



# The MRCP PACES Handbook

Second Edition

Saira Ghafur, Parminder K. Judge,  
Richard Kitchen, Samuel Blows

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# Foreword

Postgraduate medical exams are significant both as career-defining milestones for the candidate and as one of the ways of ensuring that doctors who progress have all that it takes to provide excellent patient care. Exams focus the minds of candidates for whom exam content and exam approach define what examiners consider important and so perhaps mould approaches to practise. The Practical Assessment of Clinical Examination Skills (PACES), the final part of the Membership of the Royal Colleges of Physicians (MRCP) examination, is a pivotal step for all aspiring physicians, and this book will be an enormous support for them. But, the *MRCP PACES Handbook* is so much more than a book for passing exams. A comprehensive book, it succinctly covers much of the necessary content, and for that it will be useful to candidates. Responses and analyses to the questions posed are logical and practical and cleverly proceed from the likely and frequent to the rarefied. It is also very well organised, demonstrating not just how to pass an exam, but also the importance of an ordered approach to clinical reasoning. Lists of key points at the ends of chapters keep the reader on track, and the well-chosen lists of references really do encourage further reading. The real strength of this handbook is that it gets to the heart of the purpose of the PACES examination.

The PACES exam reflects the work done through MRCP UK to develop assessments of all the skills necessary for excellent clinical practice. Of course, this includes physical examination, recognition of clinical signs and differential diagnosis, the stuff of medical examinations for many years. Now, assessments have become more patient centred and more holistic and are designed to demonstrate explicitly the expectation of modern physicians and good medical practice. Clinical judgement, communication, managing patient concerns and maintaining patient welfare are four of the seven skills assessed through the PACES exam, and these skills are very well covered in this handbook.

Not only are technical points covered, but by including, for example, outlines of themes explored, suggestions for candidates and expectations for both candidates and patients, this handbook provides a succinct but in-depth outline of some of the difficult but crucial aspects of medical practice.

The simplicity of approach and the clear language throughout make it clear that this is not a book for the 'superficial learner' wanting a 'quick fix' for an impending exam, but one that encourages understanding and a mature approach to developing all the skills necessary for becoming a good physician.

PACES, the final part of the MRCP exam, is what it says: practical. And reflecting the essence of the PACES exam, this handbook is a supremely practical 'vade mecum'. It is a handbook that not only will be essential for those approaching their PACES exam or those considering becoming a physician, but also will be a useful and continuing guide to 'practise after PACES' for those entering higher training after PACES, and it will be useful for anyone who teaches postgraduate or undergraduate medicine.

This is a book that will be bought and used to help aspiring physicians through their PACES, will encourage development of a patient-centred approach to care and then, for many years after, will stay in easy reach.

**Dr Fiona Moss, CBE, MD, FRC**  
**Dean of the Royal Society of Medicine**



# Preface

Observe, record, tabulate, and communicate. Use your five senses. Learn to see, learn to hear, learn to feel, learn to smell, and know that by practice alone you can become an expert.

**Sir William Osler, FRS, FRCP**

The Practical Assessment of Clinical Examination Skills (PACES) should be seen as the defining moment of your medical training when you have been accepted into the fold – the final hurdle you need to jump over before gaining the elusive title of ‘Medical Registrar’. The wise words from Sir William Osler sum up the skills needed to achieve this goal.

This book is designed to be a comprehensive revision aid that will be invaluable to candidates studying for their PACES examination. The book is designed to be used at the patient’s bedside to guide clinical examination, as well as having enough detail to cover pertinent points in the case in question.

Each of the chapters contains hints and tips on how to tackle each of the examinations and a summary at the end to consolidate key learning points. The cases represent a collection of those which are most frequently encountered in the PACES examination and are set out in a standardised format. Each chapter has been reviewed by senior clinicians within the specialty. The information within the handbook has been updated and is supported by evidence-based literature and supporting guidelines.

Passing PACES requires not only an in-depth knowledge of the subject of the case but also the fine art of being able to present in a concise and coherent manner. Each case contains a section on presentation which will aid the candidate in perfecting this skill.

Excellent communication skills are the cornerstone of every doctor’s interaction with patients and colleagues and are essential in passing the PACES examination. Uphold the legacy of Dr Kate Granger, who initiated the #hellomynameis campaign, and treat every patient with the utmost respect, kindness and compassion.

PACES is feared as the unachievable milestone; the key to passing the exam is hard work and sheer dedication. The blood, sweat and tears will all be worth it in the end when you have attained the highly sought after title of Membership of the Royal Colleges of Physicians of the United Kingdom (MRCP UK).

If you follow in the footsteps of those who have gone before you, you are guaranteed to succeed.

Good luck – go forth and conquer!

**SG, PKJ, RK and SB**

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The authors all worked together at the University Hospitals of Leicester as junior doctors, during which time they all passed the PACES examination and developed their interest in medical education and evidenced-based medicine.

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# List of Abbreviations

A2	aortic component of heart sound 2
AA	amyloid A
ABG	arterial blood gas
ABPA	allergic bronchopulmonary aspergillosis
ACE	angiotensin-converting enzyme
ACF	antecubital fossa
ACPA	anticitrullinated protein antibodies
ACTH	adrenocorticotrophic hormone
ADH	antidiuretic hormone
ADLs	activities of daily living
ADPKD	autosomal dominant polycystic kidney disease
AF	atrial fibrillation
AFB	acid-fast bacilli
AFP	alpha-fetoprotein
AKI	acute kidney injury
AL	amyloid L
ALD	adrenoleukodystrophy
ALD	alcoholic liver disease
ALP	alkaline phosphatase
ALS	amyotrophic lateral sclerosis
AMA	antimitochondrial antibody
AMTS	abbreviated mental test score
ANA	antinuclear antibody
APKD	adult polycystic kidney disease
AR	aortic regurgitation
ARB	angiotensin receptor blocker
ARDS	acute respiratory distress syndrome
AS	aortic stenosis
ASA	aminosalicylic acid
ASD	atrial septal defect
AV	arteriovenous
AVA	aortic valve area
AVR	aortic valve replacement
AXR	abdominal X-ray
AVSD	atrioventricular septal defect
BCC	basal cell carcinoma
BP	blood pressure
BTS	British Thoracic Society
CABG	coronary artery bypass graft
CCF	congestive cardiac failure
CDT	<i>Clostridium difficile</i> toxin
CF	cystic fibrosis

CFTR	cystic fibrosis transmembrane conductance regulator
Ch	chromosome
CK	creatine kinase
CKD	chronic kidney disease
CLL	chronic lymphocytic leukaemia
CML	chronic myeloid leukaemia
CMV	cytomegalovirus
CN	cranial nerve
CO	carbon monoxide
COMT	catechol-o-methyltransferase
COPD	chronic obstructive pulmonary disease
COX	cyclo-oxygenase
CRF	chronic renal failure
CRP	C-reactive protein
CSF	cerebrospinal fluid
CT	computerised tomography
CVA	cerebrovascular accident
CXR	chest X-ray
DCCT	Diabetes Control and Complications Trial
DEXA	dual-energy X-ray absorptiometry
DMARD	disease-modifying antirheumatoid drug
DNase	deoxyribonuclease
DVLA	Driver and Vehicle Licensing Agency
EBUS	endobronchial ultrasound
EBV	Epstein–Barr virus
ECG	electrocardiogram
EDM	end-diastolic murmur
EMA	endomysial antibody
EMG	electromyogram
ENT	ear, nose and throat
ESR	erythrocyte sedimentation rate
ESRD	end-stage renal disease
FBC	full blood count
FEV1	forced expiratory volume in first second
FiO <sub>2</sub>	fraction of inspired oxygen
FSH	follicle-stimulating hormone
FVC	forced vital capacity
GBM	glomerular basement membrane
GBS	Guillain–Barré syndrome
GGT	gamma-glutamyl transpeptidase
GH	growth hormone
GI	gastrointestinal
GIM	general internal medicine
GNRH	gonadotrophin-releasing hormone
GP	general practitioner
HCM	hypertrophic cardiomyopathy
HIV	human immunodeficiency virus
HLA	human leucocyte antigen
HPOA	hypertrophic pulmonary osteoarthropathy



IBD	inflammatory bowel disease
IBS	irritable bowel syndrome
ICD	implantable cardioverter defibrillator
ICU	intensive care unit
IE	infective endocarditis
IGF-1	insulin-like growth factor 1
IIH	idiopathic intracranial hypertension
IL	interleukin
IMCA	independent mental capacity advocate
INO	internuclear ophthalmoplegia
INR	international normalised ratio
ITP	idiopathic thrombocytopenic purpura
IV	intravenous
IVDU	intravenous drug user
JAK2	Janus kinase 2
JVP	jugular venous pressure
KCO	carbon monoxide transfer coefficient
LA	left atrium
LABA	long-acting beta-agonist
LAMA	long-acting muscarinic antagonist
LBBS	left bundle branch block
LDH	lactate dehydrogenase
LEMS	Lambert–Eaton myaesthenic syndrome
LFT	liver function test
LH	luteinising hormone
LKM	liver kidney microsomal
LMN	lower motor neurone
LMWH	low-molecular-weight heparin
LP	lumbar puncture
LTOT	long-term oxygen therapy
LUQ	left upper quadrant
LV	left ventricle
LVF	left ventricular failure
LVH	left ventricular hypertrophy
LVOTO	left ventricular outflow tract obstruction
MAOB	monoamine oxidase B
MCPJ	metacarpophalangeal joint
MCV	mean cell volume
MDT	multidisciplinary team
MHC	major histocompatibility complex
MMF	mycophenolate mofetil
MND	motor neurone disease
MR	mitral regurgitation
MRC	Medical Research Council
MRI	magnetic resonance imaging
MS	multiple sclerosis
MVP	mitral valve prolapse
NCS	nerve conduction study
NICE	National Institute for Health and Care Excellence

NIH	National Institutes of Health
NIPPV	noninvasive positive-pressure ventilation
NOAC	novel oral anticoagulant
NSAID	nonsteroidal anti-inflammatory drug
NSCLC	non-small-cell lung cancer
NSTEMI	non-ST elevation myocardial infarction
NYHA	New York Heart Association
OCP	oral contraceptive pill
OD	once daily
OGD	oesophagogastroduodenoscopy
ON	once nightly
OSA	obstructive sleep apnoea
OT	occupational therapist
P2	pulmonary component of heart sound 2
PACES	Practical Assessment of Clinical Examination Skills
PBC	primary biliary cirrhosis
PD	peritoneal dialysis
PDA	patent ductus arteriosus
PEG	percutaneous endoscopic gastrostomy
PET	positron emission tomography
PFT	pulmonary function test
PLS	primary lateral sclerosis
PMA	progressive muscular atrophy
PPI	proton pump inhibitor
PPM	permanent pacemaker
PR	pulmonary regurgitation
PRN	as needed (pro re nata)
PRV	polycythaemia rubra vera
PTH	parathyroid hormone
PUVA	psoralen and ultraviolet A
QOL	quality of life
RA	rheumatoid arthritis
RF	rheumatoid factor
RIF	right iliac fossa
RRT	renal replacement therapy
S1	heart sound 1
S2	heart sound 2
S3	heart sound 3
S4	heart sound 4
SA-AG	serum ascites-albumin gradient
SAA	serum amyloid A
SABA	short-acting beta-agonist
SAH	subarachnoid haemorrhage
SALT	speech and language therapist
SAP	serum amyloid P
SBP	spontaneous bacterial peritonitis
SCC	squamous cell carcinoma
SCD	sudden cardiac death
SCL-70	antitopoisomerase antibodies



SCLC	small-cell lung cancer
SIADH	syndrome of inappropriate antidiuretic hormone secretion
SIGN	Scottish Intercollegiate Guidelines Network
SLE	systemic lupus erythematosus
SOL	space-occupying lesion
SSc	systemic sclerosis
SVCO	superior vena caval obstruction
SVD	structural valve deterioration
TAVI	transcatheter aortic valve implantation
TB	tuberculosis
TBLB	transbronchial lung biopsy
TFT	thyroid function test
TIA	transient ischaemic attack
TLC	total lung capacity
TLCO	transfer factor of the lung for carbon monoxide
TNF	tumour necrosis factor
TOE	transoesophageal echocardiogram
TPO	thyroid peroxidase
TR	tricuspid regurgitation
TRH	thyroid-releasing hormone
TSH	thyroid-stimulating hormone
tTG	tissue transglutaminase
U1-RNP U1	ribonucleoprotein
U&Es	urea and electrolytes
UC	ulcerative colitis
UDCA	ursodeoxycholic acid
UIP	usual interstitial pneumonia
UKPDS	United Kingdom Prospective Diabetes Study
UMN	upper motor neurone
US	ultrasound
USS	ultrasound scan
VATS	video-assisted thoracoscopic surgery
VF	ventricular fibrillation
VP	ventriculoperitoneal
VSD	ventricular septal defect
VT	ventricular tachycardia
VTE	venous thromboembolism
WHO	World Health Organization



# The PACES Examination

The Practical Assessment of Clinical Examination Skills (PACES) consists of five 20-minute stations that make up a carousel. The examination lasts for a total of 125 minutes, which includes a 5-minute break between each of the stations.

Stations 1 (respiratory/abdominal) and 3 (neurology/cardiovascular) are clinical scenarios lasting 10 minutes each. A maximum of 6 minutes is allowed for examination, followed by 4 minutes for discussion. Candidates will be given written instructions prior to examining the patient.

Stations 2 (history-taking skills) and 4 (communication skills and ethics) last 20 minutes each. In these stations, 14 minutes will be allowed for history taking or communicating with the patient. One minute for reflection is provided to consolidate your thoughts prior to a 5-minute discussion with the examiners.

Station 5 (integrated clinical assessment) involves two brief clinical scenarios of 10 minutes each. Eight minutes are allocated for focused history taking, examination and responding to the patient's concerns. The remaining 2 minutes are for discussing examination findings, diagnosis and patient's concerns with the examiners. Candidates can be assessed on a wide range of clinical problems.

There will be two examiners present at each station. Candidates will be assessed in seven key areas. They are not all assessed at every station. The key areas include

1. Physical examination
2. Identifying physical signs
3. Clinical communication
4. Differential diagnosis
5. Clinical judgement
6. Managing patient concerns
7. Managing patient welfare

Examiners will mark candidates as satisfactory, borderline or unsatisfactory in each area. It is necessary to reach a minimum standard in each of these areas, as well as meet the overall pass mark.

## REFERENCE

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Membership of the Royal Colleges of Physicians of the United Kingdom. (n.d.) PACES examination format. Available from <https://www.mrcpuk.org/mrcpuk-examinations/paces/paces-examination-format>. Accessed 25 July 2016.

# Top Tips for PACES Success: An Examiner Speaks

I have been an examiner for the Practical Assessment of Clinical Examination Skills (PACES) since dinosaurs walked the earth (or at least since the exam started). The following views are entirely my own and based on my observations of hundreds of candidates I have seen during that time.

- **The cases are usually straightforward with clear-cut physical signs.**

Centres are actively discouraged from selecting rare or esoteric conditions. Patients with physical signs sometimes come to the exam on an annual basis, and occasionally their physical signs may become less obvious over time. If this is the case, an allowance will be made during 'calibration'; a process prior to the exam where the two examiners at that station examine the patient independently, compare findings and make an assessment of the abnormalities a candidate might be reasonably expected to find.

- **I am often asked, 'What is the standard required to pass PACES?'**

There are of course strict marking criteria for each station, but the majority of examiners are 'jobbing' physicians; we don't expect super specialist levels of knowledge. For most of the time, we are examining cases outside our own speciality. For me, the overall benchmark is, 'Would I feel comfortable with this doctor running the general medical take in my hospital?'

- **Practise, practise, practise!**

I am afraid it's all too easy to spot a candidate who isn't wholly comfortable doing a physical examination. Get a senior colleague to critique your exam technique: it is important that you have a structured approach that you could do almost automatically.

- **You will not pass unless you can identify physical signs.**

Part of this is pattern recognition: once you have felt polycystic kidneys, you are likely to correctly identify them in the future. The longer and more diverse your clinical experience is, the more likely you will be to detect physical signs because you have seen them before. When you think about it, the types of physical sign you are likely to come across in each station are fairly limited, so when preparing for PACES, sit down and make a list of your deficiencies and make sure you have seen all the common abnormalities at least once, even if that means visiting other hospitals. I am not generally a fan of commercial courses, but this may be an area where they can help if your exposure to abnormal physical signs is limited. The other common problem is candidates who 'look but do not see'; in other words, they go through the motions of the examination almost by rote but do not find the abnormalities they are ostensibly looking for.

- **Not every patient in the exam will have something to find.**

So don't make things up! Abdominal and chest cases particularly may only have a scar to find. In general, if you can't find anything wrong, then you are probably correct.

- **Station 5 is problematic.**

Station 5 carries a lot of marks, and the time available is limited. You have to be able to talk and examine at the same time; the good news is that the tasks you are given and any examination findings are usually pretty straightforward. Don't try to examine too many systems at once; read the introductory statement and allow that to focus your history/discussion and examination. The examiners realise you will not be able to do everything, so concentrate on what you think is important.

- **Timing is everything.**

When practising, make sure that you take 6 minutes to complete your abdominal, respiratory, cardiology and focused neurological examinations. Finish too early and you are likely to have missed something and exposed yourself to extra questioning. Too late and you will have failed to pick up the appropriate signs. In general, in the talking stations you should have plenty of time to complete the required task, so go through the history or the communication issue in a steady, structured way, clarifying things where necessary and avoiding repetition. Try not to finish early; if you do, you are very likely to have missed something. When you are sitting outside the station in the 5 minutes after reading the clinical information, list your structured approach to problem, questions to cover in the history-taking station and important issues that need to be addressed (and questions to be answered) in communications skills.

- **Don't waste time.**

Especially on unnecessary 'peripheral' examinations; spending 3 minutes examining for a collapsing pulse will not give you time to palpate the precordium and listen to the heart properly. If you discover a displaced apex and a diastolic murmur, you can go back and do it later.

- **Honesty is the best policy.**

Don't make up physical signs because you think they should be there or you think that's what the examiner wants. In the talking stations, if you don't know the answer to the surrogate's question, say so, but explain why and how you will obtain the information.

- **Don't beat about the bush.**

Candidates who are not confident about things will sometimes be deliberately vague when describing their findings; this is unlikely to help. The examiners will press you to clarify things, so it's best to come clean early on, describing what you have found, what it could be and why.

- **Always answer the question.**

In the talking station, the surrogate will have been primed with questions or concerns. A good candidate may answer most of these during the consultation. But if asked a direct question by the surrogate, you should usually try to answer it there and then. Occasionally, you may feel you have not gathered enough information to do so, in which case you can say so and return to the question later, but do not forget that failure to address the 'patient' concerns is a common cause of poor marks in Stations 2 and 4.

- **Create an impression.**

Wear comfortable but smart clothing; it's not a fashion show, but you should look professional – so no jeans or short skirts. Don't weigh yourself down with equipment; all you need is a stethoscope. Everything else will be provided, and



bringing more stuff just means a greater chance of dropping it or leaving it behind at one of the stations.

- **Read the introductory statement carefully.**

Particularly in the neurological station, it should tell you which area to focus on; it is impossible to do a complete neurological examination in 6 minutes. In the other stations, it is likely to guide questioning, particularly when assembling the list of possible causes of the physical findings.

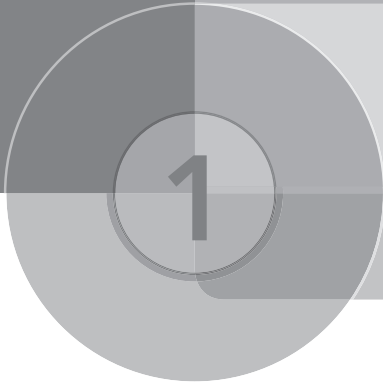
- **Be kind to the patient and try to look as though you are enjoying yourself.**

Not an easy thing to do, but handling the patient with courtesy and good humour will set you in good stead with the examiners and show that you are an experienced clinician.

- **Finally, the examiners genuinely want you to pass.**

You may not believe it or feel like it at the time, but it's true. We have been in your position ourselves at some stage and realise how nerve racking it is. You will need to meet the required standard, but we will give you every opportunity to demonstrate that you can, given the confines of the examination structure. During questioning, the examiner may try to lead you towards a diagnosis; this is not a plot to try to trick you into saying the wrong thing but is usually a genuine attempt to make you clarify your views on a clinical problem.

**Anonymous reviewer**



# Station 1: Respiratory

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## HINTS FOR THE RESPIRATORY STATION

- Observe the patient from the end of the bed and comment on chest expansion and respiratory rate.
- Look for clues around the bedside, e.g. inhalers, spacer device, peak flow metre, nebuliser.
- Be aware of the different types of inhalers and the colour design and contents of each inhaler type.
- Always examine the contents of the sputum pot if present.
- If the patient is on oxygen, comment on delivery method and the percentage inspired if appropriate.
- Examine the patient starting from the back, as findings are more likely to be picked up from the back (important, if time is running out).
- Try to look for underlying causes of the pathology identified.
- Look for signs of cor pulmonale where appropriate.
- Complete the examination by telling the examiner you would like to check oxygen saturations, peak flow and spirometry results where appropriate.
- Use ward rounds and clinics as opportunities to practise your clinical examination skills and with presenting cases.

## BRONCHIECTASIS

Please examine this patient who has a cough productive of thick sputum.

### FINDINGS

- *General:* Age (may help with underlying diagnosis), cachexia, dyspnoea, inhalers, sputum pot (check for thick sputum  $\pm$  haemoptysis), nebulisers
- *Peripheral:* Clubbing, signs of cor pulmonale, long line, tunnelled central venous catheter
- *Chest:* Coarse inspiratory crackles (which may clear with coughing), wheeze, situs inversus, scars from previous operations

### PRESENTATION

On examination, this patient has evidence of bilateral coarse inspiratory crackles to the midzones and a wheeze. She also has evidence of finger clubbing, and the sputum pot by her bedside has thick dark-green-coloured sputum. The diagnosis in this patient is bronchiectasis. The patient has no evidence of cachexia, situs inversus or previous operations. In this patient, the most likely cause may be as a result of previous infection, and I would like to investigate this further.

- Comment on the patient's productive cough.
- Look for a sputum pot and be sure to look at the contents.
- Look for an obvious underlying cause, including: possible signs of cystic fibrosis (CF) and chronic obstructive pulmonary disease (COPD).
- If there is no obvious underlying cause, mention that the top three causes are postinfective, COPD and Cystic Fibrosis.



## INVESTIGATIONS

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- Diagnosis
  - Chest X-ray (CXR): May be normal, dilated bronchi with thickened walls ('tramline' shadowing), ring shadowing.
  - High-resolution CT (HRCT): Bronchial wall dilatation with possible thickening, airway size is greater than accompanying artery size ('signet ring' appearance).
  - Sputum: Send for culture and sensitivities (*Pseudomonas aeruginosa* is important), atypical screen, acid-fast bacilli (AFB) and fungal cultures.
  - Spirometry: May show an obstructive picture (usually forced expiratory volume in first second [FEV1]/forced vital capacity [FVC] ratio <0.7).
  - Bloods: Check serum immunoglobulin levels (immunoglobulin [Ig] A, IgG, IgM) to rule out immunodeficiency as a cause, *Aspergillus* serology (total IgE, IgE to *Aspergillus* and *Aspergillus* precipitins).
- Investigations for the underlying cause of bronchiectasis
  - Cystic fibrosis: Two measurements of sweat chloride and cystic fibrosis transmembrane conductance regulator (CFTR) genetic mutation analysis.
  - Immunodeficiency: Immunoglobulin screen, functional antibody levels.
  - Allergic bronchopulmonary aspergillosis (ABPA) screen.
  - Ciliary dysfunction (e.g. primary ciliary dyskinesia): Saccharin test or ciliary electron microscopy to assess ciliary function.
  - Mechanical obstruction: Computerised tomography (CT) of the chest will locate any obstructive lesions, e.g. foreign body, lymph node compression or tumour.

## MANAGEMENT

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- Specialist physiotherapy with a daily routine for patients
- Antibiotics
  - Treatment of acute exacerbations often requires intravenous antibiotic therapy with antipseudomonal agents (piperacillin, ceftazidime, carbapenems, aminoglycosides).
  - Oral ciprofloxacin may be used.
  - Long-term antibiotics may include nebulised colistin and oral macrolides.
- Bronchodilators
  - In those with airflow limitation
- Mucolytics
  - Carbocysteine
- Surgery
  - Bronchiectasis is rarely sufficiently localised to be amenable to surgery but may be an option in a very small minority of cases.

## QUESTIONS

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1. What is bronchiectasis?
  - Abnormal and permanently dilated airways with bronchial wall thickening.
  - This is manifest as a cough with the production of thick sputum.

**2. What are the causes of bronchiectasis?**

- Postinfective bronchial damage: Severe bacterial and viral pneumonias, including measles and pertussis, tuberculosis (TB) and nontuberculous mycobacterial infections
- Mucociliary clearance defects: Cystic fibrosis, primary ciliary dyskinesia, Kartagener's syndrome, Young's syndrome
- Immunodeficiency: Primary (immunoglobulin deficiency) and secondary (human immunodeficiency virus [HIV] infection)
- Mechanical: Obstruction (tumour, foreign body)
- Immunological response: Allergic bronchopulmonary aspergillosis

**3. What is the differential diagnosis of bilateral lower-zone crackles?**

- Bronchiectasis: Coarse crackles heard in early–mid inspiration
- Lung fibrosis: Fine, late (end-inspiratory) crackles; look for clubbing, dry cough and cyanosis, as well as a cause, such as connective tissue disease
- Pulmonary oedema: Fine/coarse bibasal crackles; look for evidence of fluid overload
- Bilateral pneumonia: Coarse crackles; look for pyrexia and bronchial breathing

**KEY POINTS**

- On examination, coarse inspiratory crackles (that clear with coughing), along with finger clubbing, are the hallmark clinical features of bronchiectasis.
- Typical radiological signs seen on HRCT include bronchial wall thickening and the classic 'signet ring' appearance of dilated bronchi.
- Ensure that you are familiar with the common causes of bronchiectasis and can list them in an ordered fashion.

**REFERENCE**

Pasteur MC, Bilton D, Hill AT. (2010) British Thoracic Society guideline for non-CF bronchiectasis. *Thorax* 65: 577.

**CHRONIC OBSTRUCTIVE PULMONARY DISEASE**

Please examine this patient with breathlessness.

**FINDINGS**

- *General:* Dyspnoea, pursed lip breathing, prolonged expiratory time, use of accessory muscles, wheeze, presence of inhalers and oxygen
- *Peripheral:* Cyanosis, tar staining on finger nails, bronchodilator (fine) tremor, flap, signs of cor pulmonale
- *Chest:* Hyperexpanded chest, hyperresonance and reduced breath sounds (over bullae), wheeze, Hoover's sign (inward movement of the lower rib cage during inspiration associated with COPD)





## PRESENTATION

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This gentleman is dyspnoeic at rest with pursed lip breathing and is on oxygen therapy at 2 L/min. He has nicotine-stained fingernails. There is a widespread wheeze audible throughout both lung fields, and the patient has evidence of cor pulmonale. I note the presence of inhalers at the bedside, including salbutamol and a combination inhaler. The diagnosis in this patient is COPD.

- Comment on external clues, including inhalers, oxygen, nebulisers and a sputum pot.
- Comment on any features of respiratory distress and ensure the respiratory rate is counted.
- Look for features of cor pulmonale (raised jugular venous pressure [JVP], loud pulmonary component of loud second heart sound [P2], peripheral oedema), polycythaemia (plethora), infection (consolidation) and Cushing's syndrome (corticosteroid use).

## INVESTIGATIONS

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- Diagnosis
  - Spirometry with bronchodilator response (to differentiate from asthma). FEV1/FVC ratio <0.7 and not fully reversible
- Arterial blood gas (ABG) (type 1 or 2 respiratory failure is possible)
  - CXR (hyperinflation, flat hemidiaphragms, bullae, large prominent pulmonary arteries)
- Complications
  - Echocardiogram (pulmonary hypertension), CXR (infection/pneumothorax/bullae)

## MANAGEMENT

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- Acute
  - Oxygen therapy (with caution and monitoring of arterial blood gases), nebulised bronchodilators, corticosteroids, antibiotics for infection, theophylline, physiotherapy, consideration of noninvasive positive-pressure ventilation (NIPPV) and intubation for more severe cases
- Chronic: Depends on the severity of airway obstruction
  - Mild
    - PRN inhalers: Short-acting bronchodilator, e.g. salbutamol
  - Moderate
    - If still breathless and FEV1 >50%, the patient can have a long-acting beta-agonist (LABA), e.g. salmeterol, or a long-acting muscarinic antagonist (LAMA), e.g. tiotropium.
    - If still symptomatic and FEV1 <50%, the patient can have a combination inhaler (LABA/steroid) or a LAMA.
    - For patients who are still symptomatic, regardless of the FEV1, they can be prescribed a combination inhaler and a LAMA. There are also newer inhalers which are a combination of a LABA/LAMA.
  - Severe
    - Consider home nebulisers, theophyllines and anti-mucolytics.

- Extras
  - Smoking cessation advice is key and should be discussed with all patients, as well as offering nicotine replacement therapy and drugs such as varenicline and bupropion to help stop. Smoking cessation therapy is monitored using carbon monoxide (CO) readings (<10 ppm at 4 weeks).
  - Other considerations include nutritional management, pulmonary rehabilitation, vaccinations (pneumococcal and influenza), long-term oxygen therapy (LTOT), surgery, social support, psychological support and palliative care input.

## QUESTIONS

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1. What is cor pulmonale? What is its significance?
  - Right-sided cardiac dysfunction secondary to pulmonary hypertension. The pulmonary hypertension must be of a respiratory cause (chronic lung disease, pulmonary vasculature disorders, neuromuscular disease affecting the respiratory system).
  - Untreated cor pulmonale causes right-sided heart failure and death.
2. What are the indications for LTOT?
  - LTOT describes oxygen given for >16 hours/day, with the aim of achieving a  $\text{PaO}_2 > 8 \text{ kPa}$ .
  - It is indicated for those patients with a  $\text{PaO}_2 < 7.3 \text{ kPa}$  on two consecutive readings at least 3 weeks apart (in a stable patient), or for a  $\text{PaO}_2 7.3\text{--}8 \text{ kPa}$  in a patient with cor pulmonale.
  - All patients with an FEV1 <30% predicted, signs of right-sided heart failure and oxygen saturations of <92% should be considered for LTOT.
  - Smoking is not a contraindication to LTOT; however, patients need to be advised of the risks associated.
3. How would you classify the severity of COPD?
  - Use the National Institute for Health and Care Excellence (NICE) guidelines on classifying the severity of COPD based on the presence of symptoms and the percentage predicted of their FEV1:
    - Mild: FEV1 >80%
    - Moderate: FEV1 50%–80%
    - Severe: FEV1 30%–50%
    - Very severe: FEV1 <30%
4. What surgical interventions can be offered to patients with COPD?
  - Bullectomy: In symptomatic patients who have a large bulla and FEV1 <50% predicted.
  - Lung volume reduction surgery (LVRS): This can be considered in patients who have upper-zone dominant emphysema, FEV1 >20% predicted,  $\text{PaCO}_2 < 7.3 \text{ kPa}$  and transfer factor of the lung for carbon monoxide (TLCO) >20%.
  - Lung transplantation.



