Chapter 2A

Lung Pathology: Respiratory Distress Syndrome and Its Complications

Editor

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Chest X-Ray Findings in Respiratory Distress Syndrome

Respiratory distress syndrome (RDS), or hyaline membrane disease (HMD), is the most common cause of respiratory distress in the premature neonate. The radiographic appearance of the lungs of neonates with RDS is distinctive and characteristic. Most neonates with this disorder demonstrate clinical findings of respiratory distress in the delivery room or during the first few hours of life. Neonates with severe disease usually present with both early clinical findings and x-ray changes indicating diffuse atelectasis. The appearance of the chest x-ray varies depending upon the grade and severity of the disease; the ventilatory support applied; the use of exogenous surfactant therapy; and other complications of prematurity, such as a patent ductus arteriosus. It is important to remember that RDS is normally a diffuse, bilaterally occurring disease, with both lung fields having a similar appearance. Pneumonia, which may mimic the x-ray picture of RDS, can occasionally be differentiated from it by the lack of a symmetrical appearance of the lung fields.

X-RAY PRESENTATION

The four characteristic features of the x-ray presentation in RDS are:

1. Reduced lung volume
2. Air bronchograms
3. Reticulogranularity
4. Increased lung opacification

Reduced Lung Volume

Atelectasis occurs because of a lack of surfactant activity or availability; as a consequence, chest expansion is reduced. This reduction in lung volume is most obvious in neonates who are not receiving continuous positive airway pressure or mechanical ventilation and who have not received exogenous surfactant. On x-ray, the diaphragms are high and sometimes domed, and the intercostal spaces are narrow. Lung expansion may be only to the fifth, sixth, or seventh thoracic vertebra rather than to the eighth or ninth, as found in the normally expanded chest.

Air Bronchograms

Air bronchograms (Figure 2A-1) are the outlines of air-filled secondary and tertiary bronchi seen over abnormal lung fields. Air bronchograms are usually visible only over the heart because it is of water density; on x-ray, air-filled bronchi are visible against a background of greater density. Because normally expanded lungs are of air density and the overlying bronchi are also of air density, air bronchograms are not seen in the lung fields on a normal chest x-ray. When atelectasis occurs with RDS, the lung density becomes water/tissue density, and lucent air is seen outlining the peripheral bronchi over the more opaque lung. With severe atelectasis, air in the lung is greatly reduced, leading to loss of the air bronchogram sign and more diffusely opaque lung fields.

Reticulogranularity

A reticulogranular pattern, or ground glass appearance, uniformly distributed throughout both lung fields is characteristic of RDS. Because of surfactant deficiency, alveoli

FIGURE 2A-1 ■ Air bronchograms extending into the lung fields are visible.

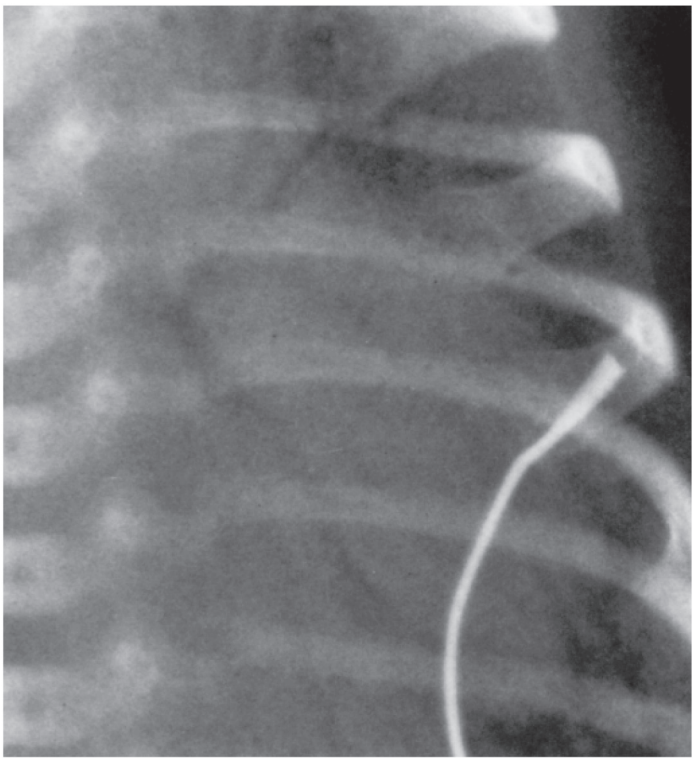


FIGURE 2A-2 ■ A fine reticulogranular pattern is present, and air bronchograms are apparent near the diaphragm.

