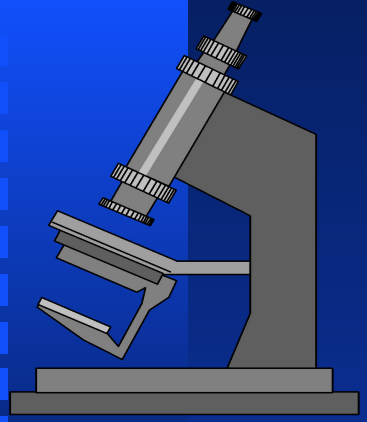
CONGENITAL AND ACQUIRED RESPIRATORY DISORDERS IN INFANTS

OBJECTIVES

- Review of Cardio-Pulmonary Development.
- Define changes that occur during transition to extra-uterine life with emphasis on breathing mechanics.
- Identify infants at risk for and who have respiratory distress
- Review of common neonatal disease states.



STAGES OF NORMAL LUNG GROWTH

Embryonic - first 5 weeks; formation of proximal airways

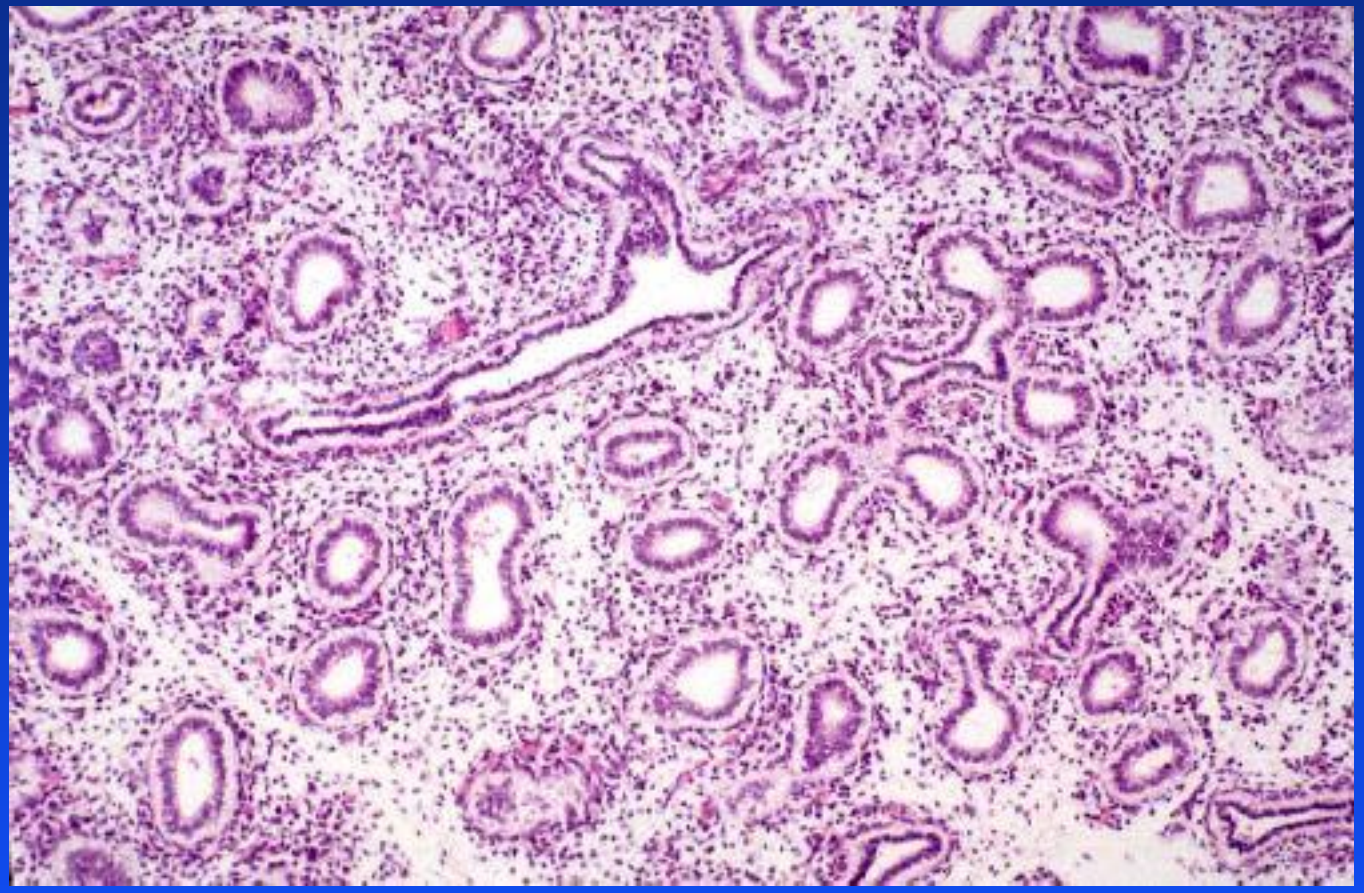
Pseudoglandular - 5-16 weeks; formation of conducting airways

Canalicular - 16-24 weeks; formation of acini

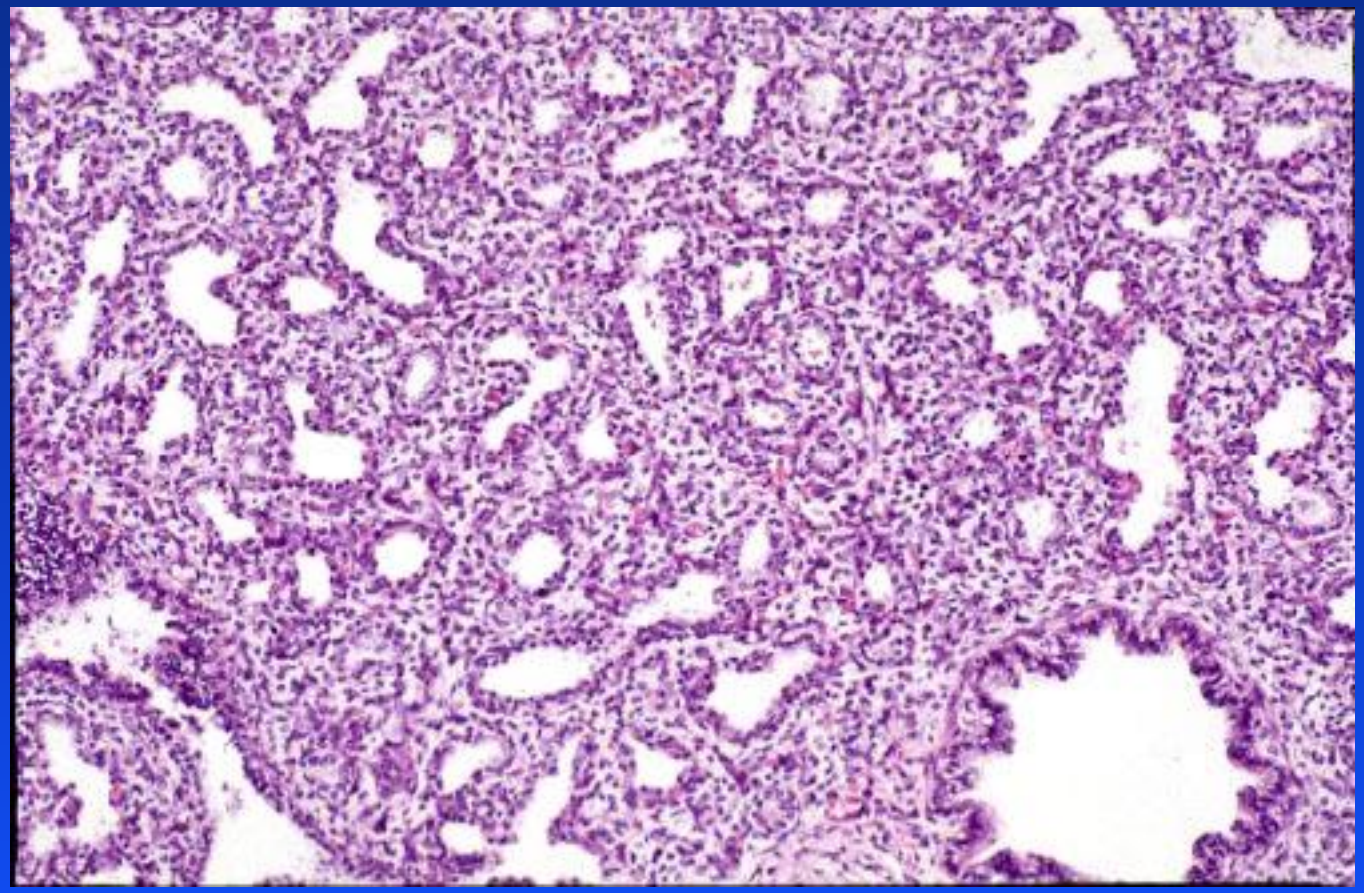
Saccular - 24 - 36 weeks; development of gas-exchange units

