GET THROUGH

MRCP: PACES

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Editor: Iñaki Bovill

Foreword by: Eric Beck

CRC Press
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Dedication

We would like to dedicate this book to our families, for their unconditional and unprecedented love. We would also like to thank our friends for their loyalty and support.

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CONTENTS

FOREWORD

Assessment drives learning! This was the mantra that underpinned the major restructuring of all parts of the MRCP(UK) examination in the 1990s, resulting in the replacement of the Oral, Long and Short Cases by PACES (Practical Assessment of Clinical Examination Skills). Leading up to this was a lengthy process of learning, observation and consultation involving other evolving medical examinations and the burgeoning new academic discipline of Medical Education, culminating in convening an international conference. I had the privilege of being part of this exciting project.

A syllabus of the knowledge and skills required by a trainee specialist in General (Internal) Medicine was drawn up. This acted as a template to ensure which different parts of the examination would best cover most of the syllabus requirements.

Reproducibility between worldwide examination centres and from one examination to another required standardization of content and marking against pre-agreed criteria by trained examiners whose training would be regularly updated. (A very few traditionalist and individualist examiners fell by the wayside.) Detailed check-list type mark sheets, customized for each station, 'drive' the five PACES stations, recording detailed performance data to give feedback to the unsuccessful candidate. The long-established and tested principles of the undergraduate OSCE (Objective Structured Clinical Examination), pioneered in Dundee, have been adapted to a postgraduate setting.

Needless to say a 'cottage industry' of courses and books has developed to familiarize candidates with the 'new' MRCP and help them prepare for it. The authors of this book are obviously 'battle hardened' and streetwise from their own successful study and preparation. The contents are up to date and certainly have authenticity. There is much practical advice on the knowledge required and how to best present it under examination conditions. They frequently remind the reader/candidate to supplement the book with individual study and of the prerequisite of practising with like-minded colleagues and willing mentors.

I commend it to readers and wish them the final ingredient for success – good luck!

Eric Beck
Former Chairman of the MRCP(UK) Part 2 Board and Former Chairman of the PACES Implementation Committee

ABOUT THE AUTHORS

Dr Rajeev Gulati qualified from university college London in 2000 with a BSc in Psychology. He achieved a Certificate of Merit for outstanding performance as well as Certificate of Merit in Clinical Pharmacology in MBBS written finals. He successfully completed the MRCP in 2003 at his first attempt. He worked as a medical registrar and then completed a year as an emergency registrar in Sydney, Australia. Rajeev subsequently backpacked around Southeast Asia. Thereafter, he moved into general practice and completed the MRCGP exams.

Rajeev currently works as a full time GP partner in North London and has an active role within medical education. He is an FY2 supervisor and is actively involved in teaching medical students and other trainees within the practice. He is currently undertaking the Teaching the Teacher (TTT) course to become a GP trainer. He completed the Certificate in Learning and Teaching (CILT) at QMUL in 2010 and is an Associate Member of the Higher Education Academy. He is an examiner for medical students at Kings College London medical school and has written books for both the MRCP and MRCGP examinations. His interests include running, travel and playing with his baby girl.

Dr Monal Wadhera qualified from Imperial College at St Mary's in 2002 with a BSc in Cardiovascular Medicine. She successfully completed the MRCP in 2005 on her first sitting. She subsequently decided on a career in General Practice. She trained on the West Middlesex Vocational Training Scheme and qualified in 2007, achieving the MRCGP with distinction. She was awarded the 'Great Expectations' Bursary by the Royal College of General Practitioners in 2007.

Monal is a GP Principal in London and has a keen interest in medical education, having completed the MA in Clinical Education at the Institute of Education. She holds posts as GP Tutor with the London Deanery and Clinical Lead in Professional Development at Hammersmith & Fulham Primary Care Trust. She was awarded the National Institute for Health Research In-Practice Fellowship by the Department of Health in 2010. She achieved a distinction in the Certificate in Learning and Teaching at Queen Mary University of London and merit in the Royal College of General Practitioners Leadership Programme. She is an examiner for medical students at Queen Mary University of London and has written previous revision guides for the MRCP and MRCGP examination. She is also a GP appraiser, which she enjoys greatly. Outside medicine, her interests include dining, reading, travelling, writing and yoga.

Dr Iñaki Bovill qualified from Charing Cross and Westminster Medical School in 1993 and subsequently trained as a General Physician Geriatrician in the North West Thames region including at St Mary's Hospital and Chelsea & Westminster

Hospital. He was appointed to the consultant staff at Chelsea & Westminster Hospital in 2004 where he has developed an interest in peri-operative medicine providing support to the General Surgeons and Orthopaedic teams (including pre-operative optimization), Rehabilitation Medicine supporting the Rehabilitation Unit at Ellesmere House and continence. His clinical interests also include all aspects of Elderly and General Medicine but particularly the complex elderly patient and polypharmacy.

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We would like to thank the National Institute for Health and Clinical Excellence for allowing us to use their material. Finally, we would like to thank the Royal Society of Medicine and Sarah Penny, Stephen Clausard and the team at Hodder Arnold for their patience and support throughout the process of preparing this book.

