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presentation of respiratory
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ESSENTIALS Respiratory disease can present in many ways, with variations attributable to many factors. The clinical presentation directs diagnostic hypothesis making, the choice of diagnostically discriminating investigations, and the most appropriate management. If a detailed history is not taken, the patient not observed carefully and examined diligently, and the information from these sources is not analysed correctly, then inappropriate investigation and management is likely. History—common symptoms of respiratory disease are breathlessness, cough, haemoptysis, and pleuritic chest pain, details of which can point to particular diagnoses. An account of environmental exposures at work and home, and of family history, is critically important in some cases. Clinical examination—environmental exposures at work and home, and of family history, is critically important in some cases, which can point to particular diagnoses, as can respiratory rate, pulse rate, and temperature. Immediate pointers to respiratory disorder are repeated cough, wheeze or stridor, painful breathing, laboured or ineffective breathing, or cyanosis, but it is crucial to remember that respiratory failure can present as a torpid or drowsy state without clear respiratory distress. Observation of the chest, followed by palpation, percussion, and auscultation must be performed systematically, and the physician who practices these skills regularly will be better at them than the one who does not. In chronic respiratory disease, where breathlessness and disability are to be assessed, walking with the patient and observing exercise tolerance and

distress (and pulse oximetry) can provide valuable information. Introduction The clinical presentation of respiratory disease is protean, with many diseases of different respiratory structures presenting in both common and rare ways. The presentation varies according to many factors—heterogeneity of disease under one diagnostic banner, the natural vagaries of one disease, comorbidities, age and family and social circumstances, and personality traits such as timidity or stoicism. The proper assessment of the clinical presentation therefore needs care and thought. The clinical presentation directs the formulation of diagnostic hypotheses, choice of discriminating investigations, and ultimately the most appropriate management. The combination of the clinical presentation and its careful assessment usually provides the crucial signal as to the seriousness or threat of an acute disorder, and the impact of disease on life and its quality likely to be caused by a chronic problem. A detailed and skilfully taken history will pick up and follow leads offered by the patient, often revealing the diagnosis even before the patient is examined. Indeed, the information from a carefully conducted clinical review by the patient often provides a more complete view of the condition and its impact than the sum of body scans and/or detailed molecular and physiological investigations. These diverse methodologies are of course complementary in good clinical practice, where at the point of management the patient's investigations need to be interpreted in the context of the clinical picture. General features are especially important in the clinical examination of a patient who has, or might have, a respiratory disorder, and they are noticeable from the start of the clinical review in the clinic or medical admissions unit. In the latter, accurate monitoring and documentation of respiratory rate, pulse rate, and temperature are essential. Serious disease may be reflected by an expression of anguish, especially when it is acute, or of dejection when it has progressed inexorably over some time, but mortal disease can also present unobtrusively. Immediate pointers to respiratory disorder, whatever its origin, are repeated cough, wheeze, or stridor, painful breathing, laboured or ineffective breathing, or evident cyanosis. Symptoms Breathlessness Breathlessness refers to an uncomfortable or distressing sensation that occurs when actual ventilation is perceived not to satisfy demand, defined by the American Thoracic Society as 'a subjective experience of breathing discomfort that consists of qualitatively distinct sensations that vary in intensity'. Breathlessness is a complex physiological phenomenon. Its perception is cerebral, with two principal components relevant to most

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Section 18 Respiratory disorders 3948 breathless patients, both of which depend on integration of information. The first allows the perception of breathlessness, and the second allows gauging of its distressfulness. In the first component, the interconnecting neural pathways allow comparison between the volume of efferent motor neurone output to drive respiration, and the volume of afferent sensory input recording the ventilatory effects achieved from juxtacapillary and stretch receptors, carotid body chemoreceptors, and muscle spindles. Breathlessness results when a mismatch occurs between the two—where the motor output (representing effort) exceeds the sensory input (the result achieved). In the second component, further connecting intracerebral neurones allow for this perception of breathlessness to be emotionally interpreted with regards to its degree of distressfulness, introducing a significant psychological component in some individuals. This model offers an understanding of how diverse disorders of the bronchi (limiting airflow) and of the lung parenchyma, pleura, and chest wall (limiting distension) cause breathlessness. It also offers an understanding of how two patients (i.e. with chronic obstructive pulmonary disease, COPD), can have the same derangement of lung function and blood gas measurements, but suffer

