Management of primary spontaneous pneumothorax: a review

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Abstract
Primary spontaneous pneumothorax is a common problem faced by doctors in medical practice. It is a significant global health problem affecting adolescent and young adults. This article will review the etiology, diagnosis and current management guidelines. It aims to improve clinical practice and compliance to the complexities of procedures involved in management.

Introduction
Pneumothorax is a thoracic disorder of an abnormal collection of air in the pleural space. It is most commonly associated with emphysema, asthma and tuberculosis. It may be spontaneous (not caused by trauma or any obvious precipitating factor), traumatic or iatrogenic. Primary spontaneous pneumothorax (PSP) occurs in young, otherwise, healthy adults without clinically apparent lung disease; secondary spontaneous pneumothorax (SSP) is a complication of pre-existing lung disease, most commonly chronic obstructive pulmonary disease (COPD). This article reviews the current management of PSP in the adult population.

Prevalence
The annual incidence of PSP is 22.7 per 100 000 and a sex ratio of 1:3.3 (women: men). It typically occurs in tall thin boys and men between the ages of 10 and 30 years, with peak age in the early 20s and rarely in patients over the age of 40. Recurrences range from 25 to 50%, with most recurrences occurring within the first year. Female gender, tall stature in men, low body weight and failure to stop smoking are associated with an increased risk of recurrence.

Risk factors
Factors that predispose patients to PSP include smoking, family history, Marfan syndrome, homocystinuria and thoracic endometriosis. Cigarette smoking is a significant factor for PSP, likely due to consequences of airway inflammation. The risk of PSP is directly related to the amount of cigarette smoking. Respiratory bronchiolitis, a form of airway inflammation associated with cigarette smoking, may contribute to the development and recurrence of PSP.

An uncommon cause and an associated increased incidence of PSP is Birt–Hogg–Dubé syndrome, an autosomal dominant condition which predisposes patients to benign skin tumours and renal cancers. The presence of lung cysts in these patients, as well as lung cyst volume, diameter and number are also associated with pneumothoraces. There is an increased frequency of PSP in patients with Marfan syndrome and homocystinuria. Catamenial pneumothorax may result from thoracic endometriosis and should be considered in women presenting with PSP temporally related to menstruation. Malnutrition in patients with anorexia nervosa may contribute to the development of PSP.

Pathophysiology
PSP is due to rupture of small, peripheral, usually apical subpleural blebs and usually subsides spontaneously as the air is resorbed. Although PSP is defined as occurring in patients without underlying pulmonary disease, many of these patients have asymptomatic blebs and bullae detected on computed tomography (CT) scans or during thoracic surgery. The mechanism of bulla formation is...
speculative. A plausible explanation is degradation of elastic fibres in the lung occurs, induced by the smoking-related influx of neutrophils and macrophages. This degradation causes an imbalance in the protease-antiprotease and oxidant-antioxidant systems. After bullae have formed, inflammation-induced obstruction of the small airways increases alveolar pressure, resulting in an air leak into the lung interstitium. The air then moves to the hilum, causing pneumothorax. As mediastinal pressure rises, rupture of the mediastinal parietal pleura occurs, causing pneumothorax.10

PSP is typically observed in tall young people without parenchymal lung disease and is thought to be related to increased shear forces in the apex. Recurrent attacks are common and can be quite disabling. Hypoxaemia occurs despite normal perfusion due to collapsed and poorly ventilated areas of lung. However, hypocapnia is unusual because underlying lung function is relatively normal, and adequate alveolar ventilation can be maintained by the contralateral lung. Acute respiratory alkalosis may exist due to pain, anxiety and/or significant hypoxaemia.

PSP can cause compression, collapse and atelectasis of the lung and may be responsible for marked respiratory distress. When the defect acts as a flap valve and permits the entrance of air during inspiration but fails to permit its escape during expiration, it effectively acts as a pump that creates the progressively increasing pressures of tension pneumothorax, which may be sufficient to compress vital mediastinal structures and the contralateral lung.