ABBREVIATIONS

ABPA	Allergic bronchopulmonary aspergillosis
ACE	Angiotensin-converting enzyme
ACTH	Adrenocorticotrophic hormone
ADH	Antidiuretic hormone
ADPKD	Adult polycystic kidney disease
A&E	Accident and emergency (department)
AF	Atrial fibrillation
AFP	α-fetoprotein
AIDS	Acquired immune deficiency syndrome
AIH	Autoimmune hepatitis
ALL	Acute lymphocytic leukaemia
ALP	Alkaline phosphatase
ALT	Alanine transaminase
AMA	Anti-mitochondrial antibody
AML	Acute myeloid leukaemia
ANA	Antinuclear antibody
AR	Aortic regurgitation
ARB	Angiotensin receptor blocker
ARMD	Age-related macular degeneration
ASD	Atrial septal defect
ASO	Anti-streptolysin O
AST	Aspartate transaminase
BCC	Basal cell carcinoma
BCG	Bacille Calmette–Guérin
b.d.	Bis in die (twice daily)
BIPAP	Bilevel positive airway pressure
BMI	Body mass index
BP	Blood pressure
CCF	Congestive cardiac failure
CF	Cystic fibrosis
CFP	Culture filtrate protein
CFTR	Cystic fibrosis transmembrane regulator
CJD	Creutzfeldt–Jakob disease
CK	Creatine kinase
CLL	Chronic lymphocytic leukaemia
CML	Chronic myeloid leukaemia
CMV	Cytomegalovirus
CN	Cranial nerve
CNS	Central nervous system
COMT	Catechol-O-methyltransferase
COPD	Chronic obstructive pulmonary disease
COX	Cyclo-oxygenase
CPA	Cerebellopontine angle
CPK	Creatine phosphokinase
CREST (syndrome)	Calcinosis, Raynaud's phenomenon, oesophageal dysmotility, sclerodactyly and telangiectasia
CRP	C-reactive protein
CRT	Cardiac resynchronization therapy
CSF	Cerebrospinal fluid
CT	Computed tomography
CVA	Cerebrovascular accident

CXR	Chest radiograph
DMARD	Disease-modifying antirheumatic drug
DNA	Deoxyribonucleic acid
DNR	Do not resuscitate (order)
DPP-4	Dipeptidyl peptidase 4
DVLA	Driver and Vehicle Licensing Agency
DVT	Deep vein thrombosis
EAA	Extrinsic allergic alveolitis
EBV	Epstein–Barr virus
ECG	Electrocardiography
ELISPOT	Enzyme-linked immunosorbent spot assay
EMG	Electromyography
ENT	Ear, nose and throat
ERCP	Endoscopic retrograde chloangiopancreato-graphy
ESAT	Early secretory antigen target
ESR	Erythrocyte sedimentation rate
FBC	Full blood count
FBG	Fasting blood glucose
FEV_1	Forced expiratory volume
FSH	Follicle-stimulating hormone
FVC	Forced vital capacity
FY1, FY2	Foundation year 1, foundation year 2
GGT	γ-glutamyl transpeptidase
GH	Growth hormone
GLP-1	Glucagon-like peptide 1
GN	Glomerulonephritis
GORD	Gastro-oesophageal reflux disease
GTN	Glyceryl trinitrate
GTT	Glucose tolerance test
HGPRT (deficiency)	Hypoxanthine–guanine phosphoribosyltrans-ferase (deficiency)
HIV	Human immunodeficiency virus
HLA	Human leucocyte antigen
HOCM	Hypertrophic obstructive cardiomyopathy
HSP	Henoch – Schonleih purpura
HSV	Herpes simplex virus
HTLV-1	Human T-cell lymphotrophic virus type I
IBD	Inflammatory bowel disease
IBS	Irritable bowel syndrome
ICD	Implantable cardiovertes–defibrillator
ICU	Intensive care unit
Ig	Immunoglobulin
IGF-1	Insulin-like growth factor
IPJ	Interphalangeal joint
IPPV	Intermittent positive pressure ventilation
ITP	Idiopathic thrombocytopaenic purpura
IV	Intravenous
IVC	Inferior vena cava
IVU	Intravenous urography
JCVI	Joint Committee on Vaccination and Immunisation
JVP	Jugular venous pressure
LDH	Lactate dehydrogenase
LDL	Low-density lipoprotein
LFT	Liver function test
LH	Luteinizing hormone
LMN	Lower motor neuron
LP	Lumbar puncture
LTOT	Long-term oxygen therapy
LVEF	Left ventricular ejection fraction
LVF	Left ventricular failure
LVH	Left ventricular hypertrophy