Alveolar - 36 weeks and up; expansion of surface area

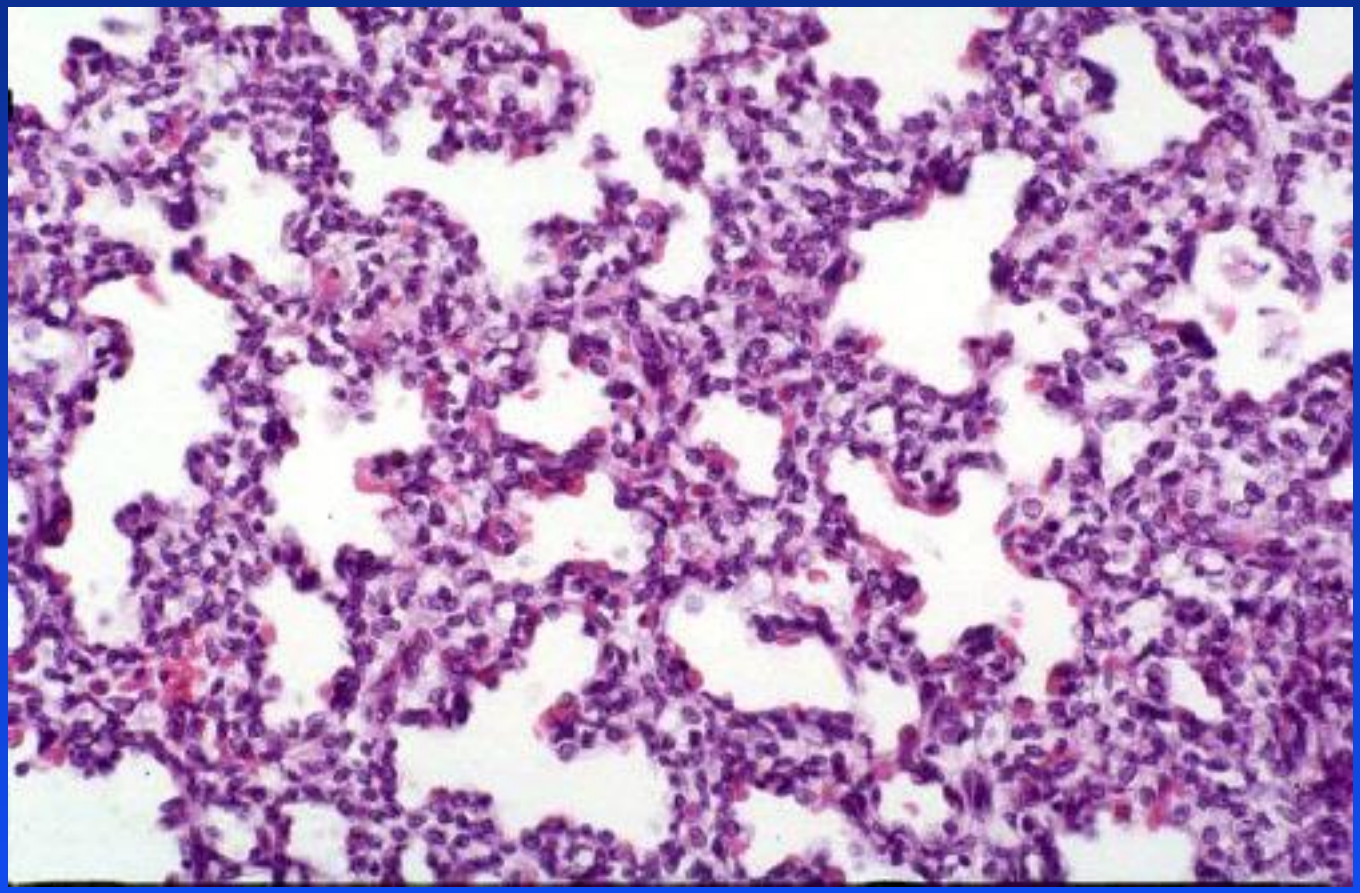
Pseudoglandular
6-16 weeks



Canalicular Phase
16-24 weeks



Saccular Phase
24-34 weeks



PHYSIOLOGIC MATURATION (Surfactant Production)

- Type 2 pneumocytes appear at 24-26 weeks
- Responsible for reduction of alveolar surface tension.
  - LaPlace's Law
- Lipid profile as indicator of lung maturity
  - L/S Ratio
  - Flourescence Polarization - FLM
- Many other factors influence lung maturation

Tests for Determining Fetal Lung Maturity[†]

Test	Positive discriminating value	Predictive value	Predictive value for prediction of pulmonary immaturity	Relative cost	Pro's and con's
L/S ratio	>2.0	95-100 percent	33-50 percent	High	Large laboratory variation
PG	"Present"	95-100 percent	23-53 percent	High	Not affected by blood, meconium. Can use vaginal pooled sample.
FSI	>47	95 percent	51 percent	Low	Affected by blood, meconium, silicon tubes.
FLM-TDx	>55	96-100 percent	47-61 percent	Moderate	Minimal inter/ intraassay variability. Simple test.
Optical density at 650 nm	OD.0.15	98 percent	13 percent	Low	Simple technique
Lamellar	30-40000	97-98 percent	29-35 percent	Low	Still investigational

L/S ratio = Lecithin sphingomyelin ratio; PG = Phosphatidylglycerol; FSI = Foam stability index; OD = Optical density.
[†] Adapted from data in the American College of Obstetricians and Gynecologists, ACOG educational bulletin #230, Washington, DC 1996.

Probability of RDS on the Basis of Gestational Age and FLM[†]

	Gestational age, weeks													
FLM	**27**	**28**	**29**	**30**	**31**	**32**	**33**	**34**	**35**	**36**	**37**	**38**	**39**	**40**
0	72	66	59	51	44	37	30	24	19	15	12	9	7	5.1
10	67	60	53	46	39	32	26	20	16	12	9.6	7.3	5.5	4.2
20	62	55	48	40	33	27	22	17	13	10	7.8	6	4.5	3.4
30	57	50	42	35	29	23	18	14	11	8.4	6.4	4.8	3.6	2.7
40	51	44	37	30	24	19	15	12	9	6.8	5.2	4	3	2.2
50	46	39	32	26	21	16	13	10	7.4	5.6	4.2	3.2	2.4	1.8
60	40	34	27	22	17	13	10	8	6	4.5	3.4	2.5	1.9	1.4
70	35	29	23	18	14	11	8.5	6.4	4.9	3.7	2.7	2	1.5	1.1
80	31	25	20	15	12	9.1	7	5.2	4	3	2.2	1.7	1.2	0.9
90	26	21	16	13	10	7.4	5.6	4.2	3.2	2.4	1.8	1.3	1	0.7
100	22	17	14	10	8	6	4.6	3.4	2.6	2	1.4	1	0.8	0.6
110	19	14	11	9	6.5	4.9	3.7	2.8	2.1	1.5	1.2	0.9	0.6	0.5
120	15	12	9	7	5.3	4	3	2.2	1.7	1.2	1	0.7	0.5	0.4
130	13	9.8	7.5	6	4.3	3.2	2.4	1.8	1.3	1	0.7	0.6	0.4	0.3
140	10	8	6.1	4.6	3.5	2.6	2	1.4	1.1	0.8	0.6	0.5	0.3	0.25
150	9	6.6	5	3.7	2.8	2.1	1.6	1.2	0.9	0.6	0.5	0.4	0.3	0.2
160	7	5.3	4	3	2.3	1.7	1.3	1	0.7	0.5	0.4	0.3	0.2	0.2
170	5.7	4.3	3.2	2.4	1.8	1.4	1	0.8	0.6	0.4	0.3	0.2	0.2	0.1
180	4.7	3.5	2.6	2	1.5	1.1	0.8	0.6	0.4	0.3	0.2	0.2	0.2	0.1
190	3.8	2.8	2.1	1.6	1.2	0.9	0.7	0.5	0.4	0.3	0.2	0.1	0.1	0.1
200	3	2.3	1.7	1.3	0.9	0.7	0.5	0.4	0.3	0.2	0.1	0.1	0.1	0.1

