Self-Assessment Colour Review

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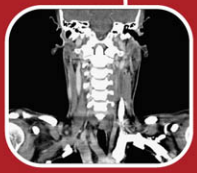
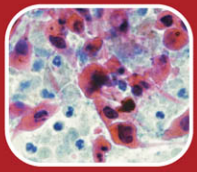
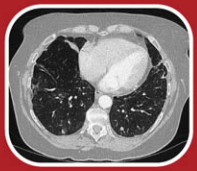
Respiratory Medicine

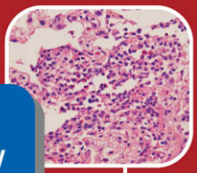
Third Edition

Stephen G. Spiro
Richard K. Albert
Jeremy S. Brown
Neal Navani









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Respiratory Medicine

Third edition

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Contents

Preface

One of the pleasures of respiratory medicine is also its greatest challenge – it has so many diseases within its subspecialty area, as well as encompassing many of the problems seen in acute medical practice, that it requires an extensive knowledge of medicine. Judging from the previous editions of this book, learning through the question and answer format has proven popular and attractive. We have completely rewritten this text and included many new cases to discuss. Those which remain have all been reviewed and updated as necessary. All the authors have an extensive international experience of general internal as well as respiratory medicine, and virtually every case is an actual clinical problem or presentation that they have encountered over the last few years.

The book is, in the same way as respiratory medicine, predominantly based on radiological presentations. There is a huge variety of radiological questions, and the reasons for any particular answer are carefully explained. The same applies to the physiological questions, an area absurdly neglected in much of today's teaching and training modules. In order to maintain the reader's interest, there is no particular sequence to the question list, and the reader can dip in and out, select a topic from the index, or just work through the book. All the main topic areas are covered: radiology, physiology, infection, malignancy, interstitial lung disease, diseases of the pleura, immunology and immunosuppression, respiratory complications of systemic diseases, hereditary conditions, sleep-disordered breathing, asthma, chronic obstructive pulmonary disease, respiratory failure, and problems around the intensive care unit. If you answer all the 200 or so questions and understand their meaning, this book, intended for trainees as well as general physicians, will, we hope, have enhanced your knowledge of respiratory medicine. And if you have enjoyed this style of education as much as we have enjoyed putting the book together, we will have succeeded in our task.

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Classification of cases

All references are to question and answer numbers

Thoracic oncology
6, 19, 22, 31, 34, 39, 41, 55, 66, 86, 96, 97, 99, 104, 107, 117, 128, 132, 133, 136, 139, 147, 149, 152, 153, 154, 157, 169, 176, 177, 178, 189, 191, 199

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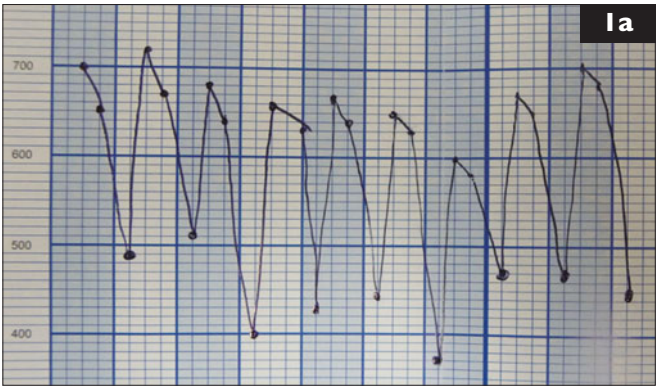
21, 51, 87, 122, 146

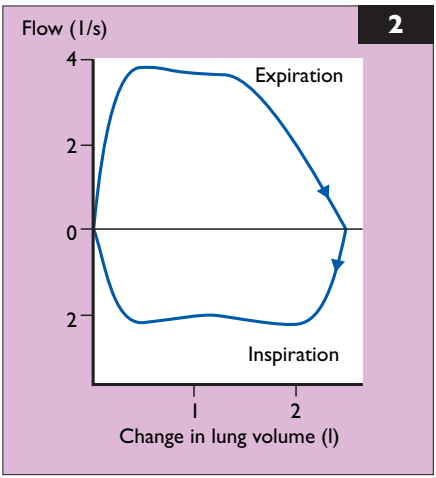
Miscellaneous
9, 10, 32, 35, 40, 50, 61, 65, 88, 118, 123, 138, 166, 172, 175, 187, 195, 196, 198, 201