MAOI	Monoamine oxidase inhibitor
MCPJ	Metacarpophalangeal joint
MCV	Mean cell volume
MEN	Multiple endocrine neoplasia
MI	Myocardial infarction
MND	Motor neuron disease
MPTP	1-methyl-4-phenyl-1,2,3,6-tetradydropyridine
MRI	Magnetic resonance imaging
MS	Multiple sclerosis
MSH	Melanocyte-stimulating hormone
MSU	Mid-stream urine
MTPJ	Metatarsophalangeal joint
NICE	National Institute for Health and Clinical Excellence
NSAID	Non-steroidal anti-inflammatory drug
o.d.	Omni in die (every day)
OGTT	Oral glucose tolerance test
Paco_2	Partial pressure of carbon dioxide in arterial blood
Pao_2	Partial pressure of oxygen in arterial blood
PAN	Polyarteritis nodosa
p-ANCA	Perinuclear anti-neutrophil cytoplasmic antibody
PBC	Primary bilary cirrhosis
PD	Parkinson's disease
PDA	Patent ductus arteriosus
PE	Pulmonary embolus
PET	Positron emission tomography
PID	Pelvic inflammatory disease
PIPJ	Proximal interphalangeal joint
PMR	Polymyalgia rheumatica
PND	Paroxysmal nocturnal dyspnoea
PNS	Peripheral nervous system
p.r.n.	Pro re nata (when required)
PRV	Polycythaemia rubra vera
PSA	Prostate-specific antigen
PSC	Primary sclerosing cholangitis
PT	Prothrombin time
PTH	Parathyroid hormone
PUVA	Psoralen combined with ultraviolet A
q.d.s	Quater die sumendus (to be taken four times daily)
RA	Rheumatoid arthritis
RAPD	Relative afferent pupillary defect
RF	Rheumatoid factor
RNA	Ribonucleic acid
RVF	Right ventricular failure
RVH	Right ventricular hypertrophy
SACD	Subacute combined degeneration of the cord
SCC	Squamous cell carcinoma
SIADH	Syndrome of inappropriate anti-diuretic hormone excess
SLE	Systemic lupus erythematosus
SRP	Signal recognition particle
STI	Sexually transmitted infection
SVC	Superior vena cava
SVCO	Superior vena cava obstruction

SVT	Supraventricular tachycardia
TB	Tuberculosis
t.d.s	Ter die sumendum (to be taken three times daily)
TEN	Toxic epidermolysis necrosis
TFT	Thyroid function test
TIA	Transient ischaemic attack
TIBC	Total iron-binding capacity
TIPJ	Terminal inter-phalangeal joint
TIPS	Transjugular intrahepatic portosystemic shunt
TLC	Total lung capacity
TNF–α	Tumour necrosis factor
TOE	Transoesophageal echocardiography
TPHA	*Treponema pallidum* haemagglutination assay
TRH	Thyroid-releasing hormone
TSH	Thyroid-stimulating hormone
U&E	Urea and electrolytes
UMN	Upper motor neurone
USS	Ultrasound scan
UTI	Urinary tract infection
UVB	Ultraviolet B
VATS	Video-assisted thoracoscopic surgery
VDRL	Venereal Disease Research Laboratory
VSD	Ventricular septal defect
VT	Ventricular tachycardia
VTE	Venous thromboembolism
WCC	White cell count
WHO	World Health Organization

INTRODUCTION

We have written this book as an aid to preparation for the MRCP PACES (Practical Assessment of Clinical Examination Skills) examination.

We have structured the book into the format of the exam, such that each chapter represents a station. We have included cases that commonly present in MRCP PACES exams.

We have also included a chapter on examination skills, which outlines how to examine each system.

The cases in this book are laid out as in the exam setting. They begin with scenario information typical of what you will receive in the exam. The structuring of the information provided for each case follows the format you will encounter in the exam. In each case discussion, we have followed a question and answer style to mimic a conversation with the examiner. This format allows you to recreate realistic scenarios for all the stations. This approach is useful in private study and for role-playing with colleagues. This can be used to practise with colleagues or on your own, and we found this a very useful approach to adopt when revising for the PACES exam ourselves.

We have included photographs throughout the book to give a useful overall impression of selected conditions. There is also a chapter of supplementary cases at the back of the book with a collection of photographs, including many on eye and skin conditions. These could arise in a variety of contexts throughout the exam, so they have been placed in a separate chapter where they are easily accessible. They include many conditions that are often used as spot diagnoses and therefore may be used to test yourself in private study or in groups. Alongside each photo we have included useful clinical information structured as possible question and answer scenarios.

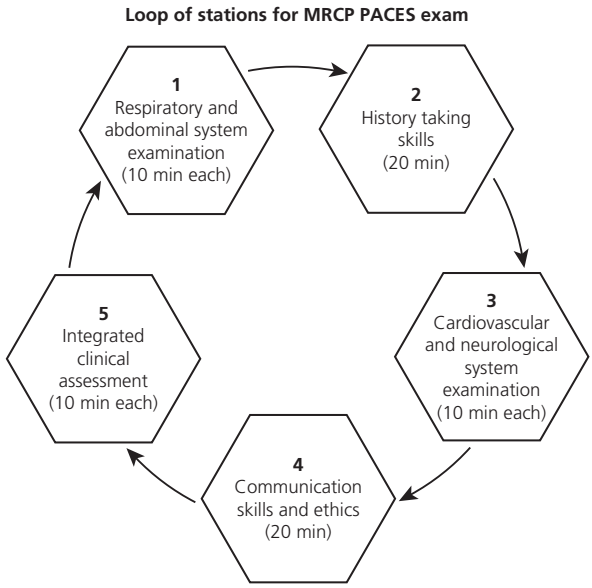
Although we have included a wide breadth of scenarios and discussion points, please remember that the content of this book is not exhaustive, and it is important to read around any topics you come across that are not included here. The further reading recommendations are sources of information that will also be useful in preparing for the exam.

About the MRCP PACES exam and preparation tips

PACES exam format

The MRCP PACES exam is the clinical part of the MRCP(UK) Part 2 exam. It can be taken before or after the MRCP(UK) Part 2 written exam as long as both Part 2 exams are taken within 7 years of passing the MRCP (UK) Part 1 written exam.

The PACES exam is structured into five clinical stations as shown below.



The five clinical stations in the MRCP PACES exam.

