Hyaline membrane disease

By: Dr. Ch Sarishma
Peadiatric Pg
• Also called Respiratory distress syndrome.
• It occurs primarily in premature infants; its incidence is inversely related to gestational age and birth weight.
• It occurs in 60-80% of infants <28 wk of gestational age, in 15-30% of those between 32 and 36 wk of gestational age, and rarely in those >37 wk of gestational age.
• The risk for development of RDS increases with maternal diabetes, multiple births, cesarean delivery, precipitous delivery, asphyxia, cold stress, and a maternal history of previously affected infants.
• The risk of RDS is reduced in pregnancies with chronic or pregnancy-associated hypertension, maternal heroin use, prolonged rupture of membranes, and antenatal corticosteroid prophylaxis.
ETIOLOGY

• Immature development of the respiratory system or inadequate amount of surfactant in the lungs.

• RDS is the leading cause of respiratory failure in preterm neonates.

• It is more common in males than females.
Pathophysiology of HMD:

1) Pulmonary immaturity—due to lack of surfactant

2) Damage to type-II alveolar cells due to asphyxia, acidosis, hypothermia, APH leads to atelectasis and alveolar collapse.
Surfactant

Produced by type II alveolar epithelial cells
Composed of lipids and surfactant proteins

Phospholipids (84%)
- Phosphatidylcholine
- Phosphatidylycerol

Surfactant proteins (8%)
- Hydrophobic protein
  - SP-B and SP-C
- Hydrophilic proteins
  - SP-A and SP-D

Neutral lipids (8%)
• With advancing gestational age, increasing amounts of phospholipids are synthesized and stored in type II alveolar cells.

• These surface-active agents are released into the alveoli, where they reduce surface tension and help maintain alveolar stability by preventing the collapse of small air spaces at end-expiration.
• Because of immaturity, the amounts produced or released may be insufficient to meet postnatal demands.

• Surfactant is present in high concentrations in fetal lung homogenates by 20 wk of gestation, but it does not reach the surface of the lungs until later.

• It appears in amniotic fluid between 28 and 32 wk of gestation.

• Mature levels of pulmonary surfactant are present usually after 35 wk of gestation.
• Synthesis of surfactant depends in part on normal pH, temperature, and perfusion.
• Asphyxia, hypoxemia, and pulmonary ischemia, particularly in association with hypovolemia, hypotension, and cold stress, may suppress surfactant synthesis.
• The epithelial lining of the lungs may also be injured by high oxygen concentrations and the effects of respirator management, thereby resulting in a further reduction in surfactant.
• Deficient synthesis or release of surfactant, together with small respiratory units and a compliant chest wall, produces atelectasis and results in perfused but not ventilated alveoli, causing hypoxia.

• Decreased lung compliance, small tidal volumes, increased physiologic dead space, and insufficient alveolar ventilation eventually result in hypercapnia.
Pathophysiology

Intrapartum asphyxia
acidosis

Familial predisposition
C-section

Prematurity

↑ Rt to Lt shunt
CLINICAL MANIFESTATIONS

• Signs of RDS usually appear within minutes of birth, although they may not be recognized for several hours in larger premature infants until rapid, shallow respirations become more obvious.

• Some patients require resuscitation at birth because of intrapartum asphyxia or initial severe respiratory distress (especially with a birth weight <1,000 g).
• Characteristically, tachypnea, prominent (often audible) grunting, intercostal and subcostal retractions, nasal flaring, and cyanosis are noted.

• Breath sounds may be normal or diminished with a harsh tubular quality, and on deep inspiration, fine crackles may be heard.

• The natural course of untreated RDS is characterized by progressive worsening of cyanosis and dyspnea.
• Untreated patients may also have a mixed respiratory-metabolic acidosis, edema, ileus, and oliguria.

• Respiratory failure may occur in infants with rapid progression of the disease.

• In most cases, the signs reach a peak within 3 days, after which improvement is gradual.
• Death can result from severe impairment of gas exchange, alveolar air leaks (interstitial emphysema, pneumothorax), pulmonary hemorrhage, or IVH.
• BPD is a form of chronic lung disease that often develops in infants with severe RDS.
DIAGNOSIS

• The clinical course, chest x-ray findings, and blood gas and acid–base values help establish the clinical diagnosis.

• On x-ray, the lungs may have a characteristic but not pathognomonic appearance that includes a fine reticular granularity of the parenchyma and air bronchograms, which are often more prominent early in the left lower lobe because of superimposition of the cardiac shadow.
Classical HMD – white out lungs
• Laboratory findings are characterized initially by hypoxemia and later by progressive hypoxemia, hypercapnia, and variable metabolic acidosis.
PREVENTION

• Avoidance of unnecessary or poorly timed early cesarean section (<39 wk) or induction of labor, appropriate management of high-risk pregnancy and labor
• Administration of antenatal corticosteroids
• Antenatal and intrapartum fetal monitoring may decrease the risk of fetal asphyxia; asphyxia is associated with an increased incidence and severity of RDS.
• Steroid administration is recommended for all women in preterm labor who are likely to deliver a fetus within 1 wk.

• Antenatal steroids act synergistically with postnatal exogenous surfactant therapy so they should be given even though surfactant therapy is effective.

• Betamethasone and dexamethasone have both been used antenatally.

• Betamethasone may reduce neonatal death to a greater extent as compared to dexamethasone.
• Antenatal steroids also reduce the need for and duration of ventilatory support and admission to a neonatal intensive care unit and the incidence of severe IVH, necrotizing enterocolitis, and developmental delay.

• Inj Betamethasone 12 mg IM every 24 hrs X 2 doses; or

• Inj Dexamethasone 6 mg IM every 12 hrs X 4 doses.
• Administration of surfactant into the trachea of symptomatic premature infants immediately after birth (prophylactic) or during the 1st few hr of life (early rescue) showed reduced air leak and mortality from RDS.