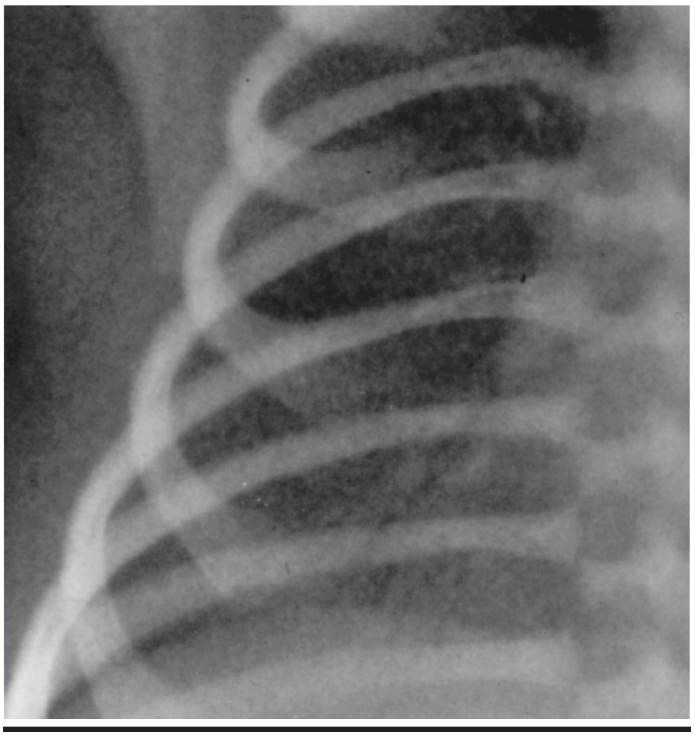
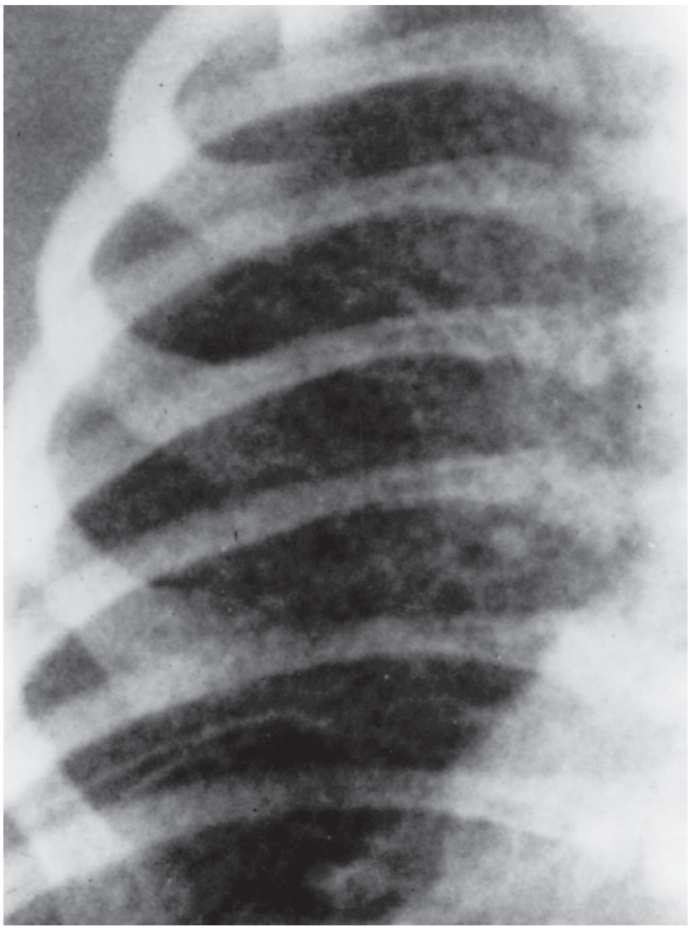