Station 1 – Respiratory (10 minutes) and abdominal (10 minutes) system examination
Station 2 – History taking (20 minutes)
Station 3 – Cardiovascular (10 minutes) and neurological (10 minutes) system examination
Station 4 – Communication skills and ethics (20 minutes)
Station 5 – Two brief clinical consultations (10 minutes each)
Each candidate passes through five clinical stations, each of which last 20 minutes each, in a clockwise fashion, and there is a 5-minute break between stations.

Exam details and tips

The PACES exam aims to test the following seven skills:

- Physical examination
- Identifying physical signs
- Communication skills
- Differential diagnosis
- Clinical judgement
- Handling patient concerns
- Ensuring patient welfare

Examination stations (Stations 1 and 3)

The examination stations aim to determine whether candidates are able to perform correct examination techniques, identify physical signs, generate differential diagnoses and suggest appropriate investigation and management

plans. The candidate has 6 minutes for each examination followed by 4 minutes of questions.

Candidates should practise carrying out a comprehensive full examination within the 6 minutes and presenting the physical findings and likely diagnosis. However, depending on the individual examiner, candidates may be interrupted with questions during their examination and may even be asked to skip bits. To avoid this becoming a problem, we would advise practising with your peers and taking the role of different types of examiners to help each other prepare. Always assume a structured examination and do not skip bits yourself unless specifically asked to do so.

History taking and communication skills and ethics stations (Stations 2 and 4)

The history taking station aims to determine whether candidates are able to adequately gather data, generate differential diagnoses, deal with any patient concerns and suggest appropriate investigation and management plans clearly to the patient. The candidate has 14 minutes for history taking, followed by 1 minute for reflection and 5 minutes of questions.

The communication skills and ethics station aims to determine whether candidates are able to clearly explain clinical information and apply their knowledge and ethics to clinical situations. The candidate has 14 minutes for the interview, followed by 1 minute for reflection and 5 minutes of questions. The scenario at this station will often involve a complex situation, which is associated with a difficult ethical dilemma, or sensitive situations.

For Stations 2 and 4, the written scenario is given to the candidate to read during the 5-minute interval. We found that this was a good opportunity to note the main points you wish to cover during the scenario. This acts as an aide-memoire, which will assist you when under pressure.

A good way to approach these stations is to adopt good verbal and non-verbal skills and use a mixture of open and closed questions. Following up on cues is also essential as this may lead you to important information.

Brief clinical consultation station (Station 5)

The brief clinical consultation station aims to determine whether candidates are able to approach a focused clinical scenario, encompassing a targeted history, examination and communication. The candidate has 8 minutes for each scenario followed by 2 minutes to describe the physical signs and differential diagnoses.

Throughout all of the stations, it is important to treat the patient with dignity and respect.

Patients

Each of the clinical stations (1, 3 and 5) will have real patients present, whereas the history taking and communication skills and ethics stations (2 and 4) tend to have actors playing the role of a patient.

Examiners

Each station has two examiners, each marking candidates separately without conferring. There is a different set of examiners for each station, giving a total of 10 examiners for the whole exam.

Marking scheme

There are structured mark sheets for each station (two for each of the examination and brief consultation stations and one for each of the history taking and communications stations). As there are two examiners at each station, there will be a total of 16 marksheets. These are given to candidates at the beginning of the exam for them to fill out their personal details (name, examination number and centre number). The mark sheets are then handed to the relevant examiners at each station.

Candidates are marked as **Satisfactory, Borderline** or **Unsatisfactory** on each of the seven defined skills listed above, which will be variably tested at each station. Each grade reflects a numerical value between 0 and 2 (unsatisfactory = 0; borderline = 1; satisfactory = 2). The MRCP(UK) board sets the pass marks for each of the seven skills and for the whole exam annually in advance. In order to pass the PACES exam, candidates need to achieve *both* the minimum marks for each skill *and* the minimum marks for the whole exam.

Borderline or Unsatisfactory results have to be accompanied by explanatory comments that can then be used to give feedback to failing candidates.

Sample mark sheets for each station and details of the pass marks are available on the MRCP(UK) website (www.mrcpuk.org/PACES/Pages/).

Results

Results are made available on the MRCP(UK) website approximately 10 days after the exam.

In cases of a 'fail', candidates are able to retake the PACES exam an unlimited number of times as long as it is within 7 years of passing the MRCP(UK) Part 1 exam.

Preparation for the exam

Go to www.mrcpuk.org/PACES/Pages/ for information about the PACES exam. The website has up-to-date details about applying for the exam, required conduct at the exam, preparation for the day and getting your results. There are also sample scenarios and mark sheets, which are well worth looking at.

Our overall advice in preparing for the exam is simple: practise, practise, practise!

Practicing on patients

It is best to practise with colleagues or show an interest during ward rounds with your senior colleagues and encourage them to viva you. The scenarios you will see in these circumstances are likely to come up in some form. Practise on patients

until the examination routines are familiar and then read around the subjects using this book.

Familiarizing yourself with the techniques under pressure

Another way to practise is when seeing patients in the accident and emergency (A&E) or outpatient department. This is a good opportunity to take a history or consider the communications and ethics around various scenarios. Discuss cases with your colleagues and read around the subjects as you go along. Remember, unlike the Part I and Part II written exams, this is a practical exam.

Exam conditions

Try to recreate exam conditions by working in pairs or small groups in which each person takes it in turns to be examined or carry out the role of examiner. Be strict with time limits and follow the marking schemes for feedback.