Abbreviations

ACE	angiotensin-converting enzyme
ADH	antidiuretic hormone
AIDS	acquired immune deficiency syndrome
ANCA	antineutrophil cytoplasmic antibody
ARDS	acute respiratory distress syndrome
BAL	bronchoalveolar lavage
BCG	Bacille Calmette-Guérin
BiPAP	bi-level positive airway pressure
BMI	body mass index
BO	bronchiolitis obliterans
BTS	British Thoracic Society
CAP	community-acquired pneumonia
CFTR	cystic fibrosis transmembrane regulator
CMV	cytomegalovirus
CNS	central nervous system
COPD	chronic obstructive pulmonary disease
CPAP	continuous positive airway pressure
CRP	C-reactive protein
CSF	cerebrospinal fluid
CT	computed tomography
DL_{CO}	diffusing capacity of the lung to carbon monoxide
DPI	dry powder inhaler
EAA	extrinsic allergic alveolitis
EBUS	endobronchial ultrasound
ECG	electrocardiograph
ELISA	enzyme-linked immunosorbent assay
ESR	erythrocyte sedimentation rate
FEV_1	forced expiratory volume in 1s
FiO_2	fraction of inspired oxygen
FRC	functional residual capacity
FVC	forced vital capacity
GINA	Global Initative for Asthma
GOLD	Global Initiative for Chronic Obstructive Lung Disease
HIV	human immunodeficiency virus
HPOA	hypertrophic pulmonary osteoarthropathy
HRCT	high-resolution computed tomography
INR	International Normalized Ratio
K_{CO}	carbon monoxide transfer coefficient
LAM	lymphangioleiomyomatosis
LEMS	Lambert–Eaton myasthenic syndrome
LIP	lymphocytic interstitial pneumonitis
MDI	metered dose inhaler
MRC	Medical Research Council
MRI	magnetic resonance imaging
OSA	obstructive sleep apnoea
$PaCO_2$	arterial partial pressure of carbon dioxide
PaO_2	arterial partial pressure of oxygen
PAS	periodic acid–Schiff
PAVM	pulmonary arteriovenous malformation
PCR	polymerase chain reaction
PEFR	peak expiratory flow rate
PET	positron emission tomography
PiO_2	inspired partial pressure of oxygen
RQ	respiratory exchange ratio (quotient)
RV	residual volume
SaO_2	arterial oxygen saturation
TB	tuberculosis
TLC	total lung capacity
VATS	video-assisted thoracoscopic surgery
VC	vital capacity
VO_2max	maximal oxygen uptake
V/Q	ventilation/perfusion



1 i. What is the investigation shown in **1a**?
ii. How should the patient be managed?



2 This flow–volume loop (**2**) was obtained from a 56-year-old patient with marked respiratory distress and an audible wheeze in whom the initial diagnosis was 'status asthmaticus'. The patient had recently been discharged from hospital after 4 weeks of treatment in intensive care for ARDS.
i. What does the flow–volume loop demonstrate?
ii. What is the likely diagnosis?
iii. What inhalation treatment might be helpful?

1, 2: Answers

1b

REDUCE ⟵ ⟶ INCREASE

TREATMENT STEPS

	Step 1	Step 2	Step 3	Step 4	Step 5
	asthma education				
	environmental control				
	as needed rapid-acting β_2-agonist	as needed rapid-acting β_2-agonist			
CONTROLLER OPTIONS		SELECT ONE	SELECT ONE	ADD ONE OR MORE	ADD ONE OR BOTH
		low-dose ICS*	low-dose ICS *plus* long-acting β_2-agonist	medium- *or* high-dose ICS *plus* long-acting β_2-agonist	oral glucocorticosteroid (lowest dose)
		leukotriene modifier**	medium- *or* high-dose ICS	leukotriene modifier	anti-IgE treatment
			low-dose ICS *plus* leukotriene modifier	sustained-release theophylline	
			low-dose ICS *plus* sustained-release theophylline		

* inhaled glucocorticosteroids ** receptor antagonist or synthesis inhibitors

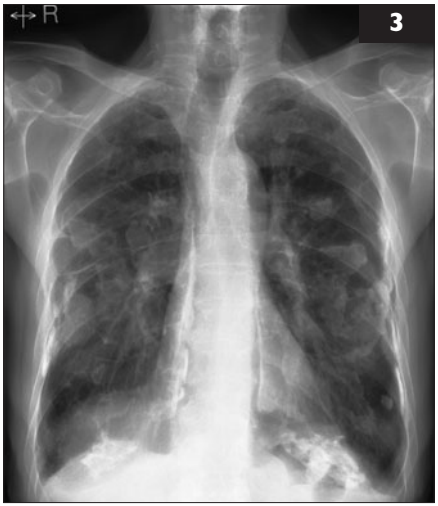
1 i. This is the daily PEFR measurement. Measurements are lowest in the early morning and highest in the afternoon as all individuals show at least a 10% diurnal rhythm for peak flow. In asthma, this is exaggerated, and a more than 20% diurnal variability is consistent with a diagnosis of asthma.
ii. Patients with asthma should be managed according to a stepwise regime (e.g. GINA; **1b**). They should be started at the most appropriate step for their symptoms and moved up when their asthma is uncontrolled and down when it is well controlled.

