

# X RAY

BY-Dr. Ritesh B. Patel



- Gas in the pleural space is termed "pneumothorax."
- A spontaneous pneumothorax is that which occurs in the absence of an external event.
- **PRIMARY SPONTANEOUS PNEUMOTHORAX**
- **Definition** — A primary spontaneous pneumothorax (PSP) is traditionally defined as a pneumothorax which presents without a precipitating external event in the absence of clinical lung disease.
- most affected patients have unrecognized lung abnormalities (mostly subpleural blebs) that likely predispose to pneumothorax .
- Thus, many experts believe that the distinction between pneumothorax in patients "without" lung disease (ie, pneumothorax with subpleural blebs, also known as PSP).
- Pneumothorax in patients with lung diseases (ie, SSP) is somewhat artificial, and that PSP and SSP may exist on either end of a continuum .

- **Epidemiology** — PSP is more common in males than females (roughly three to six times higher).
- **Pathogenesis and risk factors**
- **Subpleural blebs** — PSP is thought to be due to small apical subpleural blebs or bullae (ie, air sacs between the lung tissue and pleura) that rupture into the pleural cavity . The mechanism of bleb/bulla formation is unknown. PSP classically occurs in tall, thin males between the ages of 10 and 30 years .
- The development of subpleural blebs is thought to be due to either increasing negative pressure or greater mechanical alveolar stretch at the apex of the lungs during growth or a congenital phenomenon in which lung tissue at the apex grows more quickly than the vasculature, thereby outstripping its blood supply.
- **Smoking (cigarette, cannabis)** — Cigarette smoking (current or past) is a significant risk factor for PSP, probably due to airway inflammation and respiratory bronchiolitis.

- **Genetic predisposition** — Genetic variants associated with PSP include HLA haplotype A2B40, alpha-1 antitrypsin (M1M2 mutations), and fibrillin 1 (FBN1) mutations .
- The autosomal dominant Birt-Hogg-Dubé syndrome (BHD; due to mutations in the folliculin gene [especially c.1300G>C, c.250-2A>G] ), hyperhomocysteinemia , alpha-1 antitrypsin and Marfan syndrome are also inherited conditions associated with pneumothorax .

## • **SECONDARY SPONTANEOUS PNEUMOTHORAX**

- Secondary spontaneous pneumothorax (SSP) is defined as pneumothorax that presents as a complication of underlying lung disease
- SSP presents in older patients (>55 years).
- Among all of spontaneous pneumothorax, 61 percent of which were due to chronic obstructive pulmonary disease (COPD), has increased by 9 percent over a 48-year period from 1968 through 2016 [[37](#)]. Rates were higher in males than females (73 versus 27 percent).

## Causes of non-traumatic spontaneous pneumothorax

| Primary pneumothorax                          | Secondary pneumothorax                           |
|-----------------------------------------------|--------------------------------------------------|
| <b>Asthenic body habitus/subpleural blebs</b> | <b>Airway disease</b>                            |
|                                               | Cystic fibrosis                                  |
| <b>Drug use</b>                               | Asthma                                           |
| Cigarette smoking                             | COPD                                             |
| Snorting cocaine                              | <b>Infection</b>                                 |
| Smoking marijuana                             | <i>Pneumocystis jirovecii</i> ( <i>carinii</i> ) |
| <b>Increased transpulmonary pressure</b>      | Tuberculosis                                     |
| Valsalva maneuver                             | Necrotizing pneumonia                            |
| Diving, military flying                       | <b>Congenital lung disease</b>                   |
|                                               | Congenital pulmonary adenomatous malformation    |
|                                               | Congenital lobar emphysema                       |
|                                               | <b>Interstitial lung disease</b>                 |
|                                               | Sarcoidosis                                      |
|                                               | Langerhans cell granulomatosis                   |
|                                               | Other                                            |
|                                               | <b>Connective tissue/inflammatory disease</b>    |
|                                               | Marfan syndrome                                  |
|                                               | Ehlers-Danlos syndrome                           |
|                                               | Juvenile idiopathic arthritis                    |
|                                               | Polymyositis or dermatomyositis                  |
|                                               | Birt-Hogg-Dubé syndrome                          |
|                                               | Other                                            |
|                                               | <b>Malignancy</b>                                |
|                                               | Primary lung cancer                              |
|                                               | Metastatic disease                               |
|                                               | <b>Airway obstruction</b>                        |
|                                               | Foreign body aspiration                          |
|                                               | <b>Thoracic endometriosis</b>                    |
|                                               | Catamenial pneumothorax                          |

COPD: chronic obstructive pulmonary disease.

