3. *Respiratory disorders*

3.1 INTRODUCTION

It would be convenient to divide lung diseases into those of airways and those of alveoli, thus identifying them according to the functions of ventilation and gas exchange, but the body refuses to be neatly classified, and conditions such as COPD straddle the fence. Lung disorders are usually divided into obstructive and restrictive disease, plus those that fit neither or both categories.

Airways obstruction is due to:

- reversible factors, e.g. inflammation, bronchospasm or mucus plugging,
- irreversible factors, e.g. fibrotic airway walls or damaged alveoli leading to loss of elastic recoil that supports the airways (Fig. 3.1),
- localized lesions, e.g. tumour or foreign body.

Restrictive disorders are characterized by reduced lung volume and reduced compliance due to impaired lung, pleura, chest wall or neuromuscular mechanisms.

3.2 OBSTRUCTIVE DISORDERS

The common disease entity of chronic bronchitis and emphysema is known as chronic obstructive pulmonary disease (COPD) or chronic obstructive airways disease. Asthma can overlap with COPD (Fig. 3.2). This convenient lumping together occurs because it is not easy to determine the relative proportions of each disorder.

3.2.1 Chronic bronchitis

Chronic bronchitis is a disease of the airways. It is characterized by excess mucus secretion and productive cough. The cough is called a

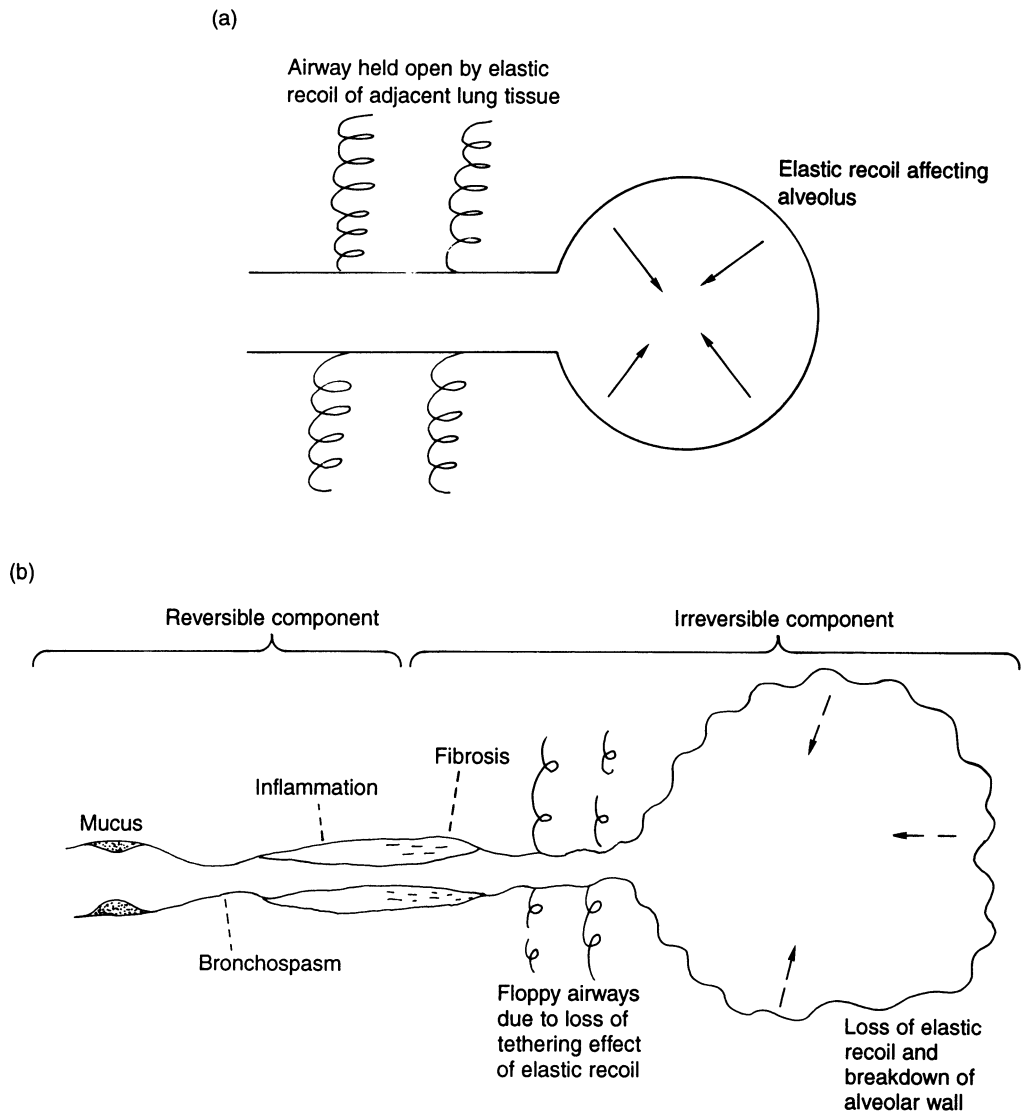

Figure 3.1 Mechanism of airways obstruction: (a) normal, (b) COPD.

smokers' cough in the early stages, but once it has occurred for three months per year for two years, it becomes the inadequate, but traditional, definition of chronic bronchitis.

Causes

Smoking is the major cause of chronic bronchitis, although other pollutants contribute. Risk factors are male sex and poverty (both associated with smoking), occupation, housing, climate and childhood respiratory illness (Clarke 1991).

Pathophysiology

Repeated inhalation of pollutants causes irritation of the sensitive lining of the airways,

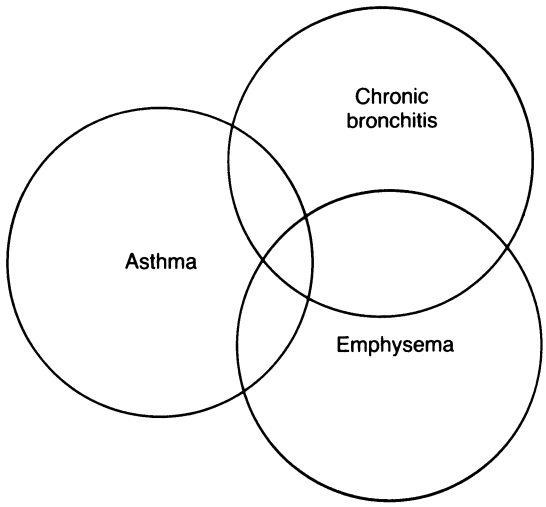

Figure 3.2 Relationship between the common forms of obstructive lung disease.

inflammation, mucus hypersecretion and sometimes bronchospasm.

Inflammation is the key process in chronic bronchitis. It causes narrowing first in the small and then the large airways. Acute inflammation resolves, but chronic inflammation leads to fibrotic changes, scarring and permanent damage.

Mucus hypersecretion is caused by rampant increase in the size and number of mucus-secreting goblet cells. Excess mucus has traditionally engaged the attention of physiotherapists, but causes little overall airways obstruction, correlates little with physiological disturbances (Faling 1986) and does not relate to mortality (Wiles and Hnizdo 1991). Breathlessness is more significant to the patient and more related to inflammatory damage and airway narrowing (Peto *et al* 1983). However, excess mucus predisposes to infection and clearance is hampered by cilia rendered inefficient by damaged epithelium, airway collapse and abnormal hydration (Smalldone 1993).

Bronchospasm is thought to be caused by acetylcholine release due to inflammatory stimulation of the parasympathetic nervous system.

Gradual patchy airway narrowing, often augmented by the floppy airways of emphysema (Gelb *et al* 1993), leads to uneven distribution of ventilation and hypoxaemia. The inexorable downhill path of advanced chronic bronchitis is shown in Fig. 3.3. Polycythaemia is red cell proliferation and represents an attempt to compensate for hypoxaemia by boosting arterial oxygen capacity. It manifests as increased red cell count, haemoglobin concentration and packed cell volume. The associated rise in blood viscosity can reduce cardiac output, hinder oxygen delivery, increase pulmonary hypertension and cause headaches. If the disadvantages of polycythaemia are greater than the advantages, venesection (bloodletting), with reinfusion of the plasma, brings temporary relief.

Capillary destruction and widespread hypoxic pulmonary vasoconstriction increases pulmonary vascular resistance (Vender 1994). Pulmonary hypertension increases the load against which the right ventricle must pump, leading to hypertrophy and dilation, a condition known as cor pulmonale. This reduces the efficiency of the heart and further impairs oxygen delivery. Right heart failure then supervenes, followed by a rise in systemic BP in order to overcome the increased right atrial pressure and maintain cardiac output. This process eventually strains the left ventricle and leads to left heart failure, but death is ultimately due to inadequate gas exchange rather than cardiac involvement (Harris 1989).

Clinical features

The natural history of chronic bronchitis spans 20–50 years, but the disease is asymptomatic at first because changes in the small airways barely affect total airways resistance. Onset of symptoms is then insidious, and a morning cough is tolerable and considered

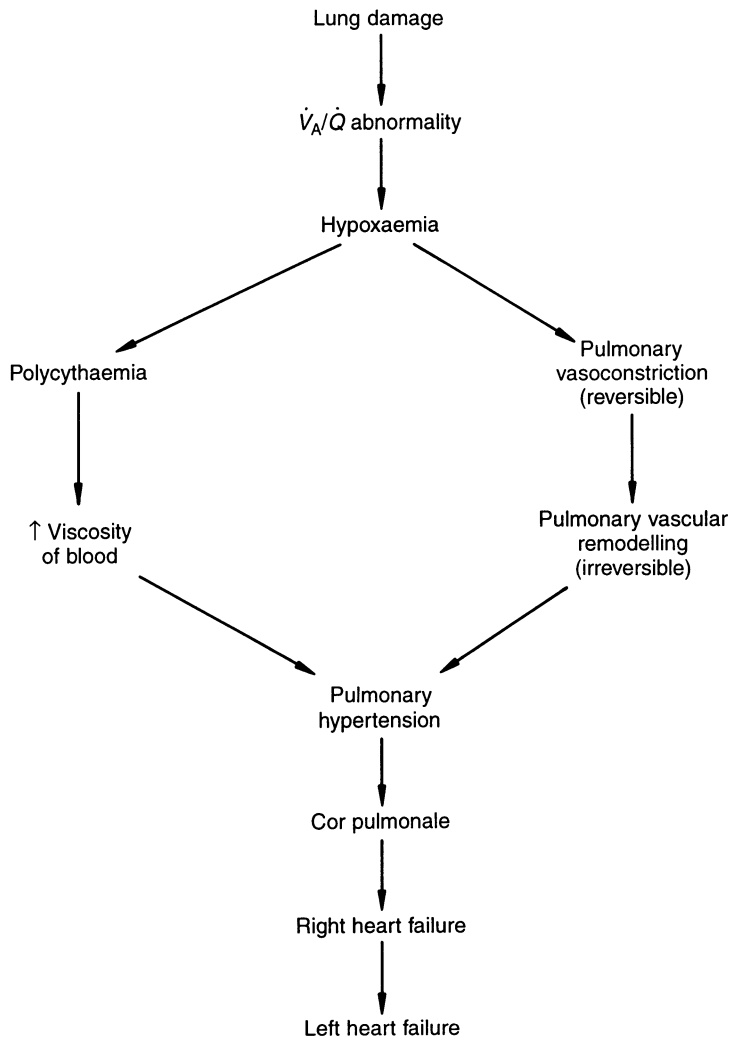

Figure 3.3 Sequence of events in COPD.

normal among smokers, who may not seek medical advice until the lungs are considerably damaged.

Patients then gradually develop breathlessness, which should be routinely measured in COPD patients (Mahler 1995), reduced exercise tolerance and fatigue. Sleep is no longer recuperative but is disturbed by breathlessness, while nocturnal oxygen desaturation plays a role in progression of the disease. Objectively, there is a rich tapestry of signs, such as laboured breathing, a plethoric or cyanotic appearance and crackles on auscultation (Piirilä *et al* 1991). FEV_1 declines by 60–120 ml a year (Hodgkin 1993, p. 65) and is a useful indicator of obstruction but relates weakly to functional impairment (Mahler 1995). X-ray signs may be insignificant or may show a feathery appearance, upper lobe diversion and later the enlarged heart of cor pulmonale.

The appearance of peripheral oedema is a turning point in the disease process, indicating a $PaO_2 < 7.3$ kPa (55 mmHg) (Stewart

and Howard 1992). Although often associated with cor pulmonale, oedema is not caused by right heart failure, but reflects impaired water handling by the kidneys due to hypoxaemia and hypercapnia (Henderson 1994) and the release of intracellular water during hypoxic dissolution of tissue matrix. Oedema is reduced by oxygen therapy (Howes *et al* 1995). Other systems affected by hypoxia include the gut, leading to the association of COPD with peptic ulcers.

3.2.2 Emphysema

Sometimes when exhausted by breathless endeavor
I wish I could sleep forever and ever.
But then this thought my longing allays:
I shall be doing it one of these days.
Carroll O'Carroll, Chicago

Emphysema usually coexists with chronic bronchitis and shares a similar aetiology, but is primarily a disease of alveoli with secondary effects on the airways. It is commonly caused by smoking. Rarely, primary emphysema is caused by congenital lack of alpha$_1$-antitrypsin.

Pathophysiology

Protein breakdown is the villain of emphysema, causing erosion of alveolar septa, dilatation of distal airspaces and destruction of elastic fibres. This leads to a hyperinflated chest by three mechanisms:

1. The walls of the terminal bronchi are normally supported by radial traction exerted by alveolar septa. Loss of elastic tissue means that during expiration compressive forces are not opposed by radial traction, and the floppy airways tend to collapse (Fig. 3.1), a phenomenon known as dynamic compression of the airways. This leads to air trapping distally and **passive hyperinflation**.
2. The patient has to maintain hyperinflation actively by sustained inspiratory muscle contraction in order to hold the airways open, a mechanism known as **dynamic hyperinflation** (McCarren 1992). This unfortunate but necessary process is achieved at the cost of excess work of breathing, barrel chest and a lung volume that can actually exceed the predicted TLC (Decramer 1989).
3. Obstructed airways mean that air from the previous breath may not be fully expelled before the next inspiration starts, creating positive pressure in the chest known as **intrinsic PEEP** (Fig. 3.4).

Figure 3.4 Development of intrinsic PEEP.

