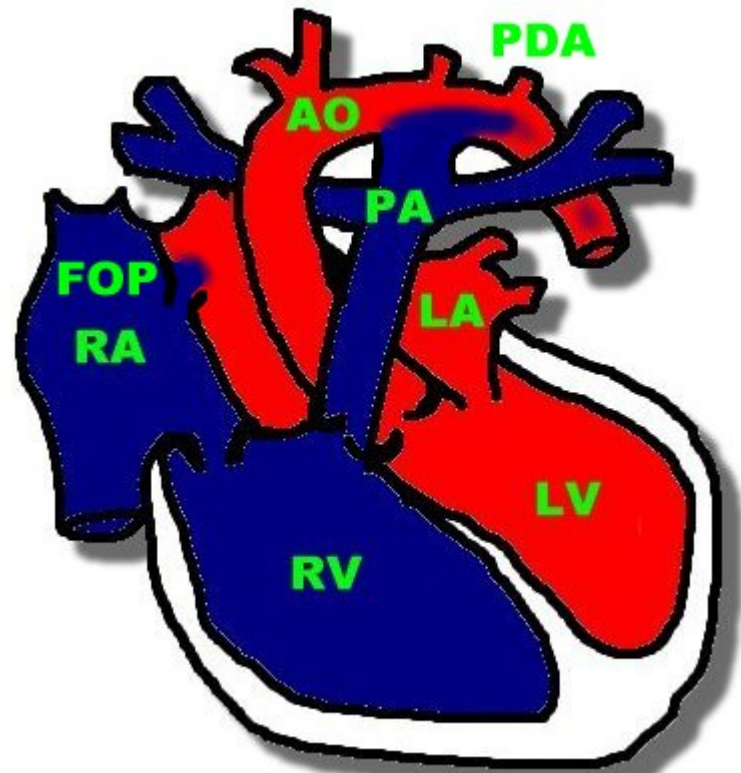
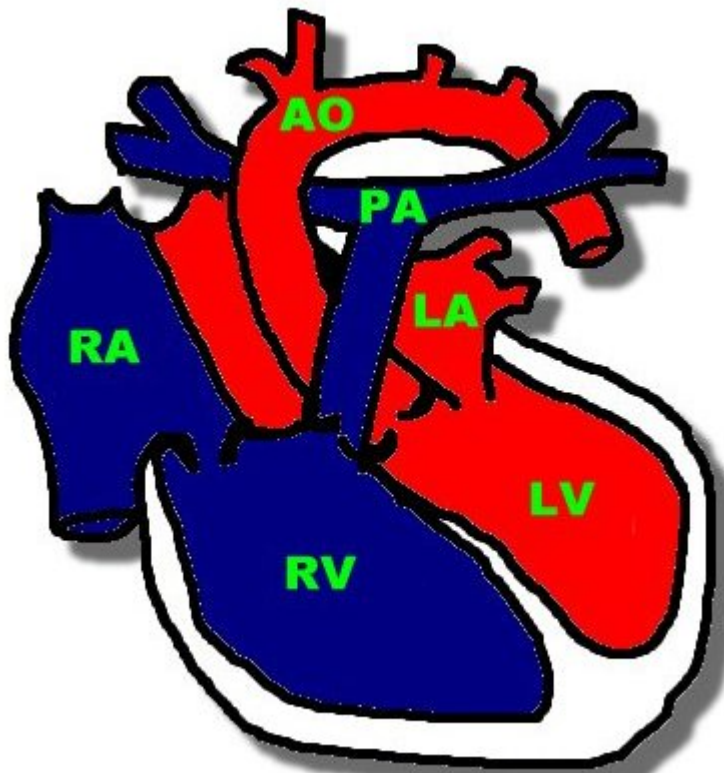


# Persistent Pulmonary Hypertension of the Newborn





# Nomenclature



- ⌘ Persistent pulmonary vascular obstruction
- ⌘ Persistent fetal circulation
- ⌘ Pulmonary vasospasm
- ⌘ Neonatal pulmonary ischemia
- ⌘ Persistent transitional circulation

# PPHN



- ⌘ Also known as Persistent fetal circulation.
- ⌘ PPHN is the failure of PVR to fall at birth.
- ⌘ The transition from fetal circulation to extra uterine circulation is not complete.
- ⌘ R-L shunting occurs through a patent ductus arteriosus and foramen ovale.

# Typically seen in:



- ⌘ Full term or post term infants
- ⌘ **Pulmonary parenchymal disease**, such as hyaline membrane disease
- ⌘ 37-41 weeks gestational age
- ⌘ within the first 12-24 hours after birth.