## KEY POINTS

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- Be able to identify what medications inhalers contain, from their colour/design.
- Ensure that you know the criteria for LTOT.

## REFERENCES

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- Hardinge M, et al. (2015) BTS guidelines for home oxygen use in adults. *Thorax* 70: i1–43.
- National Institute for Health and Care Excellence. (2010) Guidance CG101. Chronic obstructive pulmonary disease in over 16s: Diagnosis and management. Available from <https://www.nice.org.uk/guidance/cg101>. Accessed 17 August 2016.
- Troosters T, et al. (2005) Pulmonary rehabilitation in chronic obstructive pulmonary disease. *Am J Respir Crit Care Med* 172: 19–38.

## CONSOLIDATION

Please examine this patient's respiratory system.

## FINDINGS

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- *General:* Oxygen, nebulisers, sputum pot, haemoptysis, herpes labialis, cough
- *Peripheral:* Tachypnoea, tachycardia, pyrexia, respiratory distress
- *Chest:* Decreased chest expansion, dullness to percussion over affected lobe, bronchial breathing, crackles, increased vocal resonance

## PRESENTATION

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This patient has evidence of consolidation at his right base. He is receiving oxygen therapy and is tachypnoeic with a respiratory rate of 28 breaths/minute. There is a sputum pot containing thick green sputum. There is dullness to percussion at the right lung base, and on auscultation, there is evidence of bronchial breathing and coarse crackles at the right base.

- Remember that in the exam situation if the information about the case is handwritten, it is likely that a patient has withdrawn from the exam at short notice and a last-minute replacement may have been recruited from the inpatient wards.
- Such a patient is likely to be more unwell than a stable patient; therefore, comment on any features of respiratory distress, note any infusions or nebulisers and check the FiO<sub>2</sub> being delivered.
- Patients at highest risk of pneumonia include immunocompromised, COPD, elderly and alcoholic patients.

## INVESTIGATIONS

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- Diagnosis
  - CXR, ABG, sputum culture, routine bloods, blood cultures, atypical pneumonia screen (urine for legionella and pneumococcal antigens, mycoplasma serology) if indicated

## MANAGEMENT

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- Use the CURB-65 score to assess the severity of the pneumonia.
  - Confusion (abbreviated mental test score [AMTS]  $\leq 8$ )
  - Urea  $>7$  mmol/L
  - Respiratory rate  $\geq 30$  breaths/minute
  - Blood pressure (systolic  $<90$  mmHg or diastolic  $<60$  mmHg)
  - Age  $>65$  years
- A score of 0 or 1: Treat as an outpatient.
- A score of 2: Possible short stay in hospital.
- A score of 3–5: Requires hospitalisation and may require critical care intervention.
- Treatment is with antimicrobials and oxygen therapy where required. In more severe cases, critical care intervention may be required.
- Antimicrobial therapy is guided by local trust guidelines, but generally use amoxicillin  $\pm$  a macrolide (to cover atypical organisms in severe cases).
- Patients should be followed up after a 6-week interval with a repeat CXR to ensure that the consolidation has resolved and there is nothing sinister underlying it.
- Complications of pneumonia can include pleural effusion, empyema or a lung abscess.

## QUESTIONS

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1. What are the common organisms causing community-acquired pneumonia?
  - Common: *Streptococcus pneumoniae*, *Haemophilus influenzae*, *Staphylococcus aureus*
  - Atypical: *Mycoplasma pneumoniae*, *Legionella pneumophila*, *Chlamydia pneumoniae*, *Chlamydia psittaci*, *Coxiella burnetii*
  - Viruses: Influenza, cytomegalovirus (CMV) and varicella-zoster
2. List possible complications of pneumonia.
  - Parapneumonic effusion, empyema, cavitation, lung abscess, septic shock, respiratory failure/acute respiratory distress syndrome (ARDS), hepatitis, haemolytic anaemia, erythema multiforme
3. What is the difference between an empyema and a complicated parapneumonic effusion?
  - An empyema is pus in the pleural cavity with a pH  $<7.2$ .
  - A complicated parapneumonic effusion has a pH  $<7.2$  but is clear.
  - A parapneumonic effusion is clear with a pH  $>7.2$ .



4. How would consolidation be differentiated from an effusion on clinical examination?
  - Tactile vocal fremitus: Sound transmission is increased through tissue (consolidation) and decreased through fluid (pleural effusion).
  - Whispering pectoriloquy: Is indicative of consolidation when whispered sounds are heard clearly through affected lung tissue.
5. Other than guiding the clinical management of a patient, what other information does the CURB-65 score give?
  - The CURB-65 score also indicates the mortality associated with the severity of the pneumonia. A higher score is associated with a higher mortality rate:
    - Score 0–1 = <5% mortality rate
    - Score 2 = 9% mortality rate
    - Score 3–5 = 15%–40% mortality rate

## KEY POINTS

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- Look closely at the information presented to the candidate, i.e. is it handwritten.
- Patients from an inpatient ward may be more unwell; comment on any respiratory distress and any infusions.
- Be aware of complications associated with pneumonia.

## REFERENCE

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National Institute for Health and Care Excellence. (2014) Guidance CG191. Pneumonia in adults: Diagnosis and management. Available from <https://www.nice.org.uk/guidance/cg191?unlid=2520898732016221192356>. Accessed 17 August 2016.

## CYSTIC FIBROSIS

This patient has presented with repeated chest infections. Please examine their respiratory system.

## FINDINGS

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- *General*: Young patient, short stature, cachexia, pallor, dyspnoea, tunnelled central venous catheter (port-a-cath) or long line, inhalers, sputum pot, pinpricks from blood glucose measurement, nebulisers, Creon tablets, percutaneous endoscopic gastrostomy (PEG) tube
- *Peripheral*: Finger clubbing, signs of cor pulmonale (in patients with severe lung disease)
- *Chest*: Coarse inspiratory crackles (which may clear with coughing), wheeze

## PRESENTATION

---

This young patient has cystic fibrosis. He appears cachectic and also has a port-a-cath *in situ*. The patient has finger clubbing and also is likely to have CF-related diabetes due to the pinpricks on his fingertips from blood glucose monitoring. On auscultation of his chest, there is evidence of coarse crackles in both lower zones in keeping with bronchiectasis.

- Comment on the patient's general appearance and look for any extrapulmonary manifestations. Some patients may have had a liver or renal transplant as well.

## INVESTIGATIONS

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- Diagnosis
  - Guthrie test.
  - Further genetic testing:  $\Delta F508$  is the most common mutation.
  - Sweat test: Sweat sodium and chloride.
- Complications
  - Chest
    - Chest X-ray: Bronchial wall thickening, mucous plugging, pneumothorax
    - HRCT: Bronchial wall thickening, mucous plugging, bronchiectasis (signet ring sign)
    - Sputum culture: Looking for common CF pathogens, atypical infections, AFB and fungal infections
    - Spirometry: Obstructive pattern
  - CF-related diabetes: Oral glucose tolerance test, Hba1c, blood sugar series and continuous glucose monitoring system
  - Pancreatic insufficiency: Faecal elastase levels
  - CF-related liver disease: Liver function tests (LFTs), coagulation screen (assess synthetic function of liver), ultrasound scan (USS) of liver.
  - Ear, nose and throat (ENT) complications (nasal polyps, sinusitis): CT scan of sinuses
  - Osteoporosis/osteopenia: Dual-energy X-ray absorptiometry (DEXA) scan, parathyroid hormone (PTH), calcium and vitamin D

## MANAGEMENT

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- Multidisciplinary team (MDT) approach
  - Respiratory physician (CF specialist), CF specialist nurse, physiotherapist, dietitian, clinical psychologist, GP, other medical teams (including endocrine and gastroenterology)
- Specialist physiotherapy
- Antibiotics (preventative nebulised antibiotics, as well as rescue courses during an acute exacerbation)
  - See bronchiectasis section.
- Mucolytics
  - DNase (nebulised), hypertonic saline
- Nutrition
  - Special diet: High calorie/high fat, vitamins and Creon.
  - Enteral feeding may be needed.



- Management of extrapulmonary complications, including CF-related diabetes and liver disease
- Psychological support

## QUESTIONS

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1. How common is cystic fibrosis in the UK population?
  - Cystic fibrosis occurs in approximately 1 in 2500 live births.
  - Cystic fibrosis is an autosomal recessive condition.
  - The chance of being a carrier in the United Kingdom is 1 in 25.
2. What organisms are commonly found in the sputum of patients with cystic fibrosis, and which are most important for prognosis?
  - *Haemophilus influenzae*, *Staphylococcus aureus* (commonly found in CF), *Moraxella catarrhalis*, *Streptococcus pneumoniae*, atypical mycobacteria, *Aspergillus fumigatus*.
  - *Burkholderia cepacia* (in particular cenocepacia), *Pseudomonas aeruginosa* and *Mycobacterium abscessus* infection are poor prognostic indicators.
3. List the extrapulmonary manifestations of cystic fibrosis.
  - Pancreatic: Malabsorption
  - Endocrine: CF-related diabetes – often associated with pancreatic insufficiency
  - Hepatobiliary: Gallstones, cirrhosis, portal hypertension
  - Intestinal: Distal intestinal obstruction syndrome (meconium ileus equivalent) which is treated with laxatives, including gastrograffin
  - Musculoskeletal: Osteoporosis, arthritis, osteopenia
  - Sinusitis and nasal polyps
  - Male infertility, delayed puberty
4. What are the respiratory complications caused by CF?
  - Infective exacerbations
  - Pneumothorax
  - Haemoptysis
  - *Aspergillus* lung disease
  - Respiratory failure
5. Are you aware of any new treatments available for CF?
  - Genetic modulators such as ivacaftor are now available for some CF patients with particular genetic defects, e.g. G551D.
  - Ivacaftor works on chromosome 7, CFTR gene, G551D defect (7% of CF patients) by making the chloride channel functional.

## KEY POINTS

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- Be able to recognise these patients, taking their age and general appearance into account.
- Recognise and comment on any extrapulmonary manifestations.
- Be aware of common respiratory tract pathogens in cystic fibrosis.

## REFERENCE

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Ramsey B, et al. (2011) A CFTR potentiator in patients with cystic fibrosis and the G551D mutation. *N Engl J Med* 365: 1663–72.

## FIBROTIC LUNG DISEASE

Please examine this patient who is breathless.

## FINDINGS

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- *General:* Dyspnoea, oxygen, cachexia, tachypnoea
- *Peripheral:* Cyanosis (peripheral and central), clubbing, signs of cor pulmonale
- *Chest:* Scar (from biopsy), reduced chest expansion, fine end-inspiratory crackles

## PRESENTATION

---

This patient has pulmonary fibrosis as evidenced by finger clubbing and fine inspiratory bibasal crackles. The likely underlying cause is systemic sclerosis, as the patient also has evidence of sclerodactyly, telangiectasiae and microstomia.

- This is a standard case which may not be included on its own in the respiratory station and may also appear in Station 5, so look for a possible underlying cause:
  - Features of rheumatoid arthritis: Swan neck and boutonnière deformities, Z-thumb, ulnar deviation at the wrist, nodules at the elbow
  - Ankylosing spondylitis: 'Question mark' posture
  - Pacemaker and amiodarone facies
  - Radiation burns and tattoos: Radiation therapy
  - Features of systemic sclerosis: Sclerodactyly, telangiectasiae, beaked nose, furrowing of the mouth
- Also look for features of Cushing's syndrome (corticosteroid use)

## INVESTIGATIONS

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- Diagnosis
  - ABG: Type 1 respiratory failure
  - Pulmonary function tests – restrictive picture, reduced transfer factor
  - CXR: Peripheral and basal reticular shadowing
  - HRCT: Ground-glass appearance
- Cause
  - Full history and examination, autoimmune screen, serum angiotensin-converting enzyme (ACE), bronchoalveolar lavage, transbronchial lung biopsy (TBLB) or surgical lung biopsy (if diagnostic doubt)





## MANAGEMENT

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- Discussion at interstitial lung disease MDT
- Smoking cessation
- Pulmonary rehabilitation
- Gastric protection (e.g. proton pump inhibitors) for any patients with symptoms of reflux disease
- LTOT
- Nutritional assessment
- Psychological support
- Palliative care

## QUESTIONS

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1. Please describe the causes of lung fibrosis and divide them into upper- and lower-zone aetiologies.
  - Upper zone
    - Berylliosis, radiation, extrinsic allergic alveolitis, ankylosing spondylitis, sarcoidosis, TB (mnemonic: 'breast')
  - Lower zone
    - Rheumatoid arthritis and other connective tissue diseases, idiopathic, drugs (methotrexate and amiodarone), asbestosis
2. What new treatments are available for the treatment of pulmonary fibrosis (usual interstitial pneumonia [UIP] pattern)?
  - Pirfenidone and nintedanib are both antifibrotics which have recently been introduced to slow down the progression of mild–moderate disease. Any patient being considered for treatment needs to be discussed at an interstitial lung disease MDT and, if they fit the required criteria, can be prescribed treatment from a specialist centre.
3. What are the pulmonary manifestations of rheumatoid arthritis?
  - Lung fibrosis (which may also be secondary to methotrexate treatment)
  - Pleural effusions
  - Intrapulmonary nodules (including Caplan's syndrome)
  - Obliterative bronchiolitis

## KEY POINTS

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- Fine end-inspiratory crackles are suggestive of fibrotic lung disease; a key differential is pulmonary oedema.
- Know the causes of pulmonary fibrosis and the lung zones they affect.
- Ensure that any management plan for a patient with pulmonary fibrosis is based around a MDT approach.
- Comment on any evidence of cor pulmonale which signifies advanced disease.

## REFERENCE

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National Institute for Health and Care Excellence. (2013) Guidance CG163. Idiopathic pulmonary fibrosis in adults: Diagnosis and management. Available from <https://www.nice.org.uk/guidance/CG163>. Accessed 8 August 2016.

## LUNG CANCER

Please examine this patient who has presented with a chronic cough.

## FINDINGS

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- *General:* Cachexia (may or may not be present), lymphadenopathy, hoarse voice
- *Peripheral:* Finger clubbing, nicotine staining
- *Chest:* Scars from previous lobectomy/pneumonectomy, radiation tattoos/burns, reduced chest expansion, tracheal deviation, reduced breath sounds, dullness to percussion, possible pleural effusion

## PRESENTATION

---

This patient appears cachectic and has finger clubbing and a hoarse voice. There are palpable cervical lymph nodes. The patient has a small blue tattoo at the front of his chest which is indicative of a radiotherapy tattoo and also has a lateral thoracotomy scar. The diagnosis in this patient would be previous lung cancer (possible recurrence with chronic cough), with the patient having had surgery and radiotherapy in the past for treatment.

Look for the following features:

- Superior vena caval obstruction (SVCO): Raised JVP, oedematous face, distended veins over chest wall and neck, respiratory distress
- Metastases
- Hepatomegaly, bony tenderness, skin lesions, cervical lymph nodes
- Paraneoplastic disorders (see below)
- Pancoast's syndrome (see below)

## INVESTIGATIONS

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- Diagnosis
  - CXR: Mass, pleural effusion, bulky hilum
  - CT scan: Look for evidence of masses, lymphadenopathy and metastases
  - Positron emission tomography (PET) scan: For evaluation and staging of lung cancers (especially pulmonary nodules, lymph nodes and distant metastases)
  - Pleural fluid cytology
  - Bronchoscopy and biopsy of mass lesions
  - Endobronchial ultrasound (EBUS)-guided biopsy: to assess mediastinal lymph nodes



- CT-guided lung biopsy: For peripheral lung lesions
- Biopsy of peripheral lesions to assess for metastases
- Thoracoscopy: To assess an exudative pleural effusion of unknown aetiology

## MANAGEMENT

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- Referral to lung MDT team.
- Dependent on staging and histology; surgical resection/chemotherapy/radiotherapy/palliation.
- Surgical resection (pneumonectomy or lobectomy) is suitable for patients with adequate lung function and no medical contraindications.
- Refer to patient's functional status using the World Health Organization (WHO) performance status.

## QUESTIONS

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1. What are the four main histological types of lung cancer and how common are each of these?
  - Lung cancer classification can be divided into small-cell lung cancer (SCLC) and non-small-cell lung cancer (NSCLC).
  - SCLC comprises 15% of all cases, whereas NSCLC (adenocarcinoma [35%–40%], squamous cell carcinoma [25%–30%] and large-cell carcinoma [10%–15%]) accounts for the remainder of lung malignancies.
  - Small-cell tumours have the worst prognosis, as they have a rapid growth rate and metastasise early.
2. Which paraneoplastic syndromes are associated with lung cancer?
  - *Small-cell lung cancer* is the most common form of cancer associated with paraneoplastic syndromes, including
    - Ectopic hormone secretion: Adrenocorticotrophic hormone (ACTH) (Cushing's syndrome) and antidiuretic hormone (ADH) (causing syndrome of inappropriate antidiuretic hormone secretion [SIADH] – low sodium)
    - Lambert–Eaton myasthenic syndrome (LEMS)
  - *Squamous cell carcinoma*: Parathyroid hormone–related peptide release, resulting in hypercalcaemia (note that hypercalcaemia is more frequently a result of bony metastases).
  - *Adenocarcinoma*: Hypertrophic pulmonary osteoarthropathy (HPOA); it results in gross finger clubbing and arthritis with radiological evidence of subperiosteal new bone formation.
3. What are the signs of Pancoast's syndrome?
  - Pancoast's syndrome is characterised by an apical lung tumour with involvement of the brachial plexus and cervical sympathetic nerves; patients may complain of pain in the shoulder/anterior chest wall and arm weakness, and have wasting of the intrinsic muscles of the hand and an ipsilateral Horner's syndrome (ptosis, miosis, anhidrosis and enophthalmos).

4. What are the clinical signs and symptoms of SVCO?
  - Oedema of the face, neck and upper body
  - Prominent neck and chest wall vessels
  - Facial plethora
  - Stridor
  - Headache (often worse on bending forward)
  - Dizziness

## KEY POINTS

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- Comment on any past treatment, including surgery and radiotherapy.
- Comment on any associated complications as detailed above.
- Be able to differentiate between a pneumonectomy and lobectomy on clinical examination.

## REFERENCE

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Lim E, et al. (2010) Guidelines on the radical management of patients with lung cancer. *Thorax* 65(Suppl III): iii1–27.

## OLD TUBERCULOSIS

Please examine this gentleman's respiratory system.

## FINDINGS

---

- *General:* May appear well or cachectic
- *Chest:* Tracheal deviation, thoracotomy scar, rib resection, decreased breath sounds with reduced chest expansion, signs of fibrosis/bronchiectasis, evidence of respiratory failure secondary to thoracoplasty
  - Look for a supraclavicular scar from a phrenic nerve crush procedure, scarring from induced pneumothoraces or a lateral thoracotomy scar

## PRESENTATION

---

This gentleman appears well with no evidence of respiratory distress. He has a scar in the left supraclavicular fossa with fine inspiratory crepitations in the left upper zone. The patient also has a left-sided chest wall deformity and a lateral thoracotomy scar. The examination findings in this patient would be consistent with old TB with a phrenic nerve crush and thoracoplasty.

## INVESTIGATIONS

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- CXR
  - Raised hemidiaphragm on the side of phrenic nerve crush procedure, upper-lobe fibrosis, areas of cavitation



- CT scan
  - Areas of fibrosis, loss of lung volume, thickened or calcified pleura, areas of bronchiectatic change
- Spirometry
  - Postthoracoplasty may show an obstructive or restrictive defect.

## MANAGEMENT

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- The patient may be completely well and not need treatment.
- They may be investigated for possible recurrence of TB (bronchoscopy for a bronchoalveolar lavage) or manifestations of old TB, such as pulmonary fibrosis.

## QUESTIONS

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1. Outline the current drug therapies available for treatment of TB, and their side effects.
  - The standard treatment for TB involves a combination of the following drugs: rifampicin, isoniazid, pyrazinamide and ethambutol. Patients are initially treated with all four drugs for the initial 2 months of therapy, followed by rifampicin and isoniazid for the subsequent 4 months; treatment is complete at 6 months.
  - Multi-drug-resistant TB is that which is resistant to rifampicin and isoniazid.
  - Drug-induced hepatitis
    - Rifampicin, isoniazid and pyrazinamide (check LFTs prior to commencement)
  - Optic neuritis
    - Ethambutol (visual acuity should be documented before starting treatment)
  - Peripheral neuropathy
    - Isoniazid (coprescribe pyridoxine)
2. Discuss past surgical treatments of TB.
  - Induced pneumothoraces
    - To collapse the affected lung; procedure repeated every few weeks
  - Phrenic nerve crush
    - To paralyse the diaphragm and cause collapse of the underlying lung
  - Plombage
    - Insertion of polystyrene balls into the chest cavity to cause collapse of underlying lung
  - Thoracoplasty
    - Ribs around the infected cavity broken and pushed inwards to collapse underlying lung
3. What tests are used in the diagnosis of TB?
  - CXR: There may be evidence of an enlarged hilum, consolidation, cavitation, pleural effusion or granulomata.
  - Sputum staining: Ziehl–Neelsen staining for acid-fast bacilli.

- Bronchoscopy and washings.
- Biopsies of extrapulmonary manifestations, e.g. lymph nodes.
- Whole-blood interferon or skin tests (Mantoux test).
- Always consider HIV testing in patients with suspected TB.

## KEY POINTS

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- Be aware of the past surgical treatments for TB and the associated clinical signs.
- Be aware of the current treatment recommendations for TB.
- Be aware of the main side effects of anti-TB therapy.

## PLEURAL EFFUSION

Please examine this patient's respiratory system. They have been complaining of worsening shortness of breath.

## FINDINGS

---

- *General:* Dyspnoea, oxygen and walking aids
- *Peripheral:* Dependent on the underlying cause (see below)
- *Chest:* Decreased chest expansion, trachea deviated away from side of effusion, stony-dull percussion note on affected side, decreased breath sounds on affected side

## PRESENTATION

---

On examination, this patient has evidence of a right-sided pleural effusion. He has reduced chest expansion and a stony-dull percussion note to the right midzone with reduced breath sounds. The likely cause in this patient would be lung cancer, as he has evidence of scars that are likely from a chest drain at the right side of his chest, has finger clubbing and is cachectic.

Search for a cause for the effusion:

- Connective tissue disease: Features of rheumatoid arthritis butterfly facial rash indicative of systemic lupus erythematosus (SLE)
- Cardiac disease: Raised JVP and ankle swelling
- Lung cancer: Clubbing, wasting, Horner's syndrome, radiation scars, tattoos, lymphadenopathy, previous drain site scars
- Liver disease: Leuconychia, palmar erythema, spider naevi, gynaecomastia, parotid swelling, ascites
- Renal disease: Arteriovenous fistulae, scars from neck lines, peritoneal dialysis catheters, renal transplant

Also look for signs of treatment:

- Scars from pleural taps and chest drains



## INVESTIGATIONS

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- CXR, pleural tap (should be US guided) sent for lactate dehydrogenase (LDH), protein, pH, amylase, glucose, cytology, microscopy and culture

## MANAGEMENT

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- Dependent on underlying cause and the treatment for that (e.g. infection, lung cancer), but can include the following options: therapeutic aspiration, chest drain insertion, thoracoscopy, long-term drain insertion and possible surgical interventions for an empyema

## QUESTIONS

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1. How would you differentiate an exudative effusion from a transudate?
  - An exudative effusion is defined by
    - An effusion albumin/plasma albumin ratio  $>0.5$
    - An effusion LDH/plasma LDH ratio  $>0.6$
    - A pleural fluid LDH  $>2/3$  the upper limit of normal serum LDH (Light's criteria)
2. What are the causes of an exudative and transudative effusion?
  - Exudative
    - The 4 I's: Infiltration (neoplasm), infection, infarction (pulmonary embolus), inflammation (rheumatoid arthritis and SLE)
  - Transudative
    - Cardiac failure, chronic renal disease, chronic liver disease, other rarer causes include Meigs' syndrome
3. List some drugs that may cause a pleural effusion.
  - Amiodarone
  - Phenytoin
  - Methotrexate
  - Nitrofurantoin
  - Beta-blockers
4. What is the main clinical indication for a thoracoscopy?
  - The main indication for a thoracoscopy is to investigate an exudative effusion of uncertain cause, as it yields a better diagnostic rate than a pleural tap and pleural biopsies can also be taken at the same time. A thoracoscopy can also be used for therapeutic purposes with drainage of the effusion and pleurodesis during the procedure.

## KEY POINTS

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- Be able to differentiate between effusion and collapse (stony-dull percussion in an effusion)
- Be aware of the causes of a pleural effusion and be familiar with Light's criteria for classifying pleural effusions.
- Look for an underlying cause for the effusion.

## PATIENT WITH PREVIOUS LUNG SURGERY

This patient is complaining of a cough; please examine their respiratory system.

### FINDINGS

- *General:* Dyspnoea, reduced chest expansion, cachexia
- *Peripheral:* Finger clubbing, peripheral and central cyanosis, reduced muscle bulk under scar site, tracheal deviation towards the side of surgery (more obvious in pneumonectomy)
- *Chest:* Thoracotomy scar, scars from drain sites, reduced expansion on affected side, dull percussion note on the affected side, decreased breath sounds on affected side

### PRESENTATION

This patient has evidence of a right-sided pneumonectomy as evidenced by the lateral thoracotomy scar, reduced chest expansion and absence of breath sounds. The absence of breath sounds suggests that the procedure has been a pneumonectomy as opposed to a lobectomy.

- Comment on the possible reason for surgery.
  - Lung cancer: Clubbing, cachexia, Horner's syndrome, radiation scars and tattoos, lymphadenopathy
  - Evidence of old TB: COPD (could have been treated with surgery for excision of large bullae and lung reduction surgery)
  - Keyhole scars from video-assisted thoracoscopic surgery (VATS)
- Note: The other scenario in this station is that you have a patient who has had a lung transplant; if this is the case, you may find that the only abnormality is a 'clamshell' scar.

### QUESTIONS

1. How would you differentiate between a lobectomy and a pneumonectomy?
  - Pneumonectomy
    - The trachea is deviated away towards the side of surgery.
    - Decreased breath sounds (or no sounds) over the whole lung field.
    - Reduced chest expansion.
  - Lobectomy
    - Trachea may be shifted away from the side of surgery.
    - Audible breath sounds from the lobes that have not been operated on.
    - Chest expansion may be reduced.





2. What are the criteria for lung surgery in lung cancer?
  - Patients must have an FEV1 >1.5l, a transfer factor >50%, no evidence of severe pulmonary hypertension and no evidence of metastatic disease (surgery is beneficial only in peripheral non-small-cell disease).
3. What are the indications for a lung transplant?
  - Patients with emphysema (usually with alpha-1 antitrypsin deficiency), idiopathic pulmonary fibrosis, idiopathic pulmonary hypertension, bronchiectasis and cystic fibrosis may be considered for surgery.
4. What are the absolute contraindications for lung transplantation?
  - Recent malignancy in the past 2 years
  - Substance abuse (alcohol, smoking)
  - Chest wall deformity
  - Poor social support
  - Psychiatric illness
  - Advanced extrapulmonary organ dysfunction
  - Noncurative infections: HIV

## KEY POINTS

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- Be able to recognise the range of thoracic scars and look for clues to guide to the underlying cause.
- Be aware of indications for lung transplantation and the major contraindications.

## REFERENCE

---

British Thoracic Society. (2001) Guidelines on the selection of patients with lung cancer for surgery. *Thorax* 56: 89–108.

## RESPIRATORY STATION SUMMARY

- Observe the patient from the end of the bed and count the respiratory rate.
- Take a good look around the bedside and pick up on any clues that may help aid your diagnosis, e.g. sputum pot, inhalers, oxygen mask.
- Look at the inhalers to elicit their contents.
- Ensure that you have time to examine both the front and the back of the chest in your examination – concentrate on the back, as you are more likely to elicit findings and especially if you are running out of time (although this should not be the case for a well-practiced candidate).
- Be able to differentiate between consolidation and an effusion on examination.
- Look for any scars which may aid you with the diagnosis.
- If you think the diagnosis is bronchiectasis and the patient has bibasal coarse crackles, ask them to cough to see if the crackles clear.
- Be able to differentiate the causes of upper- and lower-zone fibrosis.
- In cases such as pulmonary fibrosis and pleural effusion, look for an underlying cause for the pathology.
- If you see a surgical scar, look for clues such as a radiotherapy tattoo to help guide your diagnosis.
- Familiarise yourself with pulmonary function test results for common respiratory conditions such as COPD.
- Be aware of British Thoracic Society (BTS), NICE and Scottish Intercollegiate Guidelines Network (SIGN) guidelines to help with the diagnosis and treatment of patients.

# 2

## Station 1: Abdominal

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## HINTS FOR THE ABDOMINAL STATION

- Read the candidate information carefully and look closely from the end of the bed for any signs that may help guide you to the specific examination; renal/haematology/liver/general abdominal.
- For a renal examination, look for evidence of renal replacement therapy (RRT). Always check for arteriovenous (AV) fistulae along the arms and look for fresh venepuncture marks at a fistula site. Also look for evidence of previous internal jugular vein lines or scars from peritoneal dialysis (PD) catheter insertions. Don't miss nephrectomy and pelvic transplant scars.
- For a liver exam, ensure that you comment on the presence of finger clubbing, leuconychia, palmar erythema, Dupuytren's contractures and tattoos if present.
- Look in the oral cavity; gum hypertrophy in ciclosporin toxicity, poor dentition in chronic liver disease and glossitis in anaemia.
- If you find evidence of any cervical lymphadenopathy, always go on to check for axillary and inguinal lymphadenopathy.
- Ensure that the patient is adequately exposed and that they are lying as flat as possible prior to palpating the abdomen.
- Always make constant eye contact with the patient when palpating their abdomen to ensure that they are not in any discomfort. Apologise if you do cause any discomfort to the patient.
- Do not panic if you cannot elicit any abnormal findings; the patient may have a normal abdomen. In this case, look for peripheral signs which may help guide you towards a diagnosis.
- If you elicit any hepatomegaly or splenomegaly, always try to comment on the size of the organomegaly. In a case of splenomegaly, this may aid you with your differential diagnosis.
- Look for evidence of peripheral oedema.

## ABDOMINAL CASE WITH A NORMAL ABDOMEN

Please examine this patient's abdomen.

### FINDINGS

- *General:* Cachexia
- *Peripheral:* Oral/perioral telangiectasia, buccal pigmentation, hypertension, facial flushing, lymphadenopathy, cushingoid features, signs of RRT
- *Abdomen:* No abnormality

If you finish your examination and have no positive findings, it is likely that you are in one of the three following scenarios:



1. You have been given a patient with a normal abdomen.
2. You may have missed a peripheral sign that points to a diagnosis such as hereditary haemorrhagic telangiectasia (HHT), Peutz–Jeghers syndrome, generalised lymphadenopathy, Cushing’s syndrome, carcinoid syndrome or a patient with end-stage renal disease (ESRD) on RRT
3. You may have missed organomegaly.

## PRESENTATION

---

This patient has oral telangiectasia and looks clinically anaemic. There is nothing abnormal to find in the abdomen. The diagnosis may be hereditary haemorrhagic telangiectasia. I would like to know this patient’s full blood count (FBC) and ask if there is a family history of this disorder.