The latter was first described during mechanical ventilation but occurs spontaneously in some COPD patients, especially during exacerbations or when breathing rapidly, e.g. during exercise. The lungs are prevented from emptying to their usual relaxed volume between inflations by an average positive pressure of 2 cmH_2O (Ninane *et al* 1993), imposing an extra load at the start of inspiration, hindering cardiac output and impairing perfusion to the labouring inspiratory muscles (Kawagoe 1994). Stabilization occurs at volumes and pressures that are higher than normal, which reduces lung compliance. The distended alveoli require greater than normal pressure for inflation, thus overturning the old concept that emphysematous lungs are hypercompliant (Macklem and Eidelman 1990).

The flat diaphragm of a hyperinflated chest is inefficient and susceptible to fatigue, as with any muscle which deviates from its resting length (Yan 1992). Extra energy is required to:

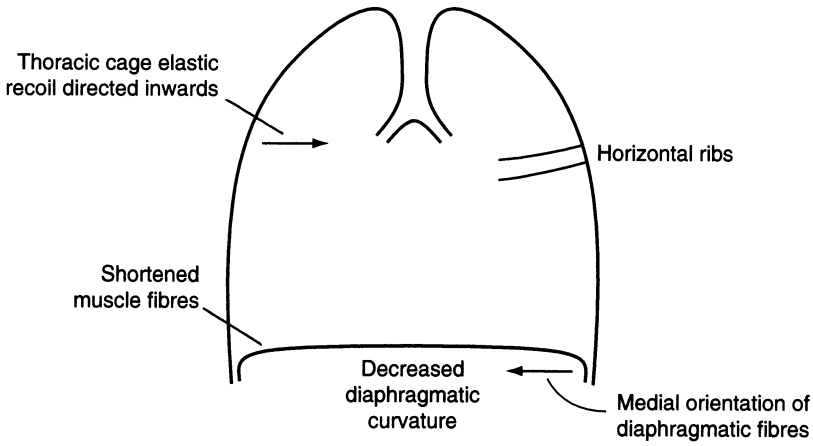

Figure 3.5 The detrimental effects of hyperinflation on the mechanics of breathing. (From Tobin, M. (1988) Respiratory muscles in disease. *Clinics in Chest Medicine*, **9**, 264, with permission.)

- compensate for this inefficient muscle contraction,
- compensate for loss of the 'bucket handle' action of the ribs (Fig. 3.5),
- compensate for reversed action of rib cage elastic recoil, which in the hyperinflated chest is directed inwards rather than outwards, thus resisting instead of assisting inspiration (Fig. 3.5),
- sustain inspiratory muscle action so that high lung volumes are maintained, alveoli being opened at a high point on the pressure–volume curve (Fig. 1.3),
- assist expiration, which has become active in an attempt to squeeze air out through floppy airways on expiration.

People with emphysema are doubly burdened because airways resistance makes it harder to breathe, while hyperinflation impairs the capacity of the inspiratory muscles to handle the added load. The flat diaphragm works paradoxically and becomes expiratory in action, the lower ribs being drawn in on inspiration (Hoover's sign). Some patients can only inhale by lifting up their entire rigid rib cage with their accessory muscles.

Normal muscle is able to respond to increased load by hypertrophy, but an emphysematous diaphragm often labours under further handicaps, such as malnutrition, so diaphragmatic weakness is common (Duranti 1995). Malnourishment is caused by excess energy demand due to the work of breathing, and impaired energy supply due to difficulty in eating when breathless. It leads to cannibalization of the respiratory muscles for their protein, further impairing ventilation. Malnutrition also accelerates the process of emphysema itself (Schlichtig and Sargent 1990).

Two types of emphysema are described, although they may coexist. Centrilobular emphysema affects the respiratory bronchioles, leaving the alveoli relatively unscathed. Panlobular/panacinar emphysema is associated with severe alpha$_1$-antitrypsin deficiency and affects the alveoli themselves, causing more extensive destruction. Breakdown of lung tissue may lead to the formation of bullae (see Glossary), which occasionally require laser ablation (Barker *et al* 1993) or surgery (Nickoladze 1992).

Clinical features

Emphysema shares many of the features of chronic bronchitis. Its hallmarks are breath-

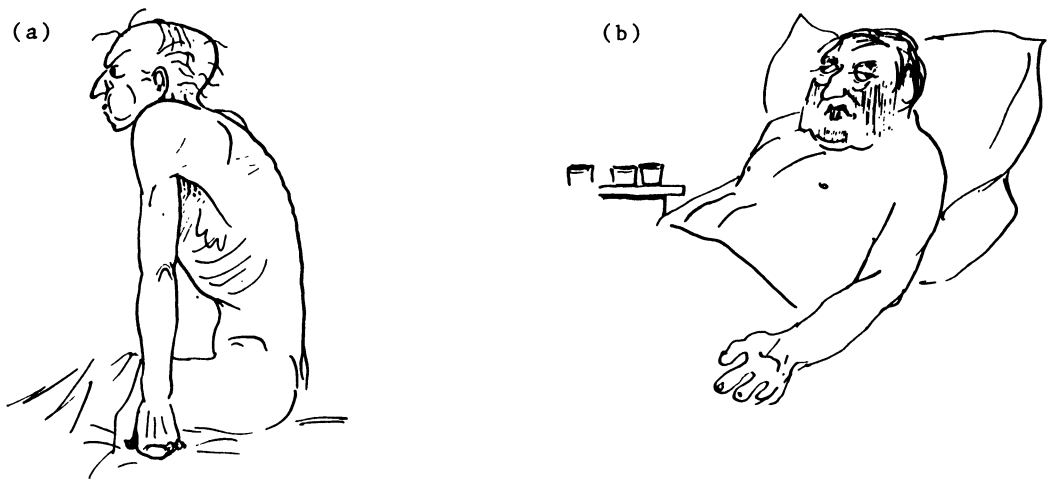

Figure 3.6 Patients with (a) pink puffer and (b) blue bloater characteristics of COPD. (From Brewis, R.A.L. (1977) *Lecture Notes in Respiratory Disease*, Blackwell, Oxford, with permission.)

lessness, laboured breathing and fatigue. Other signs are weight loss, barrel chest, forced expiration with pursed lip breathing, prolonged expiration with I:E ratio at 1:3–1:4, and quiet breath sounds. Soft-tissue recession and other signs of laboured breathing are evident, fossae are prominent and the neck appears sculpted with the skin draped over the bones (Fig. 2.2), because of inspiratory effort and malnutrition with the associated decreased skin-fold thickness (Tobin, 1988). Patients may lean forwards on their elbows in order to force the diaphragm into a more efficient dome shape and stabilize the shoulder girdle for optimum accessory muscle action.

Gas delivery to areas of capillary destruction leads to a high $\dot{V}_A/\dot{Q}$ ratio, and airways obstruction leads to a low $\dot{V}_A/\dot{Q}$ ratio, but relative preservation of gas exchange is a feature of emphysema in the early stages, $\dot{V}_A/\dot{Q}$ match being maintained partly by capillary and airways damage compensating for each other, and partly by development of collateral ventilation (Morrell 1994).

The radiograph shows signs of hyperinflation (Fig 2.5), but CT scanning quantifies emphysematous changes more accurately (Morgan 1992), showing areas of low density, blood vessel attenuation and sometimes bullae. Airways obstruction causes expiratory flow limitation as shown by a reduced FEV_1. A low gas transfer distinguishes it from chronic bronchitis and is the most sensitive test for emphysema, a reduced TLCO becoming evident before X-ray or physical changes (Hodgkin 1993, p. 66).

3.2.3 Chronic bronchitis with emphysema

People with COPD can potentially demonstrate 89 symptoms (Kinsman *et al* 1983).

The blood gas picture is seen anywhere along a spectrum with the following extremes (Fig. 3.6):

- the 'pink puffer' (PP) patient maintains near-normal blood gases at the expense of breathlessness and weight loss,
- the 'blue bloater' (BB) patient abandons the fight for normal blood gases and feels less breathless, but pays for symptomatic relief with oedema, inadequate gas exchange and a mortality rate twice as

high as the PP patient (Clague and Calverley 1990). Some BB patients may tolerate a $PaCO_2$ of over 12 kPa (90 mmHg) for years (Hodgkin 1993, p. 436).

It was originally thought that repeated hypoventilation in BB patients desensitized their chemoreceptors to hypercapnia so that they became dependent on low oxygen tension as a stimulus to breathe. But more recent studies put the blame on respiratory muscle fatigue, the patient 'choosing' the wise option to hypoventilate (Bégin, 1991). PP patients sometimes show a tendency to rush at activities, and breathless management techniques are particularly effective (Chapter 6).

Half of all COPD patients aged over 50 years have cardiovascular disorders (Hodgkin 1993, p. 66) because of related pathology and the mutual association with smoking. Cardiac function plays a significant role in oxygen delivery in people with COPD. Despite preservation of blood gases and more efficient tissue oxygen extraction in the PP patient, oxygen delivery is impaired because of reduced cardiac output (Donahoe *et al* 1992). Low cardiac output prolongs pulmonary transit time, which further allows these patients to escape hypoxaemia. BB patients have more unsaturated arterial blood, but often have normal cardiac output and may be less hypoxic at tissue level. In other words, 'pink puffers' may be only superficially pink and 'blue bloaters' only superficially blue. These terms are hardly complimentary and are not appropriate bedside terminology, but are useful as graphic descriptions of individual responses to COPD.

Exacerbation of COPD occurs when the severity of signs and symptoms increases, especially breathlessness, airways obstruction and sputum production (Ball 1995). Causes are viral infection, sometimes bacterial infection, or non-microbial inflammation due to pollution or allergy, which may be moderated by steroids. Secretions may compromise airway function (Wanner 1990), and physiotherapy is often required to help the patient clear secretions and reduce the work of breathing.

Medical management of COPD is discussed in Chapter 4, and physiotherapy management in Chapters 5 and 6.

3.2.4 Asthma

Asthma is more common, more serious and more treatable than is generally thought. Mortality increased by one-third during the 1980s (Lawrence 1995), and the disease kills on average five people a day in England and Wales (Finfer and Garrard 1993), many of them young. Asthma is now the only treatable life-threatening condition in the Western world with a rising death rate, with up to 86% of asthma deaths being preventable (GRASSIC 1994). People die because they, their relatives or doctors do not see asthma as a potentially fatal disease, or grasp the importance of prevention, or recognize deterioration.

Asthma is a chronic inflammatory condition of the airways, characterized by undue responsiveness to stimuli that are normally innocuous, a mechanism known as hyperreactivity. It is distinguished by the variability and reversibility of its presentation, which makes evaluation of severity difficult. It shares with COPD the common pathology of small airways obstruction, but the differences are shown in Table 3.1. Patients suffer recurrent airway narrowing, which usually reverses spontaneously or with treatment.

Physical examination is a hit-and-miss affair because of the episodic nature of the condition and the fact that the symptoms of wheeze, breathlessness and cough are common respiratory complaints. Diagnosis is made from a history of recurrent acute attacks and confirmed by respiratory function tests. If the peak flow varies by 15%, either diurnally, after exercise, or after broncho-

Table 3.1 Distinguishing features of asthma and COPD

	Asthma	*Chronic bronchitis*	*Emphysema*
History	?Family history ?Allergy ?From childhood	Smoking Gradual history	
Provocation of symptoms	Weak stimulus, e.g. cold air	Strong stimulus, e.g. infection	
Variation of symptoms	Much	Little	
Cough at night	Wakes coughing	Wakes then coughs	None
Response to bronchodilators	Yes	Sometimes	Rarely
Response to steroids	Yes	Occasionally	Rarely

dilator treatment, this is considered diagnostic of asthma.

Causes and pathophysiology

A combination of genetic and environmental factors leads to the development of asthma. Predisposing factors include anxious parents, history of a stressful birth and a single gene which causes atopy. Atopy is predisposition to allergic disease and is the main risk factor for developing asthma. An atopic person may become asthmatic if exposed to the house-dust mite, other allergens or occupational pollutants. Asthma is more common in urban areas, implicating pollution (Cogswell 1994). Passive smoking or viral infection damage epithelium so it is more sensitive to allergens. Two phases of response occur (Fig. 3.7):

1. Sensitization stage (which only occurs in atopic people): exposure to allergens, especially in fetal or early life, stimulates production of excess immunoglobulin E (IgE) antibodies in the serum. IgE become fixed to mast cells, which then react to antigens and release bronchoconstrictor substances such as histamine.
2. Hyperreactive stage: continued exposure leads to chronic low-grade inflammation of the airways, causing bronchial hyperreactivity. Once asthma has developed, removal from exposure to the allergen, if delayed, does not always prevent continuing asthma.

The mechanism for the persistence of asthma is thought to be localized because asthmatic recipients of transplanted lungs lose their asthma, while non-asthmatic patients who receive asthmatic lungs develop the disease (Corris and Dark 1993).

Once hyperreactivity is established, other factors which may or may not be related to the original cause can trigger an asthma attack, for example:

- exercise,
- allergenic foods, e.g. dairy products, eggs, wheat, nuts, additives, cola or other acidic drink,
- drugs, such as NSAIDs, beta-blockers, aspirin (Empey 1992),
- stress, through multiple CNS interactions (Busse 1995),
- chest infection, especially in infants,
- warm-blooded pets,
- pollen,
- car exhaust,
- premenstruation (Cross 1994),
- night time,
- hyperventilation (Groen 1979),
- frustrated expression of emotion (Groen 1979),
- weather, especially change in temperature (Rossi *et al* 1993),
- smoking.

