CHRONIC OBSTRUCTIVE PULMONARY DISEASE **MEDICAL APPENDIX**

DEFINITION

1. Chronic obstructive pulmonary disease (COPD) is a chronic, slowly progressive disorder of the lungs characterised by an increased resistance to air flow.

2. COPD is now the generally preferred term for the disease and has been accepted by most authorities including the British Thoracic Society. It supersedes a number of previously-used clinical labels and acronyms, such as chronic obstructive airways disease (COAD), chronic airflow limitation (CAL), non-reversible obstructive airways disease (NROAD), etc. The term was coined to encompass both major phenomena of the condition, i.e. the narrowing of airways and the destruction of lung tissue.

3. The components of COPD are chronic bronchitis and emphysema, either of which may predominate. In addition there is often a variable degree of hyper-reactivity of the airways, which may be temporarily reversible with bronchodilator treatment.

4. **Chronic Bronchitis** In this component of COPD there is hypersecretion of mucus, and it is defined clinically as the presence of cough productive of sputum on most days for at least three months of two successive years, in a person in whom other causes of chronic cough have been excluded.

5. **Emphysema** This term refers to a condition of abnormal dilatation of the terminal air spaces of the lung with destruction of their walls. Emphysema is difficult to diagnose radiologically and is more usually identified retrospectively, at post-mortem. Two varieties are recognised; centrilobular and panacinar emphysema.

6. **Airways hyper-reactivity** This element is usually also present to a variable extent in COPD, causing a constriction of the airways which is partly reversible by medication. This component shares some of the features of chronic incompletely reversible asthma and indeed the distinction has not yet been clarified precisely. Most patients with COPD show some temporary improvement after using bronchodilators and terms such as chronic asthmatic bronchitis and adult wheezy bronchitis reflect this apparent overlap.

7. The distinction between this aspect of COPD and chronic incompletely reversible asthma is important mainly in relation to the prognosis for progression and mortality, which is considerably worse in COPD. In addition it is virtually certain that the pathogenesis of asthma and COPD is entirely different.

8. Some patients with chronic bronchitis and/or emphysema show no signs of airways obstruction. They are excluded from the diagnosis of COPD, as are those with completely reversible airways obstruction (i.e. asthma). Similarly, the term COPD does not include obstruction of the airways due to specific pathology such as bronchiectasis, post-tuberculous fibrosis, pulmonary oedema and obliterative bronchiolitis.

9. COPD is a major cause of chronic morbidity and mortality and is currently the fourth leading cause of death in the world. Further increases in its prevalence and mortality can be predicted in the coming decades.

CLINICAL MANIFESTATIONS

10. The onset of COPD is insidious. Typically it gradually manifests itself in the fifth, sixth or seventh decades of life, and may present with chronic productive cough and recurrent, increasingly frequent bronchial infections in which the sputum becomes purulent and increased wheeze is experienced. On the other hand the main complaint may be gradually increasing breathlessness on effort. Usually the patient reports a combination of these two types of symptom.

11. There is usually a gradual progression of disability over a period of years with increasingly frequent absence from work, reduced exercise tolerance and increased restriction of activities. The sedentary patient may fail to notice breathlessness until a significant proportion of lung function has been permanently lost.

12. Early in the progress of the disease there may be few clinical signs and cases of mild to moderate severity are difficult to diagnose on clinical grounds alone. Even in moderately advanced cases routine chest x-rays often fail to show significant changes.

13. The most practical and accurate screening of at-risk patients is by means of spirometry, whereby a reduction in the forced expiratory volume in 1 second (FEV_1) may be identified. Spirometry yields other measurements of lung function which further refine diagnostic and therapeutic issues, e.g. Forced Vital Capacity (FVC).

14. As a general rule, patients' activity is significantly restricted when the FEV_1 falls to about 50% of predicted normal, while those in whom the FEV_1 is less than 30% of predicted normal are usually severely incapacitated.

15. Early diagnosis of COPD is important in order to give advice on stopping smoking and so avoid further irreversible damage to the lungs.

16. Complications include acute respiratory infections, pneumothorax, particularly in older patients, and pulmonary embolism. Left ventricular failure may occur, possibly due to myocardial hypoxia, and is particularly dangerous as maintenance of cardiac output is essential to help compensate for low arterial oxygen.

17. The major change encountered in the blood is a raised haematocrit (secondary polycythaemia). This increases blood viscosity and contributes to the flow resistance in the vascular bed of the lung. Some patients with severe COPD develop marked pulmonary hypertension, right ventricular dysfunction, and tricuspid incompetence (cor pulmonale).