## QUESTIONS

---

1. What is the mode of inheritance of Peutz–Jeghers syndrome?
  - Autosomal dominant, caused by mutations in the STK11 (also known as LKB1), which is a tumour suppressor gene
2. What are the manifestations of Peutz–Jeghers syndrome?
  - This condition is characterised by dark-coloured freckling from mucocutaneous lesions predominantly on the face (around lips and mouth), oral mucosa and peripheries. It also causes polyps within the gastrointestinal (GI) tract which may be complicated by bleeding, intestinal obstruction and chronic pain and have a high risk of malignancy during their lifetime (particularly breast, colorectal, pancreatic, stomach ovarian, lung and small bowel).
3. What is the mode of presentation of hereditary haemorrhagic telangiectasia?
  - Recurrent epistaxis in childhood with red spots on lips, tongue, fingertips  $\pm$  family history.
  - It is also known as Osler–Weber–Rendu syndrome.
  - It is characterised by multiple telangiectasia.
  - The patient is at risk of haemorrhage from AV malformations, particularly pulmonary and cerebral.
4. What is the mode of inheritance of hereditary haemorrhagic telangiectasia?
  - It has an autosomal dominant mode of inheritance. More than 80% of cases are due to mutations in ENG or ACVRL1 genes. MADH4 gene mutations have also been reported.

## KEY POINTS

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- If you are unable to elicit any positive findings, you may have been given a ‘normal abdomen’ as your case.
- Ensure that you look carefully for the presence of any peripheral signs to help guide your diagnosis.
- Do not make up any findings.

## CHRONIC LIVER DISEASE

Please examine this patient who has presented with abdominal swelling.

### FINDINGS

- *General:* Cachexia, poor dentition, muscle wasting, loss of body hair, gynaecomastia, testicular atrophy, tattoos, pedal oedema
- *Skin:* Excoriation marks, purpura, spider naevi (most likely to be seen on the chest/back)
- *Hands:* Clubbing, palmar erythema, leuconychia, Dupuytren's contractures
- *Face:* Icterus, parotid swelling, pallor
- *Abdomen:* Caput medusae, ascites, hepatomegaly, hepatic bruit, splenomegaly, evidence of previous ascitic taps/drains

### PRESENTATION

On examination, this patient has evidence of chronic liver disease. There is leuconychia and palmar erythema. The patient is icteric and there are excoriation marks. There is marked ascites with prominent superficial veins over the abdominal wall.

This is a common presentation, so be sure to look for an underlying cause. The most common cause is alcoholic liver disease (ALD).

### CAUSES

- ALD
  - Parotid swelling, cachexia, Dupuytren's contracture
- Viral hepatitis
  - Tattoos, injection sites
- Wilson's disease
  - Kayser-Fleischer rings, Parkinsonism, cognitive impairment, features of heart failure
- Haemochromatosis
  - Slate-grey pigmentation, evidence of diabetes mellitus, features of heart failure, arthritis
- Alpha-1 antitrypsin deficiency
  - Shortness of breath, hyperinflated chest, evidence of cor pulmonale, clubbing (bronchiectasis)

### INVESTIGATIONS

- Bloods
  - FBC, urea and electrolytes (U&Es), liver function tests (LFTs) including albumin, gamma-glutamyl transpeptidase (GGT), coagulation screen, hepatitis serology, caeruloplasmin, ferritin, autoantibodies (antimitochondrial antibody [AMA],



anti-smooth muscle, anti-liver kidney microsomal [LKM], antinuclear antibody [ANA], alpha-fetoprotein (AFP), thyroid function tests (TFTs), coeliac screen, alpha-1 antitrypsin levels, glucose

- Ascitic fluid
  - Gram stain and cell count ( $>250$  white cells/mm<sup>3</sup> is indicative of spontaneous bacterial peritonitis [SBP]), protein concentration, culture
- Imaging
  - Ultrasound scan (USS): Check for hepato-/splenomegaly, confirm ascites
  - Doppler flow studies of the hepatic/portal vein: Rule out thrombosis
  - Computerised tomography (CT) abdomen (adds little if USS is normal)

## MANAGEMENT

---

Dependent on the underlying cause

- Alcohol
  - Alcohol avoidance, diuretics, vitamin B compound, thiamine, oesophagogastroduodenoscopy (OGD) to look for oesophageal varices
- Hepatitis
  - Hepatitis C: Antiviral agents
- Wilson's disease
  - Chelation: Penicillamine, trientine
- Haemochromatosis
  - Venesection (to a ferritin level of below 50 µg/L)
- Alpha-1 antitrypsin deficiency
  - Generally supportive; advise not to smoke. Alpha-1 antitrypsin may be an option.

All these patients should be considered for liver transplantation on an individual basis if severity dictates.

## QUESTIONS

---

1. What are the signs of decompensated chronic liver disease?
  - Encephalopathy: Grade 1 (altered mood/behaviour) to grade 4 (coma)
  - Asterixis
  - Jaundice
  - Ascites
  - Hepatic fetor
  - Constructional apraxia: Unable to draw a five-pointed star
2. What are the reversible causes of hepatic encephalopathy?
  - Alcohol
  - Drugs
  - GI haemorrhage
  - Infection
  - Constipation

## 3. How is ascites investigated?

- The protein level of the ascites is used to split the aetiology by transudative and exudative causes. However, the serum ascites-albumin gradient (SA-AG) is more accurate for diagnosis of the cause. SA-AG is calculated as 'serum albumin concentration minus ascites albumin concentration'.
- SA-AG  $\geq 11$  g/L
  - Cirrhosis, cardiac failure, nephrotic syndrome
- SA-AG  $< 11$  g/L
  - Malignancy, pancreatitis, tuberculosis

Further investigations are also helpful for diagnosis:

- Neutrophil count and microscopy and culture for spontaneous bacterial peritonitis (neutrophil count  $> 250$  cells/mm<sup>3</sup> is diagnostic of SBP)
- Amylase for pancreatitis
- Cytology for malignancy

## 4. What are the possible indications for liver transplantation in an adult?

- Acute causes
  - Paracetamol poisoning (acetaminophen poisoning)
  - Other drugs, e.g. isoniazid, phenytoin, sodium valproate
  - Acute hepatitis
  - Epstein–Barr virus (EBV) and cytomegalovirus (CMV)
- Chronic causes
  - Alcoholic liver disease
  - Primary biliary cirrhosis
  - Primary sclerosing cholangitis
  - Chronic viral hepatitis
  - Wilson's disease
  - Budd–Chiari syndrome
  - Hepatic malignancy

## KEY POINTS

---

- Look out for examination findings which will guide you to the underlying diagnosis.
- In chronic liver disease, many patients will have a shrunken cirrhotic liver rather than hepatomegaly.
- Be able to grade the various stages of hepatic encephalopathy.

## REFERENCE

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Moore KP, Aithal GP. (2006) Guidelines on the management of ascites in cirrhosis. *Gut* 55: 1–12.





## GENERALISED LYMPHADENOPATHY

Please examine this patient's abdominal system.

### FINDINGS

---

- *General:* Evident lumps and bumps, purpura, pallor, cachexia
- *Peripheral:* Lymphadenopathy (cervical, supraclavicular, axillary), arthritis, enlarged tonsils
- *Abdomen:* Splenomegaly, hepatomegaly, inguinal lymph nodes

### PRESENTATION

---

This patient has marked lymphadenopathy. There are palpable nodes in the cervical, axillary and inguinal regions. There are purpuric lesions present on both arms. This could be a lymphoproliferative disorder with thrombocytopenia.

A difficult case, this could be picked up when examining the neck or following palpation of an enlarged spleen (and possibly liver). If following examination of the abdomen you feel that the patient could have a lymphoproliferative disorder, do not be afraid to re-examine the neck and axillae for nodes.

### INVESTIGATIONS

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- Diagnosis
  - Blood tests (FBC, lactate dehydrogenase [LDH], blood film, viral screen, autoimmune screen, LFTs), lymph node biopsy, bone marrow aspirate and trephine, chest X-ray and sputum for acid-fast bacilli
- Staging
  - CT/positron emission tomography (PET), lumbar puncture

### MANAGEMENT

---

- Dependent on the cause.
- The main objective of treatment of high-grade lymphoproliferative disease is to achieve a cure.
- Long-term remission may be achieved in low-grade disease.
- It is also important to manage symptoms accordingly.

## QUESTIONS

---

1. What is the differential diagnosis of generalised lymphadenopathy?
  - Lymphoproliferative disease: Chronic lymphocytic leukaemia (CLL), acute lymphoblastic leukaemia, Hodgkin's and non-Hodgkin's lymphoma
  - Viral disease: Includes human immunodeficiency virus (HIV), EBV and CMV
  - Other infections: Includes tuberculosis, brucellosis and toxoplasmosis
  - Inflammatory disease: Sarcoidosis, rheumatoid arthritis, systemic lupus erythematosus (SLE)
2. How are Hodgkin's and non-Hodgkin's lymphoma differentiated pathologically?
  - Through the presence of Reed–Sternberg cells in Hodgkin's lymphoma. These are characteristic binucleate cells found on light microscopy of a biopsy.
3. What are 'B-symptoms' and what are their significance?
  - These include weight loss (>10% in 6 months), unexplained fever >38.0°C and night sweats. B-symptoms are included in the Ann Arbor staging classification of non-Hodgkin's and Hodgkin's lymphoma and indicate a poorer prognosis.
4. Tell me about CLL.
  - This is due to a monoclonal proliferation of lymphocytes, usually B-cells.
  - It is most commonly suspected from a routine blood test, with a raised white cell count (lymphocytosis). It may also present with signs of bone marrow failure (fatigue, infections, bleeding) or constitutional symptoms (such as weight loss, fevers, fatigue and malaise).
  - Investigations may include a CT scan, lymph node biopsy and bone marrow biopsy. CLL is staged by the Binet system (Stage A: <3 groups of enlarged lymph nodes, no anaemia or thrombocytopenia; Stage B: ≥3 groups of enlarged lymph nodes, no anaemia or thrombocytopenia; Stage C: anaemia and/or thrombocytopenia). Cytogenetic testing can give information on how the disease is likely to progress, and can therefore guide treatment.
  - Stage A disease can initially be managed with a watch and wait approach. More advanced disease requires earlier intervention with cytotoxic chemotherapy (such as fludarabine and cyclophosphamide) and monoclonal antibodies (such as rituximab, a CD20 antibody). B-cell receptor signalling pathway inhibitors such as ibrutinib and idelalisib are also now available. Rarely, a bone marrow transplant is carried out.
  - Complications of CLL include bone marrow failure, autoimmune haemolytic anaemia, recurrent chest infections and acute transformation (Richter's syndrome).

## KEY POINTS

---

- When examining a seemingly normal abdomen, do not forget to feel for inguinal lymph nodes.
- Be able to give a wide differential for lymphadenopathy.
- Remember that a haematological cause is not the only cause of lymphadenopathy.



## REFERENCES

---

- Binet JL, et al. (1981) A new prognostic classification of chronic lymphocytic leukemia derived from a multivariate survival analysis. *Cancer* 48(1): 198–206.
- Woyach JA, Johnson AJ. (2015) Targeted therapies in CLL: Mechanisms of resistance and strategies for management. *Blood* 126(4): 471–7.

## HEPATOSPLENOMEGALY

Please examine this patient's abdominal system.

## FINDINGS

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- *General*: Purpura, pallor, jaundice
- *Peripheral*: Enlarged tonsils, lymphadenopathy (cervical, supraclavicular, axillary), features of chronic liver disease
- *Abdomen*: Splenomegaly (note degree of enlargement), hepatomegaly, ascites, inguinal lymph nodes

## PRESENTATION

---

This patient has 10 cm splenomegaly, with 4 cm hepatomegaly. There is no evident lymphadenopathy and no peripheral features of chronic liver disease. The most likely diagnosis is a myeloproliferative disorder.

A common case, the most likely causes are chronic liver disease and haematological malignancy; be sure to look for signs of both of these. After the enlarged liver and spleen have been palpated, reexamine for peripheral signs if necessary.

## INVESTIGATIONS

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If the diagnosis is chronic liver disease, investigate as necessary (see chronic liver disease case). Otherwise, consider the following tests:

- Blood tests
  - FBC, LDH, blood film, viral screen, autoimmune screen, LFTs, renal function, serum and urine electrophoresis
- Imaging
  - Abdominal ultrasound, CT/PET scan
- Invasive
  - Lymph node biopsy, bone marrow aspirate and trephine, other biopsy site (e.g. renal biopsy for amyloidosis)

## MANAGEMENT

---

- Dependent on the cause

## QUESTIONS

---

1. What is the differential diagnosis of hepatosplenomegaly?
  - Chronic liver disease with portal hypertension
  - Lymphoproliferative disease: Chronic lymphocytic leukaemia, Hodgkin's and non-Hodgkin's lymphoma (in acute lymphoblastic leukaemia the spleen is not usually greatly enlarged)
  - Myeloproliferative disease: Chronic myeloid leukaemia (CML) (and acute myeloid leukaemia), polycythaemia rubra vera (PRV), essential thrombocythaemia and myelofibrosis
  - Other haematological disease: Thalassaemia, sickle cell disease
  - Viral disease: Includes HIV, EBV, CMV and hepatitis B/C (may cause chronic liver disease)
  - Other infections: Includes malaria, brucellosis, toxoplasmosis and leptospirosis
  - Inflammatory disease: Sarcoidosis
  - Infiltrative disease: Glycogen storage disease, amyloidosis
  
2. What are the myeloproliferative disorders?
 

A group of conditions caused by abnormal myeloid stem cell proliferation in the bone marrow. They are generally distinguished from each other by the type of cell which is most affected:

  - Red blood cells: Polycythaemia rubra vera
  - White blood cells: Chronic myeloid leukaemia
  - Platelets: Essential thrombocythaemia
  - Fibroblasts: Myelofibrosis
  
3. How might a patient with CML present, and how would you diagnose this?
  - Chronic myeloid leukaemia is the uncontrolled growth of myeloid cells. Therefore, red cells, white cells and platelets can all be affected. It could present as a result of the effect on these cells (fatigue, infection, bleeding), or with constitutional symptoms, abdominal pain (splenomegaly) or by chance.
  - For diagnosis, blood tests (raised white cell count) are relevant, but cytogenetic testing of bone marrow is usually required. On such chromosome analysis, the Philadelphia chromosome (a translocation between chromosomes 9 and 22, resulting in oncogenic fusion of the BCR-ABL1 gene [a type of tyrosine kinase]) is present in 95% of cases of chronic myeloid leukaemia. This chromosomal translocation forms the basis of targeted treatment with tyrosine kinase inhibitors (such as imatinib).
  
4. Tell me about amyloidosis.
  - This is a multisystem disease that results from extracellular deposition of abnormal proteins.
  - There are two main types: amyloid L (AL) and amyloid A (AA).
  - AL results from abnormal light-chain production and most commonly occurs on its own, although it can also be seen alongside myeloma. Deposition can affect the heart (cardiomyopathy and heart failure), kidneys (renal failure), nerves (peripheral neuropathy), gut (malabsorption) and clotting function.



It is mainly diagnosed on biopsy, although a serum amyloid P (SAP) scan is sometimes carried out. Treatment is similar to that for myeloma. The complications can also be treated (for example, with a renal transplant).

- AA results from abnormal deposition of serum amyloid A (SAA) protein, which is an acute-phase protein. It occurs as a secondary process to chronic inflammatory disorders (such as rheumatoid arthritis and familial Mediterranean fever) and chronic infections. It most commonly affects the liver, spleen and kidneys. It is again diagnosed by biopsy. The condition often improves with treatment of the underlying inflammatory disorder.

## KEY POINTS

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- Aim to decide between liver and haematological disease when giving a primary diagnosis.
- Look for clues of these two groups throughout the examination.
- Don't be afraid to re-examine for peripheral signs if necessary.

## REFERENCE

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Pinney JH, Hawkins PN. (2012) Amyloidosis. *Ann Clin Biochem* 49(Pt 3): 229–41.

## MULTIPLE ABDOMINAL SCARS

This patient has had an operation. Please examine their abdomen.

## FINDINGS

---

- *General*: Cachexia, cushingoid features
- *Peripheral*: Oral ulceration, pallor, finger-prick marks (signs of glucose testing), gum hypertrophy and hypertension (side effects of ciclosporin)
- *Abdomen*:
  - Multiple surgical scars (most commonly midline)
  - Stoma sites (past/current)
  - Evidence of previous PD catheter exit site or removal (see case on RRT) or repeated ascitic drainage
  - Mention that you would wish to perform a rectal examination
  - Comment on nutritional state

## PRESENTATION

---

This patient has inflammatory bowel disease (IBD) as evidenced by cachexia, clinical anaemia and multiple scars on the abdomen suggesting fistulae, bowel obstruction or abscess drainage. A per rectal examination may be useful to look for fistulae and abscesses.

## INVESTIGATIONS

- Stool cultures × 3 (microscopy/culture/*Clostridium difficile* toxin [DCT]) (in acute presentations).
- Bloods including FBC, C-reactive protein (CRP), LFTs (evidence of associated liver disease, e.g. autoimmune hepatitis primary biliary cirrhosis).
- A faecal calprotectin test (evaluates bowel inflammation); a negative test rules out inflammatory bowel disease.
- Abdominal X-ray to rule out toxic megacolon (in acute presentations).
- Sigmoidoscopy/colonoscopy and biopsy (for histological confirmation).
- Magnetic resonance imaging (MRI) enterocolysis (MRI of the small bowel) or bowel ultrasound (in expert hands) can be used if Crohn's is suspected and to detect small bowel strictures.

## MANAGEMENT

- Medical
  - Immunosuppression including steroids, 5-aminosalicylic acid (ASA), disease-modifying agents such as azathioprine/mercaptopurine/methotrexate and biological agents
- Surgical
  - Indications include fistulae, strictures and failure to respond to medical therapy
- Nutritional support and elemental and low-residue diets
- Psychological support

## QUESTIONS

1. What are the differences between ulcerative colitis (UC) and Crohn's disease?

### Differences between Crohn's Disease and Ulcerative Colitis

	Crohn's disease	Ulcerative colitis
Distribution	Patchy ('skip lesions')	Continuous
Depth	Transmural	Superficial
Area involved	Whole gastrointestinal tract with predilection for terminal ileum and anus	Large bowel with predilection for rectum
Smoking	Higher risk	Lower risk
Fistulae and stenosis	Common	Rare

2. What are the extra-intestinal manifestations of inflammatory bowel disease?
  - Skin: Erythema nodosum, pyoderma gangrenosum, aphthous ulceration
  - Joints: Seronegative arthritides (mostly large-joint arthritis and sacroiliitis)
  - Eye: Uveitis, episcleritis/scleritis, conjunctivitis
  - Hepatobiliary: Primary sclerosing cholangitis (more likely in UC), cholangiocarcinoma
  - Renal: Oxalate stones

3. What screening tests should you consider before initiating biological therapies?
  - History and examination (looking specifically for features of tuberculosis)
  - Hepatitis serology
  - HIV test
  - Chest X-ray (CXR)
  - T-spot test (QuantiFERON® test is adequate if patient is not on steroids)
4. What biological agents are available for the treatment of IBD?
  - Ulcerative colitis
    - Infliximab and golimumab are recommended by the National Institute for Health and Care Excellence (NICE) for the treatment of moderate to severe active UC in adults who have had a poor response to conventional therapy (including steroids and mercaptopurine or azathioprine), or if they have been unable to tolerate or have contraindications to such therapy.
  - Crohn's disease
    - Infliximab and adalimumab are recommended by NICE for adults with severe active Crohn's disease who have not responded to conventional therapy, or who have been unable to tolerate or have contraindications to conventional therapy.
  - A novel agent called vedolizumab is now available for those who fail first-line treatment; this is for both UC and Crohn's disease.

## KEY POINTS

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- Look for the extra-intestinal manifestations of inflammatory bowel disease to help guide the underlying diagnosis.
- Familiarise yourself with the key treatments available for the management of these conditions.
- Be aware that midline scars may not just be a sign of IBD – see case on RRT, if there's a midline scar – do not miss a kidney–pancreas transplant.

## REFERENCES

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- National Institute for Health and Care Excellence. (2012) Guidance CG152. Crohn's disease: Management. Available from <https://www.nice.org.uk/guidance/cg152>. Accessed 2 May 2016.
- National Institute for Health and Care Excellence. (2013) Guidance CG 166. Ulcerative colitis: Management. Available from <https://www.nice.org.uk/guidance/cg166/chapter/1-recommendations>. Accessed 2 May 2016.
- National Institute for Health and Care Excellence. (2015) Guidance TA329. Infliximab, adalimumab and golimumab for treating moderately to severely active ulcerative colitis after the failure of conventional therapy. Available from <https://www.nice.org.uk/guidance/ta329/chapter/1-Guidance>. Accessed 2 May 2016.
- Parente F, et al. (2002) Bowel ultrasound in assessment of Crohn's disease and detection of related small bowel strictures: A prospective comparative study versus x ray and intraoperative findings. *Gut* 50(4): 490–5.

## PALPABLE KIDNEYS

This patient has presented with haematuria. Please examine their abdomen.

### FINDINGS

- *General:* Signs of anaemia, hypertension and renal replacement therapy
- *Arms:* Arteriovenous fistulae/grafts
- *Face:* Gum hypertrophy from immunosuppressants
- *Fundoscopy:* Hypertensive retinopathy and evidence of Von Hippel–Lindau syndrome
- *Chest/neck:* Scars from previous tunnelled dialysis catheters
- *Abdomen:* Enlarged kidneys (bilateral flank masses), hepatomegaly (associated with autosomal dominant polycystic kidney disease [ADPKD]), renal transplant and nephrectomy scars
- *Legs:* Oedema

### PRESENTATION

The diagnosis is autosomal dominant polycystic kidney disease. On inspection, there is the appearance of fullness in the flanks. On palpation, there are bilateral bimanually ballotable masses in the flanks. It is possible to get above the masses, and each moves with respiration.

### TIPS

Ensure that you comment on any evidence of renal replacement therapy, e.g. the presence of a tunnelled vascular catheter, a peritoneal dialysis catheter or an AV fistula in the arms. In addition, on abdominal examination you may note a J-shaped renal transplant scar in the iliac fossa and/or a subsequent midline scar from where the enlarged kidneys may have been removed.

### INVESTIGATIONS

- Urinalysis
  - Haematuria and proteinuria, signs of infection
- Ultrasound abdomen
  - Gold standard investigation to assess renal size, cysts, obstruction, liver cysts
- CT abdomen
  - To look for malignancy if clinically indicated
- MRI
  - To assess renal size and volume (if indicated)
- Echocardiogram
  - Examine for mitral valve prolapse (MVP), aortic disease and assess ventricular function





- Cerebral angiogram
  - Evidence of berry aneurysms associated with ADPKD.
  - Only indicated if a first-degree relative with ADPKD had a subarachnoid haemorrhage/bleeding aneurysm. Routine scanning in all is not indicated.
- Genetic testing
  - Autosomal dominant polycystic kidney disease (ADPKD 1 and 2)

## MANAGEMENT

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- Counselling
- Regular surveillance
- Monitor renal function, treat hypertension and manage chronic kidney disease (CKD) and complications
- Consider nephrectomy if appropriate
- Vasopressin antagonists – tolvaptan – inhibits binding of vasopressin to V2 receptors, reducing cell proliferation, cyst formation and fluid excretion. NICE recommends this if PKD with CKD stage 2 or 3 at the start of treatment with evidence of rapidly progressive disease
- Renal replacement therapy if required
- Investigation of first-degree relatives

## QUESTIONS

---

1. What is the genetic basis of polycystic kidney disease?
  - ADPKD
    - Affects adults
    - 75% of patients have liver cysts by the seventh decade
    - Chromosome 16 mutation in PKD 1: Accounts for 85% of cases; carries highest risk of developing ESRD
    - Chromosome 4 mutation in PKD 2: Accounts for ~15% of cases; ESRD develops later than with PKD1
  - Autosomal recessive PKD (ARPKD)
    - More severe liver involvement than ADPKD
2. List some other manifestations of ADPKD.
  - Cystic disease
    - Liver, spleen, pancreas
  - Berry aneurysm
    - Risk of subarachnoid haemorrhage (SAH) if ruptures (treatment includes blood pressure [BP] control, lipid lowering and smoking cessation)
  - Pain/haematuria
    - If ruptured cyst, stone, infection or renal cell carcinoma

- Renal cell carcinoma
    - Risk of malignant cyst transformation
  - Valvular disease
    - MVP, aortic valve disease
  - Hypertension
    - LVH
  - Gastrointestinal
    - Colonic diverticulum formation, herniae
3. What are the indications for nephrectomy in ADPKD?
- Recurrent infections
  - Chronic pain
  - Recurrent haematuria
  - GI pressure symptoms, e.g. early satiety
  - Size/creating space for transplantation
4. List some other renal cystic disorders.
- Unilateral
    - Benign renal cysts
    - Renal cell carcinoma
    - Polycystic kidney disease
  - Bilateral
    - Bilateral renal cell carcinoma
    - Amyloidosis
    - Von Hippel–Lindau syndrome (see below for further information regarding this condition)
    - Tuberous sclerosis

### **Von Hippel–Lindau syndrome**

Autosomal dominant condition

Caused by mutations in the VHL gene (tumour suppressor gene)

Features

- Angiomata develop in retina (may develop retinal haemorrhages and cause visual loss if left untreated), brain and spinal cord, liver, kidney and pancreas
- Cerebellar haemangioblastoma (lateral lobes)
- Pheochromocytoma
- Renal cell carcinoma
- Endolymphatic sac tumors (tumours of the inner ear, causing hearing loss, tinnitus and balance problems)
- Usually requires annual screening of eyes, kidneys and urinary peptides (for pheochromocytoma), intermittent brain imaging

### **KEY POINTS**

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- Look for any scars that may indicate a nephrectomy.
- Look for any signs of renal replacement therapy.
- Know the causes of cystic renal disease.



## RENAL REPLACEMENT THERAPY

Please examine this patient's abdomen.

### FINDINGS

- *General*: Cushingoid appearance
- *Hands*:
  - Evidence of finger-prick testing: Suggestive of diabetes (could be the cause of ESRD or have arisen post-transplant)
  - Fine tremor: Could signify tacrolimus toxicity
- *Arms*: AV fistulae (radiocephalic [seen at the wrist], brachiocephalic [seen around the antecubital fossa] or brachiocephalic [palpable medially from around the antecubital fossa (ACF) to towards the axilla]): Feel for thrills *and* listen for a bruit to determine if still functional; look for fresh venepuncture marks. If there is no palpable thrill but a bruit is audible, this could signify a synthetic graft.
- *Face*: Corneal arcus, gum hypertrophy (secondary to ciclosporin)
- *Neck/chest*: Scars from internal jugular vein catheters, tunnelled-catheter exit site scar, parathyroidectomy scar (look carefully for this: it will be hidden in the crease at the base of the neck)
- *Abdomen*:
  - Peritoneal dialysis catheter or scars from previous catheters (look for previous exit site scars: usually lateral to the umbilicus, and if removed, there may be a small longitudinal scar infra-umbilically).
  - Midline laparotomy scar or posterior subcostal scar: Evidence of previous nephrectomy (possibly bilateral).
  - Note that a midline laparotomy scar could also be present if the patient has had a simultaneous kidney–pancreas transplant (usually the renal graft is situated in the left iliac fossa in these patients).
  - Palpable mass in iliac fossa (graft may have been removed, so mass may not be palpable; scars can be very faint).

### PRESENTATION

This patient has had a renal transplant as evidenced by the presence of a renal transplant in the right iliac fossa (RIF). This is a functioning transplant as the patient does not have evidence of recent dialysis (fistula not recently needled/no PD catheter present) or fluid overload.

Comment on the following:

- Renal replacement therapy
- Any other forms, e.g. vascular catheter, PD catheter, AV fistula (are they being actively used?)
- Fluid status
- Signs of fluid overload (raised jugular venous pressure [JVP], peripheral oedema)
- Immunosuppression
  - Ciclosporin: Gum hypertrophy
  - Corticosteroids: Cushingoid features
  - Skin lesions: Suggestive of previous skin cancers/removals (squamous cell carcinomas [SCCs]/basal cell carcinomas [BCCs])

Try to link with a possible underlying cause for renal failure:

- Diabetes
  - Injection sites on abdomen/lipodystrophy, pinprick marks on fingertips
- ADPKD
  - Bilateral ballotable masses in flanks
- Alport's syndrome
  - Hearing aids

## MANAGEMENT

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- Maintenance immunosuppression usually consists of a calcineurin inhibitor (CNI) (tacrolimus or ciclosporin) and an antiproliferative (azathioprine or mycophenolate mofetil), with or without steroids.
- Side effects include (this is not an exhaustive list of complications; see British National Formulary [BNF] for further details)
  - Tacrolimus (calcineurin inhibitor): Tremor, alopecia, diabetes, hypertension, renal impairment, renal failure, blood disorders
  - Ciclosporin A, as for tacrolimus except gingival hypertrophy, hypertrichosis, hyperlipidaemia,
  - Mycophenolate mofetil: Diarrhoea, nausea, vomiting, leucopenia, lymphopenia, anaemia, thrombocytopenia, foetal toxicity, infections, malignancy (especially skin)
  - Azathioprine: Hypersensitivity reactions, dose-related bone marrow suppression, liver impairment, infections, pancreatitis
  - Prednisolone: Thin skin, easy bruising, muscle wasting/weakness, diabetes, hypertension, osteoporosis, pancreatitis, infections, glaucoma, cataracts, heart failure
  - Rapamycin (mTOR) inhibitors (sirolimus, everolimus): Hypertension, impaired healing, venous thromboembolism (VTE), interstitial lung disease, proteinuria, hyperlipidaemia, reversible male infertility
- All these except prednisolone give an increased risk of malignancy.
- Screening should be similar to that for the general population, e.g. breast, colon screening.
- Annual skin checks should be undertaken with a dermatologist for signs of SCC and BCC.

## QUESTIONS

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1. What are the most common causes for a renal transplant?
  - Diabetes mellitus
  - Glomerulonephritis
  - ADPKD
  - Hypertension
2. What investigations would you request in a patient with a renal transplant admitted with a rise in serum creatinine?

- Blood tests: Renal function, FBC, bone profile, LFTs, CRP if signs of infection + blood cultures/virology (e.g. CMV, BK virus polymerase chain reaction [PCR]), immunosuppression levels to check for toxicity
  - Urine: Dipstick (check for haematuria, proteinuria, leucocytes and nitrites), urine microscopy, culture and sensitivity, urine for quantification of proteinuria (if applicable), virology for BK virus
  - Ultrasound scan of the transplant with Dopplers of the vessels: To assess for obstruction, renal perfusion, possible renal artery stenosis and renal vein thrombosis
  - Transplant biopsy: If no other cause found, to rule out rejection and look for any other cause
3. What are the signs of a failing transplant?
- Progressively declining renal function
  - Proteinuria
  - Tenderness over the transplant graft
  - Fluid overload
  - Interstitial fibrosis, tubular atrophy or vascular changes in a renal transplant biopsy
4. What do you know about the manifestations of Alport's syndrome (AS)?
- Alport's syndrome is a genetic disease in which there is defective type IV collagen.
  - There are three main types – X-linked (the most common), in which males are more severely affected than females; autosomal dominant; and autosomal recessive
  - Manifestations
  - Kidneys
  - Initially presents with microscopic haematuria and then proteinuria develops. About 50% of males with X-linked AS require dialysis or transplantation by ~25 years and about ~90% by aged 40 years.
  - Ears
    - Sensorineural hearing loss
  - Eyes
    - Anterior lenticonus can lead to slow progressive visual loss.
  - Management
    - Angiotensin-converting enzyme (ACE) inhibitors or angiotensin receptor blockers (ARBs) to try to delay onset of ESRD
    - Hearing aids as needed

## KEY POINTS

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- Look closely for any scars which may represent a renal transplant.
- If there is a renal graft present, comment on whether you believe it is functioning.
- Look for other forms of renal replacement therapy.
- Know the common causes for a renal transplant and how to manage a patient presenting with acute transplant dysfunction.

