Chronic Obstructive Pulmonary Disease

Chronic obstructive pulmonary disease (COPD) is characterised by airflow obstruction that is not fully reversible. The airflow limitation is usually progressive and is associated with an abnormal inflammatory response of the lungs to noxious particles or gases.\[1\] The airflow obstruction is due to a combination of airway and parenchymal damage. COPD is now the preferred term for patients with airflow obstruction who were previously diagnosed as having chronic bronchitis or emphysema.\[2\]

Airflow obstruction is defined as a reduced post-bronchodilator FEV1/FVC ratio (where FEV1 is forced expiratory volume in 1 second and FVC is forced vital capacity), such that FEV1/FVC is less than 0.7. If FEV1 is 80% or more of predicted normal, a diagnosis of COPD should only be made in the presence of respiratory symptoms - eg, breathlessness or cough.\[2\]

The respiratory drive is normally largely initiated by PaCO\_2 but in COPD hypoxia can be a strong driving force, which can therefore be reduced if the hypoxia is corrected.

Epidemiology\[2\]

The damage to the lungs is the result of chronic inflammation, which is usually the result of tobacco smoke. Other factors, particularly occupational exposures, may also contribute to the development of COPD.

- An estimated three million people are affected by COPD in the UK. COPD is underdiagnosed. 60-85% of patients, mainly with mild-to-moderate disease, are thought to remain undiagnosed.\[3\]
- In the UK, it is estimated that more than 3 million people currently have COPD, and an estimated 2 million people have COPD which remains undiagnosed.\[4\]
- Most patients are not diagnosed until they are in their fifties. COPD is closely associated with levels of deprivation - rates of COPD are higher in more deprived communities.
- COPD is often associated with comorbidities, particularly cardiovascular disease, lung cancer, osteoporosis, muscle weakness and cachexia.\[5\]

Presentation

See also the separate articles on Respiratory System - History and Examination and Diagnosing COPD.

- A diagnosis of COPD should be considered in patients over the age of 35 who have a risk factor (generally smoking) and who present with exertional breathlessness, chronic cough, regular sputum production, frequent winter 'bronchitis' or wheeze.\[2\]
- COPD may also cause abnormal weight loss, effort intolerance and ankle oedema.

Smoking: an up-to-date smoking history, including pack years smoked (number of cigarettes smoked per day, divided by 20, multiplied by the number of years smoked) should be documented for everyone with COPD. An assessment of their 'readiness to change' should also be made.

Signs

- Respiratory distress: tachypnoea, breathlessness on exertion, increased use of accessory muscles of respiration, pursed lip breathing.
- Abnormal posture: patients may lean forward and rest their arms on the table to ease breathing.
- Drowsiness, flapping tremor and mental confusion (these are features of elevated carbon dioxide levels).
- Other signs include being underweight, ankle oedema, cyanosis, hyperinflation of the chest, downward displacement of the liver, relatively quiet vesicular breath sounds, wheezing, prolonged forced expiratory time.
Disease severity and staging

- Disability in COPD can be poorly reflected in the FEV1. A more comprehensive assessment also includes:
  - Degree of airflow obstruction and disability.
  - Frequency of exacerbations.
  - Prognostic factors such as breathlessness (Medical Research Council (MRC) dyspnoea scale), carbon monoxide lung transfer factor, health status, exercise capacity, body mass index (BMI), partial pressure of oxygen in arterial blood (PaO₂) and presence of cor pulmonale. See article on Breathlessness for explanation of MRC dyspnoea scale.

- Investigate symptoms that seem disproportionate to the spirometric impairment, using a CT scan or lung transfer factor for carbon monoxide testing.

- The BODE index (BMI, airflow Obstruction, Dyspnoea and Exercise capacity index) should be used to assess the prognosis when the component information is available: measurement of the BODE index includes measurement of BMI, FEV1 as a percentage of predicted, dyspnoea (modified MRC score) and exercise tolerance (six-minute walking distance).

- Assess the severity of airflow obstruction by FEV1 as a percentage of predicted:
  - Stage 1 - mild: 80% or above (symptoms should be present to diagnose COPD in people with mild airflow obstruction).
  - Stage 2 - moderate: 50-79%.
  - Stage 3 - severe: 30-49%.
  - Stage 4 - very severe: below 30% (or FEV1 less than 50% but with respiratory failure).

The GOLD (Global Initiative on Obstructive Lung Disease) classification of severity is also still used:

- Stage 0: at risk; chronic cough and sputum production. Spirometry is normal.
- Stage I: mild COPD; mild airflow limitation (FEV1/FVC) less than 70% but FEV1 80% or more than predicted; usually, but not always, chronic cough and sputum production.
- Stage II: moderate COPD; worsening airflow limitation (FEV1 50-79% predicted) and usually progression of symptoms, with shortness of breath, especially on exertion.
- Stage III: severe COPD; further worsening of airflow limitation (FEV1 30-50% predicted), increased shortness of breath, and repeated exacerbations.
- Stage IV: very severe COPD; severe airflow limitation (FEV1 less than 30% predicted) or the presence of chronic respiratory failure.

Differential diagnosis

- **Asthma**: diagnosed by establishing reversibility or variability of airflow obstruction either by spirometry or peak flow measurements after treatment with a bronchodilator or steroid.
- Other diagnoses to consider are congestive heart failure, bronchiectasis, allergic fibrosing alveolitis, pneumoconiosis, asbestosis or other restrictive lung conditions, tuberculosis, lung cancer, obliterative bronchiolitis, bronchopulmonary dysplasia, anaemia or generally poor physical condition.

