Evaluation of a Cyanotic Newborn

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(Special thanks to John Kinsella)
SBN Lecture Series
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Background: CHD

- Congenital heart disease (CHD): 8/1000 live births
  - 2/1000 will present at <1 year of age
  - 25% with additional non-cardiac anomalies

- Critical CHD: 3.5/1000 live births

- In raw numbers, approximately 32,000 children born with CHD

- 14,000 of these children have critical heart disease
<table>
<thead>
<tr>
<th>Congenital Heart Defect</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect (VSD)</td>
<td>16% (VSD is the most common congenital heart defect)</td>
</tr>
<tr>
<td>Pulmonary stenosis with intact ventricular septum</td>
<td>7.5 – 12%</td>
</tr>
<tr>
<td>Tetralogy of Fallot (TOF)</td>
<td>8-10% (TOF is the most common cyanotic heart disease beyond infancy)</td>
</tr>
<tr>
<td>Atrial septal defect (ASD)</td>
<td>6-11%</td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td>5-10% (TGA is the most common cyanotic heart disease presenting in the first week of life)</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>4-10% in full term infants</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>5-8%</td>
</tr>
<tr>
<td>Atrioventricular septal defects</td>
<td>2-5%</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous return</td>
<td>1-2.5%</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>1-4%</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>1.5% (HLHS is the 2nd most common cyanotic heart disease presenting in the first week of life)</td>
</tr>
</tbody>
</table>
## Recurrence Risk of Congenital Cardiac Diseases

<table>
<thead>
<tr>
<th>Cardiac Lesion</th>
<th>Previous Sibling Affected</th>
<th>Father Affected</th>
<th>Mother Affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>Marfan’s syndrome</td>
<td>-</td>
<td>50</td>
<td>50</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>2</td>
<td>3</td>
<td>15-17.9</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>2</td>
<td>2</td>
<td>6.5</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>3</td>
<td>2</td>
<td>9.5-15.6</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>2.5</td>
<td>1.5</td>
<td>4.6-11</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>3</td>
<td>2.5</td>
<td>4.1</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>-</td>
<td>-</td>
<td>14.1</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>2.5</td>
<td>1.5</td>
<td>2.6</td>
</tr>
</tbody>
</table>
Genetic Syndromes: CHD

- Trisomy 21: AVSD (30%)
- Turner (XO): Coarctation, AS
- Cri-du-chat (5p-): VSD
- Williams (microdel. 7)-supravalvular AS, PPS
- Noonan’s (AD Chr 12)—Pulmonary stenosis
Maternal Causes: CHD

- **Systemic illnesses**
  - Diabetes: HCM
  - SLE: Complete heart block

- **Intrauterine infection**
  - Rubella: PDA, septal defects, PPS

- **Toxicology**
  - Anticonvulsants—AS, PS, Coarctation
  - Excessive alcohol—Septal defects
Board Question

- Match the congenital heart disease with the syndrome, associated clinical anomalies or maternal condition

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Congenital Heart Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Webbed neck, congenital lymphedema, prominent ears, broad chest</td>
<td>A: Interrupted aortic valve</td>
</tr>
<tr>
<td>Hypotonia, protuberant tongue, flat facial profile, duodenal obstruction</td>
<td>B: Coarctation of the aorta</td>
</tr>
<tr>
<td>Severe hypocalcemia and seizures, cellular immunity deficit, and infections</td>
<td>C: Dysplastic pulmonary valve</td>
</tr>
<tr>
<td>Growth retardation, microcephaly, long philtrum, and smooth upper lip</td>
<td>D: AV canal</td>
</tr>
<tr>
<td>Cryptorchidism, small penis, webbed neck, pectus excavatum, “Turner-like”</td>
<td>E: VSD</td>
</tr>
</tbody>
</table>
Cyanosis

- Bluish discoloration due to arterial oxygen desaturation of any cause

- Occurs when concentration of capillary deoxyhemoglobin exceeds 3-5 gm/dL
  
  **NOTE:** defined by absolute amount of reduced Hb (NOT the oxygen saturation)
Cyanosis

- $\text{Hb} = \text{reduced Hb} + \text{HbO}_2$

- $\text{O}_2\text{saturation} = \frac{\text{HbO}_2}{(\text{HbO}_2 + \text{reducedHbO}_2)}$

