

RECOMMENDATIONS OF THE SPANISH SOCIETY OF PULMONOLOGY AND THORACIC SURGERY (SEPAR)

Guidelines for the Diagnosis and Treatment of Spontaneous Pneumothorax

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This is the fourth update of the guidelines for the diagnosis and treatment of pneumothorax published by the Spanish Society of Pulmonology and Thoracic Surgery (SEPAR).

Spontaneous pneumothorax, or the presence of air in the pleural space not caused by injury or medical intervention, is a significant clinical problem. We propose a method for classifying cases into 3 categories: partial, complete, and complete with total lung collapse. This classification, together with a clinical assessment, would provide sufficient information to enable physicians to decide on an approach to treatment.

This update introduces simple aspiration in an outpatient setting as a treatment option that has yielded results comparable to conventional drainage in the management of uncomplicated primary spontaneous pneumothorax; this technique is not, as yet, widely used in Spain.

For the definitive treatment of primary spontaneous pneumothorax, the technique most often used by thoracic surgeons is video-assisted thoracoscopic bullectomy and pleural abrasion.

Hospitalization and conventional tube drainage is recommended for the treatment of secondary spontaneous pneumothorax.

This update also has a new section on catamenial pneumothorax, a condition that is probably underdiagnosed.

The definitive treatment for a recurring or persistent air leak is usually surgery or the application of talc through the drainage tube when surgery is contraindicated.

Our aim in proposing treatment algorithms for the management of pneumothorax in these guidelines was to provide a useful tool for clinicians involved in the diagnosis and treatment of this disease.

Key words: Spontaneous pneumothorax. Bullae. simple aspiration. Chest drainage. Video-assisted thoracoscopy. Treatment.

Normativa sobre el diagnóstico y tratamiento del neumotórax espontáneo

Se presenta la cuarta puesta al día de la "Normativa sobre diagnóstico y tratamiento del neumotórax", de la Sociedad Española de Neumología y Cirugía Torácica (SEPAR).

La presencia de aire dentro de la cavidad pleural de causa no traumática o iatrógena es un problema clínico relevante. Se propone un método de cuantificación del neumotórax mediante su clasificación en parcial, completo o total, que junto a la valoración clínica parece suficiente para adoptar las diversas actitudes terapéuticas.

En la presente actualización se incorpora la aspiración simple ambulatoria, como método equiparable en resultados al drenaje convencional, para el tratamiento del neumotórax espontáneo primario no complicado, cuyo uso no está todavía muy extendido en España.

Para el tratamiento definitivo del neumotórax espontáneo primario, la cirugía videotoracoscópica con bullectomía y abrasión pleural es la técnica más ampliamente utilizada por la mayoría de cirujanos torácicos.

En el tratamiento del neumotórax espontáneo secundario se recomienda el ingreso y la colocación de drenaje torácico convencional.

Se ha introducido también una referencia al neumotórax catamenial, probablemente infradiagnosticado.

En caso de recidiva o fuga aérea persistente, el tratamiento definitivo suele ser el quirúrgico o el uso de talco a través del drenaje en caso de contraindicación.

Los algoritmos de estrategia terapéutica aquí propuestos pretenden convertirse en una herramienta de trabajo útil para todos los implicados en el diagnóstico y tratamiento de esta enfermedad.

Palabras clave: Neumotórax espontáneo. Bullas. Aspiración simple. Drenaje torácico. Videotoracoscopia. Tratamiento.

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Manuscript received October 2, 2007. Accepted for publication January 30, 2008.

Introduction

With this fourth update of the recommendations for the diagnosis and treatment of pneumothorax published by the Spanish Society of Pulmonology and Thoracic Surgery (SEPAR) we have tried to fulfill the 2 primary objectives

of such guidelines: *a*) to help the pulmonologist, thoracic surgeon, or other professional interested in the subject to take the most appropriate action to resolve a specific clinical problem; and *b*) to define SEPAR's official position on this significant clinical problem.

The focus of these guidelines is the treatment of spontaneous pneumothorax, defined as the presence of air in the pleural space not caused by injury or medical intervention. In our opinion, the distinction between primary and secondary spontaneous pneumothorax remains useful (despite the obvious presence in the former of pathological abnormalities) because this internationally accepted terminology precisely indicates and defines the respective characteristics of these 2 clinical entities.

In the SEPAR guidelines published 5 years ago, chest drainage methods based on small-bore catheters and one-way valves were introduced as therapeutic options for short stay patients and even outpatients. This update simplifies the criteria for classifying and quantifying pneumothoraces on radiographs, and introduces simple aspiration—a procedure as yet not widely used in Spain—as a simple, safe, and effective technique that yields results comparable to those of conventional chest drainage in the management of uncomplicated first primary pneumothoraces. There is still some debate concerning the definitive treatment of primary pneumothorax between the interventional pulmonologists, who propose conventional simple thoracoscopy with insufflation of talc, and the chest surgeons, who favor resection of bullae and mechanical pleural abrasion or partial pleurectomy using video-assisted thoroscopic surgery (VATS).

This update also includes a new section on catamenial pneumothorax, a condition that is probably underdiagnosed and perhaps for this reason is often treated with poor results.

We believe that these guidelines should become a useful working tool in hospital emergency departments, and will be of interest to emergency physicians, internists, general and thoracic surgeons, and pulmonologists, as well as any other professionals involved in the diagnosis and treatment of this prevalent disease. Although pneumothorax rarely endangers the patient's life except in the case of tension pneumothorax or when the condition is caused by a severe underlying lung disease, we believe that interdisciplinary cooperation and the use of the treatment algorithms proposed in these guidelines will improve the care of these patients.

Pathophysiologic and Etiologic Characteristics

Concept

We define pneumothorax as the presence of air in the pleural space leading to loss of the negative subatmospheric intrapleural pressure and partial or total lung collapse.

Classification

Pneumothoraces can be classified by etiology as either spontaneous or acquired (iatrogenic or traumatic). There are 3 types of spontaneous pneumothorax: *a*) primary,

when there is no apparent lung disease; *b*) secondary, when there is an underlying pleural or pulmonary disease; and *c*) catamenial, when the condition is related to the menstrual cycle.