AETIOLOGY

18. By far the most important cause of COPD is cigarette smoking. There is some relation to the total dose of tobacco inhaled so the age of starting smoking is significant, and the number of cigarettes smoked over the years. Conventionally this is recorded as "pack-years" (one pack-year = 20 cigarettes a day for one year). The tar content of the cigarettes however does not appear to influence the condition. Pipe and cigar smoking may also cause the disorder, but to a much lesser extent.

19. There seems to be a varying susceptibility to smoking-induced lung damage in the population and only 15-20% of smokers will develop COPD. The reason for this is unclear.

20. Normally, the lung is protected from the damaging effect of inflammation by the elastase inhibitor, alpha-1 antitrypsin (AAT). This protein protects the alveolar walls from destruction by leukocyte elastase, a substance released by neutrophils activated by inflammatory processes. If AAT is deficient or absent the end result may be emphysema. Cigarette smoke may overwhelm the body's ability to produce AAT and in addition may directly inactivate it.

21. A rare inherited autosomal recessive disorder causes a deficiency of AAT and occurs in 1 in 3000 live births. Familial AAT deficiency accounts for less than 5% of all cases of COPD.

22. Adverse climatic factors do not have any bearing on the aetiology of COPD.

23. In the healthy individual a progressive decline in lung function is inevitable with age, but in patients with COPD who continue to smoke this decline is accelerated. Stopping smoking slows the abnormal rate of decline significantly and may extend life-span.

24. **Risk factors** A number of risk factors influence the development and course of COPD.

    24.1. **Gender** Some research suggests that men are more at risk than women, after standardizing for smoking. However this relationship is by no means certain and the incidence of COPD in women is increasing as more succumb to the effects of long-term smoking.

    24.2. **Race** Afro-Caribbean and Chinese races seem to have a reduced susceptibility to developing COPD.

    24.3. **Environmental pollution** There is evidence that particulate urban air pollution and gases contribute to COPD. The particulates mainly originate from the incomplete combustion of solid fuels and diesel, and the main gaseous components (which may also be encountered in industry) include the various oxides of sulphur, nitrogen and carbon; hydrocarbons and ozone. However the contribution of atmospheric pollution is small, compared with the effect of smoking.

    24.4. **Occupation** Occupational exposure to coal and silica dust may increase an individual's susceptibility to COPD. Other occupations at risk include workers in contact with cadmium, construction workers who handle cement, metal workers and grain handlers, but again the effect of smoking far outweighs any influences from the work environment.

    24.5. **Birth weight and childhood respiratory infections** Low birth weight may predispose to COPD in the adult, as may repeated childhood respiratory infections. The reason is unknown, although both factors may help determine the maximum lung function achieved in adolescence before the natural age-related decline begins.

24.6. **Adult respiratory infections** There is no evidence that attacks of respiratory infection such as acute bronchitis in adulthood lead to COPD. Once COPD is established however, repeated episodes of respiratory infection, either viral or bacterial, may accelerate the decline in lung function.

24.7. **Socioeconomic deprivation** There is evidence that COPD is more prevalent in areas of socioeconomic deprivation. This may simply be due to the higher proportion of the population who smoke, or to the increased likelihood of occupational exposure.

24.8. **Indoor pollution** In some countries, for example New Guinea, long-term exposure to fumes from domestic cooking devices in a poorly-ventilated environment may cause the condition, and in these populations COPD is more common in women than in men.

24.9. **Passive smoking** While there is some evidence that respiratory symptoms and respiratory infections may be more common in children in households where one or both parents smoke there is at present no clear evidence that exposure to passive smoking in adulthood can cause COPD.

CONCLUSION

25. Chronic obstructive pulmonary disease is a progressive condition characterised by chronic airway obstruction due to chronic bronchitis, emphysema and a variable degree of airways hyper-reactivity.

26. The condition has an insidious onset and symptoms frequently go unrecognised by the patient until significant irreversible respiratory damage has occurred.

27. It is caused almost exclusively by smoking, although certain occupations such as coal mining may predispose to it, and urban atmospheric pollution contributes to it. Rarely, it may be caused by an inherited abnormality. It is not caused by exposure to adverse climatic conditions or by respiratory infections in adulthood.

28. Once established the pathological changes are irreversible and prevention of disability by early diagnosis and cessation of smoking is of prime importance.

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