- Occurs at different saturations depending on Hb level

- So, if an anemic infant has a Hb of 8 g/dL and one would expect cyanosis to appear if there is a 5 g reduced Hb/dL, the HbO2 must be 3. The O2 saturation at which the infant would appear cyanotic is $\frac{3}{8} = 37\%$

- Similarly, if a polycythemic infant had a Hb of 22 g/dL, the HbO2 at which the infant will appear cyanotic is 17. The O2 saturation at which the infant would appear cyanotic is... 77\%
Board question

- Match the maximum saturation at which the described infant become cyanotic

<table>
<thead>
<tr>
<th>Infant Type</th>
<th>Hemoglobin Concentration</th>
<th>Saturation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polycythemic infant with Hb of 21 gm/dL</td>
<td>Saturation of 100%</td>
<td></td>
</tr>
<tr>
<td>Normal infant with Hb of 16 gm/dL</td>
<td>Saturation of 81%</td>
<td></td>
</tr>
<tr>
<td>Anemic infant with Hb of 9 gm/dL</td>
<td>Saturation of 50%</td>
<td></td>
</tr>
</tbody>
</table>
Pathophysiology

- Who are the players involved in the cyanosis?
- Where is there a defect in the system?
  - Heart
    - Is there a problem with the delivery of blood to the lungs?
    - Is there a right to left shunt?
  - Lungs
    - Is there a problem with oxygenation as it passes through the lungs?
  - End organs
    - Is there a shift with oxygen dissociation?
  - Blood
    - Is there a defect in the oxygen carrying capacity?
HEART

- Factors favoring R to L shunting
  - Abnormal RV compliance
  - Atretic or stenotic pathways
  - Elevated PVR
- Single ventricle physiology
- AV discordance
Extrapulmonary Right-to-Left Shunt
Causes of Hypoxemia in Neonatal Lung Disease

Extrapulmonary

- Right-to-left shunting across “fetal” channels
  - Foramen ovale (FO)
  - Patent ductus arteriosus (PDA)
LUNGS

- Anatomic barriers
- Mechanical
- Oxygen diffusion capacity is compromised
- Hypoxia
- Lung ventilation distribution and atelectasis
Lung Disease Causing Hypoxemic Respiratory Failure in the Newborn

- Meconium aspiration syndrome
- Conditions characterized by diffuse, homogeneous parenchymal disease (RDS, pneumonia)
- Pulmonary hypoplasia (oligohydramnios, congenital diaphragmatic hernia [CDH])
Causes of Hypoxemia in Neonatal Lung Disease

Intrapulmonary

- Intrapulmonary shunting: Pulmonary arterial blood that reaches pulmonary venous side without passing through ventilated areas of the lung

- Ventilation:perfusion (V/Q) inequality: Imbalance between ventilation and perfusion; increased dead-space ventilation
Intrapulmonary Shunt and V/Q Mismatch
Relationship of Lung Disease to Pulmonary Hypertension

- Pulmonary vascular resistance (PVR) increases at lung volumes above and below functional residual capacity
  - Underinflation in RDS
  - Overinflation in MAS

- Extremes of lung volume contribute to high pulmonary vascular resistance and extrapulmonary shunting
PVR Increases at Lung Volumes Below and Above FRC
Lung Recruitment With HFOV Augments Response to INO in Newborns With PPHN and Lung Disease

Kinsella et al, J. Pediatrics, 1997
Hb has more affinity for curves to the left, which means POORER oxygen delivery (LEFT, L or LOW suggesting LOW acid, LOW paCO2, LOW 2,3 DPG, LOW temp)

Hb has less affinity of oxygen towards the right, which means BETTER oxygen delivery (RIGHT, R for release with Inc 2,3 DPG, Inc H+ and Inc Temp)
BLOOD

- Hemoglobinopathies
  - Sickle cell disease
  - Methemoglobinemia: KNOW THIS: the iron of Hb changes from ferrous (reduced) to ferric (oxidized), decreasing ability of Hb to bind O2.
    - So you get a normal paO2, but decreased oxygen saturations
  - Thalassemia

- Anemia
- Polycythemia
Initial Evaluation of the Term Newborn with Cyanosis

Prenatal
- Prenatal ultrasound study results
- History of oligohydramnios and duration
- History of fetal brady/tachyarrhythmia
- Maternal illnesses, drugs, medications
- History of fetal distress
- Risk factors for infection