Epidemiology

Spontaneous pneumothorax remains a significant health problem, and the incidence varies considerably.¹⁻⁴ Gupta et al¹ estimated an overall incidence for this disease in England of 16.8 per 100 000 population per year (24 for men and 9.8 for women), and an incidence for patients requiring hospitalization of 11.1 per 100 000 per year (16.6 for men and 5.8 for women). Melton et al³ found the age adjusted incidences of primary and secondary pneumothorax in the United States of America to be 4.2 and 3.8 cases per 100 000, respectively. The incidence of primary spontaneous pneumothorax was 7.4 per 100 000 in men and 1.2 per 100 000 in women (a male/female ratio of 6.2:1), and that of secondary spontaneous pneumothorax was 6.3 per 100 000 in men and 2 per 100 000 in women (a male/female ratio of 3.2:1). Bense et al⁴ found an incidence of primary pneumothorax in Sweden of 18 to 28 per 100 000 in men and 1.2 to 6 in women. It is important to bear in mind that the incidence of this entity is not fully known since the condition can be asymptomatic, but the prevalence has been shown to be increasing.¹ It is important to note, with respect to the age of onset, that the peak incidence of primary pneumothorax is among young people, whereas the incidence of secondary pneumothorax is highest in the population over 55 years of age.^{1,3}

Recurrence rates reported in the literature vary widely depending on the treatment applied and the follow-up period used in the study.⁵ Although it is impossible to predict the general course of spontaneous pneumothorax, studies have shown that the recurrence rate of primary pneumothorax is approximately 30% (range, 16%-52%) with follow-up in some studies for as long as 10 years⁵; recurrence rates of secondary pneumothorax range from 40% to 56%.^{6,7} In most cases, recurrence occurred during the 6 months following the first episode.⁸ Sadikot et al,⁹ who followed 153 patients with primary pneumothorax for 54 months, observed a recurrence rate of 39% during the first year and reported that pneumothorax was contralateral in 15% of the patients studied. When a second episode of pneumothorax is not treated appropriately, the possibility of a third increases considerably. Voge and Anthracile¹⁰ reported a 28% rate of recurrence following a first episode. Of these cases, 23% presented a third episode, and 14% of this 23% had a fourth episode. The overall rate of recurrence in that study was 35%.

Among the risk factors for recurrence of spontaneous pneumothorax, Lippert et al⁶ mentioned pulmonary fibrosis on chest radiographs, age over 60 years, and an increased height-to-weight ratio (evidence level 2-). According to the British Thoracic Society (BTS), smoking, height in male patients, and age are the main risk factors for recurrence in primary pneumothorax, while in secondary pneumothorax the factors are age, pulmonary fibrosis, and

emphysema.² It is important to emphasize that the BTS pleural disease group found a relationship between recurrence of pneumothorax and continued smoking (evidence level 3).²

Recent mortality rates in the United Kingdom were 0.62 per million per year for women and 1.26 per million per year for men.¹ Among patients aged over 55, the same authors reported mortality of 1.8% and 3.3% among hospitalized men and women, respectively (evidence level 3).¹ With respect to the relationship between mortality and type of pneumothorax, primary spontaneous pneumothorax is rarely fatal,¹ but death is more common in secondary pneumothorax because of the underlying lung disease and the patients' impaired respiratory reserve.¹¹ In patients with chronic obstructive pulmonary disease (COPD), mortality ranges between 1% and 7%. Videm et al⁷ found that mortality in patients with COPD was 4 times higher with each successive episode (evidence level 2+).

Etiology and Pathophysiology

Primary spontaneous pneumothorax is unrelated to a specific precipitating event and occurs in a person who in theory has no underlying lung disease. It is associated with smoking and an asthenic or leptosomic physical constitution or morphologic biotype.¹² Smoking is an important risk factor associated with respiratory bronchiolitis, which has been detected in 88% of smokers with primary pneumothorax.¹³ The risk among men who smoke more than 20 cigarettes per day is 12.3%, a figure that contrasts with 0.1% in nonsmokers and correlates directly and exponentially with tobacco consumption (evidence level 2++).⁴ It should be noted that their conclusions apply equally to women. Bense et al⁴ observed that the relative risk of pneumothorax among smokers as compared to nonsmokers was 22 times higher for men and 9 times higher for women. The surprising finding is that, in spite of this clear relationship between spontaneous pneumothorax and smoking, 80% to 86% of patients continue to smoke after a first episode.¹⁴ Patterns of family inheritance, although uncommon, may also influence risk. Several studies have analyzed the familial incidence of spontaneous pneumothorax and of primary pneumothorax in particular. Contrary to popular belief, physical activity does not appear to influence the onset of primary pneumothorax, and there is no clear evidence of any relationship with changes in weather or atmospheric pressure (evidence level 2+).¹⁵ Primary pneumothorax has, however, occasionally been linked to specific sporting and work activities (evidence level 3).¹⁶

Secondary spontaneous pneumothorax can be caused by many different diseases, including infectious processes (*Pneumocystis jiroveci*, *Mycobacterium tuberculosis*, and necrotizing pneumonia), interstitial lung disease, connective tissue disease, Langerhans cell histiocytosis, lymphangioleiomyomatosis, cystic fibrosis, and COPD. Cystic fibrosis and COPD are the most commonly reported associated conditions.¹¹

While the pathophysiologic mechanisms of spontaneous pneumothorax are still poorly understood,^{17,18}

it is thought that primary pneumothorax is caused by the formation and subsequent rupture of subpleural bullae.⁵ The hypothesis has been advanced that structural changes in the lung parenchyma (generally called emphysema-like changes) give rise to a higher pressure gradient from the apex to the base of the pleural space, which in turn increases intraalveolar pressure and results in considerable distension of the apical subpleural alveoli. This leads to the formation and subsequent rupture of cysts, a process associated with the degradation of elastic fibers in the lung caused—particularly in smokers—by mediators released by neutrophils and macrophages. This degradation causes an imbalance between proteases and antiproteases and oxidants and antioxidants, resulting in irreversible damage to the lung parenchyma, emphysema, and the formation of pulmonary bullae. After a bulla forms, inflammation of the small airways leads to an increase in alveolar pressure resulting in rupture followed by leakage of air through the lung interstitium into the hilum.¹¹ In one study, emphysema-like changes were found on computed tomography (CT) scans in 81% of nonsmoking non- α_1 -antitrypsin deficient men with a previous episode of primary pneumothorax and in only 20% of the controls with no history of pneumothorax.¹⁹ These changes were located in the apices of the upper pulmonary lobes and presented as localized areas of low attenuation measuring at least 3 mm in diameter, sometimes delimited by thin walls. Despite the evidence of emphysema-like changes found in patients with primary spontaneous pneumothorax, some authors maintain that these changes are not the sole cause of this disease. It is also possible that an association of factors may trigger the pneumothorax and give rise to a persistent air leak. These may include distal airway inflammation due to a hereditary predisposition, anatomic abnormalities, or smoking (which plays an important role) as well as the diffuse bilateral subpleural emphysema-like changes and the porosity of the visceral pleura observed in these patients.^{18,20}

Jordan et al²¹ observed bullae in the CT scans of 46% to 52% of the patients they studied, and Lesur et al²² in 80%. Using thoracoscopy, Janssen et al²³ found emphysema-like changes in 69% to 77% of cases, and Donahue et al²⁴ found them in 90%. Sihoe et al²⁵ detected bilateral bullous lesions on CT scans in 54% of cases, and Ikeda et al²⁶ in 93% of patients who underwent bilateral surgery. However, in a study comparing patients with first-time primary pneumothorax and patients with recurrent primary pneumothorax, the number of bullae detected in the patients with recurrent disease was not higher, an observation that led the authors to suggest that such lesions do not constitute the sole risk factor.²³ Horio et al²⁷ reported that bullectomy alone without chemical pleurodesis or pleurectomy does not prevent recurrence.