## REFERENCES

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- National Institute for Health and Care Excellence. (2016) British National Formulary. Available from <https://www.evidence.nhs.uk/formulary/bnf/current/>. Accessed 18 August 2016.
- Renal Association. (2011) British Renal Association guidelines on the post-operative care of the kidney transplant recipients. Available from <http://www.renal.org/guidelines/modules/post-operative-care-of-the-kidney-transplant-recipient#sthash.UPL7Z6si.dpbs>. Accessed 18 August 2016.
- Renal Association. (n.d.) Alport syndrome – Patient information. Available from <http://rarerenal.org/clinicianinformation/alport-syndrome/>. Accessed 18 August 2016.

## SPLENOMEGALY

Please examine this gentleman's abdominal system.

## FINDINGS

---

- *General*: Pallor, purpura
- *Peripheral*: Lymphadenopathy
- *Abdomen*: Enlarged spleen
- Be aware of how to differentiate splenomegaly from an enlarged kidney:
  - Features of a spleen: Mass in the left upper quadrant (LUQ) (moves towards RIF with respiration), unable to get above a spleen on examination, dull to percussion, nonballotable, presence of a palpable splenic notch
  - Features of a kidney: Moves minimally with respiration, can get above a kidney, resonant to percussion, ballotable, no notch palpable

## PRESENTATION

---

This patient has evidence of splenomegaly as there is a palpable mass in the left upper quadrant which moves inferomedially with respiration. I am unable to palpate above it and there is also a splenic notch palpable. On peripheral examination, there is evidence of pallor and bruising. There is no palpable lymphadenopathy.

Look for other signs leading to the underlying cause of the splenomegaly:

- Felty's syndrome
  - Rheumatoid hand signs
- Portal hypertension
  - Signs of chronic liver disease
- Infective endocarditis
  - Murmur, splinter haemorrhages
- CML/CLL/lymphoma
  - Lymphadenopathy (CLL/lymphoma), pallor

Splenomegaly can be classified by the degree of enlargement (but remember that causes of massive splenomegaly may be moderate to start with):

- Moderate enlargement (11–20 cm)
  - Rheumatological disease
    - Rheumatoid arthritis, SLE, Sjogren's syndrome



- Infection
  - Schistosomiasis, malaria, leishmaniasis, EBV
- Haematological
  - Lymphoma, leukaemia, myeloproliferative disease, haemolytic anaemia
- Massive enlargement (>20 cm)
  - Myelofibrosis
  - Chronic myeloid leukaemia
  - Visceral leishmaniasis

## INVESTIGATIONS

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- Bloods
  - FBC and blood film, haemolysis screen, LFTs, U&Es, CRP, blood cultures, malaria screen, virology for EBV, autoantibodies including rheumatoid factor
- Biopsy
  - Bone marrow aspirate, lymph node biopsy, splenic biopsy
- Imaging
  - USS abdomen, CT chest/abdomen/pelvis, echocardiogram if infective endocarditis suspected

## MANAGEMENT

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- This is dependent on the underlying cause.

## QUESTIONS

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1. What is the function of the spleen?
  - Removal of old/damaged red blood cells
  - Storage of platelets
  - B- and T-lymphocyte-mediated immune function
2. What advice should be given to a patient undergoing a splenectomy?
  - Preoperative vaccinations: Pneumococcal, meningococcal, *Haemophilus influenzae*
  - Lifelong prophylactic penicillin
  - Annual influenza vaccinations
  - Wear a medic alert bracelet
  - Advice regarding seeking medical attention if unwell
3. What are the indications for a splenectomy?
  - Hypersplenism: Autoimmune destruction of blood cells (for example, idiopathic thrombocytopenic purpura [ITP])
  - Mass effect of spleen
  - Traumatic rupture
  - Haematological malignancies (rarely)
  - Congenital haemolytic anaemias: Hereditary spherocytosis, elliptocytosis

## 4. Tell me about PRV.

- This is a primary polycythaemia, due to a fault in the bone marrow. It is the result of uncontrolled proliferation of mainly red blood cells, often due to a mutation in the Janus kinase 2 (JAK2) gene. Myeloid leucocytosis, thrombocytosis and splenomegaly can also occur.
- PRV may present with a raised haemoglobin level on a routine blood test. It can also present with thrombosis (or more rarely, bleeding), headache, sweating and pruritus.
- Investigation would initially be with a full blood count. Other (secondary) causes of polycythaemia should be excluded. If PRV is suspected, JAK2 mutation testing is carried out on a blood test. Imaging (CT/ultrasound) is used to visualise the spleen. Bone marrow biopsy is helpful for diagnosis.
- Diagnosis is by the World Health Organization (WHO) diagnostic criteria 2008.
- Management is targeted at lowering the risk of thrombosis. This includes venesection, aspirin and myelosuppression. JAK2 inhibitors are being introduced/under clinical trial. Management of symptoms such as pruritus should also be considered.
- Alongside thrombosis, a major complication of PRV is transformation to acute myeloid leukaemia.

## KEY POINTS

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- Be sure to measure splenomegaly to aid clinical diagnosis and be aware of how to differentiate between a palpable kidney.
- Be sure to look for peripheral signs to identify an underlying cause.

## REFERENCE

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Tefferi A, Thiele J, Vardiman JW. (2009) The 2008 World Health Organization classification system for myeloproliferative neoplasms: Order out of chaos. *Cancer* 115(17): 3842–7.





## ABDOMINAL STATION SUMMARY

- In this station, do not forget the normal abdomen. Remember the value of peripheral signs, e.g. lymphadenopathy; you may have your diagnosis before you lay your hand on the abdomen.
- Don't forget to ask the patient if there is any pain in the abdomen before you begin palpation. This is basic, but it can be embarrassing if you hurt the patient, and in all likelihood will result in a fail.
- Apologise if you cause the patient any discomfort.
- If possible, establish a cause for the patient's organomegaly to demonstrate to the examiners that you are thinking one step ahead.
- Demonstrate to the examiners that you are thinking ahead by inspecting for side effects of any treatments that the patient is receiving, e.g. the cushingoid patient with a renal transplant.
- Positioning is very important; ensure the patient is flat when you palpate the abdomen.
- Make your examination slick and quick, as you may run out of time before you finish it, but at the same time, don't be in a rush (a skill that can only be learnt with practice).
- Try not to be put off by unusual opening statements, for example, 'Please examine this patient with hypertension' (the opening statement will generally be relevant, e.g. a patient with renal disease/ESRD).
- If you palpate an organ, always ensure that you percuss it.
- Know how to clinically differentiate a spleen from a kidney (this is a PACES classic).
- Do not make up any findings.



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# 3

## Station 2: History Taking

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## HINTS FOR THE HISTORY-TAKING STATION

- The history-taking station is often thought of as being one of the easier stations; this misconception often costs candidates marks and sometimes their pass. You should not take this station lightly. You should see this station as an opportunity to excel, which may in turn give you breathing space if you have a bad station elsewhere.
- Use your 5 minutes before you enter the room wisely; think about differential diagnoses and the pertinent questions you may ask to exclude or include them.
- Pay very close attention to what you are being asked to do. Commonly, you will only be asked to find out more about the problem; you are probably not going to be expected to explain to the patient what you think is going on (unless they specifically ask you to), so focus on taking your history.
- You must always be polite and courteous to the patient.
- Do not forget simple things like introducing yourself.
- It is very important to establish a baseline. Does the patient know why they have come to see you?
- Start with open questions, as the patient will give you a great deal of information in the first 3 minutes. Try not to interrupt them.
- Demonstrate to the examiners that you are organised by taking a structured history.
- Don't forget risk factors for your differential diagnoses.
- If the patient has brought a piece of paper with their medication on it, go through it with them; do not merely assume that it is up to date. Ask if there are any other medications? Ask about any specific allergies they may have.
- Be vigilant for any hidden agendas the patient may have (addressing their concerns specifically is a good way to flush this out).
- Remember to address the patient's ideas, concerns and expectations.
- Never forget psychosocial elements, as they are important when developing your problem list.
- Summarise appropriately and make it obvious that you are doing this, so the examiners know.

## PATIENT WITH AN ABNORMAL BLOOD RESULT

Mr Warner is a 40-year-old male who on attendance at his well man clinic was found to have an adjusted serum calcium of 2.75 mmol/L. All other blood tests were within normal parameters. He seems to have few symptoms. He is known to have hypertension and takes ramipril 5 mg once daily (OD); he is otherwise well. He is an office worker, fully independent and lives with wife and three children. He drinks 20 units of alcohol a week and does not smoke.

Examination revealed a clear chest and a soft and nontender abdomen. A few small lymph nodes were palpated in his neck.

## KEY POINTS FOR THE PATIENT

- You are 40 years old and, as far as you are concerned, fit and healthy.
- You only went to the well man check-up because your wife has been nagging you.



- You cannot think of any obvious symptoms, but on direct questioning, you admit to having a troublesome dry cough.
- You have not coughed up any blood.
- You think you are becoming a little breathless on exertion.
- You have a good appetite, have not lost any weight and have not had fevers or night sweats.
- Your joints are generally sore.
- Your general practitioner (GP) felt something in your neck, but you're not sure of the significance of this.
- You have no other symptoms beyond this.
- You have high blood pressure but no other medical problems. You take ramipril and have not recently used any over-the-counter medications.
- You are not allergic to any medications.
- You do not smoke and never have.
- You drink 20 units of alcohol a week.
- You cannot think of any specific family history; both your parents are alive and generally well.

## SUGGESTIONS FOR THE CANDIDATE

---

- Ensure that you introduce yourself and check the patient's name and age.
- Ask the patient why they have come to see you.
- Ask the patient what they know about what has happened so far.
- Ask about any symptoms the patient may have.
- When the patient denies any symptoms, be sure to clarify this. Proceed to direct questioning if necessary, working through a list of differentials.
- When finding out about the cough, cover other respiratory symptoms and consider both lung cancer and sarcoidosis as potential causes of the hypercalcaemia. Ask about constitutional symptoms of malignancy or systemic illness.
- Ask about arthralgia, as this is a feature of sarcoidosis.
- Take a drug history, including over-the-counter medications, and ask about allergies.
- Ask about family history.
- Take an appropriate social history, being sure to enquire about smoking.
- Be sure to summarise appropriately.
- Ask the patient about any particular concerns or expectations they might have.
- Explain to the patient that you cannot be sure of a diagnosis, but sarcoidosis is most likely (if asked directly about malignancy, you cannot definitively rule this out at this stage).
- Explain to the patient that a chest X-ray and some specialist blood tests are required, and that you will see them next week with the results. You will likely need to do a biopsy of the lymph nodes.

## THEMES EXPLORED

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- It is important to have a list of differentials for common abnormal blood test results (renal failure, hypercalcaemia and raised alkaline phosphatase [ALP], among others).

- Tie the symptoms (and signs) elicited into this list. Lymphadenopathy points towards malignancy and sarcoidosis, and this link is furthered with the presence of a dry cough.
- Look at the case as a whole; the young age, lack of social history and lack of constitutional symptoms in this patient swing the balance towards sarcoidosis.

## RELEVANT INFORMATION

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### CAUSES OF HYPERCALCAEMIA

- Hyperparathyroidism (primary and tertiary)
- Malignancy (bony metastases, myeloma, paraneoplastic syndromes)
- Drugs (thiazides, vitamin D excess)
- Dehydration
- Sarcoidosis
- Thyrotoxicosis
- Spurious result

### PRIMARY HYPERPARATHYROIDISM

Most commonly caused by a parathyroid adenoma. Can also be caused by parathyroid hyperplasia and malignancy (rare). Can be associated with multiple endocrine neoplasia. Often presents by chance from blood tests, although can present with symptoms of hypercalcaemia.

On blood tests, serum calcium is raised. The serum parathyroid level is either raised or (inappropriately) normal.

Treatment is most commonly surgical (excision of the adenoma or hyperplastic glands).

### MANIFESTATIONS OF SARCOIDOSIS

- Pulmonary: Bilateral hilar lymphadenopathy, pulmonary fibrosis
- Cutaneous: Subcutaneous nodules, erythema nodosum, lupus pernio
- Ocular: Uveitis, glaucoma, soft tissue orbital mass, keratoconjunctivitis sicca
- Cardiac: Heart block and arrhythmias, cardiomyopathy
- Joints: Polyarthritits
- Facial: Parotid swelling
- Abdominal: Splenomegaly, hepatomegaly
- Neurological: Cranial neuropathy (e.g. Bell's palsy), meningitis
- Metabolic and renal: Hypercalcaemia, hypercalciuria, renal impairment, hypothalamic involvement
- General: Fever, fatigue

## REFERENCE

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Pallan S, Rahman MO, Khan AA. (2012) Diagnosis and management of primary hyperparathyroidism. *BMJ* 344: e1013.



## PATIENT WITH BACK PAIN

Mr Quimby is a 78-year-old gentleman who complains of back pain. He has had this for 4 months, mainly in the thoracic region. There are no exacerbating or relieving factors. He says it is worse at night. He is also complaining of fatigue and lethargy.

Examination is unrevealing. He is thin and has a soft and nontender abdomen with no obvious organomegaly, normal heart sounds and a clear chest. A full blood count (FBC) has found a haemoglobin of 109 g/L with a mean cell volume (MCV) of 85 fL (white cell count, platelets and renal function were normal).

### KEY POINTS FOR THE PATIENT

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- You have had back pain in the middle of your back for 4 months, and you feel it is getting worse.
- Your pain is worse at night-time.
- Your pain doesn't move anywhere and nothing makes it better or worse.
- Paracetamol has been ineffective for the pain.
- You have been lethargic for some time now and are generally listless to the extent that your garden is overgrown, as you don't have the energy to tend it.
- You don't weigh yourself, but your clothes are slightly looser.
- You have had no change in bowel habit.
- You have no new urinary symptoms; you pass urine twice at night and that is normal for you.
- You have not noticed any pains anywhere else.
- You have no chest symptoms. You do not sweat at night and have not had any fevers.
- Your only past medical problem is that of prostate trouble, and the GP gave you a tablet for that (you can't remember the name) about a year ago. Your urine stream is currently fine.
- To your knowledge, you have not had any trauma to the back.
- You are a retired factory worker.
- You are an ex-smoker, stopping 25 years ago. You don't drink alcohol.
- You live with your son.
- With the lethargy, you find it hard to have an active life. You used to play bowls, but the back pain has put an end to this. You can still walk reasonably well.
- You are concerned that you may have cancer, as your mother died of lung cancer.

### SUGGESTIONS FOR THE CANDIDATE

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- Ensure that you introduce yourself and check the patient's name and age.
- Ask the patient if they know why they have come to see you and what has happened so far.
- Start with an open question about his back pain.
- Find out where the pain is, the nature and if there is any radiation.
- Establish the onset, course and duration of symptoms.

- Establish if there are any exacerbating or relieving factors.
- Ask if there is any particular time when the symptoms are worse.
- Make sure you ask about red-flag symptoms: weight loss, systemic upset, night sweats, fevers and neurological symptoms in both upper and lower limbs (given the location of the pain).
- Screen for malignancy with your systems review.
- Take a thorough past medical history.
- Enquire about family history.
- Ask about the patient's social situation and ensure that you ask about the impact on their life.
- Be sure to summarise appropriately.
- Ask the patient about any particular concerns that they might have.
- Outline the investigations that are required, including blood tests and spinal imaging/bone scan.

## THEMES EXPLORED

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- In a patient of this age group with back pain, malignancy is likely and needs to be asked about. Other red-flag symptoms are of relevance.
- The blood results here also point towards malignancy; be sure to take note of this. Knowledge of how to proceed with investigations is required.

## RELEVANT INFORMATION

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### RED-FLAG SIGNS/SYMPTOMS FOR BACK PAIN

- Age <20 or >55
- Pain at night
- Constant/worsening pain
- Constitutional features of malignancy (weight loss, appetite loss, night sweats)
- History of malignancy
- Thoracic pain
- Pyrexia
- Bilateral lower limb neurological symptoms
- Sphincter disturbance

### CAUSES OF BACK PAIN IN AN ELDERLY PATIENT

- Degeneration (osteoarthritis)
- Vertebral collapse (osteoporosis)
- Solid malignancy metastases (a primary malignancy is less likely)
- Myeloma
- Paget's disease
- Trauma





## TUMOURS MOST LIKELY TO METASTASISE TO BONE

- Breast
- Lung
- Prostate
- Kidney
- Thyroid

## PATIENT WHO HAS COLLAPSED

Mr Foyle is a 31-year-old male who has collapsed twice within the last few weeks. Between the episodes he has felt well. Mr Foyle sustained no injury from either collapse. He has been referred by his GP, who stated that an electrocardiogram (ECG) done in the practice was 'normal'.

There are no positive findings on examination, with a regular pulse of 64 beats/minute. His blood pressure is 116/70 with no postural drop. All his blood tests (FBC, urea and electrolytes [U&Es], liver function, bone profile, magnesium and glucose) have been normal. An ECG taken by the nurse in clinic has found normal sinus rhythm with no abnormalities.

## INSTRUCTIONS FOR THE PATIENT

- You are 31 years old and, to your knowledge, fit and healthy.
- You initially collapsed 4 weeks ago. This occurred while you were standing at work (and had been for a couple of hours). You felt 'light-headed' for about a minute before the event.
- You collapsed again 2 weeks ago. This occurred following a game of football with friends. You felt 'light-headed' after standing up in the changing rooms and collapsed about 30 seconds later. You felt 'fine' during the game itself.
- You remember being sweaty before both events, although you didn't notice any unusual sensations, smells or tastes.
- During both episodes, you report that you lost consciousness very briefly, and awoke on the floor. Your friends told you that during the second episode, you awoke very quickly.
- After resting for half an hour, you felt 'back to normal' in both cases. You have a good memory of what happened during both episodes.
- Your work colleagues and friends reported that there was no shaking of your arms or legs after you had collapsed. They reported that your eyes were closed through both events.
- You did not bite your tongue and were not incontinent of urine or faeces.
- You sustained no injury during either episode. You had no arm or leg weakness afterwards.
- You have not sustained a head injury recently.
- You have not had chest pain.
- You have not felt breathless at all.

- You have not been aware of your heart beating more than usual, or having an unusual rhythm.
- You have not had episodes like these collapses before.
- You have otherwise been well recently. You have not lost any weight or had a recent infection.
- You have no past medical history of note.
- You do not take any prescribed medications. You have not taken any over-the-counter medications recently. You have no known allergies.
- You do not take illicit or herbal drugs.
- You used to smoke 10 cigarettes a day, but quit 6 months ago.
- You drink approximately 15 units of alcohol per week and haven't been drinking excessively recently.
- No one in your family has ever had any similar episodes. No one in your family has died suddenly and unexpectedly.
- You live at home with your wife and son, who is 3 months old. On close questioning, you feel sleep deprived, as you are awoken several times throughout the night.
- You work in a factory. You have recently started working on a new production line. On close questioning, this involves you standing for long periods of the day, more than previously. You have also been busier at work and don't always get regular breaks.
- You do drive, although not every day, as you walk to work.
- You are concerned that you have a serious illness; a friend you used to play football with collapsed while playing and died from a 'heart problem'.

## SUGGESTIONS FOR THE CANDIDATE

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- Ensure that you introduce yourself and check the patient's name and age.
- Ask the patient if they know why they have come to see you and what they know about what has happened so far.
- Start with an open question about the episodes of collapse.
- Ask about the patient's experience of these events (before, during and after), but also about any collateral history that they can provide from others who were present. If you feel you have insufficient collateral history, discuss contacting witnesses of the events.
- Ensure that you ask about symptoms relevant to a collapse of cardiac cause, such as palpitations and chest pain.
- Ensure that you ask about features of a seizure, such as aura, incontinence and tongue biting.
- Demonstrate to the examiners that you are working through your list of differential diagnoses.
- Check on past medical history, including previous episodes of collapse.
- Take a drug history and be sure to ask about allergies.
- Ask about illicit drug and alcohol usage.
- Ask about the patient's social situation and the role this could be playing in these episodes.
- Ask about the patient's work and the impact this could have had on these episodes.
- Ask about family history of collapse and sudden death.



- Be sure to summarise appropriately.
- Ask the patient about any particular concerns that they might have.
- On making a likely diagnosis of vasovagal syncope, offer reassurance. There is no evidence here of a serious cardiac disorder (remember, this is the patient's main concern), given the normal ECG and identified triggers of the episodes.
- Explain that further investigations are not needed at this time.
- Offer lifestyle advice in terms of ensuring an adequate fluid intake. Offer occupational advice in advising sitting where possible at work and trying to take breaks.
- Offer advice that the patient is able to drive currently, as there is an identified trigger to the events (standing, low fluid intake) and the episodes occurred while standing.

## THEMES EXPLORED

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- Episodes of collapse have a wide differential diagnosis, and it is important to explore a variety of symptoms in order to make a diagnosis. It is important to rule out serious causes of collapse, such as cardiac disorders and seizures.
- Lifestyle and occupational advice can form a significant part of the management plan; this needs to be communicated to the patient.
- Despite episodes of collapse, patients may be able to drive according to Driver and Vehicle Licensing Agency (DVLA) guidelines.

## RELEVANT INFORMATION

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### DIFFERENTIAL DIAGNOSIS OF COLLAPSE

- Cardiovascular: Vasovagal syncope, situational syncope, orthostatic hypotension, carotid sinus hypersensitivity, hypovolaemia, arrhythmia, cardiac outflow obstruction (such as aortic stenosis and hypertrophic cardiomyopathy)
- Neurological: Seizure/epilepsy
- Metabolic: Hypoglycaemia, Addisonian crisis
- Psychological: Anxiety

### FEATURES OF UNCOMPLICATED VASOVAGAL SYNCOPE

- No features of another diagnosis (such as prolonged seizure activity)
- Postural reasons: Prolonged standing, occurring *after* exercise
- Provoking factors: Pain, emotion
- Prodrome: Sweating, nausea

## REFERENCES

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- Driver and Vehicle Licensing Agency. (2016) Assessing fitness to drive – A guide for medical professionals. Available from <https://www.gov.uk/government/publications/assessing-fitness-to-drive-a-guide-for-medical-professionals>. Accessed 5 July 2016.
- National Institute for Health and Care Excellence. (2014) Guidance CG109. Transient loss of consciousness ('blackouts') in over 16s. Available from <https://www.nice.org.uk/guidance/cg109>. Accessed 5 July 2016.

## PATIENT WITH A COUGH

Miss Jones is a 36-year-old female who has been troubled by a persistent dry cough for the last 6 months. She has tried a salbutamol inhaler and has stopped smoking, although this has not led to an improvement in her symptoms. She has a peak flow of 440 L/minute with no reversibility. Spirometry does not show an obstructive deficit. There are no positive findings on examination. All her blood tests have been normal, and a recent chest X-ray found no abnormalities.

### INSTRUCTIONS FOR THE PATIENT

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- You are 36 years old and, to your knowledge, fit and healthy.
- You have had a cough for a long time; it is really starting to bother you.
- Your cough is worse at night-time.
- The cough is nonproductive, and you have never coughed up any blood.
- Your breathing is fine; you play badminton once a week and have noticed no change in your fitness levels.
- You are not wheezy.
- You have no chest pain or palpitations.
- You have not lost weight.
- You sleep with one pillow.
- On close questioning, you experience some indigestion and a feeling of acid in your mouth.
- You do not have any abdominal pain.
- You have no past medical history.
- You take a progesterone-only contraceptive pill and have not missed any doses. You are not on any other medications. You have no allergies.
- You do not think you are pregnant.
- Your mother has had a blood clot in her leg.
- You did not have any serious childhood illnesses.
- You are a lawyer. Your job is important to you and keeps you very busy. You work late commonly and often eat on the run, sometimes very late in the day.
- You used to smoke when you were out with friends but gave this up 6 months ago.
- You have no pets.
- You drink approximately 35 units of alcohol per week and occasionally more, dependent on client functions.
- You are concerned that you have a serious illness.
- You are worried, as the cough is keeping you awake at night, affecting your sleep and your performance at work.

### SUGGESTIONS FOR THE CANDIDATE

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- Ensure that you introduce yourself and check the patient's name and age.
- Ask the patient if they know why they have come to see you and what they know about what has happened so far.
- Start with an open question about her cough.



- Ask her specifically about when the cough is worse, and about sputum and blood production.
- Ensure that you ask about other respiratory symptoms.
- Demonstrate to the examiners that you are working through your list of differentials (so specifically ask about symptoms of reflux, a common cause of cough). Also ask about a postnasal drip and any symptoms of sinus congestion.
- Clues such as the cough worsening on lying down and after eating help point towards the diagnosis of acid reflux.
- Check on past medical history, including childhood infections.
- As in any respiratory history, take an occupational history and a smoking history.
- Enquire about pets and other common triggers for asthma.
- Take a drug history and be sure to ask about allergies.
- Ask about family history.
- Perform a systems review (nonspecific symptoms could be the only clue that the patient has a lymphoma or sarcoidosis causing their cough).
- Ask about their social situation and ensure that you ask about the impact on their life.
- Be sure to summarise appropriately.
- Ask the patient about any particular concerns that they might have (particularly relevant here).
- On making a likely diagnosis of reflux disease, offer reassurance and a trial of proton pump inhibitor (PPI), with follow-up in clinic in 1 month. Consider an oesophagogastroduodenoscopy (OGD) with regard to the duration of symptoms.
- Offer lifestyle advice in terms of reducing alcohol intake and eating at sensible times.

## THEMES EXPLORED

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- The final diagnosis may be from a different 'system' than the presenting complaint; be prepared to 'think outside the box'.
- It is important to know the 'red-flag' symptoms which may require urgent referral and endoscopic examination.
- Lifestyle advice may form a significant part of the management plan; this should be communicated to the patient.

## RELEVANT INFORMATION

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### RED-FLAG SYMPTOMS/SIGNS REQUIRING URGENT UPPER GI ENDOSCOPY

- Chronic gastrointestinal (GI) bleeding
- Unintentional weight loss
- Dysphagia
- Persistent vomiting
- Iron deficiency anaemia
- Epigastric mass
- Suspicious barium meal result
- Age >55

## MANAGEMENT OF GASTRO-OESOPHAGEAL REFLUX DISEASE

- Endoscopy in those with red-flag symptoms.
- Consider testing for *Helicobacter pylori*.
- Lifestyle advice: Healthy eating, weight reduction and smoking cessation.
- Full-dose PPI for 4–8 weeks.
- Give a PPI at the lowest effective dose for those in whom symptoms recur.
- In those with severe oesophagitis, a full-dose PPI may need to be continued.
- Use a histamine-2 receptor antagonist in those in whom a PPI is ineffective.

## REFERENCES

- National Institute for Health and Care Excellence. (2014) Gastro-oesophageal reflux disease and dyspepsia in adults: Investigation and management. Available from <https://www.nice.org.uk/guidance/cg184>. Accessed 5 July 2016.
- National Institute for Health and Care Excellence. (2015) Guidelines for suspected cancer: Recognition and referral. Available from <https://www.nice.org.uk/guidance/ng12>. Accessed 5 July 2016.

## PATIENT WITH DIARRHOEA

Mr Wentworth is a 22-year-old male who complains of loose stools. He works as a car mechanic and is finding that his symptoms are disrupting his job. Examination is unremarkable. He is thin and has a soft and nontender abdomen, normal heart sounds and a clear chest. Preliminary blood tests have been sent, and he has had two stool samples that have come back negative on microscopy and culture.

## INSTRUCTIONS FOR THE PATIENT

- You have had loose stools for 6 months.
- You generally open your bowels once a day.
- You have never noticed any blood or mucus.
- You have lost 1 stone of weight in this period (this is not intentional).
- You get a bloating sensation in your abdomen but do not have pain.
- You complain of fatigue.
- You do suffer with regular mouth ulcers.
- You went to see your GP 2 years ago with an intensely itchy rash, which cleared of its own accord.
- You have a good appetite and do not suffer with night sweats.
- You have no past medical history and do not take any medications.
- You are a smoker of 20 cigarettes/day.
- You don't drink alcohol.
- You find your symptoms embarrassing, and they are interfering with your work.
- You have not had any foreign travel or any infectious contacts.
- You eat a 'normal' diet and have not altered this at any time.
- You are not aware of any specific family history.



## SUGGESTIONS FOR THE CANDIDATE

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- Ensure that you introduce yourself and check the patient's name and age, as well as asking the patient why they have come to see you and what has happened so far.
- Start with an open question about his loose stools.
- Specifically, ask about how often it occurs, stool consistency and if there is any blood or mucus.
- Ask about abdominal pain, nausea and vomiting and weight loss.
- Enquire specifically about any extra-intestinal symptoms or associations of coeliac disease:
  - Lethargy (anaemia).
  - Dermatitis herpetiformis.
  - Low-impact bone fractures due to osteoporosis.
  - Be aware of the predisposition to malignancy, particularly small bowel lymphoma. Ask about constitutional symptoms.
- In order to assist in ruling out irritable bowel syndrome, ask about anxiety and depression, and ensure that there is no evidence of other functional illness (e.g. functional dyspepsia).
- Enquire about family history of coeliac disease.
- Ask about foreign travel.
- Ask about infectious contacts.
- Ask about recent antibiotic use.
- Always ask about alcohol intake where malabsorption is suspected.
- Ask about the patient's social situation and the impact of the symptoms on their life.
- Be sure to summarise appropriately.
- Ask the patient about any particular concerns that they might have.
- Outline the investigations that will be required (blood tests, possible upper GI endoscopy), and the management given a diagnosis of coeliac disease (dietary change, dietician referral).

## THEMES EXPLORED

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- Coeliac disease is common in this age group and should be top of a list of differential diagnoses. Inflammatory bowel disease is less likely given the frequency of stool.
- It is important to ask about extra-intestinal manifestations and associations of coeliac disease; the history of dermatitis herpetiformis here helps to confirm the primary diagnosis.

## RELEVANT INFORMATION

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### CONDITIONS ASSOCIATED WITH COELIAC DISEASE

- Dermatitis herpetiformis
- Hyposplenism
- Immunoglobulin (Ig) A deficiency
- Type 1 diabetes mellitus
- Autoimmune thyroiditis
- Primary biliary cirrhosis (PBC)

## COMMON CAUSES OF MALABSORPTION

- Coeliac disease
- Chronic pancreatitis (including cystic fibrosis)
- Carcinoma of the pancreas
- Crohn's disease (terminal ileal disease)
- Iatrogenic causes, including small bowel resection and radiation enteritis

## DIAGNOSIS AND MANAGEMENT OF COELIAC DISEASE

- Serological testing: Tissue transglutaminase (tTG) antibody, endomysial antibody (EMA) (while patient is on a normal diet).
- A faecal calprotectin test evaluates bowel inflammation; a negative test rules out inflammatory bowel disease.
- Duodenal biopsy is required to confirm the diagnosis in adults. This will show villous atrophy and increased intraepithelial lymphocytes in coeliac disease.
- A strict gluten-free diet is the only effective treatment.
- Adherence to such a diet can be improved with input from an expert dietician and from support by a coeliac disease patient advocacy group (such as Coeliac UK).