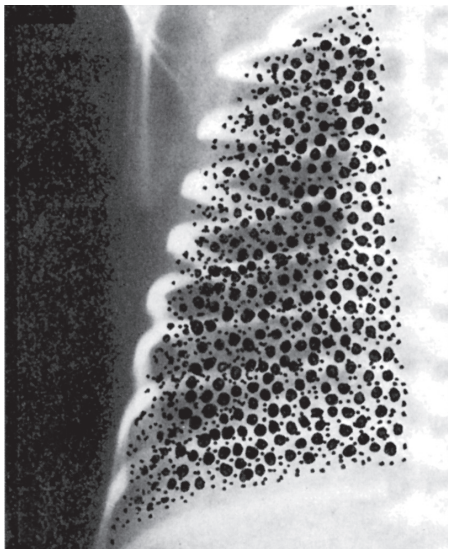
FIGURE 2A-3 ■ Rounded spherical lucencies of the Type I bubble can be seen, as well as air bronchograms.



throughout the lung have high surface tension, and some are collapsed. The alveoli that remain distended have lower surface tension; air that enters the lung will go to the area of lowest pressure, that is, the partially distended alveoli. The atelectatic alveoli have high surface tension, requiring high opening pressures. When inspiration occurs, the terminal airways (ducts leading to the alveoli and the terminal bronchioles) distend because they are more elastic than the collapsed alveoli. The expanded alveoli and terminal airways against a background of atelectasis appear on x-ray as a fine reticulogranular pattern (Figure 2A-2).

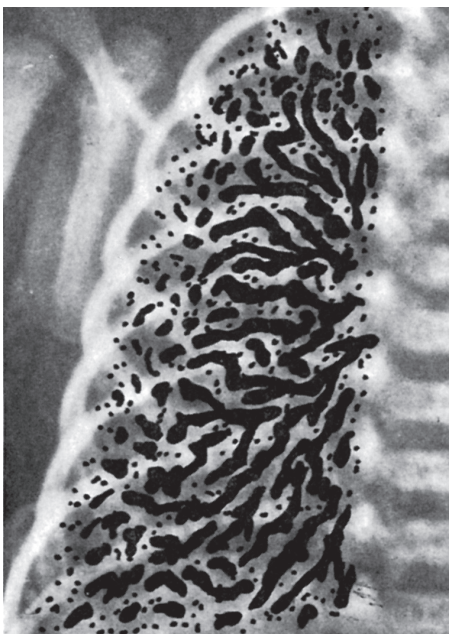
A more distinct granular or bubble pattern may be seen in neonates with RDS. Fine spherical bubbles of 1 to 2 mm in diameter have been designated Type I bubbles consistent with respiratory distress syndrome (Figures 2A-3 and 2A-4).[1] These bubbles are seen bilaterally and represent a stage of lung disease in which there is widespread alveolar collapse. Inspiration results in overdistention of the terminal airways, bronchioles, and alveolar ducts rather than of the more high- pressure alveolar units. This x-ray appearance is usually

FIGURE 2A-4 ■ Diagrammatic representation of Type I bubble commonly found in infants with RDS.



From: Swischuk LE. 1977. Bubbles in hyaline membrane disease: Differentiation of three types. *Radiology* 122(2): 417. Reprinted by permission.

FIGURE 2A-5 ■ Diagrammatic representation of Type II bubble demonstrating air in the interstitium radiating out from the hilus of the lung.



From: Swischuk LE. 1977. Bubbles in hyaline membrane disease: Differentiation of three types. *Radiology* 122(2): 417. Reprinted by permission.

seen in the ventilated neonate, but has also been seen in larger neonates with RDS who are not receiving mechanical ventilation.