The pathophysiology of secondary spontaneous pneumothorax is multifactorial and remains poorly understood. Air enters the pleural space after alveolar rupture caused by peripheral lung necrosis or some other mechanism related to the underlying lung disease responsible in each case.^{2,20}

Diagnostic Assessment

It is important to ascertain whether the pneumothorax is primary or secondary, whether it is a first episode or a recurrence, and whether or not it is accompanied by pleural effusion. The overall care plan for this disease should, therefore, include a complete and accurate medical history and a physical examination.

Evaluation and quantification of the size and volume of the pneumothorax by chest radiography should be complemented by assessment of the patient's clinical status, determination of the type of spontaneous pneumothorax, and identification of possible active air leaks. The findings will provide a basis for choosing the most appropriate therapeutic approach.^{2,28,29}

Clinical Presentation and Physical Examination

The extent to which the lung has collapsed and the reduction in ventilatory capacity caused by such collapse correlates with the volume of air in the pleural space. However, the symptoms reported by patients with secondary spontaneous pneumothoraces are more closely related to their pulmonary respiratory reserve than to the degree of collapse.

The typical profile of the patient with primary spontaneous pneumothorax is a tall, thin or asthenic, and usually male young person aged between 20 and 30 years.¹¹ There is no relationship between clinical presentation and the size of the pneumothorax (evidence level 2+).² The condition is asymptomatic in 10% of patients and in such cases it may be detected during a routine medical examination or evaluation for another disease. It has even been shown that 46% of patients with primary pneumothorax wait more than 2 days after onset of symptoms before consulting a physician (evidence level 3).² In 80% to 90% of cases, pneumothorax occurs when the patient is at rest or their level of physical activity is very low. The condition tends to affect individuals with specific physical characteristics (a leptosomic morphological constitutional type) and psychological makeup (type A behavior pattern characterized by an emphasis on competition, impatience, distrust, hostility, and dedication to work) (evidence level 3). The patient presents with acute or subacute one-sided pleuritic chest pain that is exacerbated by deep respiratory movements and coughing fits and alleviated by shallow breathing and immobilization. They report a certain degree of sudden onset dyspnea, sometimes accompanied by irritative cough.²⁰ All of these symptoms are exacerbated by respiratory movements. If the pneumothorax is small, pain may be the only symptom and even this may disappear within hours of onset, usually within 24 hours, even when the condition remains untreated and does not resolve.¹¹ The results of physical examination in patients with primary pneumothorax vary. They may be almost normal (although tachycardia is a common finding in patients with small pneumothoraces) or they may include reduced movement of the chest wall on the affected side, decreased or absent breath sounds, increased resonance to percussion, and a reduction in voice projection.¹¹ Spontaneous pneumothorax

usually affects the right lung, and is bilateral in fewer than 10% to 15% of patients.^{9,11}

The clinical signs and symptoms of secondary pneumothorax are more intense and severe. Intense dyspnea, respiratory failure, and scant respiratory reserve are all symptoms that may be life-threatening and require immediate treatment. Some of the most clinically significant symptoms that may develop, depending on the underlying disease, include ipsilateral chest pain, varying degrees of hypoxemia with or without hypercapnia, and systemic hypotension. Due to the presence of underlying disease, physical examination does not reveal very much in these patients; the absence of breath sounds, a hyperresonant percussion note, and diminished vocal vibrations are the main findings.⁴

Both primary and secondary spontaneous pneumothorax are occasionally accompanied by pleural effusion (10%-20%) of greater or lesser volume. This effusion, which is usually eosinophilic, is caused by irritation of the pleura by the entrance of air, and is, on rare occasions, accompanied by hemothorax.

In short, clinical parameters are not reliable indicators of the size of the pneumothorax (evidence level 2+).² With respect to clinical status, according to the criteria published by the American College of Chest Physicians²⁸ a pneumothorax is clinically stable when the patient's respiratory rate is under 24 breaths/min, heart rate is between 60 and 120 beats/min, systemic blood pressure is normal, room air oxygen saturation is above 90%, and the patient can speak in whole sentences between breaths. The BTS guidelines add absence of dyspnea as another marker of clinical stability.²

Additional Testing

Certain additional tests are important in the diagnosis of pneumothorax.

Imaging studies before treatment. Diagnosis is confirmed by imaging studies, primarily a plain chest film using a standard projection (posteroanterior radiograph obtained during forced inspiration with the patient in a standing position). The image will show the visceral pleural line clearly defined by increased opacity and the absence of distal lung markings. International guidelines do not recommend routine use of the forced expiratory maneuver for diagnosis (grade B recommendation).^{2,28,29,30}

Although CT scans can reveal the underlying pathophysiologic lesions that cause spontaneous pneumothorax, the ACCP does not recommend the routine use of this imaging technique for patients with first-time primary or secondary pneumothoraces.²⁸ CT may, however, be useful for evaluating patients with recurrent secondary pneumothorax, to determine the best treatment for persistent air leaks or to plan a surgical intervention. The BTS recommends using CT when required to differentiate between pneumothorax and bullous lung disease, when aberrant tube placement is suspected and when the plain chest radiograph is difficult to read owing to the presence of subcutaneous emphysema (grade C recommendation).²

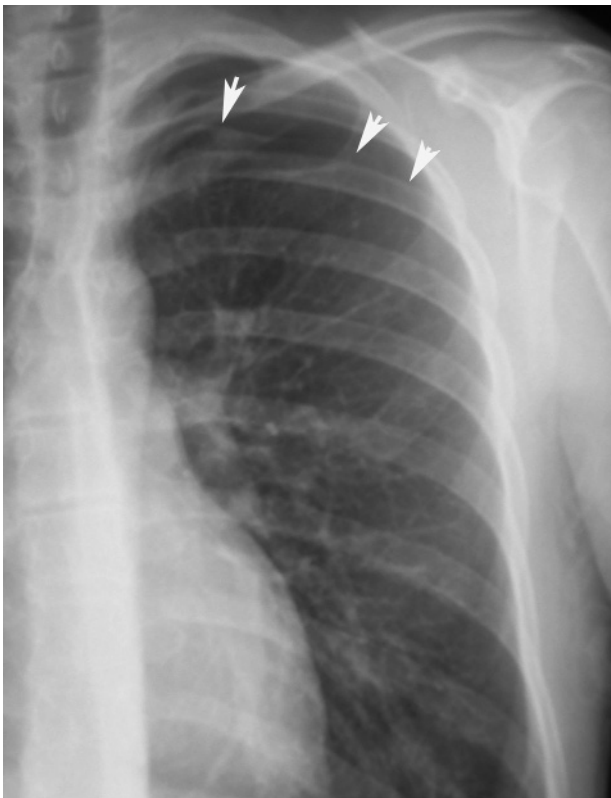


Figure 1. Partial pneumothorax in the left lung.