## REFERENCE

Mooney PD, Hadjivassiliou M, Sanders DS. (2014) Coeliac disease. *BMJ* 348: g1561.

## PATIENT WITH JAUNDICE

Mrs Valdes is a 64-year-old female with jaundice. She first noticed this after returning from India 2 months ago, and since then it has gradually worsened. She also feels lethargic to the extent that she is struggling to make it through her days at work as a primary school teacher. She has a past history of hypothyroidism, for which she takes levothyroxine 150 µg once daily, and hypercholesterolaemia, for which she takes simvastatin 40 mg once nightly (ON). She is a moderate drinker, consuming one gin and tonic every other evening.

On examination, she is icteric, she has palmar erythema and there are five spider naevi visible on her anterior chest wall. Her abdomen is soft and nontender, and she has no palpable organomegaly. Her blood tests demonstrate deranged liver function tests (LFTs), and an ultrasound scan (USS) of the liver has been arranged.

## INSTRUCTIONS FOR THE PATIENT

- You first noticed the jaundice while you were in India, but you have been feeling generally unwell for 3 months, with tiredness and general lack of energy.
- Your urine is a normal colour and your motions have not changed.
- You have no abdominal pain, abdominal distension, nausea, vomiting, diarrhoea, fever or indigestion.
- You have not lost weight and your appetite is OK.





- You have severe itching that has been worsening for some time.
- You have hypothyroidism and take tablets for this. You are also taking a statin for high cholesterol.
- You are fit and well otherwise.
- You work as a teacher and have done so since leaving university.
- Your husband is your only sexual partner.
- You were well throughout your stay in India, where you spent 2 weeks on holiday in Goa.
- You have a single gin and tonic every other evening.
- You have never been involved in intravenous (IV) drug use. You do not have any tattoos.
- You have never had any operations or blood transfusions.
- You are a nonsmoker.
- Your mother has hypothyroidism and pernicious anaemia, and she takes steroids but you're not sure why.
- No one else in the family has been jaundiced.
- You live with your husband and two children.
- The GP has done some blood tests and organised a scan of your abdomen and sent you to see the specialist.
- You are worried it might be cancer.

## SUGGESTIONS FOR THE CANDIDATE

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- Ensure that you introduce yourself and check the patient's name and age.
- Ask the patient why they have come to see you and what has happened so far.
- Start with an open question about the jaundice.
- Ask specifically about the course of the illness and associated symptoms.
- Specifically enquire about the colour of the patient's urine and stool.
- Ask about other gastrointestinal symptoms.
- Demonstrate to the examiners that you are aware of the risk factors for jaundice by asking about alcohol, sexual history, intravenous drug abuse, blood transfusions, occupational history and travel history.
- Take a drug history and be sure to ask about allergies.
- Ask about family history and establish if there are any other symptoms of autoimmune disease in this patient.
- Ask about social situation and ensure that you ask about impact on their life.
- Be sure to summarise appropriately.
- Ask the patient about any particular concerns and expectations that they might have.
- Communicate to the patient that further investigations are required. Medication can be offered to treat the itching.

## THEMES EXPLORED

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- Painless jaundice is often caused by malignancy, although other causes need to be considered, especially in younger patients. PBC is possible here; clues include the hypothyroidism and hypercholesterolaemia.

- It is important to establish the patient's concerns. Mrs Valdes is less likely to have a malignancy here, but this cannot be ruled out until after imaging.
- Symptom control is important, as well as making a diagnosis. Mrs Valdes's itching should be addressed.

## RELEVANT INFORMATION

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### COMMON AUTOIMMUNE ASSOCIATIONS OF PBC

- Sjögren's syndrome
- Autoimmune thyroid disease
- Rheumatoid arthritis
- Systemic sclerosis
- Diabetes mellitus

### INVESTIGATION OF SUSPECTED PBC

- Deranged LFTs (raised ALP early in the disease, raised bilirubin later)
- Liver screen to rule out other causes, including USS of liver (or computerised tomography [CT] abdomen)
- Positive antimitochondrial antibody (M2 antibody is specific)
- Raised serum immunoglobulins, especially IgM
- Diffuse architectural change on liver ultrasound
- Hepatic granulomas, lymphocytic infiltrates and cirrhosis on liver biopsy

### CONTROL OF DISEASE

- Early and midstages of disease are managed with ursodeoxycholic acid (UDCA) to change the balance of toxic and nontoxic bile salts (it is the autoimmune destruction of small bile ducts and the accumulation of toxic bile salts which causes cirrhosis; UDCA slows the progression of disease).
- Late stage of disease may require liver transplant.

### MANAGEMENT OF ITCH IN CHOLESTASIS

- Topical agents ineffective.
- Cholestyramine is used first line (bile salt sequestrant).
- Rifampicin is used second line. Liver function needs to be closely monitored on this drug.
- Naltrexone, an opioid antagonist, is third line.
- Sertraline is fourth line.

## REFERENCE

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European Association for the Study of the Liver. (2009) EASL clinical practice guidelines: Management of cholestatic liver diseases. *J Hepatol* 51(2): 237–67.



## PATIENT WITH JOINT PAINS

Mrs Leadbitter is a 34-year-old female who over the past 2 months has developed pains in the small joints of her hands. She is a secretary and is finding typing increasingly difficult. She also complains of stiffness in her fingers. She has type 1 diabetes mellitus. Her blood sugars are well controlled. On examination, she has tender joints in both hands, but otherwise, there is nothing abnormal to find.

### INSTRUCTIONS FOR THE PATIENT

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- You have pain in both of your hands, particularly the joints around the knuckles and the wrist.
- This pain started 2 months ago, and you feel it is getting worse.
- It is an aching pain.
- You have noticed that your fingers and hands are also stiff in the morning. This stiffness goes away as the day progresses.
- None of your other joints are affected.
- You have been feeling more tired recently, but put this down to working extra hours.
- You have not lost any weight.
- You do not have any rashes.
- You have had no problems with your eyes.
- You have no rashes, and your nails are fine.
- You do not have chest pain, but you have noticed the beginnings of a dry cough. You have no problems with your breathing.
- On close questioning, you do have a dry mouth.
- You have insulin-dependent diabetes and are meticulous with control of your blood sugars.
- Other than the diabetes, you are fit and healthy.
- You take insulin as a basal bolus regimen, simvastatin 40 mg once daily and ramipril 2.5 mg once daily.
- You do not have any allergies to medications.
- Your sister has pernicious anaemia.
- You live with your husband and three children.
- You work as a secretary for a law firm.

### SUGGESTIONS FOR THE CANDIDATE

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- Introduce yourself and check the patient's name and age.
- Ask the patient why they have come to see you.
- Ask the patient what they know about what has happened so far (establish baseline).
- Establish which joints are affected. Are the symptoms in both hands? Are any other joints affected?
- Ask specifically about stiffness and clarify when it occurs.
- Ask about any lumps and bumps (nodules).

- Make sure you ask about symptoms of inflammatory bowel disease in order to help rule out an enteropathic arthritis.
- Ask about urinary symptoms to help rule out reactive arthritis (although more commonly an oligoarthritis or large-joint arthritis).
- Demonstrate that you know the associations of inflammatory arthropathies by asking about
  - Dry mouth and eyes (Sjögren's syndrome)
  - Cough and dyspnoea (pleural effusions and lung fibrosis)
  - Cardiac symptoms (pericarditis and pericardial effusions)
  - Systemic symptoms (weight loss and lethargy)
  - Rashes (psoriasis)
  - Ophthalmic symptoms (episcleritis, scleritis and anterior uveitis)
- Ask about neurological symptoms in the hands, as carpal tunnel syndrome can develop in both rheumatoid arthritis and osteoarthritis.
- Take a thorough past medical history
- Take a thorough drug history and be sure to ask about allergies.
- Family history is important.
- Occupational history is important in this case.
- Try to ascertain the functional status of the patient.
- Be sure to summarise appropriately.
- Ask the patient about any particular concerns and expectations that they might have.
- Discuss a plan in terms of investigations and follow-up.

## THEMES EXPLORED

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- This case demonstrates the link between autoimmune diseases, with the patient having an established diagnosis of type 1 diabetes. There is also a family history of pernicious anaemia.
- This case gives the candidate the opportunity to show that they know both the causes and associations of inflammatory arthritides.
- A key point is to distinguish early between inflammatory and mechanical arthritis.

## RELEVANT INFORMATION

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### DIAGNOSTIC CRITERIA FOR RHEUMATOID ARTHRITIS

- Rheumatoid arthritis was previously diagnosed using the American Rheumatism Association 1987 criteria. However, new criteria were published in 2010: the American College of Rheumatologists' criteria. For details on these, see the rheumatoid arthritis case in Station 5 of this book.
- Recent evidence suggests trying to identify patients early in the disease process, so that treatment with disease-modifying agents can be instituted early. National Institute for Health and Care Excellence (NICE) guidance recommends suspecting rheumatoid arthritis in those with 'persistent synovitis' (meaning it has been present for a few weeks) where no other underlying cause is obvious.



## PHARMACOLOGICAL MANAGEMENT OF RHEUMATOID ARTHRITIS

- NICE guidelines now suggest using two disease-modifying antirheumatoid drugs (DMARDs) initially. This is generally methotrexate alongside another drug. DMARDs are commenced soon after diagnosis. A short course of glucocorticoids is also recommended to control symptoms quickly.
- After symptoms are controlled, the aim is to reduce DMARDs to their lowest effective dose.
- In those whom a combination of DMARDs is not effective, and in whom disease activity is severe, biological agents (including anti-tumour necrosis factor (TNF) monoclonal antibodies) are considered.
- Analgesics should be used where pain is problematic. Paracetamol is recommended first line, and weak opioids can also be considered. Nonsteroidal anti-inflammatory drugs (NSAIDs) and cyclo-oxygenase (COX)-2 inhibitors are likely to be effective, but they should be used for the shortest period of time possible to prevent side effects.

## REFERENCES

- National Institute for Health and Care Excellence. (2013) Clinical knowledge summary for rheumatoid arthritis. Available from <http://cks.nice.org.uk/rheumatoid-arthritis#!topicsummary>. Accessed 5 July 2016.
- National Institute for Health and Care Excellence. (2015) Guidance CG79. Rheumatoid arthritis in adults: Management. Available from <https://www.nice.org.uk/guidance/cg79/chapter/Recommendations#pharmacological-management>. Accessed 5 July 2016.

## PATIENT WITH LEFT ARM WEAKNESS

Mrs Abedney is a 62-year-old female, who developed left arm weakness 8 days ago, which has subsequently not improved. She was fit and healthy prior to this. There is no family history of cardiac disease; she is a retired bank clerk and lives with her husband. She is a nonsmoker. On examination, she has an upper motor neurone lesion affecting her left arm. Her blood pressure in clinic was 160/95. She has been told that she has had a stroke, and has initially been commenced on high-dose aspirin, atorvastatin and ramipril. Her serum cholesterol and random glucose are pending.

## INSTRUCTIONS FOR THE PATIENT

- You were fit and healthy up to this event.
- You are right-handed.
- Two weeks ago, you noticed some tingling in your left arm. Since then, it has gradually become weaker and the whole of the left arm is now affected.
- The weakness is getting worse and not improving.
- The sensation in your left arm remains intact.
- Your right arm is normal.
- Your legs are normal.
- Your speech has been normal.

- You have not experienced any headaches or visual symptoms.
- You have not sustained a head injury.
- Your swallowing is normal.
- Your bladder and bowel functions have been normal.
- You have lost a stone in weight over the past 6 months. This was not planned.
- You have been increasingly tired in the last 3 months.
- You have no urinary, respiratory, cardiac or gastrointestinal symptoms.
- You have no family history of note.
- You are a nonsmoker and drink 2 units of alcohol/day.
- You saw your GP 7 days ago (1 day after the onset of your symptoms), and she said you had a 'mini-stroke' or transient ischaemic attack (TIA). She gave you some tablets and said she would refer you to a specialist.
- You are yet to have a scan of your head.
- The weakness has left you dependent on your husband to manage the house, and he now has to help you get ready in the mornings.
- You are concerned that despite treatment for a week, your symptoms have worsened. You feel a burden on your husband.

## SUGGESTIONS FOR THE CANDIDATE

---

- Ensure that you introduce yourself and check the patient's name and age.
- Ask the patient why they have come to see you and what has happened so far.
- Ask the patient about their symptoms and establish a history of presenting complaint (you will note that the onset is gradual and the symptoms progressive, not in keeping with a stroke).
- Check the patient's handedness.
- Ask the patient about other neurological symptoms.
- Ensure that you take a thorough past medical history, including asking about risk factors for stroke. Take a drug history.
- Ask about the patient's social situation, including the impact of the symptoms on their life.
- Make sure that you do a thorough systems review, particularly asking about any symptoms that might suggest an underlying malignancy (especially once you have established there is a suggestion of weight loss and lethargy) or multiple sclerosis (ask about prior episodes of visual disturbance/incontinence and ataxia).
- Ask the patient about any particular concerns and expectations that they might have. Social support is important, as well as making a medical plan for the patient.
- Explain to the patient that cerebral imaging is crucial here for diagnosis. Explain that you will expedite this and see the patient again this week.

## THEMES EXPLORED

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- The history taken here leads you to think that the diagnosis could be a space-occupying lesion.
- The diagnosis may be different from the GP's initial impression and from what the patient may have been told. Keep an open mind.
- Address the patient's concerns, that investigations are required to make a diagnosis, and that social support is available.



## RELEVANT INFORMATION

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### CAUSES OF SPACE-OCCUPYING LESIONS

- Benign tumour
- Malignant primary tumour
- Cerebral metastasis
- Colloid cyst
- Aneurysm
- Abscess
- Chronic subdural haematoma

### CANCERS MOST COMMONLY METASTASISING TO THE BRAIN

- Breast
- Lung
- Melanoma
- Haematological malignancies (leukaemia and lymphoma) commonly cause leptomeningeal metastases.

### IMPACT ON DRIVING

- In this case, the patient should not drive until a diagnosis is made, at which point advice can be taken from the DVLA.
- The DVLA's rules are quite complex. A craniotomy requires at least a 6-month period off driving, with varying rules for benign tumours and at least 1–2 years off for malignant tumours post-treatment.

## REFERENCES

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- DVLA. (2016) Assessing fitness to drive – A guide for medical professionals. Available from [https://www.gov.uk/government/uploads/system/uploads/attachment\\_data/file/517268/Fitness\\_to\\_drive.pdf](https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/517268/Fitness_to_drive.pdf). Accessed 5 July 2016.
- Giglio P, Gilbert MR. (2010) Neurologic complications of cancer and its treatment. *Curr Oncol Rep* 12(1): 50–59.

## PATIENT WITH VISUAL DISTURBANCE

Mr Atherton is a 69-year-old male who has gradually developed blurred vision over the past 3 days. He also complains of a nonspecific headache. He has a past history of hypertension, atrial fibrillation and hypercholesterolaemia, and had an OGD 1 year ago which demonstrated peptic ulcer disease.

On examination, he looks generally well; his pulse is irregularly irregular, his blood pressure is 145/83 and his heart sounds are normal. Neurological assessment is normal. His GP thinks he is having transient ischaemic attacks. His medications are as below:

- Aspirin 75 mg OD
- Simvastatin 40 mg ON

- Bisoprolol 10 mg OD
- Candesartan 8 mg OD
- Lansoprazole 30 mg OD
- Quinine sulphate 300 mg ON

## INSTRUCTIONS FOR THE PATIENT

---

- You have had visual disturbance for a few days. Your vision is generally blurred, and you cannot read the small print of the newspaper as well as you used to.
- The visual disturbance is getting worse.
- You have not had anything like this before.
- You don't wear glasses and last went to the optician 1 year ago, when your vision was reported to be normal.
- You also have a headache which has come on at the same time.
- This headache hasn't particularly bothered you, but it is not normal and you notice that it hurts when you wash your hair in the shower.
- You are also noticing pain when you chew.
- You don't have any floaters or flashing lights, or any symptoms of a 'curtain coming down'.
- You don't have any nausea or vomiting.
- There has been no recent head injury.
- Your legs, face and speech have been fine, although you have pain in both shoulders. Swallowing and bladder and bowel function are normal.
- Your doctor says you have high blood pressure, high cholesterol and an irregular heart beat, and you take some tablets for all these (you don't have any allergies).
- You had some abdominal pain last year and were found to have a stomach ulcer.
- You live with your wife, who has dementia. You are her main carer.
- You worked as a banker in the past.

## SUGGESTIONS FOR THE CANDIDATE

---

- Ensure that you introduce yourself and check the patient's name and age.
- Ask the patient why they have come to see you and what they know about what has happened so far.
- Ask an open question to start with about the blurred vision.
- Ask about floaters, flashing lights, diplopia, eye pain and the use of glasses.
- Ask specifically about the headache and what precipitates it.
- Ask about other neurological symptoms.
- Show the examiners you are working through your differentials:
  - Glaucoma (sudden onset, nausea and vomiting)
  - Space-occupying lesion (early morning headache, worse on sneezing/coughing/leaning forward)
  - Cerebrovascular disease (sudden-onset symptoms that improve with time, risk factors)
  - Temporal arteritis (headache, scalp tenderness)





- Once you establish the likelihood of temporal arteritis, demonstrate to the examiners that you know it is associated with polymyalgia rheumatica and ask about symptoms such as fatigue and joint stiffness.
- Take a thorough past medical history, as well as a drug history, including allergies.
- Ask about family history.
- Ask about the patient's social situation and ensure that you ask about the impact on their life.
- Be sure to summarise appropriately.
- Ask the patient about any particular concerns or expectations that they might have.
- Explain to the patient that while further investigations are required, treatment (corticosteroids) needs to commence immediately, as the treatment would be sight saving.
- Remember to discuss the potential risks of high-dose steroids. These include steroid-induced diabetes, gastro-oesophageal reflux disease (GORD), osteoporosis, avascular necrosis and steroid-induced psychosis.

## THEMES EXPLORED

---

- Have an open mind as to the diagnosis (this case is about ensuring that you take your own thorough history and not to be swayed by the provisional diagnosis from the GP).
- Associations are important; once you establish the likelihood of giant cell arteritis, show the examiners that you know it is associated with polymyalgia rheumatica.
- A treatment plan should be communicated to the patient where prompt action is required.

## RELEVANT INFORMATION

---

### GIANT CELL ARTERITIS

- A large artery inflammatory granulomatous arteritis.
- It presents with the following symptoms/signs:
  - Headache, either occipital or unilaterally over the temporal region
  - Scalp/temporal tenderness
  - Tenderness of the temporal/occipital arteries
  - Jaw claudication
  - Sudden visual loss
  - Constitutional symptoms: Malaise, fever
- Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are likely to be elevated on testing. Biopsy of the temporal artery may give a definitive diagnosis (skip lesions are common).
- Early treatment with high-dose corticosteroids is imperative. If a patient has visual symptoms, they should be commenced on prednisolone 60 mg OD and seen urgently by an ophthalmologist. Without visual symptoms, the starting dose is 40–60 mg prednisolone OD. Corticosteroids can be tapered down slowly, but treatment with steroids for at least 1–2 years is usual. Provide osteoporosis prophylaxis.

- Low-dose aspirin (75 mg OD) with a PPI for gastric protection should also be commenced if the patient is not already on these medications and does not have contraindications.

## POLYMYALGIA RHEUMATICA

- A disease affecting the muscles around the shoulder, neck and hips. It is far more common in patients over 70. It presents with the following symptoms/signs:
  - Morning muscle stiffness, lasting over 45 minutes
  - Muscle tenderness around the shoulder, neck and hips
  - Fatigue
  - Fever
  - Anorexia and weight loss
- ESR and CRP are generally raised on testing, while creatine kinase (CK) is normal. It should be distinguished from other illnesses, including rheumatoid arthritis.
- As with giant cell arteritis, treatment is with corticosteroids. However, lower doses are required; start at prednisolone 15 mg OD. This is tapered down slowly over 1–3 years. Provide osteoporosis prophylaxis to cover the corticosteroids and for gastric protection.

## REFERENCES

- National Institute for Health and Care Excellence. (2013) Clinical knowledge summary: Polymyalgia rheumatic. Available from <http://cks.nice.org.uk/polymyalgia-rheumatica#!scenario>. Accessed 5 July 2016.
- National Institute for Health and Care Excellence. (2014) Clinical knowledge summary: Giant cell arteritis. Available from <http://cks.nice.org.uk/giant-cell-arteritis#!scenario>. Accessed 5 July 2016.

## PATIENT WITH WEIGHT LOSS

Miss Alfonso is a 29-year-old student who has lost 3 stone in weight over the last 6 months. The weight loss has been unintentional. She complains of associated reduced appetite and lethargy. She is a first-year nursing student and is finding it difficult to complete her studies. A systems review was unremarkable. She has no past medical history except for recurrent urinary tract infections (UTIs), for which she has received several courses of antibiotics and made a good recovery.

Examination reveals a thin female. Her abdomen is soft and nontender, with a clear chest and normal heart sounds. Preliminary blood tests have been sent but are not yet available.

## INSTRUCTIONS FOR THE PATIENT

- You have lost 3 stone in weight in the last 6 months, going from 12 to 9 stone.
- You have not been intending to lose weight.
- You have noticed that you have become increasingly thirsty, to the extent that you take a jug of water to bed.



- You wake up twice at night to pass urine.
- You have noticed that you are passing more urine more often than before.
- You have had two courses of antibiotics in the last 6 months for urinary tract infections.
- You don't have any other symptoms.
- Prior to this, you were fit and healthy. You no take regular medications and have no allergies.
- Your mother has thyroid problems, but there is no other family history.
- You are a nursing student and live with friends (who have also been concerned regarding your weight loss and symptoms).
- You don't smoke or drink alcohol.
- You are worried you may have diabetes.

## SUGGESTIONS FOR THE CANDIDATE

---

- Ensure that you introduce yourself and check the patient's name and age.
- Ask the patient if they know why they have come to see you and what has happened so far.
- Start with an open question about her weight loss.
- Quantify the weight loss and associated symptoms.
- Demonstrate to the examiners that you are covering symptoms of potential causes of weight loss by performing a thorough systems review and particularly looking for the following features:
  - Inflammatory bowel disease: Ask about diarrhoea (including blood and mucus), constipation, mouth ulcers, abdominal pain and perianal disease.
  - Malignancy (including haematological): Ask about bruising, bleeding, night sweats, fever and a thorough systems review to unveil potential sites of malignancy.
  - Diabetes mellitus: Polyuria, polydipsia, nocturia and recurrent infections.
  - Hyperthyroidism: Irritability, eye symptoms, heat intolerance, tremor, sweating and palpitations.
  - Addison's disease: Dizziness and abnormal pigmentation.
  - Inflammatory conditions: Enquire about back pain, joint pain and rashes.
  - Chronic infections (including TB): Ask about travel history and infectious contacts. Ask about night sweats, haemoptysis and fever.
  - Psychiatric: Enquire about the patient's mood.
  - Drug and smoking history.
- Ask about the patient's social situation and ensure that you ask about impact on her life.
- Be sure to summarise appropriately.
- Ask the patient about any particular concerns that they might have (particularly relevant here).
- Suggest that diabetes mellitus is certainly a possible cause, although further investigations are required to diagnose this and rule out other causes.
- Give some initial information about diabetes mellitus. This might include that if she has diabetes, this will likely be type 1, which would require lifelong insulin therapy (starting immediately).

## THEMES EXPLORED

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- Autoimmune diseases are commonly linked, especially in the Practical Assessment of Clinical Examination Skills (PACES) setting.
- Weight loss is not always due to malignancy, especially in young patients. However, it is important to mention this in a differential, with the likelihood higher the older the patient is.
- Cases with a wide differential can be covered in the time period, so don't become disheartened when given a nonspecific symptom.

## RELEVANT INFORMATION

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### WORLD HEALTH ORGANIZATION CRITERIA FOR DIAGNOSIS OF DIABETES MELLITUS

Criteria include either of the following:

- Symptoms of hyperglycaemia and a raised venous glucose (fasting  $\geq 7$  mmol/L or random  $\geq 11.1$  mmol/L)
- Raised venous glucose on two separate occasions (fasting  $\geq 7$  mmol/L or random  $\geq 11.1$  mmol/L or oral glucose tolerance test 2-hour value  $\geq 11.1$  mmol/L)

Note that diabetes can now also be diagnosed with a raised HbA1c; a HbA1c  $\geq 6.5\%$  (48 mmol/mol) is diagnostic in a symptomatic patient, or alongside a raised blood glucose (as above). Of note, this level is also used as the treatment target level in adults to minimise the risk of vascular complications.

### AUTOIMMUNE DISEASE HUMAN LEUCOCYTE ANTIGEN ASSOCIATIONS

- HLA-B27: Ankylosing spondylitis
- HLA-DR3: Type 1 diabetes mellitus, Graves' disease, Hashimoto's thyroiditis, Addison's disease, systemic lupus erythematosus (SLE), myasthenia gravis
- HLA-DR4: Type 1 diabetes mellitus, Hashimoto's thyroiditis, rheumatoid arthritis

## REFERENCES

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- Gough SCL, Simmonds MJ. (2007) The HLA region and autoimmune disease: Associations and mechanisms of action. *Curr Genomics* 8(7): 453–65.
- National Institute for Health and Care Excellence. (2015) Guidance NG17. Type 1 diabetes in adults: Diagnosis and management. Available from <https://www.nice.org.uk/guidance/ng17>. Accessed 5 July 2016.
- World Health Organization. (2006) Definition and diagnosis of diabetes mellitus and intermediate hyperglycaemia. Available from [http://who.int/diabetes/publications/Definition%20and%20diagnosis%20of%20diabetes\\_new.pdf](http://who.int/diabetes/publications/Definition%20and%20diagnosis%20of%20diabetes_new.pdf). Accessed 5 July 2016.
- World Health Organization. (2011) Use of glycated haemoglobin (HbA1c) in the diagnosis of diabetes mellitus. Available from [http://www.who.int/diabetes/publications/report-hba1c\\_2011.pdf](http://www.who.int/diabetes/publications/report-hba1c_2011.pdf). Accessed 5 July 2016.



## HISTORY-TAKING STATION SUMMARY

- This station may give you an opportunity to score highly, but you should *not* take it for granted. Remain focused and target this station for scoring well (which will allow you some leeway on the stations you find more challenging).
- Do the simple things well, be polite and courteous and introduce yourself.
- Use your 5 minutes before you enter the room wisely; plan an approach to the case and look to structure your approach.
- Start with an open question, as the patient may well give you the full history and all the salient points; if they don't, then you will still have time to fill in the gaps.
- Try not to interrupt the patient, but if they are rambling, you must do so subtly and politely; you must practise this beforehand.
- Remember to look for hidden agendas, particularly if it is an apparently easy case (e.g. is it a chronic hypertensive who is noncompliant with meds because of impotence brought on by beta-blockers but who is too embarrassed to tell you this).
- Use all the time you are given. It would be embarrassing to have to sit in silence for 5 minutes if you finish early. Summarising is a useful tool, as it will remind you of things that you still need to ask.
- Allergies are an important question, as you cannot manage your patient until you know if they are allergic to specific medicines.
- Read the instructions very carefully; do not start explaining the management plan unless it specifically asks you to. The likelihood is that they will only want you to take a history. The examiners may then ask you what your approach will be when they question you.
- An easy station to practise, you should do it in groups with one person being the patient, one being the candidate and the others being the examiners.



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# 4

## Station 3: Neurology

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## HINTS FOR THE NEUROLOGY STATION

- Ensure that you pay close attention to the lead-in statement, as this will guide your examination. Once you have decided on a system, be efficient with your time for this.
- Neurology cases lend themselves to being spot diagnoses (e.g. myotonic dystrophy, Parkinson's disease [PD], old polio and myasthenia gravis). Therefore, be alert from the moment you walk into the room.
- Looking around the bed is very relevant in neurology (e.g. calipers, walking aids, devices for measuring forced vital capacity [FVC]).
- If you spot an obvious deformity (claw hand, wrist drop, Charcot's joint), be sure to direct your examination towards this.
- When commenting on power, use the Medical Research Council (MRC) scale to grade it.
- Neurology is often thought of as being a particularly difficult station, but if you work on the premise that you are trying to find the site of the lesion (e.g. cerebrum, cerebellum, brainstem, spinal cord, anterior horn, peripheral nerve or muscle) and then consider the differential diagnosis, you will demonstrate to the examiners that you are organised and pragmatic.
- Have set routines for examining the cranial nerves and the peripheral nervous system (upper limb and lower limb), but be prepared to adapt your examination (e.g. you may be asked to examine just cranial nerves III–VII). Neurology particularly lends itself to this type of adjustment.
- You will earn yourself extra marks if you look for the underlying pathology; e.g. blood glucose pinprick marks, granuloma annulare and cataracts in a case of peripheral neuropathy indicate that diabetes is the underlying cause.
- If you are unable to elicit reflexes, you must demonstrate that you have attempted to elicit them with reinforcement.
- When assessing sensation, be sure to check for normal sensation on the chest first. Compare normal sensation with the peripheral sensation (i.e. ask if the sensation is different to the sensation on the chest, not merely 'Can you feel it?').
- It is important to decide early whether to assess sensation in a dermatomal or a peripheral ('glove and stocking') pattern. This is not always possible. Therefore, start in a dermatomal distribution and move on to test glove and stocking pattern if peripherally confluent.
- Although the patients may have been examined many times and will be well versed in the routines, you must give clear instructions about what you want them to do (remember that they're not allowed to help you).
- At the end of the examination, ask to examine other relevant neurological areas (including fundoscopy).
- If you believe that the diagnosis is related to vascular disease, then also mention that you would like to examine the cardiovascular system for risk factors.





## CEREBELLAR SYNDROME

Please examine this patient who has had problems with his balance, and recurrent falls over the past 6 months.

### FINDINGS

---

- *General:* Action tremor
- *Neurological:* Ataxic gait, truncal ataxia, hypotonia, pendular reflexes, dysmetria, dysdiadochokinesis, nystagmus, slurred speech, heel–shin ataxia
- *Extras:*
  - Features of multiple sclerosis (MS) (spastic paraparesis, sensory disturbance, internuclear ophthalmoplegia [INO])
  - Stigmata of chronic liver disease
  - Friedreich’s ataxia (young patient, wheelchair/walking aids, pes cavus, kyphoscoliosis, absent ankle jerks with upgoing plantars)

### PRESENTATION

---

This man has cerebellar dysfunction. He has an ataxic gait with nystagmus and dysarthria. He also has multiple stigmata of liver disease. The most likely cause is excess alcohol consumption.