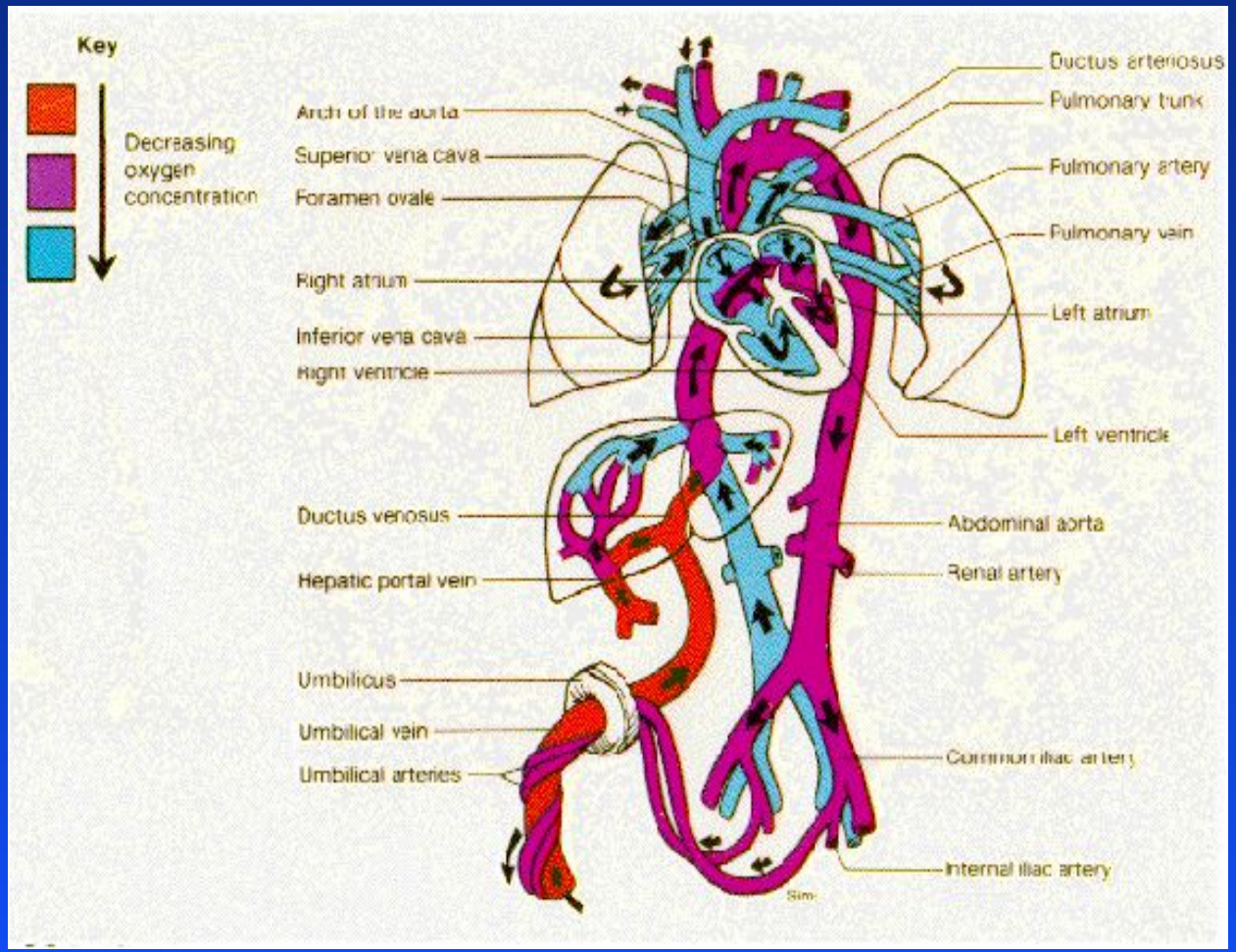
[†] Reproduced with permission from: Pinette, MG, Blackstone, J, Wax, JR, Cartin, A. Fetal lung maturity indices – A plea for gestational age – specific interpretation: A case report and discussion. Am J Obstet Gynecol. 2002; 187:1721. Copyright © 2002 Elsevier Science.

RDS = respiratory distress syndrome
FLM = TDx-FLM , fluorescence polarization test, result given as percent

Maturational Factors

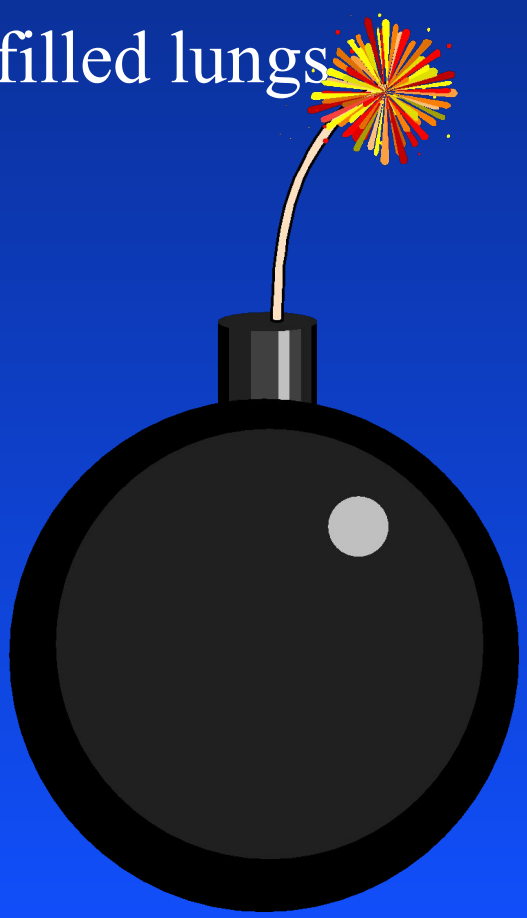
- Stimulation
  - Glucorticoids, ACTH
  - Thyroid Hormones, TRF
  - EGF
  - Heroin
  - Aminophyline,cAMP
  - Interferon
  - Estrogens
- Inhibition
  - Diabetes (insulin, hyperglycemia, butyric acid)
  - Testosterone
  - TGF-B
  - Barbiturates
  - Prolactin

FETAL CIRCULATION



TRANSITION TO EXTRA-UTERINE LIFE

- Fetal Breathing
- Instantaneous; liquid filled to air filled lungs
- Maintenance of FRC
- Placental blood flow termination
- Decreased PVR
- Closure of fetal shunts



MECHANICS OF BREATHING

- Respiratory Control Center...CNS
  - Metabolic Needs
- Negative pressure breathing
- Compliance and Resistance
  - Inspiratory Muscles
  - Rib Cage
    - “Compliability becomes a liability”

Signs of Respiratory Distress

- **Tachypnea**
- **Intercostal retractions**
- **Nasal Flaring**
- **Grunting**
- **Cyanosis**

When is it abnormal to show signs of respiratory distress?