PACES courses

We would recommend doing a PACES course. Besides putting yourself into the exam situation, it also provides the opportunity to see patients with less common clinical conditions that you may not have seen on the wards.

Location

PACES exams are carried out at various centres around the country. Often candidates have to travel long distances and stay overnight. It is important to plan ahead, ensure that you know where the exam is and, if possible, rehearse the walk to the hospital. Hospital departments can be difficult to find at the best of times.

On the day

Ensure that you set out to arrive early in case there are delays. Examiners can pick up on a candidate who is stressed and tired. It is therefore important to arrive at the exam well rested and as relaxed as possible.

PART I

PRACTICE EXAM STATIONS

1 STATION 1 RESPIRATORY AND ABDOMINAL SYSTEM

RESPIRATORY SYSTEM

Pulmonary fibrosis

Describe your clinical findings

On examination, this patient is short of breath at rest with a respiratory rate of 18 breaths/minute. She has rheumatoid arthritis of the hands, as evidenced by wasting of the small muscles of the hands, ulnar deviation and deformity of the fingers. Her fingers are also clubbed and she is cyanosed. There are several purpura on the arms suggestive of steroid use. Chest expansion is reduced. On auscultation of the chest there are bilateral, fine inspiratory crackles at both lung bases.

These findings suggest a diagnosis of pulmonary lung fibrosis. In view of the rheumatoid hands, the likely underlying diagnosis is rheumatoid disease.

TOP TIPS

- Look for other signs of autoimmune disease, rash such as butterfly rash of SLE or changes in hands and face associated with systemic sclerosis.
- Look for cushingoid features arising from the use of steroids.

What do you understand by the term pulmonary fibrosis?

This condition occurs when there is abnormal and excessive deposition of fibrotic tissue in the lung parenchyma, resulting in impaired gas transfer.

What are the causes of interstitial lung disease?

- Cryptogenic fibrosing alveolitis
- Connective tissue disease:
  - rheumatoid lung disease
  - systemic sclerosis
  - SLE
  - polymyositis
  - dermatomyositis

  - Sjögren's syndrome
  - mixed connective tissue disease
- Ankylosing spondylitis
- Sarcoidosis
- Asbestosis
- Silicosis
- Extrinsic allergic alveolitis
- Radiation
- Drugs such as bleomycin, nitrofurantoin and amiodarone

What are the causes of upper lobe fibrosis?

Remember: BREADTHS

- **B**eryliosis
- **R**adiation, when the upper lobe is not shielded
- **E**xtrinsic allergic alveolitis
- **A**nkylosing spondylosis
- **A**llergic bronchopulmonary aspergillosis
- **D**rugs
- **T**uberculosis
- **H**istiocytosis X
- **S**ilicosis
- **S**arcoidosis

What are the causes of lower lobe fibrosis?

Remember: SCRAD

- **S**cleroderma
- **C**ryptogenic fibrosing alveolitis
- **R**heumatoid arthritis
- **R**adiation, when the lower lobe is not shielded
- **A**sbestosis
- **D**rugs

With what symptoms and signs do patients with pulmonary fibrosis present?

- Shortness of breath
- Non-productive cough
- Clubbing
- Malaise
- Weight loss
- Cyanosis
- Fine bilateral inspiratory crackles
- Signs of associated disease

What drugs are linked to pulmonary fibrosis?

- Methotrexate
- Cyclophosphamide

- Amiodarone
- Gold
- Nitrofurantoin
- Sulfonamides

How would you investigate this patient?

Blood tests

- ESR – raised
- RF and ANA – can be positive
- Hypergammaglobulinaemia
- Arterial blood gases – to assess the degree of hypoxia

Imaging

- CXR shows bilateral basal reticulonodular changes.
- High-resolution CT scan shows 'honeycombing'.

Other tests

- Lung function tests:
  - restrictive pattern: $FEV_1/FVC > 0.8$
  - reduced diffusion capacity (K_{co})
- Broncho-alveolar lavage:
  - lymphocytes suggest a good prognosis and good response to steroids
  - neutrophils suggest a poor prognosis and poor response to steroids
- Lung biopsy may be needed

How would you manage this condition?

- Smoking cessation if the patient is a smoker
- Avoid environmental causes
- Stop any medications thought to cause pulmonary fibrosis
- Provide the patient with influenza and pneumococcal vaccination
- Immunosuppressant drugs such as steroids, cyclophosphamide and azathioprine
- Lung transplantation if disease progresses and the patient is fit for surgery
- Long-term oxygen therapy for patients with significant hypoxia

What are the pulmonary manifestations of rheumatoid disease?

- Interstitial lung disease
- Pleural effusion
- Pulmonary arteritis
- Pulmonary nodules
- Obliterative bronchiolitis

What tests on pleural fluid in a patient with a pleural effusion suggest rheumatoid disease?

- RF +ve
- Exudate (high in protein)

- Raised LDL
- Low glucose
- Reduced C3 and C4
- WCC < 5000/μL

What are the complications of pulmonary nodules in patients with rheumatoid disease?

- The nodules have a predilection for the upper lobes
- They often resemble cancer and tuberculosis
- Complications include:
  - haemoptysis
  - pleural effusion
  - bronchopulmonary fistulas
  - can become infected and cavitate
  - can rupture into the pleural space leading to a pneumothorax

What do you know about Caplan's syndrome?

- This is the combination of pulmonary nodules in rheumatoid disease and coal worker's pneumoconiosis (due to mining dust).
- Lung function tests show a mixed restrictive and obstructive picture.
- Patients are treated with steroids.

