Premature neonatal lung disease

RADIOLOGICAL MANIFESTATIONS OF DISEASE AND TREATMENT COMPLICATIONS

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Introduction

- 12% of all live births are premature (<37 weeks gestation)
- 14% increase in preterm birthrate since 1990 (multiple birth rate, lowered threshold of fetal viability)
- 60% of preterm births occur in Africa and South Asia

Blencowe H, et al. National, regional and worldwide estimates of preterm birth. The Lancet, June 2012.

Introduction

- premature newborns NICU (cost, expertise)
- complications of prematurity: ICH, NEC, sepsis
  - lung disease most common cause of morbidity
- improved survival - surfactant replacement, mechanical ventilation (modern ventilators, HFOV, ECMO)
- acute and chronic pulmonary disease and altered familiar radiologic patterns of disease.

- ClarkRH,et al. Lung injury in neonates: causes, strategies for prevention, and long-term consequences. *J Pediatr* 2001
- CourtneySE, et al. High-frequency oscillatory ventilation versus conventional mechanical ventilation for very-low-birth-weight infants. *N Engl J Med* 2002

Objectives

- Respiratory distress syndrome (RDS)
- RDS and surfactant era
- “old” bronchopulmonary dysplasia (BPD)
- “new” BPD
- Air leak complications

Neonatal Respiratory Distress Syndrome/*surfactant deficiency disorder*

- RDS - clinical expression of surfactant deficiency
- premature or term infants (DM mothers)
- respiratory distress shortly after birth (< 24 hrs)
- *Hyaline membrane disease* – histologic appearance necrotic alveolar cells, fibrin - line terminal bronchioles
- natural history of RDS modified: corticosteroids, prophylactic/rescue surfactant replacement, sophisticated assisted ventilation techniques

Martin J, et al.. Births: final data for 2002. *Natl Vital Stat Rep*2003

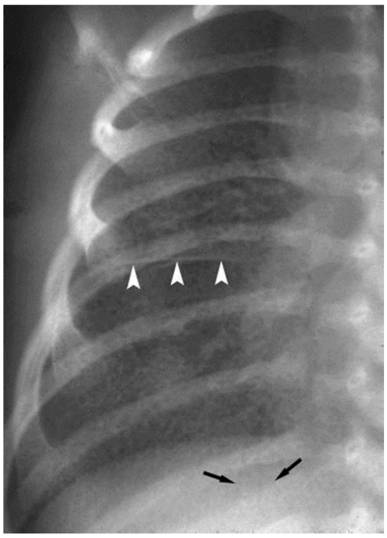
Radiologic Features ('classic" RDS)

- Surfactant deficiency - bilateral diffuse symmetric
- alveolar atelectasis – volume loss
- collapsed alveoli, transudate interstitium - reticulogranular, ground glass opacification
- obscuration pulmonary vessels, air bronchograms
- severe cases – dense consolidation (white-out)

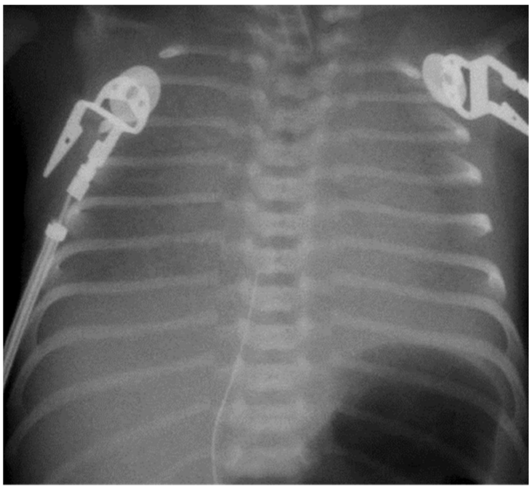
- mild RDS evolution typically: reticulogranular - generalized hazy opacities – clearing days to 2–3 weeks

ClevelandRH. A radiologic update on medical diseases of the newborn chest. *Pediatr Radiol*1995

