Toledo Children's Hospital
On the campus of The Toledo Hospital

# Heart of the Matter: Transport of the Infant with Suspected Cardiac Disease

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## Case Presentation

- Term male infant born to a 20 y.o. primagravida
- Pregnancy, labor and delivery were unremarkable
- After birth the infant was noted to be tachypneic (RR 70) and mildly cyanotic (O2 sat 78% in room air).
- The referring hospital is concerned and call for advice and to request a transport team.

## Case Presentation Continued

- On arrival of the team the infant has a RR of 90 breaths per minute and is breathing shallow; is mottled and poorly perfused and has a O2 sat of 55% in 100% hood. His temperature is 99, his heart rate is 180 bpm and he is lethargic.
- The local nurses cheer when you walk in and move away from the bedside in relief.

## Initial Assessment

ALWAYS

- A – airway
- B – breathing
- C – circulation

- Concurrently you will be asking a problem directed history

## Differential Diagnosis of Cyanosis

- Respiratory (lack of oxygen delivery to blood)
  - Obstruction
  - Infection
  - Pneumothorax
- Hematologic (decrease oxygen carrying capacity)
  - Bleeding
  - Abnormal hemoglobin
- Cardiac
  - Structural disease (mixing)
  - Poor cardiac function (lack of oxygen delivery to tissues)

## Maternal Risk factors
for Newborn Problems

- Respiratory
- Infection
- Hematological
- Cardiac

## Maternal Risk factors
for Newborn Problems

- Maternal risk factors for respiratory problems in the newborn – maternal infection, meconium stained fluid, bloody amniotic fluid, maternal diabetes.
- Maternal risk factors for infection in the newborn – recent or current maternal infection (GBS, UTI, viral, maternal temp or elevated WBC), length of ROM.
- Maternal risk factors for hematological problems in the newborn – family history of bleeding disorders

## Maternal Risk Factors Continued

- Risk factors for cardiac problems:
  - Congenital infection
  - Family history of congenital heart disease
  - Maternal diabetes
  - Prenatal exposure to alcohol, drugs or medications

## Cardiac Risk Factors
### Congenital Infection

- Maternal Rubella infection – infection in the first half of pregnancy has 50% chance of infecting the fetus – septal defects and myocardial disease.
- Any congenital viral infection - myocarditis

## Cardiac Risk Factors
### Family History of Congenital Heart Disease

- Mom with CHD – increases risk 6-15 fold (AS, AVC)
- Father with CHD – increases risk 1-3 fold (AS)
- Previous child with CHD – increase risk 2-3 fold (EF, VSD, AS)
- Family history of genetic syndromes:
  - Trisomy 21 - AVC, TOF, VSD
  - Trisomy 18 - VSD
  - Trisomy 13 - VSD
  - Turners - CoA
  - 22 q11 deletion - DiGeorges – Truncus Arteriousis

## Cardiac Risk Factors
### Maternal Diabetes

- Pre-pregnancy diabetes - exposure to high glucose levels in the first trimester:
  - TGA, VSD
- Pre-pregnancy and gestational diabetes – exposure to high glucose levels in the third trimester:
  - Diabetic cardiomyopathy – left ventricular outflow tract obstruction in severe cases

## Cardiac Risk Factors
### Prenatal Exposure to Alcohol, Drugs or Medications

- Alcohol – VSD, TOF
- Valproic acid – HLHS, PAt without VSD, VSD
- Dilantin – PS, AS, CoA
- Trimethadione – TGA, TOF, HLHS
- Accutane – TGA, TOF, Truncus, DORV, HLHS
- Amphetamines – VSD, TGA
- Progesterone, estrogen – VSD, TOF, TGA
- Maternal PKU – 15% with variable defects

## Typical Clinical Presentation
of the infant with cyanotic cardiac disease

- Normal pregnancy, labor and delivery
- Present from birth to about 1 week of age as the PDA closes
- Cyanosis that does not improve with oxygen
- Many have no other obvious anomalies

## Characteristic Physical Findings

- General appearance -
- Respiratory rate and effort -
- Color -
- Murmur -
- Perfusion -

## Physical Findings
### General Appearance

- Is the newborn comfortable or in distress?
- Is the newborn active or lethargic?
- Does the newborn look:
  - AGA – are their other anomalies?
  - LGA – infant of a diabetic mother?
  - SGA – congenital infection, chromosomal problems?
- Is the newborn diaphoretic?

