Clinical Practice Review Respiratory disease

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In this article...

- Characteristics and prevalence of chronic obstructive pulmonary disease
- Signs and symptoms and the role of spirometry and assessment tools
- Understanding disease progression and prognosis

COPD 1: pathophysiology, diagnosis and prognosis

Key points

Chronic obstructive pulmonary disease is a common respiratory condition, with significant mortality and morbidity

The disease is an umbrella term for a group of conditions involving progressive and irreversible lung damage

Symptoms are often referred to as a 'smoker's cough' or a natural part of ageing rather than an incurable disease

Early and accurate diagnosis can make a real difference to patients' lives

Care and management plans should reflect the highly individual nature of the disease Author Stephen Gundry is COPD nurse, Newcastle Hospitals NHS Foundation Trust.

Abstract Chronic obstructive pulmonary disease is a common and incurable respiratory condition that is largely preventable and treatable, pharmacologically and non-pharmacologically. It involves progressive and permanent damage to lung structures, leading to symptoms of breathlessness, cough, wheeze and sputum production. Early diagnosis and treatment allows patients to benefit from symptom-relieving treatment to maximise their quality of life. This article, the first in a two-part series, describes its pathophysiology, diagnosis and prognosis.

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hronic obstructive pulmonary disease (COPD) is a common respiratory condition, affecting 4.5% of people over the age of 40 in the UK (statistics.blf.org.uk/copd). The British Lung Foundation estimates 1.2 million people have been diagnosed with COPD, and this is thought to represent a third of people who have the disease, many are as yet undiagnosed. COPD is responsible for nearly 30,000 deaths a year or around 5.3% of all UK deaths; in Europe, the UK lags only behind Denmark and Hungary in mortality rates for COPD, and ranks 12th worldwide in terms of deaths per million population a year (statistics.blf.org.uk/copd).

Lung health is one of the priorities in the NHS Long Term Plan, as part of a recognition of the needs of patients with long-term conditions, including COPD (NHS England, 2019). The plan includes a commitment to improve the availability and quality of spirometry to support accurate and timely diagnosis, and highlights the value of pulmonary rehabilitation and the need to expand the scope of rehabilitation programmes to include more patients. It also acknowledges the importance of patients receiving the correct inhaled medication

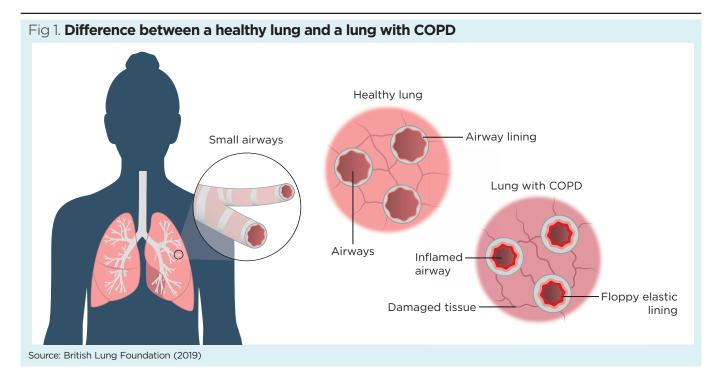
and the need for medication reviews, including the correct use of inhalers.

Although COPD is a significant concern countrywide, BLF figures show its distribution is uneven across the UK population; prevalence is highest in the North East, North West and Scotland, and the disease is three times more common in the most deprived populations compared with populations that are least deprived (statistics. blf.org.uk/copd).

While the prevalence of COPD is rising, and more people have a diagnosis than ever before, since early 2000 the number of new diagnoses has been slowing (statistics.blf. org.uk/copd). However, it is not clear whether this is due to a fall in the number of people developing COPD or changes to record-keeping practice. One possible explanation could be the ongoing reduction in tobacco smoking over recent decades, but this is not a reason for complacency and early diagnosis of COPD is essential. The insidious onset of the disease means patients may dismiss early symptoms, such as a cough and subtle increases in breathlessness, as normal age-related changes or 'smoker's cough', instead of a serious condition that needs medical assessment.

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BLF figures also show around 10% more men than women have a COPD diagnosis. Although the difference is slowly narrowing, this has been a long-term finding and reflects the greater incidence of smoking among men over previous years.

What is COPD?

The Global Initiative for Chronic Obstructive Lung Disease (2018) defines COPD as "a common, preventable and treatable disease that is characterised by persistent respiratory symptoms and airway limitation due to airway and/or alveolar abnormalities usually caused by significant exposure to noxious particles of gases". It is an evolving condition that progresses over time, although the rate of progression is widely heterogeneous and varies unpredictably from one individual to another (GOLD, 2019). COPD is an umbrella term that covers:

- Emphysema (loss of alveolar structure);
- Chronic bronchitis (long-term inflammation of the airways and mucus hyper-secretion).

The persistent respiratory symptoms consistent with the disease reflect the permanent changes that take place in the lung structures and include breathlessness, cough and sputum production.