What do you know about extrinsic allergic alveolitis (EAA)?

- In this condition, there is a hypersensitivity reaction affecting the lung parenchyma causing diffuse, granulomatous inflammation in response to repeated inhalation of organic antigens in dust.
- Patients can present acutely, sub-acutely or with chronic respiratory problems.
- Acute presentations are usually due to Type III (immune complex-mediated) hypersensitivity reactions causing a pneumonitis.
- Chronic presentations are usually due to Type IV (cell mediated) hypersensitivity reactions and lead to fibrosis.
- Several antigens are responsible but the provoking antigen in an individual can be difficult to identify.
- EAA can be associated with many occupations and hobbies as seen in the table below.

Table 1 Classification and causes of EAA

Name	Antigen
Bird fancier's lung	Avian proteins in bird feathers and faeces
Farmer's lung	Mouldy hay containing *Saaccharopolyspora rectivirgula*
Cheese-worker's lung	Mouldy cheese containing *Penicillum casei*
Malt-worker's lung	Mouldy malt containing *Aspergillus clavatus*
Mushroom-worker's lung	Mushroom compost containing thermophilic actinomycetes
Chemical-worker's lung	Trimellitic anhydride, diisocyanate and methylene diisocyanate found in plastics, polyurethane foam and rubber manufacturing
Hot-tub lung	*Mycobacterium avium* found in poorly maintained hot tubs

How do patients with EAA present?

(a) Acute presentation

- Symptoms start 4–8 hours after exposure to the antigen and resolve within days.
- The duration and severity of symptoms are related to the level of exposure. Low-level acute exposure may produce mild symptoms for only a few hours. In very severe cases, patients may develop life-threatening respiratory failure with cyanosis, respiratory distress and high fever.
- Symptoms include:
  - flu-like illness with fever
  - dry cough
  - shortness of breath
  - chest tightness
  - anorexia
  - malaise
  - headache
  - generalized aches and pains
- Signs include:
  - fever
  - tachypnoea
  - bilateral basal fine inspiratory crackles.

(b) Sub-acute presentation

- Patients who present sub-acutely may have a history of repeated acute attacks.
- Symptoms tend to be gradual and less severe, although severe life-threatening episodes can occur.
- After the exposure is removed, it can take weeks or months for symptoms to resolve.
- Symptoms include:
  - cough
  - shortness of breath
  - fatigue
  - anorexia
  - weight loss
  - recurrent pneumonia
- Signs are as for acute presentation.

(c) Chronic presentation

- Patients with chronic disease usually have permanent lung damage. Removal of the antigen may have minimal improvement on symptoms.
- Symptoms include:
  - weight loss
  - worsening shortness of breath
- Signs include:
  - cyanosis
  - clubbing

  - tachypnoea
  - bilateral basal inspiratory crackles
- Chronic hypoxia leads to pulmonary hypertension with right heart failure.

What blood test would you request, in addition to those listed above for pulmonary fibrosis?

- Serum antibodies or precipitans may be detectable e.g. IgG antibody to pigeon gammaglobulin but many have no detectable antibodies.
- Inflammatory makers may be raised but this is non-specific.

What other tests may you undertake?

- CXR: can be normal or show reticular opacities in upper fields in the acute or sub-acute forms. Upper lung fibrosis with loss of lung volume is seen in chronic disease.
- High resolution CT.
- Lung function tests reveal a restrictive defect.
- Bronchoalveolar lavage reveals lymphocytosis and the CD4/CD8 ratio is <1.
- Lung biopsy may be needed and shows the characteristic histopathological features.

How do you manage patients with EAA?

- Avoidance of allergen exposure is important in patients with acute and chronic EAA. Allergen avoidance usually results in recovery for patients presenting with acute EAA. This nlay require a change in occupation.
- LTOT to treat hypoxaemia.
- Corticosteroids may be indicated for severe acute and subacute presentations as well as for chronic forms. However, steriods do not alter the long-term outcome.

What do you know about asbestos and lung disease?

- Asbestos exposure can affect people working in ship building, lagging, construction workers and those working in factories in which asbestos products are manufactured.
- Asbestos exposure can cause a wide range of lung disease such as:
  - pleural plaques
  - diffuse pleural thickening
  - pleural effusions
  - asbestosis
  - increased risk of lung cancer and mesothelioma

Asbestos and the lung

Pleural plaques

- Appear ~20 years after asbestos exposure.
- Usually asymptomatic but can cause mild restrictive lung disease.
- Can develop on the pleura, diaphragm, mediastinum and pericardium.

Diffuse pleural thickening

- Mainly affects the lung bases.
- Causes exertional dyspnoea.
- Lung function tests show restrictive lung disease with reduced total lung capacity.

Asbestosis

- Appears ~20 years after asbestos exposure.
- Fibrosis occurs mainly in the lower lobes.
- Patients can present with exertional dyspnoea.
- Examination reveals clubbing and fine inspiratory crackles.
- Lung function tests show restrictive lung disease and reduced gas transfer.
- There is an increased risk of lung cancer and mesothelioma.
- Patients are entitled to industrial compensation.