2 i. The flow–volume loop demonstrates a reduction in the maximum inspiratory and expiratory flows, which both plateau over a large proportion of the FVC manoeuvre and a low FVC <3 l. The reduction in flow is more marked during inspiration, and this is typical of narrowing of the extrathoracic trachea.
ii. The initial diagnosis of asthma should be avoided on the basis of the physical signs, which, in addition to stridor, are most marked in inspiration but often audible during expiration. A simple test to detect upper airway obstruction is the ratio of FEV_1 to peak flow, i.e. FEV_1 (ml)/PEFR (l/min). This is usually less than 10, but in upper airway obstruction the peak flow is affected most, e.g. 2000 ml ÷ 150 l/min = 13.

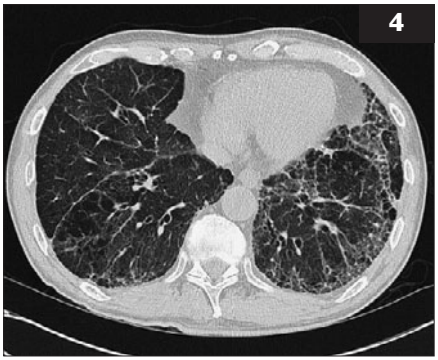
The data would also be compatible with a high tracheal or laryngeal area of narrowing/collapse, as after a tracheostomy.

Another cause of this presentation is a high tracheal tumour, which, because of its slow onset, may mimic asthma. However, the wheeze will be fixed and the symptoms constant, unlike in asthma.
iii. A mixture of oxygen (21%) and helium (79%) as helium is less dense than nitrogen, allowing a greater flow of gas.

3 A 74-year-old male has previously worked as a plumber and has been exposed to asbestos. He has not previously had any respiratory problems and now has a chest X-ray (**3**) before an elective hip replacement.
i. What does it show?
ii. What further action is required?
iii. How else can asbestos exposure affect the lungs?



4 This 65-year-old male has a 4-month history of a dry cough and exertional breathlessness.
i. Describe the appearances on HRCT (**4**).
ii. What clinical features might you expect to find on examination?
iii. What are the associations and complications of this condition?
iv. What is the treatment?



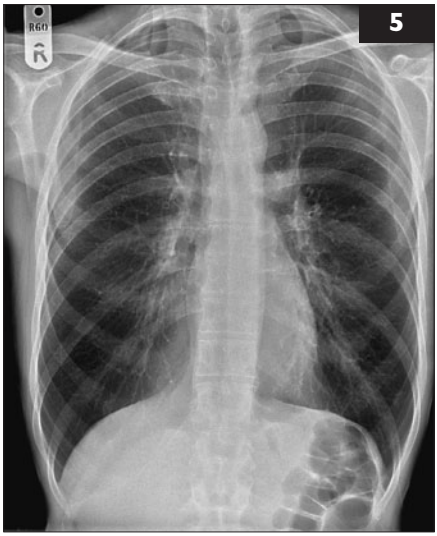
3, 4: Answers

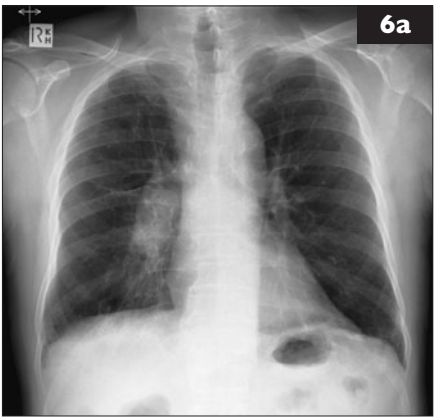
3 i. The chest X-ray demonstrates extensive pleural plaques. These are evident both above the hemi-diaphragms and also along the ribs throughout the thorax.
ii. This is a benign condition and rarely causes symptoms. The plaques occur after a latent period of approximately 20–40 years following asbestos exposure and almost exclusively involve the parietal pleura. Plaques may grow over time, but as they are not considered premalignant no further action is required.
iii. Asbestos may affect the lung in several ways (*Box*).