- **When tachypnea, retractions, flaring, or grunting persist beyond one hour after birth.**
- **When there is worsening tachypnea, retractions, flaring or grunting at any time.**
- **Any time there is cyanosis**

Causes of Neonatal Respiratory Distress

- **Obstructive/restrictive - mucous, choanal atresia, pneumothorax, diaphragmatic hernia.**
- **Primary lung problem - Respiratory Distress Syndrome (RDS), meconium aspiration, bacterial pneumonia, transient (TTN).**
- **Non-pulmonary**
  -hypovolemia/hypotension, congenital heart disease, hypoxia, acidosis, cold stress, anemia, polycythemia

Infants at Risk for Developing Respiratory Distress

- **Preterm Infants**
- **Infants with birth asphyxia**
- **Infants of Diabetic Mothers**
- **Infants born by Cesarean Section**
- **Infants born to mothers with fever, Prolonged ROM, foul-smelling amniotic fluid.**
- **Meconium in amniotic fluid.**
- **Other problems**

Evaluation of Respiratory Distress

- **Administer Oxygen and other necessary emergency treatment**
- **Vital sign assessment**
- **Determine cause-- physical exam, Chest x-ray, ABG, Screening tests: Hematocrit, blood glucose, CBC**
- **Sepsis work-up**

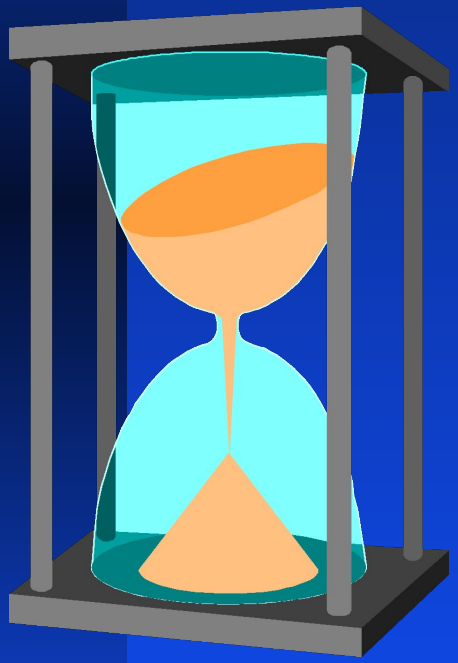
Principles of Therapy

- **Improve oxygen delivery to lungs-- supplemental oxygen, CPAP, assisted ventilation, surfactant**
- **Improve blood flow to lungs-- volume expanders, blood transfusion, partial exchange transfusion for high hematocrit, correct acidosis (metabolic/respiratory)**
- **Minimize oxygen consumption-- neutral thermal environment, warming/humidifying oxygen, withhold oral feedings, minimal handling**

DISEASE STATES

- Respiratory Distress Syndrome
- Transient Tachypnea of the Newborn
- Meconium Aspiration Syndrome
- Persistent Hypertension of the Newborn
- Congenital Pneumonia
- Congenital Malformations
- Acquired Processes

RESPIRATORY DISTRESS SYNDROME



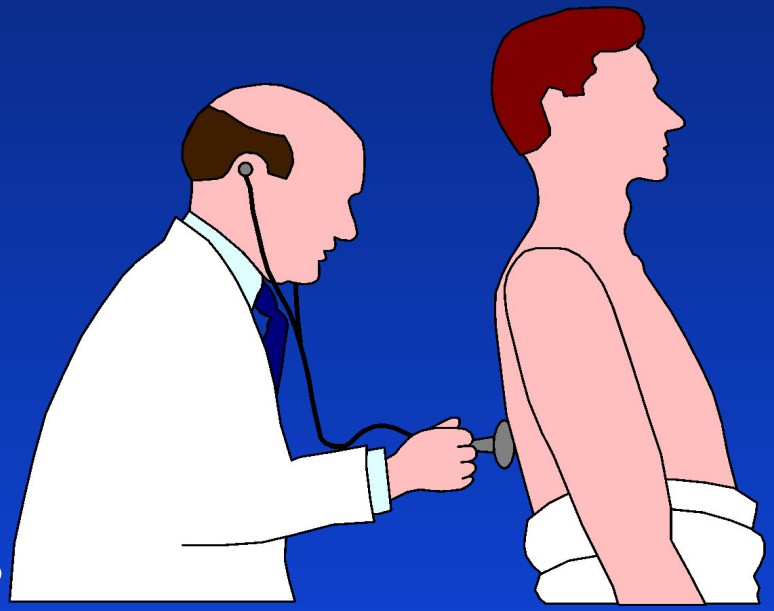
Surfactant Deficiency

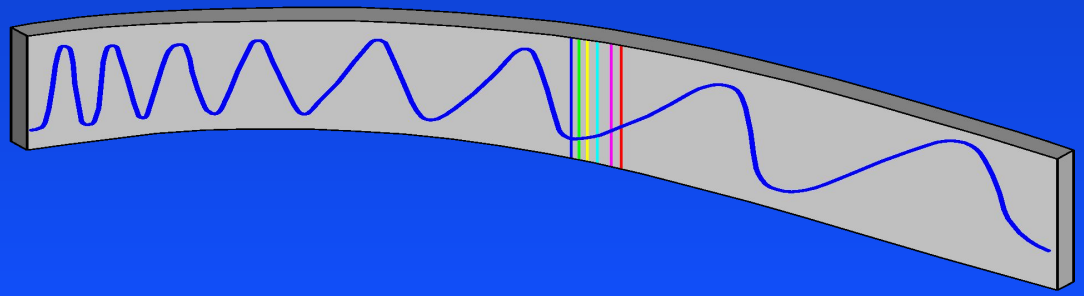
Tidal Volume Ventilation

Pulmonary Injury Sequence

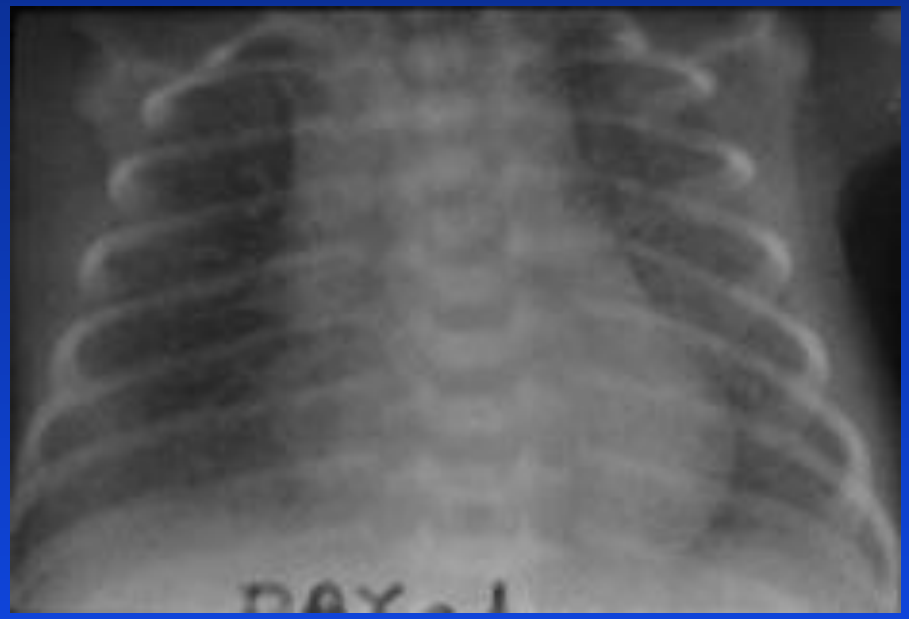
CLINICAL FEATURES OF RDS

- Tachypnea/Apnea
- Dyspnea
- Grunting/Flaring
- Hypoxemia
- Radiographic Features
- Pulmonary Function Abnormalities



