Respiratory Distress Syndrome (Hyaline Membrane Disease)

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INCIDENCE

- RDS occurs primarily in premature infants.
- Its incidence is inversely related to gestational age and birth weight.
- It occurs in 60–80% of infants less than 28 wk of gestational age, in 15–30% of those between 32 and 36 wk, in about 5% beyond 37 wk, and rarely at term.

Risk Factors

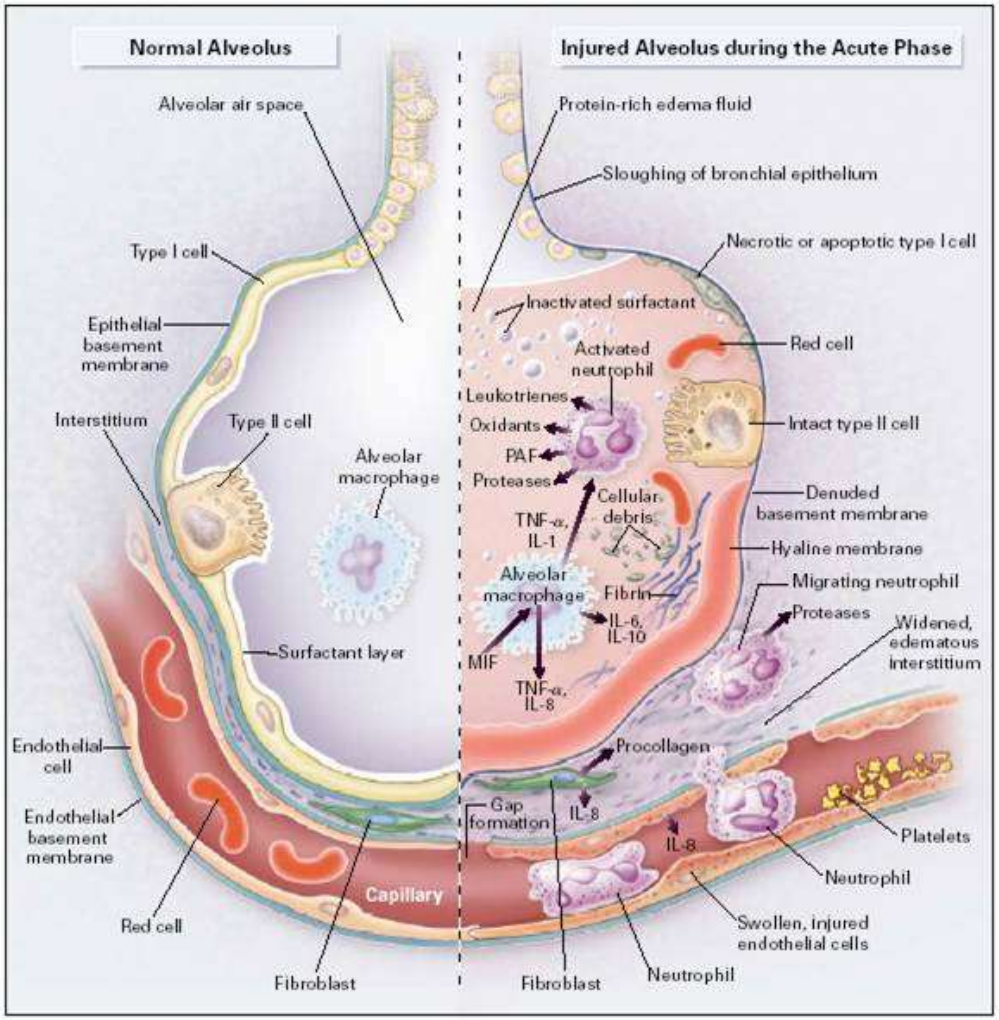
- Maternal diabetes.
- Multiple births.
- Cesarean section delivery.
- Precipitous delivery.
- Asphyxia.
- Cold stress.
- History of previously affected infants.
- The incidence is highest in preterm male or white infants

