INTRODUCTION

Definition of pneumothorax:

Pneumothorax is the accumulation of extra-pulmonary air within the chest, most commonly from leakage of air from within the lung.[1]

Etiology:

Pneumothorax can be spontaneous, traumatic, iatrogenic or catamenial.[2]

Primary spontaneous pneumothorax occurs without trauma or underlying lung disease. Examples include those in teenagers and young adults, most frequently in males who are tall.[2]

Secondary spontaneous pneumothorax occurs when there is an underlying lung disease. Examples are those occurring with pneumonia, lung abcess, infarcts, asthma, foreign bodies in the lung , cystic fibrosis , malignancies involving the lung and graft versus host disease with bronchiolitis obliterans .[2]

Traumatic pneumothorax can occur due to chest or abdominal blunt trauma or penetrating trauma tearing a bronchus or abdominal viscus with leakage of air into the pleural space.[2]

Iatrogenic pneumothorax can result from needle aspiration of the pleural space, tracheostomy, subclavian line placement, thoracentesis, trans-bronchial biopsy, mechanical or non- invasive ventilation or other procedures.[2]

Catamenial pneumothorax is related to menses and is associated with diaphragmatic defects and pleural blebs: it is uncommon.[2]

Pneumothorax is usually unilateral; bilateral pneumothorax is seen in neonatal period and rarely in Mycoplasma Pneumoniae infection, tuberculosis and after lung transplantation. Pneumothorax may have associated pleural fluid which can be serous (hydrothorax), pus (pyopneumothorax) or blood (hemopneumothorax).[2]

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Table (1): Causes of pneumothorax in children).[1]

Spontaneous
• Primary idiopathic—usually resulting from ruptured subpleural blebs
• Secondary blebs
• Congenital lung disease:
− Congenital cystic adenomatoid malformation − Bronchogenic cysts − Pulmonary hypoplasia*
• Conditions associated with increased intrathoracic pressure:
− Asthma − Bronchiolitis − Air-block syndrome in neonates − Cystic fibrosis − Airway foreign body
• Infection:
− Pneumatocele − Lung abscess − Bronchopleural fistula
• Diffuse lung disease:
− Langerhans cell histiocytosis − Tuberous sclerosis − Marfan syndrome − Ehlers-Danlos syndrome
• Metastatic neoplasm—usually osteosarcoma (rare)
Traumatic
• Non-iatrogenic
− Penetrating trauma − Blunt trauma − Loud music (air pressure)
• Iatrogenic
− Thoracotomy − Thoracoscopy, thoracentesis − Tracheostomy − Tube or needle puncture − Mechanical ventilation

* Associated with renal agenesis, diaphragmatic hernia, amniotic fluid leaks.

Pathogenesis:

Although intra-pleural pressures are negative throughout most of the respiratory cycle,[3] air does not enter into the pleural space because the sum of all partial pressures of gases in the capillary blood averages only 93.9kPa (706 mmHg). Hence, net movement of gases from capillary blood into the pleural space would require pleural pressure lower than -54 kPa (i.e lower than -36 cmH_2o) which hardly never occur in normal circumstances.[4] Hence if air is present in the pleural space one of three events must have occurred: 1) communication between alveolar spaces and pleura ; 2) direct or indirect communication

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between atmosphere and pleural space ; 3) presence of gas producing organisms into the pleural space .[4,5]

