Cardiac Assessment of the Neonate

Shabib Alhadheri, MD
Congenital Heart Disease

- Incidence:
  - 8-12 of 1000 live births (~ 1%)
  - CHD is responsible for 3-5% of deaths during the first week of life and up to 33% of deaths during neonatal period
  - 50% of children with CHD present before 12 months of age
  - About one-third of discharged infants from regular nurseries turn to have significant CHD

- Heart Defects
  - Acyanotic Lesions
  - Cyanotic Lesions
  - Obstructive Lesions
Development of Fetal Heart

A
- atrium
- ventricle
- sinus venosus
- conus cordis

B
- atrium
- ventricle
- sinus venosus
- conus cordis

C
- atrium
- truncus arteriosus
- ventricle
Normal Anatomy & Physiology

- Atria collect blood
- Ventricles pump blood
- Blue on right, pink on left
- Pulmonary/RV pressure = $\frac{1}{4}$ systemic pressure
- Circulation in series
- No mixing allowed!
Fetal Anatomy & Physiology

- Brain and heart get highest oxygenated blood (from UV)
- Lower body gets less oxygenated blood (from RV)
- Pulmonary = systemic pressure
- Lowest flow at aortic isthmus
- Both lungs receive only 10% of CO and have the highest vascular resistance
- Since the blood is oxygenated in the placenta, the O2 sat in the IVC (70%) is higher than that in the SVC (40%)
- The highest pO2 is found in the umbilical vein (32 mm Hg)
Waste from Fetus

Placenta

Food and Oxygen from Mother

Umbilical Cord

Umbilical Vein

Umbilical Arteries

Ductus Arteriosus

Aorta

Foramen Ovale

Lung

Pulmonary Artery

Ductus Venosus

Liver

Left Kidney

Oxygen-rich Blood

Oxygen-poor Blood

Mixed Blood
Fetal Circulation

- Parallel circulations
- Mixing of venous returns
- High resistance of the pulmonary circulation
- Low resistance of the placental circulation
- Presence of shunts
- Right ventricular dominance
- Non-compliant behavior of ventricular filling in utero
- Limited range of HR over which CO can be maintained
Transitional Circulation

- Cord is clamped, baby cries
  - Systemic vascular resistance (SVR) jumps
  - LV end diastolic pressure (LVEDP) increases
  - Lungs expand
  - Pulmonary capillaries see high pO2
  - Pulmonary vascular resistance (PVR) falls
  - Ductus arteriosus constricts
Pulmonary Vascular Resistance

- PVR 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.
Examination Challenge

- The constant changes of the hemodynamics makes cardiac assessment of the neonate a challenge
  - Changes from fetal circulation
  - Changes ductal flow
  - Decrease pulmonary vascular resistance
  - Increase systemic vascular resistance
High Index of Suspicion

- Suspicious family and perinatal history
- How does the baby look?
  - Color
  - Respiratory status
  - Dysmorphic features
  - O2 Saturations (pre/post-ductal)
  - Blood pressure (BP) from different extremities
  - Feeding difficulties
- On exam:
  - Heart murmur (in the delivery room never good)
  - Irregular rhythm
  - Diminished femoral pulses
  - Peripheral perfusion
Perinatal History

- **Maternal age:** Trisomy 21

- **Congenital Infections:**
  - Rubella
  - HIV
  - Coxsackie B virus, CMV, HSV

- **Maternal medical conditions:**
  - IDM (risk is 3-4x)
  - Maternal SLE
  - Maternal PKU

PDA, pathologic PPS infantile cardiomyopathy
infantile myocarditis

PDA, TGA, Hypertrophic cardiomyopathy, VSD
Congenital complete heart block

TOF, VSD, CoA
Perinatal History

- **Teratogens:**
  - Amphetamine  VSD, PDA, ASD, TGA
  - Valproic acid  ASD, VSD, AS, CoA, PA/IVS
  - Hydantoin  PS, AS, CoA, PDA
  - Alcohol  ASD, VSD
  - Lithium  Ebstein’s anomaly
  - Retinoic acid  Conotruncal abnormalities (TOF, TA)
  - Oral contraceptives  VSD, TGA, TOF
  - Warfarin  PDA, PPS

- **Mode of delivery and ease of transition to extruterine life:**
  Prematurity, Apgar score, birth weight (SGA due to intrauterine infections “e.g. Rubella”, LGA as in IDM) etc.
Family History

- Congenital heart disease
  - No family history: 1%
  - Sibling with CHD: 5%
  - Two siblings/parent with CHD: 5-10%
- Premature unexpected death
- Arrhythmias: Long QT syndrome
- Cardiomyopathy
- Genetic conditions/Syndromes: Trisomy 21, Marfan syndrome, Noonan syndrome
Auscultation.. Cardiac Cycle