### CAUSES OF CEREBELLAR SYNDROME

---

- Multiple sclerosis
- Alcoholic cerebellar degeneration
- Posterior fossa space-occupying lesion
- Brainstem vascular lesion
- Inherited ataxias (e.g. Friedreich’s) (note that the history would be longer than 6 months)
- Paraneoplastic syndromes
- Drugs (phenytoin)

### INVESTIGATIONS

---

- Dependent on the presumed cause of cerebellar signs
- If multiple sclerosis is suspected,
  - Cranial and spinal imaging (magnetic resonance imaging [MRI])
- Lumbar puncture (LP) (oligoclonal bands and protein in cerebrospinal fluid [CSF])
- Visual-evoked potentials

## MANAGEMENT

---

- Dependent on the cause of cerebellar syndrome

## QUESTIONS

---

1. How would you identify the site of a cerebellar lesion from clinical findings?
  - The cerebellum is divided into a midline vermis and two cerebellar hemispheres.
  - Disease of the vermis leads to truncal ataxia and ataxic gait.
  - Disease of a hemisphere causes ipsilateral dysmetria, dysdiadochokinesis, an intention tremor and fast-beat nystagmus towards the lesion.
  - Multiple sclerosis (demyelination) can cause a global deficit.
2. What is Friedreich's ataxia?
  - Autosomal recessive disorder; a trinucleotide repeat on chromosome 9.
  - Degeneration of the spinocerebellar tract resulting in cerebellar signs.
  - Corticospinal tract damage and peripheral nerve degeneration lead to absent ankle jerks with extensor plantars.
  - Pes cavus, scoliosis and diabetes are common features. Other features include cardiomyopathy, cataracts and sensorineural deafness.
  - Think of this in a younger patient.

## KEY POINTS

---

- Prepare a specific examination for the cerebellar syndrome.
- Split the examination into two parts. First, ensure that you have demonstrated that the patient has cerebellar signs. Second, ensure that you have demonstrated that you have looked for specific underlying causes.

## HEMIPARESIS

This patient has developed weakness. Please examine their upper limbs.

## FINDINGS

---

- *General:* Walking aids, percutaneous endoscopic gastrostomy (PEG) tube, wasting/oedema on affected side, upper limbs held in flexion, lower limbs held in extension
- *Peripheral:* Bruising (on warfarin), amiodarone facies, irregularly irregular pulse



- *Neurological:*
  - Increased tone
  - Reduced power (use MRC grade for power)
  - Hyperreflexia and extensor plantars
  - Decreased sensation
  - Hemianopia
- *Extras:*
  - Observe for upper motor neurone (UMN) facial nerve lesion.
  - Note any dysphasia/dysarthria.
  - Neglect/visual or sensory inattention.
  - Mention that you would like to perform a full cardiovascular examination.

## PRESENTATION

---

This patient has a right-sided upper motor neurone lesion affecting arm, leg and face. The likely cause is a stroke, but I would include space-occupying lesion and demyelination in my differential diagnosis. I would like to examine his cardiovascular system, including blood pressure readings, in order to look for a predisposing cause. He mobilises with a frame.

- Comment on the side of stroke and the areas involved.
- Comment on any obvious underlying predisposing factors.
- Comment on functional status.

## INVESTIGATIONS FOR STROKE

---

- Neuroimaging (computerised tomography [CT]/MRI)
- Blood pressure
- Fasting glucose/cholesterol
- Electrocardiogram (ECG)/24-hour tape
- Echocardiogram
- Carotid artery Doppler

## MANAGEMENT OF STROKE

---

- Thrombolysis (provided meets criteria).
- High-dose aspirin for 2 weeks, followed by clopidogrel for secondary prevention.
- Use a tool such as the NIH Stroke Scale to assess patient functionality at onset of stroke and at regular intervals after onset.
- Speech and language assessment.
- Management of predisposing factors and secondary prevention.
- Rehabilitation (occupational therapist [OT] and physiotherapy).
- Psychological and nutritional support.
- Measure progress with tools, e.g. Modified Rankin Score to assess prognosis.

## QUESTIONS

---

1. What extra investigations are merited in young patients with a proven stroke?
  - Thrombophilia screen
  - Homocystine levels (postulated to promote atherosclerosis leading to stroke)
  - Bubble echo (looking for a patent foramen ovale)
2. Do you know of any alternative anticoagulants to warfarin in patients who may have suffered a thrombotic stroke and have atrial fibrillation?
  - Factor 10A inhibitors, e.g. apixaban and rivoroxaban.
  - Dabigatran is a direct thrombin inhibitor (Factor IIa).
3. What is 'lateral medullary syndrome'?
  - This is also known as Wallenberg's syndrome; it is caused by a brainstem stroke in the territory of the vertebral or posterior inferior cerebellar artery.
  - Clinical features include
    - Ipsilateral signs: Horner's syndrome, nystagmus, facial sensory impairment, ataxia and diplopia
    - Contralateral signs: Pain and temperature loss over opposite arm and trunk (spinothalamic tract)
4. Discuss the advantages and disadvantages of novel anticoagulant therapies versus warfarin.
  - Advantages
    - Rapid onset of action – no need for bridging therapy.
    - Fewer interactions with other medications/foods.
    - Provided patients are compliant, they are always in the therapeutic window (unlike warfarin).
    - No need for repeated blood tests to monitor the international normalised ratio (INR).
  - Disadvantages
    - Not safe in renal failure – dose dependent on estimated glomerular filtration rate (eGFR). Consult the British National Formulary (BNF) before prescribing.

## KEY POINTS

---

- Establish the diagnosis and the cerebral area(s) affected.
- Mention to examiners that you would like to examine the cardiovascular system (for risk factors).
- Look for evidence of disability caused by the hemiparesis and comment on this.

## REFERENCE

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National Institute for Health and Care Excellence. (2010) Guidance QS2. Stroke in adults. Available from <https://www.nice.org.uk/guidance/qs2/resources/stroke-in-adults-58292707525>. Accessed 24 August 2016.



## MYOTONIC DYSTROPHY

This patient, who is undergoing preoperative evaluation for elective surgery, is noted to have bilateral ptosis. Please examine their face and proceed.

### FINDINGS

- *General:* 'Myopathic facies', wasting of sternocleidomastoid muscle group, speech (nasal or dysarthric), pinprick marks on fingertips from blood glucose testing
- *Face:* Bilateral ptosis, wasting of facial muscles with hollowing of temporal fossae and cheeks, frontal baldness, smooth forehead, cataracts
- *Hands:* Generalised weakness and wasting of upper limbs
- *Key points:* The candidate must be able to demonstrate myotonia.
  - *Grip myotonia:*
    - Ask the patient to quickly clench their fist as tightly as possible and then immediately release. This will demonstrate a slow release instead of rapid finger extension.
    - This can also be demonstrated by shaking the patient's hand, which will demonstrate slow release of grip.
  - *Percussion myotonia:* This can be demonstrated by using a tendon hammer to tap the thenar eminence, which will display a muscle twitch, followed by a slow relaxation of the muscle group.

### PRESENTATION

On examination, this patient has evidence of myotonic dystrophy. This is demonstrated by the myopathic facies: elongated face, wasting of temporal muscles, frontal balding and bilateral ptosis. I have demonstrated evidence of myotonia, as there was slow release of grip after shaking his hand and percussion myotonia on tapping the thenar eminence.

- Comment on any evidence of complications (see below).

### DIAGNOSIS

- Electromyography (EMG)
- Muscle biopsy
- Genetic analysis: Expansion of cytosine–thymine–guanine (CTG) triple repeat on long arm of chromosome 19

### MANAGEMENT

- Reduction of myotonia using drugs such as phenytoin, mexiletine and carbamazepine (these drugs are sodium channel blockers; they reduce myotonia but may increase weakness)
- Identification and treatment of associated complications (see below)
- Genetic counselling
- Avoidance of general anaesthesia where possible

## QUESTIONS

---

1. List some complications associated with myotonic dystrophy.
  - Cardiac
    - Dilated cardiomyopathy, cardiac arrhythmias
  - Respiratory
    - Risk of aspiration due to muscle weakness; may require noninvasive ventilation due to myotonia affecting respiratory muscles
  - Gastrointestinal
    - Dysphagia, delayed gastric emptying
  - Endocrine
    - Increased risk of diabetes, thyroid dysfunction
  - Reproductive
    - Testicular atrophy, infertility
  - Other
    - Cataracts
  
2. What is the genetic basis of this condition?
  - Autosomal dominant
  - Trinucleotide-repeat disorder showing genetic anticipation (expansion of an unstable CTG trinucleotide repeat in the myotonic protein kinase gene)
  - Affects a gene located on chromosome 19
  
3. What problems are associated with general anaesthesia?
  - Sedatives and neuromuscular blocking drugs may lead to cardiorespiratory complications and delayed recovery from anaesthesia.
  - Depolarising neuromuscular blocking agents should be avoided, e.g. suxamethonium (can lead to induced myotonia).
  
4. What changes would be detected on EMG in myotonic dystrophy?
  - Electromyography shows the electrical potential generated by muscles when they are neurologically or electrically activated.
  - Myotonic dystrophy produces high-frequency activity that varies, producing a whining sound on the loudspeaker ('dive-bomber').
  
5. Describe what is meant by *anticipation*?
  - Anticipation is a phenomenon whereby as a genetic disorder is passed on to the next generation, the symptoms of the genetic disorder become more apparent at an earlier age with each generation.

## KEY POINTS

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- Try to identify myotonic dystrophy early from the characteristic facial changes.
- Be sure to examine for specific signs of myotonia (grip and percussion).
- Look for evidence of extraneurological complications.



## OCULAR PALSIES

Please examine this patient's eyes, as they have been complaining of double vision.

Note: The term *ocular palsy* refers specifically to the loss of function of an ocular muscle due to pathology in the nerve supplying it. We have also considered other causes of diplopia in this section.

### FINDINGS

- *General:* Walking aids, eye patch, ptosis
- *Neurological:*
  - Third (oculomotor) nerve palsy: Complete ptosis, eye looks down and out, pupil may be dilated and unreactive dependent on the cause
  - Fourth (trochlear) nerve palsy: Causes weakness of downward movement of eye, causing vertical diplopia (rare)
  - Sixth (abducens) nerve palsy: Inability to abduct affected eye
  - Complex ophthalmoplegia: A combination/no specific nerve involvement
  - INO: Impaired adduction, unilaterally or bilaterally
- *Extras:*
  - Stigmata of diabetes mellitus
  - Myasthenia gravis: Bilateral ptosis, fatiguability, thymectomy scar
  - Graves' disease: Proptosis, neck lump/scar
  - Multiple sclerosis: Spastic paraparesis, cerebellar signs, walking aids/wheelchair
  - Mitochondrial diseases: Hearing aid, proximal myopathy, ataxia, pacemaker (cardiomyopathy)
  - Miller-Fisher syndrome: Peripheral neuropathy, ataxia, areflexia

### PRESENTATION

This patient has a complex external ophthalmoplegia, as evidenced by diplopia in directions of gaze that are not attributable to a single nerve lesion. The likely cause is myasthenia gravis, as this patient also has bilateral ptosis, demonstrable fatiguability and a midline thoracotomy scar which would be consistent with a prior thymectomy.

- Where possible, try to work out which nerve is the culprit, though bear in mind that the patient may have a complex ophthalmoplegia.

### INVESTIGATIONS

- Neuroimaging if a nerve lesion is suspected. MRI is most helpful, as it is important to obtain good views of the brainstem and posterior fossa.
- Investigations for causes of mononeuritis multiplex (see the section on mononeuropathy).
- Investigations for myasthenia gravis and thyroid disease in a complex ophthalmoplegia.

## MANAGEMENT

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- Dependent on the underlying cause

## QUESTIONS

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1. What are the causes of an oculomotor nerve palsy?
  - 'Surgical': These causes generally affect the pupil.
    - Posterior communicating artery aneurysm
    - Space-occupying lesion in midbrain/sphenoid wing/near cavernous sinus
    - Haemorrhage
  - 'Medical': These causes often do not affect the pupil.
    - Causes of mononeuritis multiplex
    - Demyelination
    - Infarction
2. What are the causes of an abducens nerve palsy?
  - Causes of mononeuritis multiplex.
  - Vascular lesion.
  - Malignancy.
  - Demyelination.
  - Infection (Lyme disease, syphilis).
  - Raised intracranial pressure ('false localising' sign).
  - Wernicke's encephalopathy can cause a bilateral abducens nerve palsy.
3. What are the causes of a complex ophthalmoplegia?
  - Nerve lesions: Demyelination, mononeuritis multiplex
  - Neuromuscular junction: Myasthenia gravis
  - Muscle: Graves' disease
  - Mitochondrial disease
4. What are the causes of an internuclear ophthalmoplegia?
  - Multiple sclerosis
  - Vascular disease

## KEY POINTS

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- Attempt to work out if the ocular palsy is due to a specific nerve lesion. If this is not possible, consider a complex ophthalmoplegia.
- The pupil may give some idea towards aetiology in an oculomotor palsy.





## PARKINSON'S DISEASE

Please examine this patient's gait and proceed. They have presented with unsteadiness.

### FINDINGS

- *General:* Patients can be in the 'off' medication state (poverty of facial expression, slowness of movement, reduced blinking, etc., are clues prior to examination) or in the 'on' medication state (excessive choreodystonic movements due to levodopa-induced dyskinesia).
- *Neurological:*
  - Resting tremor (4–6 Hz), rigidity (can be cog-wheeling in presence of rest tremor), bradykinesia (progressive decrement of amplitude of repetitive movements such as finger tapping).
  - Gait: Ignition failure, short stride length, festination, freezing.
  - Soft and monotonous speech.
  - Micrographia.
  - Rigidity can be demonstrated with concurrent activity (synkinesis – waving the right arm when assessing for the left). Distraction can bring out rest tremor – arm tremor is sometimes only revealed when testing gait or asking the patient to do a cognitive task like serial seven subtraction.
- *Extras:*
  - Advanced therapies: Infusion pumps – apomorphine (subcutaneous) or duodopa (via percutaneous endoscopic jejunostomy [PEJ]), deep brain stimulation (implantable pulse generator or chest or abdominal wall – resembles cardiac pacemaker).
  - Full assessment should also include mood, cognition (nonmotor symptoms) and functional status.
  - Demonstrate that you are aware of Parkinson-plus syndromes by assessing eye movements and cerebellar signs.

### PRESENTATION

This patient has evidence of Parkinson's disease. There is a resting tremor, as well as rigidity and bradykinesia. I note an apomorphine infusion being administered via a pump.

- This is a common presentation at the neurology station and should be recognisable from the classic triad of parkinsonism: bradykinesia, rigidity and a resting tremor.
- Be sure to discuss the functional status of the patient and consider causes other than just Parkinson's disease.
- Red flags for Parkinson-plus syndromes include symmetrical onset, early falls (patient in a wheelchair), lack of levodopa responses, supranuclear gaze palsy (limited downgaze) and cerebellar signs.

## CAUSES OF PARKINSONISM

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- Parkinson's disease.
- Drugs (neuroleptics).
- Parkinson-plus syndromes: Progressive supranuclear palsy (Steele–Richardson–Olszewski syndrome), multisystem atrophy (Shy–Drager syndrome).
- Stroke or space-occupying lesion affecting the basal ganglia.
- Postencephalitis.
- Wilson's disease.
- Other disorders resembling parkinsonism include dementia (including normal-pressure hydrocephalus) and tremulous conditions like benign essential tremor (action rather than rest tremor, i.e. postural and kinetic tremor).

## INVESTIGATIONS

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- Parkinson's disease is generally a clinical diagnosis.
- Supportive evidence may be provided by a therapeutic trial of levodopa or an apomorphine challenge.
- Cerebral imaging may be appropriate where other diagnoses require exclusion.
- A dopamine transporter (DaT) scan can be used to support a diagnosis of Parkinson's disease or to rule out drug-induced parkinsonism (but cannot distinguish idiopathic PD from PD-plus syndromes).

## MANAGEMENT

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- Multidisciplinary team (MDT) approach
  - Specialist, physiotherapist, OT, specialist nurse, speech and language therapist (SALT)
- Levodopa formulations
- Other medications
  - Dopamine agonists, e.g. rotigotine patch, ropinirole, pramipexole
  - Anticholinergics: Trihexyphenidyl (for tremor)
  - Selegiline, rasagiline (monoamine oxidase B [MAOB] inhibitors)
  - Tolcapone (rarely used due to hepatic side effects), entacapone, opicapone (new) (catechol-o-methyltransferase [COMT] inhibitors)
  - Apomorphine (dopamine agonist)
- Complications
- Assess disability and cognition regularly; treat depression
- Surgery
  - For example, deep brain stimulation of subthalamic nucleus or internal segment of globus pallidus may be considered for those with motor fluctuations or levodopa-induced dyskinesia.



## QUESTIONS

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1. What are the characteristic features of Parkinson's disease?
  - The triad of parkinsonism (bradykinesia, rigidity, resting tremor).
  - In Parkinson's disease, these features begin asymmetrically and generally affect the upper limbs first. There is fluctuation in severity, with the patient being better some days than others.
  - Autonomic dysfunction, loss of postural reflexes and balance and changes in higher mental functioning are also common later in the illness.
2. What drugs other than L-dopa are used in the management of Parkinson's disease?
  - Dopamine agonists (e.g. ropinirole): May be used as first-line therapy, especially in younger patients (due to the decreased risk of dyskinesias), or as an add-on to levodopa.
  - Anticholinergics (e.g. trihexyphenidyl): Useful for tremor.
  - Monoamine oxidase B inhibitors (selegiline): May be helpful with motor symptoms.
  - COMT inhibitors (entacapone): Help to decrease immobility by shortening the 'off' time associated with L-dopa.
  - Apomorphine: A parenteral dopamine agonist that can be helpful with 'on-off' effects. It may be given via a syringe driver.
3. What is the pathology underlying Parkinson's disease?
  - Degeneration of the substantia nigra dopaminergic neurones in the basal ganglia. The hallmark is the presence of Lewy bodies.
4. Describe the potential problems patients need to be counselled about before commencing dopamine agonists.
  - Impulse control disorders
    - Hyperreligiosity
    - Gambling
    - Hypersexuality
    - Compulsive eating

## KEY POINTS

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- Have a specific examination prepared for Parkinson's disease.
- Look for the triad of parkinsonism early, and then search for extra features.

## REFERENCE

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Benabid AL, et al. (2009) Deep brain stimulation of the subthalamic nucleus for the treatment of Parkinson's disease. *Lancet Neurol* 8(1): 67–81.

## PERIPHERAL NEUROPATHY

Please examine this patient who has painful legs.

### FINDINGS

- *General:* Walking aids, diabetic shoes, prostheses, insulin pen
- *Neurological:*
  - 'Glove and stocking' sensation loss
  - All sensory modalities should be tested (use a 128 Hz tuning fork to assess vibration sense, as dorsal columns are usually affected first)
  - Wasting, weakness, areflexia
- *Extras:*
  - Finger-prick testing, cataracts, ulcers, Charcot's joints, callus (diabetes mellitus)
  - Clawing of the toes, pes cavus (Charcot-Marie-Tooth disease)
  - Amiodarone facies
  - Anaemia (B12 deficiency)
  - Evidence of alcohol abuse
  - Evidence of arthritis and rashes (vasculitis)

### PRESENTATION

This patient has a predominantly sensory peripheral neuropathy, as evidenced by lack of sensation bilaterally to midcalf for all modalities. There is no evidence of any ulceration or callus formation. The most likely underlying cause is diabetes mellitus, as evidenced by finger pulp pricks from capillary blood glucose testing.

- It is important to first ensure that the patient has a peripheral neuropathy rather than a mononeuropathy or mononeuritis multiplex.
- This is a common Practical Assessment of Clinical Examination Skills (PACES) case, and looking for a cause is crucial. Try to decide if the neuropathy is predominantly motor or sensory, as this should help to elucidate the aetiology.

### CAUSES OF NEUROPATHY

- Predominantly sensory neuropathy
  - Diabetes mellitus
  - Alcohol
  - Drugs
  - Vitamin deficiencies (B1, B12)
  - Uraemia
- Predominantly motor
  - Guillain-Barré syndrome
  - Malignancy
  - Charcot-Marie-Tooth disease
  - Porphyria
  - Lead poisoning



- Other causes include paraneoplastic syndromes, paraproteinaemia, vasculitis and infections (human immunodeficiency virus [HIV], Lyme disease).
- Some cases are idiopathic.

## INVESTIGATIONS

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- Full drug and alcohol history
- Blood tests: Full blood count (FBC) (including mean cell volume [MCV]), urea and electrolytes (U&Es), liver function tests (LFTs) (including gamma-glutamyl transpeptidase [GGT]), vitamin B12 and folate, glucose, thyroid function tests (TFTs), autoimmune screen and immunoglobulins, hepatitis screen, HIV screen, Lyme serology, syphilis serology
- Urine: Dip for glucose and protein, Bence–Jones protein
- Imaging: Chest X-ray (CXR)
- LP and CSF study: Protein and CSF virology
- Nerve conduction studies

## MANAGEMENT

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- Dependent on the aetiology; remove any precipitants and treat the cause.

## QUESTIONS

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1. What is an autonomic neuropathy?
  - A neuropathy of the autonomic nervous system.
  - May present alone or in conjunction with a motor or sensory neuropathy. The most common cause is diabetes.
  - May present with postural hypotension, impotence, urinary retention, diarrhoea/constipation and a Horner's syndrome.
2. What is Charcot–Marie–Tooth disease?
  - A hereditary, sensory and motor neuropathy. Also known as peroneal muscular atrophy.
  - Usually starts at puberty with foot drop and weak legs.
  - The peroneal muscles are the first to atrophy, with upper limb signs appearing at a later stage.
  - There is muscle wasting, pes cavus and a bilateral foot drop (high-stepping gait). Reflexes are often absent. Sensory loss is variable.
  - The most common form is inherited in an autosomal dominant manner.
3. Which drugs can cause a peripheral neuropathy?
  - Amiodarone
  - Gold
  - Isoniazid
  - Metronidazole

- Nitrofurantoin
  - Phenytoin
4. What is tabes dorsalis? Describe the gait associated with this.
- Tabes dorsalis is a demyelinating condition affecting primarily the nerves in the dorsal/posterior columns of the spinal cord. These nerves normally help maintain proprioception, vibration and discriminative touch. The gait is 'high stepping' due to a lack of proprioception.

## KEY POINTS

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- Establish the diagnosis early to allow a cause to be found.
- Be sure to present a list of possible aetiologies to the examiner, taking into account whether the neuropathy is predominantly motor or sensory.
- The most likely causes in PACES include diabetes and Charcot–Marie–Tooth disease.

## MONONEUROPATHIES

Please examine this patient who has paraesthesiae in his left hand.

## FINDINGS

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### CARPAL TUNNEL SYNDROME

- Wasting of the thenar eminence.
- Weakness of LOAF (lumbricals, opponens pollicis, abductor pollicis brevis, flexor pollicis brevis).
- Sensory loss over the lateral 3½ digits.
- Maximal wrist flexion for 1 minute may elicit symptoms (Phalen's test).
- Tapping over the nerve at the wrist induces tingling (Tinel's test).
- Look for a scar from carpal tunnel release surgery and evidence of diabetes, hypothyroidism, acromegaly and rheumatoid arthritis.

## SPECIFIC NERVE PALSIES

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### ULNAR NERVE PALSY

- Wasting of the hypothenar eminence (thenar eminence spared), claw hand, guttering on the dorsal aspect of the hand.
- Look for wasting of the medial aspect of the forearm (note low/high lesions, hand less clawed in high lesion).
- Weakness of abduction and adduction (test for Froment's sign) of the fingers and adduction of the thumb.
- Sensory loss over the medial 1½ digits.
- Look for scars (fracture dislocation) and osteoarthritis at the elbow.



## RADIAL NERVE PALSY

- Wrist drop, weakness of wrist extension; if the wrist is passively extended, intrinsic muscles of the hand should be intact.
- Impaired grip strength.
- Sensory loss over the first dorsal interosseous.
- Look for scars at the elbow (fracture/dislocation), and note if the patient uses crutches.

## COMMON PERONEAL NERVE PALSY

- Foot drop on inspection (leading to a high-stepping gait).
- Weakness of dorsiflexion and eversion of the foot.
- All reflexes will be intact.
- Sensory loss over the lateral dorsum of the foot.
- Look for evidence of compression around the fibular neck.

## PRESENTATION

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This patient has wasting of the thenar eminence in the left hand, indicating a median nerve lesion. This is supported by weakness of opposition of the thumb. There is sensory loss over the lateral 3½ digits with sparing of the palm. Tinel's test is positive. There is a scar over the anterior wrist. The patient also has a symmetrical arthritis affecting the hands with a swan neck deformity of two digits, but no evident skin or nail changes. The likely diagnosis is carpal tunnel syndrome of the left wrist on a background of rheumatoid arthritis.

- Functionality is crucial and should always be tested and commented on. Try to find a cause for the lesion.
- In all mononeuropathies, it is important to look for causes of mononeuritis multiplex.

## INVESTIGATIONS

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- Single nerve lesions are often a clinical diagnosis. Neurophysiology can be used to confirm the diagnosis and to assess severity.

## MANAGEMENT

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- Splints and physiotherapy can be helpful. Surgery is sometimes used, especially in carpal tunnel syndrome where surgical decompression of the flexor retinaculum is a simple and definitive treatment.

## QUESTIONS

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1. What is the differential diagnosis of a foot drop?
  - Common peroneal nerve palsy, peripheral neuropathy (especially Charcot-Marie-Tooth disease), sciatic nerve palsy, L4/5 radiculopathy (prolapsed lumbar disc) and lumbosacral plexopathy

2. What are the causes of mononeuritis multiplex?
  - Wegener's granulomatosis.
  - Amyloidosis.
  - Rheumatoid arthritis.
  - Diabetes mellitus.
  - Sarcoidosis.
  - Polyarteritis nodosa.
  - Leprosy.
  - Carcinomatosis.
  - Note that WARDS PLC is a useful way of remembering this list.
3. What are the causes of wasting of the small (intrinsic) muscles of the hand?
  - Resembles an ulnar nerve lesion, but with thenar wasting and weakness also
  - Causes include
    - Anterior horn cells disease, e.g. poliomyelitis
    - Radiculopathy, e.g. trauma, prolapsed disc
    - Plexopathy, e.g. brachial plexus injury, Pancoast's tumour, cervical rib
    - Peripheral nerve lesions
    - Muscle, e.g. disuse atrophy

## KEY POINTS

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- Initially look for any evident wasting or deformity.
- Examine all motor and sensory areas supplied by the nerve to localise the lesion.
- Search for a possible cause.

## MOTOR NEURONE DISEASE

Please examine this patient who presents with weakness.

## FINDINGS

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- Amyotrophic lateral sclerosis (ALS)
  - Suspect when UMNs and lower motor neurones (LMNs) occur concurrently in a limb with weakness.
  - Degree of weakness depends on the number of muscles affected and distribution of motor neurone loss.
  - Reflexes usually exaggerated (UMN signs) in the presence of muscle wasting and fasciculation (LMN signs).
  - Sensation is unaffected throughout.
  - Involvement of lower cranial nerves causes a pseudobulbar palsy.
- Progressive muscular atrophy (PMA)
  - Flaccid weakness, as only LMNs affected
  - Fasciculations and wasting





- Decreased or absent reflexes
- Plantars down going
- Primary lateral sclerosis (PLS)
  - UMN signs only
  - Usually begins in lower limbs (spastic gait)
  - Exaggerated reflexes
- Progressive bulbar palsy
  - Only lower cranial nerves affected (IX, X, XII).
  - 'Donald Duck'/nasal speech.
  - Weakness of palatal muscles results in swallowing difficulties.

## PRESENTATION

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The most likely diagnosis is motor neurone disease (MND). There is generalised wasting and fasciculation. Tone is increased with generalised weakness. Reflexes are brisk/reduced/absent. Leg reflexes are brisk and ankle clonus is present. There are bilateral extensor plantar responses.

## INVESTIGATIONS

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- Diagnosis is mainly clinical, based on a high index of suspicion from the collection of signs and symptoms.
- EMG
  - Abnormally slowed conduction due to reduction in the number of viable motor axons/anterior horn cells to activate the muscle(s) involved
- Nerve conduction studies
  - Normal sensory nerve conduction and abnormal motor nerve conduction
  - Reduced muscle action potentials
  - Repetitive stimulation: Decremental response with slow repetitive stimulation
- MRI
  - To exclude other causes for the symptoms, e.g. cervical myelopathy/spondylosis or cord compression

## MANAGEMENT

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- Supportive measures
  - Physiotherapy, occupational therapy, speech therapy.
  - Swallowing and nutritional support (nasogastric [NG]/PEG feeding).
  - Respiratory support: Noninvasive positive-pressure ventilation (NIPPV), tracheostomy and invasive ventilation.
  - Advanced care planning is an important part of the patient's treatment.
- Specific therapy
  - Riluzole: A glutamate inhibitor that acts by inhibiting voltage-gated sodium channels
  - Increases survival compared with placebo by roughly 3 months; no significant effect on muscle strength or neurological function seen

## QUESTIONS

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1. What are the disease variants of MND?
  - Amyotrophic lateral sclerosis
    - Approximately 50% of cases; combined UMN and LMN signs; mostly sporadic (90%–95%)
    - Familial form: Copper/zinc superoxide dismutase (SOD-1) gene mutation on chromosome 21
  - Primary lateral sclerosis
    - Rare; affects upper motor neurones only; has the best prognosis but can later progress to ALS
  - Progressive muscular atrophy
    - Approximately 25% of cases; affects anterior horn cells only – therefore signs in distal muscle groups
  - Progressive bulbar palsy
    - Approximately 25% of cases; worst prognosis; affects suprabulbar nuclei and lower cranial nerves, resulting in speech and swallowing difficulties (increased risk of aspiration)
2. What is the prognosis for patients with MND?
  - No known cure; usually fatal within 3–5 years of diagnosis. Cause of death is usually aspiration pneumonia and/or ventilatory failure.
3. What is the differential diagnosis for MND?
  - Degenerative: Cervical cord compression, cervical spondylosis
  - Inflammatory/traumatic/inherited: Syringomyelia, spinal muscular atrophy
  - Infectious: Polio, syphilis
  - Malignant/paraneoplastic
4. What are the other causes of absent ankle jerks and extensor plantar responses?
  - Hereditary cerebellar ataxias: Friedreich's ataxia, spinocerebellar ataxia
  - Syphilitic taboparesis
  - Subacute combined degeneration of the cord
  - Conus medullaris pathology
  - Combined pathologies, e.g. peripheral neuropathy from any cause in a patient with cervical spondylosis

Note that question 4 is a 'classic' PACES question and should be committed to memory.

## KEY POINTS

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- Note any external features that may indicate the diagnosis (fasciculations, NIPPV machine).
- Note the patient's speech.
- It is crucial to check the plantar response.



## REFERENCE

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National Institute for Health and Care Excellence. (2011) TA20. Guidance on the use of riluzole (Rilutek) for the treatment of motor neurone disease. Available from <https://www.nice.org.uk/guidance/ta20/resources/guidance-on-the-use-of-riluzole-rilutek-for-the-treatment-of-motor-neurone-disease-2294449469125>. Accessed 24 August 2016.

## MULTIPLE SCLEROSIS

Please examine the legs of this patient who has progressive difficulty in walking.