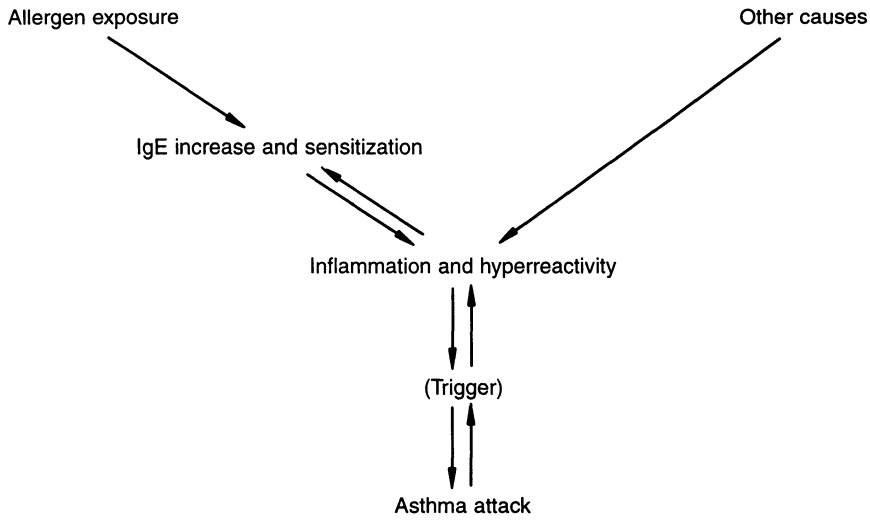

Figure 3.7 Development of asthma.

Some factors may be cause, effect or both. Anxiety, depression and social isolation are associated with asthma (Ramsay 1994), and gastro-oesophageal reflux (GOR) is common (Jack 1995) due to microaspiration of acid into the upper airway triggering bronchospasm, and/or reflux being caused by relaxation of the smooth muscle of the cardiac sphincter due to bronchodilators (Miles and Ayres 1993).

Allergic asthma, known as **extrinsic**, is common during early life in atopic people. **Intrinsic** asthma occurs with normal IgE levels, develops in adulthood, is more fulminant and less responsive to treatment.

Inflammation and hyperreactivity lead to airways obstruction by:

- mucosal oedema,
- bronchospasm,
- gelatinous mucus plugging.

The more severe the asthma attack, the greater the small airways obstruction (as shown by $\dot{V}_A/\dot{Q}$ mismatch), compared with large airways obstruction (as shown by spirometry). Small airways obstruction lasts longer than large airways obstruction, and it is suggested that more prolonged anti-inflammatory medication should be given than would be indicated by spirometry alone (Ferrer *et al* 1993). Persistent inflammation leads to fibrosis of the airway walls and irreversibility.

Classification and clinical features

Mild chronic asthma manifests as an intermittent dry cough, often at night, or a morning wheeze once or twice a week. Peak flow varies by less than 25%. Even when asymptomatic, peripheral airways resistance can be fives times normal (Wagner 1992), and severe attacks are possible.

Severe chronic asthma means frequent exacerbations and symptoms that significantly affect quality of life. Peak flow varies by more than 25%. Daily anti-inflammatory drugs are required. The most severe form is unstable or brittle asthma, which shows greatly fluctuating peak flows, persistent symptoms despite multiple drug treatments, and unpredictable severe falls in lung function, often without a known precipitating factor (Garden and Ayres 1993).

Acute asthma reflects failure of preventive management or exposure to a noxious stimulus. The large airways are obstructed by bronchospasm and the small airways by oedema and mucus plugging. Asthma

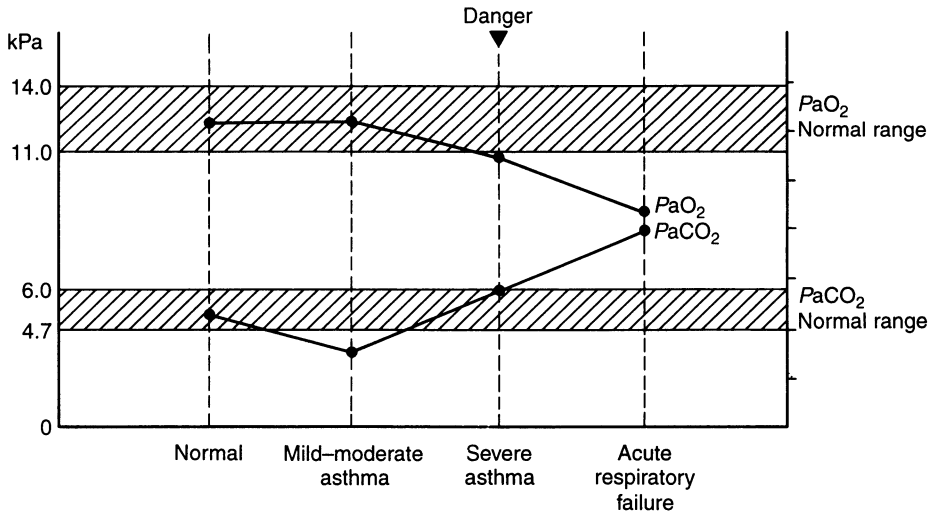

Figure 3.8 Progressive changes in blood gases during acute severe asthma. (From Smith, M. (1982) In case of emergency. *Nursing Mirror*, March 17, with permission.)

attacks are associated with breathlessness, rapid breathing and abdominal paradox. The work of breathing is increased by airflow resistance up to 15 times normal (Mador 1991) and hyperinflation to keep the narrow airways open (Wheatley 1990). Hyperinflation renders the diaphragm ineffectual and the inspiratory muscles are forced to work throughout inspiration and expiration. $\dot{V}_A/\dot{Q}$ mismatch reduces PaO_2, and rapid breathing reduces $PaCO_2$. If oximetry shows SaO_2 below 92%, respiratory failure is likely and blood gas monitoring is required (Carruthers and Harrison 1995).

The effort to breathe out against obstructed airways is reflected in prolonged expiration, as shown by decreased peak flow readings. Patients feel as if they are struggling to breathe through a narrow straw, and many are extremely frightened.

> *All one's strength, that one feels becoming weaker and weaker, is concentrated into one last effort to take one slight breath that will allow the respiration to continue.*
>
> Ruiz 1993

Severe acute asthma may present in one of two ways. Most commonly, the condition develops slowly, often after several weeks of wheezing. Alternately, the attack is sudden, especially if there has been poor drug control, and this so-called asphyxic or catastrophic attack can be fatal within minutes.

Deterioration can be deceptive, and paradoxically the patient may appear less distressed as the condition worsens. Medical help should be sought if the patient shows:

- respiratory rate > 25/min, then decreasing as the patient tires,
- ↑ $PaCO_2$ as the patient tires (Fig. 3.8),
- heart rate > 110, then bradycardia if the patient deteriorates,
- peak flow < 40% of the predicted value, or < 200 l/min if the predicted value is not known,
- ↓ response to bronchodilator, i.e. < 10% improvement in peak flow or FEV_1,
- difficulty in speaking,
- loss of wheeze, and silent chest on auscultation if the airflow is too slow to oscillate the airways, indicating worsening obstruction or failing ventilatory muscle

function, with decreased respiratory effort and exhaustion,
- pallor and sweating,
- pulsus paradoxus (the difference between the highest and lowest systolic blood pressure) < 10 mmHg,
- cyanosis, which represents a life-threatening attack (Carruthers and Harrison 1995),
- exhaustion, confusion, drowsiness, coma.

Very breathless patients cannot produce reliable peak flow readings, and for those too breathless to speak, the peak flow meter can exacerbate bronchospasm (Fanta 1992). Ventilatory failure during severe acute asthma is usually attributed to inspiratory muscle fatigue, but sudden deaths without obvious exacerbation of airflow obstruction have been associated with impaired respiratory drive related to depressed mood (Allen *et al* 1994).

Status asthmaticus is an imprecise term that is sometimes used interchangeably with severe acute asthma, but specifically describes an asthma attack prolonged over 24 hours, leading to dehydration and exhaustion. When the term is used accurately it does not mean the most dangerous form of acute asthma, which can lead to rapid death (Kallenbach *et al* 1993).

Exercise-induced asthma is present in 80% of asthma sufferers, and in some is the only manifestation. Hyperventilation during exercise, especially in cold weather, leads to evaporation, hyperosmolality and cooling of airway surface liquid, and then bronchospasm (Widdicombe 1992). Bronchospasm normally occurs 5–10 minutes after exertion, recovery is usually complete 30 minutes later, and a refractory period of about an hour follows in 50% of patients (Sterk 1993). Bronchospasm can also occur during exertion (Beck *et al* 1994). Warm-up and cool-down periods help to prevent the rapid airway rewarming which can cause oedema. Other preventive measures include β_2-stimulants or sodium cromoglycate taken 20 minutes before exercise, and on cold days a scarf worn over the mouth to warm the inspired air.

Nocturnal asthma occurs in 80% of asthmatics (Douglas 1993), interferes with sexual intercourse and causes a marked early morning dip in peak flow (Fig. 3.9). Many people develop tolerance to nocturnal symptoms, and lack of awareness of the treacherous diurnal variation leads to underdiagnosis and avoidable deaths. Suggested trigger factors are either an exaggerated bronchial response to cold bedrooms, gastro-oesophageal reflux, reduced lung volume in the supine posture, allergens in bedding, the timing of drug administration or hormonal circadian rhythms.

Occupational asthma may take weeks or even years to develop. Symptoms usually worsen during the week and ease at weekends, but some patients may need several days of not working before improvement is noticed, which confuses the diagnosis.

Education and prevention

Self-management can prevent the majority of asthma deaths and allow most people with stable chronic disease to be largely free of symptoms rather than accept them as inevitable. Education based on booklets and peak flows improves knowledge but does not necessarily change behaviour, and personal instruction has been found the most effective approach (Make 1994). Substantial changes in behaviour have been shown, for example, by a brief three-hour education programme in the community for those recently discharged (Yoon *et al* 1993). Patients are more motivated when the emphasis is on management of symptoms rather than general knowledge (Osman *et al* 1994).

The advantage of education during hospitalization is that motivation is greatest during an acute episode. The disadvantages are that there is often little time, and information may not be easily absorbed during this anxious period. It is best to motivate patients

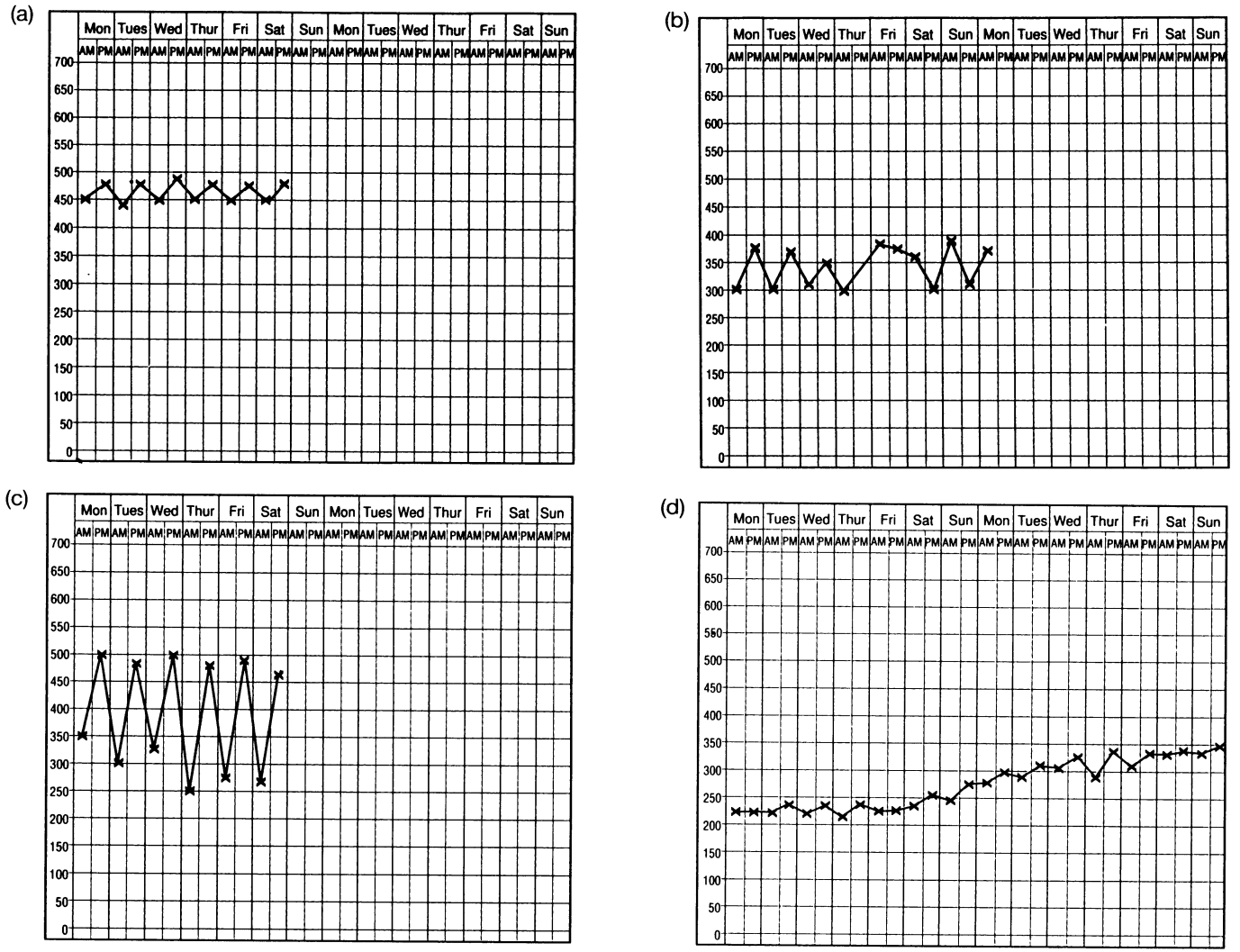

Figure 3.9 Serial peak flow readings: (a) normal diurnal variation, (b) increased diurnal variation indicative of chronic asthma, (c) early morning dip, representing nocturnal asthma and/or impending asthma attack, (d) steroid trial demonstrating significant reversibility. (From Hubbard, J. (1992) Use of the peak flow meter in asthma, *Resp. Dis. Pract.*, **9**(4). Reproduced by kind permission of Hayward Medical Communications Ltd, 44 Earlham Street, London WC2H 9LA.)

in the acute phase, provide sufficient information for them to understand that prevention is the key, help them identify their own needs and offer some follow-up education. Collaborative self-management has shown the following outcomes (Make 1994):

- ↓ accident and emergency department visits and hospitalization,
- ↓ medication use,
- ↓ symptoms,
- ↑ compliance with treatment,
- ↑ quality of life.