Mesothelioma

- Develops from mesothelial cells in the pleura or, less commonly, the peritoneum.
- Asbestos is responsible for the vast majority of malignant mesotheliomas.
- Crocidolite (blue asbestos) is the most toxic, followed by amosite (brown asbestos) and then cryosolite (white asbestos) which is the least toxic.
- There is a latent period, usually ~30 or more years, between asbestos exposure and mesothelioma development.
- Patients can present with chest pain, shortness of breath, dry cough or weight loss.
- Diagnosis is made by pleural biopsy.
- Treatment is with surgery and radiotherapy.
- Patients are eligible for industrial compensation.

Lung cancer

- The risk of lung cancer is increased 5-fold.
- The combination of asbestos and smoking increases the risk of cancer 55-fold.

Name some other occupational lung diseases

Table 2 Occupational Lung Diseases

Name	Caused by Exposure to
Silicosis	Silicon dioxide
Berylliosis	Beryllium
Byssinosis	Cotton dust, flax and hemp
Coal worker's pneumoconiosis	Mining dust

Bronchiectasis

Describe your clinical findings

On examination, this patient is cachectic and tachypnoeic, with a respiratory rate of 18 breaths/minute. He has bilateral clubbing of the fingers and cyanosis. The sputum pot contains copious amounts of green-coloured sputum. On auscultation of the chest, there are left (or right) crepitations in the lower zones and widespread wheeze.

These findings suggest a diagnosis of bronchiectasis.

What is bronchiectasis?

This is a chronic infection of the bronchi and bronchioles leading to abnormal, permanent dilation of the airways.

What are the causes of bronchiectasis?

- Bronchial obstruction due to bronchial tumour, external compression from lymphadenopathy or inhalation of a foreign body
- Respiratory infection in childhood (measles, pertussis and tuberculosis)
- Cystic fibrosis
- Marfan's syndrome
- Kartagener's syndrome
- Hypogammaglobulinaemia
- Allergic bronchopulmonary aspergillosis (ABPA)

How do you investigate patients with bronchiectasis?

Tests to confirm bronchiectasis

- Sputum culture and cytology
- Chest radiograph (CXR) shows tramlines and ring shadows (thickened bronchial walls)
- High-resolution computed tomography (CT) scan shows thickened, dilated bronchi that are larger than the adjacent vascular bundles ('signet' sign)

Tests to determine the underlying cause

- Bloods to check immunoglobulin levels and detect hypogammaglobulinaemia
- Skin prick test for *Aspergillus* to check for aspergillosis precipitins
- Sweat test to detect cystic fibrosis (CF)
- Electron microscopy of cilia from nasal or tracheal mucosa to check for Kartagener's syndrome
- Bronchoscopy to exclude malignancy or foreign body

What are the common pathogens that cause infections in bronchiectasis?

- *Haemophilus influenzae*
- *Staphylococcus aureus*
- *Pseudomonas aeruginosa*

How do you manage patients with bronchiectasis?

- Physiotherapy to aid postural drainage
- Antibiotics for short-term exacerbations; long-term antibiotics may be needed

- Bronchodilators to help airflow obstruction
- Inhaled or oral steroids may be helpful in certain cases
- Surgery with lobe resection can be curative in patients with localized disease

List some complications of bronchiectasis

- Pneumonia
- Empyema
- Pleural effusion
- Cor pulmonale
- Haemoptysis
- Amyloidosis
- Cerebral abscess due to haematogenous spread of infection

What are the features of Kartagener's syndrome?

- Kartagener's syndrome is an autosomal recessive condition where the primary defect is in the structure and function of cilia. Features include:
  - bronchiectasis
  - dextrocardia
  - situs invertus
  - infertility
  - sinusitis
  - otitis media
  - dysplasia of the frontal sinuses

Cystic fibrosis

Describe your clinical findings

On examination this young patient appears underweight. He is tachypnoeic with a respiratory rate of 18 breaths/minute. He has bilateral clubbing of his fingers and a productive cough. The sputum pot contains a large amount of green sputum. Chest expansion is reduced. On auscultation of the chest, there are crepitations over the lower zones of the left (± right) lung. There is a widespread polyphonic expiratory wheeze.

These findings suggest the patient has bronchiectasis. In a young patient, the likely diagnosis is CF.

What do you know about CF?

- Autosomal recessive disorder affecting 1 in 2500 Caucasians.
- 1 in 20 are carriers.
- Most cases are due to a mutation at position 508 on chromosome 7, which results in a deletion of phenylalanine. The chloride channel, CF transmembrane regulator (CFTR), is defective and chloride secretion from cells is therefore reduced.
- As a result, exocrine secretions have a high concentration of sodium and a low concentration of chloride and water. The secretions are thickened and viscid and so block the lumens of ducts and organs, such as the gut.

What are the clinical features of the disease?

Respiratory

- Nasal polyps
- Haemoptysis
- Recurrent chest infections
- Bronchiectasis
- Sinusitis
- Respiratory failure
- Pneumothorax
- Pulmonary fibrosis
- Cor pulmonale
- ABPA

Gastrointestinal

- Rectal prolapse
- Meconium ileus
- Steatorrhoea
- Pancreatitis
- Gallstones
- Biliary cirrhosis
- Liver cirrhosis and portal hypertension resulting from biliary strictures

Other

- Arthropathy
- Diabetes
- Infertility in males
- Sub-fertility in females
- Failure to thrive

What investigations would you perform?

- Heel prick test at birth showing raised level of immunoreactive trypsin
- Sweat sodium test shows sodium > 60 mmol/L
- Genetic testing

Name some pathogens that cause acute exacerbations in patients with CF

- *Staphylococcus aureus*
- *Haemophilus influenzae*
- *Pseudomonas aeruginosa*
- *Burkholderia cepacia*

How do you manage patients with CF?

