



NCCU CLINICAL GUIDELINES
SECTION: 2

RESPIRATORY PROBLEMS AND MANAGEMENT

Section 2: Respiratory problems and management
Pulmonary interstitial emphysema (PIE)
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PULMONARY INTERSTITIAL EMPHYSEMA (PIE)

The pulmonary air leak syndromes (pneumomediastinum, pneumothorax, pulmonary interstitial emphysema and pneumopericardium) comprise a spectrum of diseases which have the same underlying pathophysiology - overdistention of alveolar sacs or terminal airways resulting in dissection of air into surrounding spaces.

PIE is the most common form of barotrauma associated with mechanical ventilation and is usually observed as a complication of RDS, although it also occurs in association with aspiration syndromes and sepsis. Whilst normally confined to intubated subjects, PIE has been described in an infant who only received nasal CPAP. PIE is important because of the significant pathophysiologic alterations it causes and that it commonly progresses to more severe forms of extra-alveolar gas leak.

EPIDEMIOLOGY

PIE is caused by a dissection of gas from the base of ruptured overdistended terminal airways or alveoli into the pulmonary interstitium. The incidence of PIE has decreased from over 30% to as low as 7% since the introduction of surfactant treatment for infants with RDS. In patients requiring conventional mechanical ventilation, the use of high rates and short inspiratory times (avoidance of end-inspiratory pressure hold) appears to reduce the incidence of gas leak, including PIE.

PATHOPHYSIOLOGY

Once rupture occurs, gas enters the adjacent interstitial tissue, where it can dissect along the perivascular (particularly pulmonary venous) sheaths to the hilum of the lung. When the volume of gas in the interstitium is large enough, the resulting perivascular pressure can obstruct the pulmonary circulation.

PRESENTATION

- May be sudden and profound or subtle
- Respiratory compromised with oxygen desaturation, asymmetrical chest wall movement / air entry, displaced apex beat, restlessness, air hunger/gasping
- Blood gas may show hypoxia, respiratory and/or metabolic acidosis
- Circulatory compromise
- Associated with preterm infants, resuscitation, MAS, RDS, positive pressure ventilation
- The development of mild PIE is rarely associated with demonstrable changes in cardiorespiratory function. When PIE is severe and diffuse there is deterioration of the respiratory status with hypoxemia and hypercarbia due to compression of the pulmonary vasculature by trapped gas.
- In a small, preterm infant trans-illumination of the chest may be indistinguishable from a large pneumothorax.

INVESTIGATIONS

- Chest X-ray: fine, bubbly appearance of lungs, sometimes with larger lucent areas.

MANAGEMENT

1. Prevent progression to other forms of gas leak by using the lowest effective airway pressures and avoid end-inspiratory hold (short inspiratory times).
2. Unilateral PIE - selective intubation of the mainstem bronchus on the unaffected side
 - Positioning of the infant with the involved side down may be beneficial.
 - Surgical resection of affected lobe may be considered when PIE has progressed to formation of large cysts.
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 - Randomized clinical trials comparing conventional mechanical ventilation with high-frequency ventilation have either failed to show a reduction in the incidence of PIE with high-frequency ventilation, or demonstrated effective treatment of PIE and/or reduce the incidence of PIE in infants with severe RDS.

COMPLICATIONS/PROGNOSIS

May resolve spontaneously, or progress to include other airleak phenomena, chronic lung disease.