It is theorized that spontaneous respiration with grunting leads to increased intraluminal pressure during exhalation, resulting in distended airways in these neonates. As atelectasis increases and ventilation is more impaired in these nonventilated neonates, this pattern may be replaced by more diffuse opacity. The Type I bubble is also frequently seen soon after the institution of mechanical ventilation or continuous positive airway pressure. In these neonates, surfactant activity or availability is diminished, the pressure within the alveoli is high, atelectasis is widespread, and the addition of positive pressure ventilation leads to distention of the more elastic terminal bronchioles and alveolar ducts rather than distention of the alveoli themselves. In these neonates, the prior x-ray picture may have been one of greater opacity; the appearance of the Type I bubble pattern may give the impression of clearing. However, this is referred to as "pseudoclearing," because, although the x-ray does look less opaque, gas exchange at the alveolar unit has not improved, and the blood gas and clinical picture remain unchanged.

The Type I bubble must be discriminated from the bubble of interstitial emphysema, the Type II bubble. The appearance of the bubble of interstitial emphysema is nodular and wormy, rather than smoothly spherical, in most cases. Also the Type II bubble is often first seen unilaterally in the hilar region radiating outward (Figures 2A-5 and 2A-6).

FIGURE 2A-6 ■ The Type II bubble of pulmonary interstitial emphysema.

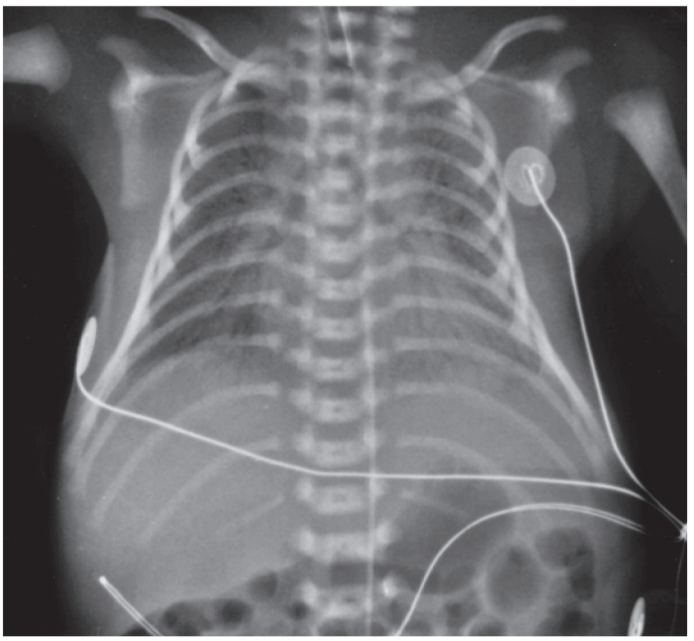


FIGURE 2A-7 ■ The chest x-ray of this neonate with RDS shows a reticulogranular pattern with air bronchograms.

There is increasing opacity in this film compared with previous films, and the heart borders are beginning to become less distinct.