very different degrees of distress due to their breathlessness and different degrees of impact on their lives. Such variability in emotional apprehension of breathlessness is important to recognize. It can valuably influence the deployment of rehabilitation programmes, or perhaps cognitive behavioural therapy, in the patient with chronic distressing breathlessness. The character of breathlessness, as related in the history—onset and duration (sudden, acute, subacute, or chronic—see Table 18.2.1), severity, and any associated symptoms—may provide clear diagnostic pointers. One should assess the intensity of the distinct sensations, the degree of distress involved, and the impact on activities of daily living. Most asthmatics clearly describe episodic breathlessness, with a feeling of chest tightness with audible wheeze, and regular early morning (c.04.00–06.00 h) worsening. Many can relate some episodes to exposures to extrinsic triggers (e.g. cats, perfumes, cigarette smoke, or occupational agents, such as flour in bakers). In chronic obstructive pulmonary disease, the typical picture is progressive exertional breathlessness over many months or years, punctuated by exacerbations of this breathlessness during bouts of winter-season, virus-initiated bronchitis, with cough and purulent sputum. Progressive breathlessness, with strict limitation of exercise, is often a feature of pulmonary fibrosis and related disorders of the pulmonary parenchyma. Progressive breathlessness over days and weeks, where the symptoms by night and in bed are worse than the day, and where there are repeated awakenings from sleep by breathlessness, and relief from sitting up, suggest heart failure. Pneumothorax often presents as truly abrupt onset of breathlessness with or without pleural pain. The character of the breathlessness is often less decisive than in the examples given here, and it is only the complete clinical picture (with the full history and clinical examination) and the chest radiograph that advances the diagnostic possibilities, and then further investigations that clarify the diagnosis. For example, most physicians are only too familiar with variable presentation of pulmonary embolism across a clinical spectrum of breathlessness syndromes, fleeting pleuritic pain, and sudden circulatory arrest. Moreover, there is always the potential for diagnostic confusion between the rare manifestation of a common disease (pulmonary oedema due to malaria in the pregnant female), the occasional respiratory manifestation of a multisystem disease (e.g. lupus pneumonitis, or infection in the immunosuppressed), the usual manifestation of a relatively rare respiratory disease (cryptogenic organizing pneumonia), or an idiosyncratic reaction to a medication such as pulmonary eosinophilia syndrome (see ‘Medication history’, later). It should also be noted that breathlessness is not always a prominent presenting symptom in cases of respiratory failure. In respiratory failure due to progressive neuromuscular disorder, chest wall deformity and severe obesity, fatigue and morning headaches due to CO₂ retention may dominate the clinical picture. Cough Cough can be productive or nonproductive, and assessment should establish the amount of sputum produced as well as its consistency and colour. The timing of the cough may be helpful in diagnosing a cause. For example, a cough in the early morning, after exercise, on exposure to allergens such as dust, pollen, animals, or cold air, may point towards an asthma diagnosis. If the patient complains of cough following a meal or when bending down, gastro-oesophageal reflux should be considered. The cough reflex is triggered by irritant and chemical receptors, predominantly of the upper respiratory tract and large airways, and cough can be the principal manifestation of many acute and chronic lung diseases, but also of other disorders (Table 18.2.2). Viral upper respiratory tract infection is the commonest cause of acute onset cough with or without sputum. This can take some 2–3 weeks to subside fully in the otherwise healthy person. In asthma, Table 18.2.1 The onset and duration of different causes of breathlessness

Onset	Causes
Immediate	Acute upper airway obstruction Cardiac arrhythmia Flash pulmonary oedema Pneumothorax Pulmonary embolism
Acute (minutes)	

to hours) Asthma exacerbation Chronic obstructive pulmonary disease exacerbation Hyperventilation syndrome Metabolic acidosis Pneumonia Pulmonary embolism Pulmonary oedema Upper airway obstruction Subacute (days) Acute interstitial pneumonias Atelectasis Pleural effusion Vasculitis Chronic Anaemia Asthma Bronchiectasis Chronic obstructive pulmonary disease Cystic fibrosis Interstitial lung disease Neuromuscular disease Obesity Occupational lung disease Pregnancy Pulmonary vascular disease Thoracic cage deformities

18.2 The clinical presentation of respiratory disease 3949 chronic obstructive pulmonary disease, and bronchiectasis, such viral infections may be accompanied by significant decline in airflow function, with breathlessness and wheeze, or respiratory failure. Cough of acute onset due to inhalation of foreign body is relatively rare, but needs consideration and investigation by bronchoscopy in adults as well as children when there is stridor (see 'Breath sounds', later) or localized inspiratory wheeze in the chest, when dry cough persists unabated, or when the chest radiograph shows asymmetric lung field volumes. A foreign body may inflate or deflate a lung or lobe, dependent on the mechanics of the obstruction. Chronic cough is usually defined as a cough lasting more than 8 weeks, and needs thorough investigation to determine its cause. Chronic cough, with little or no sputum, can often be due to gastro-oesophageal reflux disease, paranasal sinus disease, or common medications. A detailed drug history is important, as drugs can cause cough directly (e.g. angiotensin-converting enzyme inhibitors) or through their effects on the lung (see later). Other features of the history should provide useful pointers to sinus disease (past history of sinus surgery, postnasal drip, nasal blockage, or periodic facial discomfort or fullness) or reflux (episodic 'heartburn', regurgitation of food, or acid brash, morning sore throats, although often related to nonacid reflux). Long-standing cough (of many years' duration) is usually due to tobacco smokers' chronic bronchitis or bronchiectasis. Milder forms of bronchiectasis (although often related to nonacid reflux), history of sinus surgery, postnasal drip, nasal blockage, or periodic facial discomfort or fullness should also be recognized and noted. Asthma is also a common cause of chronic or periodic cough, and in some instances this symptom overshadows wheeze. There may be episodic 'heartburn', in others the diagnosis may require bronchoscopic biopsy to demonstrate the eosinophilic bronchitis typical of asthma. Chronic or periodic cough in some instances overshadows wheeze as a symptom. The possibility of tuberculosis must be addressed by chest radiograph, microscopy, and culture of any sputum, since clinical examination of the lungs is rarely useful (except in advanced cases). Likewise, the clinical features of surgically curable carcinoma of the bronchus are not reliable, hence smokers with a new or change in cough require CT scanning of the thorax and/or bronchoscopy. Other infections, such as lung abscess due to various bacterial infections or fungal infections with localized endemicity (e.g. coccidioidomycosis and paracoccidioidomycosis in the Americas) need the same approach as for the already-mentioned tuberculosis, supplemented as appropriate by serology for certain mycoses, CT scanning, or bronchoscopic samplings for microscopy and culture. A prominently productive cough usually suggests bronchial disorder, such as bronchiectasis or others as just described. In bacterial infection, sputum changes colour to yellow or green owing to the presence of granulocytes and neutrophil myeloperoxidase (green); brown or rusty-coloured sputum is seen in pneumococcal pneumonia; frank pus may be expectorated in cases of lung abscess or bronchiectasis. Parenchymal lung disease may also cause productive cough, as in mucinous adenocarcinoma or tuberculosis. Experienced clinicians recognize that pneumonia may occur without cough and often without sputum, and where the clinical picture is dominated by systemic disturbance (fever, chills, anorexia, or confusion in older people), and with or without other

