

## section 2.6.1: End stage lung disease

### General description of the disease and its progression

Lung diseases are classified by two groups according to physiology:

- **Obstructive:** blocks or obstructs ventilation by a combination of mucous secretions, airway spasm, airway inflammation or alveolar destruction – conditions include COPD, cystic fibrosis, sarcoidosis, overlap syndrome (COPD and sleep apnoea)
- **Restrictive:** neuromuscular (eg MND), thoracic deformities, mesothelioma, interstitial lung disease.

Most patients referred to palliative care services with an end stage chronic lung disease will have a diagnosis of COPD. COPD is currently one of the major causes of death globally and is the fifth leading cause of death for males and the seventh most common cause of death among females in Australia in 2003.<sup>1</sup> The incidence is increasing in women due to the uptake of smoking. Patients with severe COPD are often disabled by their disease for longer, and have a mortality rate comparable to that of many common cancers. COPD can be viewed as a disease with similarities to cancer, however with a longer pattern of trajectory.

### Staging

There is currently no consensus on what end-stage is – however the needs of patients are usually complex, they have a BODE score of 7-10<sup>2</sup> and poor functional status. The course of disease is generally marked by increasingly frequent hospital admissions with exacerbations and increasingly disabling dyspnoea. Significant weight loss and the onset of respiratory failure are often evident in the last 6-12 months of life.

The overall prognosis for a patient with COPD depends on the severity of lung disease and whether the patient continues to smoke. The BODE index, a simple multidimensional grading system, is better than the FEV<sub>1</sub> at predicting the risk of death for patients with COPD.<sup>3</sup>

### Main symptoms in the last year of life

- **Dyspnoea:** increases over time and on minimal exertion (eg even with conversation) – the Medical Research Council (MRC) dyspnoea scale is a useful measurement tool to assess deterioration
- **Cough:** productive or non productive, is usually productive and worse in the morning – can cause rib fractures especially in patients with steroid induced osteoporosis
- **Psychiatric morbidity:** depression and/or anxiety are very common and have been estimated to affect 20-60% of COPD patients<sup>4</sup>
- **Weight loss and anorexia** are common problems in advanced COPD
- **Cor pulmonale:** right sided heart failure secondary to chronic lung disease, clinical features are elevated JVP, hepatomegaly and peripheral oedema
- **Sleep disturbance** is extremely common in this population – contributed to by dyspnoea (especially orthopnoea) anxiety and cough

### Specific considerations for end stage disease

- patient requirements for community-based supports are reviewed
- assessment of caregiver needs
- discussion regarding end of life decision making and advance directives is facilitated
- refer to clinical pathways that have been developed in your hospital

## section 2.6.1: End stage respiratory failure

### General description of the disease and its progression

A clinical term used to describe failure of the lungs to maintain oxygenation. There are 2 types of respiratory failure:

- **Type 1:** this mainly relates to hypoxaemia (low blood oxygen levels) without hypercapnia (elevated carbon dioxide levels) – it can be found in patients with asthma, pulmonary fibrosis, pulmonary oedema and pulmonary embolus
- **Type 2:** this refers to hypoxaemia with hypercapnia and is associated with alveolar hypoventilation often evident with neuromuscular disorders (eg MND), kyphoscoliosis, COPD

### Blood gas measurements

In advanced COPD measurement of blood gases is important. This test should be performed in patients with FEV<sub>1</sub> <30% predicted or with clinical signs suggestive of right heart failure. Clinical signs include central cyanosis, ankle swelling, and an increase in the jugular venous pressure.

Clinical signs of hypercapnia include morning headache on waking which resolves within a few hours, confusion, lethargy, somnolence, bounding peripheral pulse and asterixis – these signs are not always a reliable indicator and may also indicate hypoxaemia. Screening patients by pulse oximetry and assessing arterial blood gases in those with an oxygen saturation (SaO<sub>2</sub>) <90% may be a useful way of selecting patients for blood gas measurement. However, pulse oximetry gives no information about CO<sub>2</sub> tensions.

### Oxygen therapy

Untreated hypoxaemia associated with COPD has a high mortality rate. Oxygen is only prescribed if a patient has a persistent Po<sub>2</sub> of <55mmHg or Po<sub>2</sub> of <60mmHg with right-sided heart failure, evidence of pulmonary hypertension or significant desaturation following a 6-minute walk test.<sup>5</sup> The flow rate is prescribed by a respiratory physician in the context of the patient's blood gas measurement. Most oxygen therapy is prescribed for at least 15 hours daily, usually during sleep and during daytime exertion.

### Opioids

There has been much debate about the use of opioids in the treatment of dyspnoea in this patient population. A recent double blind randomized cross-over study<sup>6</sup> has shown that a small dose of slow release opioids taken once daily can reduce the severity of this difficult symptom. Some patients prefer to take an "as required" low dose of morphine syrup. Constipation should be treated prophylactically.

### Respiratory rehabilitation

This is now recognized as a vital part of COPD treatment. It has been shown to improve quality of life, exercise tolerance and mastery of disease when used in moderate stage disease. It is entirely appropriate for many of the elements of a respiratory rehabilitation programme to be modified for patients in the palliative stage of the disease. Breathing retraining and relaxation techniques for example can be used up until the terminal phase.

### Special considerations

- encourage consideration of end of life decision making and advance directives
- role of diuretics for symptom relief
- for specific symptom management see Therapeutic Guidelines – Palliative Care<sup>7</sup>

- presence of co-morbid illness such as IHD, PVD, diabetes, osteoporosis and presence of lung cancer
- volunteer support may be valuable as the patient may not be able to be left alone for any length of time

## references

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- <sup>1</sup> Australian Institute of Health and Welfare, Incidence and prevalence of chronic diseases, see [www.aihw.gov.au](http://www.aihw.gov.au)
  - <sup>2</sup> Celli BR, Cote CG, Marin JM, et al. (2004) The body-mass index, airflow obstruction, dyspnea, and exercise capacity index in chronic obstructive pulmonary disease. *N Engl J Med*; 350:1005-1012.
  - <sup>3</sup> Celli BR, Cote CG, Marin JM, et al. (2004) The body-mass index, airflow obstruction, dyspnea, and exercise capacity index in chronic obstructive pulmonary disease. *N Engl J Med*; 350:1005-1012
  - <sup>4</sup> Norwood R (2006) Prevalence and impact of depression in chronic obstructive pulmonary disease patients *Curr Opin Pulm Med*. Mar;12(2):113-7
  - <sup>5</sup> Young I, Crockett A, McDonald C (2005) Adult domiciliary oxygen therapy- position statement of the Thoracic Society of Australia and New Zealand *MJA* 182 (12): 621-626
  - <sup>6</sup> Abernethy A, Currow D, Frith P, Fazekas B, McHugh A, Bui C(2003) Randomized, double blind, placebo controlled crossover trial of sustained release morphine for the management of refractory dyspnoea *BMJ* 327:523-528.
  - <sup>7</sup> Therapeutic Guidelines- Palliative Care, Version 2 (2005), Therapeutic Guidelines Ltd, Victoria, Australia.