- Physiotherapy to aid postural drainage and breathing techniques
- Antibiotics – can be oral, nebulized or IV
- Bronchodilators for symptomatic relief

- Pancreatic supplements to reduce the risk of malabsorption
- Recombinant human DNAse, which degrades DNA in the bronchial secretions
- Immunizations for measles, influenza and pneumococcus
- Heart and lung transplant
- Gene therapy
- Genetic counselling for couples

Pleural effusion

Describe your clinical findings

On examination this patient is comfortable at rest. His respiratory rate is 12 breaths/minute. Chest expansion is reduced on the left side and the percussion note is stony dull at the left base. On auscultation of the chest, vocal resonance and breath sounds are reduced. There is an area of bronchial breathing above the area of the dullness.

These findings are suggestive of a pleural effusion.

What are the other cases of dullness at a lung base?

- Pleural effusion
- Collapse
- Consolidation
- Pleural thickening
- Raised hemi-diaphragm (can occur with hepatomegaly)

What are the causes of pleural effusion?

Exudates (protein > 30 g/L)	Transudates (protein < 30 g/L)
● Lung malignancy (primary or secondary)	● Cardiac failure
● Mesothelioma	● Nephrotic syndrome
● Pneumonia	● Liver cirrhosis
● Tuberculosis	● Meigs' syndrome
● Pulmonary embolus	● Hypothyroidism
● Rheumatoid arthritis	
● SLE	
● Lymphoma	

What is Meigs' syndrome?

- This is a right-sided pleural effusion due to an ovarian fibroma.

What drugs can cause a pleural effusion?

- Methysergide
- Methotrexate
- Nitrofurantoin

How would you investigate this patient?

- Pleurocentesis
- Pleural biopsy
- CXR (anterior–posterior and lateral)
- CT scan of thorax to confirm pleural effusion
- Bronchoscopy if malignancy considered

What tests would you do on the pleural fluid?

Biochemistry

- Protein concentration: > 30 g/L suggests an exudate and < 30 g/L suggests a transudate.
- LDH is raised in empyema, malignancy, TB, RA and SLE.
- Amylase is raised in pancreatitis, malignancy and pneumonia.
- Glucose is reduced in empyema, malignancy, TB, RA and SLE.
- RF or ANA may be positive in pleural fluid in autoimmune conditions.

Microbiology and culture

- Gram or Ziehl–Neelsen stain

Cytology

- Malignant cells may be present in neoplastic conditions.
- Red blood cells suggest malignancy, pulmonary infarction or TB.
- Neutrophils are raised in effusions arising from pneumonia.
- Lymphocytes are raised in infection, malignancy, autoimmune conditions and TB.
- Eosinophils are raised in pneumothorax and asbestos-related effusions.

What are Light's criteria?

An exudate is considered present when:

- The ratio of pleural fluid albumin to the plasma albumin > 0.5.
- The ratio of pleural fluid LDH to the plasma LDH > 0.6.
- The pleural fluid LDH is > two-thirds of the normal level of plasma LDH.

Pneumonectomy and lobectomy

Describe your clinical findings

Pneumonectomy

On examination, this patient is comfortable at rest. There is a thoracotomy scar on the left chest wall, which is flattened. The trachea and apex beat are deviated to the left side. Chest expansion is reduced on the left (affected) side and the percussion note is dull. On auscultation of the chest, the breath sounds are reduced.

These findings suggest a left-sided pneumonectomy.

On examination this patient is comfortable at rest. There is a thoracotomy scar on the left chest wall, which is flattened. The trachea is central and the apex beat is not displaced. Chest expansion is normal but the percussion note is dull in the left lower zone. On auscultation of the chest, the breath sounds are reduced.

These findings suggest a left-sided lobectomy.

What are the indications for pneumonectomy or lobectomy?

- Carcinoma
- Pulmonary nodules
- Bronchiectasis
- TB – before chemotherapy was available

Chronic obstructive pulmonary disease (COPD)

Describe your clinical findings

On examination this patient is short of breath at rest (may have oxygen cylinder) with a respiratory rate of 18 breaths/minute. The fingers have nicotine stains, but there is no carbon dioxide retention flap of the hands. He is cyanosed and his lips are pursing during expiration. He has a plethoric appearance and is using his accessory muscles of respiration. The chest is hyperinflated and chest expansion is normal. There is a tracheal tug. The percussion note is resonant. On auscultation of the chest, the breath sounds are reduced and have a prolonged expiratory phase. There is widespread expiratory wheeze.

These findings suggest a diagnosis of COPD due to cigarette smoking.

What is COPD?

- COPD is a term encompassing the spectrum of disease from chronic bronchitis to emphysema. It is a chronic, progressive disease.
- Chronic bronchitis is a clinical diagnosis. It is defined as a cough productive of sputum on most days for more than 3 months of 2 consecutive years in the absence of other diseases causing sputum production.
- Emphysema is a pathological diagnosis. There is permanent enlargement of the airway distal to the respiratory bronchioles with the destruction of alveolar walls.

What are the causes of emphysema?

- Smoking is the commonest cause.
- Industrial dust exposure (coal dust in mine workers or cadmium exposure) can cause emphysema.
- α1-antitrypsin deficiency is rare but should be considered in a young patient who does not smoke.