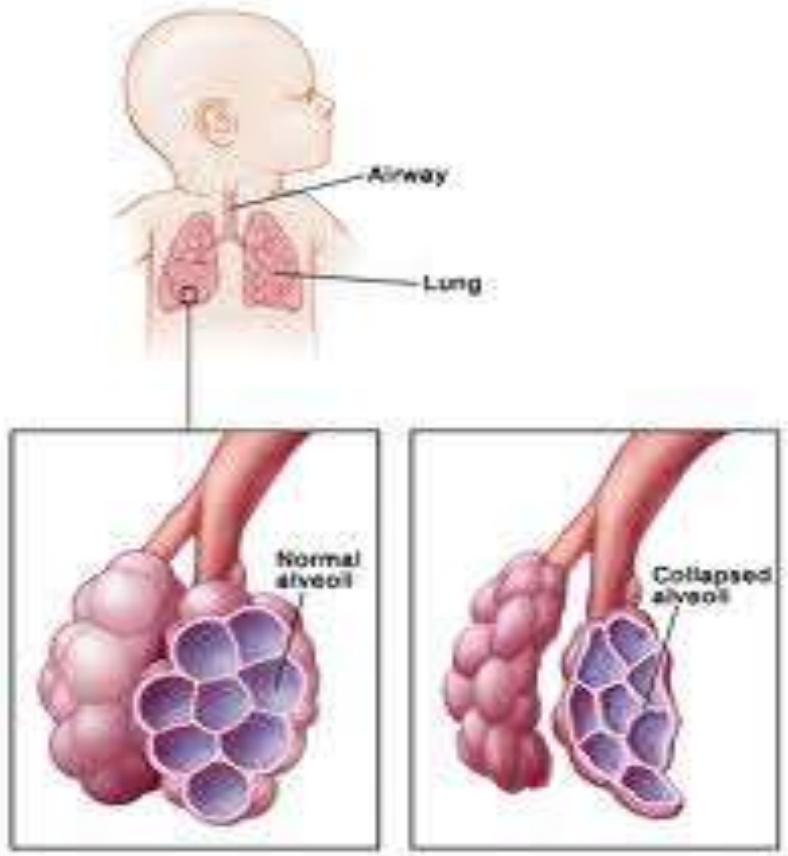
The risk of RDS is reduced:

- In pregnancies with chronic or pregnancy-associated hypertension.
- Maternal heroin use.
- Prolonged rupture of membranes.
- Antenatal corticosteroid prophylaxis.

ETIOLOGY AND PATHOPHYSIOLOGY

- Surfactant deficiency (decreased production and secretion) is the primary cause of RDS.
- With advancing gestational age, increasing amounts of phospholipids are synthesized and stored in type II alveolar cells .
- they reduce surface tension and help maintain alveolar stability by preventing the collapse .
- . Mature levels of pulmonary surfactant are usually present after 35 wk .
- Though rare, genetic disorders may contribute to respiratory distress .


Normal Alveolus
Injured Alveolus during the Acute Phase
Alveolar air space
Type I cell
Epithelial basement membrane
Interstitium
Type II cell
Alveolar macrophage
Surfactant layer
Endothelial cell
Endothelial basement membrane
Red cell
Capillary
Fibroblast
Protein-rich edema fluid
Sloughing of bronchial epithelium
Necrotic or apoptotic type I cell
Inactivated surfactant
Activated neutrophil
Red cell
Leukotrienes
Oxidants
PAF
Proteases
Intact type II cell
Cellular debris
Denuded basement membrane
TNF-α, IL-1
Hyaline membrane
Alveolar macrophage
Fibrin
Migrating neutrophil
IL-6, IL-10
Proteases
MIF
TNF-α, IL-8
Widened, edematous interstitium
Procollagen
Gap formation
IL-8
IL-8
Platelets
Neutrophil
Swollen, injured endothelial cells
Fibroblast
Neutrophil