- **COPD** is the most common cause of SSP .
- Patients with COPD may also be at higher risk for iatrogenic pneumothorax (eg, venous catheterization, mechanical ventilation), particularly when there is a significant amount of underlying emphysema or air trapping.
- It is more common in person with FEV1 less than 1 lit and FEV1/FVC is < 40 %.
- **Cystic fibrosis:** The risk of pneumothorax in CF increases with the severity of lung function abnormalities.
- **Lung malignancy (primary and metastatic)**
- **Necrotizing lung infections** — SSP can complicate the course of necrotizing pneumonia due to *Pneumocystis jirovecii* (ie, pneumocystis pneumonia [PCP]), TB, bacteria, and less often fungi or other microorganisms, including severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2)
- **Pneumocystis jirovecii** – Unilateral and bilateral SSP can be seen in patients with PCP, most often in patients with human immunodeficiency virus (HIV) . In the era of antiretroviral therapy (ART), the frequency of pneumothorax less common .

- **Bacterial pneumonia** – SSP has been associated with bacterial pneumonias caused by *Staphylococcus*, *Klebsiella*, *Pseudomonas*, *Streptococcus pneumoniae*, and anaerobic organisms.
- SSP in the setting of bacterial pneumonia is more likely to be unilateral than bilateral and can be associated with extension of bacterial infection into the pleura and development of empyema, giving the appearance of a hydropneumothorax.
- **Tuberculosis** – SSP occurs in 1 to 3 percent of patients hospitalized with pulmonary TB . The pneumothorax is usually due to rupture of a tuberculous cavity into the pleural space.
- **Cystic lung disorders (other than emphysema)** — Pneumothorax is common in lung conditions associated with cysts.

|                                                                                                                                                                                                              |
|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Amyloidosis <sup>¶</sup>                                                                                                                                                                                     |
| Bronchopulmonary dysplasia (lung cysts likely a consequence of infection)                                                                                                                                    |
| Congenital/genetic (eg, <b>Birt-Hogg-Dubé syndrome</b> , Down syndrome, neurofibromatosis, Proteus syndrome)                                                                                                 |
| Erdheim Chester disease                                                                                                                                                                                      |
| Fire-eater's lung                                                                                                                                                                                            |
| Fungi (eg, coccidioidomycosis, <i>Pneumocystic jirovecii</i> )                                                                                                                                               |
| Hypersensitivity pneumonitis                                                                                                                                                                                 |
| Hyper immunoglobulin-E syndrome (due to recurrent staphylococcal pneumonia)                                                                                                                                  |
| Light chain deposition disease <sup>Δ</sup>                                                                                                                                                                  |
| <b>Lymphangiomyomatosis</b> (sporadic or related to tuberous sclerosis complex)                                                                                                                              |
| <b>Lymphoid interstitial pneumonia</b> <sup>◇</sup> (eg, associated with Sjögren's disease, immunodeficiency)                                                                                                |
| Paragonimiasis                                                                                                                                                                                               |
| Primary and metastatic tumors (eg, lung adenocarcinoma, metastatic gastrointestinal and genitourinary adenocarcinoma, lymphoma, mesenchymal cystic hamartoma, metastatic sarcomas, pleuropulmonary blastoma) |
| Recurrent respiratory papillomatosis (involving lung parenchyma in adult)                                                                                                                                    |
| Sjögren's disease (eg, due to Light chain deposition disease or lymphoid interstitial pneumonia)                                                                                                             |
| Smoking-related interstitial lung disease (desquamative interstitial pneumonia, <b>pulmonary Langerhans cell histiocytosis</b> , respiratory bronchiolitis interstitial lung disease)                        |
| Smoking-related small airways injury                                                                                                                                                                         |
| Staphylococcal pneumonia                                                                                                                                                                                     |

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- **Clinical presentation** – Pneumothorax (gas in the pleural space) should be suspected in patients with acute dyspnea and pleuritic chest pain, particularly when an underlying risk factor is present .
- •While young, thin, smoking males are more likely to have primary spontaneous pneumothorax (PSP; ie, that associated with subpleural blebs in the absence of an underlying disorder) and older patients are more likely to have secondary pneumothorax (SSP; ie, as a complication of an underlying lung disorder), this division is not absolute, and underlying disorders are not always readily apparent.
- •Arterial blood gas analysis may be normal but classically reveals hypoxemia and a respiratory alkalosis; acute hypercapnic respiratory acidosis can occur rarely and is an ominous sign.