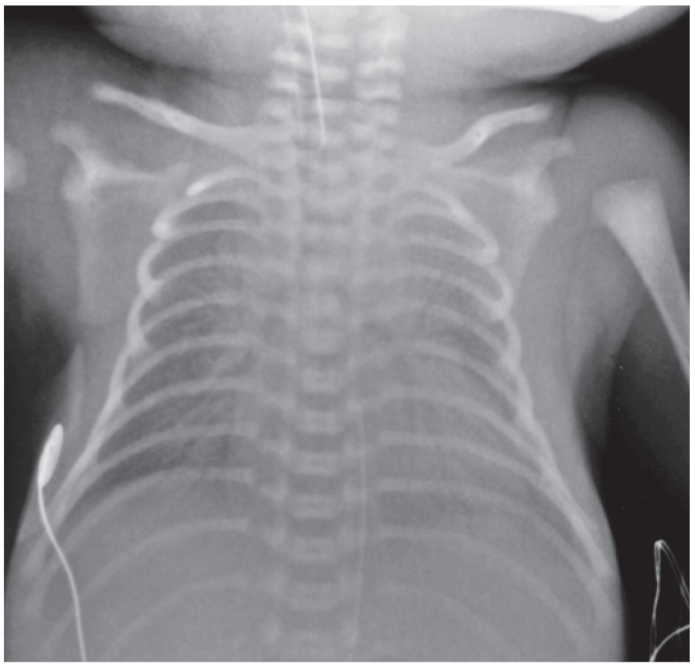
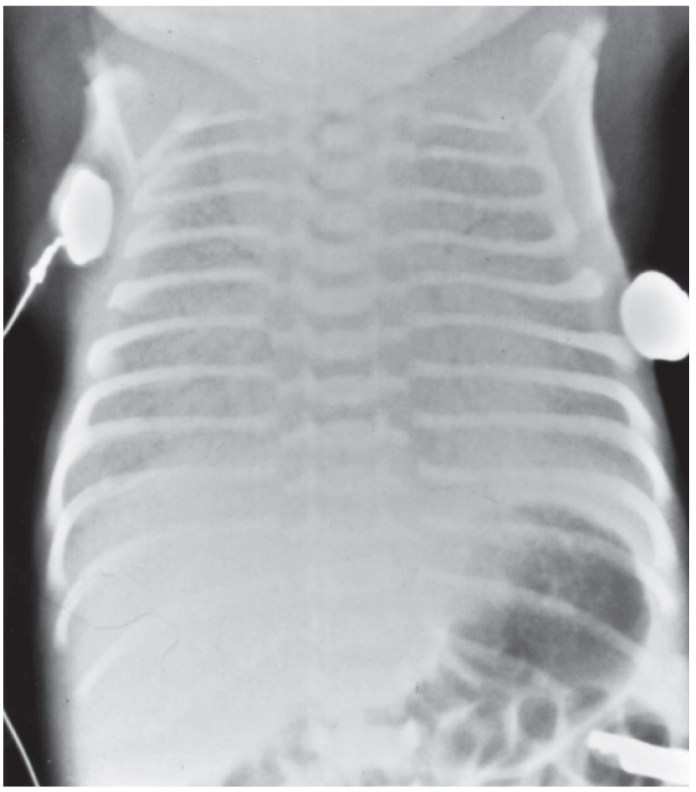


FIGURE 2A-8 ■ Diffuse opacification with loss of distinct heart borders.



Increased Lung Opacification

When diffuse opacification becomes apparent in the chest x-ray of the neonate with RDS, it is usually as a result of nonexpanded alveoli with little or no terminal airway aeration. Initially, the heart borders may be visible, but the x-ray may progress to complete loss of visualization of the heart borders or a "whiteout" (Figures 2A-7 and 2A-8).

Successive chest x-rays may gradually grow more opaque, preceded by a granular chest x-ray, but in some instances, the first x-ray may display this opaque appearance in the neonate with severe disease. Another cause of increasingly opaque lungs on chest x-ray may be pulmonary edema resulting from left-to-right shunting across the ductus arteriosus. Clinical evaluation for signs of a patent ductus arteriosus should be carried out when this chest x-ray picture is seen. Pulmonary edema may also occur secondary to "leaky capillaries," with increased capillary permeability leading to fluid leak into the alveolar spaces. As the alveoli fill with fluid, their appearance on the chest x-ray becomes more opaque. It has been suggested that the lung volumes of neonates with pulmonary edema will be greater than those of neonates with RDS because in edema the alveolar units are not collapsed but rather are filled with fluid. Expiratory films can also give the impression of increasing lung opacity, but recognition of a trachea deviated to the right, domed diaphragms, and a stable clinical picture allow for easy discrimination. Massive bilateral pulmonary hemorrhage can also lead to sudden near or total lung opacity. Generally, this problem is rapidly recognized clinically (Figure 2A-9).