Airway
Lung
Normal alveoli
Collapsed alveoli

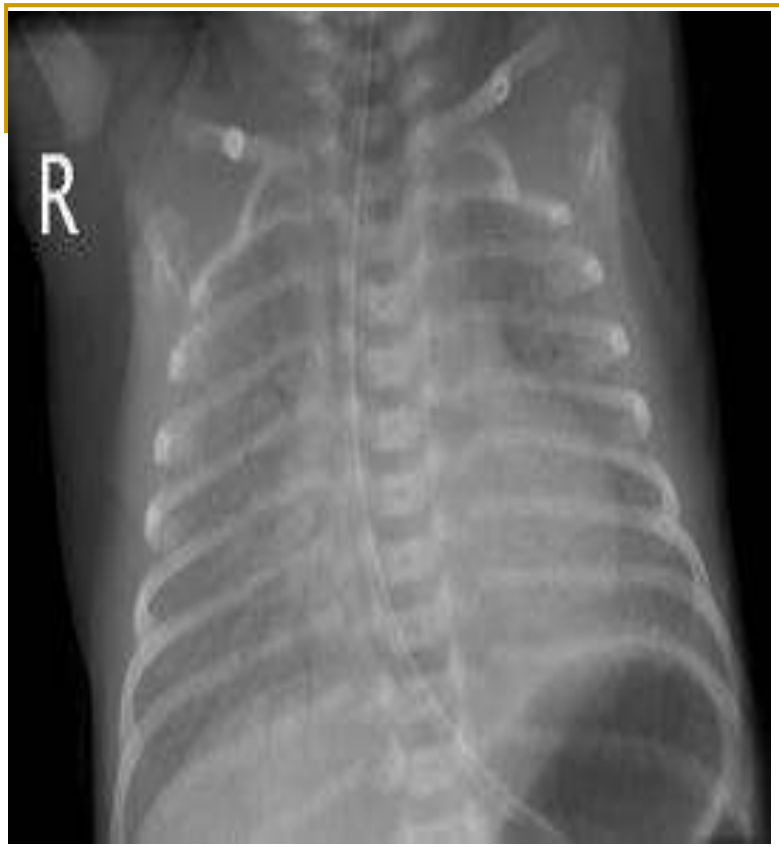
CLINICAL MANIFESTATIONS

- Signs of RDS usually appear within minutes of birth.
- The characteristic hyaline membranes are rarely seen in infants dying earlier than 6–8 hr after birth .
- The natural course of untreated RDS is characterized by progressive worsening of cyanosis and dyspnea.
- In most cases, the symptoms and signs reach a peak within 3 days.

- Improvement is often heralded by spontaneous diuresis and the ability to oxygenate the infant at lower inspired oxygen levels or lower ventilator pressures .
- . Death is rare on the 1st day of illness, usually occurs between days 2 and 7, and is associated with alveolar air leaks (interstitial emphysema, pneumothorax), pulmonary hemorrhage, or IVH .
- Mortality may be delayed weeks or months if BPD develops in mechanically ventilated infants with severe RDS.

DIAGNOSIS

- The clinical course, x-ray of the chest, and blood gas and acid-base values help establish the clinical diagnosis .
- X-ray, the lungs may have a characteristic, but not pathognomonic appearanc .
- A fine reticular granularity of the parenchyma and air bronchograms.
- Early-onset sepsis may be indistinguishable from RDS .
- In pneumonia manifested at birth, the chest roentgenogram may be identical to that for RDS .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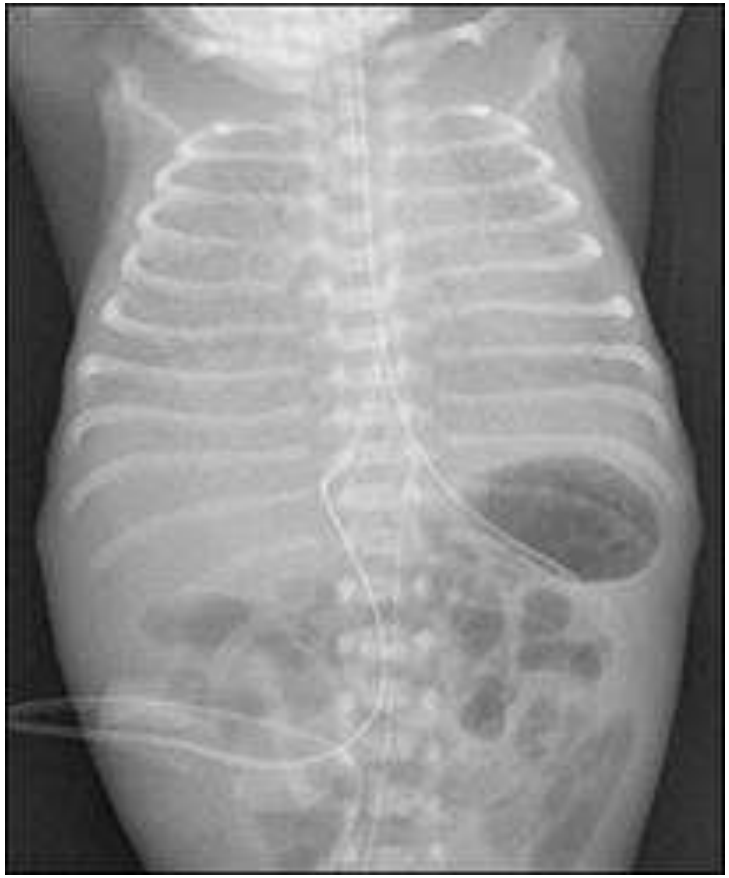