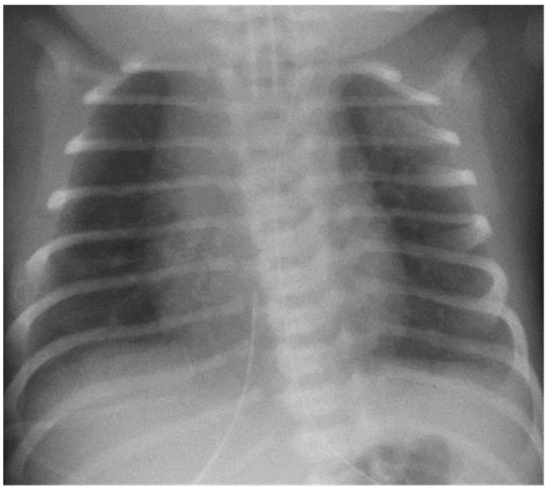
Mild RDS



Severe RDS



RDS post surfactant



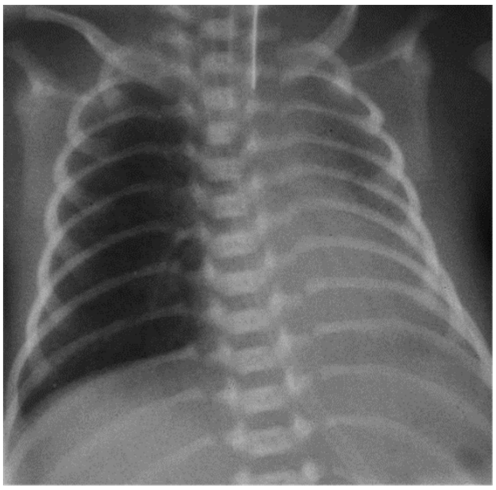
Radiologic Features of RDS with surfactant

- surfactant replacement - "classic" RDS radiographic findings less common
- RDS radiographic patterns with surfactant administration - complicates image interpretation
- particularly when surfactant has been administered before baseline imaging

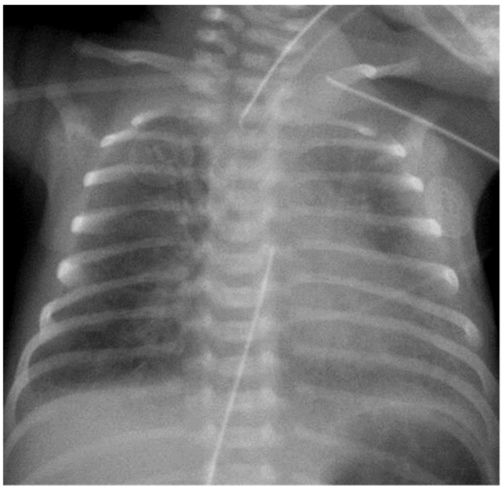
Radiologic Features of RDS with surfactant

- Localized segmental hyperinflation – may produce cystic lucencies - mimic interstitial air leak/PIE
- unilateral improvement - resulting in hyperlucent lung with contralateral mediastinal shift - mimic tension pneumothorax.

Unilateral distribution of surfactant



Asymmetric surfactant effect

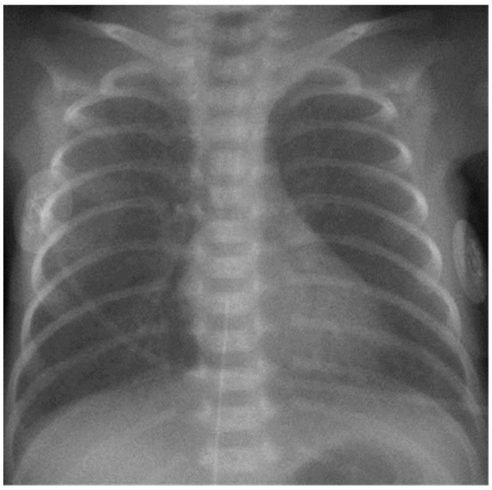


Radiologic Features of RDS with surfactant

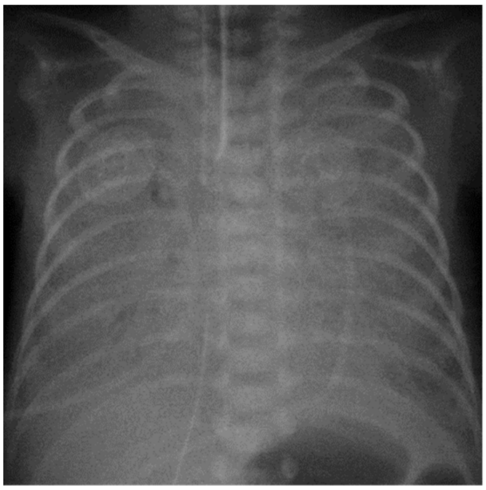
- Pulmonary hemorrhage - rare complication
- clinically acute respiratory decompensation after initial surfactant response
- radiograph - sudden dense airspace consolidation
- mechanism unclear - improved ventilation, decreased pulmonary vascular resistance, promotes left-to-right shunting through ductus arteriosus

van Houten J, et al. Pulmonary hemorrhage in premature infants after treatment with synthetic surfactant: an autopsy evaluation. *J Pediatr* 1992