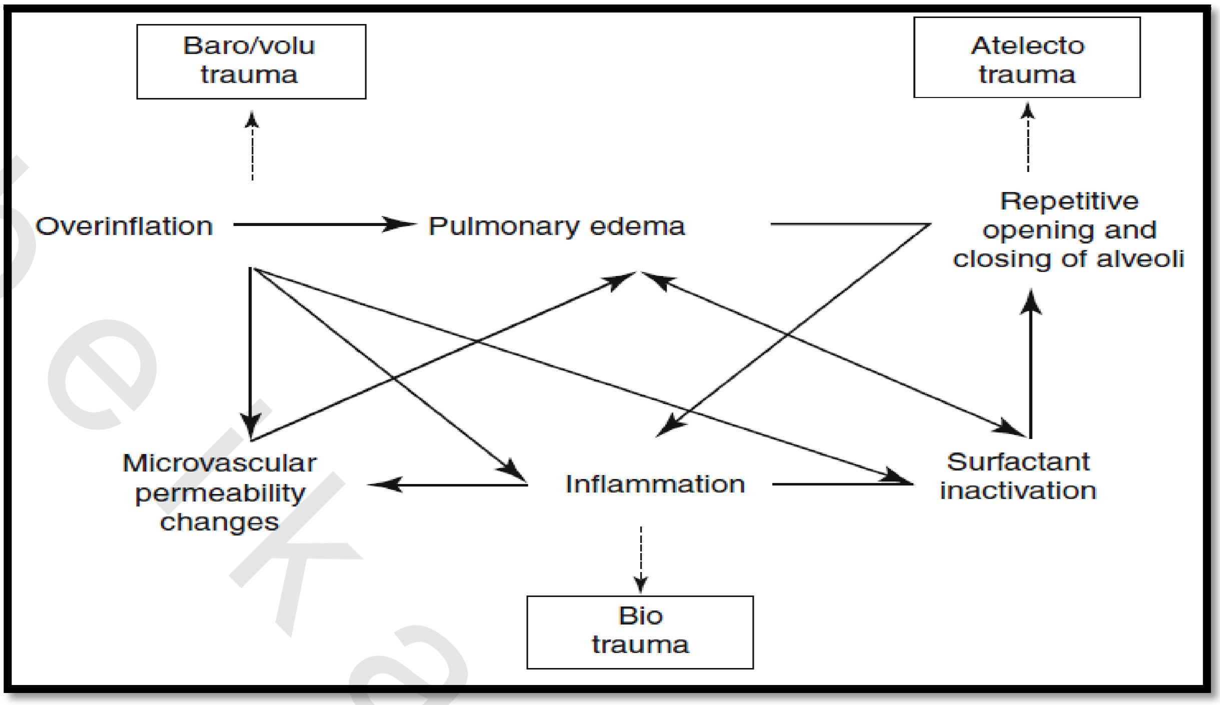

Figure (1): Pathophysiological mechanisms of ventilator induced lung injury. Three major mechanism lead to ventilator induced lung injury (1) barotrauma/volutrauma. (2) atelectotrauma. (3) biotrauma. There is a reinforcing interaction between the different mechanisms leading to a number of vicios circles.[6]

Pneumothorax can be classified in simple, communicating or tension pneumothorax. A simple pneumothorax occurs when there is accumulation of air without any communication to the atmosphere and without causing a shift of the mediastinum. A communicating pneumothorax (sucking chest wound) occurs when there is an associated defect in the chest wall. This defect may cause paradoxical chest wall movement (collapse during inhalation and expansion during exhalation) along with the sonorous sound of air entering and exiting the wound. A tension pneumothorax occurs when the progressive accumulation of air causes a shift of the mediastinum to the opposite hemithorax causing a subsequent compression of the contralateral lung and great vessels. Communicating and tension pneumothoraces may result in rapid onset of hypoxia, acidosis and shock.[7]

The tendency of the lung to collapse, or elastic recoil, is balanced in the normal resting state by the inherent tendency of the chest wall to expand outward, creating negative pressure in the intra-pleural space. When air enters the pleural space, the lung collapses. Hypoxemia occurs because of alveolar hypoventilation, ventilation-perfusion mismatch, and intrapulmonary shunt. In simple pneumothorax, intra-pleural pressure is atmospheric, and the lung collapses up to 30%. In communicating or tension pneumothorax, continuing leak causes increasing positive pressure in the pleural space, with further compression of the lung, shift of mediastinal structures toward the contralateral side, and decreasing in venous return and cardiac output.[1]

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Incidence, risk factors and mortality:

In the United States the incidence of spontaneous pneumothorax is approximately 7.4 to 18 cases per 100000 boys and 1.2 to 6 cases per 100000 girls, with male to female ratio of about 2:1.[7] This ratio appears to be reversed, however, below the age of 9 years.[9] Mean age of presentation has been reported to be 14 to 15.9 years, with one pediatric cohort reporting mean age at the diagnosis of 13.8 years.[10,11] It appears to present most typically in tall thin males with low body mass index. Mortality is considered low in children.[8]

Although the incidence of secondary pneumothorax in children is not well described, those with asthma and cystic fibrosis are considered at particular risk.[12,13] The probability of pneumothorax is thought to increase as the lung function decreases , and the mortality is considered higher because of decreased reserve of the diseased lung.[14] Infectious causes such as Pneumocystitis jirovecii pneumonia in immune disorders and necrotizing pneumonias (anaerobic gram negative or staphylococcal) are associated with higher incidence of pneumothorax.[14] Individuals with underlying connective tissue disease, such as Marfan Syndrome, Ehlers – Danlos, and ankylosing spondylitis, are also at a higher risk.[15]

In the setting of thoracic trauma, pneumothorax occurs in one third of pediatric cases, with the majority of these having associated intra-thoracic and extra-thoracic injuries, and only one third occurring in isolation.[16]

Iatrogenic pneumothorax is an unfortunate consequence of medical procedures. A recent review found that 57% of the procedures that lead to pneumothorax at teaching hospital were performed under emergent conditions. The most frequent procedure types were central venous catheterization (43.8%) of iatrogenic pneumothorax, thoracentesis (20.1%) and barotrauma due to mechanical ventilation (9.1%).[17] The internal jugular and subclavian approaches to central venous access are considered to have the highest risk of pneumothorax.[18] A systemic review reported an overall incidence of pneumothorax resulting from thoracentesis of 6%, with 34.1% requiring chest tube insertion.[19] The introduction of real- time ultrasound guidance has been shown to reduce the incidence of pneumothorax for both central venous access and thoracentesis in adults .[20,21] A recent meta-analysis of the pediatric literature of ultrasound guided central venous access, however, failed to show any statistical difference in adverse outcome.[22]

Clinical features:

Spontaneous pneumothorax usually presents with sudden onset of pleuritic chest pain, shortness of breath and occasionally dry cough. The onset is usually at rest and there is no association with exercise. Older children may describe the initial chest pain as sharp and latter as steady ache. In secondary spontaneous pneumothorax, other symptoms may occur related to the underlying disease process e.g. pyogenic lung infection. In most cases symptoms resolve within 24 hours even in the absence of treatment. In case of asthmatics, shortness of breath without chest pain appears to be the most common presentation. Rarely there may be a family history of pneumothorax.[23]

Clinical findings range from normal to acute respiratory distress. A simple pneumothorax occupying less than 15% of thoracic cavity is difficult to detect on clinical examination. [14] The size of pneumothorax, however, correlates poorly with the clinical manifestations. Chest wall movement may be reduced. Percussion of the affected thorax may be hyper-resonant. Auscultation may demonstrate decreased breath and heart sounds. Surgical emphysema is a rare finding. The presence of marked tachycardia, hypotension, cyanosis, neck vein engorgement, contralateral tracheal deviation, or a cardiac arrest with electro-mechanical dissociation suggests pneumothorax under tension .[(24)]

The severity of respiratory distress of a child with a pneumothorax depends on their age, pulmonary reserve, and the etiology of pneumothorax and whether the pleural gas is under tension. As a general rule, signs and symptoms are more marked in secondary spontaneous pneumothorax where the pulmonary reserve is lower. [(24)] Typically infants present respiratory distress with marked subcostal recession, intercostal indrawing and widespread accessory muscle use particularly in the neck. A high proportion of infant pneumothoraces present under tension. In a child receiving positive pressure ventilation, the onset of pneumothorax will be heralded by an abrupt deterioration of gas exchange and hypotension.[(25)]

Diagnosis of pneumothorax:

The diagnosis of pneumothorax in critical illness is established from patient's history, physical examination and radiological investigations. The factors related to the underlying lung disease are important in the history. Examination of respiratory and cardiovascular systems may help establish the diagnosis of pneumothorax. However, examination findings may vary according to the size of pneumothorax and the presence of limited cardiorespiratory reserve, with patients with small pneumothorax (one involving <15% of the hemi-thorax) possibly having normal physical examination.[(26)]

Careful inspection and repeated auscultation of the chest is therefore crucial. Contralateral tracheal deviation, hyper-resonance percussion over the chest and decreased breathing sounds might be noted. Reduction in tidal volumes during pressure controlled ventilation and increased airway pressure with volume controlled ventilation might be found. A pulsus-paradoxus on the arterial trace and increased central venous pressure from central venous catheterization may be observed .[(26-29)]

Arterial blood gas measurements are frequently abnormal in patients with pneumothorax, with arterial oxygen tension (PaO_2) being less than 10.9 kPa (80 mmHg) in 75% of patients.[(30)] The presence of underlying lung disease along with the size of pneumothorax predicts the degree of hypoxemia.[(30)] Arterial PaO_2 was below 7.5 kPa (55 mmHg) and arterial carbon dioxide tension ($PaCO_2$) above 6.9kPa (50mm Hg) in 16% of cases of secondary pneumothorax in the largest reported series.[(31)] Pulmonary function tests are weakly sensitive measures of the presence or size of pneumothorax and are not recommended.[(32)]

It should be noted that many of the above findings are nonspecific and have not been a reliable indicator of pneumothorax. The radiological data thus remains the gold standard for diagnosis of pneumothorax. [(27)]

Chest X ray:

Chest radiographs have traditionally been the first test ordered for suspected pneumothorax, but in the era of bedside ultrasonography this is no longer the standard of

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care.[33] In the critically ill, the traditional postero- anterior expiratory film is not practical and thus the supine or semi –recumbent anterior-posterior film is frequently obtained. [34] Expiratory chest radiographs were recommended previously in cases in which small pneumothorax was suspected and not confirmed by standard views, however, these do not increase the diagnostic yield and are especially challenging to obtain correctly in the ventilated patients.[35]

Radiologic findings of a pneumothorax in the supine patient may present in several ways. In addition to the classic findings of lung collapse in or around apices, in the supine position air can have a propensity to collect in a subtle fashion along the anterior space without a clear lung edge finding. As additional air accumulates, it can track further around the chest and result in the classic deep sulcus sign.(Figure :2)[36]

