4.8 Pneumothorax in CF

Authors: Urs Bürgi, Lars C. Huber, Christian Benden

1. INTRODUCTION

- From registry data the annual incidence of spontaneous pneumothorax in CF is 0.64% (one in 167 patients per year). This complication is associated with
  - older age (>21 years of age) and
  - advanced airways disease (FEV1 <40%)
- The hypothesis for the pathophysiological mechanism of a pneumothorax is similar to that of other lung disease with spontaneous pneumothorax:
  - secretions plugged in the distal airways → air trapping in the alveoli through a check-valve mechanism → elevated alveolar pressure → mechanical damage and rupture of the tissue.
- Factors associated with spontaneous pneumothorax in CF are presented in **Table 1**.
  - The relevance of smoking in CF patients has not been investigated. However, since more than 90% of non-CF patients presenting with spontaneous pneumothorax are smokers, cigarette smoke is likely an additional risk factor also in CF patients.
  - The use of CF-specific inhalation therapies has been associated with development of pneumothorax but causality is unclear.
  - Chronic pulmonary infection with *Staphylococcus aureus* appears to be protective, which is in strong contrast to the development of massive haemoptysis **(see also Chapter "*Haemoptysis*")**.

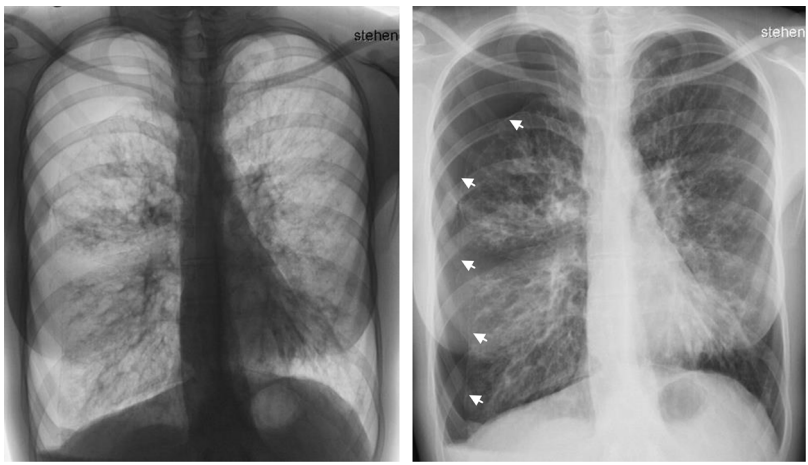
Table 1: Factors associated with spontaneous pneumothorax in CF (adapted from[1])

Risk factors	**Odds ratio (OR)**
Pseudomonas aeruginosa (in sputum)	2.27
Dornase alpha	2.05
Burkholderia cepacia complex (in sputum)	1.78
Inhaled tobramycin	1.60
ABPA	1.48
FEV_1 < 30%	1.46
Pancreatic insufficiency	1.39
Massive haemoptysis	1.37
Aspergillus (in sputum)	1.33
Protective factors	
Staphylococcus aureus (in sputum)	0.90

2. DIAGNOSIS

- The diagnosis of a pneumothorax is straightforward in the presence of suggestive symptoms or signs and is achieved by conventional CXR. In unusual cases (i.e. atypical localization and small size) a chest CT scan is needed.

Figure 1: Pneumothorax of the right lung – margins of the collapsed lung indicated by arrowheads *(Courtesy of PD Dr. T. Frauenfelder, Department of Radiology, University Hospital Zurich).*