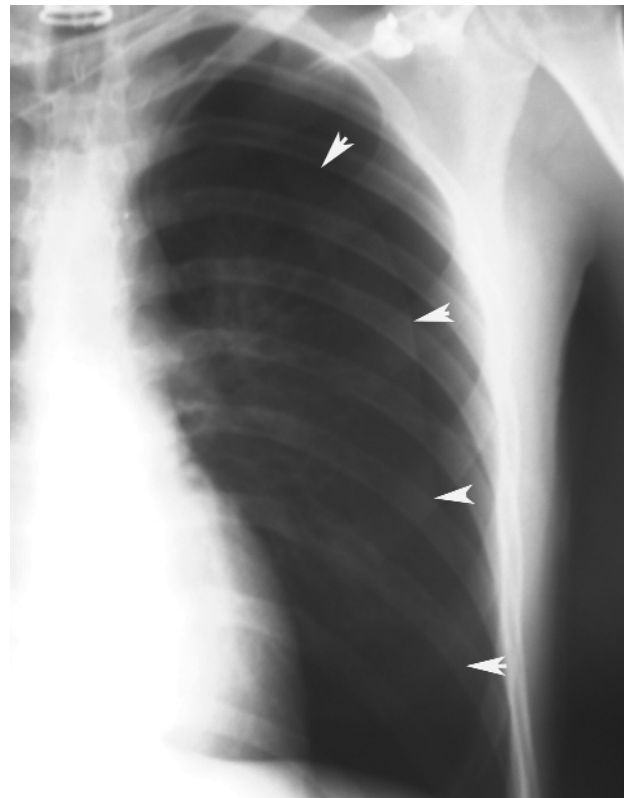


Figure 2. Complete pneumothorax in the left lung.

Resting arterial blood gas analysis-pulse oximetry. In primary spontaneous pneumothorax, the results of arterial blood gas analysis may reveal an increase in the alveolar-arterial oxygen gradient and acute respiratory alkalosis.¹¹ When a pneumothorax occupies some 25% or more of the hemithorax, the abnormal ventilation-perfusion ratio in the affected area may cause hypoxemia and increase the alveolar-arterial oxygen gradient. Resting arterial blood gas analysis is not routinely recommended in patients with primary pneumothorax.

Lung function assessment. Lung function testing is not recommended as a method for evaluating the size or presence of pneumothoraces, the functional status of patients with ongoing pneumothorax, or as part of their pretreatment assessment.

Electrocardiogram. Electrocardiographic abnormalities are rare in patients with pneumothorax and this procedure is not routinely recommended in patients with primary spontaneous pneumothorax.

Quantification of Spontaneous Pneumothoraces on Chest Radiographs

As there is no agreement about the best way to determine the size of a pneumothorax, the degree of lung collapse, or the volume of accumulated air, many different methods

and classifications have been used. These include the Light index¹² based on the ratio of the cubed diameters of the lung and hemithorax ($\% \text{ pneumothorax} = 100 [1 - \text{lung diameter}^3 / \text{hemithorax diameter}^3]$), and the method proposed by Rhea et al³¹ based on the calculation of the average interpleural distance and its subsequent adjustment using a nomogram. However, nearly all of these methods underestimate the volume of pneumothoraces and neither of the 2 most recent international guidelines use the percentage of collapsed lung to guide treatment.^{2,28} The ACCP consensus statement defines a pneumothorax as “small” when the distance between the apex of the lung and the thoracic cupola is less than 3 cm and “large” when this distance exceeds 3 cm.²⁸ The BTS guidelines, on the other hand, consider a pneumothorax to be “small” when the separation between the lung margin and the chest wall is less than 2 cm and “large” when this distance is greater than 2 cm.² In a recent editorial, Henry³² discussed the possibility of a practical method for quantifying the size of pneumothoraces. He proposed that a pneumothorax be defined as large when there is complete dehiscence between the visceral pleura and the chest wall, and partial when the dehiscence is incomplete.

Indeed, it is difficult to assess the size of a pneumothorax either in centimeters, given the growing use of digital imagery, or in percentages since the lung does not deflate in a constant and uniform manner. We therefore propose classifying pneumothoraces using simple anatomical and

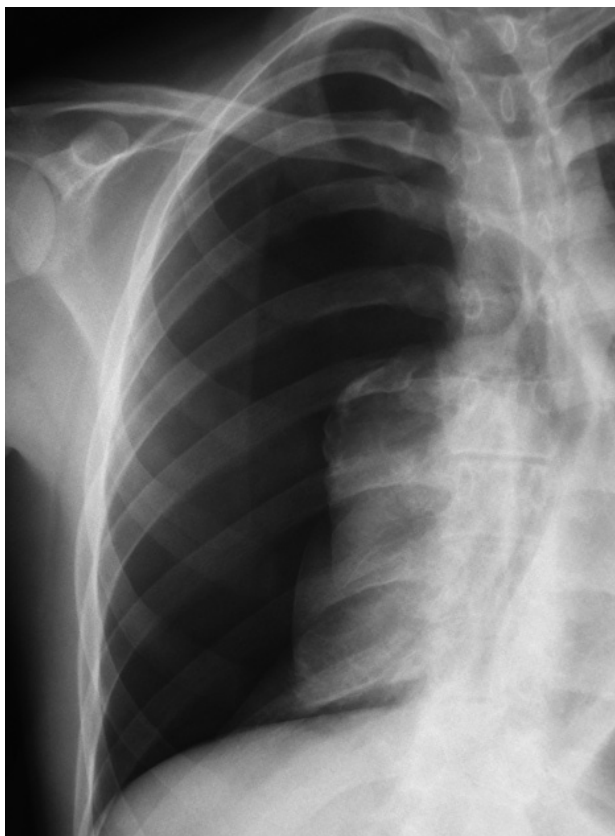


Figure 3. Complete pneumothorax with total lung collapse in the right lung.

morphological criteria, as follows: *a*) partial, if the visceral pleura is separated from the chest wall along only part of the pleural space, generally apical (Figure 1); *b*) complete, when the visceral and parietal pleuras are completely separated from the base to the apex of the pleural space without total lung collapse (Figure 2); and *c*) complete with total lung collapse, when there is such collapse and also uniform formation of a stump (Figure 3). This method for quantifying the size of pneumothoraces on plain radiographs is simple, easy, and quick, and together with the clinical assessment of the patient's condition provides enough information to support a decision about the optimum treatment strategy.