Education should include the following:

1. Identification of precipitating factors using a diary (Fig. 3.10) to identify the individual pattern of asthma, then prevention when possible, e.g. intensive vacuum cleaning and special bedding to protect against house dust mite (Owen *et al* 1990), avoiding insecticides, keeping pets out of bedrooms, avoiding the not-uncommon scenario of an inhaler in one hand and a cigarette in the other.
2. For people with stable asthma, twice-daily peak flow readings are encouraged, using peak flow diaries available from drug manufacturers. Plans to short-circuit the referral system and self-admit to hospital should be prearranged if the peak flow is less than 40% of the patient's normal after medication. Comprehensive prehospital care can reduce asthma deaths to zero (Cochrane 1995).
3. For women with a family history of asthma, minimal intake of allergenic foods during pregnancy and while breast feeding, and similar care of the baby's diet in the first year of life.
4. Food exclusion diets in appropriate patients, which can reduce medication by over a half (Lewith 1995).
5. Understanding the action and administration of drugs, and the importance of taking preventive drugs, such as steroids, even when feeling well. At present fewer than half of asthmatics take their drugs as prescribed (Couriel 1994b).
6. Mastering relaxation (Freedberg *et al* 1987).
7. Stress reduction by techniques such as biofeedback (Peper 1992), meditation (Lehrer *et al* 1986), hypnotherapy (Morrison 1988a) or yoga (Singh *et al* 1990). Acupuncture can reduce the need for medication, and homeopathy is useful if the trigger is identified (Lewith 1995). Complementary therapies are reviewed by Lane and Lane (1991).
8. For those with nocturnal asthma, trying different sleeping positions, room temperatures, drug timings and a stress-free period before bed.
9. Avoidance of room humidifiers, which nurture the house dust mite and collect moulds.
10. Prevention of gastro-oesophageal reflux (p. 87).
11. Learning the warning signs of an exacerbation, e.g. reduced exercise tolerance, waking at night, prolonged wheeze after waking, declining or variable peak flows, reduced effectiveness of bronchodilator.
12. Individual written guidelines on how to respond to warning signs, and an understanding of when to increase drug dosage, take oral steroids, or seek medical assistance, e.g:
    (a) if the peak flow falls < 70% of the patient's normal, take regular bronchodilators and double inhaled steroids,
    (b) if the peak flow falls < 50%, start oral steroids and contact the doctor,
    (c) if the peak flow falls < 40%, seek urgent advice (Couriel 1994b).

ASTHMA DIARY
Times when I felt extra breathless or wheezy

Date	Time of day or night	What made me breathless or wheezy?	What did I do to help myself?	How much did it help?

Figure 3.10 Example of a diary for the self-management of asthma.

13. Advice for high-risk patients is to keep with them at all times their devices for self-administration of drugs, information bracelet (BTS 1990) and a note from their GP for ambulance personnel to administer unrestricted oxygen. Spare inhalers should be kept in the car and at work.
14. Joining an organization, such as the National Asthma Campaign, which provides diary cards and educational material (Appendix C).

Breathing techniques

Certain breathing manoeuvres can be used to aid relaxation and give patients some control. Patients can practice them regularly and put them into practice when an attack is anticipated. The emphasis is on gentle changes in breathing, not deep breathing which can exacerbate bronchospasm (Lim *et al* 1989). All patients should be encouraged to find their own forms of relaxation and stress management, e.g. the profound relaxation achieved with meditation has shown positive outcomes (Fried 1993, p. 234).

Girodo *et al* (1992) show how simple relaxed abdominal breathing can reduce the intensity of symptoms and increase physical activity. Innocenti (1974) describes how patients can gain control by learning to change back and forth between abdominal and upper chest breathing, and to alter, breath by breath, the rate and depth of breathing. Weissleder (1976) claims that asthmatic attacks can be aborted by teaching the 'complete breath technique', which consists of the following instructions, with the patient in supported sitting:

- listen to and feel the quality of your breathing, including any wheeze,
- inhale through your nose, slowly enough to eliminate the wheeze, while increasing the depth of your breathing temporarily to compensate for the slow breath,
- adjust the rate as breathing becomes comfortable and there is less hunger for air, a hunger which will also be modified by feeling in control,
- observe and modify any muscle tension,
- gradually increase the depth of breathing in three different segments, progressing upwards from abdominal, lateral costal and upper chest expansion, then reduce the depth in reverse order,
- recheck muscle tension,
- progress to unsupported sitting and standing positions.

The physiotherapist can start by breathing alongside the patient, but the aim is for patients to recognize their ability to manage their own condition.

Exercise

Asthma and exercise have a difficult relationship. Exercise can directly trigger an acute episode, but numerous asthmatic athletes have won Olympic medals, and aerobic training, with bronchodilator prophylaxis, has shown the following outcomes:

- ↓ bronchospasm, ↑ peak flow, ↓ wheeze (Dean *et al* 1988),
- ↓ breathlessness (Cochrane and Clark 1990),
- ↑ work capacity (Gong 1992),
- ↑ confidence (Robinson *et al* 1992),
- ↓ exercise-induced asthma by greater fitness, which reduces minute ventilation at high intensity work loads (Cochrane and Clark 1990).

Other physical measures

It has been claimed that the inspiratory muscles can be damaged during an acute attack and that this risk can be reduced by strengthening these muscles. Six months of inspiratory muscle training, using a pressure-threshold device for half-an-hour five days a

week, showed evidence of reduced symptoms, medication use and hospitalization (Weiner *et al* 1992).

Some patients who are anxious or have poorly controlled asthma show muscle tension and poor posture. The physiotherapist can help the patient deal with anxiety, ask for a drug review and work on stretching and postural exercises. Patients who have developed a habitually hyperinflated chest as a carry-over from acute episodes benefit from advice to emphasize expiration (not to be encouraged during acute episodes, when hyperinflation is necessary to hold open the obstructed airways).

Coughing can relieve or exacerbate asthma (Young *et al* 1991) and the physiotherapist can advise on effective coughing or cough suppression (p. 139) as appropriate.

Outcome measures have traditionally comprised respiratory function tests, complication rates, hospitalization and death rates, but functional measures most usefully relate to physiotherapy outcomes. Quality of life questionnaires have been described by Rowe (1993) and Juniper (1993).

Drug management

Charts describing the step-by-step recognition and drug management of chronic and acute asthma are available (BTS 1993). Underuse, overuse and inappropriate use of drugs is common. Historically, treatment relied on the sticking plaster of bronchodilators for symptom relief, but it is now understood that medication should hinge on regular preventive medication in the form of mast cell stabilizers (p. 99) or steroids, with intermittent bronchodilators if symptoms recur.

Patients find bronchodilators attractive, but they do not prevent long-term inflammatory damage to the airways. The paradox is that they can be beneficial immediately but detrimental in the long run. Even in the acute state, over-reliance may delay seeking medical assistance. In the chronic state, regular use, especially overuse, can smother symptoms so that a wheeze no longer acts as a warning to avoid the offending stimulus. Prolonged inflammation develops which can double hyperreactivity and increase morbidity and mortality (Cockcroft *et al* 1993). Surveys on asthma deaths invariably implicate underuse of steroids in the fatal attack (Neville *et al* 1991).

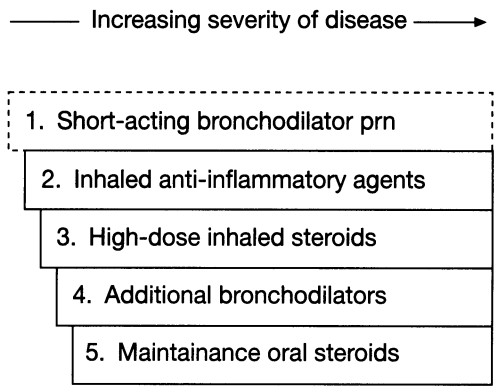

Figure 3.11 Drug management of chronic asthma. If 'as required' inhaled bronchodilators (1) are needed more than once a day, inhaled anti-inflammatory drugs (2, 3) are given. If symptoms persist, a second bronchodilator (4) is given. A small percentage of people with severe chronic asthma need oral steroids (5).

The frequency of the need for bronchodilators provides a useful marker for adjusting prophylactic treatment. A stepwise protocol is advocated for drug management (Fig. 3.11). Accurate monitoring is essential. After diagnosis, peak flow should be measured within 30 minutes of waking and in the evening, then drugs adjusted until either the normal predicted value or 'best peak flow' is achieved. If the initial value is less than 80% predicted, a two-week course of steroids, and sometimes bronchodilators, may be needed to find the 'best peak flow'.

People with acute asthma may need high concentrations of oxygen, high-dose nebulized bronchodilators in small frequent doses

(Bennett 1991) and oral or intravenous steroids (Neville *et al* 1991). Antibiotics are rarely indicated.

The characteristics of asthma make it susceptible to non-compliance with treatment. It is a chronic condition with long periods of remission, drug regimes may show no immediate benefit and inhalers are conspicuous and sometimes difficult to operate. Education that considers the patient's individual needs, health beliefs and lifestyle is likely to succeed, especially with adolescents.

Physical management of an asthma attack

Fear of an acute episode is reduced if the patient is given the following advice:

1. Sit upright, or lean slightly forwards resting the arms on a table, or sit astride a chair backwards with the arms resting on the chair's back.
2. Sit near fresh but not cold air.
3. Drink sips of warm liquid.
4. Breathe through the nose unless breathlessness makes this impossible.
5. If there is dizziness with tingling hands and feet, try to slow down the breathing.
6. Practise previously-learned techniques of relaxation, abdominal breathing and control over breathing. These should be begun at the first intimation of an acute episode.
7. Raise the resting respiratory lung volume by starting inspiration slightly early, which may help to open the narrowed airways (Innocenti 1974). The extra elastic work imposed by hyperinflation is offset by less airflow resistance so that total work is reduced (Wheatley 1990). However, many patients will have already achieved optimal hyperinflation spontaneously, and close observation of the patient's breathing pattern is required if this technique is chosen.

Relatives can be shown how to apply acupressure to the bronchospasm acupuncture points: CV.17 between the nipples, Co.4 on the dorsal thumb web, Li.3 on the dorsal space between first and second metatarsals, to be pressed or massaged alternately each side.

Vibrators over the chest wall can aid relaxation. Some find it helpful to cuddle a not-too-hot hot water bottle. Some benefit from rhythmic slow percussion to help promote relaxation. Other techniques to reduce the work of breathing are described in Chapter 5.

Mucus may or may not be a problem (Strunk 1993). Slow percussion has the benefit of both promoting relaxation and clearing secretions. If secretions are too thick to shift, warm humidification with normal saline may be indicated (Phillips and Millard 1994), but this can increase airflow resistance in children or those with severe airways obstruction, so close observation for desaturation or bronchospasm is required.

Patients who continue to tire will need mechanical assistance. Low-to-medium levels of CPAP will relieve the inspiratory muscles from their relentless work of holding open the obstructed airways (Shivaram *et al* 1987). If CPAP is not available, IPPB can be used to ease the work of breathing. If positive pressure aids are needed, the radiograph should be checked in case of pneumothorax. Mechanical ventilation for asthma is discussed in Chapter 11 and asthma in children in Chapter 12.

3.2.5 Bronchiectasis

Bronchiectasis is characterized by chronic irreversible dilatation and distortion of the bronchi. This originates from an inflammatory insult, e.g.:

- inhalation of unwanted material such as gastric contents or a peanut,
- severe respiratory infection, particularly if repeated.

The incidence of bronchiectasis is diminishing in countries where living standards are rising and children are vaccinated

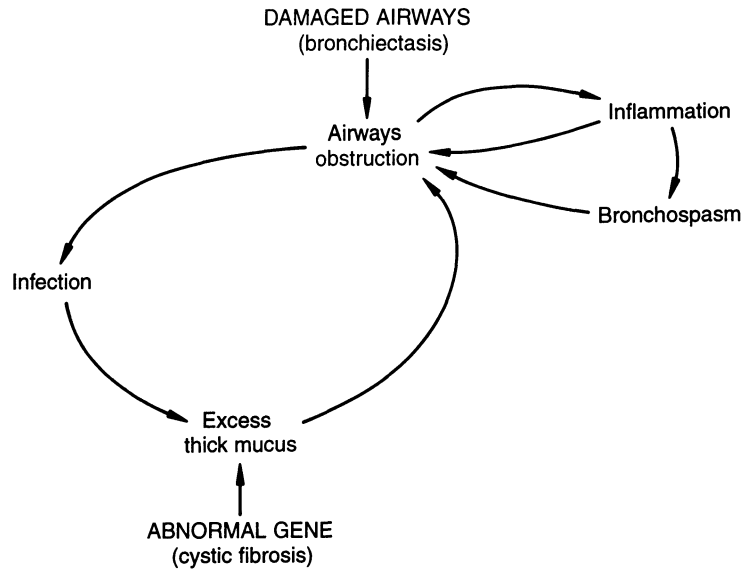

Figure 3.12 Vicious cycle that augments the processes of cystic fibrosis and bronchiectasis.

against diseases such as whooping cough and measles.

The **pathophysiology** involves destruction of the elastic and muscular components of the airway walls. This is most damaging if it occurs in early childhood before the respiratory tract is fully developed. The warm, moist environment within the lung combines with excess mucus to set up a vicious cycle of infection, destroyed cilia, disorganized airways, persistent inflammation and further obstruction (Fig. 3.12). Infection and inflammation release toxic chemicals which interfere with lung defences. Inflammation can cause bronchospasm, which augments the cycle. The process may be complicated by abscess formation. A third to a half of patients show progressive destruction (Munro 1992), leading to pulmonary hypertension and cor pulmonale.

Clinical features include coarse wheezes and crackles, due to secretions and collapsing airways on expiration (Piirilä *et al* 1991), finger clubbing, dyspnoea and fatigue. Coughing is less efficient than normal because of corrugated airways, but voluminous quantities of purulent sputum are produced, sometimes with haemoptysis caused by mucosal ulceration. Radiological signs, if present, are usually localized, showing parallel tramlines representing thickened airway walls and cystic ring shadows representing dilated airways seen end-on. There may be patchy areas of overinflation, consolidation and atelectasis.

Medical treatment is based on the liberal prescription of antibiotics, which help control infection but not the persistent inflammation which may be progressively destroying the airways (Shum *et al* 1993). Other drugs are inhaled steroids, which reduce inflammation and the volume of sputum (Elborn *et al* 1992), and bronchodilators for patients with demonstrable hyperreactive airways. Surgical resection is occasionally indicated for localized and disabling disease.