Pulmonary hemorrhage



Pulmonary hemorrhage post surfactant



Bronchopulmonary Dysplasia

"Classic" BPD

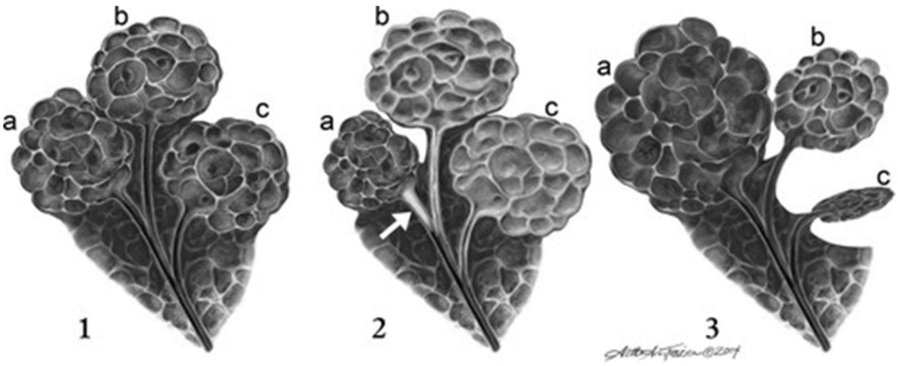
- chronic lung disease in premature infants (34 weeks, 2235g) with RDS
- treated with positive-pressure mechanical ventilation (> 3 days during first 2 weeks of life)
- required supplemental oxygen (beyond 28 days of life)
- developed characteristic radiographic abnormalities
- significant pulmonary dysfunction during first year of life
- Alveolar septal fibrosis the predominant residual feature

Northway WH Jr, et al. Pulmonary disease following respirator therapy of hyaline-membrane disease: bronchopulmonary dysplasia. *N Engl J Med*1967

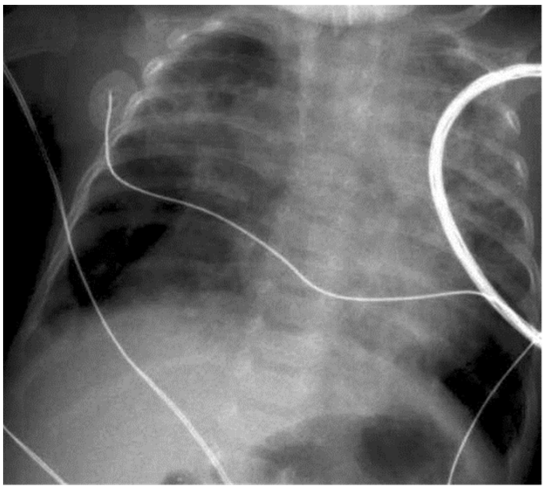
Radiologic criteria of "Classic" BPD

- stage I (2–3 days) reticulogranular/mild RDS
- stage II (4–10 days), near complete opacification
- stage III (10–20 days) small round lucencies (cysts) alternating with irregular opacity
- stage IV (> 1 month) larger lucencies alternating with thin strands of increased opacity, "bubbly lungs"

Proposed model for the pathogenesis of long-standing healed BPD



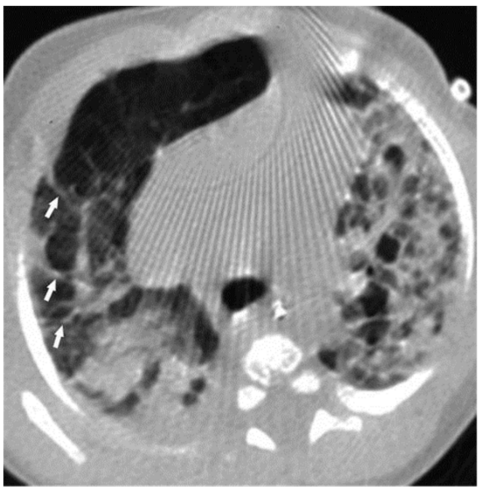
"Classic" severe BPD



Chest CT

- greater sensitivity than radiography,
- regional air trapping
- reticular and linear opacity - thickened interlobular septa,
- Subsegmental, lobar atelectasis,
- fibrosis;
- vascular attenuation with reduced bronchoarterial diameter ratios;
- bronchial wall thickening without bronchiectasis; and
- bullae or pneumatoceles