Delivery
- History of positive pressure ventilation in DR
- Meconium stained amniotic fluid
- Hemorrhage
- Trauma
- Apgar score
Physical Examination

Respiratory Distress (retractions, grunting, nasal flaring)
Suggests lung parenchymal disease (↓ compliance), airway disease or metabolic acidemia

No Significant Respiratory Distress (tachypnea alone)
Suggests hypoxemia caused by cyanotic heart disease without lung disease
Role of Pulse Oximetry

Preductal / postductal SaO₂

Preductal SaO₂ = postductal SaO₂
1) Intrapulmonary shunt: (PVR<SVR)
2) Cyanotic CHD with left-to-right PDA: (Ductal-dependent Qp: e.g. PS, tricuspid atresia, Ebstein’s)
3) PPHN: extrapulmonary shunt at FO: (PVR>SVR with closed DA)

Preductal SaO₂ > postductal SaO₂
1) PVR>SVR with R→L PDA: PPHN with MAS, RDS, CDH, or idiopathic
2) Ductal-dependent Qs: HLHS, AS, IAA, coarctation
3) Anatomic pulm. vasc. disease: alveolar-capillary dysplasia, pulmonary venous stenosis, TAPVR with obstruction.

Preductal SaO₂ < postductal SaO₂
1) TGV with pulmonary hypertension
2) TGV with coarctation of aorta
Response to Supplemental Oxygen

Acute response to high FiO₂ (hood, mask)

- Minimal or transient change in SaO₂
  - Cyanotic heart disease, PPHN
- Marked improvement in SaO₂
  - Parenchymal lung disease, CHD with ductal-dependent systemic blood flow
**CXR and Laboratory Evaluation**

**Chest Radiograph**
- Hypoxemia out of proportion to radiographic changes
- CHD with ductal-dependent pulmonary blood flow or extrapulmonary right-to-left shunting with PPHN

**ABG, CBC, 4 ext. BP**
- ABG: Assess respiratory and metabolic acidemia
- CBC: For evidence of infection
- BP: Ductal-dependent systemic blood flow and closing PDA: (e.g. coarctation, IAA)
Meconium Aspiration Syndrome
**Hyperoxia test**

- ABG pre-ductal sample from the right radial artery

<table>
<thead>
<tr>
<th>FiO2 0.21</th>
<th>FiO2 1.00</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>pO2 50 - 150</td>
<td>pO2 50 - 150</td>
<td>Cardiac</td>
</tr>
<tr>
<td>pO2 50 - 150</td>
<td>pO2 150 – 300</td>
<td>Pulmonary</td>
</tr>
<tr>
<td>pO2 50 - 150</td>
<td>pO2 &gt; 300</td>
<td>Normal</td>
</tr>
</tbody>
</table>
Role of Echocardiography

R→L shunting at PDA and/or FO with normal LV performance
Consider iNO use after effective lung recruitment.

Anatomic abnormalities
Ductal-dependent systemic blood flow:
HLHS, critical AS, coarctation, IAA

Functional abnormalities
LV dysfunction with mitral insufficiency:
L→R atrial shunting with R→L ductal shunting=pulmonary venous hypertension
? RV dependent systemic blood flow
What do you want to know from an echo??

- Structural abnormalities
- Heart function: LV dysfunction
- Shunts
  - Intracardiac (extrapulmonary)
    - PFO
    - PDA
- Measurement of PA pressure
  - Tricuspid regurgitation
Cardiopulmonary Interactions in PPHN

- **Pulmonary Vascular**
  - (Structural changes; altered reactivity to dilator and constrictor stimuli)
  - Right-to-left shunting at PDA and FO
  - Hypoxia, hypercarbia, acidosis
  - Lung
    - ↓ Lung volume
    - ↓ Compliance
    - ↑ Intrapulmonary shunt

- **Heart**
  - RV pressure overload and LV dysfunction

- **PVR**
  - SVR ↓
Relationship of Cardiac Dysfunction to Pulmonary Hypertension

- Left ventricular dysfunction causes mitral insufficiency, increased left atrial end diastolic pressure, and pulmonary venous hypertension.
- Increasing pulmonary blood flow does not improve systemic oxygenation in this setting.
Lesion Classification