## Physical Findings
### Respiratory Rate and Effort

- Tachypnea but with minimal distress:
  - tissue hypoxia results in anaerobic metabolism producing lactic acid. This results in a metabolic acidosis. The body's compensation is to lower CO2 by increasing respiratory rate.
- In respiratory distress (tachypnea with grunting, flaring, retracting):
  - Inability of the respiratory system to compensate for the metabolic acidosis.
    - Concurrent respiratory disease
    - Unrelenting metabolic acidosis – decreased cardiac function
    - Exhaustion

## Physical Findings
### Color

- Cyanosis:
  - Degree – saturation usually < 85% to be seen 3-5 gm of unsaturated hemoglobin
  - Distribution – upper vs. lower central vs. peripheral
  - must have a right to left shunt
- Pallor:
  - seen in vasoconstriction from circulatory shock
- Jaundice – prolonged physiologic
  - Severe congestive heart failure
  - Hypothyroidism (PS)

## Physical Findings
### Murmur

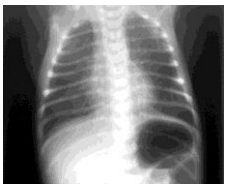
- S1 – usually normal in newborns
- S2 – closure of the pulmonary and aortic valves
  - Should be loud and single – pulmonary hypertension
  - Soft and single – suggest atretic aortic or pulmonary valve
- Murmur at birth – suggests valvular stenosis, PDA or severe regurgitation. VSDs are usually silent
- No murmur does not rule out heart disease

## Physical Findings

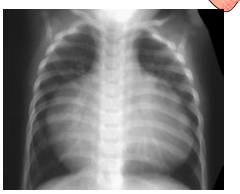
Perfusion

- Capillary filling time – should be 2-3 seconds
- Blood pressure – mean should be about gestational age
- Peripheral pulses – evaluate both upper and lower (femoral) pulses
  - Bounding pulses
    - PDA
    - AR
    - AV fistula
  - Weak or absent femoral pulses
    - CoA
  - The presence of femoral pulses at birth does not rule out a CoA

## Evaluating the CXR

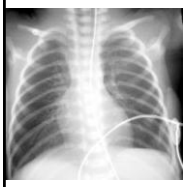


Normal heart size

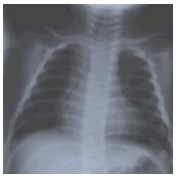


Cardiomegaly
> 60 % of the chest

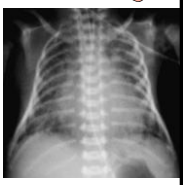
## Pulmonary Blood Flow



Normal vascular markings

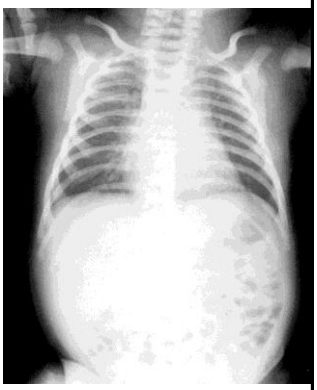


Decreased vascular markings

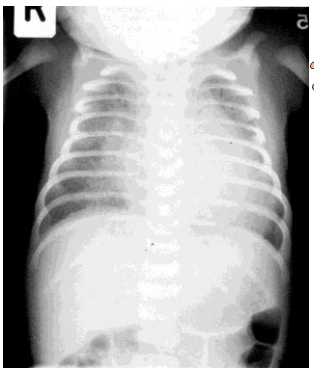


Increased vascular markings

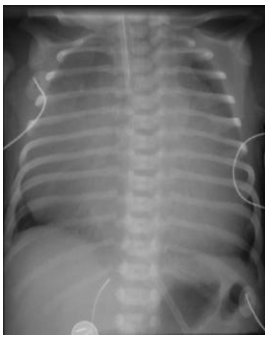
- Heart size:
- Lung perfusion:
- Diff. Diagnosis:



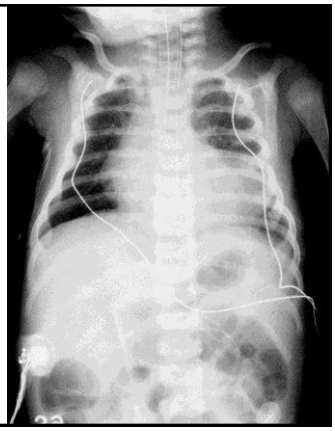
- Heart size:
- Lung perfusion:
- Diff. Diagnosis:



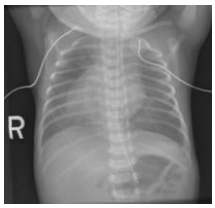
- Heart Size:
- Lung Perfusion:
- Diff. Diagnosis:



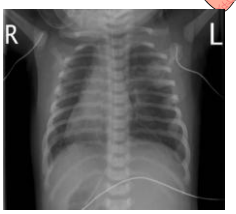
- Heart size:
- Lung perfusion:
- Diff. Diagnosis:



## Dextrocardia




Dextrocardia with situs solitus
(Incidence of CHD is 95-98%)




Dextrocardia with situs inversus
(Most common malposition- CHD unlikely)

## Blood Gases

| | Arterial | Capillary | Venous |
|---|---|---|---|
| pH | accurate | accurate | lower |
| PO2 | accurate | variable | lower |
| PCO2 | accurate | accurate | higher |
| HCO3(calculated) | accurate | accurate | accurate |

## Blood Gases

- In congenital heart disease typically:
  - compensated or partially compensated metabolic acidosis
  - Arterial PO2 usually low - < 50
  - Arterial PO2 < 30 suggests TGA
- If PCO2 is rising despite the metabolic acidosis, the newborn is going into respiratory failure – be ready to intubate!

## Stabilization of the Infant with Suspected CHD – General Approach

- ABC first:
- 100% oxygen to get sats >75%
- Support ventilation as needed
- Support perfusion with volume and inotropes
- Rule out other causes of cyanosis
- If sats still < 75% - start Prostaglandins

## Stabilization of the Infant with Suspected CHD

- Oxygen -
- Saturation monitoring -
- Fluids -
- Prostaglandin -
- Assisted ventilation -

## Oxygen

- Oxygen is a drug – use it with respect
- Use only enough oxygen to get the sats 75-85% - abolish anaerobic metabolism
- Oxygen (high PO2) is a pulmonary vasodilator
  - May worsen pulmonary congestion in HLHS
- Oxygen (high PO2) is a stimulus for the PDA to close
  - May worsen ductal dependent lesions by speeding up closure of the PDA

## Saturation Monitoring

- Oxygen saturation reflects tissue oxygenation and usually does not correlate with PO2 – because of fetal hemaglobin
- Concurrent monitoring of the upper and lower extremities may help to detect CoA.
- May be confused with pulmonary hypertension – also shunts right to left across the PDA.

## Fluids

- Needed if poorly perfused or evidence of shock
- NS, albumin boluses
- Glucose - watch for and treat hypoglycemia
  - stress causes epinephrine release which increases utilization of glucose
  - Anaerobic metabolism utilizes more glucose than aerobic metabolism to produce enough energy
- PRBC to treat anemia – optimize oxygen carrying capacity
- Other colloids (FFP, cryprecipitate) to treat other organ dysfunction (DIC)

## Prostaglandin

- Prostaglandin infusion:
  - Purpose is to open the PDA if a ductal dependent lesion is suspected
  - Can be initiated before a definitive diagnosis is established especially if the saturation does not increase to >75% despite oxygen, effective ventilation, fluids and ruling out other causes of cyanosis
  - Need a secure IV preferably a central line (UAC or UVC)

## Prostaglandin Continued

- Side effects –
  - Apnea – be prepared to intubate
  - Fever – should also be on antibiotics for possible sepsis
  - Hypotension – have volume and inotropes available
  - Flushing
  - Seizures
  - Hypocalcemia, platelet dysfunction

## Assisted Ventilation

- Intubate if:
  - Impending respiratory failure
  - High suspicion of HLHS – goal is to **underventilate** to create an acidosis that will increase PVR and force blood from the lungs to the body
  - Consider if initiating prostaglandin

## Special Transport Issues

- If on prostaglandin, may need to intubate prior to leaving the referring hospital
- Have a UVC or a least 2 PIVs in place for the prostaglandin infusion.
- Limit the use of oxygen to “as much as is necessary” to prevent a metabolic acidosis. Saturation > 75% in a infant **without distress** is acceptable.

## Special Transport Issues

- Take your time - do not scoop and run. Make sure you have a secure airway and IV access before ‘getting on the road’
- It is better to start prostaglandin if unsure than have the PDA close on transport
- Do not be shy about asking for help – communicate with your base hospital frequently