Physiotherapy is by education in sputum clearance. Hydration must always be attended to, and an exercise programme may be sufficient for moderate disease, but other measures (Chapter 5) are needed for most patients. Much encouragement is needed to

help patients set up a life-long programme that is both effective and suited to their lifestyle. Thereafter occasional checks are needed.

3.2.6 Cystic fibrosis (CF)

CF is a chronic progressive obstructive disorder affecting the exocrine glands. It is the commonest lethal inherited disease among white people. It is acquired as an autosomal recessive disorder, so that only people who have two faulty genes – one from each parent – develop the disease. Two carriers have a one-in-four chance of having an affected baby and a two-in-four chance that a baby will be a carrier. The pattern of CF management has been changed by identification of the rogue gene, prenatal diagnosis and transplantation. A child born with CF in the 1990s is expected to survive to middle age, but the disease is still eventually fatal, and treatment is aimed primarily at improving the quality of life. With skilled management, a high proportion of adults live fulfilled lives. The diagnosis is suspected if infants show failure to thrive or repeated chest infections. Suspicions are heightened if a blood test reveals increased levels of the antibody IRT (Harris 1991), and confirmation is by a test for abnormally salty sweat.

Pathophysiology

In most cells the abnormal gene is dormant, but in epithelial cells it is switched on. This impairs ion and water transport across epithelial surfaces of the body, causing dehydration of secretions and obstruction of various body lumens. In the gut there is malabsorption secondary to pancreatic insufficiency, and in the lungs sodium and chloride ions cannot escape from the epithelial cells into the airways in order to maintain hydration of mucus, which becomes thick and sticky.

The respiratory component determines the quality of life and is the usual cause of death. The lungs are structurally normal at birth, then become caught up in a vicious cycle that is similar to that in bronchiectasis (Fig. 3.12), but set off by tenacious mucus which encourages bacterial adherence. CF is progressive, whereas the course of bronchiectasis varies.

Infection is commonly caused by bacteria, but viruses and fungi play undervalued roles. The patient's own inflammatory mediators directed against the colonizing organisms not only fail in their task, but actually cause tissue breakdown, a by-product of which is excess DNA within the cells, leading to even thicker secretions. The viscid mucus, instead of helping clear bacteria, impairs defence, and intractable infection becomes established in early life, leading to a smouldering course of bacterial colonization punctuated by exacerbations.

The range of bacteria is curiously restricted, but *Staphylococcus aureus* causes significant damage, and acquisition of *Pseudomonas/Burkholderia cepacia* poses a particular threat because the organism is unresponsive to many antibiotics and its presence may herald rapid deterioration and premature death (Govan *et al* 1993). Preventive measures against *cepacia* include segregation and minimal social contact, at great personal cost for those who have previously socialized freely. Even sibling separation is tolerated by some families.

Aggravating factors are inflammation, especially in severe disease, and bronchospasm. Malnutrition contributes to impaired respiratory defence and is related to deteriorating lung function, leading to a spiral of decline in both. Pneumothorax occurs in up to 10% of children and 20% of adults, due to rupture of a subpleural bleb or bulla (Noppen *et al* 1994).

As more patients are surviving to adulthood, new clinical difficulties have arisen. Liver and gall bladder problems develop, pancreatic fibrosis can lead to diabetes and dehydration, and vasculitis can affect joints, skin and brain. Bronchial artery hypertrophy may lead to pulmonary haemorrhage. Respir-

atory and cardiac failure eventually supervene.

Clinical features

> *Coughing and spluttering like an old man does not endear one to the general public, and neither does the popular misconception that one is scattering infections round like confetti . . . My fingers are like spoons and I can't wear nail polish . . . It doesn't do too much for one's confidence to know that one has probably got halitosis – so I tend to talk to people sideways on . . .*
>
> Hall 1984

This fictionalized account of the experience of CF underlines the antisocial nature of the disease. Clinical features include incessant coughing, delayed puberty, flatus, increasing breathlessness and relentless weariness. Breath sounds are clear in the early stages, becoming wheezy as a bronchiolitis-like process develops in the small airways, then exhibiting widespread crackles. Other signs are similar to bronchiectasis, with the addition of small stature and sometimes hepatomegaly and GOR. Growth is stunted because of energy imbalance, energy supply being reduced by malabsorption and often anorexia, and energy demand increased by excess work of breathing. The fact that patients do not often look ill means that they have to cope with others not expecting them to suffer fatigue and other invisible problems (Eigen *et al* 1987).

The radiograph is normal at first, then shows patchy opacities in the apical regions, then signs of widespread bronchiectasis, emphysema and, finally, cor pulmonale. If chest pain occurs, it may be due to pleural inflammation, strain from excessive coughing or pneumothorax. Exacerbation is indicated by weight loss or worsening respiratory signs and symptoms.

Males are sterile. Women can have children at some medical risk to themselves, but their child is unlikely by adolescence to have a mother.

In later stages, FEV_1 declines, PaO_2 falls and eventually $PaCO_2$ rises. The inexorable deterioration is anticipated by patients, who each respond in their individual way. They often form strong attachments to each other, which provide comradeship but can be devastating when one of them dies.

Medical management

Prevention Screening is possible at three stages. Carrier screening helps when making decisions about reproduction, and if two carriers want to have a child, they can be offered *in vitro* fertilization, genetic screening and implantation of a healthy embryo. Prenatal diagnosis provides information on which to base decisions about continuing a pregnancy. Neonatal screening leads to early diagnosis and more effective treatment. Screening usually occurs only after the birth of the first, unexpected, cystic child or if there is a family history of CF.

Research into gene therapy is proceeding at a dizzy pace and if successful could provide a virtual cure for the disease by halting it in its tracks. The accessibility of the airway makes CF suitable for gene therapy, which involves inhalation of a normal copy of the gene to sufficient lung epithelial cells to replace the defective gene. Treatment would be required monthly because of the turnover of these cells, and the damage that had already occurred would not be reversible.

Education When a baby with CF is born, education for the parents should begin immediately:

- no smoking in the home,
- CF children are of normal intelligence and should go to normal schools,
- within their limitations they should take part in normal physical activities,
- they are not infectious,

- they should share with their healthy siblings the disciplines and standards of the family,
- the lifelong treatment routine is time consuming, and preplanning can ensure attention for siblings and prevent isolation for the family.

Self-help groups are valued by adolescents, young adults and parents.

Medication At present the backbone of medical management is an unremitting onslaught against bacterial infection. Antibiotics are used aggressively, but complete eradication of infection is impossible. High doses are required to compensate for difficulty in reaching the lung through areas of poor perfusion or obstructed airways. Access is by peripheral or central venous line or subcutaneous venous reservoir (Davies *et al* 1991), often managed from home with back-up support. Nebulized antibiotics are preferred by some patients, but are time consuming, polluting and variable in effect (Mukhopadhyay *et al* 1994).

The drug DNase is a clone of the gene responsible for breaking down DNA, and when given as an aerosol can reduce secretion viscosity and improve pulmonary function by 10–15% (Hardy 1993). It is also helpful if there is intractable atelectasis (Shah *et al* 1994). Aerosolized alpha$_1$-antitrypsin helps replenish the patient's defences (Briars and Warner 1993). Aerosolized amiloride, a sodium channel blocker, helps restore normal hydration to secretions but requires up to four treatments a day and has been challenged as no better than inhaled saline (Middleton *et al* 1993). Anti-inflammatory therapy may reduce the inflammation associated with infection (Konstan *et al* 1990). Asthma is often present, and bronchodilators improve lung function if hyperreactivity can be demonstrated (Eggleston 1991). Mucolytic drugs are considered ineffective in CF (Fiel 1993).

Nutrition Malnutrition is associated with poor survival (Heijerman 1993). Nutritional support is essential in childhood to prevent impaired growth, and a calorie intake up to 150% of normal is the goal. Enzyme supplements are usually required throughout life to make up for pancreatic insufficiency. Supplementary feeds may be used in advanced disease.

Surgery Pneumothoraces are managed by chest tube drainage if minor, but recurrent pneumothoraces require thoracoscopic pleurodesis (Noppen *et al* 1994). More intrusive intervention precludes future lung transplants because of pleural adhesions.

Transplantation of heart, lung and/or liver can transform a chair-ridden patient into an active individual within weeks of the operation. Most of the pulmonary problems of CF can be eliminated because donor lungs do not have the genetic abnormality. But the obstacles are formidable, including the stress of waiting, life-long immunosuppressive drugs for the successful, and dashed hopes for the unsuccessful (Whitehead and Leval 1994). Selection criteria include life expectancy < 2 years, oxygen dependence, $FEV_1 < 30\%$ predicted, cor pulmonale and high motivation. The availability of transplantation has raised difficult moral issues. Gentle palliative management may now be supplanted by vigorous gastrostomy feeding, mechanical ventilation and other heroics to keep an increasingly desperate patient alive.

Patients must be free to make their own choices, especially in the later stages. Options include nocturnal oxygen therapy (Coates 1992), nasal ventilation (Regnis 1994) or palliation.

Physiotherapy

Once cystic fibrosis has been diagnosed, physiotherapy is started immediately, with the intention of minimizing the cycle of excess secretions and airway damage. Physio-

therapy is the most gruelling and least tolerated aspect of treatment and shows adherence rates below 50% (Abbott *et al* 1994). Daily treatment regimes produce no immediate improvement in well-being, and sputum is the only reinforcement to encourage this repetitive task. Parents of CF children are compliant with treatment, but less than half of CF adults believe physiotherapy to be effective (Fong 1994), and some physiotherapists consider that it is not necessary for all patients (Samuels *et al* 1995). Treatment to clear secretions is best individualized according to patient preference, and should achieve maximum effectiveness with minimum burden on an already stressed family.

Physiotherapy should take place after any prescribed bronchodilator treatment to open the airways, and before nebulized antibiotics to encourage absorption through mucus-filled airways. Details of sputum clearance techniques are in Chapter 5, with aspects specific to CF outlined below.

Positive expiratory pressure, the active cycle of breathing techniques and autogenic drainage are popular because they allow independence.

If postural drainage is the chosen treatment, drainage time is about 15 minutes in younger children, more in older patients or if there are excessive secretions. Length of time depends on fatigue, patient preference, whether there are scant or copious secretions and the effectiveness of other measures. History and symptoms should be checked for GOR, because the head-down postural drainage position exacerbates symptoms and should be avoided (Button *et al* 1994). Percussion and vibrations can be included if they produce more sputum or the patient finds them effective. Some authorities consider them unnecessary (Sutton *et al* 1985), but the combination of all three techniques has shown positive outcomes (Reisman 1988).

Exercise has the advantage that most patients enjoy it and will actually do it (Abbott *et al* 1994). It usually complements other techniques, but in less severe cases can be the primary treatment (Andréasson *et al* 1987). Patients with severe disease are less likely to benefit, partly because of fatigue and partly because little extra tidal volume can be superimposed on hyperinflated lungs. Swimming is especially beneficial, but patients should choose their favourite activity, which may be trampolining for children and weight-lifting or jogging for adults.

Exercise training has the added benefit of improving breathlessness, well-being, clinical status and lung function (Dodd 1991). For training, patients exercise to a pulse rate of 50–75% of that obtained during maximum exercise capacity, with the duration and frequency tailored to the individual but aiming at a minimum 30 minutes four times a week. Successful exercise training requires regular contact with a physiotherapist and commitment from patient and family.

Inspiratory muscle endurance is normally elevated because of the extra work required to breathe through obstructed airways, but it has been claimed that inspiratory muscle training improves exercise tolerance (Sawyer and Clanton 1993).

Interesting data have emerged suggesting that abdominal breathing with biofeedback can reduce airways obstruction (Delk *et al* 1993).

The optimal frequency for physiotherapy is not known (Eigen *et al* 1987), but it is usually performed twice daily, with variations depending on quantity of secretions and whether there is remission or exacerbation. Treatment is best continued until sputum is no longer expectorated or a rest is needed.

Both parents should be given advice and support until children are able to manage themselves, with a check on treatment techniques every three months. Early independence should be encouraged, with young children actively participating in their treatment, and 10-year-olds encouraged to stay

with friends overnight and do their own treatment. Older children are advised against cough suppression, by which they sometimes conceal their illness from peers.

The 'best' treatment is not always the most effective in the teenage years, when it may not be followed. Management is best negotiated, with the physiotherapist 'complying' with the patient's wish rather than the other way round. People with CF are particularly worth listening to because they are medically streetwise and understand much about their treatment. When patients are hospitalized, motivation is enhanced by simple measures such as offering a choice of treatment times and techniques.

Outcome measures are based on quality-of-life measures such as exercise tolerance, questionnaires or well-being scales (Orenstein and Kaplan, 1991).

Precautions

Patients should not be prevented from coughing, but also not exhorted unnecessarily, because excessive coughing causes collapse of central airways with impairment of sputum clearance (Zapleta *et al* 1983), and may accelerate emphysematous changes. The active cycle of breathing provides a more controlled means of sputum clearance, although it still causes a degree of airway closure.

When using CPAP or nasal ventilation (Chapter 5), high pressures are not necessary for respite from fatigue and should be avoided because of the risk of pneumothorax.

If haematemesis develops from oesophageal varices associated with liver cirrhosis, all physiotherapy, except abdominal breathing, is contraindicated until bleeding is controlled.

Blood streaking of sputum is common in CF and should be disregarded, but frank haemoptysis should be reported and physiotherapy temporarily halted.

Measures to prevent cross-infection include separate treatment areas, scrupulous hand washing, single patient use of PEP and flutter devices and the covering of sputum pots.

Longer lifespans mean that osteoporosis is emerging as a problem in young adults (Bachrach *et al* 1994), sometimes represented by increased kyphosis. Loss of bone mineral is due to steroids, chronic respiratory acidosis and, for older patients, the limited diets that were advised in the past.

It is not known if short periods of oxygen desaturation are harmful, but those with an $FEV_1 < 50\%$ predicted are likely to desaturate during exercise. They need supervised exercise testing and should exercise at a level that avoids desaturating by more than 5% or an absolute level below 80% (Dodd 1991). In advanced disease, added oxygen allows longer periods of exercise and may limit pulmonary hypertension (Marcus 1992). Fluids and free access to the salt shaker are needed in hot weather because of the high salt concentration in sweat.