Clinical presentation

PSP often develops when the patient is at rest. Patients usually complain of a sudden onset of dyspnoea and pleuritic chest pain. The severity of symptoms is primarily related to the volume of air in the pleural space, with dyspnoea being more prominent if the pneumothorax is large. Characteristic physical findings when there is a large pneumothorax include reduced chest excursion on the affected side, diminished breath sounds and hyperresonant percussion. Subcutaneous emphysema may be present. Evidence of laboured breathing and haemodynamic compromise (e.g. tachycardia and hypotension) and deviation of trachea to the contralateral side suggest a possible tension pneumothorax which warrants emergency decompression.

Imaging

The presence of a pneumothorax is established by demonstrating a white visceral pleural line on the chest X-ray (CXR). The visceral pleural line defines the interface of the lung and pleural air; it is either straight or convex towards the chest wall. In most cases, no pulmonary vessels are visible beyond the visceral pleural edge; and there may be flattening or inversion of the diaphragm on the affected side. Inspiratory and expiratory films have equal sensitivity in detecting pneumothoraces; hence, a standard inspiratory CXR is sufficient in most cases.11

Lung sonography has rapidly emerged as a reliable technique in the evaluation and diagnosis of a pneumothorax.12 It is most commonly used in situations where a diagnosis must be made emergently by the bedside, such as for an intensive care unit patient or a trauma patient in the emergency department. Sonographic signs include ‘lung sliding’, ‘B-lines’ or ‘comet tail artefacts’, ‘A-lines’ and ‘the lung point sign’.13 Ultrasound has 81% sensitivity and 100% specificity, which is higher than an upright CXR for the detection of a pneumothorax.13

The underlying lung parenchyma should be examined for the presence of underlying lung disease that would suggest a SSP. CT scanning is not recommended routinely unless abnormalities are noted on the CXR or underlying lung disease is suspected. CT may be useful when an abnormal chest tube placement or a loculated pneumothorax is suspected.

Diagnosis

The diagnosis of PSP is suggested by the patient’s history and confirmed by detection of a visceral pleural line on the CXR without underlying lung disease. Differential diagnosis includes the causes of spontaneous pneumothorax in patients who have undiagnosed underlying lung disease (e.g. COPD, interstitial lung disease, lung cancer, cataminal pneumothorax and lymphangioleiomyomatosis). In patients with a persistent air leak or recurrent spontaneous pneumothorax, a CT scan should be considered, as well as a lung biopsy at the time of thoracoscopy.

Management

Current treatment management guidelines for PSP are variable, with sparse evidence from randomized controlled trials. According to the American College of Chest Physicians (ACCP), the British Thoracic Society (BTS) and the Spanish Society of Pulmonology and Thoracic Surgery (SEPAR), the initial management of PSP is directed at removing air from the pleural space with subsequent management directed at preventing recurrence and smoking cessation.9,14,15 Treatment options include observation, supplemental oxygen, needle aspiration of intrapleural air, chest tube insertion (i.e. tube thoracostomy) and thoracoscopy. The choice of procedure depends on patient characteristics and clinical circumstances.

A current trial being undertaken aims to determine if conservative management in PSP is effective and safe compared with invasive management, and whether it reduces the risk of recurrence.16 One possible mechanism for a lower recurrence rate with conservative management is that healing of the pleural defect may be facilitated by allowing the lung to stay collapsed initially followed by slow re-expansion (Fig. 1).17

Minimal symptoms

Patients who are clinically stable and having their first episode of PSP can be administered supplemental oxygen and observed if they are not breathless and their pneumothorax is small (≤2–3 cm between the lung and chest wall on a CXR).18 Observation should last 6 hours, and supplemental oxygen given to facilitate the resorption of the pleural air. If a repeat chest radiograph excludes progression of the pneumothorax after this time, reliable patients with ready access to emergency medical services can be discharged home with outpatient follow-up in 2–4 weeks.
Symptomatic pneumothorax

Patients who are clinically stable and having their first PSP should undergo needle aspiration if their pneumothorax is large (>3 cm rim of air on CXR) or if they are symptomatic with chest pain or dyspnoea. Marked breathlessness in a patient with a small PSP may herald tension pneumothorax.