PROGRESSION OF CHANGES

Four gradations in the progression and severity of x-ray changes seen in RDS have been outlined.[2] The first, Grade 1, consists of a fine granularity with some air bronchograms visible. Grade 2 is characterized by a more apparent, distinct, and coarse granularity to the lung fields, with more extensive air bronchograms. Grade 3 is characterized by increasing opacity, with decreasing air bronchograms and granularity. Heart borders are still visible in Grade 3. In Grade 4, diffuse bilateral opacification is present, with lack of apparent heart borders and loss of air bronchograms—a "whiteout" on chest x-ray.

The use of exogenous surfactant will change the x-ray and clinical course of RDS. Following surfactant administration, improved lung expansion and clearing lung fields with only a mildly hazy background are seen in many cases. In others, normal pulmonary radiolucency may follow surfactant administration. A small percentage of neonates are nonresponders, and their clinical and x-ray course changes little after surfactant administration.

FIGURE 2A-9 ■ The lungs of this premature neonate with bilateral pulmonary hemorrhage appear bilaterally opaque, although the right is more opaque than the left.

Marked clinical deterioration occurred before this film was taken, and large amounts of frank blood were suctioned from the endotracheal tube.

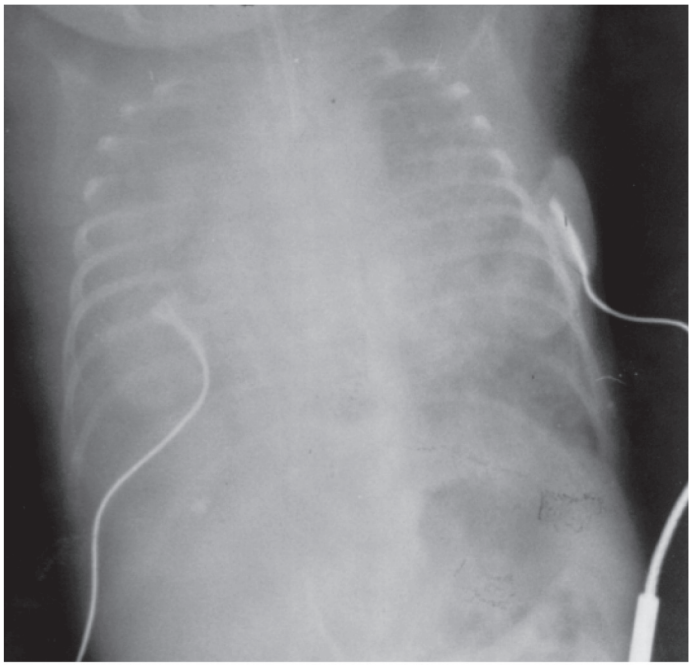
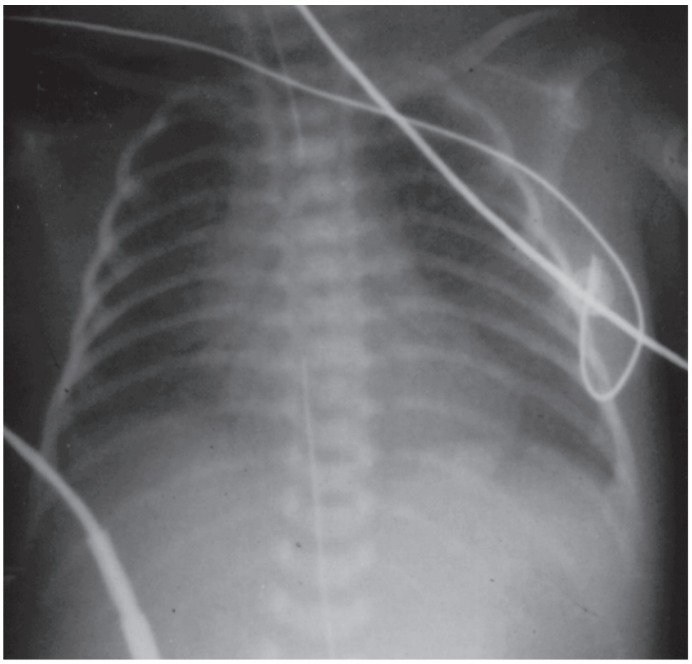


FIGURE 2A-10 ■ The mild, diffuse, bilateral haze characteristic of neonates in the recovery phase of RDS.