3.2.7 Primary ciliary dyskinesia

Primary ciliary dyskinesia is often misdiagnosed as CF. It is an inherited condition characterized by an uncoordinated and ineffectual ciliary beat, causing recurrent infection of ears, sinuses and lungs. It is suspected in children with a perpetually runny nose, glue ear and frequent chest infections. Regular physiotherapy delays the onset of bronchiectasis (Baum 1990).

3.2.8 Allergic bronchopulmonary aspergillosis

Aspergillosis is an infection manifesting mainly in the lung as allergic bronchopulmonary aspergillosis, which is an allergic reaction to the *Aspergillus* fungus. It occurs in 10% of people with CF, up to 20% of asthmatics and often in cavitating lung diseases, such as TB.

Patients present with malaise, weight loss, fever, haemoptysis and a cough productive of brown, rubbery mucus casts sometimes in the shape of the bronchial tree. The disorder may continue for years with episodes of pulmonary infiltration and wheezing, sometimes leading to fibrosis and cor pulmonale.

Treatment is by inhaled steroids (Seaton 1994), antifungal agents delivered bronchoscopically or percutaneously, or for those with adequate lung function, surgical resection (Jackson *et al* 1993).

3.3 RESTRICTIVE DISORDERS

3.3.1 Pneumonia

Pneumonia is acute inflammation of lung parenchyma, which fills alveoli with inflammatory products, creating consolidation. When alveoli are filled with something other than air, this is termed consolidation. The precipitating factor for pneumonia is a breach in lung defences by infective or chemical agents. The disease is a common cause of death because of its predilection for the elderly and immunosuppressed. Other risk factors are heart disease, acute stroke, poor nutrition, smoking and alcoholism.

Clinical features are fever, chills, breathlessness and often dehydration. If localized, the affected area shows decreased expansion, a dull percussion note, bronchial breath sounds, opacity on X-ray and sometimes a pleural rub. There may be a dry cough at first, which can become productive of purulent and sometimes rusty blood-stained sputum. The structure of the lung is preserved and complete resolution is possible, although sensitized nerve endings sometimes leave a dry, irritating cough.

Treatment is by oral or intravenous fluids, oxygen if indicated and antibiotics if there is bacterial infection. In the acute stage when consolidation is not yet resolved, physiotherapy is limited to positioning for $\dot{V}_A/\dot{Q}$ matching (p. 111) and sometimes CPAP if hypoxaemia persists despite 40% oxygen (Brett and Sinclair 1993). There is overlap between different types of pneumonia, but the usual classifications are described below, with an explanation of modified physiotherapy when appropriate.

Bronchopneumonia

Bronchopneumonia is patchy and scattered, often favouring the lower lobes. It is common in the immobile and elderly. Early signs are dullness to percussion and barely-perceptible fine crackles which persist despite deep breathing. Physiotherapy is based on hydration and early mobility. Other measures may be needed to increase lung volume or improve gas exchange. When the disease is resolving, some patients need assistance or advice to clear secretions.

Lobar pneumonia

When pneumonia is confined to a lobe, localized pleuritic pain and bronchial breathing are added features. Physiotherapy is as above plus relief of pain, for example, by transcutaneous nerve stimulation.

Pneumocystis carinii pneumonia (PCP)

If a person's defence mechanisms are weakened by HIV or medication given after organ transplant, he or she is vulnerable to opportunist infection by organisms that are not normally pathogenic. PCP is a manifestation of this and is a common first AIDS-defining illness in people with HIV, especially in those who smoke.

Invading organisms damage the alveolar lining, and a foamy exudate interferes with gas exchange. Clinical features include a dry cough, breathlessness, hypoxaemia and the features of stiff lungs. Patients are wasted from diarrhoea, malabsorption, dysphagia and mouth ulcers. They may or may not be pyrexial, because immunocompromised people cannot always mount a fever in

response to infection. Some patients may be reluctant to take a full inspiration in case it brings on coughing or pleuritic pain. Auscultation may be normal or show fine scattered crackles. The radiograph may be normal at first, if immune deficiency delays the appearance of an inflammatory response, but later signs are a perihilar haze, progressing to diffuse symmetrical shadowing and air bronchograms (Fig. 2.10). Reduced peak flows sometimes indicate a degree of bronchospasm. Sudden deterioration raises suspicions of a pneumothorax.

Drug treatment is most effective if started early. High-dose steroids, co-trimoxazole or pentamidine are given.

Physiotherapists are involved in the following ways:

1. Diagnostic procedures, such as bronchoscopy, can be avoided by using the more comfortable technique of inducing sputum from the lower respiratory tract for analysis. After starvation and mouth cleaning to reduce oral contamination, 20–30 ml of hypertonic (3–5%) saline is ultrasonically nebulized over 10–20 minutes. This irritates the airways to produce secretions, and the hypertonicity of the saline draws water into the airways (Miller *et al* 1991). The patient is asked to breathe through an open mouth during nebulization, to take occasional deep breaths and gargle with sterile water before coughing so that oral pathogens do not contaminate the specimen. Side-effects are bronchospasm, breathlessness, oxygen desaturation and nausea. TB is common in patients with HIV, and a well-ventilated room is needed to minimize cross-infection from coughing. Ideally, two sputum specimens are obtained because the second is thought to originate from deeper in the lung. The sputum should be clear, non-purulent and resemble saliva. A simpler diagnostic test is oxygen desaturation during exercise (Chouaid *et al* 1993).
2. Physiotherapists may be involved in administering nebulized drugs to the lung parenchyma. A nebulizer is needed that can deliver particle sizes of 2–5 μm and incorporates a filter to reduce environmental contamination. The room needs an extractor fan or venting system. It is best that patients lie down to ensure that the upper lobes are not excluded (Thomas *et al* 1990). The side-effects of coughing and bronchospasm can be reduced by prior bronchodilator inhalation (Harrison and Laube 1994).
3. Patients need support when in the grip of acute breathlessness because they are intensely frightened and often think that they are dying. Physical assistance is described on p. 154. Positioning depends on the patient's choice, but when breathlessness is severe, minimal handling is preferred. Reassurance can be given because although AIDS patients know that they have a fatal disease, death rarely occurs during episodes of acute breathlessness.
4. Help with mobilization may be needed because of weakness.
5. Immune function can be improved by relaxation and exercise which incorporates endurance and strengthening (Lang 1991).
6. Neuropathic pain can be relieved by massage, which is especially important in this group of people, to whom touch is still sometimes denied.
7. CPAP (p. 115) at pressures of 5–10 cmH_2O improves gas exchange in patients with severe PCP (Miller and Semple, 1991) and buys time for discussion with the patient about further treatment. If necessary, some patients may choose mechanical ventilation while antimicrobial drugs have time to work. Ventilated patients with PCP rarely have a secretion problem, and physiotherapy is based on

positioning for $\dot{V}_A/\dot{Q}$ matching, maintenance of lung volume and comfort.

Patients are prone to infections and extra care is needed with hand washing and sterilization of respiratory equipment. Patients are often undernourished, dehydrated and stressed. They need autonomy and attention to their many individual needs. In the later stages, they need particular attention to physical comfort.

People with AIDS may have other respiratory complications, such as lung abscesses and pleural effusion, and other problems such as diarrhoea, dry skin, oral herpes, Kaposi's sarcoma (p. 83) or a sore mouth, which may make eating difficult and oxygen therapy intolerable. Necrotic lung tissue may rupture and cause a pneumothorax, which is notoriously difficult to treat (Light 1993).

Body fluids known to contain the HIV are blood, semen, vaginal secretions and urine. The virus has not been found in normal sputum in enough quantities to pose a risk, but any sputum may contain blood and any patient may be HIV-positive. Precautions against transmission include covering cuts or abrasions with waterproof plasters, wearing gloves during contact with body fluids and wearing masks and protective eyewear during suction.

Nosocomial pneumonia

Pneumonia which develops in a patient hospitalized for over 48 hours is considered to be hospital-acquired or nosocomial pneumonia. It is the leading cause of hospital-related mortality (Heyland 1994) and involves a different spectrum of pathogens from community-acquired pneumonia. The disease may be caused by cross-infection between patients, usually carried by staff, or endogenous infection arising from other colonized sites in the patient, such as a tracheostomy or the gut. A major cause is misuse of broad spectrum antibiotics (Fiorentini 1992).

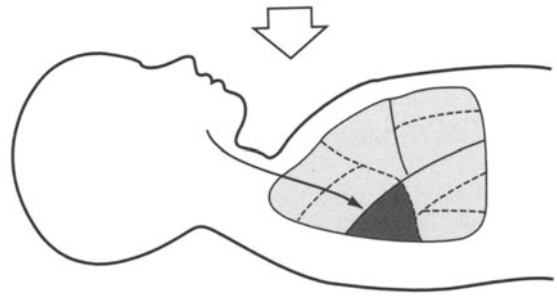

Figure 3.13 Area of aspiration pneumonia for a supine patient.

Legionella pneumonia

Pneumonia that occurs in local outbreaks, especially in relation to cooling systems, or develops after a trip abroad, raises suspicions of Legionnaire's disease, a community-acquired pneumonia with a mortality of 5–10%.

Aspiration pneumonia

People who have inhaled unfriendly substances, such as vomit or gastric acid, need immediate physiotherapy to clear their lungs. Aspiration (inhalation) sets up a vicious pneumonitis which corrodes the alveolar–capillary membrane and leaves a legacy of pulmonary oedema, haemorrhage, necrosis and the classic signs of pneumonia. Aspiration pneumonia should be suspected in anyone after a period of unexpected unconsciousness or in those with swallowing difficulties. The area most likely to be affected is the apical segment of the right lower lobe because this is dependent when the patient is supine (Fig. 3.13).

Signs of acute aspiration include coughing or choking, added sounds on auscultation, gurgly voice or loss of voice, tachycardia and sometimes change of colour. Chronic aspiration is caused by dysphagia or a poor gag reflex, and is suspected in patients with recurrent right lower lobe pneumonia, spiking temperatures, excess oral secretions, reluctance to eat or drink, weight loss or

GOR. Silent aspiration during sleep is common in elderly people (Kikuchi *et al* 1994).

3.3.2 Interstitial lung disease

The umbrella term 'interstitial lung disease' covers a bewildering array of conditions. Over 200 disorders, often related to immune disturbances and/or exposure to toxic agents, are associated with generalized inflammatory changes, to which the lungs are particularly susceptible. Inflammation leads to alveolitis, which may resolve or progress to fibrosis, leading to thickened alveolar septa and small, stiff lungs. Stiff lungs lead to increased elastic recoil (usually) and fewer functioning alveoli (always). Smoking augments the lung injury. Examples are described below.

- **Fibrosing alveolitis** is the commonest interstitial lung disease and kills half of its sufferers within five years of diagnosis (Bois 1992). It may have no obvious cause or may be the end result of other disorders, including those described below (which may or may not be classified separately). There is widespread fibrosis of the alveolar walls and eventually severe impairment.
- **Asbestosis** is lung fibrosis which can occur 20 years after asbestosis exposure.
- **Pneumoconiosis** is lung fibrosis caused by inhaling mineral dust such as coal.
- **Bird fancier's** or **farmer's lung** are allergies which produce fever and malaise some hours after exposure. Lung fibrosis may develop if the patient and offending antigen do not remove themselves from each other.
- **Systemic lupus erythematosis** (SLE) is a connective tissue disorder characterized by exacerbations and remissions, including joint pain. The lung may become involved by developing pleurisy, pleural effusion and fibrosis.
- **Scleroderma** is a connective tissue disorder which is confined to the skin at first, but often progresses to internal organs including the lung.
- **Rheumatoid disease** is a systemic disease of connective tissue, manifesting usually as inflamed joints and sometimes including 'rheumatoid lung', i.e. pleural effusion, nodules, predisposition to infection and fibrosis.
- **Sarcoidosis** is a multisystem disorder of unknown cause, often presenting in young people with widespread variation in severity including sometimes lung fibrosis.

Clinical features

The lungs have a large reserve capacity and the following symptoms only emerge after considerable injury has occurred:

1. Shallow breathing to ease the elastic load, and rapid breathing to sustain ventilation.
2. Dry unproductive cough.
3. Fine end-inspiratory crackles on auscultation, due to sudden opening of peripheral airways.
4. X-ray signs of irregular 'ground glass' lung fields at first, then a reticular or honeycomb appearance as alveoli are pulled apart to form cystic spaces (Fig. 2.9). In advanced cases the shrinking lung pulls up the diaphragm to an exaggerated dome shape.
5. PaO_2 is reduced due to $\dot{V}_A/\dot{Q}$ mismatch, and $PaCO_2$ is reduced due to rapid shallow breathing. Hypercapnia is rare even in severe disease.
6. Respiratory function tests show impaired gas transfer and reduced lung volumes, both static (VC and TLC) and dynamic (FEV_1).
7. Dyspnoea may become progressively incapacitating.

Treatment

Early diagnosis is possible with CT scanning. Patients may not respond to medication, but

symptoms are sometimes alleviated by immunosuppressive drugs or steroids. Oxygen may be helpful, and portable oxygen can improve exercise tolerance. Single lung transplant offers hope for people with severe irreversible disease.

Physiotherapy is aimed at reducing the work of breathing (Chapters 5 and 6) and maintaining functional activities within the limits of dyspnoea, desaturation and fatigue. Help with positioning is appreciated in the late stages. In the unlikely event of a patient being mechanically ventilated, bag-squeezing should be used minimally because the non-compliant lungs are at risk of pneumothorax.

3.3.3 Pleurisy

Pleurisy is inflammation of the pleural membranes, sometimes associated with lobar pneumonia. The parietal pleura is highly sensitive, and pleurisy causes a wicked localized pain, leading to rapid shallow breathing, a stony, dull percussion note and pleural friction rub. The pain can sometimes be eased by heat or transcutaneous nerve stimulation.

Dry pleurisy may or may not develop into a pleural effusion. This brings relief from symptoms as the raw pleural membranes are separated by fluid.

3.3.4 Pleural effusion

The pleural space normally contains 7–14 ml of fluid (Bartter 1994). Pleural effusion is excess fluid in the pleural cavity, caused by disturbed osmotic or hydrostatic pressure in the plasma, or changes in membrane permeability. Twenty-five per cent of pleural effusions are due to malignancy (Bartter 1994); other causes include heart, renal or liver failure, abdominal or cardiac surgery, pneumonia or TB.