Several methods are used to perform simple aspiration, ranging from venous catheters to small-bore chest tubes that can be removed once re-expansion of the lung is confirmed. Needle (14–16-G) aspiration is as effective as large-bore (>20 Fr) chest drains and may be associated with reduced hospitalization and length of stay. Aspiration is most easily accomplished with a commercially available thoracentesis kit. An 18-G needle with an 8 to 9 Fr catheter is inserted into the pleural space; the catheter is threaded deeper into the pleural space; and then the needle is withdrawn. Air is manually withdrawn through the indwelling catheter until no more can be aspirated. Once no further air can be aspirated from the pleural space, a Heimlich (i.e. one-way) valve or a closed stopcock can be attached, and the indwelling catheter secured to the chest wall. Those with a Heimlich valve can be discharged home with follow-up within 2 days. Those with a stopcock should have a CXR 4 hours later and, if adequate lung expansion has occurred, the catheter can be removed. Following an additional 2 hours of observation, another CXR should be performed. If the lung remains expanded, the patient can be discharged.

It is assumed that there is a persistent air leak if there is still no resistance after 4 L of air has been aspirated and the lung has not expanded. In that situation, thoracoscopy should be performed. Alternatively, if thoracoscopy is not readily available, a chest tube should be inserted. One of the advantages of aspiration over tube thoracostomy is that the patient need not be hospitalized, whether the catheter is removed after the aspiration or left attached to the Heimlich valve.

When to insert a chest tube (tube thoracostomy)

Patients who are clinically stable with recurrent PSP, a concomitant haemothorax (i.e. a haemopneumothorax), bilateral pneumothoraces

Fig. 1. Flowchart of management of spontaneous pneumothorax (adapted from MacDuff et al. with permission).

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or who are clinically unstable should undergo chest tube insertion. Most patients with PSP can be managed successfully with a small chest tube (≤22 Fr) or chest catheter (≤14 Fr). If the patient is anticipated to have a bronchopleural fistula with a large air leak or requires positive-pressure ventilation, a 24–28 Fr standard chest tube may be used. The chest tube can be connected to a water seal device, with or without suction and left in position until the pneumothorax resolves. If the pneumothorax fails to resolve with a chest tube, suction can be applied. Suction is thought to remove air from the pleural cavity at a rate that exceeds the egress of air through the breach in the visceral pleura and to subsequently promote healing by apposition of the visceral and parietal pleural layers. Optimal suction pressures are −10 to −20 cm H₂O (compared with normal intrapleural pressures of between −3.4 and −8 cm H₂O, according to the respiratory cycle), with the capacity to increase the air flow volume to 15–20 L/min. Applying suction too early after chest tube insertion may precipitate re-expansion pulmonary oedema, especially in patients with PSP that may have been present for more than a few days.

Chest drain insertion has a number of important complications such as injury to organs, bleeding and infection. Surgical emphysema is a complication of chest drainage and occurs in the context of a malpositioned, kinked, blocked or clamped chest drain. It can also occur with an imbalance between a large air leak and a relatively small-bore chest drain. It usually subsides spontaneously after a few days, but it may occasionally lead to acute airway obstruction or thoracic compression leading to respiratory compromise. A tracheostomy, skin incision decompression and insertion of a large-bore chest drain may be required.