# Primary PPHN

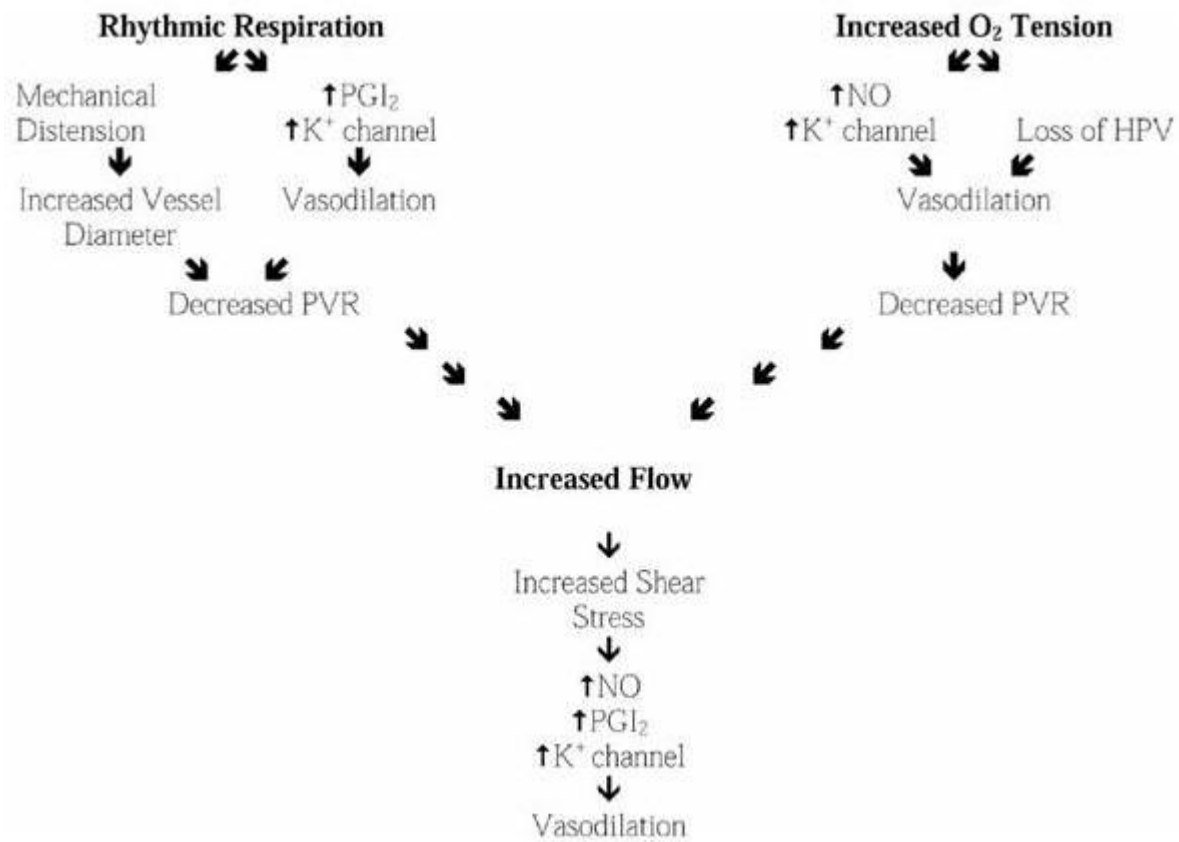


## ⌘ Classical PPHN

☑ idiopathic

⌘ Hypoxemia develops in a baby with normal lungs.

⌘ Breath sounds and CXR are usually normal.



# Possible causes



- ⌘ chronic intrauterine hypoxia
- ⌘ asphyxia
- ⌘ maternal ingestion of prostaglandin
  - ⌘ premature ductal closure
- ⌘ hypoglycemia
- ⌘ hypothermia
- ⌘ maternal hypertension
- ⌘ hyaline membrane disease

# Prostaglandin ingestion



- ⌘ Mothers who took aspirin near term caused repeated intrauterine closure of the ductus with redirection of blood into the pulmonary vasculature.

# Secondary PPHN



⌘ PPHN secondary to lung disease.

- ☑ meconium aspiration syndrome
- ☑ congenital diaphragmatic hernia
- ☑ group B streptococcal pneumonia
- ☑ respiratory distress syndrome
- ☑ sepsis
- ☑ Hypoplasia
- ☑ Hyaline membrane disease

# In Utero



- ⌘ Fetal gas exchange occurs through the placenta instead of the lungs.
- ⌘  $PVR > SVR$  causes blood from the right side of the heart to bypass the lungs through the ductus arteriosus and foramen ovale.