respiratory symptoms such as pleuritic pain or breathlessness. In some pneumonias (e.g. pneumocystis in the immunosuppressed), breathlessness is regularly the cardinal respiratory symptom. Cough, despite the predominance of airways receptors in its production, can be an important and distressing feature of parenchymal lung disease such as idiopathic pulmonary fibrosis, when opioids may provide the most useful relief. Cough may also feature in pleural effusion or pneumothorax, perhaps because of the bronchial distortion caused. Haemoptysis

Haemoptysis is a frightening symptom, hence presentation to the physician is often rapid, albeit that the amount of blood is frequently overestimated by the patient. Careful enquiry will almost always exclude epistaxis, haematemesis, or other upper airway bleeding (gums, pharynx). There are several important causes (Table 18.2.3). In developing countries, tuberculosis and AIDS-related diseases are the commonest causes, whereas in industrialized nations these are lung cancer, bronchiectasis, infection, and pulmonary embolism. Haemoptysis can occur in simple bouts of acute infective bronchitis, and is a well-recognized feature of pneumonia, as may occur for instance with pneumococcal or klebsiella infection. Haemoptysis, often periodic and coinciding with infective exacerbation, is a common feature of bronchiectasis. Though the bleeding can be relatively profuse, it almost invariably subsides spontaneously and quickly with antibiotic treatment. Haemoptysis is an important presenting feature of carcinoma of the bronchus, tuberculosis, or pulmonary embolism, when the quantity of blood per se may not be impressive. If haemoptysis occurs without clinical features of acute bronchial or pulmonary infection, clinical review needs to be followed by definitive investigation. A CT pulmonary angiography will prove or exclude pulmonary embolism; it will give some clear indication whether carcinoma or tuberculosis are likely, indicating the need for proceeding to bronchoscopy or microscopy for mycobacteria in sputum. The CT can also identify or suggest other rarer diagnoses such as pulmonary arteriovenous malformation, or some forms of pulmonary vasculitis (e.g. polyangiitis with granulomatosis, where pulmonary nodules with or without cavitation may be shown). Diagnosis of Goodpasture's syndrome or lupus depends on clinical suspicion, and serology for antiglomerular basement membrane (anti-GBM) and antinuclear autoantibodies.

Table 18.2.2 Causes of cough

Cause	Example
Upper airway	Inhaled foreign body
Paranasal sinus disease	Upper respiratory tract infection
Upper respiratory tract tumour	Airways disease
Asthma	Cough variant asthma
Lung disease	Bronchial carcinoma or adenoma
Inflammatory parenchymal lung disease (e.g. sarcoidosis, idiopathic pulmonary fibrosis, hypersensitivity pneumonitis)	Inhalation of noxious fumes or gases
Pneumonia (some)	Gastro-oesophageal reflux
Medication	ACE inhibitors
Functional	

Section 18 Respiratory disorders 3950 In massive haemoptysis, immediate resuscitation is paramount, and treatment may run concurrently with or precede definitive investigation. While blood loss can be substantial, the greatest risk is from asphyxiation owing to widespread clotting of blood in the airways rather than blood loss per se, and efforts should be made to prevent blood flooding the unaffected lung. This is best performed, where available, via rigid bronchoscopy under general anaesthetic, allowing the use of topical vasoconstrictors or of balloon tamponade. In practice this is rarely immediately available, and large volume blood loss can make visualization virtually impossible. Double lumen endotracheal tubes allow isolation of the affected lung, and where bleeding does not settle, radiological therapeutic embolization of a bronchial artery or arteriovenous pulmonary malformation can be performed as appropriate or, in extremis, surgical resection of the bleeding lobe.

Chest pain Pain associated with respiratory disease is usually pleuritic in nature. Caused by inflammation or irritation of the parietal pleura (the visceral pleura is insensate), it is sharp and 'catching' in nature, and made worse by anything that increases