## FINDINGS

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- *General*: Walking sticks, wheelchair, catheter, ataxic gait, dysarthria, mood (depressed/elated)
- *Eyes*: Internuclear ophthalmoplegia, optic neuritis, central scotoma, loss of colour vision, relative afferent pupillary defect (RAPD), nystagmus
- *Neurological*: Spastic paraparesis, cerebellar signs

## PRESENTATION

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This patient has an ataxic gait. Examination of her lower limbs shows increased tone and brisk reflexes. Further examination shows that the patient also has evidence of bilateral internuclear ophthalmoplegia. Putting these findings together, this patient is likely to have a diagnosis of multiple sclerosis.

- Comment on the patient's functionality by looking for general findings, e.g. walking aids, the presence of a catheter and the patient's general affect.

## INVESTIGATIONS

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- CSF analysis
  - Immunoglobulin (Ig) G oligoclonal bands on electrophoresis
- Visual-evoked potentials
  - Delayed response
- MRI
  - Highlighting areas of demyelination

## MANAGEMENT

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- Multidisciplinary approach/patient education/MS support group information
- Acute relapse
  - IV methylprednisolone: This may help to reduce the duration and severity of the relapse but will not alter the course of the disease.

- Disease-modifying drugs
  - Interferon beta-1a
  - Interferon beta-1b
  - Glatiramer acetate
  - Azathioprine
  - Natalizumab (for highly active relapsing–remitting disease)
- Symptomatic treatment
  - Spasticity: Physiotherapy, baclofen, tizanidine
  - Urinary dysfunction: Oxybutynin, catheterisation
  - Constipation: Laxatives, enemas
  - Pain: Amitriptyline, carbamazepine, gabapentin
  - Fatigue: Amantadine
  - Depression: Support groups, selective serotonin reuptake inhibitors (SSRIs)

## QUESTIONS

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1. What is multiple sclerosis?
  - Multiple sclerosis is a chronic inflammatory autoimmune disease of the central nervous system. The diagnosis depends on demonstrating at least two demyelinating lesions in the brain or spinal cord on MRI, separated in time and space.
2. What are Lhermitte’s sign and Uhthoff’s phenomenon?
  - Lhermitte’s sign: Flexion of the neck causing an ‘electric shock’–like sensation in the trunk and limbs; this occurs in cervical spondylosis as well as MS.
  - Uhthoff’s phenomenon: An increase in the severity of symptoms (mainly visual), commonly precipitated by an increase in temperature or by exercise.
3. How is multiple sclerosis classified?
  - Relapsing–remitting
    - Affects approximately 85% of MS sufferers
  - Secondary progressive
    - Follows a period of relapsing–remitting MS
  - Primary progressive
    - 15% of cases; progressive deterioration from the start
  - The classification of MS becomes important when considering the role of disease-modifying antirheumatoid drugs (DMARDs). Currently, these drugs can help reduce the severity and frequency of relapses in relapsing–remitting MS and secondary progressive MS.

## KEY POINTS

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- A combination of spastic paraparesis and cerebellar signs is likely evidence of multiple sclerosis.
- Always comment on the patient’s functional status.



## REFERENCE

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National Institute for Health and Care Excellence. (2014) Guidance CG186. Multiple sclerosis in adults: Management. Available from <https://www.nice.org.uk/guidance/cg186/resources/multiple-sclerosis-management-of-multiple-sclerosis-in-primary-and-secondary-care-35109816059077>. Accessed 24 August 2016.

## SPASTIC PARAPARESIS

Please examine this patient's legs neurologically, as they are finding it difficult to walk.

## FINDINGS

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- *General:* Walking aids, scissoring gait
- *Neurological:* Increased tone, clonus, decreased power, hyperreflexia, extensor plantars and sensory loss. Signs may be present only below a particular spinal level.
- *Other:* Evidence of cerebellar syndrome
  - Cachexia and other evidence of malignancy

## PRESENTATION

---

On examination, this patient has signs suggestive of a spastic paraparesis. This is evidenced by hypertonia, hyperreflexia and decreased power throughout the lower limbs. There is tenderness over the T12 vertebra with a sensory level present, making spinal cord compression a likely cause.

- It is important to exclude acute spinal cord compression, so tell the examiners you would ask about bladder and bowel symptoms, and would offer to perform a pulmonary regurgitation (PR) examination.
- Always check gait and assess the functional status of the patient.

## CAUSES OF A SPASTIC PARAPARESIS

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- Compression
  - Tumour, osteoarthritis, trauma/fracture, central disc prolapse
- Transverse myelitis
  - Multiple sclerosis (cerebellar signs), inflammatory and vascular disorders
- Degenerative
  - Hereditary spastic paraparesis, motor neurone disease (absence of sensory signs/ combination of UMN and LMN signs), Friedreich's ataxia (cerebellar signs)
- Infective
  - HIV myelopathy
- Others
  - Cerebral palsy, subacute combined degeneration of the cord

## INVESTIGATIONS

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- MRI of the spine is the gold standard imaging test.
- Other investigations include FBC, vitamin B12/folate levels, erythrocyte sedimentation rate (ESR)/C-reactive protein (CRP), syphilis serology, CSF protein and oligoclonal bands, myeloma screen and tumour markers.

## MANAGEMENT

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- The most urgent treatment is needed for spinal cord compression:
  - If malignancy is the cause, give dexamethasone. This is followed by radiotherapy or surgery.
  - Surgery is the mainstay of treatment for other causes.
  - Further management of this presentation depends on the underlying cause, although an MDT approach focusing on neurorehabilitation may be helpful.

## QUESTIONS

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1. What malignancies are most likely to cause spinal cord compression?
  - The most likely cause is metastasis from lung, breast or prostate cancer. Kidney and thyroid primaries also commonly metastasise to bone.
  - Multiple myeloma should be considered a cause of cord compression.
  - Intrinsic spinal malignancy is rare.
2. What is transverse myelitis?
  - Inflammation of the spinal cord, characterised by axonal demyelination. Generally, the inflammation is across the thickness of the cord.
  - Symptoms come on over a period of hours to weeks.
  - The most common cause is multiple sclerosis. Viral and other infections can be implicated. Often, no cause is found.
  - Steroids and plasma exchange may be helpful in management, although more crucial is neurorehabilitation.
3. What is the pathology in hereditary spastic paraparesis?
  - Axonal degeneration that is maximal in the terminal portions of the longest descending and ascending tracts, including the crossed and uncrossed corticospinal tracts to the legs and fasciculus gracilis. Neuronal cell bodies of degenerating fibres are preserved, and there is no evidence of primary demyelination.

## KEY POINTS

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- When finding a spastic paraparesis, it is important to look for a spinal level.
- Be sure to assess gait and examine the spine locally.
- Excluding acute cord compression is essential.



## VISUAL FIELD DEFECTS

Please examine this patient who has recently developed a visual disturbance and, as a result, finds that they are bumping into things.

- Sit at the same height as the patient. Ask the patient to cover each eye in turn.
- Assess both nasal and temporal fields and all four quadrants. Look carefully at where the field of vision starts/finishes and which areas of the field are lost, i.e. nasal/temporal, superior/inferior.
- Use the information collected to establish which pattern of visual field defect the patient has.
- Be sure you are familiar with the visual pathway anatomy to enable you to ascertain the site of the lesion, as the nature of the visual field defect is determined by the site of the lesion along the visual pathway.

## PRESENTATION

This patient presents with a history of bumping into objects and visual impairment. They have visual loss affecting the same half of the visual field in each eye. This is known as a homonymous hemianopia.

## CLASSIFICATION OF VISUAL FIELD DEFECTS

- Classification of defects depends on whether one or both eyes are affected, which half of the visual field is affected, i.e. temporal, nasal or both (heteronymous or homonymous hemianopia), and the extent of field affected in each eye relative to the other (congruity).
- Optic nerve lesions: Result in partial or complete visual loss on the side of the lesion.
- Optic chiasm lesions: Result in both temporal fields being lost.
- Optic radiation lesions: Result in homonymous field defects that depend on the location of the lesion in the temporal or parietal lobe.
- Temporal lobe lesions lead to superior homonymous quadrantanopias.
- Parietal lobe lesions lead to inferior homonymous quadrantanopias.
- The further back towards the visual cortex the defect, the greater the degree of congruity (i.e. both eyes affected to the same degree).
- Visual cortex or optic radiation lesions result in a homonymous hemianopia.

## QUESTIONS

1. What are the causes of a homonymous hemianopia?
  - Vascular: Stroke
  - Trauma
  - Tumour
  - Infection: Encephalitis
  - Demyelination: MS

2. What are the causes of an optic nerve lesion?
  - Trauma
  - Inflammatory: Demyelination, optic neuritis
  - Compression: Raised intraocular or intracranial pressure
  - Vascular: Acute optic artery ischaemia
  - Metabolic: B12 deficiency, diabetes, alcohol excess
  - Inherited conditions, e.g. Leber's optic neuropathy
3. What are the common causes of a bitemporal hemianopia?
  - Usually occur as a result of a lesion of the optic chiasm
    - Pituitary fossa tumour
    - Craniopharyngioma
    - Suprasellar meningioma

## KEY POINTS

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- Remember to assess both temporal and nasal visual fields for both eyes.
- Be familiar with visual pathways so the site of the lesion can be identified.

## NEUROLOGY STATION SUMMARY

- Neurology is a difficult station, particularly if you don't look well practiced. You must practise your examination technique until you really are slick and confident performing the examination.
- As ever, particular emphasis should be paid to observation. In neurology, many diagnoses can be made from the end of the bed (Parkinson's/myasthenia gravis/myotonic dystrophy). When this is the case, don't relax. You still need to demonstrate all the signs to the examiner.
- Looking around the bed is just as important in neurology as any other station (e.g. look for forced expiratory volume in first second [FEV1] monitors in myasthenia gravis).
- Don't panic if you don't know the diagnosis; work out where the lesion is and use a pathological sieve to create a differential list.
- Be aware of how to demonstrate specific signs for specific syndromes (e.g. myotonia in myotonic dystrophy). Your examiners will be impressed if you look confident and rehearsed while doing this.
- Go into the station prepared to do a range of examinations. You may have to examine the arms, the legs, the eyes or the cranial nerves or conduct a special examination.
- Don't forget to reinforce the reflexes if you think that they are absent.
- Don't forget to check sensation on the chest prior to commencing limb examination, so that comparison can be made. *But*, remember to check that it feel's normal on the chest wall and not just whether the patient can feel you touching.
- Asking the patient to walk so that you can assess their gait is a key skill in neurology. Be aware of the different types of gait and the different syndromes that each gait may indicate.
- When presenting your case, first tell the examiners the syndrome or the likely site of the lesion, followed by the likely cause.



# 5

## Station 3: Cardiology

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## HINTS FOR THE CARDIOLOGY STATION

- Be alert to any peripheral signs, as these will help guide you to the diagnosis by the time you reach for your stethoscope.
- Check the radial pulse. Comment on rate, rhythm, character or absence.
- Look at the carotid pulsation from the end of the bed; a 'dancing carotid' signifies Corrigan's sign seen in aortic regurgitation (AR).
- Examine the jugular venous pressure (JVP) with the patient resting at a 45°; use the hepatojugular reflex if not visible.
- Giant V-waves in the JVP may indicate tricuspid regurgitation (TR).
- Feel for a parasternal heave (indicative of right ventricular hypertrophy).
- Check for a mitral valvotomy scar or subcutaneous implantable cardioverter defibrillator (ICD) under the left breast. These can be easily missed.
- Always comment on the position of the apex beat and demonstrate the position by marking it out on examination, and re-establish its location in the left lateral position before auscultation.
- If you cannot feel the apex beat in the fifth intercostal space/midclavicular line, the patient may have evidence of cardiomegaly or dextrocardia.
- Listen from the end of the bed for an audible click from metallic heart valves, and comment on any evidence of valvular regurgitation or endocarditis.
- On auscultation, there may be more than one murmur; try to identify the dominant one.
- Ensure that you expose the legs up to the thighs at the end of the examination to look for any scars from vein grafts and evidence of peripheral vascular disease, and to check for peripheral oedema.
- Comment on any evidence of infective endocarditis (IE), heart failure and other significant comorbidities.

## AORTIC STENOSIS

This patient has had episodes of collapse and chest pain. Please examine their cardiovascular system.

### FINDINGS

- *General:* Dyspnoea
- *Peripheral:* Slow-rising pulse, low systolic blood pressure, narrow pulse pressure
- *Chest:* Palpable thrill over the aortic area, heaving apex beat, ejection systolic murmur loudest over the aortic region radiating to the carotids, soft or absent A2, reversed splitting of S2, S4

### PRESENTATION

This patient has aortic stenosis (AS) as evidenced by a slow-rising pulse, narrow pulse pressure, heaving apex beat and an ejection systolic murmur that radiates to the carotids. I believe in this case it is a severe stenosis, as the aortic second heart sound is inaudible and I can hear a fourth heart sound. There is no evidence of congestive cardiac failure (CCF).



- Look for evidence of congestive cardiac failure, a displaced apex and associated mitral regurgitation (MR) (or Gallavardin's phenomenon due to radiation of the murmur through the left atrium (LA) to the apex mimicking MR), and signs of pulmonary hypertension (left parasternal heave, loud A2).
- Try to comment on the severity of the murmur.

## DIFFERENTIAL DIAGNOSIS

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- Aortic sclerosis (normal A2)
- Hypertrophic cardiomyopathy (HCM) (look for jerky pulse, double apical impulse, thrill and ejection systolic murmur loudest at left lower sternal edge)
- Systolic flow murmur (soft, heard in all valve areas, especially in young women)
- Pulmonary stenosis
- Patient prosthesis mismatch; e.g. an aortic prosthetic valve is too small for the patient (presents with same symptoms and signs as AS)
- Radiotherapy

## INVESTIGATIONS

---

- Diagnosis
  - Electrocardiogram (ECG) (large LA, voltage left ventricular hypertrophy [LVH], left bundle branch block [LBBB]).
  - Chest X-ray (CXR) (may be normal, or an unfolded aorta).
  - Echocardiogram is the investigation of choice (transoesophageal echocardiogram [TOE] is rarely helpful unless concomitant mitral valve [MV] abnormality). Ascertain concomitant valvular lesions, left ventricle (LV) dimensions and function (provides prognostic information).
- To determine severity of AS, Doppler echo is preferred.
  - Cardiac catheterisation (underlying coronary artery disease)

## MANAGEMENT

---

- Medical
  - Regular follow-up and echocardiograms (timing for intervention)
  - Diuretics, digoxin, angiotensin-converting enzyme (ACE) inhibitors or angiotensin receptor blockers (ARBs) for heart failure
  - Statins for secondary prevention of atherosclerotic events (statins do not affect progression of disease)
- Surgical
  - Aortic valve replacement (AVR) ( $\pm$  coronary artery bypass graft [CABG] if indicated) is the definitive treatment of symptomatic severe aortic stenosis. Surgery improves quality and quantity of life.
  - Balloon valvuloplasty has a limited role in adult AS, as restenosis occurs within 6–12 months and complication rates exceed 10%. It is only very rarely used as a bridge to surgery or transcatheter aortic valve implantation (TAVI) in unstable patients, or in pregnancy or as a palliative procedure.

- TAVI considered if open AVR is too high risk (transfemoral or apical). Main complications include stroke, pacemaker insertion and vascular complications (including paravalvular regurgitation). One-year survival is 60%–80%, depending on severity of comorbidities.

## QUESTIONS

---

1. What are the causes of aortic stenosis?
  - Common
    - Calcific degeneration (or 'calcific AS')
    - Bicuspid valve
  - Rare
    - Rheumatic fever
    - HCM
    - Congenital (other than bicuspid): Supravalvular stenosis (Williams syndrome)
2. What are the indications for surgery?
  - Severe stenosis (valve area  $<1.0 \text{ cm}^2$  on echo, peak arteriovenous [AV] velocity  $>4 \text{ m/second}$  [corresponding to a mean AV gradient of 40 mmHg])
  - Symptoms (angina/collapse/dyspnoea/heart failure) with moderate stenosis (valve area  $<1.5 \text{ cm}^2$  on echo, peak velocity  $>3 \text{ m/second}$ )
  - Critical AS = valve area  $<0.8 \text{ cm}^2$
3. What differentiates severe aortic stenosis from aortic sclerosis?
  - Difficult! They lie on a continuum.
  - In sclerosis, there is a normal pulse pressure and character, normal A2 component and little murmur radiation.
4. What happens to the loudness of the murmur with progressive stenosis severity?
  - Murmur intensity is dependent on the flow turbulence through the valve and the cardiac output. Thus, in critical AS with a failing ventricle, cardiac output will fall and the murmur will be soft, but A2 will be absent.
5. Should exercise testing be performed?
  - Exercise testing should not be performed in symptomatic patients with AS when the aortic velocity is  $\geq 4 \text{ m/second}$  or mean pressure gradient is  $\geq 40 \text{ mmHg}$  but is helpful for prognosis in 'asymptomatic' patients to unmask functional limitation.

## KEY POINTS

---

- Aortic stenosis is an ejection systolic murmur which is loudest over the aortic area and radiates to the carotids.
- Comment on the peripheral signs associated with this murmur.
- Ensure that you know the indications for surgical treatment, as well as the medical management.



## REFERENCE

---

Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). (2012) Guidelines on the management of valvular heart disease (version 2012). *Eur Heart J* 33: 2451–96.

## AORTIC REGURGITATION

This patient complains of progressive breathlessness. Examine the patient's cardiovascular system.

## FINDINGS

---

- *General*: Dyspnoea
- *Peripheral*: Collapsing (water hammer) pulse, wide pulse pressure
- *Chest*: Displaced and hyperdynamic apex beat, high-pitched early diastolic murmur (heard best in fixed expiration at the left sternal edge or sometimes the right sternal edge)
- *Eponymous signs*:
  - Corrigan's sign: 'Dancing carotid'
  - De Musset's sign: Head nodding
  - Quincke's sign: Pulsation at nail-bed when pressed
  - Traube's sign: Pistol-shot sound heard over the femoral arteries
  - Austin Flint murmur: Mitral stenosis murmur due to impingement of the anterior mitral valve leaflet by the regurgitant jet

## PRESENTATION

---

This patient has aortic regurgitation as evidenced by the early diastolic murmur which is heard loudest at the left sternal edge with the patient sitting forward. This patient also has a collapsing pulse and evidence of Corrigan's sign.

- Comment on the severity of the aortic incompetence.
- In severe cases, there may be a widened pulse pressure, soft S2, short end-diastolic murmur (EDM), S3 sound and evidence of left ventricular failure (LVF).
- Comment on the possible underlying cause of AR.

## AETIOLOGY OF AR

---

- Acute
  - Infective endocarditis
  - Aortic dissection
- Chronic
  - Congenital aortic valve malformation (e.g. bicuspid, quadricuspid)
  - Aortic root dilatation
  - Prior endocarditis
  - Rheumatic fever
  - Iatrogenic: Following balloon dilatation or TAVI

- Connective tissue disease
  - Arthritis, systemic lupus erythematosus (SLE), ankylosing spondylitis, Ehlers–Danlos syndrome, osteogenesis imperfecta
  - Marfan’s syndrome: High-arched palate, tall, arachnodactyly autosomal dominant condition due to mutations of protein fibrillin-1 located on chromosome 15
- Seronegative arthritides
  - Ankylosing spondylitis, reactive arthritis
- Syphilitic aortitis
  - A rare cause, may be associated with Argyll Robertson pupils

## INVESTIGATIONS

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- CXR
  - Cardiomegaly (‘coeur en sabot’), evidence of LVF
- ECG
  - Lateral T-wave inversion, left ventricular hypertrophy
- Echo  $\pm$  TOE
  - To determine if aortic regurgitation is due to valve or root disease and to assess left ventricular size and systolic function
  - Doppler colour is used to view the regurgitant jet and pulsed-wave Doppler to assess diastolic flow reversal in the descending aorta
- Computerised tomography (CT) or magnetic resonance imaging (MRI)
  - Of aorta if aortic root disease
- Cardiac MRI
  - Recommended in patients with Marfan’s or if an enlarged aorta is found on echo (especially in those with bicuspid valves)

## MANAGEMENT

---

- In acute cases (infective endocarditis and aortic dissection), urgent surgery may be needed.
- In chronic cases, regular monitoring of the patient by a cardiologist to determine when a valve replacement is appropriate, depending on the patient’s symptoms and clinical findings.
- Medical
  - Regular follow-up and echocardiograms (timing for intervention).
  - Diuretic therapy for heart failure.
  - Vasodilating agents such as ACE inhibitors, ARBs or dihydropyridine calcium channel blockers (e.g. amlodipine, felodipine, nifedipine), especially if hypertension is present.
  - In Marfan’s syndrome: Beta-blockers may slow aortic root dilatation and reduce risk of aortic complications (pre- and postoperative). Consider offering screening to first-degree relatives of affected individuals.
- Surgical
  - AVR  $\pm$  CABG
  - TAVI considered if open AVR too high risk (transfemoral or apical)



## QUESTIONS

---

1. List the findings which would determine the need for surgery in aortic regurgitation.
  - Acute severe symptomatic AR; requires emergency surgery.
  - Symptomatic patients with severe AR (dyspnoea, New York Heart Association [NYHA] II–IV, angina) regardless of left ventricular systolic function.
  - Surgery is also indicated for patients with severe AR undergoing surgery for another indication (e.g. CABG, mitral valve surgery or aortic surgery).
  - Asymptomatic patients with severe AR.
2. List some other manifestations of Marfan's syndrome.
  - Ectopia lentis (upwards lens dislocation), arm span > height, dural ectasia, pectus excavatum, joint laxity, scoliosis, pes planus (Ghent criteria)
3. How often should asymptomatic patients with Marfan's be screened?
  - Patients with Marfan's, especially those with a dilated aorta, should be screened annually with an echocardiogram to monitor the proximal aorta/aortic root.
4. What is an Austin Flint murmur?
  - This is a low-pitched, rumbling mid-diastolic murmur which is a sign of severe aortic regurgitation. It is attributed to the fluttering of the anterior mitral valve leaflet caused by a severe regurgitant stream.

## KEY POINTS

---

- Aortic regurgitation is an early diastolic murmur which is heard loudest over the left sternal edge with the patient sitting forward.
- Ensure that you can recognise the eponymous signs associated with this murmur.
- Know the common acute and chronic causes of this murmur.

## REFERENCE

---

Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). (2012) Guidelines on the management of valvular heart disease (version 2012). *Eur Heart J* 33: 2451–96.

## EISENMENGER'S SYNDROME

Please examine this patient who presents with dyspnoea and orthopnoea.

## FINDINGS

---

- *General:* Cyanosis
- *Peripheral:* Clubbing, raised JVP
- *Chest:*
  - Left parasternal heave

- Loud single P2 audible (and palpable)
- Possibly features of a ventricular septal defect (VSD), an ejection systolic click in the pulmonary area, an audible early diastolic murmur in the pulmonary area due to pulmonary regurgitation (PR) and/or a pansystolic murmur at the left sternal edge (LSE) due to tricuspid regurgitation (secondary to pulmonary hypertension)
- Possible wide fixed-split first heart sound (atrial septal defect [ASD])

## PRESENTATION

---

This patient has features suggestive of Eisenmenger's syndrome. They are peripherally cyanosed with clubbing. There is a left parasternal heave. On auscultation, there is a loud pulmonary second heart sound with an audible ejection click in the pulmonary area.

## INVESTIGATIONS

---

- Blood tests
  - Full blood count (FBC) (for polycythaemia), renal (for CKD) and liver function (hepatic congestion/cholestasis), iron stores (iron deficiency anaemia), uric acid (for gout)
- ECG
  - Signs of right ventricular hypertrophy with P-pulmonale
- CXR
  - Increased pulmonary vascular markings, prominent right ventricle, large rheumatoid arthritis (RA)
- Echocardiogram
  - To assess the cause and degree of shunt (VSD/patent ductus arteriosus [PDA]) and the presence of valvular disease (TR/PR), and to calculate pulmonary artery pressures and the extent of pulmonary hypertension
- Pulmonary function tests with volume and CO<sub>2</sub> diffusion studies
- Cardiac catheterisation
- To assess the degree of shunting, and to calculate pressure gradients and pulmonary pressures

## MANAGEMENT

---

- Ideally, treat defects early to prevent this syndrome from developing and avoid situations that make symptoms worse, such as high altitude.
- Avoid smoking
- Medical
  - Oxygen: Symptomatic benefit (no prognostic benefit)
  - Vasodilators: Prostacyclin, bosentan, sildenafil
  - Diuretics to treat right heart failure
  - Iron supplementation to avoid iron deficiency anaemia
  - Contraceptive measures strongly advised and pregnancy should be avoided, as it carries high maternal and foetal mortality rates
  - Venesection, but only if symptoms/signs of hyperviscosity syndrome





- Surgical
  - Not beneficial once Eisenmenger's is present, as the damage to the lung vasculature is irreversible.
  - Palliative procedures are available.
  - Combined heart–lung transplantation is an option.

## QUESTIONS

---

1. What is Eisenmenger's syndrome?
  - Eisenmenger's results from a large left-to-right shunt/cardiac defect causing increased pulmonary blood flow, pulmonary vessel injury, increased pulmonary vascular resistance and resultant pulmonary hypertension.
  - This leads to reversal of the shunt, causing either a unidirectional right-to-left shunt or a bidirectional shunt.
  - This is clinically manifest as cyanotic heart disease.
2. What are the causes of Eisenmenger's syndrome?
  - Large nonrestrictive VSD
  - Nonrestrictive PDA
  - Atrioventricular septal defects
  - Large uncorrected or surgically created systemic-to-pulmonary shunts for treatment of congenital heart disease
3. What are the potential complications of Eisenmenger's syndrome?
  - Secondary polycythaemia and hyperviscosity
  - Arrhythmias
  - Heart failure
  - Stroke
  - Haemoptysis
  - CKD
  - Hyperuricaemia and gout
  - Hypertrophic osteoarthropathy
  - Sudden cardiac death (SCD)

## KEY POINTS

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- Ensure that you comment on the presence of cyanosis and clubbing.
- Be aware of the underlying causes of this condition.
- Be aware that once Eisenmenger's develops, treatment of the underlying defect is purely medical.

## REFERENCE

---

Baumgartner H, et al. (2010) ESC guidelines for the management of grown-up congenital heart disease (new version 2010). Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPIC); ESC Committee for Practice Guidelines (CPG). *Eur Heart J* 31(23): 2915–57.

## HYPERTROPHIC CARDIOMYOPATHY

This patient, who has had episodes of syncope and palpitations, has been told he has a murmur. Please examine his cardiovascular system.

### FINDINGS

- *General:* Dyspnoea
- *Peripheral:* Bifid jerky pulse, large A-wave visible in JVP, low systolic blood pressure, narrow pulse pressure
- *Chest:* Palpable thrill over the aortic area, heaving double apical impulse, ejection systolic murmur loudest over the left sternal edge (accentuated by Valsalva manoeuvre, softer with squatting), S4, normal A2
- *Extra features:* Look for evidence of congestive cardiac failure, murmur of MR, ICD in left subclavicular fossa

### PRESENTATION

This patient has HCM as evidenced by a jerky pulse, double-impulse apex beat and an ejection systolic murmur that radiates to the carotids but a normal A2 component. There is no evidence of congestive cardiac failure.

- Mention the murmur and comment on severity. Comment on lack/presence of signs of congestive cardiac failure.

### DIFFERENTIAL DIAGNOSIS

- Aortic stenosis
- Flow murmur
- Pulmonary stenosis
- Hypertensive heart disease

### INVESTIGATIONS

- ECG
  - Voltage LVH, deep Q-waves, ST elevation, interventricular conduction delays
- Chest X-ray
- Echo
  - Investigation of choice, looking for an LV outflow tract obstruction (LVOTO) defined as a peak Doppler LV outflow tract gradient of  $\geq 30$  mmHg at rest, but if  $\geq 50$  mmHg, invasive treatment should be considered. Other echo features: Septal asymmetrical hypertrophy, systolic anterior motion (SAM) of the mitral valve, MR, diastolic LV dysfunction.
- Cardiac MRI
  - Demonstrates severity and distribution of hypertrophy and wall fibrosis and LV function



- Holter monitor
  - Nonsustained ventricular tachycardia (VT)
- Exercise tolerance test
  - Blood pressure response, looking for inadequate rise or paradoxical systolic drop with exertion
- Cardiac catheterisation
  - Aortic outflow gradient, coronary artery disease
- Genetic testing
  - Recommended for patients fulfilling diagnostic criteria for HCM with testing of relatives if a causative mutation is found

## MANAGEMENT

---

- Medical
  - Regular follow-up and echocardiograms
  - Beta-blocker/(verapamil) ± disopyramide (Nifedipine and other dihydropyridine calcium channel blockers are potentially harmful for patients with LVOTO)
  - Diuretic therapy only for heart failure (avoid digoxin in patients with LVOTO)
  - Septal alcohol ablation
  - Dual-chamber pacemaker insertion
  - ICD insertion
- Surgical
  - Septal myomectomy (Morrow procedure) ± mitral valve surgery
  - Heart transplant considered for moderate–severe refractory heart failure without LVOTO

## QUESTIONS

---

1. What is the cause of the dynamic outflow gradient?
  - It is caused by systolic anterior motion of the mitral valve due to local underpressure (Venturi effect), exacerbated by the septal hypertrophy.
2. What are the risk factors for SCD?
  - Family history of SCD with HCM at a young age
  - Unexplained syncope in young age (<30 years)
  - Personal history of ventricular fibrillation (VF)
  - VT or nonsustained VT
  - Maximum LV wall thickness  $\geq 30$  mm
  - Hypotensive response to exercise (a fall of at least 20 mmHg from peak pressure)
  - LVOTO
3. What do you know about the genetics of HCM?
  - Sixty percent of HCM is inherited as autosomal dominant, and most mutations occur in the cardiac sarcomere protein genes or arise spontaneously, producing a *de novo* mutation (usually of the  $\beta$ -myosin heavy-chain gene), and 5%–10% are due to other genetic disorders.

- The majority of cases are due to mutations in the  $\beta$ -myosin heavy-chain gene (MYH7, chromosome 14) and cardiac myosin-binding protein C gene (MYBPC3, chromosome 11).
  - Less commonly affected genes include cardiac troponin I (TNNT3) and T (TNNT2) and  $\alpha$ 1-tropomyosin (TPM1) and myosin light-chain 3 (MYL3).
  - Therefore, a detailed family history and first-degree relative screening should be advised.
4. When would an ICD be indicated?
- Should be considered if the 5-year risk of SCD is 4%–6% and definitely indicated if the 5-year risk is  $\geq$ 6% using the European Society of Cardiology (ESC) HCM risk score calculator

## KEY POINTS

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- HCM is associated with an ejection systolic murmur heard loudest at the left sternal edge.
- Key clinical findings are a bifid jerky pulse, a double apical impulse and a palpable thrill.
- This is an autosomal dominant inherited condition which is associated with sudden cardiac death.

## REFERENCE

---

Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). (2014) ESC guidelines on diagnosis and management of hypertrophic cardiomyopathy. *Eur Heart J* 35(39): 2733–79.

## MITRAL STENOSIS

This patient complains of breathlessness. Please examine their cardiovascular system.