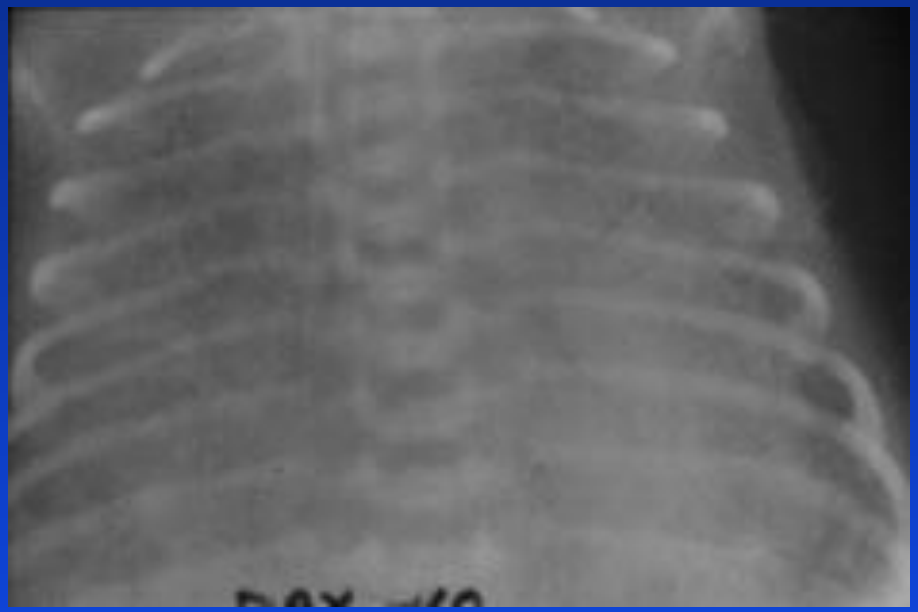


Early RDS

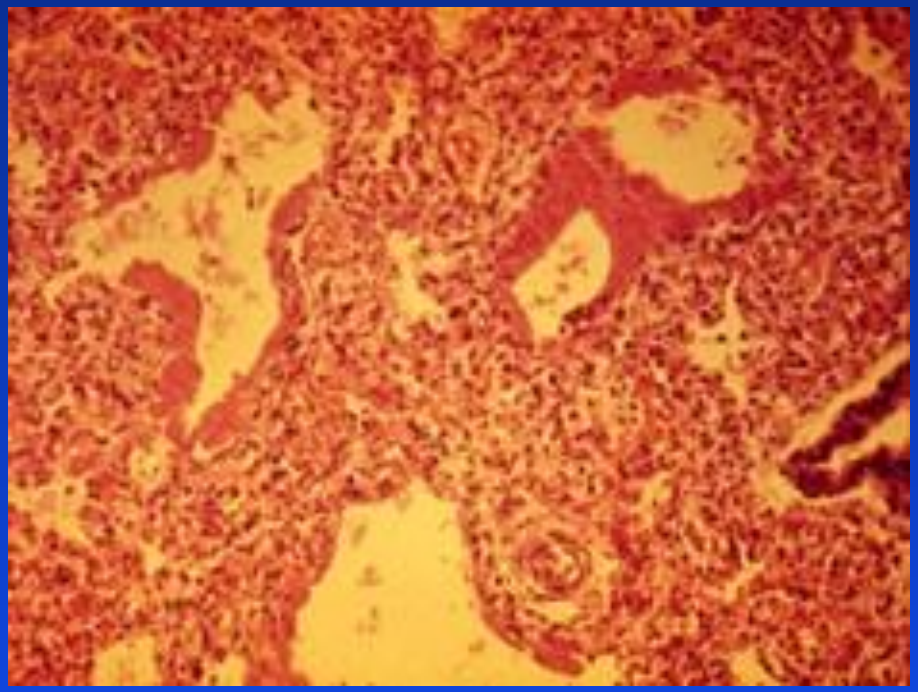


Progressive RDS

Late RDS



Hyaline Membrane Disease



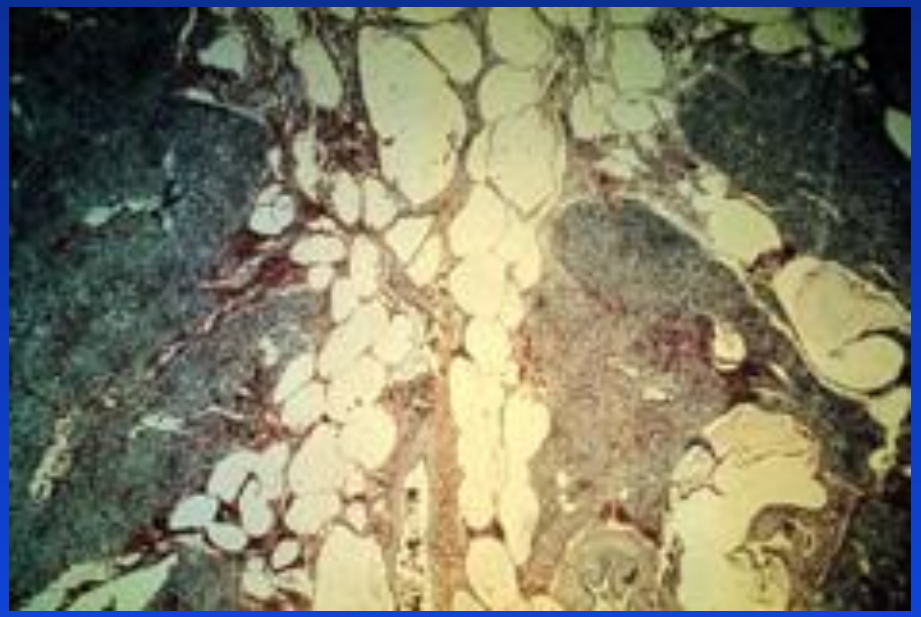
THERAPY FOR RDS

- Oxygen - maintain PaO2 > 50 torr
- Nasal CPAP
- Intermittent Mandatory Ventilation
- Surfactant Replacement
- High Frequency Ventilation
- Intercurrent Therapies

