
  - Typically present due to cyanosis
  - Loud second heart sound (Anterior Aorta closing)
  - Murmur only if associated lesions (PS, VSD)

- Most common cyanotic lesion to present as a newborn (first 24 hrs)
- Large first born male
- Incidence of 0.3/1000 live births
- Oxygenation is dependent on “mixing” of blood via PFO and PDA
Transposition of the Great Vessels

- Chest Film
  - “Egg on a string”
  - Increased pulmonary blood flow
- ECG
  - Normal
Transposition of the Great Vessels

- Management
  - Improve mixing
    - PGE₁
    - Balloon Septostomy
  - Surgical Correction

- Prognosis
  - Freedom from re-operation after ASO at 15 years is 82%
  - Most common sequelae are pulmonary stenosis and aortic insufficiency.
  - Too early to determine incidence of late arrhythmia.
  - Coronary issues are seen in up to 8% of patients
Balloon Atrial Septostomy in TGA
A 16 month old id brought in by his parents with a complaint of pallor and lethargy. They give a history of a congenital heart problem that was supposed to have surgery, but it has not happened yet. On examination you note a cyanotic, lethargic and anxious toddler with a pulse oximeter reading of 38%. Heart rate is 154 bpm, BP is 65/30 mmHg and respirations are 36 per minute. CXR is shown below. All of the following are indicated EXCEPT:

1. IV or IM morphine.
2. IV Propranolol
3. IV Epinephrine
4. Oxygen as tolerated
Tetralogy of Fallot
Tetralogy of Fallot

- Most Common cyanotic heart disease
- Incidence of 0.4/1000 live births
- NEED to KNOW: spells, boot shaped heart, right to left shunt at ventricular level
- Four components
  - Ventricular septal defect
  - Pulmonic stenosis
  - Over-riding aorta
  - Right ventricular hypertrophy
Tetralogy of Fallot

- Presentation depends on degree of pulmonary stenosis and timing of ductal closure.
  - Typically present with a murmur or cyanosis in first two weeks of life.
  - Most commonly diagnosed pre-natally
  - Variable cyanosis
Tetralogy of Fallot

- Chest Film
  - “Boot Shaped heart”
  - Decreased pulmonary blood flow
- ECG
  - RAD and RVH
Tetralogy of Fallot

- Management
  - May need PGE1 and surgical intervention as a newborn
  - if inadequate pulmonary blood flow
  - Primary repair vs. aorto-pulmonary shunt is controversial
  - NOT FOR BOARDS: Cath lab palliations: PDA stent, pulmonary valve balloon valvuloplasty or even stenting of the RVOT
Tetralogy of Fallot

- Prognosis
  - Incidence of re-intervention is high
  - Pulmonary insufficiency
  - Aortic insufficiency
  - Late arrhythmia
Tetralogy of Fallot

- Prognosis
  - Incidence of re-intervention is high
  - Pulmonary insufficiency
  - Aortic insufficiency
  - Late arrhythmia
Hypercyanotic or “tet” spells.

- Pathology
  - Something causes increased heart rate and hypercontractile state of RVOT
  - This leads to decreased oxygen delivery, acidosis and worsening hypercontractile state.
Acyanotic baseline- Hb normal

Agitation/↑RVOT obstruction ↓SVR

Acute ↓ pulmonary flow

↓Oxygen delivery

Metabolic acidosis

↑PVR  ↓SVR

↓RVOT obstruction
Morphine
Propranolol

Treatment

↑SVR
Knee chest
Phenylephrine

↑Oxygen delivery
Volume
PRBC
Oxygen
Bicarbonate
Hypercyanotic or “tet” spells.