"Classic" severe BPD



"New" BPD

- very immature neonates (less than 30 weeks, <1200g), (did not survive previously)
- antenatal glucocorticoid administration, postnatal surfactant therapy, "gentler" ventilation
- synergy of oxidant injury and mechanical ventilation no longer considered major trigger
- Perinatal factors (low-grade chorioamnionitis) influence lung maturation likely play an important role in the pathogenesis of BPD
- fundamentally an inhibition of acinar and vascular growth during vulnerable stage of lung development

Jobe AJ. The new BPD: an arrest of lung development. *Pediatr Res*1999

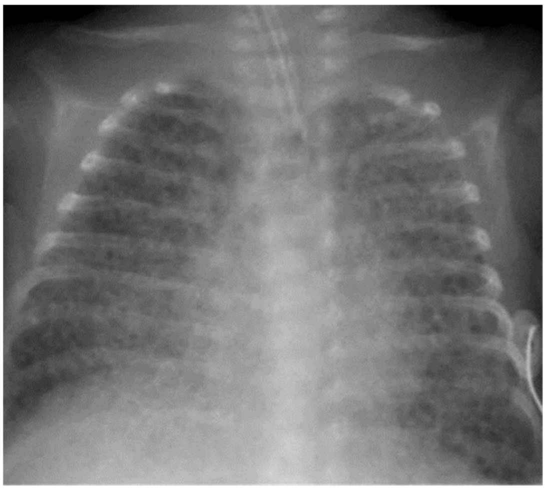
Lung development

- pseudoglandular phase (6–16 weeks gestation): airways to the level of the terminal bronchioles
- canalicular phase (16–28 weeks gestation): alveolar ducts(type II pneumocytes - surfactant) develop from respiratory bronchioles, thinning of the pulmonary interstitium allows gas exchange
- saccular phase (28–34 weeks gestation): increase in terminal sacs, thinning of interstitium, proliferation of the capillary bed, early development of true alveoli
- alveolar phase: 36 weeks gestation until 18 postnatal months,
- Factors influencing lung maturation (glucocorticoids, antepartum stressors)

Radiology of "new" BPD

- four radiographic stages of BPD less commonly observed
- normal/near normal initial chest radiograph
- gradual and subtle progression hazy reticulogranular opacity
- uniform pattern of coarse interstitial opacities without cystic lucencies
- eventually "bubbly" lungs (symmetric diffuse smaller cysts)

"New" BPD



Definition of BPD and Diagnostic Criteria

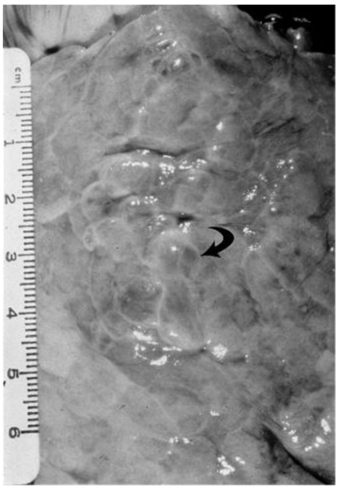
Gestational Age	Less Than 32 Wk	Greater Than 32 Wk
Time point of assessment	36 wk PMA* or discharge to home, whichever comes first; treatment with >21% oxygen for at least 28 d plus	>28 d but <56 d postnatal age or discharge to home, whichever comes first; treatment with >21% oxygen for at least 28 d plus
Mild BPD	Breathing room air at 36 wk PMA or discharge, whichever comes first	Breathing room air by 56 d of postnatal age or discharge, whichever comes first
Moderate BPD	Need† for <30% oxygen at 36 wk PMA or discharge, whichever comes first	Need† for <30% oxygen at 56 d postnatal age or discharge, whichever comes first
Severe BPD	Need† for ≥30% oxygen and/or positive pressure (PPV* or nasal CPAP) at 36 wk PMA or discharge, whichever comes first	Need† for ≥30% oxygen and/or positive pressure (PPV or nasal CPAP) at 56 d postnatal age or discharge, whichever comes first