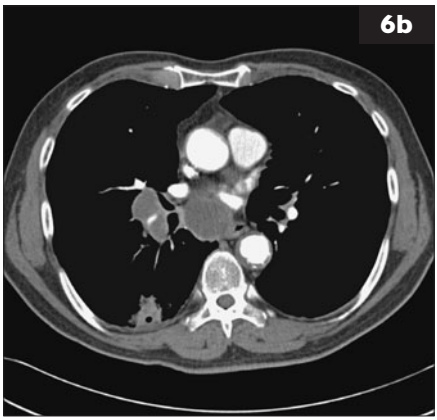
Asbestos-related pulmonary manifestations
- Pleural plaques
- Benign pleural effusion
- Diffuse pleural thickening
- Rounded atelectasis (Blesovsky sign)
- Asbestosis (pulmonary fibrosis)
- Mesothelioma
- Non-small-cell lung cancer

4 i. The HRCT axial cut demonstrates reticular opacities, traction bronchiectasis, and honeycombing in a subpleural and basal distribution. Honeycombing is recognized by the presence of one or more rows of clustered cysts (<5 mm in diameter) and implies a poor prognosis. The features are characteristic of idiopathic pulmonary fibrosis. HRCT is a very reliable method of making the diagnosis, and lung biopsy (which would demonstrate usual interstitial pneumonitis) is not required in most cases.
ii. The examination features would include digital clubbing (25–50% of cases) and fine end-inspiratory 'Velcro-like' crackles at the bases. The pansystolic murmur of tricuspid regurgitation, a raised jugular venous pressure, and peripheral oedema may herald the development of pulmonary hypertension.
iii. No cause for idiopathic pulmonary fibrosis has been discovered. It may, however, be associated with connective tissue diseases (e.g. rheumatoid arthritis, scleroderma and mixed connective tissue diseases) and in this context has a better prognosis. The condition is linked to an increased incidence of lung cancer and is a cause of respiratory failure and secondary pulmonary hypertension. The 5-year survival is worse than for many extrathoracic cancers, at 20–40%.
iv. The treatment options are limited. A combination of *N*-acetylcysteine, prednisolone, and azathioprine is recommended to reduce the rate of decline of lung function, although it does not improve mortality. Lung transplant is rarely performed for patients with idiopathic pulmonary fibrosis, since most patients present over the age of 60 (the cut-off for transplantation in the UK).

5 i. Describe this chest X-ray (**5**) of a 74-year-old smoker.
ii. How would you judge the severity of the condition?







6 i. How would you stage this patient's lung cancer (**6a, 6b**)?
ii. How reliable is CT for identifying metastatic mediastinal disease?
iii. What is the maximum size of so-called normal nodes on CT scanning?

COPD severity	GOLD criteria (2008) FEV_1 % predicted	National Institute for Health and Clinical Excellence criteria (2004) FEV_1 % predicted
Mild	>80%	50–80%
Moderate	50–80%	30–49%
Severe	30–49%	<30%
Very severe	<30% or <50% with chronic respiratory failure	–

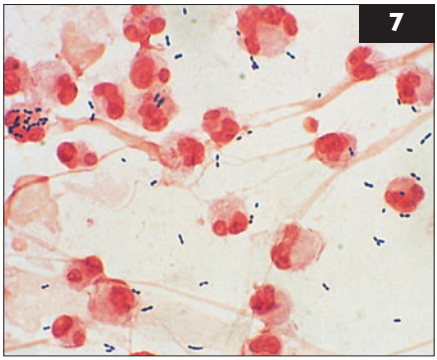
5 i. The posteroanterior chest X-ray shows hyperinflated lung fields, flattening of the diaphragms, a general reduction in lung density, and a peripheral pruning of vascular markings consistent with COPD.
ii. The severity of COPD is most usefully assessed by spirometry. The GOLD (Global Initiative for Chronic Obstructive Lung Disease) guidelines define COPD from when the post-bronchodilator FEV_1/FVC ratio is <0.7 (*Table*).

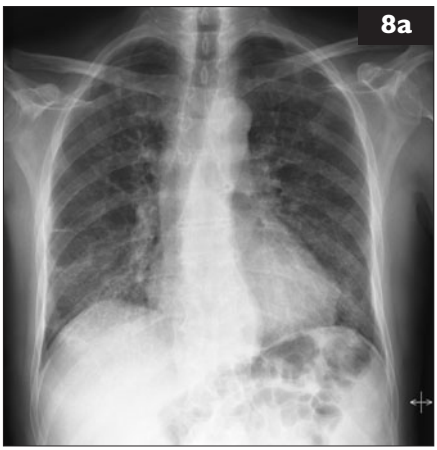
Other factors that predict the prognosis and exacerbation rate include transfer factor for carbon monoxide, breathlessness (according to the MRC scale), exercise capacity, BMI, PaO_2, and cor pulmonale.