- **S1** = Mitral and Tricuspid valve closure (start of systole)
- **S2** = Aortic and pulmonary valve closure (end of systole)
  - A2 closes before P2
  - P2 moves with respiration
  - S2 splits with inspiration, closes with expiration
- **S3, S4** (gallops)
  
  S3: sudden deceleration of flow during rapid ventricular filling, benign
  S4: rapid ventricular filling during atrial contraction, heard in CHF & CM
Cardiac Auscultation of Neonates

- S1 is loud at birth and decreases in intensity during the first 48 hours of life
- S1 may be split
- A systolic ejection click along the LSB may be present
- S2 is single at birth due to elevated PVR, which splits by 48 hours after birth
- One third to three fourth of neonates have been described to have a murmur
In the Newborn Nursery

*Innocent Murmurs to Hypoplast*

Innocent murmurs
Patent ductus arteriosus
Pulmonic stenosis (mild to critical)
Aortic stenosis (mild to critical)
Small VSDs
*(NOT LARGE VSDs)*
*NOT ASD*
Nursery Screening

- Prenatal concerns
- Dysmorphic features
- Physical examination
  (don't forget LL pulses)
- Four extremities BPs
- Pulse oximetry (pre & postductal)
  - EKG
  - CXR
First Week of Life

Ductal-dependent lesions

HLHS/HRHS
Critical aortic stenosis
Severe coarctation of the aorta
Interrupted aortic arch
Critical pulmonic stenosis
Where to Listen?

Don't forget:
Both axillae, back, over the head and liver
Cardiac Exam - Auscultation

- Do you hear both S1 and S2?
- Are there any additional heart sounds?
  - Gallop (S3 or S4)
  - Click
  - Pericardial rub
  - Murmur
- Does S2 split normally?
Classification of Heart Murmurs

Systolic Ejection

Holosystolic (Regurgitant)

Early diastolic

Mid diastolic (Rumble)

Continuous
Pathologic Murmurs

- Aortic Valve Stenosis
- Aortic Insufficiency
- Patent Ductus Arteriosus
- Ventricular Septal Defect
- Pericarditis
- Mitral valve prolapse
Don’t Forget the Pulses

- **Normal**: Equal full upper/lower extremity pulses with no delay
- **Coarctation**: Diminished and delayed femoral pulses
- **Aortic Stenosis**: Diminished pulses - decreased stroke volume
- **Bounding pulses**: Diastolic runoff as in moderate-severe AI, PDA or BT shunt
Down Syndrome 
(Trisomy 21)

- First described in 1866, and the first cytogenetic confirmation was reported in 1959
- Incidence: ~ 1 in 660 live births
- Non-disjunction (94%), mosaicism (3%) and translocation (3%)
- Risk increases with maternal age (15-29 yrs, 1 in 1500; 30-34 yrs, 1 in 800; 35-39 yrs, 1 in 270; 40-44 yrs, 1 in 100; > 45 yrs, 1 in 50)
- **CHD (~ 40-50%)**
  - Endocardial cushion defect (AVSD/AVC) - ~ 40-50%
    ~ 50% of patients with AVC have Down Syndrome
  - Others: VSD, TOF, PDA, COA, almost never D-TGA
- **Other abnormalities:** duodenal atresia, hypothyroidism, gut malrotation and annular pancreas, atlanto-occipital dislocation
Turner Syndrome

- Bicuspid aortic valve (30%)
- Coarctation of the Aorta (10%)
Trisomy 13 (Patau’s) Syndrome

- Incidence of CHD ~90%
- VSD, PDA, Dextrocardia
Trisomy 18 (Edward’s) Syndrome

- Incidence of CHD ~99%
- VSD, PDA, PS, DORV
Noonan Syndrome

- Short stature, webbed neck, low posterior hair line, pectus excavatum, cubitus valgus, and cryptorchidism in males
- ? ‘Male Turner syndrome”
- Sporadic, may be autosomal dominant
- Mental retardation seldom severe
- Cardiac defects (~ 50%):
  - *Valvar pulmonic stenosis* (dysplastic valve) {75%}
  - *PPS, ASD, VSD, PDA, CoA, TOF*
  - *Hypertrophic cardiomyopathy*
  - *Other: ECG shows leftward or superior frontal QRS axis, despite the presence of severe RVH*
**VATER/VACTERL**
- Vertebral anomalies
- Anal atresia
- Cardiac: VSD
- T-E fistula
- Renal anomalies
- Limb anomalies: radial dysplasia