Air Leak Phenomena

- Ventilation – airway barotrauma and volutrauma
- rupture bronchioloalveolar junctions, gas perivascular and peribronchial spaces - *pulmonary interstitial emphysema* (PIE)
- distinguished from true *emphysema*: permanent expansion of alveoli with absence of fibrosis.
- Gas dissect centrifugally: subpleural blebs, pneumothorax
- Centripetal: pneumomediastinum, -pericardium, systemic air embolism

EffmannE, Martin LD. Lymphatic air embolism: a proposed hypothesis. *Pediatr Radiol*1995

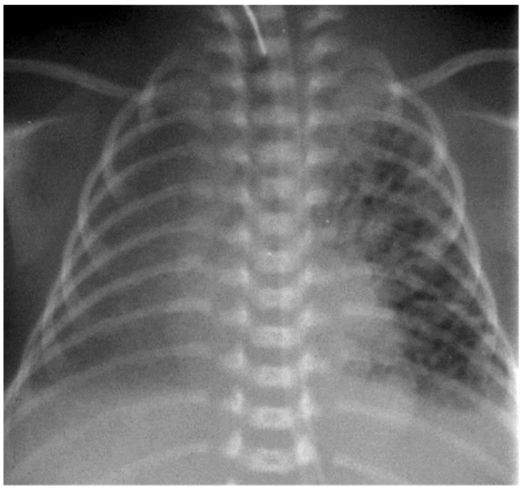
Acute PIE.



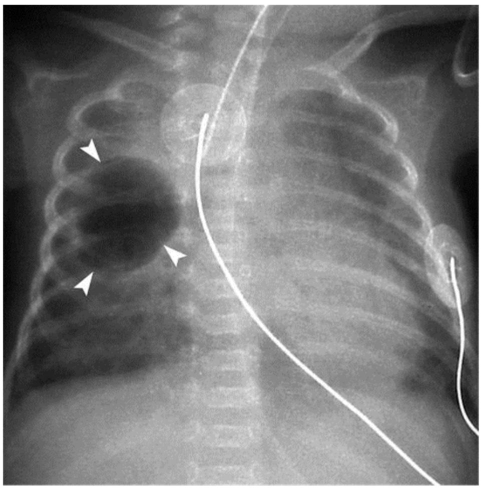
Radiology of acute PIE

- tubular and cystic lucencies – not branching pattern of air bronchograms
- focal or diffuse, unilateral or bilateral
- unilateral PIE: pulmonary overexpansion and contralateral shift of mediastinum
- focal PIE: single or multiple well-defined thin walled cystic air collections - *pseudocysts*

Unilateral acute PIE



Focal acute PIE

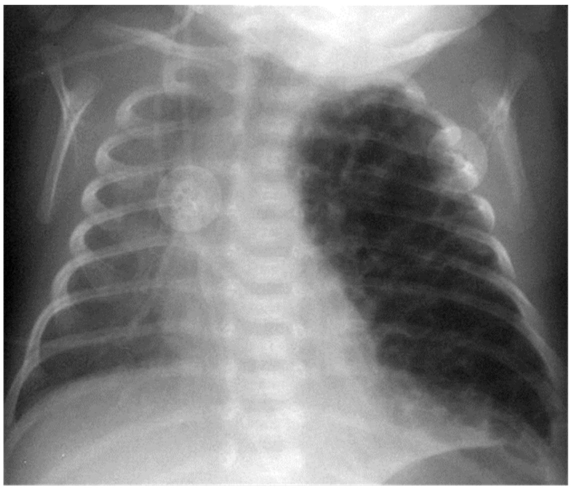


Persistent pulmonary interstitial emphysema

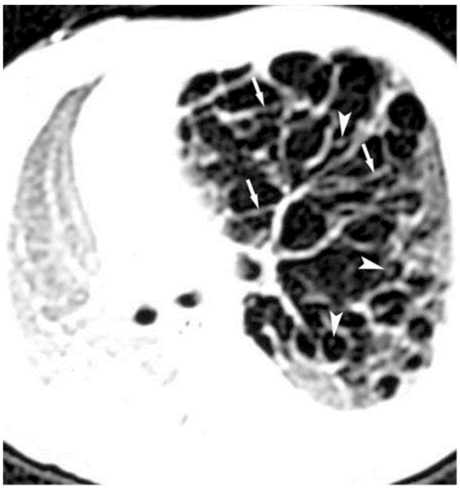
- PIE lasts >1 week
- focal or diffuse
- cysts are composed of fibrous walls
- lobar persistent PIE - expanding masslike aggregate of smooth-walled cysts
- may compress adjacent lung parenchyma and cause mediastinal displacement