pleural friction (e.g. deep inspiration, coughing). Most often that irritation arises from lung disease with extension to the pleura—infection, infarction, tumour—but primary pleural disease is also seen—infection (e.g. from subdiaphragmatic or blood-borne spread), asbestos (inflammatory effusions and mesothelioma), tumour (e.g. metastatic from breast or other cancer), serositis (in lupus or rheumatoid disease), trauma (fractured rib, pneumothorax and its tube drainage). Pleuritic pain, with audible pleural rubs on auscultation (Table 18.2.4), occurs with or without the presence of pleural fluid. By contrast, pleural effusions resulting from transudation from oedematous lungs in heart failure produce neither pleuritic pain nor audible rub, hence their presence under such circumstances suggests compounding disorder such as pulmonary embolism and infarction. Pleuritic pain needs to be distinguished from chest wall pain due to injury of the ribs, or associated muscles from unusual use, trauma, or from forceful repeated coughing. Such chest wall pain can cause diagnostic confusion, particularly when cough is a prominent feature. The absence of audible pleural rub and of pleural effusion, together with marked localized tenderness on palpation at the site of the pain, suggests chest wall disorder. In Bornholm's disease (epidemic pleurodynia due to Coxsackie virus strains B1–5) there is pleuritic-type pain with accompanying malaise and fever and marked local chest tenderness, but normally no audible rub. Chest pain of a nonpleuritic type is not often a feature of lung disease. The exceptions are when tumour invades sensitive tissues in the thorax to cause a dull but often severe pain, and sarcoidosis, where inexplicable fleeting chest pains often occur. Other symptoms Respiratory disease may also present with nonthoracic symptoms, and the dominant features vary. Examples include constitutional symptoms (chills and prostration in pneumonia; fever and sweating in thoracic empyema; drowsiness, headache, and fatigue in respiratory failure; daytime somnolence in sleep apnoea); gastrointestinal symptoms (anorexia and weight loss in lung cancer and tuberculosis); neurological symptoms (headache, weakness, seizures, or cerebellar signs in cerebral metastases; neuromuscular disorder in lung cancer or respiratory muscle weakness; seizure and hyponatraemia in inappropriate antidiuretic hormone syndrome); musculoskeletal symptoms (underlying connective tissue disease). Table 18.2.4 The onset and duration of different causes of pleuritic pain

Onset	Cause
Acute (respiratory)	Costochondritis Muscular pain Pleural infection Pneumonia Pneumothorax Pulmonary embolism Rib fractures
Acute (other)	Angina Aortic dissection Gastro-oesophageal reflux Myocardial infarction Oesophageal rupture Pericarditis Sickle cell crisis
Chronic	Autoimmune disease (systemic lupus erythematosus, rheumatoid arthritis) Benign musculoskeletal pain Malignant pleural disease Pleural infection Recurrent pulmonary embolism Sarcoidosis (often flitting)

Table 18.2.3 Causes of haemoptysis

Cause	Examples
Infection	Abscess Acute purulent bronchitis Aspergillus and other fungi Bronchiectasis Pneumonia (typical and atypical) Tuberculosis (active and inactive)
Malignancy	Benign tumours and adenoma Carcinoid tumour Kaposi's sarcoma in AIDS Lung cancer Pulmonary and bronchial metastases
Vasculitides	Churg–Strauss Goodpasture's syndrome Lung transplant rejection (capillaritis) Polyangiitis with granulomatosis Systemic lupus erythematosus
Cardiovascular	Heart failure with pulmonary oedema Mitral stenosis Pulmonary arteriovenous malformations Pulmonary embolism Clotting disorders Anticoagulation Disseminated intravascular coagulation Haemophilia Thrombocytopaenia
Other	Endometriosis Foreign body aspiration Iatrogenic (transbronchial biopsy; surgery) Idiopathic pulmonary haemosiderosis Trauma (inhalation; contusion)

18.2 The clinical presentation of respiratory disease 3951 tissue or collagen vascular diseases in interstitial lung disease); and dermatological manifestations, which may present before respiratory symptoms become evident (e.g. erythema nodosum in sarcoidosis). Other features of the