- **Treatment**
  - Comfort
  - Oxygen (if possible, better not to upset them)
  - IV Fluids
  - IV beta blockers
  - Best IV starter, consider IM sedation first.
  - Beta blockers once IV is in.
- **Sedation**
  - Narcotics, typically morphine
  - Ketamine
  - Intubation/Anesthesia
- **Maneuvers to increase SVR**
  - Knee to chest
  - Vasoconstrictors like phenylephrine
  - Not inotropes
- **Surgery**
Tetralogy of Fallot

- Associations
  - DiGeorge Syndrome
  - Down Syndrome
Truncus Arteriosus
Truncus Arteriosus

- Ventricular septal defect
- Aorta and pulmonary artery arise from common trunk
- “Truncal” valve often abnormal
  - Stenotic +/- regurgitant
- Incidence of 0.1/1000 live births
- **NEED to KNOW:** mixes at the ventricular outflow, can have higher SATs and large heart with some failure – **NOT REALLY THAT CYANOTIC**
Truncus Arteriosus

- Presentation
  - 2 weeks-2 months when PVR drops
    - Congestive heart failure
    - Murmur: systolic @ LUSB (Often present at birth)
    - Single S2
  - Chest film
    - Increased pulmonary flow, cardiomegaly
Truncus Arteriosus

- **Treatment:** Surgical repair – includes VSD closure with LV to the aorta, detach the pulmonary arteries and an RV to PA conduit
- **Associations:** DiGeorge syndrome
Tricuspid Atresia
Tricuspid Atresia

- **NEED to KNOW:** single ventricle pathway with a characteristic ECG (unlikely to test in detail)

- **Presentation:**
  - Depends on size of VSD (and presence of TGA)
  - Cyanosis: if VSD small
  - CHF: if VSD large
  - Typically will have systolic murmur
Tricuspid Atresia

- Chest Film
  - Pulmonary blood flow determined by size of VSD
  - Large VSD will have cardiomegaly and increased pulmonary blood flow
- ECG
  - LAD, LVH
  - Northeast AXIS
- Treatment
  - Staged palliation to Fontan
Total Anomalous Pulmonary Venous Return- Infracardiac
Total Anomalous Pulmonary Venous Return- Infracardiac

- Incidence of 0.1/1000 live births – KNOW: very sick soon after birth with venous congestion
- Presentation:
  - Cyanosis (profound)
  - Respiratory distress
  - PE: No murmur
- Chest Film
  - Normal heart size
  - Severe pulmonary edema
- Treatment
  - Emergent surgery
Total Anomalous Pulmonary Venous Return - Chest films

Infracardiac with obstruction

Supracardiac, without obstruction
Ebstein’s Anomaly of the TV
Ebstein’s Anomaly of the TV

- Apical displacement of hinge point of tricuspid valve leaflets
- Incidence of 0.1/1000 live births
- Presentation - **KNOW:** HUGE hearts with cyanosis, SVT

**Cyanosis**
- Fixed split S₂, systolic murmur
- ECG: RAE, WPW (Wolf Parkinson White), RBBB
- CXR: Wall to wall heart

**Associations**
- ASD
- WPW
- Maternal lithium
Ebstein’s Anomaly of the TV
Pulmonic Stenosis
(presenting with cyanosis, AKA Critical PS)

- Incidence of 0.7/1000 live births
- Presentation
  - Cyanosis if stenosis is severe
  - Opening click and systolic murmur
  - ECG: RVH
  - CXR: Decreased pulmonary blood flow
- Treatment!
  - PGE1 if cyanotic in newborn period
  - Balloon valvuloplasty
  - Surgical valvotomy
Pulmonic Stenosis
(presenting with cyanosis, AKA Critical PS)

NEED to KNOW: balloon valvuloplasty is a good palliation, obvious murmur
Acyanotic Heart Disease

- Left to right shunt lesions
- Physiology
  - Obstruction = congestion, cardiac hypertrophy and CHF
  - L->R shunt – inefficiency and incr pulmn blood flow
  - Regurgitation = volume load on the hear and CHF
- Obstructive lesions without shunt
  - Valve stenosis or leakage
  - Coarctation
  - Pulmonary artery obstruction
Patent Ductus Arteriosus