General Approach to Management

The aims in the management of pneumothorax are to reexpand the lung, minimize morbidity, prevent recurrence, and provide appropriate treatment for symptoms. In patients with secondary pneumothorax, the underlying disease is also treated. While the solution lies in the appropriate use of existing treatments, this does not mean that research into new approaches should be abandoned.

General Measures

One of the principal therapeutic measures in these patients is adequate analgesia for relief of pain caused by the pneumothorax itself and the treatment applied.

There is no evidence that confining the patient to bed favors lung reexpansion or air reabsorption, and complete bed rest can lead to complications in patients with spontaneous pneumothorax. When bed rest is indicated, routine prophylaxis for thromboembolism is important in all patients with risk factors. Physical or pharmaceutical preventive measures should be used, such as administration of low-molecular-weight heparin in appropriate doses (grade B recommendation).³³

Spontaneous reabsorption of pneumothoraces occurs at rates from 1.25% to 1.8% (50-75 mL) of the total volume of air in the pleural space per day and can be accelerated by as much as a factor of 4 by the administration of supplemental oxygen. Hospitalized patients waiting for assessment and patients admitted for a simple conservative treatment should receive supplemental oxygen, which reduces the partial pressure of nitrogen in the pleural capillaries and increases the rate of reabsorption of air from the pleural space (grade B recommendation).² The BTS recommends administration of supplemental high flow oxygen (for example, 10 L/min), while emphasizing that care should be taken in patients with COPD because of the risk of hypercapnia (grade B recommendation).²

Tobacco cessation should be one of the priorities of patients with spontaneous pneumothorax as it is an important preventive and therapeutic measure with respect to both the onset and recurrence of this condition as well as lung function impairment in general.

Recommendations for Specific Situations

Due to the increase in air travel and likewise in high risk sports and professions, the SEPAR guideline on respiratory disease and air travel, defines pneumothorax as a contraindication for air travel and stipulates that patients should not fly until complete reexpansion of the lung has been achieved.³⁴ Patients with pneumothorax should not, therefore, be permitted to fly until 72 hours after a pleural drainage tube is removed and then only if a chest radiograph obtained 48 hours after removal of the tube has confirmed that the problem has resolved (grade C recommendation).³⁵ The Aerospace Medical Association states that, optionally, some airlines can accept the transport of passengers with chest tubes.³⁶ In such cases, since it is difficult to guarantee continuous aspiration during the flight, a Heimlich valve assembly should be installed. In exceptional cases, evacuation of a pneumothorax may be necessary during a flight, and this procedure must be performed by trained personnel.

According to the BTS guidelines, commercial airlines advise individuals to avoid air travel for 6 weeks after an episode of primary spontaneous pneumothorax and stress that patients should not fly until resolution has been confirmed.² Although there is no evidence that recurrence is caused by flying, the consequences of a pneumothorax occurring during a flight could be serious because of the lack of medical care. Restrictions on flying may be more justified in patients for whom pneumothorax is associated with higher risk, such as smokers and patients with underlying lung disease (secondary spontaneous pneumothorax). In patients with secondary pneumothorax who have not been

treated surgically, air travel should be avoided for 1 year after an episode (grade C recommendation).²

Patients with a history of pneumothorax who have not been treated surgically should also be advised against practicing high risk sports, such as diving (grade C recommendation).² The BTS guidelines on the respiratory aspects of fitness for diving recommend that such patients should avoid diving until they have undergone bilateral surgical thoracotomy with apical pleurectomy or pleural abrasion.³⁷ VATS is not included in this criteria because the rate of recurrence reported—between 5% and 10%—in some case series is unacceptable for this high-risk activity. Therefore, bilateral thoracotomy with apical pleurectomy (associated with a 0.5% rate of recurrence³⁸) or bilateral VATS with apical pleurectomy (with a 0% recurrence rate³⁹) should be the first-line interventions in these patients.

Follow-up and Monitoring

No follow-up is required when spontaneous pneumothorax has fully resolved. However, patients with untreated spontaneous pneumothorax and outpatients with chest tubes should be followed by the appropriate specialist physicians depending on their availability at different levels of the health care system.

Treatment of Primary Spontaneous Pneumothorax

Primary pneumothorax affects individuals without apparent lung disease and should not, therefore, be considered a serious disease. In most cases, the condition resolves with observation or minimally invasive procedures and its impact on the activities of daily living of these patients is slight.

The chief objective of the management of primary pneumothorax is elimination of the intrapleural air, either by observation in the case of partial pneumothorax or by evacuation using any of several different methods when the pneumothorax is complete and/or there is total lung collapse. The secondary aim is to prevent recurrence when there is a high probability of recurrence or when recurrence could potentially be serious. We will now analyze the therapeutic approach step by step on the basis of a treatment algorithm (Figure 4).

Notes on the Treatment Algorithm for Primary Spontaneous Pneumothorax.

1. Observation should be the first-line treatment for patients who have a partial pneumothorax and no dyspnea (grade B recommendation).⁴⁰ Persistent dyspnea in a patient with a partial pneumothorax should raise the suspicion of an underlying disease (and therefore secondary spontaneous pneumothorax) or an increase in the size of the pneumothorax. Some authors recommend observation in the emergency department for 3 to 6 hours, after which the patient can be discharged if a repeat plain chest radiograph confirms that the pneumothorax is stable (grade D recommendation).² These patients do not require hospitalization, unless they live at a great distance from