history **Past medical history** The past medical history should include questions on premature birth (bronchopulmonary dysplasia), a history of childhood asthma or atopy, recurrent infections, connective tissue disease, previous cancers, cardiac disease, allergies, previous thoracic and gastro-intestinal surgery, as well as nasal polyps and other upper airway pathology. Recent surgery or immobilization predisposes to venous thromboembolism. Cigarette smoking can have devastating results on respiratory health, pulmonary disease and emphysema, and lung cancer. Evidence for inherited disease may emerge from the family history. Affected siblings with premature emphysema due to $\alpha 1$ -antitrypsin or with cystic fibrosis exemplify autosomal recessive disease. Dominant inheritance, but with variable penetration of disease, can sometimes be traced in deep vein thrombosis and pulmonary embolism—for example, resistance of coagulation factor V to protein C inhibition (factor V Leiden). Atopic asthma with rhinitis and eczema is a common polygenic clinical complex which can show familial aggregation. It is important to consider environmental respiratory disease due to exposures at work and home. Such factors include: contact with others with respiratory infection (e.g. tuberculosis, mycoplasma, influenza); place of residence or travel predisposing to acquisition of infections such as coccidioidomycosis, legionella, and tuberculosis; the presence at home of pet animals (e.g. cats and dogs) which can trigger asthmatic reactions or underlie hypersensitivity pneumonitis (e.g. psittacines or pigeons). A thorough history of occupational exposures is required, and prompting is often needed for jobs performed years previously. Asbestos causes inflammatory effusions, benign pleural thickening, mesothelioma, pulmonary fibrosis, and carcinoma of the bronchus. Coal and stone mining cause pneumoconiosis and silicosis. Several inorganic agents (e.g. isocyanates, acid anhydrides) and organic agents (e.g. bread flour, antibiotics) can cause occupational asthma; beryllium exposure in electronics manufacturing causes sarcoid-like granulomatous disease; acute isocyanate exposures can cause pulmonary oedema. Spores from thermophilic bacteria and fungi on vegetable matter are an important agricultural cause of hypersensitivity pneumonitis (e.g. farmer's lung). Industrial accidents include inhalation of noxious gases and fumes. **Medication history** Previous or current drug use—prescribed, or otherwise—can be an important cause of lung symptoms and diseases, notably cough, interstitial disease, and bronchospasm. β -adrenergic blockers exacerbate asthma, sometimes catastrophically. Aspirin and non-steroidal inflammatory drugs can cause asthma and rhinitis in susceptible individuals, thought to be caused by an anomaly in the arachidonic acid metabolizing cascade and increased production of pro-inflammatory cysteinyl leukotrienes. Chemotherapy agents and immunosuppressive or anti-inflammatory regimens (e.g. bleomycin, tyrosine kinase inhibitors, methotrexate) can cause or exacerbate diffuse lung diseases, or can predispose to both pathogenic and opportunistic pulmonary infections (e.g. prolonged high-dosage corticosteroids and pneumocystis, anti-TNF therapy and tuberculosis). Drugs (e.g. dapsone, isoniazid) as well as parasitic worms can cause pulmonary eosinophilia syndromes, and interstitial changes can be seen with some cardiac drugs (e.g. amiodarone, statins). Use of illicit drugs must also be considered. They may cause injury directly, as seen in smoking of marijuana (bullous emphysema) and crack cocaine (interstitial disease, or 'crack lung'), and sedatives causing respiratory failure, or by association (e.g. intravenous drug abuse with dirty needles causing lung abscess by haematogenous spread, or opportunistic infections in those with advanced HIV). A thorough drug history includes over-the-counter or homeopathic preparations (e.g. St John's Wort), and oxygen therapy (long-term and ambulatory). **Clinical examination** The purpose of physical examination is to elicit signs of disease. The underlying diagnosis is often evident from thorough history taking, and signs can be observed from the moment the patient comes into view. Observations such as obesity, cachexia, obvious skeletal deformities, and the smell of cigarette

smoke are all instantly apparent. Immediate pointers to respiratory disorder, whatever its origin, are repeated cough, wheeze, or stridor, painful breathing, laboured or ineffective breathing, or evident cyanosis. A thorough physical examination of the patient using a systematic approach is essential. Accurate monitoring and documentation of respiratory rate, pulse rate, and temperature are essential for any patient who is acutely unwell. The sputum pot can be inspected for the colour, consistency and (in some cases) smell of the sputum therein. The four basic procedures of respiratory examination are inspection, palpation, percussion, and auscultation. Inspection The trachea and lymph nodes are better assessed by palpation, but obvious abnormalities such as a tracheostomy, substantial tracheal tug, or massive lymphadenopathy are apparent on observation. Other signs include engorgement of veins in the neck and superior chest accompanying superior vena cava obstruction; poor dentition is especially relevant when considering pleural infection; conjunctival pallor of anaemia; Horner's syndrome (ptosis, miosis, and anhidrosis), a rare accompaniment to respiratory disease but one that can occur in Pancoast's tumour or as a rare complication of intercostal tube insertion. Breathing pattern The use of accessory muscles of respiration is seen when diaphragm function is insufficient to maintain adequate airflow, and is classically seen in patients with severe airflow obstruction (chronic obstructive pulmonary disease, asthma). Accompanying signs include a barrel-shaped chest; reduced cricosternal distance; tracheal tug on inspiration; purse-lipped breathing (airway splinting by creating positive expiratory airway pressure); prolonged expiration. The diaphragm, already flattened by hyperinflated lungs, may even reduce

Section 18 Respiratory disorders 3952 the intrathoracic volume by pulling in the lower ribs to which it attaches (Hoover's sign). Inspiratory stridor and supraclavicular recession are seen in significant obstruction of the larger airways or larynx, either acute (epiglottitis, inhaled foreign body, laryngeal oedema) or chronic (intra- or extraluminal growths). Fast, deep breathing is consistent with severe pneumonia, other inflammatory parenchymal lung disease (diverse alveolitis), pulmonary embolism, or pulmonary oedema. Waxing and waning of respiratory rate and volume, and including spells of apnoea, can occur in severe heart failure, in neurological disease, or at high altitude (Cheyne-Stokes breathing). A patient's insistence on sitting upright is a regular feature of heart failure with pulmonary oedema (limiting dependent alveolar oedema to the lower zones of the lungs) or severe asthma or chronic obstructive pulmonary disease (gaining mechanical advantage by fixing the upper thorax with rigid arms clutching the bedside). By contrast, many patients with pulmonary embolism and inflammatory lung disease (pneumonia, interstitial disease) seem ready to lie fairly flat on their back or side. The classic deep, laboured, sighing breathing of Kussmaul respiration is characteristic of metabolic acidosis, while shallow, often erratic, respiration is seen in states of anxiety or dysfunctional breathing. The presence of abdominal paradox indicates significant diaphragm weakness (e.g. neuromuscular disease, neuropathy in lupus, the inherited myopathy of Pompe's disease), and usually requires both hemidiaphragms to be affected. Cyanosis Cyanosis is a blue discoloration of the skin (especially nail beds, ear lobes, and lips) and mucous membranes of the mouth, traditionally said to occur when there is greater than 5 g/dl of deoxygenated haemoglobin in the blood, but can be seen at levels lower than this. It indicates a failure of oxygenation through a variety of circulatory, ventilatory, neurological, or haematological causes, although is often not seen until the blood oxygen saturations drop below 85%. Polycythaemia and anaemia, respectively, increase and decrease the likelihood of visible cyanosis for any degree of hypoxaemia, and significant hypoxaemia can occur without clinically detectable cyanosis, emphasizing the value of simple