• *CPAP started at birth is as effective as prophylactic or early surfactant and is the approach of choice for the delivery room management of a preterm neonate at risk for RDS.*
Assessment of fetal lung maturity:

1. **Lecithin spingomyelin ratio:**
   
   The risk of RDS is very low if L/S ratio is > 2.

2. **Lamellar body** counts in the amniotic fluid have also been used as a rapid and inexpensive test to determine FLM.
   
   - Lamellar bodies are “packages” of phospholipids produced by type II alveolar cells, and are present in amniotic fluid in increasing numbers with advancing gestational age.
   
   - A count of >50,000 lamellar bodies/microliter predicted lung maturity.
Gastric aspirate Shake test

- Take a test tube
- Mix 0.5 ml gastric aspirate +
  1 ml absolute alcohol
- Shake for 15 seconds

Allow to stand 15 minutes.

  if < 1/3\text{rd} test is Negative (HMD)

  if > 2/3\text{rd} test is Positive (Maturity)
TREATMENT

• The basic defect requiring treatment in RDS is inadequate pulmonary exchange of oxygen and carbon dioxide; metabolic acidosis and circulatory insufficiency are secondary manifestations.

• Early supportive care of premature infants, especially in the treatment of acidosis, hypoxia, hypotension and hypothermia, may lessen the severity of RDS.
Supportive treatment

• Therapy requires careful and frequent monitoring of heart and respiratory rates, oxygen saturation, Pao2, Paco2, pH, serum bicarbonate, electrolytes, glucose, hematocrit, blood pressure, and temperature.

• To avoid hypothermia and minimize oxygen consumption, the infant should be placed in an incubator or radiant warmer, and core temperature maintained between 36.5 and 37°C.
• Calories and fluids should initially be provided intravenously.
• For the 1st 24 hr, 10% glucose solution with additional amino acids in extremely premature infants, should be infused.
• Electrolytes should be added on day 2 in the most mature infants and on days 3-7 in the more immature ones.
• Fluid volume is increased gradually over the 1st wk. Excessive fluids (>140 mL/kg/day) contribute to the development of patent ductus arteriosus (PDA) and BPD.

• Warm humidified oxygen should be provided at a concentration initially sufficient to keep arterial oxygen pressure between 50 an 70 mm Hg (91-95% saturation) in order to maintain normal tissue oxygenation while minimizing the risk of oxygen toxicity.
• If oxygen saturation cannot be kept >90% at inspired oxygen concentrations of 40-70% or greater, applying CPAP at a pressure of 5-10 cm H2O via nasal prongs is indicated and usually produces a rapid improvement in oxygenation.

• CPAP reduces collapse of surfactant-deficient alveoli and improves both FRC and ventilation–perfusion matching.
• Infants with respiratory failure or persistent apnea require assisted mechanical ventilation.

• Reasonable measures of respiratory failure are: arterial blood pH <7.20, arterial blood Pco2 of 60 mm Hg or higher, and oxygen saturation <90% at oxygen concentrations of 40-70% and CPAP of 5-10 cm H2O.

• The goal of mechanical ventilation is to improve oxygenation and elimination of carbon dioxide without causing pulmonary injury or oxygen toxicity.
Definitive treatment

• Surfactant deficiency is the primary pathophysiology of RDS.

• Source of surfactant: bovine, porcine, calf lung

• Dose: 4ml/kg body weight given in 4 aliquots.
• subsequent doses are given at 12-hour intervals, if needed.
• It is administered during brief disconnection from the ventilator, in quarter doses through a feeding tube that is cut to a length just slightly longer than that of the endotracheal tube.
• The baby is ventilated for at least 30 seconds, or until stable between quarter doses.
• Changes in positioning of the infant during administration are routine and intended to facilitate distribution.
Steps of administration...
Steps of administration..
Steps of administration..

- >20 G needle
Steps of administration..
Steps of administration..
• Immediate effects of surfactant replacement therapy include improved alveolar-arterial oxygen gradients, reduced ventilatory support, increased pulmonary compliance, and improved chest radiograph appearance.

• In neonates with RDS who fail CPAP, treatment with endotracheal surfactant should be initiated immediately after intubation.
Complications of surfactant therapy include:

- transient hypoxia,
- hypercapnia,
- bradycardia and hypotension,
- blockage of the endotracheal tube, and pulmonary hemorrhage.
Chest radiograph before and after surfactant
• **Strategies for weaning infants from ventilators vary widely and are** influenced by lung mechanics as well as the availability of ventilatory modes (pressure support).

• Once extubated, many infants are transitioned to nasal CPAP to avoid postextubation atelectasis and reduce re-intubation.

• Synchronized nasal intermittent ventilation decreases the need for re-intubation in premature infants.
• High flow (1-2 L/min) or warmed, humidified high-flow (2-8 L/min) nasal cannula oxygen is commonly used to support term and near-term infants following extubation and to wean premature infants from nasal CPAP.
Complications of RDS

• IVH manifested by apnic attacks
• L → R shunt through PDA leads to CHF
• DIC leading to hemarrhagic infarction in the brain and pulmonary hemorrhage
• Prolonged ventilation leads to air leak syndrome and BPD
• Retinopathy of prematurity d/t prolonged hyperoxia.
PROGNOSIS

• Early provision of intensive observation and care of high-risk newborn infants can significantly reduce the morbidity and mortality associated with RDS and other acute neonatal illnesses.

• Antenatal steroids, postnatal surfactant use, and improved modes of ventilation have resulted in low mortality from RDS.

• Mortality increases with decreasing gestational age.
THANK YOU