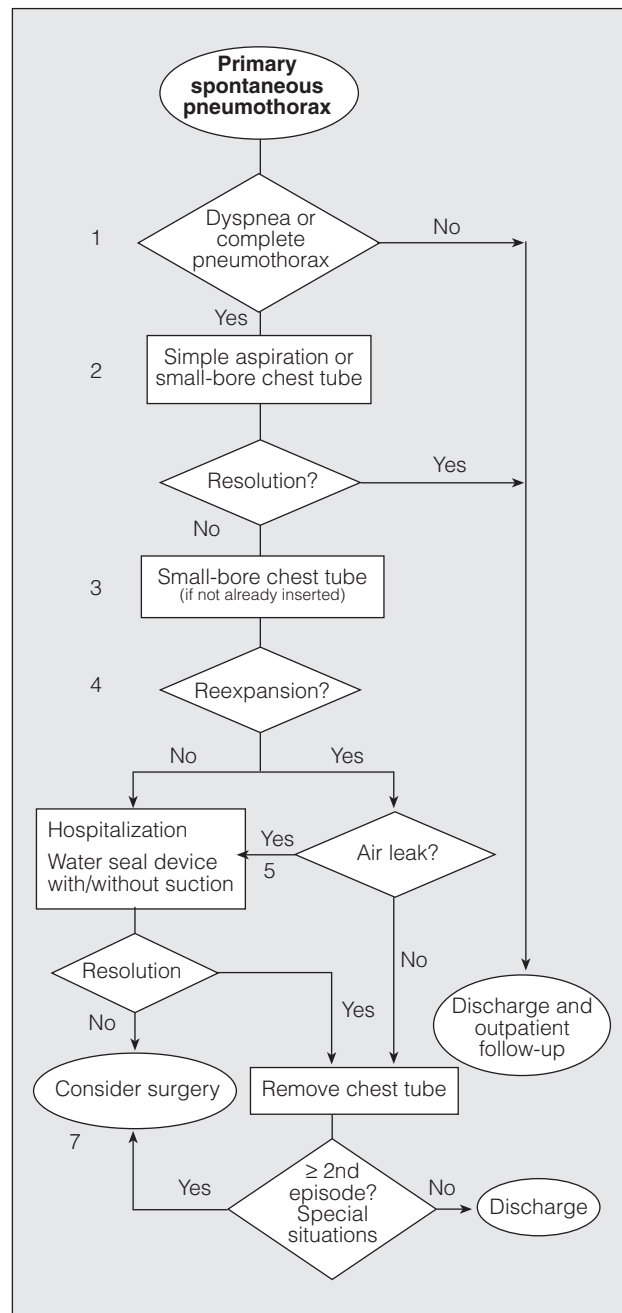


Figure 4. Treatment algorithm for primary spontaneous pneumothorax.

the hospital or would have serious difficulties in reaching an emergency department.²⁸ They should always receive clear instructions to return to the emergency department if they should have difficulty breathing.

Although, at a reabsorption rate of 1.25% to 1.8% per day, a 25% pneumothorax should take 20 days to resolve,⁴¹ there is insufficient evidence to support a specific schedule for reviewing the patient’s condition. The ACCP expert panel recommends a follow-up visit between 2 and 14 days,²⁸ and it seems reasonable to us that at least 1 chest

radiograph should be obtained to confirm resolution (grade D recommendation).

2. Although simple aspiration is not a widely used procedure in Spain—only 1 article has been published on this topic in Spanish in the last 10 years⁴²—several randomized clinical trials have shown this procedure to be as effective in both the short and long term as chest tube drainage (grade A recommendation).⁴³⁻⁴⁵ Moreover, it is associated with less pain and patients do not have to be admitted to hospital. The rate of immediate resolution ranges between 50% and 88%, results similar to those achieved with chest tube drainage. Using simple aspiration, primary spontaneous pneumothorax can be managed in a large number of patients without hospitalization.

In primary pneumothorax, the BTS guidelines recommend repeating simple aspiration when the first attempt is unsuccessful, especially when only a small volume of air was removed in the first aspiration (grade B recommendation).²

Several methods are used to perform simple aspiration, ranging from venous catheters to small-bore chest tubes that can be removed once reexpansion of the lung is confirmed. If the patient is discharged from the emergency department with an indwelling small-bore catheter, the procedure is considered to be chest drainage.³⁹ In other words, the difference between simple aspiration and small-bore tube drainage is the time the tube remains in place. Both methods are equally valid in the management of complete primary spontaneous pneumothorax, particularly when small-bore tube drainage is used as an outpatient treatment.⁸ Before a patient is discharged, a chest radiograph should always be obtained to confirm reexpansion of the lung.

3. Most studies have found no differences in the success rate obtained between small-bore catheter drainage ("14F) and intercostal tube drainage ("16F). The first-line treatment is, therefore, chest drainage with small-bore catheters for both in-hospital and outpatient management of this condition (grade B recommendation).⁴⁶⁻⁴⁷ Moreover, small-bore catheters are easier to insert and cause less discomfort.

4. The catheter can be withdrawn and the patient discharged once the air leak has resolved and lung reexpansion has been confirmed radiographically.

5. When an air leak persists after complete reexpansion of the lung, the patient should be connected to a one-way valve or underwater seal system. This strategy usually achieves resolution of the pneumothorax within an average of 48 hours (grade B recommendation).⁴¹ Valve systems make possible outpatient treatment in patients able to cooperate who live near the hospital.

6. There is no evidence that the early use of suction with pleural drainage speeds up the resolution of pneumothoraces.⁴⁸ Conclusions based on clinical trials of the application of suction indicate that it provide no benefit after pulmonary lobectomy and in most cases delayed resolution of the air leak (grade A recommendation).⁴⁹ The use of suction is often decided on an empirical basis when there is a persistent air leak or the lung fails to reexpand, but there is no consensus on how much pressure should be used. The risk of pulmonary edema caused by reexpansion is anecdotal and should not delay the use of

suction when this strategy is considered necessary.⁸ When used, suction should always be carried out in a setting where there is appropriate supervision and monitoring.²

Another aspect as yet little studied is the influence of physiotherapy on the resolution of pneumothorax. It would appear reasonable to hypothesize that, in the absence of a significant air leak, respiratory physiotherapy should favor resolution of the condition and the elimination of air from the pleural space. There is no pathophysiologic basis for the popular belief and conventional medical practice of ensuring bed rest in patients with spontaneous pneumothorax, including those with chest drainage.^{2,28}

A chest radiograph should always be obtained to confirm reexpansion of the lung. There is no evidence supporting the need to clamp the drainage tube or wait for a specific interval before removing the tube after reexpansion of the lung has been confirmed (grade B recommendation).⁴⁸ Noppen⁵⁰ recommends waiting 12 hours after the last evidence of an air leak (grade D recommendation), although this recommendation is of little use since the interval that elapses before air leaks are assessed in such patients is rarely less than 24 hours.

In various countries, including Spain, many physicians prefer to clamp the drainage tube for a short period before removal (47% in the ACCP expert panel²⁸). The objective is to detect small or intermittent leaks and to render the installation of a new drain unnecessary if such leaks are found. Since clamping a chest drain is a potentially dangerous procedure, it should only be carried out under the supervision of trained medical personnel who can unclamp it if necessary. A drainage tube should never be clamped for more than 6-12 hours (grade D recommendation).²⁸

7. In most patients with primary pneumothorax and an air leak, the condition resolves in under 2 weeks.⁵¹ However, it is common practice to refer the patient to a thoracic surgeon or to contact a thoracic surgery department to arrange definitive surgical treatment because of the zero mortality rate and low morbidity associated with such surgery. There is, however, no scientific evidence to support this practice and, consequently, it may be just as appropriate to proceed in any other way based on organizational criteria.