Investigations

- There is no single diagnostic test for COPD. The diagnosis is therefore based on a combination of history, examination and confirmation of the presence of airflow obstruction, using spirometry.
- The presence of airflow obstruction should be confirmed by performing post-bronchodilator spirometry. Spirometry is considered the gold standard test for diagnosing COPD. See also the separate article Spirometry Calculator.
- In patients with clinical and spirometry evidence of COPD, reversibility testing adds little to management.
- Initial evaluation should also include CXR to exclude other diagnoses (investigate abnormalities using a CT scan), FBC (to identify anaemia or polycythaemia) and BMI calculation.
- In younger patients, or in those who are not exposed to cigarette smoke or other factors known to be associated with COPD, consider a genetic cause such as alpha-1-antitrypsin (A1AT) deficiency.
Management

- See the separate articles on Management of Stable COPD, Acute Exacerbations of COPD, Use of Oxygen Therapy in COPD and Pulmonary Rehabilitation.
- An effective COPD management plan includes prevention (reduction of risk factors, particularly smoking cessation), assessment and monitoring of disease and its progression, and prevention of infection (all patients with COPD should be offered pneumococcal vaccination and an annual influenza vaccination). [2]
- Consider referring people with excessive sputum to a physiotherapist. Consider referring people to social services and occupational therapy if they have difficulties with activities of daily living or disability. [4]

Referral [2]

Referral for advice, specialised investigations or treatment may be appropriate at any stage of disease, not just for people who are severely disabled. Possible reasons for referral include:

- Diagnostic uncertainty.
- Suspected severe COPD.
- The individual requests a second opinion.
- Onset of cor pulmonale.
- Assessment for oxygen therapy, long-term nebuliser therapy or oral corticosteroid therapy.
- Bullous lung disease.
- Rapid decline in FEV1.
- Assessment for pulmonary rehabilitation.
- Assessment for lung volume reduction surgery or lung transplantation.
- Dysfunctional breathing.
- Onset of symptoms at age under 40 years or a family history of A1AT deficiency.
- Symptoms disproportionate to lung function deficit.
- Frequent infections.
- Haemoptysis.

Indications for surgery

- Refer patients who are breathless, have a single large bulla on a CT scan and an FEV1 of less than 50% predicted for consideration of bullectomy.
- Refer people with severe COPD for consideration of lung volume reduction surgery if they remain breathless with marked restrictions of their activities of daily living, despite maximal medical therapy (including rehabilitation), and meet all of the following:
  - FEV1 greater than 20% predicted.
  - PaCO2 less than 7.3 kPa.
  - Upper lobe predominant emphysema.
  - Carbon monoxide lung transfer factor greater than 20% predicted.

- Lung transplantation:
  - Consider referring people with severe COPD for assessment for lung transplantation if they remain breathless with marked restrictions of their activities of daily living despite maximal medical therapy.
  - Considerations include age, FEV1, PaCO2, homogeneously distributed emphysema on CT scan, elevated pulmonary artery pressures with progressive deterioration, comorbidities and local surgical protocols.

Complications

- Chronic hypoxaemia causes slowly progressive pulmonary hypertension with the development of right ventricular hypertrophy and possible cor pulmonale.
- Pneumothorax.
- Respiratory failure.
- Arrhythmias, including atrial fibrillation.
- Infection.
Secondary polycythaemia.

Prognosis[2]

The BODE index has been shown in a number of studies to be a better predictor of exacerbations, hospital admissions and mortality than using only FEV1. See Reference 6 below for explanation of BODE index.[6]

- COPD is progressive and patients deteriorate but the natural history of the disease varies in different people.
- COPD is the fifth leading cause of death in the UK. More than 90% of COPD-related deaths occur in the over-65 age group. COPD is an important comorbidity in those dying from other smoking-related diseases, especially ischaemic heart disease and lung cancer.
- Five-year survival from diagnosis is 78% in men and 72% in women with clinically mild disease (defined as not requiring continuous drug therapy), but falls to 30% in men and 24% in women with severe disease defined as requiring oxygen or nebulised therapy.
- The mean age of death of patients with severe COPD is 74.2 years compared with 77.2 years in patients with mild disease and 78.3 years in individuals who did not have COPD.
- In patients who stop being exposed to cigarette smoke and other noxious substances the disease may continue to progress but the rate of declining lung function may slow.[1]
- Repeated exacerbations lead to irreversible decline in lung function and efforts should therefore be made to reduce exacerbations. Patients who have frequent exacerbations have a more rapid decline in lung function, poorer quality of life, and greater mortality.[8]
- Increased rates of hospital admissions for exacerbations are associated with increasing risk of death.[9]

Prevention

- Smoking cessation and restriction of other potential risk factors - eg, occupational dusts and chemicals.
- Reduce the risk of exacerbations - eg, influenza and pneumococcal immunisation.

Further reading & references

1. Documents & Resources; Global Initiative for Chronic Obstructive Lung Disease (GOLD)
2. Chronic obstructive pulmonary disease; NICE Clinical Guideline (June 2010)
4. Chronic obstructive pulmonary disease; NICE CKS, November 2010 (UK access only)

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