pulse oximetry as part of the examination. Central cyanosis is a more reliable sign of poor blood oxygenation than peripheral cyanosis, which can occur when there is poor peripheral circulation alone. In chronic respiratory disease, where breathlessness and disability are to be assessed, walking with the patient and observing exercise tolerance and distress can provide particularly valuable information. This can be supplemented by recording the haemoglobin-oxygen saturation by simple pulse oximetry before, during, and after some modest exercise in the clinic room. An uncommon but important cause of cyanosis is methaemoglobinemia, due to oxidation of haemoglobin by a variety of drugs, or due to inherited haemoglobin M or deficiency of methaemoglobin reductase. Methaemoglobin binds oxygen poorly and can lead to symptoms of hypoxaemia, with fatigue and dizziness, which can be treated by intravenous methylene blue (depending on cause). Extrapulmonary features Impairment of cerebral function Just as significant hypoxaemia can occur without evident cyanosis, it is important to realize that respiratory failure can present as a torpid or drowsy state, and without clear respiratory distress—as in severe chronic obstructive pulmonary disease, asthma, sedative overdose, brain stem disorders and encephalitis, neuromuscular disorder, and extreme obesity. Arterial blood gases are therefore a vital investigation in the torpid or drowsy patient, providing vital information on possible respiratory failure or metabolic acidosis. Cerebral disturbance as the presenting feature in respiratory failure is one important example of unexpected presentation. Peripheral stigmata The hands can be a tremendous source of clinical signs in respiratory disease. Clubbing (Table 18.2.5) may occur without the presence of disease, but about half of all cases are seen in patients with non-small cell lung cancer. In hypertrophic pulmonary osteoarthropathy (clubbing, periostitis of the long bones, and arthritis) patients present with clubbing and sore ankles or wrists. Unilateral wasting of the thenar eminence is seen in apical lung tumours involving or compressing the brachial plexus (Pancoast's tumour). Table 18.2.5 Causes of clubbing

Respiratory	Cardiovascular	Gastrointestinal	Haematological	Other
Chronic suppurative lung disease	Cyanotic congenital heart disease	Inflammatory bowel disease	Hodgkin's lymphoma	Idiopathic empyema
Subacute bacterial endocarditis	Cirrhosis of the liver	CML	Hereditary bronchiectasis	Atrial myxoma
Hepatopulmonary syndrome	Myelofibrosis	Thyroid acropachy	cystic fibrosis	Coeliac disease
Axillary artery aneurysm	lung abscess	Tropical sprue	Secondary hyperparathyroidism	tuberculosis
Hepatocellular carcinoma	Pachydermoperiostitis	Pulmonary fibrosis	Pregnancy	Yellow nail syndrome
Oesophageal cancer	Mesothelioma	AV malformations	Note—There are many other causes of clubbing that appear in the literature, but the main causes are listed here.	

18.2 The clinical presentation of respiratory disease 3953 tumour), whereas bilateral signs point to underlying neuromuscular conditions (respiratory muscle weakness, aspiration). The skin should be examined for rashes associated with respiratory disease (Table 18.2.6), and the legs for erythema nodosum (tuberculosis, sarcoidosis), oedema (right heart disease), and unilateral swelling (DVT). Hyperpigmentation of the lower thighs may be seen in those with severe emphysema (Dahl's sign), owing to pressure from the arms when the patient is in the 'tripod' position. A crude assessment of gas exchange can be made by measuring the oxygen saturations and observing for asterixis (the flap of carbon dioxide retention). Orthopnoea is usually readily elicited, and measuring the saturations with the patient both supine and erect may detect platypnoea and orthodeoxia, seen in a variety of cardiac, pulmonary, or hepatic diseases where there is right to left shunting that is more marked in the upright position (although a rare phenomenon). Other peripheral stigmata are listed in Table 18.2.7. Skeleton and muscles Signs may include kyphoscoliosis as the basis of respiratory failure; immobile stiff neck and back of ankylosing spondylitis as the basis for

progressive upper lobe fibrosis; stiff and deformed joints of the hand in rheumatoid disease as the basis for pulmonary nodules, fibrosing alveolitis, and pleural effusion; muscle wasting and weakness from motor neurone disease as the basis of respiratory failure; diplopia and muscle fatigability of myasthenia as the basis for respiratory Table 18.2.6 Skin manifestations of respiratory diseases

Disease	Skin manifestation
Allergy and anaphylaxis	Urticaria
Arteriovenous malformations	Telangiectasia
Dermatomyositis	Heliotrope rash
Eczema	Red bumpy rash becoming scaly and dry
Illicit drug use	Scarred veins of forearms (and elsewhere)
Lung cancer	Palpable nodules
Neurofibromatosis	Cafe-au-lait spots; neurofibromas
Paracoccidioidomycosis	Violaceous facial lesions
Pneumonia (esp. Mycoplasma)	Erythema multiforme
Pneumothorax	Subcutaneous emphysema
Psoriasis	Plaques
Rheumatoid arthritis	Nodules; livedo reticularis; vasculitic rash
Sarcoidosis	Erythema nodosum; lupus pernio
Scleroderma	Telangiectasia; sclerodactyly
SLE	Malar rash; discoid lesions
Tuberculosis	Erythema nodosum; lupus vulgaris
Tuberous sclerosis	Shagreen patches; ash-leaf spots; adenoma sebaceum
Vasculitis	Purpuric rash; palpable purpura