- Too much pulmonary blood flow—50%
  - Acyanotic

- Too little pulmonary blood flow
  - Cyanotic

- Obstructive physiology
  - Low cardiac output—variable cyanosis

- Transposition/mixing physiology
  - Variable to severe cyanosis

- Combinations
Ductal Dependence

PDA

- Pulmonary blood flow
- Mixing
- Systemic blood flow
Pulmonary Obstruction

- Not ductal dependent
  - TOF
  - Pulmonary stenosis

- Ductal dependent
  - Critical PS
  - Severe TOF
  - Pulmonary atresia
  - Tricuspid atresia
Systemic Obstruction

- Not ductal dependent
  - AS
  - Coarctation

- Ductal dependent
  - Critical AS
  - Critical coarctation
  - IAA
  - HLHS
## BOX 43-9. Classification of Congenital Heart Disease

<table>
<thead>
<tr>
<th>Severe Cyanosis Caused by Separate Circulations and Poor Mixing</th>
</tr>
</thead>
<tbody>
<tr>
<td>D-Transposition of the great arteries</td>
</tr>
<tr>
<td>D-Transposition of the great arteries and ventricular septal defect</td>
</tr>
<tr>
<td>Double-outlet right ventricle with subpulmonary ventricular septal defect (Taussig-Bing)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Severe Cyanosis Caused by Restricted Pulmonary Blood Flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>Double-outlet right ventricle with subaortic ventricular septal defect and pulmonary stenosis</td>
</tr>
<tr>
<td>Tricuspid atresia</td>
</tr>
<tr>
<td>Pulmonary atresia with intact interventricular septum</td>
</tr>
<tr>
<td>Critical pulmonary stenosis</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
</tr>
<tr>
<td>Single ventricle with pulmonary stenosis</td>
</tr>
<tr>
<td>Persistent pulmonary hypertension</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Mild Cyanosis Caused by Complete Mixing with Normal or Increased Pulmonary Blood Flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total anomalous pulmonary venous connection</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
</tr>
<tr>
<td>Single ventricle without pulmonary stenosis</td>
</tr>
<tr>
<td>Double-outlet right ventricle with subaortic ventricular septal defect</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Systemic Hypoperfusion and Congestive Heart Failure with Mild or No Cyanosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic stenosis*</td>
</tr>
<tr>
<td>Coarctation of the aorta and aortic arch interruption</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
</tr>
<tr>
<td>Multiple left heart defects</td>
</tr>
<tr>
<td>Single ventricle with subaortic stenosis or coarctation of the aorta</td>
</tr>
<tr>
<td>Myocardial diseases: cardiomyopathy and myocarditis*</td>
</tr>
<tr>
<td>Cardiac tumor*</td>
</tr>
<tr>
<td>Arteriovenous malformation*</td>
</tr>
<tr>
<td>Hypertension*</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Acyanosis with No or Mild Respiratory Distress</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal murmurs</td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
</tr>
<tr>
<td>Ventricular septal defect*</td>
</tr>
<tr>
<td>Atrial septal defect</td>
</tr>
<tr>
<td>Endocardial cushion defect*</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
</tr>
<tr>
<td>Aortopulmonary window*</td>
</tr>
<tr>
<td>I-Transposition of the great arteries</td>
</tr>
<tr>
<td>Arteriovenous malformation</td>
</tr>
<tr>
<td>Hypertension</td>
</tr>
</tbody>
</table>

*No symptoms with mild forms of disease.

*Congestive heart failure can develop as left-to-right shunt increases with decrease in pulmonary vascular resistance.
Algorithm for diagnosis of CHD

Five “T’s”, “DO” and “ESP” = TGA, Tetralogy, TAPVR, Tricuspid atresia, truncus arteriosus, DORV, Ebstein’s, Single ventricle, and pulmonary atresia
<table>
<thead>
<tr>
<th></th>
<th>Ductal PBF</th>
<th>Ductal SBF</th>
<th>Mixing</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cardiac Output</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Perfusion</td>
<td>Normal</td>
<td>Decreased</td>
<td>Normal to decreased</td>
</tr>
<tr>
<td>Acidosis</td>
<td>Normal</td>
<td>Severe</td>
<td>Mild</td>
</tr>
<tr>
<td><strong>Cyanosis</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Amount</td>
<td>Moderate</td>
<td>Mild</td>
<td>Severe</td>
</tr>
<tr>
<td></td>
<td>70-80%</td>
<td>80-90%</td>
<td>40-70%</td>
</tr>
<tr>
<td>Differential</td>
<td>None</td>
<td>?Decreased</td>
<td>Rare</td>
</tr>
<tr>
<td></td>
<td>lower ext</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>CXR</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Heart size</td>
<td>Normal</td>
<td>Increased</td>
<td>Normal</td>
</tr>
<tr>
<td>Vascularity</td>
<td>Decreased</td>
<td>Normal to</td>
<td>Increased</td>
</tr>
<tr>
<td></td>
<td>increased</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Transposition of the Great Arteries