The following are indications for surgical treatment of primary spontaneous pneumothorax taking into account the risk of recurrence or serious complications: a second ipsilateral pneumothorax, first contralateral or simultaneous bilateral pneumothorax, first episode of tension pneumothorax, significant spontaneous hemothorax, and a high risk profession or activity (pilots, divers, parachutists, etc).²

Definitive Treatment of Primary Spontaneous Pneumothorax

None of the methods used to cure pneumothorax or prevent its recurrence have to date been shown to be clearly superior to any other. When recommending one procedure over another, however, we should evaluate the risk-benefit ratio for each. Chemical pleurodesis with sclerosing agents is less effective than surgical procedures and is not recommended for primary spontaneous pneumothorax.⁴⁶

Thoracotomy with total pleurectomy is the method associated with the lowest rate of recurrence, but it would be a very aggressive intervention for the treatment of primary pneumothorax (grade D recommendation).²⁸ VATS provides similar results to thoracotomy with respect to prevention, and is associated with fewer complications and a better cosmetic result (grade C recommendation).⁵² Bullectomy with pleural abrasion is the technique most often used by most thoracic surgeons (grade D recommendation).²⁸

Intrapleural instillation of talc via a chest drainage tube or thoracoscopy has been used successfully,²⁰ but experimental studies and some experts have raised reasonable doubts about the safety of this procedure.^{53,54} For this reason, talc pleurodesis is not recommended in young patients, who represent the majority of primary pneumothorax patients.

Treatment of Secondary Spontaneous Pneumothorax

As mentioned, patients with secondary spontaneous pneumothorax have an underlying pleural or pulmonary disease (most often COPD, with or without bullae), and the treatment of pneumothoraces in these patients is conditioned by their underlying disease.

As with primary spontaneous pneumothorax, observation remains the first-line treatment in patients who have partial pneumothorax and no dyspnea, although this clinical picture generally occurs in patients with moderate-to-severe COPD and hospitalization is usually necessary in such cases. Patients with secondary spontaneous pneumothorax more often require chest drainage, and additional treatment to induce pleurodesis should be considered because, in addition to the underlying disease, the pneumothorax itself is a predictive factor for mortality in patients with COPD.⁷ The estimated recurrence of secondary spontaneous pneumothorax ranges from 40% to 56%.⁶

A more aggressive approach is generally considered more appropriate in secondary than in primary pneumothorax, but no randomized trials have been undertaken and no consensus exists concerning the specific treatment that should be used in such cases. The ACCP consensus statement recommends that all patients who present with a first episode of secondary spontaneous pneumothorax should be treated with chest drainage followed by pleurodesis to prevent recurrence, although 19% of these experts recommended not performing pleurodesis until the second episode of pneumothorax.²⁸ Both the BTS and the Belgian Society of Pneumology recommend simple aspiration as an initial treatment in cases of partial pneumothorax in patients with mild lung disease, although they recognize that most of these patients will require chest drainage.^{2, 29} The BTS recommends removal of the chest drain once the lung has reexpanded and all air leaks have resolved and reserves pleurodesis for patients with a persistent air leak or recurrent pneumothorax.² In contrast, in patients with secondary spontaneous pneumothorax, the Belgian Society of Pneumology recommends implementing treatment to prevent recurrence after a first episode because of the high

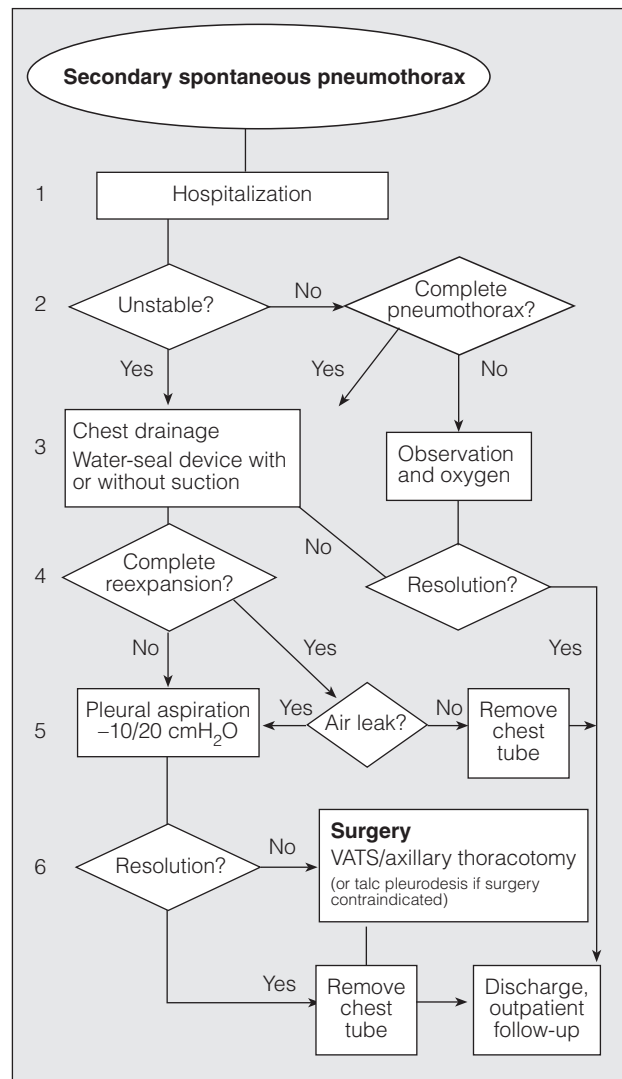


Figure 5. Algorithm for the treatment of secondary spontaneous pneumothorax. VATS indicates video-assisted thoracoscopic surgery.

incidence of recurrence and the exponential increase in mortality in such cases.²⁹

There is even less agreement on the best method of pleurodesis to be used when this procedure is indicated. The ACCP and the Belgian Society of Pneumology both recommend a minimally invasive technique, such as thoracoscopy or VATS, as a first-line approach because of lower morbidity.^{28,29} They reserve axillary thoracotomy with pleural abrasion as a second choice. However, the BTS recommends thoracotomy as a first-line strategy and reserves VATS for patients unable to tolerate open surgery.²

Although no large body of evidence exists, autologous blood-patch pleurodesis may represent an alternative treatment in selected patients who have clinically significant underlying lung disease and persistent air leaks. This option would be chosen precisely to avoid treatments associated with higher rates of morbidity and mortality.^{55,56}

We will now analyze a step by step approach to management based on a treatment algorithm (Figure 5).