Table 18.2.7 Peripheral signs in respiratory disease

Hands	Face	Neck	Legs	Sign Association
Sign Association	Sign Association	Sign Association	Tar staining	Smoking
Central cyanosis	Hypoxaemia	Lymphadenopathy	Malignancy; infection (tuberculous, viral)	Oedema
Right heart failure, hypoproteinaemia	Clubbing	see Table 18.2.5	Conjunctival pallor	Anaemia
Elevated jugular venous pulse	Cor pulmonale	Erythema nodosum	Tuberculosis, sarcoidosis	Peripheral cyanosis
hypoxaemia	Horner's syndrome	Pancoast's tumour; trauma	Suprasternal notch scar	Previous mediastinoscopy
Unilateral swelling	Venous thromboembolism	Small muscle wasting	Neuromuscular disease;	Pancoast tumour
Pharyngeal injection	Infection, HIV seroconversion	Supraclavicular scar	Phrenic nerve crush (tuberculosis)	Dahl's sign (darkened and thickened skin on lower thighs and elbows)
Emphysema	Asterixis	carbon dioxide retention	Malar flush	Systemic lupus erythematosus; mitral stenosis
Tremor	anxiety states	Parotid enlargement	Uveoparotid fever (Herefordt's syndrome)	(sarcoidosis)
Koilonychia	Iron deficiency anaemia	Microstomia	Systemic sclerosis	Leukonychia
Protein deficiency (pleural effusions)	Xerostomia	Sjorgren's syndrome	Raynaud's phenomenon	Connective tissue diseases
Oral telangiectasia	Hereditary haemorrhagic telangiectasia	Sclerodactyly	Systemic sclerosis	Nasal crusting
Granulomatosis with polyangiitis	Joint deformity	Rheumatoid arthritis	Saddle nose	Granulomatosis with polyangiitis; polychondritis

Section 18 Respiratory disorders 3954 failure. A full neurological examination is essential where there are signs or symptoms of respiratory muscle weakness. Cardiovascular signs The heart and lungs are intimately connected, and disease in one organ not infrequently produces effects in the other. Specific cardiac examination findings can help determine the presence of, or indicate the severity of, respiratory disease. Other than findings associated with left ventricular dysfunction (pulmonary oedema, third heart sound), and perhaps with the exception of mitral stenosis in cases of haemoptysis (owing to pulmonary venous hypertension), most signs in respiratory disease relate to the development of right heart dysfunction or pulmonary hypertension (Table 18.2.8). One should also look for signs of deep vein thrombosis as the origin for pulmonary embolus. Palpation The neck is examined for lymphadenopathy, tracheal deviation (best assessed at the suprasternal notch), and the carotid pulsation. Aside from examination of areas of pain or tenderness (rib fracture, cough trauma, vertebral collapse), or obvious or patient-reported lumps, the palpation of the chest is divided predominantly into the assessment of expansion and detection of chest wall vibrations (transmission of the voice, or fremitus). The hemithorax with disease always expands less than the healthy hemithorax. Anything between the lung and the chest wall (air, fluid, or solid) will decrease the transmission of vibrations to the chest wall, while consolidated lung increases

transmission and hence the detection of vocal fremitus. The same information can be obtained during auscultation by the assessment of vocal resonance. Palpation of the heart can reveal displacement of the apex beat, ventricular heaves, and valvular thrills. In women, a thorough breast examination is essential in cases of pleural effusion or lung masses, and abdominal examination can be useful in selected cases.

Percussion Percussion is a simple but valuable clinical technique, at its best in defining a region of intense thoracic dullness and thereby indicating a pleural effusion, haemothorax, or empyema. It can also suggest, with variable accuracy, the presence of pneumothorax with resonant percussion note, or lobar collapse or consolidation with dull percussion note. Pneumonic lung, while being largely solid with fluid, still retains some aeration in most cases and is therefore less dull than in effusion. Percussion of the heart provides information about displacement, enlargement (dilatation or pericardial effusion), and pneumopericardium (diminished area of cardiac dullness).

Auscultation Auscultation is essential to establishing a secure clinical diagnosis. It should, however, be recognized that several important lung diseases do not produce auscultatory signs. In some this relates to their early phase (e.g. bronchial carcinoma); in others it typifies the diseases through their course (e.g. sarcoidosis, diverse nodular lung diseases as opposed to diffuse or segmental disorders, some diffuse pneumonias such as pneumocystis, and tuberculosis). Findings on clinical examination can be unreliable in small to moderate pneumothorax and in pulmonary embolism. There are three general categories of sound that should be listened for, namely breathing sounds, pleural sounds, and the quality of the transmitted voice.