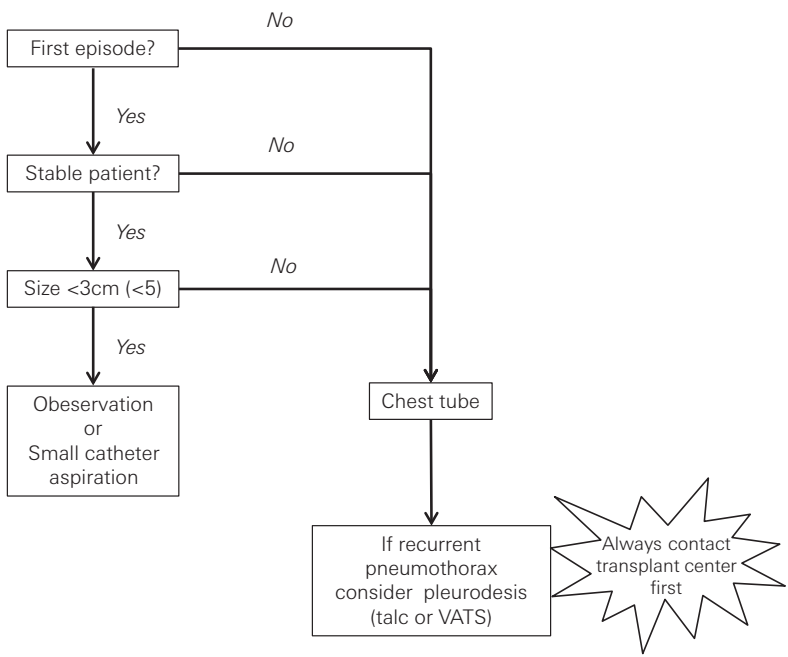
3. MANAGEMENT

- The management of pneumothorax in CF is based on retrospective data and expert opinion due to lack of clinical trials that guide proper therapies.
- The 4 main factors which influence treatment are:
  1) the size of pneumothorax
  2) the severity of the disease
  3) the stability of the patient and
  4) whether this is the first or a recurrent pneumothorax
- A pneumothorax is considered small if the distance between apex and cupola is <3cm (or <5cm, depending on the reference). These patients may be managed in an outpatient setting whereas all patients with a large pneumothorax should be admitted to the hospital. A management algorithm is provided in **Figure 2**.
- *Pleurodesis* is recommended for recurrent large pneumothoraces. **The transplant center/surgeon should always be contacted previous to pleurodesis, as it may complicate a future lung transplantation (see also Chapter "*Transplantation*")**.
  - *Surgical pleurodesis* (video-assisted thoracic surgery, VATS) is the preferred method.
  - *Chemical pleurodesis* (i.e. talc) is also an effective alternative if surgery is not tolerated by the patient.

- Although contralateral pneumothorax commonly develops in the course of disease, prophylactic pleurodesis on the opposite side is not recommended.

- *Airway clearance therapy* **(see also Chapter "*Physiotherapy*")**:
  - In small pneumothoraces the daily airway clearance therapy can be continued with exception of methods that employ high positive pressure such as IPV (intermittent percussive ventilation), PEP (positive expiratory pressure) and oscillating PEP.
  - In cases of large pneumothoraces all airway clearance methods should be stopped.
- The *aerosolized medication* can be continued even in large pneumothoraces.
- *BiPAP-therapy* should be withheld if possible until the pneumothorax has resolved.
  - In patients with concomitant hypercapnic respiratory failure, the development of a pneumothorax might result in a management dilemma. This might particularly be true for patients listed for lung transplantation. In such patients, BiPAP-therapy has been reported to be used without increasing the size of the pneumothorax. However, in most of these cases, chest tube drainage is appropriate since use of BiPAP-therapy appears to be safe after insertion of a chest drain.
- *Activities* like flying, lifting weights and performing spirometry should be postponed for a minimum of 2 weeks after resolution of the pneumothorax (see also Chapter *"Traveling, altitude, diving"*). Aerobic exercise can be continued.
- After resolution, *associated causes* (such as ABPA) should be evaluated and treated.

Figure 2: Management algorithm for spontaneous pneumothorax.



4. OUTCOME

- A pneumothorax has adverse effects on lung function and is associated with high health-care costs.
- Patients may die acutely as a consequence of a pneumothorax.
  - The estimated mortality is 6.3-14.3%
  - The 2-year mortality rate following a pneumothorax is high (49% versus 12% of patients without pneumothorax).
- Patients with recurrent or refractory pneumothoraces should be evaluated for lung transplantation.
  - Previous pleurodesis is not an absolute contraindication for lung transplantation but this might be handled center-specifically. For this reason, **the transplant center/surgeon should always be contacted previous to pleurodesis in a CF patient that is a potential candidate for lung transplantation.**

5. REFERENCES

1. Flume PA. Pneumothorax in cystic fibrosis. Current opinion in pulmonary medicine 2011;17:220-5.
2. Curtis HJ, Bourke SJ, Dark JH, Corris PA. Lung transplantation outcome in cystic fibrosis patients with previous pneumothorax. J Heart Lung Transplant 2005;24:865-9.
3. Flume PA, Mogayzel PJ, Jr., Robinson KA, Rosenblatt RL, Quittell L, Marshall BC. Cystic fibrosis pulmonary guidelines: pulmonary complications: hemoptysis and pneumothorax. Am J Respir Crit Care Med 2010;182:298-306.
4. Flume PA, Strange C, Ye X, Ebeling M, Hulsey T, Clark LL. Pneumothorax in cystic fibrosis. Chest 2005;128:720-8.