In the recovery phase of RDS, the x-ray appearance of the lungs presents as a mild, diffuse, bilateral haze (Figure 2A-10). In neonates whose course has been typical and who have not received surfactant, the recovery phase occurs between day 4 and 7 of life and is complete by day 10. The hazy appearance is thought to be due to the increased recruitment of mildly injured and edematous alveoli. The hazy appearance is lost over one to two weeks, and the lung fields return to their normal lucency.

An atypical x-ray appearance of the lungs in RDS has been described but is rarely seen. In some cases, the lower lobes show a more distinct pattern of change than the upper lobes. It is theorized that the upper lobes mature earlier than the lower lobes and therefore demonstrate less severity. This atypical appearance has been documented more commonly in larger premature neonates with milder disease. Positioning can also lead to apparent lobar differences in RDS. Prolonged supine positioning may lead to greater underaeration of the most dependent portion of the lung; the posterior lower lobes will appear more dense or opaque on x-ray. Prolonged positioning on one side leads to improved aeration of the elevated lung, and the x-ray may show increased density of the dependent lung. If, during administration, exogenous surfactant instillation is inadvertently greater in one lung than in the other, an asymmetrical picture may occur.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of the x-ray picture of respiratory distress syndrome must include pneumonia, Group B streptococcal being the most common. Because some alveoli contain inflammatory exudate, they will appear more opaque on x-ray than those that are air filled. This will give a pattern of reticulogranularity to the chest x-ray. If the alveoli extensively fill with exudate or fluid, the lungs may become more opaque. These similarities make it difficult to discriminate between the premature neonate with respiratory distress syndrome and the infant with pneumonia on the basis of the chest x-ray alone (Figure 2A-11). Other historical, clinical, and diagnostic information must be evaluated.

In some premature neonates, retained lung fluid may mimic RDS on chest x-ray when it presents with a diffuse, bilateral hazy or granular picture. Often there is an L:S ratio or lung profile indicating pulmonary maturity or treatment with maternal steroids that helps in attempting to determine the etiology of the respiratory distress. Lung expansion is frequently normal to increased due to fluid in the alveoli, and the clinical and blood gas picture is more consistent with retained lung fluid as a diagnosis.

X-ray studies contribute to a volume of information gathered during a period of rapid adjustment of the neonate to extrauterine life and to his particular disease process. Although

FIGURE 2A-11 ■ This chest x-ray shows a diffuse, bilateral reticulogranular pattern with air bronchograms and lack of clear definition of the heart borders.