Clinical features include a stony dull percussion note, decreased breath sounds over the affected area, and increased vocal resonance over the upper level of the fluid. A fluid line on X-ray is usually confirmatory, often tracking up the pleura laterally or seeping into the fissure. CT scanning distinguishes pleural effusion from pleural thickening. A large effusion displaces the mediastinum and causes breathlessness.

General treatment is directed at the cause, plus symptomatic relief of breathlessness by needle aspiration (thoracocentesis), performed slowly to avoid 're-expansion pulmonary oedema' (Kam *et al* 1993). Surgery may be needed for a thickened restrictive pleura.

Malignant invasion of the pleura is less responsive to treatment, but can be palliated by repeated aspiration, although most effusions re-accumulate within one to three days. Other options include chest tube drainage with local instillation of drugs, the more comfortable pleuroperitoneal shunt, or pleurodesis (Reid and Rudd 1993).

Physiotherapy is limited. Deep breathing exercises cannot expand lungs under pressure from fluid (Dechman *et al* 1993), but mobilization of the patient helps to prevent atelectasis, and positioning can be used to optimize gas exchange. People with moderate unilateral effusion benefit from side-lying with the fluid uppermost, because both ventilation and perfusion are greater in the lower lung, thereby optimizing $\dot{V}_A/\dot{Q}$ matching. Large effusions show improved PaO_2 with the effusion downwards to minimize compression of the unaffected lung (Chang *et al* 1989).

Transudates are clear, low-protein, straw-coloured pleural fluids associated with heart failure and fluid imbalance. **Exudates** are cloudy, high-protein fluids associated with malignancy and infection. **Haemothorax** is blood in the pleura and is managed by treating the cause, plus tube drainage if necessary.

Empyema is pus in the pleural cavity following nearby infection. It is often a complication of pneumonia, bronchiectasis, abscess or chest surgery. The patient may be

asymptomatic or toxic, depending on the offending organism and volume of pus. Early intervention for acute empyema reduces the risk of chronicity and protracted treatment. Local and systemic antibiotics are indicated. Pus can be drained and the lung re-expanded by continuous drainage into an underwater seal system (p. 192) or, for easier mobility, into a drainage bag. The pus may be thick and require aspiration with strong suction or lavage. Other options are open drainage with rib resection, or for an established empyema with a thickened pleura, a full thoracotomy with decortication (Kaplan 1994). Patients needing surgery are often malnourished and debilitated.

3.3.5 Pneumothorax

> *When the pneumothorax happened I was totally conscious of the puncturing of the thorax, the unbearable pain, the cold sweat, being afraid that the final moment would come quicker than the help that you could give me.*
>
> Ruiz 1993

The relationship between the lungs and chest wall is normally maintained by negative pressure which sucks together the two layers of pleura. If either layer is ruptured, air rushes into the pleural space, causing a pneumothorax. The lung shrivels towards the hilum in proportion to the amount of air rushing in, not necessarily symmetrically. Air continues to escape into the pleura until pressure is equalized or the collapsing lung seals the offending hole.

Clinical features are diminished breath sounds, characteristic X-ray signs (Fig. 2.7), rapid breathing by a reflex arc with afferents carried in the vagi, and pain in 75% of patients (Light 1993).

Types of pneumothorax

Spontaneous pneumothorax The apex of the upright lung is subject to greater mechanical stress than the base because the weight of the lung pulls down on it. A spontaneous pneumothorax often occurs in this region, especially in tall, thin young men who are thought to grow faster than their pleura is able to keep up with. Although 'spontaneous', many patients are smokers and have blebs on X-ray (Light 1993).

Secondary pneumothorax A pneumothorax may occur secondary to puncture from a fractured rib, inaccurate insertion of a cannula, high-volume positive pressure ventilation, or rupture of an emphysematous bulla.

Tension pneumothorax A pleural tear sometimes functions as a valve so that air enters the pleural space but cannot escape, causing a pneumothorax under tension. Increasing pressure displaces the mediastinum and impairs venous return, causing respiratory distress and circulatory collapse. Recognition and management is discussed on p. 263.

Treatment

To drain or not to drain? This depends on the size of the pneumothorax and medical opinion. Usually, a small symptomless pneumothorax can be left to heal itself, a moderate first pneumothorax is managed by needle aspiration, and a pneumothorax over 20% or causing breathlessness requires tube drainage over several days.

Chest tube drainage (p. 192) may require suction to help oppose the pleural surfaces and seal the leak more rapidly, but sudden lung re-inflation is avoided to prevent re-expansion pulmonary oedema. Once the air leak has ceased, i.e. when there is no more bubbling in the drainage bottle, the drain is clamped for 24 hours and then removed, provided there is no recurrence, as indicated by X-ray. High levels of inspired oxygen will speed resolution by increasing the absorption

of pleural air fourfold (Light 1993). Recurrence is less likely if sclerosing agents are instilled through the chest drain. Surgical intervention (p. 185) is necessary if these measures fail or if the condition is bilateral.

Physiotherapy is based on mobilization and regular position change. Lying on the side of the pneumothorax may help seal the leak and facilitate lung expansion (Zidulka *et al* 1982), but this is uncomfortable if there is a chest drain, and might cause desaturation with a large pneumothorax because of $\dot{V}_A/\dot{Q}$ mismatch (p. 111).

Mobilization should be particularly rigorous if the cause is, for example, a stab wound (Senekal 1994), but not too enthusiastic immediately after surgery in case the pleura becomes unstuck.

Precautions include avoidance of positive pressure techniques (CPAP, intermittent positive pressure breathing or bag-squeezing) if there is no chest drain.

3.3.6 Neuromuscular and skeletal disorders

Impairment of respiratory drive, muscle power or rib cage mechanics restricts expansion and reduces vital capacity. These conditions may also impair secretion clearance because of a weak cough and reduced mechanical movement of the lung (Mier *et al* 1990).

Pathophysiology and clinical features

Skeletal disorders such as kyphoscoliosis upset the configuration of the diaphragm and force it to work from an inefficient position against the load of a stiff chest wall.

Ankylosing spondylitis is a systemic disease which affects breathing because of a rigid thoracic cage and kyphotic spine. Chest wall compliance is impaired, but lung compliance and diaphragmatic movement are preserved, although occasionally fibrosis and bullous disease damage the lung. The chest X-ray shows apparent hyperinflation because the chest wall becomes fixed in an inspiratory position.

If a patient has generalized muscle weakness, this usually involves the respiratory muscles. Respiratory muscle weakness may go undetected if limb weakness reduces mobility, and ventilatory failure may arrive unexpectedly (Tobin 1988). The most important step in assessment of inspiratory muscle weakness is to think of it as a possibility.

Bilateral paralysis or severe weakness of the diaphragm show the following signs:

- orthopnoea unexplained by heart disease,
- accessory muscle activity unexplained by lung disease,
- paradoxical inward abdominal motion during inspiration, especially in supine when the impaired diaphragm is unable to counteract the weight of the abdominal contents,
- postural fall in VC of about 50% in supine compared with upright (Tobin and Yang 1990),
- symptoms of nocturnal hypoventilation, such as disturbed sleep, morning headache and daytime somnolence,
- less-specific symptoms, such as breathlessness or recurrent chest infections.

Bilateral diaphragmatic paralysis is like removing a portion of the chest wall. When upright, patients exhale by contracting the abdominal muscles, which push up the diaphragm, then relaxing them to allow passive inspiration. Breathing difficulties worsen during sleep.

Unilateral diaphragmatic paralysis shows nocturnal hypoxaemia due to $\dot{V}_A/\dot{Q}$ mismatch in supine, unilateral abdominal paradox on sniffing, and one raised hemidiaphragm on X-ray, this last sign being more obvious than with bilateral paralysis.

Severe cases of neuromuscular and skeletal disorders may lead to type II respiratory

failure which is characteristically associated with hypercapnia (p. 88). Cor pulmonale may develop.

Physiotherapy

Treatment for excess work of breathing or sputum clearance may be needed (Chapter 5), but specific measures are described below.

Upright positioning to facilitate breathing is advised for patients with muscle weakness, and manual support will assist coughing (p. 139). Regular position change and incentive spirometry help to prevent atelectasis.

Swallowing dysfunction is suspected if there is excess salivation, lack of elevation of the larynx on swallowing and deterioration after meals. Risk of aspiration is reduced by avoiding neck extension, maintaining head and chest elevation, and periodic turning from side to side. Dysphagia may lead to weight loss, which further limits mobility, and dehydration, which further limits secretion clearance. Swallowing problems often develop insidiously, but dysphagia usually parallels or shortly follows the development of speech problems. A speech therapist can provide an accurate assessment.

Hypertonic abdominal muscles may occur with some neurological conditions, which inhibits full inspiration and further reduces lung volume. Postures which encourage inhibitory control over spasticity will modify this.

Severe muscle weakness leads to hypercapnia, initially during sleep (Green and Moxham 1993). Nasal ventilation may be appropriate in certain circumstances. Some patients with progressive diseases may accept mechanical assistance if respiratory deterioration is due to a temporary relapse, but if deterioration is progressive, weaning may be impossible. For non-progressive neuromuscular disease, ageing may reduce lung function to the point where mechanical assistance can make life more comfortable.

If respiratory failure is due to bulbar weakness, the airway needs protection and some patients will accept intubation or tracheostomy with a cuffed tube, which will partially prevent aspiration and allow for mechanical ventilation. Impaired communication, due to the disease or mechanical ventilation, can be the most devastating aspect for the patient and family, and a reliable communications system is essential.

3.4 OTHER DISORDERS

3.4.1 Cancer

Lung cancer has shown the greatest rise in mortality for any tumour in the UK this century (Spiro 1993). It is the most common cancer for men worldwide, and is showing an alarming increase in women. Tumour cells usually invade the central airways at bronchial bifurcations, which are the areas most vulnerable to bombardment by inhaled carcinogenic agents.

Clinical features are breathlessness, haemoptysis, clubbing, cough, unresolving pneumonia, weight loss and pain. Recurrent pneumonia in a smoker is a suspicious sign. A large tumour of a main bronchus may produce no radiological change until the lung collapses, but stridor or monophonic wheeze may be heard. Hoarseness indicates involvement of the recurrent laryngeal nerve, which may impair speech and cough.

General treatment is aimed at inflicting the greatest damage to the cancer with the least damage to the patient, but the disease is usually disseminated at presentation and five-year survival is under 10% (Michie 1994). Surgery, radiotherapy and chemotherapy meet with limited success, especially for small cell lung cancer. Other tumours (squamous cell, large cell and adenocarcinoma) may be operable if localized.

Malignant airways obstruction can be palliated and sometimes a lung can be temporarily

reexpanded by cryotherapy, laser resection, localized radiotherapy, stenting to splint open the airway or a combination of these (Rudd 1994). Stenting may cause an irritating cough, which can be eased by nebulized bronchodilators.

A spreading tumour may obstruct the superior vena cava, causing oedema, headache, difficulty breathing, sometimes stridor and faintness on bending down. Drugs, radiotherapy and raising the head of the bed may temporarily relieve the symptoms.

Kaposi's sarcoma (KS) is a vascular tumour which affects the skin and connective tissue, and occurs in immunocompromised people. Pulmonary KS affects the parenchyma, lymph nodes or pleura, and manifests as hypoxaemia, pleural effusion, nodular signs on X-ray, breathlessness and sometimes respiratory failure. Up to a quarter of people with AIDS develop KS (Miller *et al* 1992), representing late-stage disease and a poor prognosis.

Mesothelioma arises in the mesothelial cells of the peritoneum, pericardium or pleura. It is caused by asbestos exposure and is always fatal. In the pleura it is associated with malignant pleural effusion and chest pain.

Physiotherapists may be involved at any stage of cancer from sputum induction (Khajotia 1991) through surgery to care of patients in the terminal stages. Weakness and fatigue are common accompaniments to the disease and its various treatments.

3.4.2 Abscess

Lung abscess is a focal collection of pus within the lung parenchyma, caused either by inhalation of septic material or airway blockage. It leads to cavitation and necrosis. Patients may have a swinging pyrexia, and the X-ray often shows a ring shadow and fluid line (Fig. 2.12). Medical treatment is by antibiotics. Physiotherapy is effective if the abscess is open, postural drainage being safe so long as the correct antibiotic is given and positioning is accurate and thorough to avoid dissemination of infection.

3.4.3 Sleep apnoea

Sleep apnoea occurs when breathing stops for periods of more than 10 seconds during sleep. Nocturnal oxygen saturation may drop to 75%, which stimulates the cortex, and the subsequent arousal is accompanied by spectacular snoring. A typical night includes 300–400 such events (Davies and Stradling 1993). The patient's quality of life is affected by morning headaches due to CO_2 retention, daytime sleepiness, poor concentration and a disgruntled spouse. The condition may lead to respiratory failure, cor pulmonale, and pulmonary and systemic hypertension (Okabe 1995). It often goes unrecognized, and physiotherapists may be the first to suspect the condition. Sleep apnoea is less common in women, possibly because progesterone is a respiratory stimulant. Ten per cent of patients with sleep apnoea have COPD, usually those with the blue bloater pattern (Chaouat 1995). The condition is exacerbated by smoking (Wetter 1994) and high alcohol intake (Jalleh 1993).

Obstructive sleep apnoea (OSA) is due to nocturnal upper airway obstruction despite respiratory effort, and worsens as the night progresses (Charbonneau 1994). It is the commonest form of the condition and is usually associated with obesity, which virtually chokes patients in their own fat when muscle tone wanes at night.

Restrictive sleep apnoea occurs in people whose breathing is already impaired by disorders such as scoliosis, ankylosing spondylitis or diaphragmatic paralysis. For those who have little respiratory reserve, the normal inhibition of accessory muscle action during sleep leads to apnoea.

Central sleep apnoea is caused by abnormal central nervous system control of breathing,

leading to lack of respiratory effort and hypoventilation. Many patients show a mixed obstructive and central pattern.

Associated conditions include the Pickwickian or obesity-hypoventilation syndrome, which is a severe form of OSA seen in markedly obese people, and nocturnal hypoventilation which is an exaggeration of the normal reduction in respiratory drive at night but is not the same as sleep apnoea.