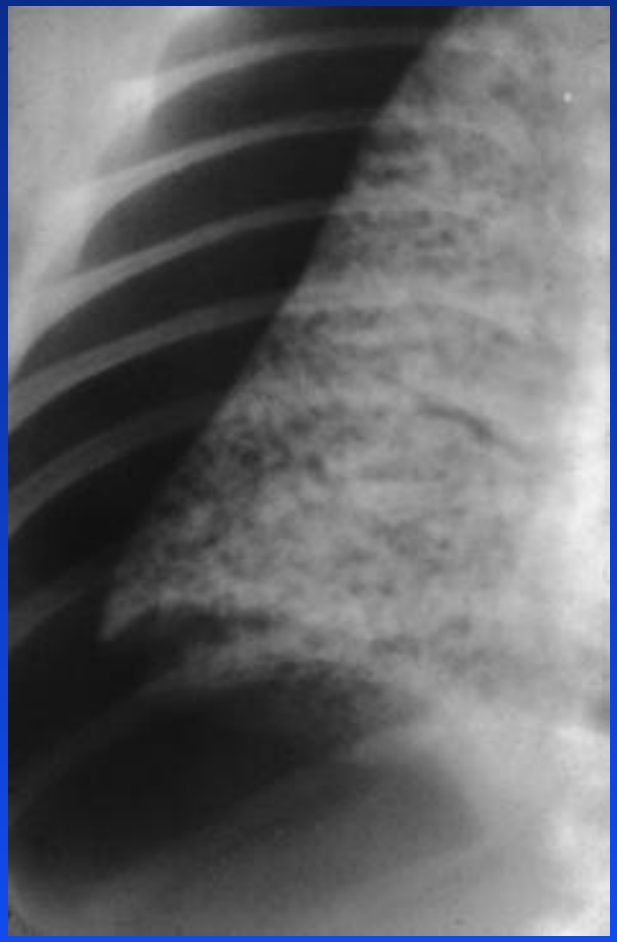
PIE

PIE Pathology

PIE Histology

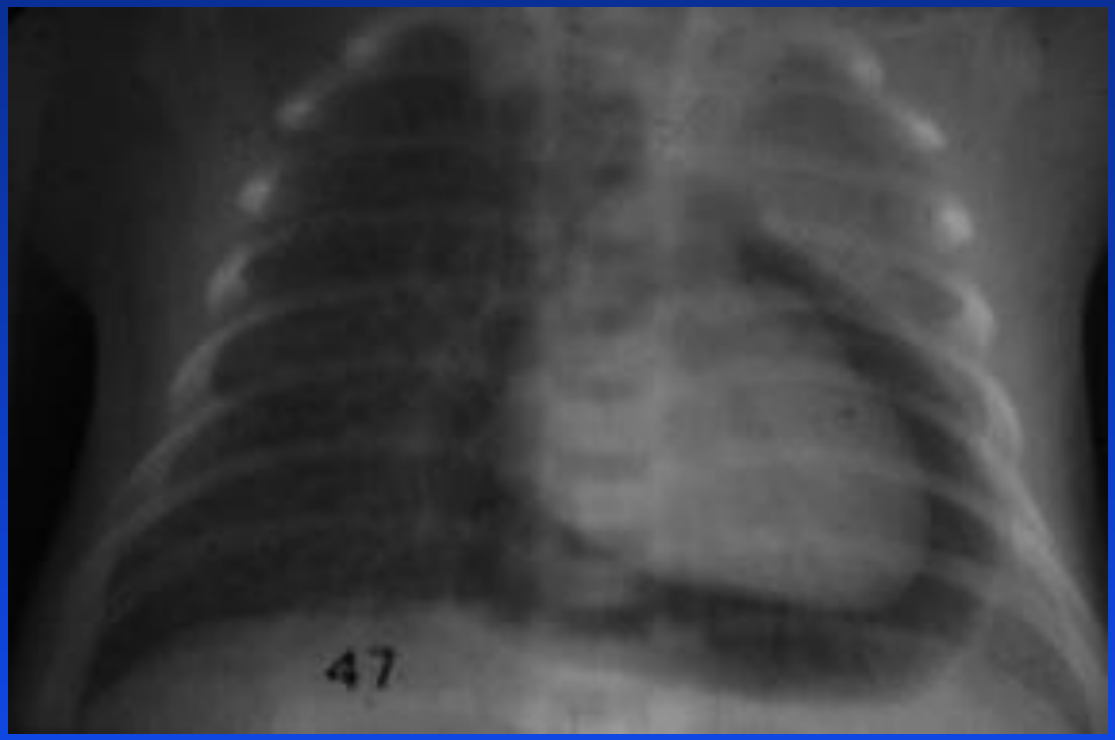


Pneumothorax/PIE

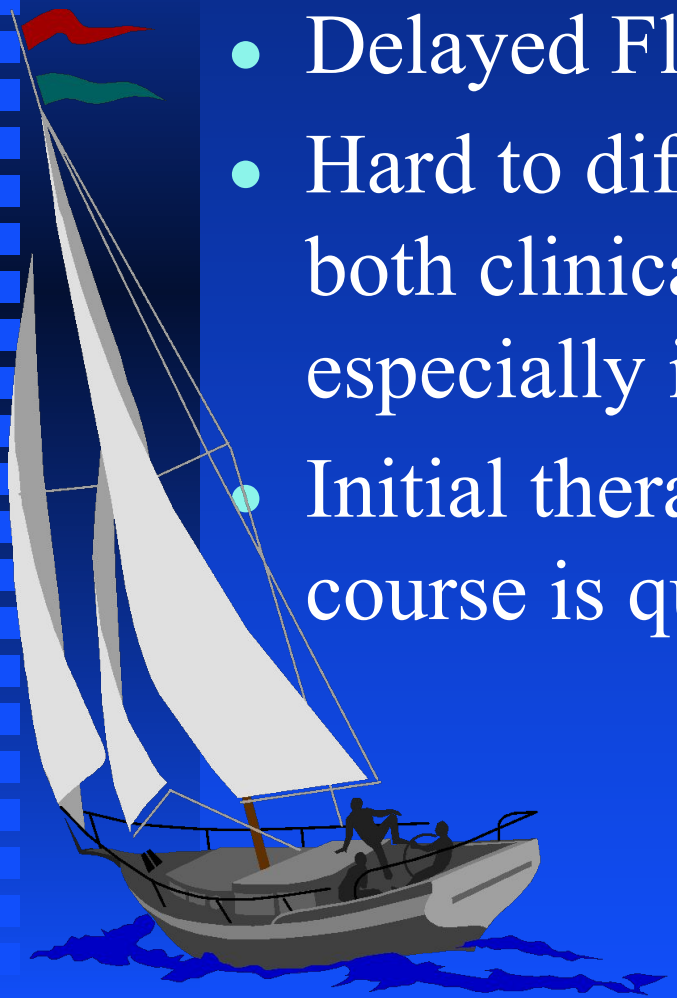


Pneumothorax

Pneumopericardium

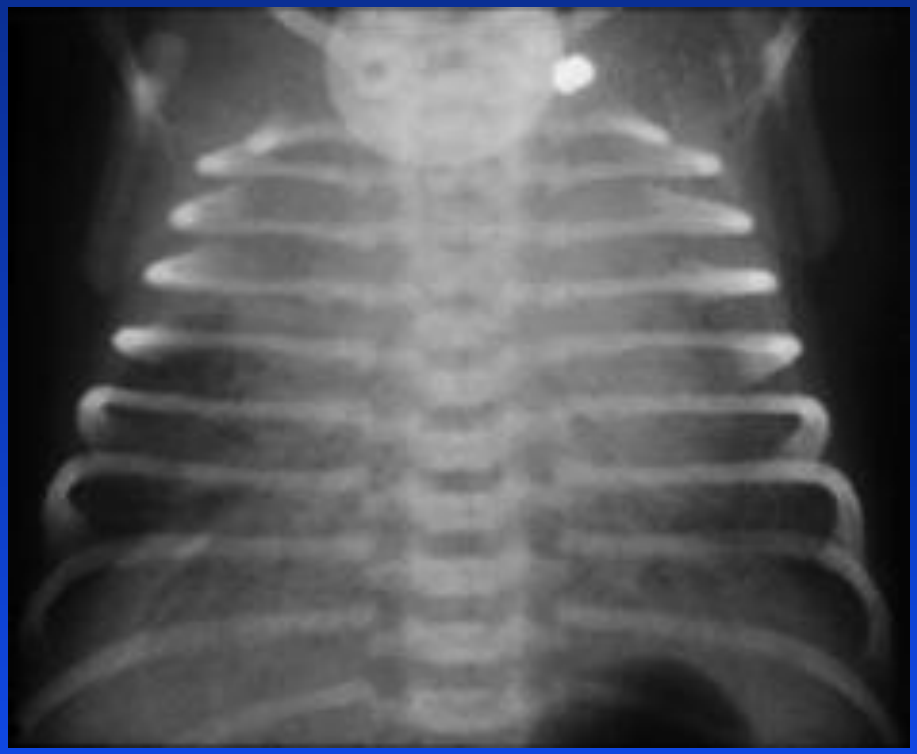


TRANSIENT TACHYPNEA OF THE NEWBORN



- Delayed Fluid Resorption
- Hard to differentiate early on from RDS both clinicaly and radiographicaly especially in the premature infant
- Initial therapy similar to RDS, but hospital course is quite different