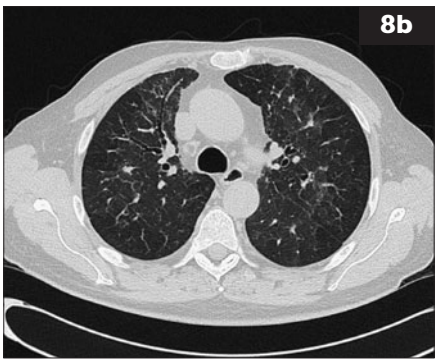
6 i. The chest X-ray (**6a**) shows a large right hilar mass and no obvious primary tumour. The primary tumour may be resectable, but only if there is no mediastinal disease present. A CT of the thorax and upper abdomen should be performed (**6b**). Such a CT scan in fact has shown the primary tumour behind the hilar mass, sitting in the apical segment of the right lower lobe. It is cavitating, suggesting a squamous cell lesion. The CT scan confirms the hilar nodal enlargement and also shows enlarged subcarinal nodes, making the patient almost certainly inoperable. In otherwise operable cases of non-small-cell lung cancer, liver metastases are seen on CT in 5% and adrenal deposits in 7% of patients.
ii. CT scanning gives a 30–40% false-positive and also a false-negative rate, and is therefore much less reliable than a PET scan, which has a specificity of 85–90% for identifying tumour.
iii. The maximum size of 'normal' nodes on a CT scan is 10 mm in the node's short axis, although this will be falsely negative in up to 40% of cases, particularly with adenocarcinomas.

7 The photomicrograph (**7**) shows a Gram stain of sputum expectorated by a 19-year-old male with symptoms of fever, rigors, and a productive cough of 2 days' duration. He has experienced four episodes of radiographically documented pneumonia and numerous episodes of acute otitis media and sinusitis in the past.

i. What does the Gram stain show?
ii. What is the most likely diagnosis?
iii. What additional diagnostic considerations are raised by this clinical presentation?







8 This 50-year-old male presented with gradually progressive dyspnoea over the course of 6 months. His mother had become unwell at that time, and he had started visiting her daily to look after her. Her chest X-ray (**8a**) is shown.

i. Describe the appearances seen on the HRCT (**8b**) of the lungs.
ii. What important history would you obtain?
iii. What is the likely diagnosis?
iv. How would you treat this condition?
v. What is the differential diagnosis of bilateral upper lobe shadowing?

7, 8: Answers

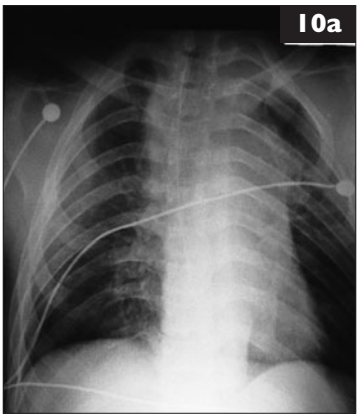
7 i. The sputum Gram stain shows numerous polymorphonuclear leukocytes, strands of mucus, and many Gram-positive, lancet-shaped diplococci. Refractile capsules are evident on many of the bacteria.
ii. The young patient has an acute pneumococcal pneumonia and has suffered numerous recurrent respiratory infections.
iii. Most patients with recurrent respiratory infections have no identifiable host defect. However, recurrent pneumonias in the same lobe or segment should suggest an anatomical abnormality. When multiple sites have been involved, disorders of mucociliary clearance such as cystic fibrosis or the ciliary dyskinesia syndrome (Kartagener's syndrome) are an important consideration.

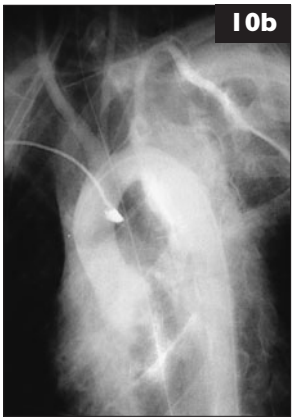
Recurrent infections with encapsulated organisms such as the pneumococcus are consistent with a defect in opsonization (e.g. complement, IgG or IgG subtype deficiencies). Deficiencies of complement components are rare and are best managed by immunization and by the early treatment of infectious complications.