- Know the difference between D and L-TGA (D has AV concordance)
- Most common cyanotic lesion in first week of life
- Greater in males 3:1
Transposition

- “egg on a string”
- Severe cyanosis at birth as a result of separate circulations and poor mixing, particularly if restrictive PFO
- Manage with PGE, Rashkind procedure if inadequate mixing, and eventual arterial switch (Jatene)
Tetralogy of Fallot

- Primary defect: underdevelopment of the right ventricular infundibulum which causes:
  - Anterior malalignment VSD
  - Pulmonary stenosis
  - Over-riding aorta
  - Right ventricular hypertrophy

- Associated with right AA(25%), 22q11 (16%), Trisomy 21 (7%), Alagille Syndrome
Tetralogy

- “boot shaped heart”
- Spectrum of clinical presentation is related to degree of RVOT
- Hypercyanotic spells, or “tet” spells
  - Balance between SVR and PVR
  - ↓PVR: comfort, feed, sedate, oxygen
  - ↑SVR: knee to chest, phenylephrine
- PGE, palliative BT shunt to eventual pulmonary arterioplasty and VSD patch
Tricuspid Atresia

- Complete absence of tricuspid valve
- With minimal VSD: leads to poor RV development because of lack of blood flow in utero, ductal dependent, requires shunt
- With VSD: VSD determines size of RV
- PA is common
Tricuspid atresia

- PBF is supplied by L to R shunt at PDA or VSD
- If baby has small VSD or PS—need PGE—may need BT shunt
- If large VSD and no PS, as PVR falls, may have increased PBF and be at risk for CHF—may need PA band
Truncus arteriosus

- Single great vessel gives rise to both aorta and pulmonary arteries
- Large VSD is always present, leading to complete ventricular mixing
- As PVR falls, further increase in PBF
Truncus arteriosus

- Cyanosis and CHF depends on PBF
- Clinical signs: low diastolic pressure, widened pulse pressure due to diastolic runoff into PA, loud murmur, increased heart size
Ebstein’s anomaly

- Tricuspid valve is displaced into RV
- Portion of RV wall above the displaced leaflets becomes “atrialized”
- TV regurgitation
- Atrial communication if 80% with R to L shunt, thus requires PDA for adequate PBF
Ebstein’s Anomaly

- If severe, cyanosis secondary to decreased PBF
- CXR shows dramatic cardiomegaly
- Treat with PGE if need to increase PBG, treat CHF, assess for airway compression, limited surgical options
TAPVR

- **Supracardiac** (most common): PV via vertical vein to innominate, azygous or directly to SVC, “snowman CXR”
- **Cardiac**: PV into RA directly or indirectly via coronary sinus
- **Infracardiac**: PV cross diaphragm and drain into portal vein or hepatic vein or IVC
- **Mixed**
<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Obstructive TAPVR</th>
<th>Nonobstructive TAPVR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patency</td>
<td>Infracardiac (subdiaphragmatic) almost always obstructive</td>
<td>Majority of intracardiac and supracardiac</td>
</tr>
<tr>
<td>Clinical findings</td>
<td>Pulmonary edema, CHF Cyanosis, respiratory distress Decreased systemic perfusion</td>
<td>Increased PBF, CHF Mild to moderate cyanosis Systolic murmur</td>
</tr>
<tr>
<td>CXR</td>
<td>Normal or small heart because decreased PBF Lung fields similar to aspiration PNA</td>
<td>Increased heart size, pulmonary congestion</td>
</tr>
<tr>
<td>ECG</td>
<td>RVH, or may be normal</td>
<td>Right axis deviation, RVH, RAE</td>
</tr>
<tr>
<td>Management</td>
<td>Medical treatment of CHF Treatment with PGE may worsen symptoms by increasing PBF, in some cases PGE may be helpful by dilating ductus venous and improving venous return, surgery is urgent</td>
<td>Medical treatment of CHF Surgery not always needed immediately</td>
</tr>
</tbody>
</table>
DORV