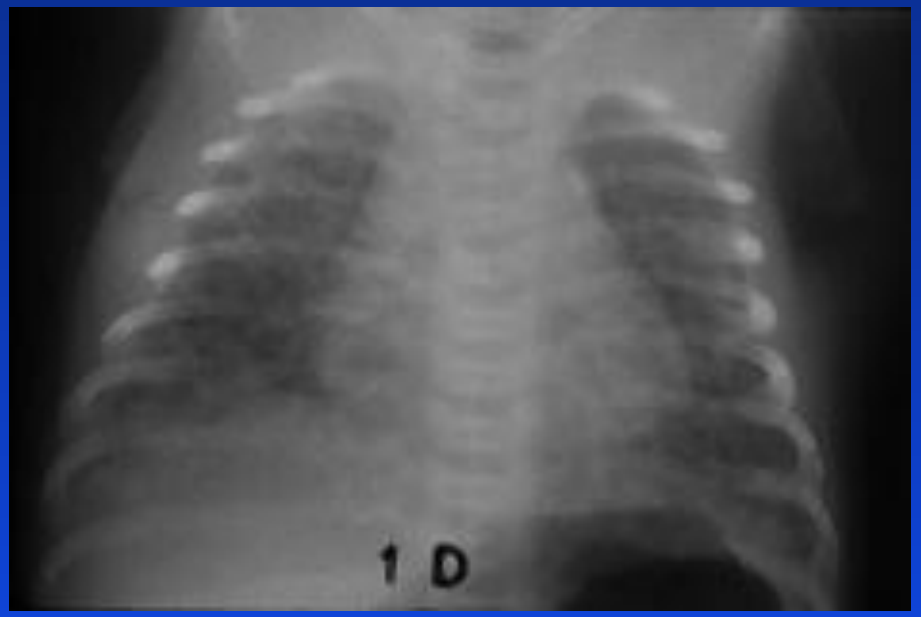
Wet Lung



MECONIUM ASPIRATION SYNDROME

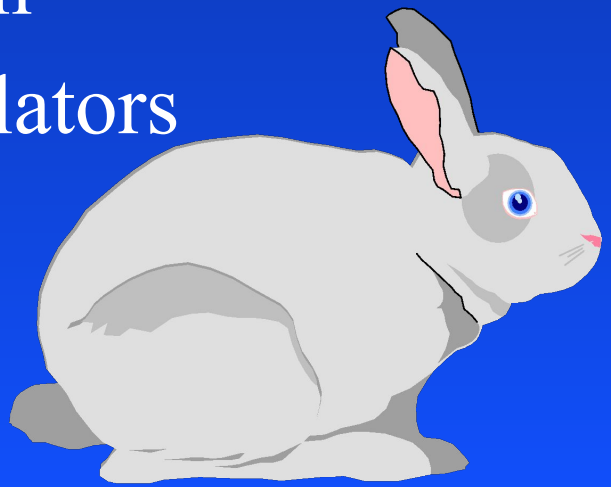
- Chemical Pneumonitis
- Surfactant Inactivation
- Potential for Infection
- Potential for Pulmonary Hypertension
- Management varies on severity

Meconium Aspiration

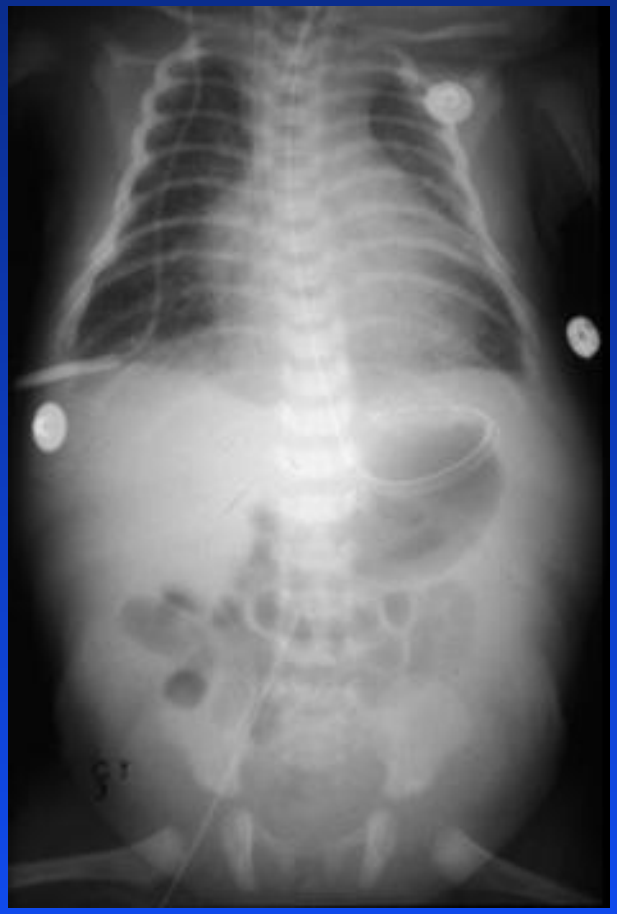


PERSISTENT PULMONARY HYPERTENSION

- Usually secondary to primary pulmonary disease state
- Pulmonary Vascular Lability
- Treat the underlying problem
- Maintain normo-oxygenation
- Selective Pulmonary Vasodilators
- Pray for good luck

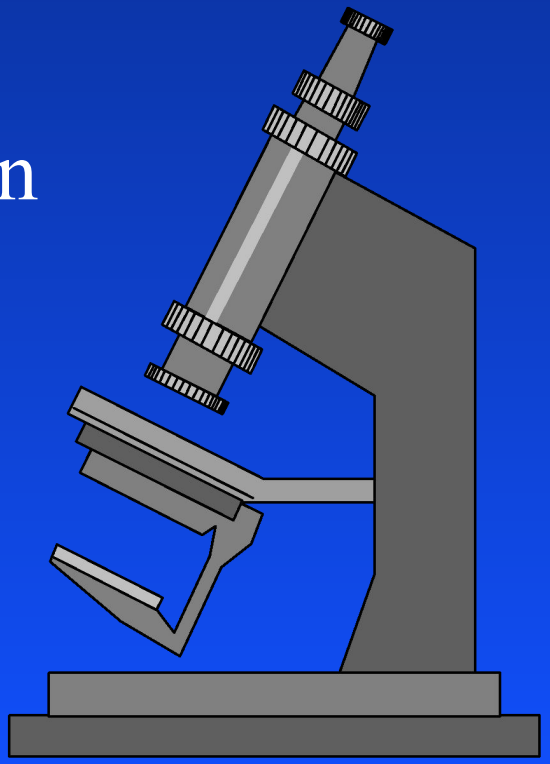


PPHN



CONGENITAL PNEUMONIA

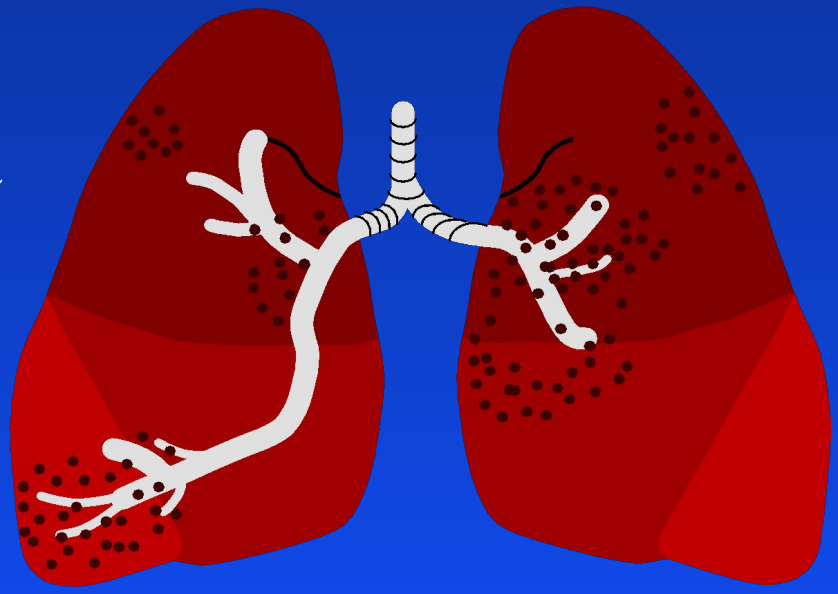
- Infectious; primarily GBS
- Amniotic Fluid aspiration
- Viral etiology
- Surfactant inactivation