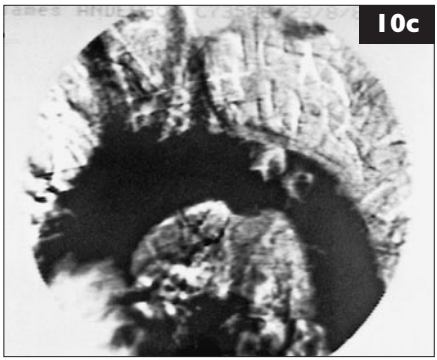
Immunoglobulin deficiencies can also be primary (inherited) or acquired due to acquired B-cell disorders, medications, or intercurrent illnesses. IgA is heavily concentrated in mucosal secretions, where it inhibits bacterial adherence. Although selective IgA deficiency is common, it rarely results in serious respiratory infection unless other deficiencies coexist. IgG deficiency (either generalized or restricted to subclasses IgG1 and IgG3) predisposes to more frequent and more severe respiratory infections. Intravenous immunoglobulin replacement may be helpful.

8 i. The HRCT (**8b**) shows diffuse ground-glass shadowing with some areas of centrilobular nodularity. The chest X-ray (**8a**) shows a mainly upper lobe nodular infiltrate.
ii. Ask about exposure to any pets or unusual dusts or chemicals. In this case, the patient had had a cockatoo that he had been feeding since his mother had become unwell. Budgerigars and pigeons are common causes of this, as are the classical irritants encountered in farmer's lung, bark-stripper's lung, etc.
iii. EAA or hypersensitivity pneumonitis may be caused by exposure to a variety of dusts and antigens, and may be confirmed by avian precipitins in the serum and removal of the source, i.e. the bird. A careful occupational history is essential.
iv. Patients must be instructed to avoid the allergen or precipitant. Oral prednisolone may be used during the initial few weeks until the patient feels better, although there is no definite evidence on optimal dose and duration.
v. The differential diagnosis includes sarcoidosis, pneumoconiosis, ankylosing spondylosis, and TB.

9 i. How does a pulse oximeter work?
ii. The accuracy of most pulse oximeters decreases below what percentage saturation?
iii. What factors affect the accuracy of pulse oximetry?







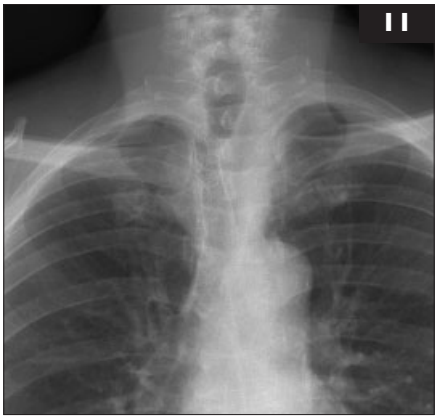
10 A 24-year-old male motorcyclist was admitted as an emergency having been involved in a road traffic accident.
i. What abnormality can be seen on the plain chest radiograph (**10a**), and what diagnosis must be suspected?
ii. What is demonstrated on the subsequent vascular imaging films (**10b, 10c**)?
iii. Where is this lesion normally encountered, what is the mechanism of injury, and what are the possible consequences?

9, 10: Answers

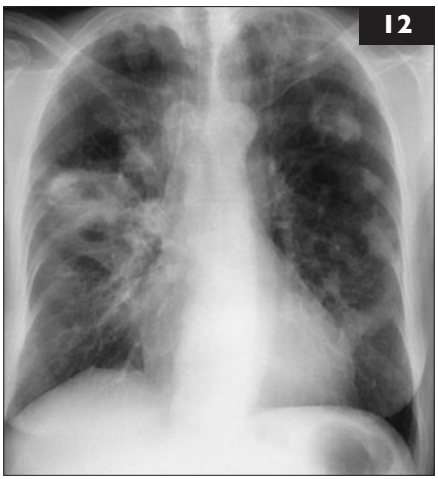
9 i. A pulse oximeter is a spectrophotometer that measures the absorption of light at two wavelengths, one in the infrared range at a wavelength of 940 nm (absorbed by oxyhaemoglobin) and the other at a wavelength of 660 nm (absorbed by reduced haemoglobin).The absorption at these two wavelengths is compared, and the percentages of oxygenated and reduced haemoglobin are calculated. The pulse oximeter compares absorption measured during systole and diastole and uses the difference to reflect only the absorption of arterial blood, thereby limiting errors induced by absorbance in venous blood and other tissues.
ii. The accuracy of most oximeters decreases below 75% saturation. Above this level, the accuracy is approximately 4%.
iii. Standard pulse oximeters are unable to distinguish dyshaemoglobins, most notably carboxyhaemoglobin and methaemoglobin, from oxyhaemoglobin. In smoke inhalation, pulse oximetry reflects absorption by both oxyhaemoglobin and carboxyhaemoglobin, and thus may overestimate the true oxygenation status. Any factor that affects assessment of the arterial pulse (e.g. motion, low-perfusion states) may distort the reading. Specific light sources, such as fluorescent or infrared lamps, can cause erroneous signals if the oximeter probe is not shielded. Dark skin pigmentation and darker coloured nail polishes can also interfere with light transmission, thereby lowering oximeter readings.