**CHARGE**
- Coloboma
- Heart: TOF, DORV, TA, VSD, aortic arch anomalies (vascular ring, IAA)
- Atresia choanae
- Retarded growth/mental
- Genital hypoplasia
- Ear anomalies
Cyanosis

- Cyanosis is a sign not a diagnosis.
- Bluish discoloration of skin, nail beds, and mucous membrane.
- Noticeable when the concentration of the deoxy-hemoglobin is at least 5 g/dl.
- Not every cyanosis is cardiac!
- Central vs. peripheral (acrocyanosis).
Causes of Cyanosis

- Primary parenchymal lung disease
  - Pneumonia, meconium aspiration syndrome
  - Problem is with $O_2$ diffusion
  - Responds to increased FiO$_2$
- Primary cardiac disease
  - Combination of decreased pulmonary flow and intracardiac mixing of “blue” and “pink” blood
  - Doesn’t respond much to increased FiO$_2$
Physiologic Classification of Cyanotic Lesions

- Intracardiac mixing of systemic and pulmonary venous returns (*increased PBF*)
  - D-TGA
  - TAPVC
  - Persistent truncus arteriosus
- Obstruction to pulmonary blood flow and intracardiac shunt (*decreased PBF*)
  - TOF
  - Tricuspid Atresia
  - Pulmonary Atresia
  - Ebstein’s anomaly (when severe)
The 5 T’s

- **TOF** (Tetralogy of Fallot)
- **TGA** (D- transposition of the great arteries)
- **Tr A** (Persistent truncus arteriosus)
- **TAPVR** (Total anomalous pulmonary venous return)
- **TA** (Tricuspid atresia “Tingle” \{single\} ventricle)
  - Single-ventricle lesions:
    - Hypoplastic left heart syndrome (HLHS)
    - Hypoplastic right heart syndrome (PA/TA)
    - Ebstein’s anomaly of the tricuspid valve (when severe)
    - Double-inlet left ventricle (DILV)
    - Unbalanced atrioventricular septal defect (unbalanced AVC)
Your Role!

- **Initiate work-up:**
  - Four extremities BPs
  - Oximetry (pre/postductal)
  - Hyperoxia test
  - CXR +/- EKG
  - ABGs

- **Initiate therapy:**
  - Don’t hesitate to start Prostaglandin \(0.0125-0.1 \text{ mcg/kg/min}\)
  - Avoid O2 unless necessary (Keep O2 sats > 75% unless associated acidosis)

- Always, when in doubt, call your cardiologist!
Cyanosis Work-Up

- Hyperoxia test
  - pO2 > 150 mm Hg --------- most likely respiratory
  - pO2 < 40 mm Hg --------- most likely cardiac
  - pO2 40-150 mm Hg --------- either possible

- Always obtain ABG/Oximetry from two areas:
  - Preductal (right arm, ear, radial artery)
  - Postductal (umbilical artery or feet)
**Cyanosis Work-Up**

**Differential Cyanosis**
Pred ductal 10% higher than postductal (pink upper and blue lower part of the body)
- PPHN
- Left heart obstructive lesions (severe AS, IAA, COA) with R→L ductal shunt
- TAPVR (Obstructed) with PPHN

**Reverse Differential Cyanosis**
Postductal 10% higher than preductal (blue upper and pink lower part of the body)
- D-TGA+PDA+PPHN
- D-TGA+PDA+COA/IAA
## Differential Diagnosis of a Cyanotic Infant

<table>
<thead>
<tr>
<th></th>
<th>Breathing Pattern</th>
<th>Metabolic acidosis</th>
<th>pCO₂</th>
<th>Response to 100% O₂</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Primary pulmonary disease</strong></td>
<td>Resp. distress</td>
<td>No</td>
<td>↑</td>
<td>↑ pO₂ &amp; SaO₂</td>
</tr>
<tr>
<td><strong>Cardiac</strong></td>
<td>Happy tachypnea</td>
<td>present</td>
<td>NL/↓</td>
<td>No change</td>
</tr>
<tr>
<td><strong>PPHN</strong></td>
<td>Resp. distress</td>
<td>+/-</td>
<td>NL/↑</td>
<td>+/-</td>
</tr>
<tr>
<td><strong>Sepsis</strong></td>
<td>+/- resp. distress</td>
<td>+/-</td>
<td>NL/↑</td>
<td>Moderate ↑ pO₂ &amp; SaO₂</td>
</tr>
</tbody>
</table>
Patent Ductus Arteriosus (PDA)