- Incidence of 0.8/1000 live births, excluding premature babies
- **NEED to KNOW:** big heart especially in small premies from huge left to right shunt, murmur is **CONTINUOUS!!**
- Common in prematurity
- **Treatment**
  - Indomethacin
    - Renal toxicity
    - Platelet dysfunction
  - Catheter Intervention
  - Surgery
Patent Ductus Arteriosus
Ventricular Septal Defect

- Incidence of 3.6/1000 live births (Most Common CHD after BAV)
- Presentation depends on size of VSD and drop in PVR
  - Small VSD presents early with a murmur
  - Large VSD presents later with signs of congestive heart failure
Ventricular Septal Defect

- **NEED to KNOW:** holosystolic murmur, pitch = size, CHF
- **Physical Exam**
  - Murmur along left sternal border
  - Harsh, holosystolic – pitch = size
  - PMI may be displaced
  - Precordium may be hyperdynamic
  - May have diastolic rumble
  - May have signs of congestive heart failure
Ventricular Septal Defect

- **Treatment**
  - Anticongestive medications
    - Furosemide
    - Diuril
    - Aldactone
    - Angiotensin Converting Enzyme inhibitor
  - Surgery
  - Catheter device closure
Ventricular Septal Defect

- **Treatment**
  - Anticongestive medications
    - Furosemide
    - Diuril
    - Aldactone
    - ACEi
  - Surgery
  - Catheter device closure
You referred a 2 month old male for cardiac evaluation of tachypnea and a murmur. He was diagnosed with a large ventricular septal defect and started on diuretics and an angiotensin converting enzyme inhibitor. He returns now at 3 months of age. The parents state he was scheduled for surgery, but they canceled it because he is asymptomatic. They ask why surgery is needed if the medicines are working. Which of the following is the most appropriate answer to their question?

1. It is safe to wait as long as the medicines are working.
2. They can defer surgery until age two, to see if the defect closes spontaneously.
3. The medicines are not approved for use in children and should be stopped.
4. Large, unoperated VSDs have a risk of developing pulmonary hypertension if not closed by one year of age.
Atrial Septal Defect

- Incidence of 0.9/1000 live births
- Generally asymptomatic but can have poor growth and increased number of infections
- Large right heart and increase pulmonary blood flow
- Typically diagnosed in first 5 years of age due to a murmur
- Treatment
  - Catheter or surgical closure.
LA angiogram shows opening
Atrial Septal Defect

- **NEED to KNOW**: fixed split $S_2$, physiologic murmur, older kids, closed by a device and rarely surgery

- Natural History
  - Risk of atrial arrhythmias
  - Sick sinus syndrome
  - Pulmonary hypertension
  - 50% mortality by age 37 yrs
  - Risk of sick sinus syndrome and atrial arrhythmias is not eliminated by closure.
ASD After Device Closure
A 10 yo boy presents with a complaint of frequent headaches. On exam you detect a long, crescendo-decrescendo murmur at the right upper sternal border, heart also between the scapulae. Blood pressure in the right arm is 180/80 and 60/40 in the right leg with a diminished femoral pulse. Which of the following is indicated to treat the blood pressure?

1. IV nitroprusside
2. Surgery to relieve supraaortic stenosis
3. Stenting of an obstruction
4. ACEi
Obstructive lesions: Coarctation

- Incidence of 0.4/1000 live births
- Presentation
  - Weak femoral pulses
  - Hypertension
  - Newborns can present with shock in first few weeks of life
  - Remember PGE₁
Coarctation

- Physical Exam
  - Hypertension, with RUE BP > LE BP
  - Murmur long crescendo-decrescendo murmur at ULSB radiating to mid scapular area (back)
- ECG
  - RVH in neonates
  - LVH in older children
- Chest Film
  - RV enlargement and pulmonary edema in newborns
  - 3 sign with enlarged ascending aorta and rib notching in older children
Coarctation