10 i. There is gross widening of the mediastinum on the plain chest radiograph (**10a**), even allowing for the anteroposterior nature of the examination. The severity of the injury and the mediastinal widening make traumatic aortic transection a definite possibility.
ii. The aortogram (**10b**) shows a bulge in the region of the distal aortic arch/proximal descending aorta. This is clearly shown on digital subtraction angiography (**10c**) to be due to disruption of the aortic wall.
iii. This lesion is characteristically seen at the aortic isthmus, i.e. the junction between the aortic arch and the descending aorta. This injury is believed to occur because the arch can move forward with deceleration whereas the descending aorta is relatively fixed. Immediate exsanguination will occur in many cases. Those individuals who reach hospital can undergo repair of the ruptured segment, usually with the insertion of a short length of prosthetic graft. Spinal cord ischaemia can occur either before or during surgery, with consequent lower limb paralysis.

11 i. What has been done to this patient whose chest X-ray (**11**) is shown here, and why?
ii. How would the patient have presented?
iii. What are the potential risks of the treatment?



12 This 72-year-old female presented with a 3-month history of malaise, sweats, weight loss, cough, and occasional haemoptysis. She developed a pneumonia that was treated with antibiotics but responded poorly, and she remained unwell with a low-grade fever. She was mildly anaemic with an ESR of 120 mm in 1 hr and a CRP value of 92 mg/l.
i. What does the radiograph (**12**) show, and what is the differential diagnosis?
ii. How would you make the correct diagnosis?
iii. How would you treat the patient?

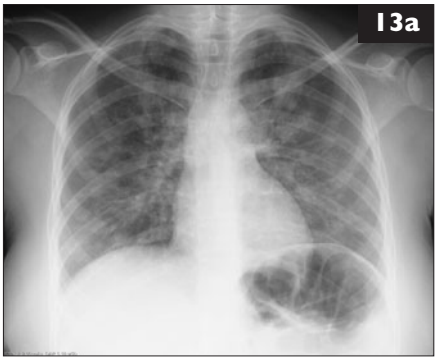


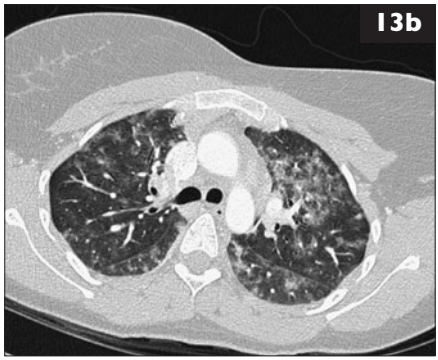
11, 12: Answers

11 i. The image shows an endotracheal stent. Stents are deployed for the management of extrinsic compression of an airway.
ii. The most likely cause of tracheal compression is a retrosternal goitre, and the patient would have presented with stridor. However, this should be managed surgically and not with a stent. Malignant masses or lymph nodes are the most common cause of more distal main airway compression and may need stenting. Although airway stents are an effective initial treatment, definitive management of the underlying lesion should be first choice, for example using surgery (although this is unlikely as the disease is usually advanced by the time major airway occlusion occurs). Radiotherapy and chemotherapy can be effective, especially in small-cell lung cancers. However, in tracheal cancers and squamous cell lung cancers, where the disease often relapses locally after chemotherapy and radiotherapy, stents can dramatically relieve symptoms.
iii. Airway stents also may become a nidus of infection and may become blocked due to secretions, resulting in a sudden deterioration in respiratory status.

12 i. The chest radiograph shows multiple lesions of variable size throughout both lung fields, most of which have cavitated. The differential diagnosis includes staphylococcal pneumonia, lymphoma, TB, cavitating squamous cell carcinoma, and a vasculitis – most likely Wegener's granulomatosis.
ii. This female had Wegener's granulomatosis. The diagnosis was made by identifying cytoplasmic ANCA in high titre in the peripheral blood. This is positive in more than 90% of cases and negative in classic polyarteritis nodosa and other vasculitides. The titres of cytoplasmic ANCA parallel disease activity and return to normal when the disease is in remission. They can predict reactivation if a rise is detected in patients in remission.
iii. Untreated disease is fatal, with a median survival of 5 months. The treatment of Wegener's granulomatosis, whether confined to the lung or more extensive with renal involvement, is with prednisolone and cyclophosphamide. In general, prednisolone is given at 1 mg/kg for 4 weeks and then slowly tailed down to a maintenance of 10 mg/day. Cyclophosphamide is commenced at 100–150 mg orally, and is then reduced by 25 mg every 2–3 months to 50 mg/day. Treatment is continued for 1 year after remission has been achieved.