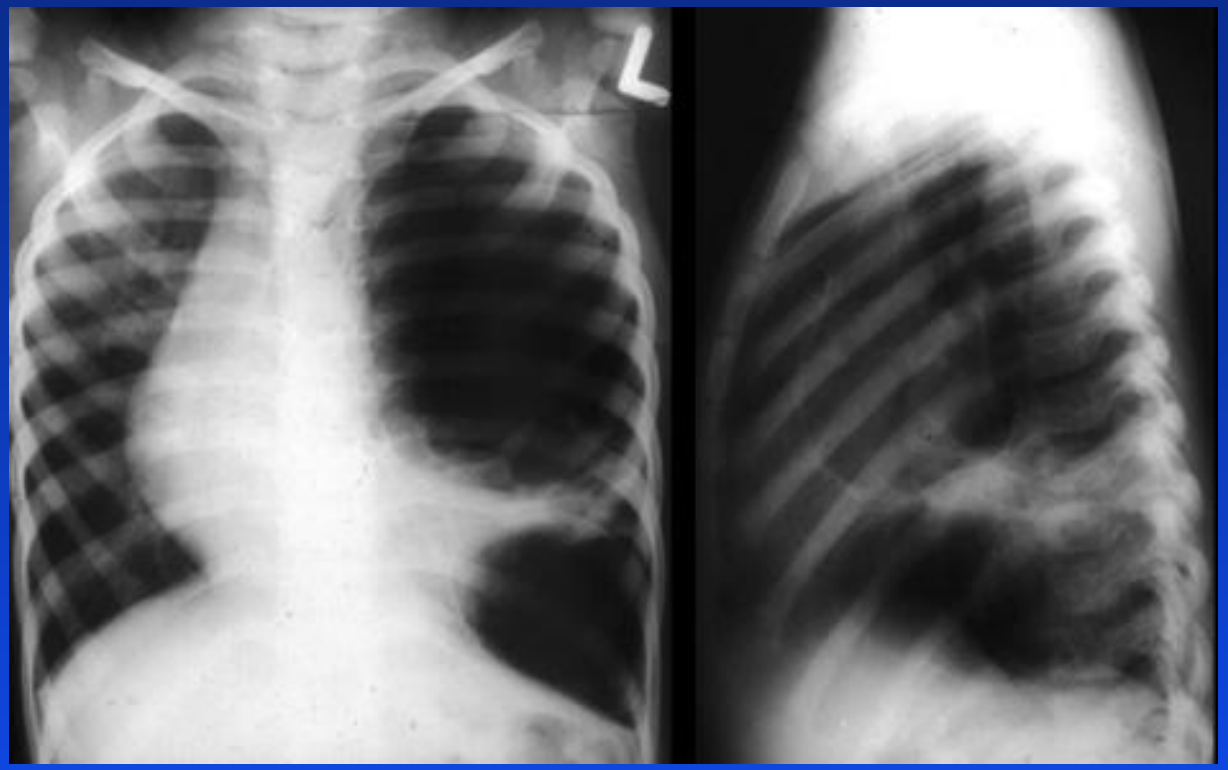
GBS Pneumonia

CONGENITAL MALFORMATIONS

- Choanal Atresia
- Tracheal Atresia/stenosis
- Chest Mass
  - Diaphragmatic hernia
  - CCAM
  - Sequestration
  - Lobar emphysema



CCAM



Lobar Emphysema

Diaphragmatic Hernia

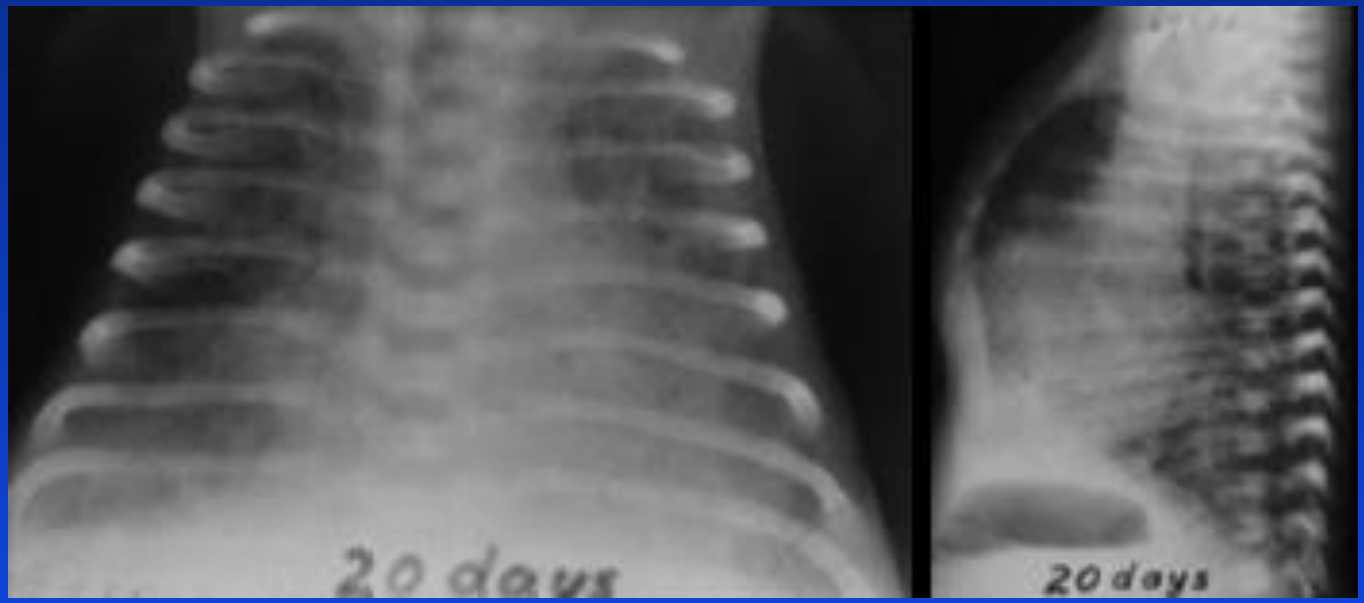
Chylothorax

Phrenic Nerve Paralysis

ACQUIRED DISEASES

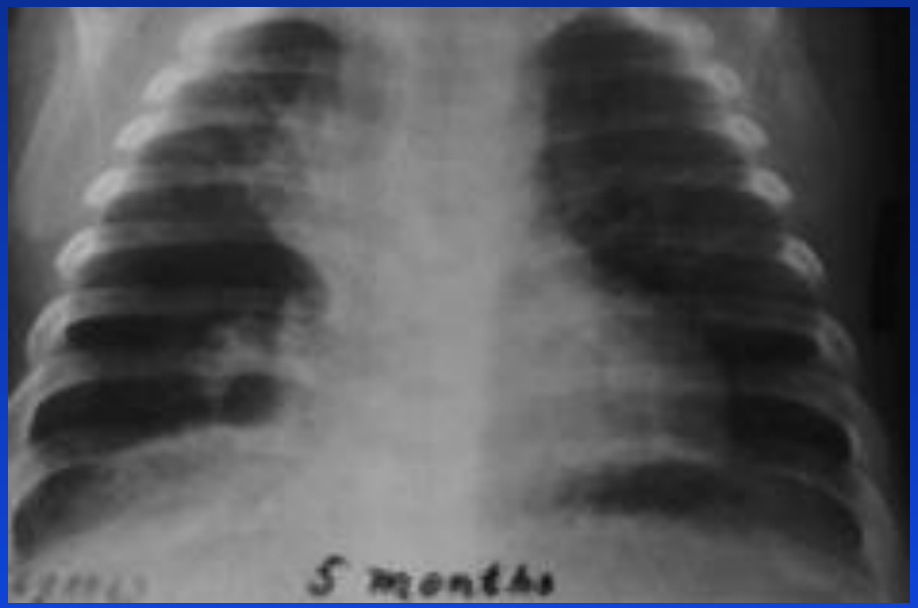
- Infections
- Bronchopulmonary Dysplasia
- Sub-glottic stenosis
- Apnea of Prematurity

Early BPD



Progressive BPD

Late BPD



APNEA

Definition: cessation of breathing for longer than a 15 second period or for a shorter time if there is bradycardia or cyanosis

Babies at Risk for Apnea

- **Preterm**
- **Respiratory Distress**
- **Metabolic Disorders**
- **Infections**
- **Cold-stressed babies who are being warmed**
- **CNS disorders**
- **Low Blood volume or low Hematocrit**
- **Perinatal Compromise**
- **Maternal drugs in labor**

Anticipation and Detection

- **Place at-risk infants on cardio-respiratory monitor**
- **Low heart rate limit (80-100)**
- **Respiratory alarm (15-20 seconds)**

Treatment

- **Determine cause:**
- **x-ray**
- **blood sugar**
- **body and environmental temperature**
- **hematocrit**
- **sepsis work up**
- **electrolytes**
- **cardiac work up**
- **r/o seizure**

Treatment

- **CPAP**
- **Theophylline/Caffeine therapy**
- **Mechanical ventilation**
- **Apnea monitor**