Once the air leak has resolved, the lung has expanded and the pleural air has been removed, the chest tube can be removed. After 12 hours of last evidence of an air leak, the chest tube is clamped and, after an additional 12 hours, a chest radiograph is obtained. Some prefer not to clamp the chest tube because this might lead to the development of a tension pneumothorax. However, if the tube is removed, a tension pneumothorax can still develop, and it is more difficult to treat if there is no chest tube in place. The chest tube can be removed if the pneumothorax has not reaccumulated.

Patients with recurrent PSP or a concomitant haemothorax (i.e. a haemopneumothorax) should be referred to a thoracic surgeon for thoracoscopy after chest tube insertion. If thoracoscopy is not readily available, chemical pleurodesis through the chest tube may be performed after drainage of the pleural space to prevent recurrence.

### Urgent decompression

If chest tube insertion is delayed in a clinically unstable patient, decompression of the pleural space can be performed using a 14-G intravenous catheter into the pleural space. A catheter or needle as long as 7 cm may be needed, depending on the thickness of the chest wall. The insertion site has traditionally been in the second or third intercostal space (ICS) at the midclavicular line, but chest wall thickness at that site may prevent access to the pleural space. Alternatively, an insertion site in the fifth ICS in the anterior axillary line or anterior to the mid-axillary line may be used.

### Persistent air leak

In patients whose lung is at least 90% expanded with a persistent air leak, treatment options include attaching a Heimlich valve to the chest tube, infusing autologous blood into the pleural space and performing video-assisted thoroscopic surgery (VATS) to oversew the area of leak and perform mechanical pleurodesis. Attaching a Heimlich valve to the chest tube usually allows rapid discharge of the patient with subsequent outpatient management. An autologous blood patch involves withdrawal of blood from a peripheral arm vein and aseptic infusion of the blood without anticoagulants into the pleural space through the chest tube. The ideal amount of blood to infuse into the pleural space is not known but usually ranges from 24 to 200 mL. After infusion of the blood, the tubing from the chest tube is draped over a hook approximately 60 cm above the patient’s chest and then down to a water seal device on the floor. The chest tube is removed 24 hours after cessation of the air leak. The blood patch technique was demonstrated to be effective in 92% of cases, but the main side effect was empyema.

For patients who have a persistent air leak and whose lung is less than 90% expanded, referral to a thoracic surgeon should be made for VATS. VATS is effective in the treatment of spontaneous pneumothorax as well as prevention of recurrent pneumothorax. With this procedure, pleurodesis is created by pleural abrasion or a partial parietal pleurectomy. When necessary, an endoscopic stapler can be used to resect bullae. Other options for eliminating bullae include electrocoagulation, laser ablation and hand sewing. The rate of recurrent pneumothorax is less than 5% after VATS with bleb/bullae resection and pleurectomy. Other techniques reported to induce pleural symphysis include parietal pleurectomy, intrapleural instillation of talc or a tetracycline derivative, laser ablation of the parietal pleura, cellulose mesh with fibrin glue and pleural abrasion with dry gauze. Thoracoscopy has essentially replaced open thoracotomy in the management of spontaneous pneumothorax for two main reasons: hospitalization is shorter and post-operative pain is less.

Open thoracotomy and pleurectomy is recommended if thoracoscopy is unavailable or has failed. Apical pleural blebs are oversewn, and the pleura is scarified. It has the lowest recurrence rate (<0.5%) and should be reserved for patients with high-risk occupations and those exposed to high variations of atmospheric pressure: deep sea divers, plane crew members, caisson workers and large truck drivers, where minimizing the risk of recurrence is paramount. Thoracotomy is associated with increased blood loss and post-operative pain, longer hospitalization and more frequent post-operative complications (12%).