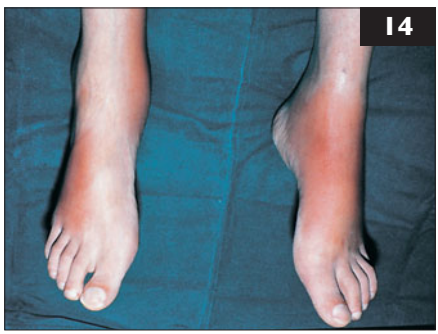
Approximately 75% of patients achieve a complete remission, and a further 15% a partial remission. Serial cytoplasmic ANCA titres should be performed to predict relapse, which can occur in up to 50% of cases. The prognosis is better for Wegener's granulomatosis that is confined to the lung.





13 This previously well 36-year-old East African female has had increasing shortness of breath for 3 weeks associated with a dry cough and some retrosternal chest pain on deep breathing. On examination, she is pyrexial at 37.7°C and cyanosed, with a respiratory rate of 40 breaths/min. Auscultation of the chest reveals some fine bilateral crepitations, and blood and sputum cultures are both negative. Her chest X-ray (**13a**) and CT scan (**13b**) are shown.
i. What is the likely diagnosis?
ii. How can the diagnosis be confirmed, and what is a likely underlying condition?
iii. What is the recommended treatment?

14 Shown here (**14**) is a swollen right ankle and diffuse macular rash in a young female with cystic fibrosis.
i. What is the diagnosis?
ii. What is the medical treatment and prognosis?



13, 14: Answers

13 i. The combination of an insidious but progressive illness with cough, marked hypoxaemia, and a chest radiograph showing bilateral perihilar infiltrates with peripheral sparing (**13a**) (confirmed as mainly upper lobe symmetrical ground-glass infiltrates with peripheral sparing on CT scan (**13b**)) are highly suggestive of *Pneumocystis jirovecii* pneumonia (formerly *Pneumocystis carinii* pneumonia or PCP). In addition, the patient belongs to a high-risk population (sub-Saharan African).

ii. *P. jirovecii* pneumonia is the classical opportunistic pneumonia affecting HIV-positive patients, especially once their lymphocyte CD4-positive count has fallen to less than 200 cells/μl. Presentation is usually a slow onset of dyspnoea associated with an unproductive cough, sometimes associated with a distinctive retrosternal pain on inspiration or coughing. The pyrexia is relatively mild, and chest signs are often limited. Patients characteristically have a fall in oxygen saturations on exercise and a marked reduction in their carbon monoxide transfer factor.

Diagnosis requires obtaining a BAL or induced sputum sample for cytology, in which the characteristic cysts can be identified by cytology using Grocott's methenamine silver staining (which stains the cysts black), or immunofluorescence with specific antibodies. A classical presentation in someone known to be HIV positive can be treated empirically, reserving invasive investigations for if the patient fails to respond. *Pneumocystis jirovecii* pneumonia also occurs in patients with other T-cell defects such as those who have had organ or bone marrow transplantation, or individuals on long-term immuosuppression.

iii. Treatment is with high-dose co-trimoxazole (120 mg/kg in divided doses daily for 3 days, and then 90 mg/kg). If the PaO_2 is <9.3 kPa (70 mmHg) or the alveolar oxygen gradient is >4.7 kPa (33 mmHg), adjuvant corticosteroids have been shown to improve outcome (e.g. 40 mg prednisolone twice a day for 5 days, followed by 40 mg for 5 days and then 20 mg for 11 days). For patients who are intolerant of Septrin (co-trimoxazole), second-line therapy is usually primaquine 15 mg once daily and clindamycin 600 mg four times a day.

14 i. The illustration shows a diffuse vasculitis involving the skin and synovium. Vasculitis of the skin may also be nodular or purpuric (without thrombocytopenia). It is associated with severe pulmonary disease and chronic *Pseudomonas aeruginosa* infection. It has been attributed to an overspill into the systemic circulation of immune complexes resulting from the hyperimmune stimulation associated with chronic pulmonary disease.

ii. Medical treatment consists of short-term, high-dose oral steroids. This will usually produce complete resolution. Immunosuppressive agents have been used, but experience with them is limited.