- **Treatment**
  - In newborns PGE1 may relieve the obstruction and restore lower body circulation.
  - Newborns may also need volume and vasoactive medications
  - *Surgery or Stenting in older children*
Coarctation

- NEED to KNOW: HTN, femoral pulses!!
- Treatment
  - Beta-blockers can be used to control hypertension in older children prior to definitive intervention!
    - Dosing should be titrated to maintain adequate lower body perfusion.
- Definitive treatment
  - Surgical resection and repair
  - Balloon angioplasty with stent placement
- Important Associations
  - Turner’s syndrome
  - Bicuspid aortic valve
Aortic Stenosis

- Incidence of 0.4/1000 live births
  - Presentation
  - Typically presents with a murmur
  - Severe aortic stenosis can present with shock or Cyanosis from RV dependent circulation
- Natural History
  - Progresses over time
  - Known cause of sudden death in young persons
- Treatment
  - Balloon valvuloplasty
  - Surgical valvotomy
  - Valve replacement
Aortic Valvuloplasty
Bicuspid Aortic Valve without stenosis

- **NEED to KNOW:** aortic stenosis can be more significant with exertion, bicuspid valves have clicks and coarct
- Incidence of 13.5/1000 live births
  - Most common congenital heart anomaly
- Presentation
  - Typically presents with a murmur, beyond toddler years.
  - Will often have an associated opening click or split first heart sound.
Bicuspid Aortic Valve

- **Natural History!**
  - Progresses over time (Both stenosis and insufficiency)
  - Annual rate of intervention 0.004 (4%/Decade)

- **Treatment**
  - Blood pressure lowering medication when AI present
  - Surgical valve replacement.
Cardiogenic Shock
(covered in critical care also)

- Inadequate delivery of blood, oxygen and nutrition despite adequate filling pressures

Presentations Vary by Age

- Neonatal
  - Birth to 2 months
- Infancy
  - One month to one year
- Childhood
  - One year to 12 years
- Adolescence
  - 13-51 years
Cardiogenic Shock

Causes

• Three categories of causes
  • Anatomic
    • Obstructive
    • Volume overload (Shunt, valve regurgitation)
    • Cyanotic
  • Functional (Myopathy)
  • Electrical (Arrhythmia)
    • Brady or tachy
Cardiogenic Shock
Presentation

- Neonatal and infant are similar
  - Tachycardia
  - Tachypnea
    - Fixed rate
    - Associated with signs of distress
      - Flaring
      - Retractions
  - Hepatomagaly
  - Poor feeding
  - Emesis
Cardiogenic Shock
Presentation

- Childhood and adolescence are similar
  - Tachycardia
  - Effort intolerance
  - Emesis and or abdominal pain
    - Often confused with gastroenteritis with dehydration
    - Tachycardia is typically out of proportion to history/ signs of dehydration
    - Tachycardia does not respond appropriately to fluid bolus
Cardiogenic Shock Treatment

- Start with the basics
  - Airway, Breathing, Circulation
  - Volume, volume, volume.
  - Remember PGE₁ in Neonates
    - Ductal dependent obstructive lesions are the most common cause of shock at 7 days of age.
Cardiogenic Shock
Treatment

- Identify and treat cause
  - Potential diagnostic tests should be tailored to initial findings, and availability.
  - ECG
    - Tachycardia or bradycardia
    - Findings suggestive of myocarditis/ pericarditis.
  - Chest Film
    - Cardiomegaly
    - Pulmonary edema
  - Echocardiogram
    - Structure and function.
A 16 yo male presents to the ED after collapsing while playing soccer. He required CPR and was cardioverted with an ICD at the field. He is awake and alert with normal vital signs. An ECHOcardiogram is most likely to show:

1. Severe pulmonic stenosis with an intact ventricular septum
2. Asymmetric hypertrophy of the ventricular septum
3. A right coronary artery arising from the left coronary cusp
4. ECHO will be normal as this is more likely to be something picked up with an ECG