# Fetal Shunts



## ⌘ Ductus arteriosus

- ☑ R-L shunting of blood from pulmonary artery to the aorta bypasses the lungs.
- ☑ Usually begins to close 24-36 hours after birth.

## ⌘ Foramen ovale

- ☑ Opening between left and right atria.
- ☑ Closes when there is an increased volume of blood in the left atrium.

# At Birth



## ⌘ First breath

- ☑ Decrease in PVR

- ☑ Increase in pulmonary blood flow and PaO<sub>2</sub>

## ⌘ Circulatory pressures change with the clamping of the cord.

- ☑ SVR > PVR allowing lungs to take over gas exchange.

- ☑ If PVR remains higher blood continues to be shunted and PPHN develops.

# Signs of PPHN



- ⌘ Infants with PPHN are born with Apgar scores of 5 or less at 1 and 5 minutes.
- ⌘ Cyanosis may be present at birth or progressively worsen within the first 12-24 hours.

# Later developments



## ⌘ Within a few hours after birth

- ☑ tachypnea
- ☑ retractions
- ☑ systolic murmur
- ☑ mixed acidosis, hypoxemia, hypercapnia

## ⌘ CXR

- ☑ mild to moderate cardiomegaly
- ☑ decreased pulmonary vasculature

# Pulmonary Vasculature



- ⌘ Pulmonary vascular bed of newborn is extremely sensitive to changes in  $O_2$  and  $CO_2$ .
- ⌘ Pulmonary arteries appear thick walled and fail to relax normally when exposed to vasodilators.
- ⌘ Capillaries begin to build protective muscle. (remodeling)

# Diagnosis



## ⌘ Hyperoxia Test

⌘ Place infant on 100% oxyhood for 10 minutes.

☒  $\text{PaO}_2 > 100 \text{ mmHg}$  parenchymal lung disease

☒  $\text{PaO}_2 = 50\text{-}100 \text{ mmHg}$  parenchymal lung disease or cardiovascular disease

☒  $\text{PaO}_2 < 50 \text{ mmHg}$  fixed R-L shunt  
cyanotic congenital heart disease or PPHN

# Hyperoxia Test (cont.)

## ⌘ If fixed R-L shunt

- ☑ need to get a preductal and postductal arterial blood gases with infant on 100% O<sub>2</sub>.
  - ☒ Preductal- R radial or temporal artery
  - ☒ Postductal- umbilical artery
- ☑ If > 15 mmHg difference in PaO<sub>2</sub> then ductal shunting
- ☑ If < 15 mmHg difference in PaO<sub>2</sub> then no ductal shunting

# Hyperoxia-Hyperventilation Test

⌘ Hyperinflate baby with manual resuscitator and 100% O<sub>2</sub> until PaCO<sub>2</sub> reaches 20-25 mmHg.

☑ PaO<sub>2</sub> = 100 mmHg with hyperinflation

☒ PPHN

☑ PaO<sub>2</sub> < 100 mmHg with hyperinflation

☒ R/O congenital heart disease with echocardiogram.

- abnormal Echo = congenital heart disease
- normal Echo = PPHN

# Echocardiography



- ⌘ R ventricle may be larger than normal.
- ⌘ Ratio of pre-ejection period (PEP) to ejection time (ET) is used to evaluate left and right ventricle performance.
  - ⊡ PPHN causes a prolonged R ventricle PEP/ET ratio
    - ⊗ increased pulmonary artery pressure
    - ⊗ increased pulmonary vascular resistance

# Echo (cont.)



- ⌘ PPHN can be identified early if R and L ventricular PEP/ET ratios are measured soon after birth.
- ⌘ Babies with R ventricular ratio  $> .50$  and L ventricular ratio  $> .38$  developed PPHN within 10-30 hours after birth.

# Cardiac Catheterization



- ⌘ In past, cardiac catheterization was used to diagnose infants with PPHN by monitoring pulmonary artery pressures.
- ⌘ Today this is not recommended because it is traumatic to the baby and it is no longer needed to make a diagnosis.

# Treatment



## ⌘ Goals:

- ☑ To maintain adequate oxygenation.
  - ☒ These babies are extremely sensitive
  - ☒ Handling them can cause a decrease in PaO<sub>2</sub> and hypoxia
  - ☒ Crying also causes a decrease in PaO<sub>2</sub>
  - ☒ Try to coordinate care as much as possible
- ☑ To maintain neutral thermal environment to minimize oxygen consumption.