- Cyanotic heart disease (total anomalous pulmonary venous return) can also mimic RDS both clinically and radiographically.
- Congenital alveolar proteinosis (congenital surfactant protein B deficiency) is a rare familial disease that manifests as severe and lethal RDS in predominantly term and near-term infants .
- In atypical cases of RDS, a lung profile (lecithin:sphingomyelin ratio and phosphatidylglycerol level) performed on a tracheal aspirate can be helpful in establishing a diagnosis of surfactant deficiency.

PREVENTION

- Avoidance of unnecessary or poorly timed cesarean section .
- Appropriate management of high-risk pregnancy and labor .
- Prediction and possible in utero acceleration of pulmonary immaturity are important preventive strategies.
- Antenatal and intrapartum fetal monitoring may similarly decrease the risk of fetal asphyxia; asphyxia is associated with an increased incidence and severity of RDS.

PREVENTION

- Administration of betamethasone to women 48 hr before the delivery of fetuses between 24 and 34 wk of gestation significantly reduces the incidence, mortality, and morbidity of RDS .
- Repeated weekly doses of betamethasone until 32 wk .
- Prenatal glucocorticoid therapy decreases the severity of RDS and reduces the incidence of other complications of prematurity.
- Prenatal dexamethasone may be associated with a higher incidence of periventricular leukomalacia than betamethasone .

TREATMENT

- The basic defect requiring treatment is :-

  1- Inadequate pulmonary exchange of oxygen and carbon dioxide.

  2- Metabolic acidosis.

  3- Circulatory insufficiency.

  4-Multidose endotracheal instillation of **exogenous surfactant** and mechanical ventilation dramatically improves survival and reduces the incidence of pulmonary air leaks.

- Calories and fluids should initially be provided intravenously.
- Excessive fluids (>140 cc/kg/day) contribute to the development of PDA and BPD .
- If an infant managed by CPAP cannot maintain an arterial oxygen tension above 50 mm Hg while breathing 70–100% oxygen, assisted ventilation is required.

Reasonable indications for mechanical ventilation use are :-

(1) Arterial blood pH < 7.20,

(2) Arterial blood Pco2 of 60 mm Hg or higher,

(3) Arterial blood Po2 of 50 mm Hg or less at oxygen concentrations of 70–100% and CPAP of 6–10 cm H2O.

(4) Persistent apnea.

- Repeated dosing is given via the endotracheal tube every 6–12 hr for a total of 2 to 4 doses.
- **Inhaled nitric oxide (iNO)** decreases the need for extracorporeal membrane oxygenation (ECMO) in term and near-term infants with hypoxic respiratory failure.
- **Metabolic acidosis** in RDS may be a result of perinatal asphyxia and hypotension.

COMPLICATIONS

- Asphyxia from obstruction of the tube.
- Cardiac arrest during intubation or suctioning.
- The subsequent development of subglottic stenosis.
- **Post umbilical arterial/ venous catheterization** :-

  Embolization, thrombosis, spasm, and vascular perforation; ischemic or chemical necrosis of abdominal viscera; infection; accidental hemorrhage; and impaired circulation to a leg with subsequent gangrene.

Bronchopulmonary dysplasia (BPD)

- Is a result of lung injury in infants have severe respiratory distress requiring prolonged periods of mechanical ventilation and oxygen therapy.
- **Treatment** of BPD includes nutritional support, fluid restriction, drug therapy, maintenance of adequate oxygenation, and prompt treatment of infection.
- Growth must be monitored because recovery is dependent on the growth of lung tissue and remodeling of the pulmonary vascular bed.

PROGNOSIS

- Early provision of intensive observation and care of high-risk newborn infants .
- Antenatal steroids, postnatal surfactant use, improved modes of ventilation, and developmentally appropriate care have resulted in low mortality from RDS (from40%to ≈10%).
- Mortality increases with decreasing gestational age