The most important cause of COPD in the UK and other western countries is exposure to tobacco smoke – usually as a result of smoking cigarettes. However exposure to any irritant, noxious airborne particles (for example, organic and inorganic dusts) and chemical fumes that can be inhaled into the lungs present an underestimated risk of COPD (GOLD, 2019).

Pathophysiology

COPD results from the combined processes of peripheral airway inflammation and narrowing of the airways. This leads to airflow limitation and the destruction and loss of alveoli, terminal bronchioles and surrounding capillary vessels and tissues, which adds to airflow limitation and leads to decreased gas transfer capacity (Fig 1). The extent of airflow limitation is determined by the severity of inflammation, development of fibrosis within the airway and presence of secretions or exudates. Reduced airflow on exhalation leads to air trapping, resulting in reduced inspiratory capacity, which may cause breathlessness (also known as dyspnoea) on exertion and reduced exercise capacity.

Abnormalities in gas transfer occur due to reduced airflow/ventilation and as a result of loss of alveolar structure and pulmonary vascular bed. Low blood oxygen levels (hypoxaemia) and raised blood carbon dioxide levels (hypercapnia) result from impaired gas transfer and can worsen as the disease inevitably progresses.

Diagnosis

National Institute for Health and Care Excellence guidance advocates early diagnosis of COPD, so that patients can benefit from symptom-relieving treatment to maximise quality of life (NICE, 2018). NICE says COPD should be suspected in people:

- Over the age of 35 who smoke;
- Who have smoked in the past and have one or more supporting symptoms, such as chronic cough, sputum production, exceptional breathlessness, wheeziness or previous 'frequent winter bronchitis'.

Accurate spirometry supports diagnosis of COPD, with obstructive lung disease confirmed by a ratio below 0.7 (70%) of forced expiratory volume per second (FEV1)/forced vital capacity (FVC) (NICE, 2018) (see Box 1). Spirometry is a reliable and valuable means of measuring lung

Box 1. Diagnosing COPD using spirometry

- Forced vital capacity (FVC): the maximum amount of air the patient can blow out from a full inspiration to full expiration during a forced blow
- Forced expiratory volume in 1 second (FEV1): the maximum volume of air the patient can blow out in the first second of a forced blow
- FEV1/FVC: the volume of air expired during the first second of a forced blow, expressed as a percentage of FVC

Source: Hughes (2017)

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function in terms of severity of airflow limitation, and of assessing and monitoring future lung function decline. It provides a useful measure of a patient's response to bronchodilator medications and helps differentiate between obstructive disease characterised by compromised airflow (as occurs in COPD) and restrictive disease of reduced lung volume due to parenchymal scarring, pleura or chest wall disease (for example, interstitial lung disease).

Recorded FEV1, as a percentage of an individual's predicted value (based on age, gender, height and ethnicity), is generally used to classify the severity of COPD. However, this correlates weakly with a patient's symptoms and functioning (GOLD, 2019), and assessment tools are important to help establish the extent of limitation imposed by COPD. There are many assessment tools available, for example:

- Medical Research Council dyspnoea/ breathlessness scale. This uses patients' perceived respiratory disability in terms of their mobility and is a simple, easily administered tool that is valid, widely adopted and quickly estimates physical limitation (Table 1);
- COPD assessment test (CAT). A widely used, validated, simple and selfcompleted instrument that helps reveal the extent of COPD impact on an individual (www.catestonline.org/).

"The individual's heightened perception of their effort of breathing determines their experience of breathlessness."

Signs and symptoms

The characteristic symptoms of COPD are:

- Breathlessness;
- Cough;
- Sputum production;
- Wheeze (Kumer and Clark, 2006).

The most obvious symptom is exertional breathlessness, as it is uncomfortable for the patient and is easily observed by clinicians making an observational assessment. The sensation of breathlessness is subjective and consists of distinct sensations that cause discomfort and distress. Breathlessness varies between individual patients and can be associated with anxiety, depression, decreased health-related quality of life and risk of mortality (Anzueto and Miravitlles, 2017). It is a complex phenomenon, chiefly the result of activation or stimulation of mechanical

Table 1. Medical Research Council dyspnoea scale/breathlessness scale

Grade	Degree of breathlessness related to activities
1	Not troubled by breathlessness except on strenuous exercise
2	Short of breath when hurrying or walking up a slight hill
3	Walks slower than contemporaries on level ground because of breathlessness, or has to stop for breath when walking at own pace
4	Stops for breath after walking about 100 meters or after a few minutes on level ground
5	Too breathless to leave the house, or breathless when dressing or undressing
C A II I D I C III I A DCD	

Source: Medical Research Council bit.ly/MRCDyspnoea

pathways associated with the increased workload of breathing (Coccia et al, 2016), and the individual's heightened perception of their effort of breathing determines their experience of breathlessness.

