

CF Guidelines - Pneumothorax

Management of Pneumothorax in Cystic Fibrosis:

The incidence of pneumothorax increases with age (overall 8%) and is a marker of severe lung disease. It carries a bad prognosis, particularly if the chest drain cannot be rapidly removed.

A high index of suspicion is needed - consider the diagnosis in case of:

- Unexpected deterioration.
- Unexplained chest pain.
- Worsening breathlessness.

Diagnosis:

- Standard inspiratory plain CXR - preferably PA.
- CT scan may be needed if extensive lung disease.

Classification- all pneumothoraces in CF are secondary, by definition:

- Spontaneous vs traumatic (blunt) vs iatrogenic.
- Size: small (< 2 cm rim of air) or large (> 2 cm rim of air).
- Tension: a pneumothorax of any size associated with evidence of cardiovascular decompensation or radiological evidence of mediastinal shift away from the pneumothorax.

Management:

- All patients will require hospital admission.
- Contact the (adult or paediatric) team responsible for CF patients at your hospital, regardless of time of admission.
- Monitor SaO₂ and give oxygen.
- Intercostal drainage (see below).
- Local anaesthesia and subsequent oral analgesia.
- Antibiotics (iv antibiotics are prudent in all but the most trivial pneumothorax).
- Gentle physiotherapy should be continued as tolerated. Techniques may need changing (no positive pressure adjuncts). Deep breathing with inspiratory holds are encouraged. Please discuss this with a senior respiratory physiotherapist.
- Non-invasive ventilation should be avoided until a functioning chest drain is in situ.

Tension pneumothorax:

This is an emergency that requires urgent treatment with a chest drain, regardless of the CF. Where life threatening prompt insertion of a intravenous cannula through the 2nd intercostals space in the mid-clavicular line can be lifesaving. In adults a cannula of at least 4.5 cm in length is recommended.

Traumatic pneumothorax:

Pneumothorax associated with significant chest trauma should be initially managed as per any other trauma patient. This aspect of management is outside the scope of these guidelines. Additional considerations including antibiotic therapy and surgical options should be considered in the light of these guidelines.

Iatrogenic pneumothorax:

Small pneumothoraces following central venous cannulation and pleural procedures should be managed as per spontaneous pneumothorax.

Options for intercostal drainage:

- Observation:
 - A small asymptomatic pneumothorax can be managed by observation alone and may resolve but in an already hypoxic patient, such a leak may cause decompensation. Patients treated with observation should be given high flow oxygen continuously to facilitate gas re-absorption unless there is blood gas evidence of CO₂ retention. This approach will generally be applicable to patients with minimal symptoms and an air rim of < 1 cm where drainage may be hazardous.

- Simple aspiration:
 - Simple aspiration may be appropriate for small, minimally symptomatic pneumothoraces. This is less likely to succeed in patients with severe lung disease which is the case with most CF patients presenting with spontaneous pneumothorax. It may be considered for iatrogenic pneumothorax in patients with relatively well preserved lungs. Guidelines for performing simple aspiration are available on the BTS website (<http://www.brit-thoracic.org.uk>).

- Intercostal tube drainage:
 - This is likely to be necessary in most CF patients with spontaneous pneumothorax. There is no evidence that tube size influences outcome. Small bore drains inserted using the Seldinger technique may be preferable in initial treatment.

- A large bore chest drain may be indicated when:
 - Initial small bore drainage fails.
 - In case of tension pneumothorax.
 - Lung adhesions are visible on CXR or suggested by prior pleural intervention – a more “surgical” approach or CT guidance is preferable.
 - Even small pneumothoraces may be symptomatic. These may need CT guided aspiration (with small bore tube).

Drains should always be connected to an underwater seal. Guidelines for insertion of an intercostal tube drain are available on the BTS website (<http://www.brit-thoracic.org.uk>). Don't forget to start appropriate IV antibiotics for respiratory infection.

Failure of resolution:

The lung may be slow to re-expand and if after 48 hours there are no signs of resolution with a continuing air leak then:

- Consider up-sizing to a large bore drain if appropriate.
- Consider suction using a high volume, low pressure (-10 to -20 cm water) system.
- Consult with thoracic surgeons. Discussion with a surgeon at a likely transplant centre for the patient may be helpful if local thoracic surgeons are in doubt about preferred surgical options.
- Within the Peninsula transfer patient to Exeter or Plymouth for further management if drain still leaking after 48 hrs or immediately if patient is in respiratory failure.

Surgery:

Surgery should be considered if no progress is being made. Discuss patients with thoracic surgeons earlier rather than later. In some centres there is a 50% mortality if

a patient has a chest drain for more than one week. Similarly, recurrences are common (> 50% ipsilateral and up to 40% contralateral) necessitating surgery.

The surgical treatment of choice is:

- VATs procedure rather than thoracotomy.
- Stapling of blebs or emphysematous leaks.
- Mild, localised abrasion pleurodesis if a pleurodesis is required at all.
- Avoid pleurectomy or a sclerosing pleurodesis with talc. These make subsequent transplant much more difficult and should be avoided if possible, although they may not be an absolute contraindication to future transplantation.
- Patients are generally advised not to fly for 6 weeks after a pneumothorax (the evidence to support this advice is scant).

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