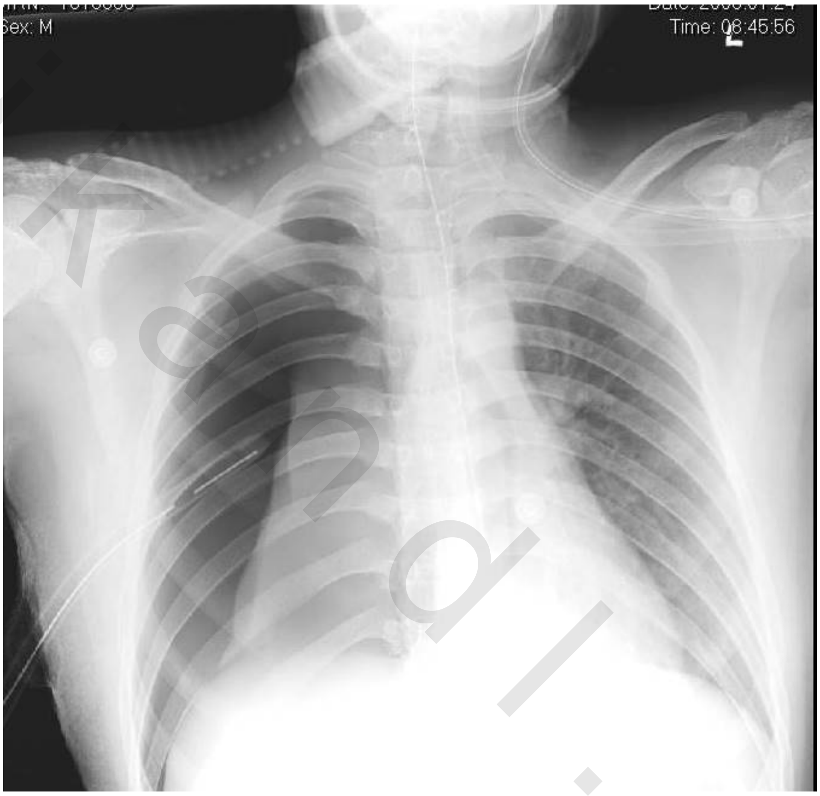

Figure (2): Deep sulcus sign revealing large right pneumothorax.

Thoracic Ultrasonography;

Ultra -sonographic assessment of pneumothorax has emerged recently as the standard of care in facilities with physicians trained in bedside ultra-sonography.[37,38] Ultrasonography has been shown to have high sensitivity (95%), specificity (100%) and diagnostic effectiveness (98%) for pneumothorax when compared with computed tomography (CT) as standard. It is useful for detection of small collections not seen on plain films and the extent of the air collection can be estimated by tracking the presence of the “sliding lung sign” over the chest wall.[39]

There are several advantages of ultrasonography over standard chest radiography and CT scanning, including the absence of radiation, portability, real-time imaging, and the

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ability to easily perform dynamic and repeat evaluations. As a result, when a critical care ultrasonography-trained physician is available, ultrasonography is the preferred first-line diagnostic test to exclude pneumothorax in the ICU.(28)

A study comparing ultrasonography to CT scan and chest radiographs for the diagnosis of occult pneumothorax showed that the use of ultrasonography detected 92% of occult pneumothoraces diagnosed with CT scan(40). Ultrasonography can also be used to confirm resolution of a pneumothorax after pleural drainage, is more sensitive then chest radiography, and allows for rapid reassessment (41) Ultrasonography has been used to rule out pneumothorax after central line placement, as well as after transbronchial biopsy. (42,43) Though there is a learning curve associated with the use of chest ultrasonography, it is relatively short.(44)

CT Scanning:

CT scan is the gold standard test for both the diagnosis and sizing of pneumothorax.(45) The technology is particularly useful in patients with significant underlying lung disease, which may obscure chest radiological findings. Specifically, CT scanning is an excellent tool to differentiate bullous lung diseases from pneumothorax, which may help avoid unnecessary drainage attempts that may result in the creation of parynchemal -pleural fistula.(46)Pneumothorax size can best be calculated with CT imaging. However, the size of pneumothorax is less important than the degree of clinical severity.(47)

Management of pneumothorax:

The cause of the pneumothorax as well as the patient's underlying disease, greatly influence the treatment course and overall prognosis in critically ill patients.(48)The primary goal during the management of a pneumothorax is to evacuate air from the pleural space and allow apposition of the lung to the chest wall. Although many patients with pneumothorax can be managed conservatively, the majority of patients in the ICU require pleural intervention.(49) The cause of pneumothoraces in critically ill patients has been shown to correlate with mortality risk. Pneumothorax secondary to barotrauma, progression to tension pneumothorax, and concurrent sepsis in a patient with pneumothorax have been significantly and independently associated with an increased risk of death in the ICU.(50) The recent pleural management guidelines by the British Thoracic Society (BTS) recommend intercostal drainage for all pneumothoraces in patients on mechanical ventilator, as well as, those exhibiting signs or suspicion of tension physiology, traumatic pneumo-or hemothorax, or post-surgical pneumothoraces .(51)

General:

Observation should be the treatment of choice for primary spontaneous small closed pneumothoraces without significant breathlessness, in a spontaneously breathing patient. Inhalation of high concentrations of oxygen may speed the resolution of a pneumothorax by reducing the partial pressure of nitrogen in the pulmonary capillaries. This should increase the pressure gradient between the pleural cavity and pleural capillaries, so increasing the absorption of air from the pleural cavity. The rate of re-absorption of spontaneous pneumothoraces is 1.25–1.8% of the volume of hemithorax every 24 h.(52)