Breath sounds There are two distinct sounds heard during respiration in the healthy chest. Bronchial breath sounds can be heard over the trachea and large airways in areas where there is little or no lung interspersed between the airways and the chest wall. They tend to be of a similar duration in both inspiration and expiration, and there is a distinct gap between the two when auscultating. Bronchial breathing heard anywhere other than as just described indicates a loss of alveolar integrity such that large airway sounds are transmitted unchanged. This can occur in lung collapse, consolidation, cavitary disease, pneumothorax, occasionally over the upper regions of pleural effusions (when it does not necessarily imply underlying consolidation), and over small areas of lung in gross pulmonary oedema or pulmonary fibrosis. Vesicular breath sounds are heard over most of the lung fields, the large volume of air in the lungs dampening the transmission of sound. They are longer and louder on inspiration than expiration, continuous in nature, louder towards the bases, and are quieter and lower pitched than bronchial sounds. Breath sounds may be globally reduced in chronic obstructive pulmonary disease and severe asthma. Regions of quiet sounds are important and can denote partial obstruction of the bronchus to the region (when no other signs may be found), an underlying large bulla in emphysema, pulmonary collapse, or pleural effusion. The quality of the transmitted voice can help distinguish these causes.

Abnormal breath sounds are known as adventitious sounds. Crackles (rales, crepitations) are short, nonmusical sounds, and are described as fine or coarse. Parallel information from the history and other physical and radiographic signs are often required to help determine a diagnosis. Fine, or dry, crackles are often said to sound like Velcro, and are produced when diseased distal airways and alveoli 'snap' open. Of short duration, they are heard only during inspiration. They classically occur (usually bilaterally) over areas of pulmonary fibrosis (often late and very fine; <10 ms). In some severe cases of fibrosis and adult respiratory distress syndrome, short musical sounds on inspiration (squawks), can accompany the crackles. Coarse, or wet, crackles are generated by fluid-filled alveoli, can be heard in inspiration and expiration, are of a lower pitch, longer duration (close to 20 ms), and are louder than fine crackles. They are heard predominantly in pulmonary oedema, pneumonia, and

Table 18.2.8 Cardiac signs in respiratory disease

Pansystolic murmur of tricuspid

regurgitation Diastolic murmur of pulmonary regurgitation Split second heart sound Loud pulmonary component of second heart sound Right ventricular third heart sound Elevated jugular venous pressure (JVP) Prominent a and v waves in JVP Right ventricular parasternal heave

18.2 The clinical presentation of respiratory disease 3955 bronchiectasis, but occur in any situation where there is excess fluid in the airways. Wheeze is a high-pitched, continuous sound with a whistling quality, (always >80 ms and usually >250 ms) generated by turbulent air travelling through the narrowed airways. It is often considerably more pronounced on expiration, as the negative intrathoracic pressure generated during inspiration helps to maintain airway patency. Polyphonic wheeze consists of several differently pitched sounds produced by airways of different calibres and is the classical finding in acute asthma, although certainly not unique to this diagnosis. A monophonic wheeze suggests a single airway is narrowed, usually by tumour (malignant or benign). Rhonchi are low-pitched wheeze-like sounds produced in the larger airways, and often have a gurgling or rattling quality. Stridor is a loud, high-pitched, continuous inspiratory sound that indicates upper airway obstruction. It should prompt urgent further investigations to establish its cause. Pleural sounds Anything which interrupts the normal smooth gliding action of the parietal pleura over the visceral pleura will create friction between the two surfaces. This can be heard as a pleural rub, a dry rubbing or crunching sound during respiration often likened to the sound made when walking in crisp snow, or the creaking of leather. Pleural inflammation, tumour, pulmonary embolus, and infection can all cause rub which is often heard only in a small part of the chest, is not modified by cough, and can vary hour by hour or day by day throughout the course of disease. Vocal sounds Vocal resonance is the auscultatory equivalent of tactile vocal fremitus, but tends to allow for easier detection of changes, especially when subtle. The significance of changes in vocal resonance parallel those of vocal fremitus, with an increase of consolidation and a reduction when anything is interposed between the lung and the chest wall (air, fluid, solid material). The normal voice has a muffled quality, while being transmitted clearly through consolidated lung. Initial investigations A plain posteroanterior (PA) chest radiograph is an essential adjunct to early diagnostic assessment in the medical admissions unit or specialist chest clinic, but note that practice varies; in rural and poor global settings, for instance, microscopy of sputum for mycobacteria is used efficiently instead of radiology in the detection of endemic tuberculosis. In dealing with any patient who is acutely unwell, pulse oximetry and (in most cases) arterial blood gas analysis (see deceptive clinical features of respiratory failure, earlier) should be performed. A haemoglobin-oxygen saturation lower than 94% signifies clinically significant hypoxaemia. Beware carbon monoxide poisoning with tissue hypoxaemia but normal pulse oximetry. Peak expiratory flow measurement is an essential adjunct in acute asthma. In the clinic, well-conducted but simple spirometry provides an excellent and objective assessment of diverse obstructive and restrictive respiratory disorders, comparing the forced expiratory volume in one second and the forced vital capacity (FEV1 and FVC) to predicted values according to age, gender, height, and ethnic origin. Repeated spirometry can provide the basis for assessing objectively the response to treatments. See Chapter 18.3.1 for further discussion. FURTHER READING Shah PL, et al. (2018). Essentials of Clinical Pulmonology. CRC Press. Von Leupoldt A, et al. (2009). Dyspnea and pain share emotion related brain network. Neuroimage, 48, 200–6. Vyshedskiy A, et al. (2005). Transmission of crackles in patients with interstitial fibrosis, heart failure and pneumonia. Chest, 128, 1468–74.