### Surgical referral

Referral to a thoracic surgeon should be sought in the following circumstances:

- Second ipsilateral pneumothorax
- First contralateral pneumothorax
- Synchronous bilateral spontaneous pneumothorax
- Persistent air leak (despite 5–7 days of chest tube drainage) or failure of lung re-expansion

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• Spontaneous haemothorax
• Professions at risk (e.g. pilots and divers)
• Pregnancy

Patient choice will play a part in decision-making. Patients may elect to undergo surgical repair after their first pneumothorax, even in those without an increased risk in the event of a pneumothorax, weighing the benefits of a reduced risk against that of chronic pain, paraesthesia or the possibility of increased costs.

Chemical pleurodesis

In patients who are unable or unwilling to undergo VATS, intrapleural injection of a chemical irritant, most commonly a tetracycline derivative (doxycycline or minocycline), or talc is another alternative. Chemical pleurodesis with tetracycline derivatives decreases the recurrence rate of pneumothorax from 20% to 25%, when compared with aspiration and drainage without pleurodesis. However, chemical pleurodesis is associated with intense chest pain in some cases, chronic pain that may limit the use of this procedure. Controversy exists whether talc should be used as the sclerosant agent in young, otherwise healthy individuals, because of safety reasons such as acute respiratory distress syndrome (ARDS) and for fear of long-term complications. Doxycycline has comparable effectiveness to talc and has not been associated with ARDS.

Discharge and follow-up

All patients discharged should be given verbal and written advice to return to the emergency department immediately if they develop further breathlessness. It is recommended that all patients should be followed up to ensure resolution of the pneumothorax, to explain the risk of recurrence and the possible later need for surgical intervention and reinforce lifestyle advice on issues such as smoking and air travel. Patients should be advised to return to work and to resume normal physical activities once all symptoms have resolved, although it is reasonable to advise deferring sports that involve extreme exertion and physical contact. Patients should be made aware of the danger of air travel in the presence of a current closed pneumothorax and should be cautioned against commercial flights at high altitude until full resolution has been confirmed on CXR. Pregnancy is an issue to be discussed with younger female patients.

Recurrence prevention

The estimated recurrence rate after the first pneumothorax is 23–50% over a 1- to 5-year follow-up period, with the highest risk occurring in the first 30 days. Recurrence rates are higher in patients who have blebs or bullae on high-resolution CT scan. Patients with recurrent PSP should undergo an intervention to prevent future recurrences once the acute air leak has resolved, the lung has expanded and the pleural air has been removed.

Patients experiencing their first PSP should have a preventive intervention if they require VATS or tube thoracostomy as part of their initial management or have a vocation in which recurrence of the pneumothorax is dangerous to the patient or others (e.g. airplane pilot, para jumper or deep sea diver). Options for preventing recurrence include pleurodesis via VATS, chemical pleurodesis via tube thoracostomy or pigtail catheter at the time of aspiration and thoracotomy. The choice of procedure is best dictated by the expertise of the practitioner at each institution. Returning to high-risk activities requires careful fitness and risk assessment by approved medical examiners in the relevant fields of occupational medicine governed by regulatory bodies, for example, Civil Aviation Safety Authority and International Marine Contractors Association. Normal lung function and a normal high-resolution CT chest are required to exclude underlying lung pathology in either lung after preventive surgery.

The strong association between smoking and an initial PSP suggests that smoking cessation may help prevent recurrent pneumothoraces. In addition, smoking may predict a higher recurrence rate with talc pleurodesis.

Conclusion

All the current guidelines – ACCP, BTS and SEPAR – recommend treatment based on the severity of symptoms and the degree of the lung collapse, as determined by chest radiographs. The current international guidelines needed updating with well-conducted randomized evidence. Initial management is directed at removing air from the pleural space, with subsequent management directed at preventing recurrence. Observation with supplemental oxygen, aspiration of intrapleural air, tube thoracostomy and VATS chemical pleurodesis with talc to prevent recurrence are the mainstays of treatment. Thoracotomy is reserved for those who are unable or unwilling to undergo VATS, in situations where VATS has failed or in high-risk vocations.

Conflicts of interest

None declared.

References