- Aorta and pulmonary artery arise from RV
- VSD is required to provide an outlet from LV, and defines type of DORV
- VSD can be subaortic (TOF or VSD), subpulmonic (TGA), doubly committed or remote
Single ventricle

- Both AV valves empty into as single ventricle
- Think tricuspid atresia physiology or HLHS physiology
- Oxygen saturations and clinical symptoms depend on balance between $Q_p/Q_s$
- If increased PBF: CHF, mild cyanosis
- If decreased PBF: similar to TOF with moderate, severe cyanosis
A full newborn has moderate respiratory distress and is cyanotic. The pO2 from the umbilical artery catheter is 80 mmHg, at the same time the pO2 from the umbilical venous catheter is 200 mmHg. What is the most likely diagnosis?

A. Persistent fetal circulation (pulmonary hypertension)
B. TGA
C. Truncus arteriosus
D. Infra-diaphragmatic TAPVR
E. HLHS
Match the time of presentation

A. TGA
B. HLHS
C. CoAorta
D. VSD

Answer: DCBA
A term 3.2 kg baby is noted to have tachypnea and poor perfusion. Septic w/u done and ABx started. Baby’s sats pre and post were 82%. O₂ was given via hood but condition worsen and baby was placed on ventilator (Vt 20 ml PEEP 5 IT 0.35 O₂ 70% rate 50). CXR showed ETT at T2 with slight increase PVMs. No infiltrate is seen. The blood gas showed pH 7.24/ pCO₂ 38/ pO₂ 46/ HCO₃ 18/-8. The sats are still running low, last reading is 78%. The most appropriate action at this time is to

A. Give surfactant  
B. Give IVF bolus  
C. Increase Vt, decrease rate & increase FiO₂  
D. Decrease Vt, decrease rate & decrease FiO₂  
E. Decrease IT, decrease PEEP & increase FiO₂

Preferred response is D.

O C R (Objective, Critique, Reference)

Objective:
To know the effect of O₂ administration on HLHS.

Critique:
Hypoplastic left heart is unique in a sense that O₂ supplementation worsens the condition due to steal of blood from PDA to go the dilated pulmonary vasculature (in response to O₂ therapy). It is therefore important to decrease vent setting and O₂ in HLHS.
During the lecture on congenital heart disease the presenter emphasizes on the use of prostaglandin in all cyanotic infants while waiting for echocardiogram. All of the following are true about prostaglandin (PGE1) EXCEPT

A. It is given as continuous drip  
B. It causes apnea, which is dose dependent  
C. Long term use cause osteopenia  
D. It causes blood flow from Pul A to Ao through PDA in HLHS  
E. It causes blood flow from Ao to Pul A through PDA in TOF

Preferred response is C.

O C R (Objective, Critique, Reference)

Objective:
To know the uses and complications of PGE1

Critique:
Long term use causes periosteal thickening not osteopenia. All other statements are true.
A 32 year-old lady with no prenatal care presents to L&D fully dilated. She delivered a 3.2 kg baby, who remained cyanotic despite oxygen therapy. You took the baby to the NICU and placed him on sat monitor. Breathing on RA, the predectal sats are 62% while postductal sats are 72%. On examination you noticed bounding pulses and tachypnea. No murmur is heard. CXR showed increase heart size with increased PVMs. EKG showed right axis. You order an urgent echo and started the PGE₁ drip. The parents are at the bedside and you are explaining about the possibility congenital heart disease. The mom tells you that she had mitral valve prolapse and father had operation for ASD last year. During the conversation mom touched the baby and baby stops breathing. You called the code and started CPR. The most likely cause for infant deterioration is

A. PGE₁ drip
B. Ductus closure
C. Laryngeal spasm
D. Subtle seizure
E. Blocked airway

Preferred response is A.