# Therapeutic Goals



- ⌘ Arterial blood gas values
- ⌘ pH 7.45 - 7.55
- ⌘ PaO<sub>2</sub> 50-100 torr
- ⌘ PaCO<sub>2</sub> 25-40 torr
- ⌘ Systemic blood pressure
- ⌘ Upper limits of normal for size and postconceptual age  
{see [Blood Pressure Chart](#)}
- ⌘ Adequate paralysis and sedation
- ⌘ Ensure adequate oxygen carrying capacity. Maintain hematocrit greater than 40%

# Medication



⌘ Tolazine (Priscoline)- pulmonary and systemic vasodilator

☑ pulmonary response needs to be assessed by giving 1-2 mg/kg through peripheral scalp vein

☑ if positive response- start continuous infusion of 0.5-1.0 mg/kg/hr

# Tolazine (cont.)



- ⌘ Monitor closely for GI bleeding, pulmonary hemorrhage and systemic hypotension.
- ⌘ May need to also give Dopamine or Dobutamine to maintain systemic blood pressure and to increase CO.

# Mechanical Ventilation



- ⌘ TCPLV (Time cycled pressure limited ventilation) may be used with PPHN.
- ⌘ Want to use low peak inspiratory pressures
- ⌘ Monitor PaO<sub>2</sub> and PaCO<sub>2</sub> with a transcutaneous monitor

# Hyperventilation



- ⌘ Hyperventilation helps promote pulmonary vasodilation
- ⌘ Respiratory Alkalosis- decrease PAP to level below systemic pressures to improve oxygenation by helping to close the shunts
  - ☑ Try to keep pH = 7.5 and PaCO<sub>2</sub> = 25-30
  - ☑ Alkalinizing agents - sodium bicarbonate or THAM

# Hyperventilation (cont.)

- ⌘ Babies often become agitated when they are hyperventilated
- ⌘ May need to administer muscle relaxants and sedation
  - ⊡ usually given pancuronium and morphine
    - ⊗ pancuronium- q 1-3 hours IV at 0.1-0.2 mg/kg
    - ⊗ morphine- continuous infusion 10 micrograms/kg/hr

# HFOV



## ⌘ High frequency oscillatory ventilation

- ☑ decrease risk of barotrauma

- ☑ effective alveolar ventilation

- ☑ alveolar recruitment

  - ☒ Nitric Oxide more effective

## ⌘ HFOV more effective in PPHN babies with lung disease

# Nitric Oxide (NO)



- ⌘ Potent pulmonary vasodilator
  - ☑ decrease pulmonary artery pressure
  - ☑ increase PaO<sub>2</sub>
- ⌘ Does not cause systemic hypotension
- ⌘ NO more effective in PPHN babies without lung disease
- ⌘ Baby must be weaned slowly off NO or may have rebound hypertension

# Effects of NO




- ⌘ NO is metabolized to nitrogen dioxide ( $\text{NO}_2$ ) which can cause acute lung injury.
- ⌘  $\text{NO}_2$  is potentially toxic.
- ⌘ NO reacts with hemoglobin to form methemoglobin.

# ECMO



- ⌘ Extra corporeal membrane oxygenation
- ⌘ Form of cardiorespiratory support that allows the lungs to rest so also called extracorporeal life support (ECLS).
- ⌘ ECMO has increased survival rate significantly

# ECMO (cont.)



⌘ ECMO is given as a last resort when everything else has failed.

## ⌘ Requirements

- ☑ > 33 weeks gestational age
- ☑ potentially reversible lung disease
- ☑ no bleeding disorders
- ☑ no intraventricular hemorrhages

# Two Routes



## ⌘ Venovenous route

☑ blood taken from R jugular vein and returned to the venous system.

## ⌘ Venoarterial route

☑ blood taken from R jugular vein and returned through R carotid artery

⌘ Gas exchange takes place as the blood is pumped through a membrane oxygenator

# Outcome



- ⌘ PPHN may last anywhere from a few days to several weeks.
- ⌘ Mortality rate is 20-50%.
  - ☑ Decreased by HFOV and NO
  - ☑ Decreased by ECMO
- ⌘ Babies treated with hyperventilation may develop sensorineural hearing loss.

# Questions About PPHN



1. Define PPHN
2. List early and late signs and symptoms of PPHN.
3. What are the treatment goals when dealing with PPHN?
4. How is NO useful?
5. What are the requirements for ECMO?
6. What is the mortality rate of PPHN