- F:M 3:1
- Common in premature infants
- Functional closure occurs within 12 hours after birth
- Anatomic closure occurs within 2-3 weeks after birth
- It represents the distal portion of the 6th embryonic aortic arch
- The hemodynamics is similar to those of VSD
Ductus Arteriosus

- **What keeps it open?**
  - Low pO2/hypoxia
  - Prostaglandins
  - Nitric oxide
  - Circulating adenosine

- **What constricts it?**
  - O2
  - Endothelin-1
  - Norepinephrine, acetylcholine, and bradykinin
  - Indomethacin, ASA
PDA... Auscultation

- Cardiac exam:
  - Bounding peripheral pulses
  - Wide pulse pressure
  - Hyperactive precordium
  - S2: normal to single and loud
  - 1-4/6 Crescendo systolic/continuous murmur "machinery" at ULSB
  - Apical Mid-diastolic rumble (with significant shunt)
PDA

- More frequent in premature babies because of decreased smooth muscle receptors and decreased sensitivity to vasoconstrictors
- Long term risks: CHF, FTT, recurrent pneumonia, endocarditis, calcification, aneurysm formation, and PVOD
PDA… Management

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

- **Large Ductus**
  * Surgery

- No residual heart disease
- D/C 1-2 yrs post op
- Coil follow up yearly
Coarctation of the Aorta

- Juxtaductal obstruction produces upper extremity hypertension and lower extremity hypotension
- Flow to lower body may be severely diminished when PDA closes
- Acidosis and CHF symptoms result from decreased lower perfusion
- Associated with bicuspid aortic valve, Turner Syndrome (XO) and aortic arch hypoplasia
Transposition of the Great Arteries (D-TGA)

- ~ 5% of all CHD
- M:F = 3:1
- A communication at atrial, ventricular, or arterial level is necessary for survival
- Associated defects: VSD, PS, COA, IAA, TA, and *coronary abnormalities*
The aorta arises from the RV, and the pulmonary artery arises from the LV. As a result, there is a complete separation of the pulmonary and systemic circulation.
**Clinical Features**

- Infants are often large for gestational age (LGA)
- Moderate to severe cyanosis from birth
- Happy tachypnea (tachypnea without retractions unless CHF develops)
- Physical findings vary with associated defect(s)
- S2 is single and loud (representing closure of the anteriorly placed aortic valve)
- Usually no murmur is heard if no other lesions exist (VSD/PS)
D-TGA

- **ABG:** severe hypoxemia +/- acidosis
- **ECG:** RAD, RVH/CVH, RAE
- **CXR:**
  - Cardiomegaly
  - Increased PVMs
  - *“Egg on a string”* appearance due to narrow mediastinum because the great vessels lie one in front of the other
D-TGA ... Management

1- Ensure mixing and improve arterial oxygen saturation:
   a. **Prostaglandin E1 (PGE1)** at 0.025-0.1 mcg/Kg/min IV infusion to maintain ductus arteriosus patency.
   {S/E: apnea, hypotension, jitteriness/seizures fever, rash, diarrhea, plts inhibition}
   b. **Rashkind** balloon atrial septostomy (BAS)

2- Treat CHF

   **Note: Oxygen can be bad ???**
   (PDA closure and pulmonary vessels dilation leading to CHF)
Balloon Atrial Septostomy (Rashkind Procedure)

Balloon-tipped catheter is inserted through the atrial septal defect (ASD)

Transposition of Great Arteries

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.
Arterial Switch Operation

Aorta

Pulmonary artery

Patch in place of old coronary artery origin.

Aorta is "switched" with the pulmonary artery.

Coronary arteries

Both transposed arteries are divided at the red dotted line.

Coronary arteries are detached from aortic valve (on right side of heart) and connected to pulmonic valve (on left side of heart).
Hypoplastic Left Heart Syndrome

- Most serious of all defects
- Staged surgical repair
- Significant morbidity/mortality
Norwood: Surgical Correction
Modified Blalock-Taussig shunt

Left: The base of the pulmonary artery is connected with the aorta, and a shunt is placed between a branch of the aorta and the other part of the pulmonary artery.

Right: A new pathway to bypass the left side of the heart is created. Blood moves through the pulmonary artery to the aorta and out to the body. Some blood moves through the Blalock Taussig shunt to the pulmonary artery, connected to the lungs.
Fontan Procedure

First stage: Bi-Directional Glenn
A graft is used to route blood flow from the superior vena cava to the pulmonary artery instead of to the right atrium.

Second stage: Fontan
Both a graft and an internal baffle (wall) are used to route blood flow from the inferior vena cava to the pulmonary artery instead of to the right atrium.
Thank You

QUESTIONS?

COMMENTS!