Donnelly LF. Localized radiolucent chest lesions in neonates: causes and differentiation. *AJR Am J Roentgenol*1999

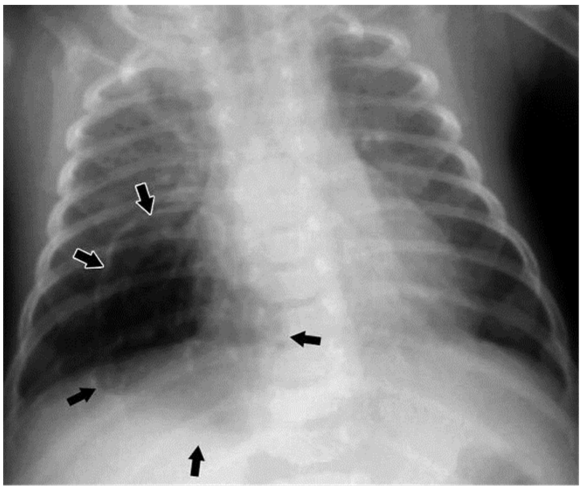
Diffuse persistent PIE



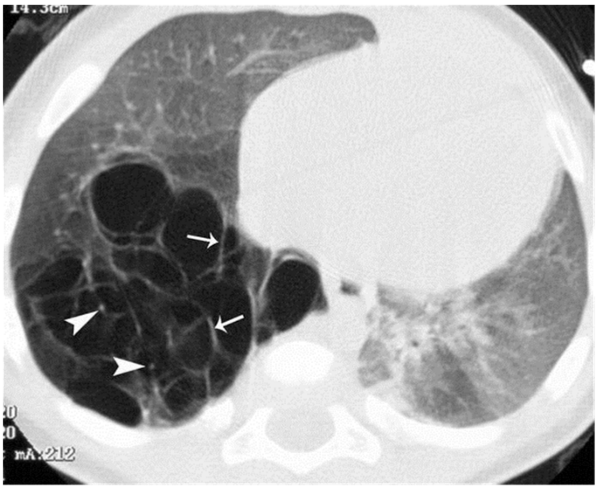
Diffuse persistent PIE



Localized persistent PIE



Localized persistent PIE



CT of the chest for evaluation of persistent PIE

- single-lobe or multilobar
- hyperexpanded cystic lucencies
- characteristic linear and dotlike structures of soft-tissue attenuation within the cysts(bronchovascular bundles surrounded by interstitial gas)

temporal relationship between acute PIE and persistent PIE usually excludes other causes (congenital lobar overinflation, cystic pulmonary airway malformation)

- persistent PIE have been reported in neonates who received only nasal CPAP
- CT is superior to radiography in characterizing pulmonary lobar involvement

Donnelly LF, et al. CT findings and temporal course of persistent pulmonary interstitial emphysema in neonates: a multi-institutional study. *AJR Am J Roentgenol* 2003

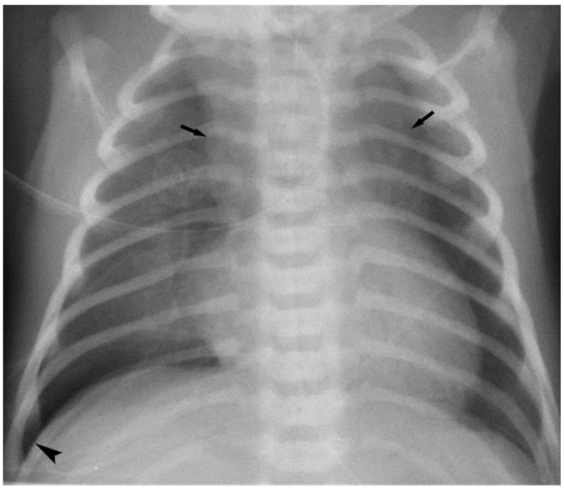
Pneumothorax

- supine infant, pleural air collects anteriorly
- pleural line often not discernible
- well-defined costophrenic sulcus (“deep sulcus sign”)
- anterior junction line - can indicate bilateral pneumothorax
- compress the thymus, “figure 8” or “pseudomass”