A cough is a persistent and troublesome symptom. Not all patients have a productive cough, though many do, and a cough is not necessarily associated with the extent of airflow limitation (GOLD, 2018). The first clue to the development of COPD may be a persistent 'smoker's cough', which patients who smoke may accept as the price of their smoking habit, rather than the onset of serious disease.

Excess sputum production is also common in COPD and results from the overproduction and hyper-secretion of mucus from goblet cells, compounded by a reduction in the elimination of mucus (Ramos et al, 2014). In the absence of other signs or symptoms, the definition of chronic bronchitis as a COPD condition is "regular sputum production for three or more months in two consecutive years". (GOLD, 2018). Changes to the characteristics of sputum are significant and the onset of bacterial infection may be indicated by the development of purulent (offwhite, yellow or green) sputum. Wheeziness is a feature of COPD that may be persistent or variable and both inspiratory and expiratory in presentation. It is described as the sound created by air escaping from narrowed airways and is typically a high-pitched whistling, but the absence of wheeze does not exclude COPD (GOLD, 2018).

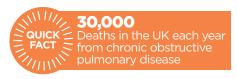
NICE describes an exacerbation of COPD as a "sustained worsening of symptoms from a person's stable state" that is beyond usual day-to-day variation and of rapid or acute onset, which can include:

- More troublesome or severe breathlessness;
- Increased sputum and purulence or a

darkening of sputum colour;

- Worsening cough;
- Increased wheeziness;
- Raised heart rate;
- Respiratory rate of 20% or more above normal;
- Upper respiratory tract infection in the last five days;
- Raised temperature with no other obvious cause (NICE, 2018).

The cause of an exacerbation may be linked to viral or bacterial infection, or be non-infective and of uncertain cause; it can also be associated with smoking or air pollution (Viniol and Vogelmeier, 2018). An exacerbation is a significant and complex event in the patient's journey, associated with increased disease progression and reduced health status (Wedzicha et al, 2014), and the aim of treatment is to mitigate the detrimental effect of the exacerbation on the patient, their need for additional health and social care and ultimately mortality.



Progression and prognosis

Although COPD is specifically defined, it is a vastly heterogeneous condition and the experience of living with it differs from one individual to another, both in its impact on quality of life and manifestation of the disease. There are no single or multidimensional indices that can accurately measure or predict prognosis, but there are numerous individual and interrelated factors that can be influential and linked to prognosis that NICE recommends taking into account when developing patient care or management plans (Box 2). Many of these may or will require specific intervention, therapy or treatment, often

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Box 2. Individual factors independently associated with disease progression and mortality

The National Institute for Health and Care Excellence recommends that from diagnosis onwards, health professionals should think about the following factors that are individually associated with prognosis:

- FEV1
- Smoking status
- Chronic hypoxia and/or cor pulmonale (structural and functional impairment of the right side of the heart)
- Low body mass index
- Severity and frequency of exacerbations
- Hospital admissions
- Symptom burden (for example, COPD Assessment Test (CAT) score)
- Exercise capacity (for example, six minute walk test)
- Gas transfer factor (TLCO) measures how lungs take up oxygen
- Whether the person meets the criteria for long-term oxygen therapy and/or home non-invasive ventilation
- Multi-morbidity

Source: NICE (2018)

necessitating referral to specialist services; for example, assessment for supplementary oxygen therapy or smoking cessation.

Treatment for COPD involves multiple pharmacological and non-pharmacological components. The cost to the NHS is considerable, amounting to over £800m a year in England (NHS, 2012). Treatment will be discussed in more detail in part 2 of this series.

End of life

The progression of COPD is heterogeneous and difficult to predict with any degree of certainty, making prognostic assessment uncertain. FEV1 is used as a measure of the severity of COPD, and low FEV1 against predicted FEV1 is associated with mortality, but not in a proportional manner so it can be used with confidence. There are numerous other factors associated with

mortality, but again these are not consistent. Low body mass index, frequency of exacerbations and hospitalisation, respiratory failure, and poor functional performance, for example, are all associated with mortality and should be considered when judging or estimating prognosis (British Medical Journal, 2018). The 'surprise question' ("would I be surprised if this patient died in the next 12 months?") is widely used to estimate prognosis, but at best performs moderately in all disease assessment and is even less reliable in non-cancer disease (Downar et al, 2017). This leaves clinicians in a dilemma about when to broach the difficult conversation around palliation, although earlier is better, allowing plans to be put in place to meet patients' wishes and needs in a timely manner.

Although COPD is a life-limiting illness, many patients will have other

comorbidities that ultimately lead to their death. Heart disease and malignancies, as well as progressive respiratory failure, are leading causes of death in patients hospitalised with COPD (GOLD, 2019). There is evidence that palliative care services can reduce physical and psychological symptoms associated with COPD, and improve patients' quality of life, but these services are lacking in both primary and secondary care (Bloom et al, 2017). NT

 The second article in this series will look at management and nursing care for COPD.

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Chronic obstructive pulmonary disease series

Part 1: pathophysiology, diagnosis and prognosis

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Part 2: Management and nursing care

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