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Symptomatic patients should not be left without intervention regardless of the size of the pneumothorax on a chest radiograph. Other considerations necessitating intervention include the need for positive pressure ventilation, planned anesthesia and surgery (nitrous oxide diffuses into air collections and increases pressure/volume), transport in or outside the hospital, and altitude changes (including air transport).(39)

Needle aspiration and intercostal chest catheter insertion:

There is an emerging body of evidence that, in adult patients with primary spontaneous pneumothorax (PSP) a single needle aspiration is not inferior to intercostal chest catheter (ICC) insertion with respect to success and recurrence rates. Moreover, needle aspiration is less invasive, more cost effective and associated with lower complication rates compared to (ICC). In the British Thoracic Society (BTS) guidelines for adult patients, it is recommended that aspiration of a maximum of 2.5 L should not be exceeded in order to avoid re-expansion pulmonary edema. Control radiographs are suggested after single aspiration to assess the presence of ongoing air leak.(53)

ICC insertion should be considered in case of:

- Age <1 year.
- Bilateral pneumothorax.
- Tension pneumothorax.
- Evidence of a big air leak.
- Pneumothorax recurrence within the first hours following aspiration.
- Co-existence of a pleural effusion especially in case of hemothorax. (53)

Furthermore, chest drain insertion should be performed in all children with iatrogenic and traumatic pneumothorax, secondary spontaneous pneumothorax (SSP) and co existing pneumo-mediastinum.(53)

The success rates of small bore ICCs are comparable to large bore ICCs, while being less painful; however, large bore ICCs are indicated when the rate of air leak exceeds the capacity of smaller ICCs. Initial chest radiograph imaging should guide the site of placement. An appropriate approach in the majority of cases is the "triangle of safty" which is bordered anteriorly by the lateral edge of pectoralis major, laterally by the lateral edge of latissmus dorsi, inferiorly by the line of the fifth intercostal space and superiorly by the base of the axilla. An alternative approach is the second intercostal space in the mid-clavicular line. Proper sedation should be given in addition to local anesthetic. After insertion, ICCs should be connected to Heimlich valve or under-water seal device.(53) Chest drain should not be removed until bubbling has ceased, and chest radiography demonstrated lung re-inflation. There is no evidence that clamping chest drain at the time of its removal is beneficial.(39)

Prevention of recurrence:

- **Surgical management:**

There are two objectives for surgical treatment of spontaneous pneumothorax. The first is to treat the underlying cause which is invariably rupture of sub-pleural bleb or bullous. This can be achieved by stapled or sutured resection.[54] The second objective is to prevent recurrence by inducing pleural symphysis either by mechanical abrasion or pleurectomy .On the basis of statistical risk of recurrence, the indications for operative intervention in spontaneous pneumothorax are as follows:

- Second ipsilateral spontaneous pneumothorax.
- First contra -lateral pneumothorax.
- Bilateral spontaneous pneumothorax.
- Persistent air leak (PAL) after 5-7 days of tube drainage
- Professions at risk as pilots and divers.[25]

- **Chemical pleurodesis:**

The instillation of many substances in the pleural space leads to aseptic pleural inflammation, dense adhesions and ultimately pleural symphysis. Although chemical pleurodesis is effective in preventing recurrent pneumothoraces in adults, use in children is limited. If this technique is to be used in children, it is desirable that future thoracotomy could be possible. Talc renders subsequent thoracotomy exceedingly difficult and should only be used in small doses. [55] Tetracycline is the recommended first line sclerosant for both PSP and SSP.[25] Chemical pleurodesis is far more painful than thoracotomy. The value of chemical pleurodesis for PSP is questionable given the potential for adverse effects and the well documented superiority of surgical pleurodesis. As a consequence, most pediatric surgeons consider the chemical pleurodesis has no role in the management of recurrent pneumothorax in children.[56]