Notes on the Treatment Algorithm for Secondary Spontaneous Pneumothorax

1. For all patients with secondary spontaneous pneumothorax hospitalization is recommended in view of the high morbidity and mortality associated with the underlying disease, irrespective of the clinical parameters or the treatment proposed (grade D recommendation).^{2,11,20,29}

2. Patients with a partial secondary pneumothorax who remain clinically and functionally stable do not require simple aspiration or small-bore catheter drainage. The main reason for hospitalizing these patients is to ensure observation and the administration of supplemental oxygen if indicated. Patients with complete pneumothorax—who represent the majority in this group—will all require chest drainage irrespective of whether their condition is stable or unstable (grade C recommendation).^{2,28,29} Simple aspiration is not recommended in such cases, even by the BTS, because of its ineffectiveness.²

3. In stable patients with complete pneumothorax, although small-bore catheter drainage ($\leq 14F$) can be used, a larger tube ($>16F$) is preferable given the presence of underlying disease, and the larger size must be used when there is a risk that the patient may require mechanical ventilation (grade C recommendation).^{2,28,29}

4. In patients under observation, a chest drain should be inserted if there is any increase in the size of the pneumothorax or if dyspnea develops. As in the case of primary spontaneous pneumothorax, an underwater seal system may be sufficient initially, and suction ($-10/20$ cm H_2) will be required if complete reexpansion of the lung is not achieved (grade C recommendation).^{2,28,29}

5. Once complete reexpansion has been achieved and there has been no evidence of air leaks for 24 hours, the chest drain can be removed, but only after resolution has been confirmed radiographically. While many physicians recommend clamping the tube for a short period before removal in case there is an immediate recurrence of the pneumothorax, no consensus has been reached on this point (grade D recommendation).²⁸ Air leaks are common and a more aggressive approach should be considered if they persist for 4 to 7 days because such strategies have been observed to result in improved efficacy and be more cost effective in the long term (grade C recommendation).^{28,30,57} In any case, all such decisions should be made on a case-by-case basis taking into account the clinical situation and organizational circumstances (the patient's age, the immediate availability of an operating room, etc) that may influence the decision about which procedure is preferable.

6. The recommended surgical intervention is abrasive pleurodesis or bullectomy with apical pleurectomy using VATS (for better visualization) or axillary thoracotomy. While thoracotomy with pleurectomy is associated with the lowest recurrence rate, this procedure is considered too aggressive to be used as an initial treatment (grade C recommendation).^{2,28,29}

Intrapleural instillation of talc via a chest tube or thoracoscopy has been shown to be effective in patients with secondary spontaneous pneumothorax and severe COPD, and this treatment is used more widely and is more

justified in this setting than in patients with primary pneumothorax.²⁰ Nevertheless, it should be reserved for patients in whom surgery is contraindicated and cases in which the prognosis for the underlying disease is poor (grade D recommendation).^{2,28,29}

Catamenial Pneumothorax

Catamenial pneumothorax is a rare condition that occurs during menstruation (although premenstrual and even intermenstrual cases have been reported).^{58,59} Between the publication of the first description by Maurer and 2004, a total of 229 cases were published, but it is likely that this condition is underdiagnosed.⁶⁰ Conventional management, which is based on pleurodesis and hormone therapy, is associated with a high failure rate.⁵⁸

The etiology of catamenial pneumothorax is poorly understood and several theories based on clinical and experimental findings have been proposed.⁶¹ The first of these postulates the presence of congenital defects in the diaphragm that allow air to pass into the pleural space during menstruation owing to permeability of the Fallopian tubes. The second hypothesis is based on the same mechanism but suggests that the diaphragmatic fenestrations could be caused by endometriosis. A third hypothesis suggests that endometriosis of the lung parenchyma could give rise to air leaks during menstruation. Finally, prostaglandin F₂, a powerful bronchoconstrictor and vasoconstrictor that could cause alveolar rupture, has been found in some women during menstruation.

Based on these pathogenic mechanisms we can identify 2 groups of patients: those who have diaphragmatic defects and those who do not.⁶² The presence of fenestrations in the diaphragm has been reported in over 50% of the cases described in the literature.⁶⁰ Consequently, most of the therapeutic strategies proposed include local treatment of the diaphragm by suture of the fenestrations, plication of the fenestrated areas of the diaphragm, resection of the membranous part of the diaphragm, or suture and pleurodesis with polyglycolic acid mesh.

No empirical treatment strategy has been clearly defined for the management of cases of catamenial pneumothorax without diaphragmatic defects. Some authors recommend conventional abrasion of the parietal pleura complemented by local treatment of the diaphragm on the basis that imperceptible defects may exist.⁵⁸

While conventional hormone therapy used to suppress ovulation is not effective in most cases, amenorrhea induced by gonadotrophin analogues prevents recurrence of pneumothorax. However, as this hormonal treatment is not tolerated for long periods, it is reserved for use as a supportive therapy after surgery to prevent recurrence during the postoperative period while adhesions are forming on the surface of the diaphragm. Treatment with gonadotropin analogues should always be prescribed and monitored by a gynecologist.

Special Circumstances

Since suppression of the menses is probably the most effective way of preventing recurrence of catamenial

pneumothorax, hysterectomy or adnexectomy in patients who require such treatment for other reasons is a definitive solution. For the same reason, recurrence of pneumothorax is unlikely during pregnancy and breastfeeding, however this effect is temporary because it is related to the associated amenorrhea.

Complications

Patients should be informed about the rare but serious immediate and delayed complications related to pneumothorax and its treatment. These include pulmonary edema caused by abrupt reexpansion of the lung in the case of a large pneumothorax or one that has persisted for several days (10%); hemopneumothorax caused by the laceration of a blood vessel during the insertion of a conventional chest tube or by tearing of the adhesions between the parietal and visceral pleura (5%); vagal reaction and systemic hypotension caused by pain or by edema secondary to lung reexpansion; intercostal neuralgia; pneumomediastinum and subcutaneous emphysema caused by alveolar rupture or incorrect pleural drainage; the presence of a tension pneumothorax (2%-3%); simultaneous bilateral pneumothorax (2%); and failure of the lung to expand, chronic pneumothorax lasting over 3 months, and persistent air leak.¹¹

Appendix

Levels of Evidence⁶³

1++ High quality meta-analyses, systematic reviews of randomized controlled trials (RCTs), or RCTs with a very low risk of bias.

1+ Well-conducted meta-analyses, systematic reviews of RCTs, or RCTs with a low risk of bias.

1? Meta-analyses, systematic reviews of RCTs, or RCTs with a high risk of bias.

2++ High quality systematic reviews of case-control or cohort studies, or high quality case-control or cohort studies with a very low risk of confounding, bias, or chance and a high probability that the relationship is causal.

2+ Well-conducted case-control or cohort studies with a low risk of confounding, bias, or chance and a moderate probability that the relationship is causal.

2? Case-control or cohort studies with a high risk of confounding, bias, or chance and a significant risk that the relationship is not causal.

3 Non-analytic studies, such as case reports and case series.

4 Expert opinion.

Grades of Recommendations⁶³

A At least 1 meta-analysis, systematic review, or RCT rated as 1++ and directly applicable to the target population; or a systematic review, RCT, or a body of evidence consisting principally of studies rated as 1+ directly applicable to the target population and demonstrating overall consistency of results.

B A body of evidence including studies rated as 2++ directly applicable to the target population and

demonstrating overall consistency of results or extrapolated evidence from studies rated as 1++ or 1+.

C A body of evidence including studies rated as 2+ directly applicable to the target population and demonstrating overall consistency of results or extrapolated evidence from studies rated as 2++.

D Evidence level 3 or 4 or extrapolated evidence from studies rated as 2+.

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