O C R (Objective, Critique, Reference)

Objective:
To know the side effects of PGE₁

Critique:
Apnea is one of the commonest side effects of PGE, others are flushing, fever & bradycardia
A 3.5 kg NB male is transferred from regional hospital with cyanosis. History reveals late deceleration which prompted CS. Apgar was 3/5/7. On arrival to your hospital, you noted the baby to be dusky with sats of 85%. On exam you noted poor air entry with gr 2/6 murmur at LSB. You immediately intubated the baby and placed him on ventilator with 100% FiO₂ and MAP of 12. A UAC is place by the NNP and the gas showed pH 7.26/ O₂ 45/ CO₂ 51/ HCO₃ 19/ -6. An urgent echo was obtained which showed no structural heart defect. You expect the Qp/Qs ratio to be

A. > 1  
B. < 1  
C. = 1  
D. same as OI  
E. same as AaDO₂

Preferred response is B.

O C R (Objective, Critique, Reference)

Objective:

To know the value of Qp/Qs ratio

Critique:

Qp/Qs ratio: Pulm BF/Sys BF, as PBF is numerator, increase flow will give a ratio > 1 or vice versa. In PPHN PBF is lower than SysBF, so ratio would be < 1.

AaDO₂ = (713 x FiO₂) – (pCO₂ / 0.8) – (paO₂).
24.) An infant with suspected congenital heart disease is transferred to your NICU. Based on data obtained at the outside hospital, you determine that the Qp / Qs ratio is < 1 (0.5). Based on this data, what type of lesion is suspected?

A. Large VSD with pulmonary edema
B. AV canal with balanced systemic and pulmonary blood flow
C. Pulmonary hypertension secondary to meconium aspiration with a pre-ductal saturation of 88% and post-ductal saturation of 60%
D. PDA at 4 days of life in an ex 28-week infant who now requires more support
E. Hypoplastic left heart at 3-days of life with post-ductal saturations of 100%.

L to R shunt if Qp/Qs > 1
R to L shunt if Qp/Qs < 1
22.) The time period when most cardiac abnormalities develop during gestation is when?

A. 1 week 
B. 4 weeks 
C. 6 weeks 
D. 8 weeks 
E. 12 weeks

The answer is D.
25.) The second most common cause of cyanotic heart disease that presents during the first week of life is what lesion?

A. TGA  
B. AV canal  
C. Coarctation of the aorta  
D. HLHS  
E. TAPVR

The answer is D.
26.) You receive a phone call from the newborn nursery to evaluate a cyanotic baby at 20-hours of life. During your initial examination, the baby is tachypneic, but otherwise comfortable. Oxygen saturations on the right foot are 75%, despite FiO2 1.0 being administered via blow by. Further physical examination demonstrates a single S2 heart sound with a Grade III/VI blowing murmur at the left sternal border. You transfer the neonate to the NICU and place the neonate under a 100% oxygen hood for approximately 15-minutes. An arterial blood gas from the left radial artery reveals a PaO2 of 45mmHg. Unfortunately, all of the cardiologists are at a national conference and the technicians are unable to be reached. You obtain an EKG and find a left superior axis. Based on these findings, what is your suspected diagnosis?

A. Truncus arteriosus
B. Tricuspid atresia
C. HLHS
D. Critical PS
E. Pulmonary atresia

The answer is B.
27.) Cataracts, microcephaly, “blueberry muffin” skin lesions, growth retardation  
28.) Arachnodactyly with hyperextensibility, inguinal hernias, lens subluxation  
29.) Situs inversus, RBC’s with Holly Jolly bodies and Heinz bodies  
30.) Elfin facies, prominent lips, blue eyes, open mouth, transient hypercalcemia  

A. Interrupted inferior vena cava  
B. VSD  
C. Supravalvular aortic stenosis  
D. PDA  
E. Aortic aneurysm
35.) A full term infant is found to be cyanotic in the newborn nursery. You place the infant on 100% oxygen and are amazed to find that the saturations in the right hand are 75% while the saturations in the right foot are 95%. Based on these findings, what is the most likely congenital heart lesion?

A. Persistent fetal circulation  
B. Transposition of the Great Vessels  
C. Truncus arteriosus  
D. Infra-diaphragmatic total anomalous pulmonary venous return  
E. Hypoplastic left heart syndrome

The answer is B
127.) The most common cardiac anomaly associated with congenital Rubella Syndrome, especially if occurs < 8-weeks gestational age is?

A). Patent ductus arteriosus  
B). Transposition of the great vessels  
C). Aortic stenosis  
D). Ventricular septal defect  
E). Atrial septal defect

The answer is A.