Diagnosis of sleep apnoea used to be a rarified activity in a few specialized centres, but is now commonly made from symptoms, history and sleep studies in hospital or at home. Sleep apnoea can worsen in hospital because of sedative drugs, the supine position and sleep deprivation.

Management of OSA is firstly by weight loss, alcohol and smoking cessation, and avoidance of sleeping supine. Nocturnal oxygen therapy is usually unhelpful (Stewart and Howard 1992).

The upper airway can be splinted open pneumatically at night with CPAP, using suggested pressures of 7.5–2.5 cmH_2O (Miljeteig and Hoffstein 1993). CPAP relieves symptoms and can reverse other problems associated with OSA, such as impotence and Cheyne–Stokes breathing, as well as acting as a catalyst to weight loss which itself may allow discontinuation of the CPAP (Bradley 1993). Education and attention to comfort are necessary to ensure compliance with CPAP.

Some patients who cannot tolerate CPAP manage with various contraptions, such as a nasopharyngeal airway, tongue retainer or mandibular positioning device. More invasive options include tracheostomy, which is poorly tolerated, reconstructive surgery, which is often disappointing, and laser therapy, which is showing some success (MacDougald 1994).

Central sleep apnoea may be helped by drugs to reduce REM sleep or stimulate respiration, supplemental CO_2 therapy (Badr *et al* 1994) or nocturnal nasal ventilation (Bott *et al* 1992). All patients must devise strategies to avoid falling asleep while driving.

3.4.4 Inhaled foreign body

Inhaled objects usually find their way into the right lung because of the more vertical direction of the right main bronchus. Clinical signs include stridor and, if there is complete obstruction, atelectasis due to absorption of trapped air. The occasional brave physiotherapist may attempt to remove the object with accurate postural drainage (Crogan 1991) but most avoid this because of the risk of shifting the offending object to a more dangerous location. More usually the foreign body is removed by bronchoscopy, following which there may be inflammatory secretions or localized collapse that need attention from the physiotherapist.

3.4.5 Pulmonary tuberculosis (TB)

TB is not a disease of the past. The incidence is now increasing in the UK and USA, and new drug-resistant strains are emerging. One-third of the world's population is infected by the TB bacillus, which may become active if the host's defence mechanisms are compromised by, for example, poor living conditions or HIV infection. The HIV pandemic has added a new dimension to TB, which is the only disease likely to be transmitted from AIDS patients to the community.

TB of the lung is the commonest form of the disease, causing three million deaths a year, which is more than any other infection (Empey 1993a). Coughing disseminates infected aerosol, which can remain suspended in the air for hours. Symptoms are fever, night sweats, cough, haemoptysis and breathlessness. The X-ray may show cavitating lesions, especially in the apices, and for miliary TB, scattered small nodules.

The tubercle bacillus is slow growing and tough, needing six months of treatment with a combination of powerful antibacterial drugs. The patient is no longer infectious after two week's treatment providing the sputum is clear of bacillus. The physiotherapist's role is usually confined to eliciting sputum specimens and devising ways to encourage exercise in an isolation cubicle. Some patients may need help in bringing up secretions. Simple paper or cloth masks are not adequate; large pleated masks should be worn and changed between patients (Philips 1992).

3.4.6 Pulmonary manifestations of systemic disease

The respiratory system is influenced by most systemic disturbances, e.g. fever increases minute volume, anaemia hinders oxygen delivery, metabolic acidosis or alkalosis affects breathing and malnutrition predisposes to infection. Breathing itself affects heart rate and BP (Fried 1993). Specific disorders are discussed below.

Cardiovascular disease

Neighbourly relations between heart and lung are reflected in their integrated response to each other's disorders, especially when intravascular pressures are affected.

Heart failure is the inability of cardiac output to meet the body's metabolic demands. It is not a diagnosis in itself but a response to heart or lung disease or heart surgery. It may be acute or chronic and is suspected if a patient with predisposing factors develops fatigue, breathlessness or oedema. Survival rates are lower than for many cancers (Dargie 1994). **Left ventricular failure** (LVF) is the commonest reason for heart failure and is usually caused by coronary heart disease. The failing left ventricle forces up pressure in the left atrium and the pulmonary vascular system behind it, leading to pulmonary oedema (extravascular water in the lungs). Interstitial pulmonary oedema barely affects lung function, but if the lymphatics become overloaded, fluid is squeezed into the alveoli, causing alveolar oedema, a widened $PA\text{-}aO_2$ gradient and hypoxaemia. Non-cardiogenic pulmonary oedema can be caused by fluid overload or increased capillary permeability due to shock or severe infection, independent of left heart function.

The main symptom of LVF with pulmonary oedema is breathlessness caused by stimulation of J receptors, leading occasionally to a misdiagnosis of asthma, from which the confusing term 'cardiac asthma' has arisen. Breathlessness due to pulmonary oedema is distinguished by orthopnoea and paroxysmal nocturnal dyspnoea. Fine crackles are heard on auscultation at the lung bases due to the popping open of alveoli squashed by peribronchial oedema. The X-ray shows an enlarged heart and the signs

described on p. 43, which are often apparent before clinical signs.

Treatment of LVF is by dealing with the cause where possible, plus oxygen, venodilator drugs to reduce filling pressures and diuretics. The role of the physiotherapist is limited to giving symptomatic relief by positioning the patient upright, with support of the feet to prevent the inexorable slide down the bed. The supine posture, that anathema for physiotherapists, is particularly unhelpful for patients with an enlarged heart because of compression of the left mid- and lower lung zones (Wiener *et al* 1990). If mechanical ventilation is necessary, patients rarely require physiotherapy other than positioning and attention to limbs.

Right ventricular failure is caused by LVF, valvular disease of the left heart or chronic hypoxic conditions, such as COPD, which lead to pulmonary hypertension and impaired emptying of the right ventricle. The term **congestive heart failure** means right and left heart failure with congestion in the pulmonary and systemic circulations.

Pulmonary embolus (PE) is a blood clot in the pulmonary vasculature. Secondary blood supply from the bronchial circulation usually keeps lung tissue viable, but this blood exudes into alveoli, causing haemoptysis and an inflammatory reaction which manifests as a sharp, localized pleuritic pain, pleural rub, breathlessness and pallor. Massive embolism causes circulatory collapse. PE is difficult to diagnose clinically, but is suspected in anyone who complains of chest pain and breathlessness, especially if there is hypoxaemia, haemoptysis, pleural rub or evidence of deep vein thrombosis (p. 172). X-ray signs are non-specific, but occasionally a wedge-shaped lesion can be identified. A $\dot{V}/\dot{Q}$ scan shows a perfusion defect (Fig. 2.14). Immediate management is by giving oxygen and placing the patient supine, thus boosting venous return to the left heart which is deprived of pulmonary artery flow (Gray 1992). Treatment is by heparin infusion or, in severe cases, embolectomy. It is unlikely that any physiotherapy treatment could dislodge the clot and, if so, the clot would move safely with the blood flow into a smaller vessel, but liaison with medical staff is advisable before active intervention because of the unstable nature of the condition.

Renal disease

Kidney disease and its treatment affect most body systems, the respiratory system being influenced by any of the following:

- fluid overload, leading to pulmonary oedema and sometimes pleural effusion,
- breathlessness associated with metabolic acidosis or pulmonary oedema,
- muscle wasting due to steroids and uraemia,
- opportunistic chest infection due to steroids and other drugs following transplantation,
- sleep apnoea associated with end stage renal disease (Kimmel *et al* 1989).

Patients in renal failure are supported by one of the following:

1. Haemofiltration, which removes toxins and excess fluid slowly and requires moderate anticoagulation.
2. Renal dialysis or haemodialysis, which is faster and can cause rapid BP changes, pulmonary and systemic inflammatory changes, wheezing, hypoxaemia due to capillary blockage, and bleeding due to anticoagulation.
3. Peritoneal dialysis, in which alkaline solution is run into the peritoneum, left there until waste products have passed into it through the semi-permeable peritoneum, then drained out. This procedure causes minimal haemodynamic disturbance, but restricts basal ventilation and is now less used. Physiotherapy should coincide with the end of the emptying cycle to ensure free diaphragmatic movement.

Precautions when working with patients on renal support are to treat lines for vascular access with respect, to be watchful of fluid volume changes or hypertension and to be aware of the risk of bleeding if patients are anticoagulated.

Liver disease

The connection between liver disorders and respiratory care include the following:

- tracheal suction is performed with caution if there is a danger of clotting disorders associated with liver dysfunction or, for nasotracheal suction, oesophageal varices associated with portal hypertension,
- impaired manufacture of albumin may disturb fluid balance,
- portal hypertension and reduced albumin cause ascites, which splints the diaphragm and leads to a restrictive lung defect,
- encephalopathy reduces the patient's ability to co-operate,
- cerebral oedema causes hyperventilation, which sometimes requires mechanical ventilation (Cowley 1993),
- asterixis is associated with liver disease,
- bilirubin in the plasma of jaundiced patients limits the accuracy of oximetry,
- some patients with grossly enlarged livers are immobile and may not even be able to roll.

Patients admitted for liver transplant are often severely debilitated. A 'Mercedes-Benz' incision incorporates a double subcostal incision and laparotomy, and close teamwork is required for effective pain relief. Postoperative complications include right basal atelectasis, pleural effusion, liver rejection and the long-term effects of immunosuppressive drugs.

Liver transplantation reverses most lung function abnormalities. It is indicated for end-stage cirrhosis, acute fulminant hepatitis or other form of liver failure. Rehabilitation is surprisingly rapid once the toxin-producing liver has been removed. Indeed, some patients may be so poisoned by their own liver that the offending organ is removed even if no donor is immediately available.

When caring for people who have alcoholic liver disease or liver damage from intended paracetamol overdose, physiotherapists need to put aside judgements about 'self-inflicted' conditions.

Sickle cell disease

Sickle cell disease deforms red cells into a sickle shape. Acute vascular occlusion leads to a sickle cell crisis, causing pulmonary vaso-occlusion and excruciating ischaemic pain. Other complaints are cough, breathlessness, chest infection or infarction and atelectasis. Physiotherapy may be indicated for these problems, or if the patient needs assistance with mobilization because of anaemia and fatigue. Epidural analgesia or other potent pain relief is advisable (Yaster *et al* 1994).

Gastro-oesophageal reflux

GOR is the involuntary passage of gastric contents into the oesophagus due to a neuronally-mediated reflex (Ing *et al* 1992). It often occurs at night, especially in people with chronic aspiration or asthma, children with spastic cerebral palsy, people with poor oral health or at the extremes of age. If allowed to enter the airways, the gastric contents cause acid damage to the mucosa. GOR is suspected if there is chronic cough or recurrent chest infection.

Symptoms include heartburn, nocturnal cough, discomfort on swallowing, morning hoarseness and regurgitation with a bitter taste in the mouth, especially after large meals, recumbency or stooping. There may be recurrent pulmonary infiltrates on X-ray. Confirmation of the diagnosis is by endoscopy, barium swallow or pH monitoring (Miles and Ayres 1993). Management is by raising the head of the bed at night, avoid-

ance of late evening meals, large meals, stooping, bending, smoking, alcohol or caffeine, weight reduction if appropriate, drug review and avoidance of aminophylline, which relaxes the cardiac sphincter.

3.4.7 Chest infection

Infection from viruses, bacteria or fungi can occur anywhere from the upper respiratory tract to the lung parenchyma. These are known non-specifically as chest infections. The term encompasses anything from acute bronchitis, a common and usually self-limiting viral infection of the upper bronchial tree, to life-threatening pneumonia. Features include fever, malaise and cough.

Chest infections are a common cause of exacerbation of lung disease, causing excess secretions and impaired mucociliary clearance (Wilson 1988). Those people most at risk are the young, the old, the immunocompromised and those with chronic lung disease. Antibiotics may help bacterial and some fungal infections, but are ineffective against viral infections. Physiotherapy is required if patients are unable to clear their secretions.

3.4.8 Respiratory failure

The main function of the respiratory system is to secure gas exchange, and **respiratory failure** is the inability to maintain blood gases within certain limits. Type I (hypoxaemic) respiratory failure is failure of oxygenation, represented by a $PaO_2 < 8$ kPa (60 mmHg). It is due to failure of the gas exchanging function of the respiratory system and can be acute, e.g. pneumonia, or chronic, e.g. COPD.

Type II (hypoxaemic and hypercapnic) respiratory failure is failure of ventilation, represented by a $PaCO_2 > 6.7$ kPa (50 mmHg) as well as $PaO_2 < 60$ mmHg. It is caused by failure of the respiratory pump and can be acute, e.g. severe acute asthma, or chronic, e.g. advanced COPD. Type II failure is also known as ventilatory failure, and is the outward clinical manifestation of muscle weakness or fatigue. If acute, it is accompanied by a fall in pH because there has been no time for renal compensation.

The process of respiration includes components other than gas exchange in the lung, but the term 'respiratory failure' is reserved for disorders that result in a disturbance of arterial blood gases only. Respiratory insufficiency is when adequate gas exchange is maintained but at great cost to the breathing mechanism (Pilbeam 1992, p. 76).

RECOMMENDED READING

BTS (1993) Guidelines for management of asthma: a summary. *Br. Med. J.*, **306**, 776–82.

Cochrane, G.M. (1995) Acute severe asthma. *Thorax*, **50**, 1–2.

Gross, N.J. (1990) Chronic obstructive pulmonary disease: current concepts and therapeutic approaches. *Chest*, **97**, 19S–23S.

Henderson, A. (1994) Chronic respiratory failure. *Practitioner*, **238**, 345–50.

Hunninghake, G.W. (1995) Approaches to the treatment of pulmonary fibrosis. *Am. J. Respir. Crit. Care Med.*, **151**, 915–18.

MacFarlane, J. (1995) Acute pneumonia in the hospital patient. *Hosp. Update*, 19–23.

McClure, J. (1993) The role of physiotherapy in HIV and AIDS. *Physiotherapy*, **79**, 388–93.

Michie, J. (1994) An introduction to lung cancer. *Physiotherapy*, **80**, 844–7.

Parker, A.E. and Young, C.S. (1991) The physiotherapy management of cystic fibrosis in children. *Physiotherapy*, **77**, 584–6.

Woodhead, M.A. (1992) Management of pneumonia. *Respir. Med.*, **86**, 459–69.