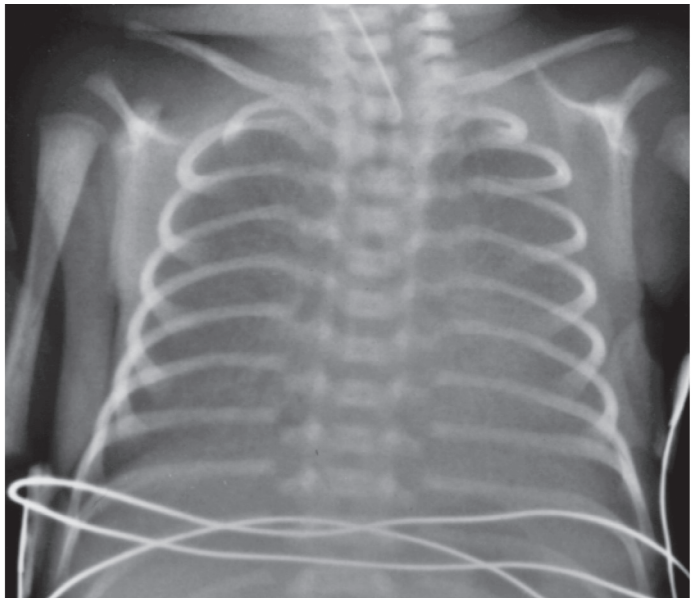
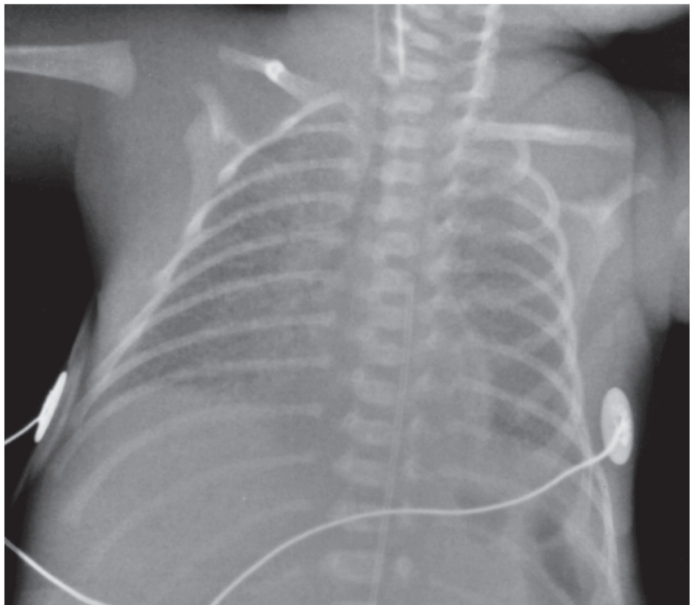


FIGURE 2A-12 ■ The first chest x-ray for a 33-week AGA neonate with respiratory distress.



x-rays serve as a guide to the patient's diagnosis and care, the history, the clinical and diagnostic data gathered, and the response to treatment must all be considered in reaching a definitive diagnosis.

CASE PRESENTATION

A 33-week gestation, 1.8 kg AGA female was born by normal spontaneous vaginal delivery to a 20-year-old gravida 2, para 1 mother. The pregnancy was uncomplicated, and the mother had received prenatal care from the tenth week of pregnancy. Her history was positive for premature delivery of her first child at 32 weeks gestation. Spontaneous rupture of membranes occurred seven hours prior to delivery, and the fluid was clear. Maternal VDRL was negative, blood type was O positive, rubella status was immune. Delivery occurred three hours after admission to the hospital, and the neonate's Apgar scores were 6 and 7 at one and five minutes. Respiratory distress and the need for oxygen were present in the delivery room. Soon after admission to the NICU, the infant was intubated and placed on mechanical ventilation because of a blood gas showing respiratory acidosis and hypoxemia in 65 percent oxygen. Upon examination prior to intubation, tachypnea, marked retractions, and poor breath sounds were evident. A CBC and differential were obtained and revealed no abnormality. Oxygenation and ventilation improved following intubation and mechanical ventilation. Figure 2A-12 shows the first chest x-ray for this patient.

X-Ray Evaluation

Indication for the x-ray was respiratory distress in a premature neonate, first admission film.

Penetration appears normal and without overexposure or underexposure.

Rotation is present to the right.

The soft tissues of the neck, chest, and extremities are of normal thickness and without emphysema. Skin folds are present over the left neck and shoulder.

The bony framework is intact, with 12 ribs bilaterally and normal vertebrae.

The trachea shows a straight air column, consistent with an inspiratory film. The tracheal bifurcation can be seen at the fourth thoracic vertebra (T4). The endotracheal tube is high, above the first thoracic vertebrae. The right and left mainstem bronchi are visible.

The hilum appears of normal size; the thymus is not visible; and the heart is of normal configuration, location, and size.

The diaphragm is at T8 on the right and T9 on the left.

The pleura reach the edges of the bony thorax, and the costophrenic angles are visible but clearer on the right than on the left.

Gastric air is present on the left.

The intercostal spaces are normal in size.

Lung fields reveal a diffuse, bilateral reticulogranular pattern with some visible air bronchograms extending beyond the heart.

The endotracheal tube is above T1. An intravascular line is present at T6 on the left side of the vertebral column, and an abdominal film (not shown) confirms an umbilical artery line.