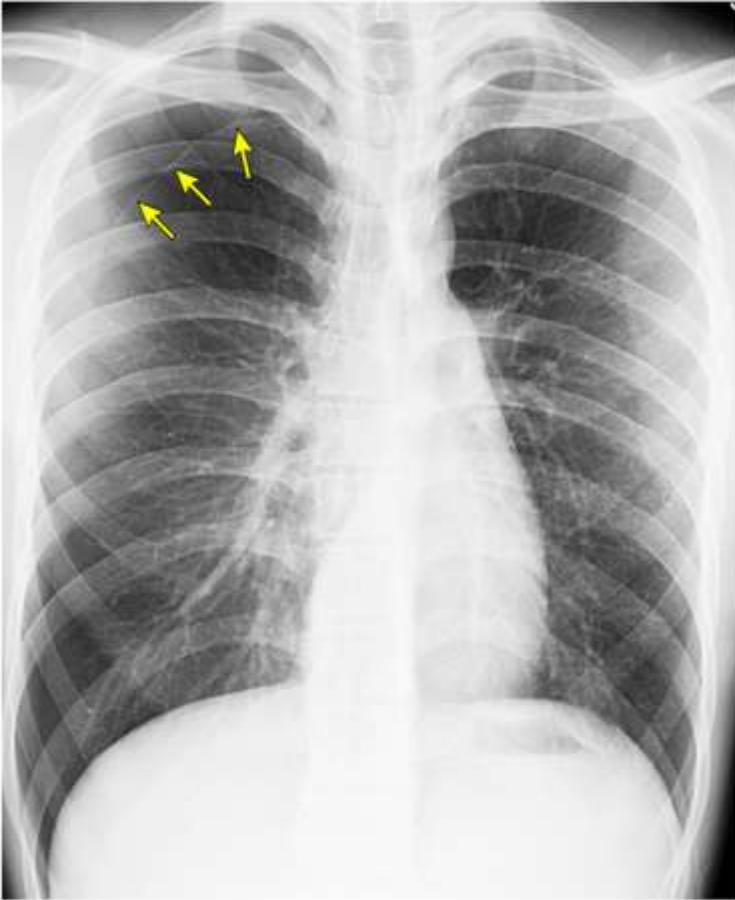
- **Pleural ultrasonography —**

- Ultrasound of the pleura is best utilized when bedside rapid imaging is needed to make the diagnosis of pneumothorax (eg, unstable patients with trauma, or patients with suspected tension) because ultrasound has been shown to be sensitive diagnostically and ultrasonography is more readily available with shorter wait times than for bedside chest radiography .
- It is also typically used for suspected pneumothorax that follows ultrasound-guided procedures (eg, thoracentesis or central venous catheterization) and is being increasingly used in critically ill patients.

- **Bedside chest radiography**

- **Chest CT** - most accurate method

Chest radiograph of spontaneous pneumothorax



Chest radiograph of a 20-year-old male with small spontaneous right pneumothorax demonstrates the characteristic convex right white visceral pleural line (arrows).

Courtesy of Nestor L. Muller, MD, PhD.

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- **Pneumothorax appearance and types** — Chest radiography (typically performed in the upright position) is the most common diagnostic imaging modality used for stable patients with suspected pneumothorax.
- The presence of a pneumothorax is established by demonstrating a white visceral pleural line on the chest radiograph. The visceral pleural line defines the interface between the lung and pleural air
- Bronchovascular markings are not typically visible beyond the visceral pleural edge unless the pneumothorax is loculated. The ipsilateral hemithorax size may be increased.

- **Chest radiography**
- **Unstable patients** — Hemodynamically unstable patients and patients with severe respiratory distress are typically those with a large or tension pneumothorax, patients with extensive trauma, or patients with significant underlying lung disease.
- Such patients are resuscitated with the emphasis on stabilization of the airway, breathing, and circulation.
- Unstable patients should also concomitantly undergo rapid bedside imaging, usually initially with ultrasound, to confirm the diagnosis before undergoing emergent needle or chest tube thoracostomy.
- **Stable patients** — Most patients suspected of having a pneumothorax who are hemodynamically stable and/or not in severe respiratory distress should undergo routine bedside chest radiography in the upright position.