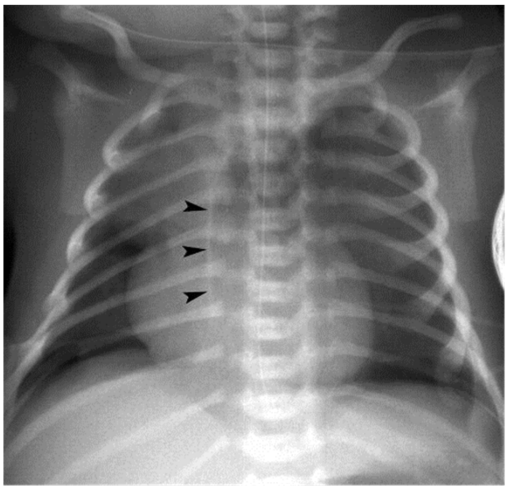
Pneumothorax

- Track the extrapleural space and outline the inferior aspect of the heart (“continuous diaphragm sign”)
- Dissect into soft tissues of the neck or chest
- pneumoretroperitoneum or -peritoneum
- Pericardial air (limited superiorly by pericardial reflection)
- Systemic air embolism

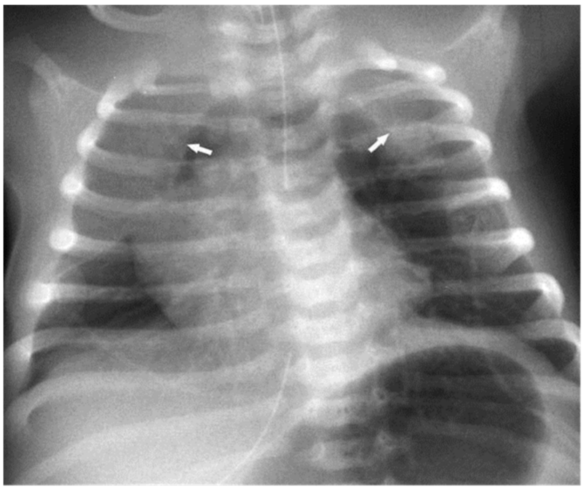
Pneumothorax



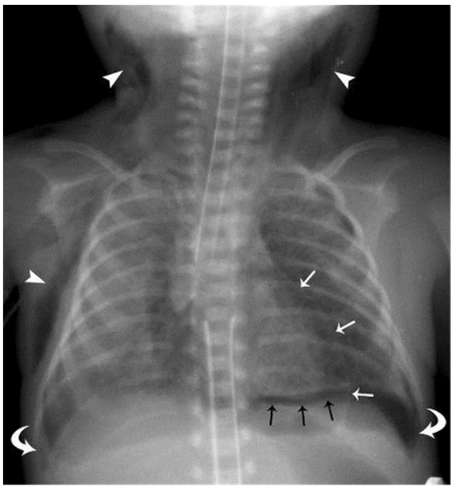
Bilateral pneumothoraces



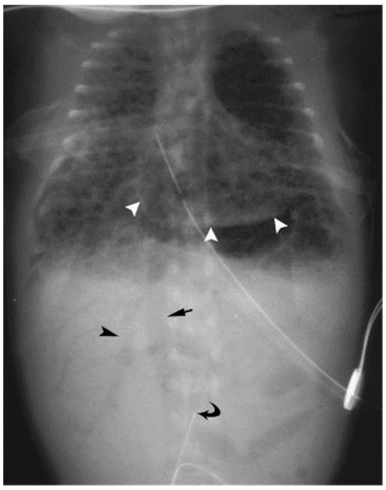
Pneumomediastinum



Extensive air leak



Systemic air embolism



Conclusions

- the natural history of lung disease in premature infants have changed due to advances in perinatal medicine
- radiology face new or perplexing expressions of once predictable disease
- interpretation of a preterm neonate chest radiograph requires appreciation of surfactant effect, impact of sophisticated ventilation and patterns of chronic lung disease.

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Thank you

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