## FINDINGS

---

- *General*: Malar flush
- *Peripheral*: Irregularly irregular pulse, raised JVP
- Chest
- Inspection/palpation: Left thoracotomy/valvotomy scar, tapping apex, palpable S1
- *Auscultation*: Opening snap of mitral valve, loud S1, low-pitched rumbling mid-diastolic murmur at the apex (heard loudest in the left lateral position)

## PRESENTATION

---

This lady has a malar flush. There is an irregularly irregular pulse but no stigmata of endocarditis. The apex beat is not displaced. There is a loud first heart sound, and there is a mid-diastolic murmur heard loudest in the left lateral position. This is mitral stenosis.

- Look for signs of pulmonary hypertension (left parasternal heave, loud/palpable P2).



## DIFFERENTIAL DIAGNOSIS

---

- Left atrial myxoma
- Parachute mitral valve
- Severe mitral annular calcification
- Thrombosed mitral valve prosthesis
- Radiotherapy

## INVESTIGATIONS

---

- Diagnosis
  - Chest X-ray: Left atrial enlargement
  - Echocardiogram  $\pm$  TOE: To assess the mitral and other valves, left atrial dimension and pulmonary artery pressures
- Complications
  - ECG
    - AF, right ventricular hypertrophy
  - Echo
    - Assess for pulmonary hypertension and evidence of endocarditis
  - TOE
    - Exclude LA thrombus
  - Right and left cardiac catheterisation
    - To measure cardiac pressures if discordance between symptoms and echo findings, e.g. wedge pressure

## MANAGEMENT

---

- Medical
  - Diuretics, long-acting nitrates (can help dyspnoea)
  - Treat AF with rate control
  - Anticoagulate all patients with warfarin where possible (target INR 2–3), especially if MS with AF, a prior embolic event or LA thrombus
- Surgical
  - Indications include clinically significant MS with a valve area  $\leq 1.0$  cm<sup>2</sup>, significant symptoms which limit normal activity, recurrent emboli, pulmonary oedema (especially in pregnancy) and deterioration due to AF.
  - Options include mitral valve replacement, open valvotomy and balloon valvuloplasty (percutaneous MV commissurotomy).
  - Percutaneous mitral commissurotomy recommended if severe MS without LA thrombus and when surgery is contraindicated or as a bridge to surgery.
  - Complications include haemopericardium, embolic phenomena, severe MR and death.

## QUESTIONS

---

1. What are the causes of mitral stenosis?
  - Rheumatic fever is the most common cause.
  - Others include

- Calcific disease (heavily calcified mitral annulus with extension to MV leaflets)
  - End-stage renal disease (ESRD)
  - Congenital lesions
  - Carcinoid tumours
  - Mucopolysaccharidoses
2. What are the complications of mitral stenosis?
- Pulmonary hypertension (leading to right-sided heart failure)
  - Haemoptysis
  - Flash pulmonary oedema
  - Emboli (stroke risk; novel oral anticoagulants [NOACs] are not licensed in MS)
  - Atrial fibrillation
  - Infective endocarditis
  - Pressure effects from an enlarged left atrium: Hoarseness (Ortner's syndrome, due to left recurrent laryngeal nerve compression), bronchial obstruction, dysphagia
3. What surgical procedures require antibiotic prophylaxis?
- Prophylaxis is generally not recommended now for surgical procedures.
  - Patients should be advised to maintain good oral health, and those at risk of infective endocarditis should be investigated and treated promptly when displaying evidence of infection.

## KEY POINTS

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- Mitral stenosis is a mid-diastolic murmur which is heard loudest at the apex in the left lateral position.
- Comment on the presence or absence of atrial fibrillation.
- Always check for the presence of pulmonary hypertension.

## REFERENCE

---

Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). (2012) Guidelines on the management of valvular heart disease (version 2012). *Eur Heart J* 33: 2451–96.

## MITRAL REGURGITATION

Please examine this patient who has shortness of breath and/or palpitations.

## FINDINGS

---

- *General*: Tachypnoea, flushed face, peripheral oedema, mitral valvotomy scar
- *Peripheral*: Raised JVP, irregularly irregular pulse
- *Chest*: Inspection/palpation: Scars (valvotomy, left thoracotomy), thrusting and laterally displaced apex, palpable thrill, parasternal heave
- *Auscultation*: Pansystolic murmur loudest at apex radiating to the axilla, soft S1, S3 ( $\pm$  gallop) in severe MR



## PRESENTATION

---

This patient has mitral regurgitation. The pulse is irregularly irregular, consistent with atrial fibrillation. The apex beat is thrusting in nature. On auscultation, there is an audible third heart sound and a pansystolic murmur, loudest at the apex, that radiates to the axilla.

- Mitral regurgitation is often associated with ischaemic heart disease and a dilated left ventricle, so it is important to look for scars from previous CABG surgery, as well as any risk factors (tar staining on the fingers, signs of diabetes, hypertension, corneal arcus and xanthelasma).

## INVESTIGATIONS

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- ECG
  - LVH, left strain pattern, evidence of ischaemia, AF
- Echocardiogram  $\pm$  TOE
  - Assess LV systolic function and dimensions, RV function, LA size, pulmonary artery pressure.
  - Additional features: Vegetations, papillary muscle/chordae tendineae rupture, other significant valvular lesions.
- Doppler flow studies
  - Size and site of regurgitant jet
- CXR
  - Normal, cardiomegaly, large left atrium, pulmonary oedema
- Cardiac catheterisation
  - If discrepancy between symptoms and noninvasive investigations

## MANAGEMENT

---

- Medical
  - Management of congestive cardiac failure
    - ACE inhibitors/ARBs, diuretics ( $\pm$  aldosterone antagonists), beta-blockers
  - Rate control therapy and anticoagulation for atrial fibrillation
  - Cardiac resynchronisation therapy with biventricular pacing
- Surgical
  - When symptomatic and LVEF  $>30\%$ , especially if evidence of cardiomegaly and raised end-systolic LV volume and resultant LV systolic dysfunction and dilatation
  - Options
    - Repair/reconstruction (valvuloplasty) is preferable (also if MR limited to posterior leaflets): Carries lower operative mortality and no need for long-term anticoagulation
    - Valve replacement: Elective or for emergency treatment of acute severe incompetence (IE/acute chordae tendineae/papillary muscle rupture) or if severe MR and undergoing cardiac surgery for another indication (e.g. CABG, AVR)
    - Percutaneous options: Valvuloplasty ring, MitraClip<sup>®</sup>

## QUESTIONS

---

1. What are the causes of mitral regurgitation?
  - Acute
    - Chorda tendineae rupture: Due to degenerative valve disease, trauma, infective endocarditis, rheumatic MV disease, mitral valve prolapse (MVP)
    - Papillary muscle rupture postmyocardial infarction (MI) (usually associated with an inferior MI)
    - Papillary muscle dysfunction due to ischaemia
    - Infective endocarditis
  - Chronic
    - Mitral valve prolapse (most common cause in developed countries) rheumatic heart disease
    - Functional MR secondary to left ventricular dilatation and systolic dysfunction resulting in
    - Lateral displacement of the papillary muscles and retraction of the valve leaflets.
    - Dilatation or calcification of the mitral valve annulus may also result in MR.
    - Infective endocarditis
    - Connective tissue disorders: SLE, rheumatoid arthritis, ankylosing spondylitis
    - Inherited disorders: Marfan's syndrome, Ehlers–Danlos syndrome, osteogenesis imperfecta, pseudoxanthoma elasticum
    - Hypertrophic cardiomyopathy
    - Radiation heart disease
  
2. What is the difference between primary and secondary MR?
  - Primary MR
    - Can be acute or chronic and includes all causes of MR in which intrinsic lesions affect one or more components of the mitral valve
    - Chronic MR can be either chronic primary or chronic secondary MR.
    - Chronic primary MR is usually degenerative, with the valve itself being diseased.
    - Chronic secondary is usually functional, so the valve is usually structurally normal.
  
3. What are the causes of primary and secondary MR, and how does the management differ?
  - Primary MR
    - Infective endocarditis causing chordae tendineae rupture or spontaneous chordae or papillary muscle rupture
    - Requires urgent surgical intervention
  - Chronic primary
    - Rheumatic heart disease mitral valve prolapse, infective endocarditis, connective tissue disease, radiation.
    - Surgery is usually curative.
  - Chronic secondary





- LV dilatation, ischaemic (e.g. MI) or nonischaemic heart disease (idiopathic dilated cardiomyopathy).
- MV surgery is not usually curative, as it's only one component of the disease, as there's often associated LV dysfunction and/or ischaemic heart disease (IHD) that also requires treatment.

## KEY POINTS

---

- Mitral regurgitation is a pansystolic murmur which is heard loudest at the apex and radiates to the axilla.
- Check for the presence/absence of atrial fibrillation.
- Be aware of the acute and chronic causes of mitral regurgitation, and be aware these can be further subdivided into primary or secondary.

## REFERENCE

---

Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). (2012) Guidelines on the management of valvular heart disease (version 2012). *Eur Heart J* 33: 2451–96.

## MITRAL VALVE PROLAPSE

Please examine this patient who has presented with a murmur.

## FINDINGS

---

- *Auscultation:*
  - Normal heart sounds
  - Midsystolic click (may or may not be present)
  - Late systolic MR murmur loudest at the left sternal edge
- The murmur is made louder by factors that decrease the volume of blood within the cardiac chambers, e.g. straining (Valsalva manoeuvre) or standing from squatting.
- There is a higher female preponderance.
- Patients are usually asymptomatic.
- The patient may have peripheral stigmata of a connective tissue disorder.

## INVESTIGATIONS

---

- CXR
  - Normal, cardiomegaly, large left atrium, pulmonary oedema
- ECG
  - May be inferior T-wave inversion, large left atrium, AF
- Echocardiogram
  - Diagnostic! Degree of thickening of the mitral valve leaflets and their displacement relative to the annulus is indicative of MVP.

## MANAGEMENT

---

- Medical
  - Palpitations controlled with beta-blocker therapy
- Surgical
  - Indications as for MR; valve repair preferred to replacement

## QUESTIONS

---

1. What are the associations of mitral valve prolapse?
  - Congenital heart disease
    - ASD, AVSD, PDA
  - Congenital disorders
    - Turner's syndrome, Marfan's syndrome, osteogenesis imperfecta, pseudoxanthoma elasticum
  - Others
    - SLE
2. What are the presenting features of MVP?
  - Usually asymptomatic but can be associated with atypical chest pain, palpitations, fatigue and dyspnoea
3. What are the complications of MVP?
  - Infective endocarditis.
  - Atrial and ventricular arrhythmias (ventricular ectopic beats are the usual cause of palpitations experienced).
  - MR: MVP is the most common cause of chronic primary MR in developed countries.
  - Cerebral emboli resulting in transient ischaemic attacks (TIAs) and/or stroke.
  - Sudden cardiac death.
4. What are the current recommendations regarding antibiotic prophylaxis dental work with MVP?
  - As patients with MVP are considered as having 'moderate risk' of infective endocarditis, antibiotic prophylaxis is not generally recommended.
  - (National Institute for Health and Care Excellence [NICE] guidance on prophylaxis against infective endocarditis in adults and children undergoing interventional procedures is currently being drafted.)

## KEY POINTS

---

- Mitral valve prolapse is a late systolic murmur which is heard loudest at the left sternal edge.
- It is more common in females.
- Be aware of the associations of this murmur.



## REFERENCE

---

Habib G, et al. (2015) 2015 ESC guidelines for the management of infective endocarditis: The Task Force for the Management of Infective Endocarditis of the European Society of Cardiology (ESC). Endorsed by: European Association for Cardio-Thoracic Surgery (EACTS), the European Association of Nuclear Medicine (EANM). *Eur Heart J* 36(44): 3075–128.

## MIXED AORTIC VALVE DISEASE

This patient has a murmur. Please examine their cardiovascular system.

## FINDINGS

---

- Features of both aortic stenosis and aortic regurgitation.
- The clinical findings should determine the predominant abnormality.

## PRESENTATION

---

This patient has mixed aortic valve disease with a predominant stenotic lesion. There is a slow-rising radial pulse and a normal blood pressure, but there are no stigmata of infective endocarditis. The apex beat is in the fifth intercostal space, midclavicular line. The heart sounds are normal, but there is a harsh ejection systolic murmur radiating to the carotids and an end-diastolic murmur heard loudest at the left sternal edge in forced expiration.

- When suspecting mixed aortic valve disease, it is crucial to look for and present the above features. Be sure to ask about the patient's blood pressure. Where possible, decide on the predominant lesion, as the clinical manifestation usually follows that.

## INVESTIGATIONS

---

- Diagnosis
  - Echocardiogram to assess the aortic valve, including valve size and gradient (also to assess the other valves, and left ventricular size and function).
  - TOE may be required to fully delineate the valvular anatomy.
  - Cardiac catheterisation can be used to assess for coronary artery disease. Left ventricular angiography, aortogram and measuring pressuring gradient from left ventricle to aorta may further aid diagnosis.
- Complications
  - ECG, echocardiogram, blood cultures if endocarditis suspected.
  - Exercise testing to assess exercise tolerance may be used to assess haemodynamic significance.

## MANAGEMENT

---

- Medical management is an option with diuretics and possibly an ACE inhibitor (predominant AR).
- Surgery or TAVI (if predominant AS with moderate AR only [not if severe AR]) should be considered.
- The indications for surgery should follow the guidelines for the predominant valvular lesion (i.e. management of AS if stenosis is the predominant lesion or AR if that is)

## QUESTIONS

---

1. What are the main causes of mixed aortic valve disease?
  - Bicuspid aortic valve
  - Degenerative calcific aortic valve disease
  - Radiotherapy
2. What will happen to the gradient and valve area with each of the predominant lesions?
  - Predominant AS: A high gradient and small valve area will be present.
  - Predominant AR: The gradient may be significantly raised due to regurgitation, but the aortic valve area (AVA) is usually relatively large.

## KEY POINTS

---

- When the murmur of aortic stenosis is detected, it is important to also listen for the diastolic murmur of aortic regurgitation (or any other murmur present).
- State which lesion is dominant in conjunction with the clinical findings.
- Management of the valve follows that of the predominant lesion.

## REFERENCE

---

Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). (2012) Guidelines on the management of valvular heart disease (version 2012). *Eur Heart J* 33: 2451–96.

## MIXED MITRAL VALVE DISEASE

This patient has a murmur. Please examine their cardiovascular system.

## FINDINGS

---

- Features of both mitral stenosis and mitral regurgitation.
- The clinical findings should determine the predominant abnormality.



## PRESENTATION

---

This patient has mixed mitral valve disease with a predominant regurgitant lesion. There are no stigmata of infective endocarditis. There is a sharp radial pulse. The apex beat is in the fifth intercostal space, towards the anterior axillary line. The heart sounds are normal with a pansystolic murmur radiating to the axilla and a mid-diastolic rumbling murmur heard loudest at the apex in the left lateral position.

- When suspecting mixed mitral valve disease, it is crucial to look for and present the above features.
- As with mixed aortic valve disease, decide on the predominant lesion, as the clinical presentation will follow that.

## INVESTIGATIONS

---

- Diagnosis
  - Echocardiogram to assess the mitral valve, including valve size and gradient (also to assess the other valves, and left ventricular size and function).
  - TOE may be required to fully delineate the valvular anatomy.
  - Right and left cardiac catheterisation further assesses valvular anatomy and concomitant coronary artery disease and measures wedge pressure.
- Complications
  - ECG (AF), CXR, echocardiogram, blood cultures if endocarditis suspected

## MANAGEMENT

---

- Anticoagulation, rate control therapy for atrial fibrillation and diuretics to treat fluid overload.
- Surgical valve replacement should be considered (according to the predominant lesion as per the guidelines for individual valve lesions earlier in this chapter).
- Mitral commissurotomy could be considered if predominant MS with only mild MR.

## QUESTIONS

---

1. What are the causes of mixed mitral valve disease?
  - Predominantly caused by rheumatic heart disease
  - Can be caused by degenerative valvular disease with increasing age
  - Radiotherapy
2. What will happen to the gradient and valve area with each of the predominant lesions?
  - Predominant MS
    - High transvalvular gradient and small valve area
  - Predominant MR
    - High transvalvular gradient due to the regurgitation, but valve area may remain relatively large

## KEY POINT

---

- Again, it is important to state which lesion is dominant.

## REFERENCE

---

Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). (2012) Guidelines on the management of valvular heart disease (version 2012). *Eur Heart J* 33: 2451–96.

## PROSTHETIC HEART VALVES

This patient attends for routine follow-up. Please examine their cardiovascular system.

## FINDINGS

---

- *General:* Dyspnoea, audible 'click' from end of bed
- *Peripheral:* Scars (midline sternotomy, lateral thoracotomy), including harvesting scars on legs/arms for concomitant CABG (more common in aortic valve replacement)
  - Jaundice, anaemia, purpura
- *Chest:*
  - Aortic valve replacement: Metallic S2 (opening/closing click), ejection systolic flow murmur (common, nonpathological), diastolic regurgitant murmur (pathological)
  - Mitral valve replacement: Metallic S1 (opening/closing click), systolic regurgitant murmur (pathological)

Note: Tissue valve replacements may demonstrate normal heart sounds (be alert to this possibility if sternotomy and no venous graft harvest scars evident).

## PRESENTATION

---

This patient has an aortic metallic valve replacement as evidenced by a midline sternotomy scar and a metallic second heart sound. The valve appears to be functioning well, as I cannot hear a murmur of aortic regurgitation and there is no evidence of congestive cardiac failure. Of note, there are no peripheral stigmata of subacute bacterial endocarditis.

- Mention the type of valve replacement and whether there is regurgitation.
- Mention stigmata of endocarditis, any evidence of leg venous graft harvest scars for CABG and signs of over-anticoagulation.
- To look for any signs of congestive cardiac failure (possibly suggestive of valve incompetence).

## INVESTIGATIONS

---

- Echocardiogram



## MANAGEMENT

---

- Metallic valves require lifelong anticoagulation (warfarin).
- Tissue valves do not usually require anticoagulation unless concomitant AF but have a shorter life span, as they are susceptible to structural valve deterioration (SVD).
- Rates of SVD decrease with age and if in the aortic position (compared with the mitral position).
- Treatment of SVD requires surgery to replace the valve.

## QUESTIONS

---

1. What are complications of prosthetic valves?
  - Early
    - Surgical complications
    - Endocarditis
  - Late
    - Thromboembolic sequelae
    - Bleeding (as a result of anticoagulation)
    - Infective endocarditis
    - Valvular or paravalvular leak (regurgitation)
    - Haemolysis (with anaemia)
    - Valvular stenosis (endothelialisation (pannus))
    - Structural failure/embolisation (very rare)
    - All the above complications can occur with metallic valves. The main risk with tissue valves is regurgitation with degeneration.
2. What influences the type of valve a patient will receive?
  - Factors to consider when deciding valve type include
    - Bleeding risk with anticoagulation and thromboembolic phenomena with a metallic valve
    - Risk of SVD with a biological valve
    - Patient/physician/surgical concerns
  - Metallic valves are used in younger patients due to their durability (they are also used if the patient is already on warfarin), unless female and planning to conceive in the future (due to high risk of thromboembolic phenomena during pregnancy regardless of the anticoagulant used).
  - Biological valves are useful in the elderly (where the patient's life expectancy is thought to be less than the durability of the valve), and those with an increased risk of bleeding if anticoagulated.
3. What types of metallic valve are available and what's their thrombogenicity risk?
  - Disc valve (e.g. Björk–Shiley's): High risk
  - Bileaflet valve (e.g. St Jude Medical): Low risk
4. What happens to the intensity of the clicks from a metallic valve when failing?
  - Decreasing intensity of the closing click

5. Is any oral anticoagulation ever required after bioprosthetic valve surgery?
  - Definitely if there's another indication for anticoagulation, e.g. AF.
  - Should be considered for 3 months following mitral or tricuspid bioprosthetic valve surgery.
  - Either low-dose aspirin or oral anticoagulation should be considered after aortic bioprosthesis for 3 months.

## KEY POINTS

---

- Listen carefully from the end of the bed for any audible 'clicks'.
- Examine for any surgical scars.
- Comment on the type of valve (tissue/metallic), any evidence of infective endocarditis and any evidence of congestive cardiac failure.
- Be aware of early and late complications of prosthetic valves.

## TRICUSPID REGURGITATION

This patient has presented with shortness of breath. Please examine her cardiovascular system.

## FINDINGS

---

- *General:* Dyspnoea, jaundice
- *Peripheral:*
  - Marked peripheral/sacral oedema, prominent JVP (giant V-waves)
  - Evidence of intravenous drug user (IVDU)
- *Chest:* Right ventricular heave, palpable P2, pansystolic murmur heard loudest at the left sternal edge in inspiration, S3, bibasal crepitations and/or pleural effusions, evidence of chronic pulmonary pathology/kyphoscoliosis
- *Abdomen:* Ascites, pulsatile hepatomegaly

## PRESENTATION

---

This patient has a diagnosis of tricuspid regurgitation as evidenced by the pansystolic murmur which is best heard at the LSE in inspiration. She also has a prominent JVP with giant V-waves. There is also evidence of pulsatile hepatomegaly which supports this diagnosis.

- The main differential diagnosis for this murmur is mitral regurgitation, which would be louder in expiration and radiates to the axilla.

## AETIOLOGY

---

Underlying causes can be divided into

- Primary
  - Congenital (Ebstein's)
  - Rheumatic heart disease





- Right-sided endocarditis (e.g. due to IVDU)
- Carcinoid syndrome
- Penetrating trauma, right-sided pacing leads
- Radiation
- Secondary
  - Right-sided heart failure (e.g. pulmonary emboli, MI)
  - Pulmonary hypertension (idiopathic or due to left-sided heart disease or cor pulmonale)
  - Eisenmenger's syndrome
  - Biventricular failure

## INVESTIGATIONS

---

- CXR
  - Pulmonary congestion or pruned vessels, hyperinflated lungs, pulmonary fibrosis, cardiomegaly (large RA)
- Echo
  - Demonstrates valvular abnormality, severity of valve disease, right ventricular dimension and function and inferior vena cava size; measures pulmonary artery pressures and any concomitant left heart disease
- Exercise testing may be useful in those with severe TR but minimal symptoms.

## MANAGEMENT

---

Identify and treat the underlying cause.

- Medical
  - Loop diuretics (for right heart failure)
  - ACE inhibitors (for systemic hypertension and heart failure)
  - Spironolactone
  - Pulmonary vasodilators may be indicated in severe TR with pulmonary hypertension
- Surgical
  - Tricuspid valve repair is usually sufficient for mild–moderate TR in those undergoing simultaneous left heart surgery.
  - In all other cases, TV repair or replacement may be performed (although replacement less frequently is performed).

## QUESTIONS

---

1. What is carcinoid syndrome?
  - Carcinoid tumours are rare neuroendocrine tumours of the enterochromaffin cells. They are mostly asymptomatic.
  - Carcinoid syndrome is evident when products of the tumour are metabolised by the liver, resulting in a collection of symptoms, including dyspnoea and wheeze (due to bronchoconstriction), flushing, diarrhoea, tachycardia, dizziness and CCF.
  - Symptoms are caused by release of serotonin, tachykinins and other vasoactive peptides into the circulatory system.

2. How is a diagnosis of carcinoid syndrome established?
  - Diagnosis is made by measurement of 24-hour urinary 5-hydroxyindoleacetic acid, which is a degradation component of serotonin.
  - Radiological methods may also be used to help identify the primary.
3. How is carcinoid syndrome treated?
  - Medical treatment
    - Loperamide: Symptomatic relief of diarrhoea
    - Octreotide or lanreotide: Somatostatin analogue; blocks the release of tumour mediators
    - Radiotherapy: Targeted to the tumour
    - Hepatic artery embolisation or radiofrequency ablation: For hepatic tumours
    - Chemotherapy
  - Surgical treatment
    - Curative resection
    - Debulking of tumour
4. What are the common organisms in infective endocarditis?
  - *Streptococcus viridans*.
  - *Staphylococcus aureus* (common in IVDUs).
  - *Staphylococcus epidermidis*.
  - *Enterococcus*.
  - Staphylococcal endocarditis has the worst prognosis, streptococcal the best.
  - Other causative organisms include
    - *Streptococcus bovis*
    - HACEK group (*Haemophilus species*, *Actinobacillus*, *Cardiobacterium*, *Eikenella*, and *Kingella*)
    - *Pseudomonas*
    - *Coxiella burnetii*
    - Fungal infections
5. What are the diagnostic criteria for infective endocarditis?
  - Modified Duke criteria, which include two major and five minor criteria

## KEY POINTS

---

- Tricuspid regurgitation is a pansystolic murmur which is heard best at the left sternal edge in inspiration.
- Ensure that you examine for peripheral signs, such as pulsatile hepatomegaly and a prominent JVP.
- Always assess for signs of right heart failure.
- Be aware of the Duke's diagnostic criteria for infective endocarditis.

## REFERENCE

---

Joint Task Force on the Management of Valvular Heart Disease of the European Society of Cardiology (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS). (2012) Guidelines on the management of valvular heart disease (version 2012). *Eur Heart J* 33: 2451–96.



## VENTRICULAR SEPTAL DEFECT

Please examine the cardiovascular system of this patient who has presented with dyspnoea.

### FINDINGS

- *General:* Well looking, often young
- *Peripheral:* Possible clubbing (can occur in non-cyanotic congenital heart disease)
- *Chest:*
  - Inspection/palpation: Laterally displaced apex, palpable thrill at LSE, left parasternal heave
  - Auscultation: Pansystolic murmur loudest at the left parasternal area (also heard at the apex), loud P2 if pulmonary hypertension

### PRESENTATION

This patient has a ventricular septal defect. The pulse is regular, and there is no evidence of infective endocarditis. The apex beat is undisplaced. There is a left parasternal heave. On auscultation, there is a pansystolic murmur loudest at the left sternal edge, but also audible at the apex.

- Comment on presence/absence of a loud P2.
- If the patient is young, the cause is likely to be congenital.
- In elderly patients, look for factors contributing to ischaemic heart disease.
- Always look for evidence of complications or previous surgical repair.

### INVESTIGATIONS

- ECG
  - Likely to be normal if small asymptomatic VSD.
  - Other features include left-axis deviation and left ventricular hypertrophy in a moderate defect *or* mainly right ventricular hypertrophy in a large defect.
- Echocardiography
  - Assess size, position of VSD, degree of shunt, LV size and function, pulmonary pressures, aortic valve function and competence.
- CXR
  - Normal in small VSDs
  - Pulmonary plethora (degree depends on size of VSD)
  - Cardiomegaly
- Cardiac catheterisation
  - Shunt assessment through oxygen saturation measurement in the right ventricle

### MANAGEMENT

- Conservative
  - Most close spontaneously (especially if muscular defects)
- Medical
  - ACE inhibitors (reduce adverse remodelling)

- Surgical
  - Surgical closure
    - Symptomatic VSD
    - Infective endocarditis
    - Post-MI (acute septal rupture)
    - Haemodynamic compromise/volume overload
    - Large shunt
    - Development of AR in perimembranous VSDs
  - Percutaneous closure
  - Perimembranous VSDs can be closed using a transcatheter approach

## QUESTIONS

---

1. What are the causes of a VSD?
  - Congenital
    - 'Lone'
    - Associated with other defects, e.g. tetralogy of Fallot, AVSD
  - Acquired
    - Post-MI (acute septal rupture)
    - Following trauma
2. What is the classification of VSDs?
  - There are four main types of defects:
    - Membranous septal (which includes perimembranous VSD, which is the most common type of VSD)
    - Muscular septal
    - Atrioventricular canal
    - Subarterial infundibular (or conal)
3. What are the complications of a VSD?
  - Infective endocarditis
  - Pulmonary hypertension
  - Eisenmenger's syndrome
  - Aortic valve leaflet prolapse and eventual aortic regurgitation (with perimembranous VSDs)
4. What are the associations of a VSD?
  - Aortic regurgitation
  - Patent ductus arteriosus
  - Coarctation of the aorta
  - Tetralogy of Fallot (VSD, overriding aorta, pulmonary stenosis, right ventricular hypertrophy)
  - Turner's syndrome
  - Trisomies
    - 21: Down's syndrome associated with endocardial cushion defects
    - 18: Edwards' syndrome
    - 13: Patau's syndrome



5. What are the potential risks associated with transcatheter closure of a VSD?
- Displacement
  - Misplacement
  - Cardiac tamponade
  - Aortic regurgitation
  - Haemorrhage
  - Complete heart block requiring pacemaker insertion
  - Need for open cardiac surgery

## KEY POINTS

---

- The murmur associated with a VSD is a pansystolic murmur, heard loudest at the left sternal edge.
- Always check for a parasternal heave.
- Patients will often be young and asymptomatic.
- Perimembranous VSDs can be closed using a transcatheter approach, but be aware of the risks associated with this.

## REFERENCE

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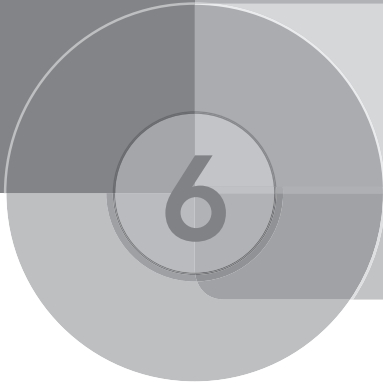
National Institute for Health and Care Excellence. (2010) Guidance IPG336. Transcatheter endovascular closure of perimembranous ventricular septal defect. Available from <https://www.nice.org.uk/guidance/ipg336>. Accessed 26 March 2016.

## CARDIOLOGY STATION SUMMARY

- The cardiac examination takes time. Don't spend too much time covering the peripheral signs, although at the same time, be sure you don't miss any (a skill that can only be learnt with practice).
- Ensure that when you listen to heart sounds and murmurs, you are seen to time them with the carotids.
- If possible, when you present your case, try to start with the diagnosis and explain why you think this is the diagnosis, and which investigations and management you would institute. This makes you sound confident, and if you really know your cardiology, you can demonstrate to the examiners that you are an excellent candidate without them having to ask any difficult questions.
- In valvular heart disease, pay attention to signs other than the murmur (e.g. character of the peripheral pulse and apex beat and association of differences in pulse pressure). It is possible you may know the diagnosis before you listen to the chest. A summary of the characteristics of some of the different murmurs is presented below.

Lesion	Site	Timing	Character	Bell or diaphragm	Position	Respiratory phase
Aortic stenosis	Aortic area	ESM	Rough/harsh	Diaphragm	Forward	Expiratory
Aortic regurgitation	LSE > apex	EDM	High pitch/decrescendo	Diaphragm	Forward	Expiratory
Mitral stenosis	Apex > axilla	Mid-diastolic murmur	Low pitch, rumbling	Bell	Left lateral	Expiratory
Mitral regurgitation	Apex > axilla	Pansystolic murmur	Blowing	Diaphragm	Left lateral	Expiratory
Tricuspid regurgitation	LSE	Pansystolic murmur	Blowing	Diaphragm	Forward	Inspiratory
Ventricular septal defect	LSE	ESM	Harsh/high pitch	Diaphragm	Forward	Expiratory

- Auscultation findings of common valvular abnormalities
  - Be well rehearsed with the manoeuvres used to auscultate specific murmurs; this is an area where candidates often look clumsy.
  - At the end of your examination, remember to inform the examiners of the other pertinent examinations that you would like to perform, e.g. fundoscopy for Roth's spots in a case of subacute bacterial endocarditis, urinalysis (looking for evidence of haematoproteinuria).
  - If you see a midline sternotomy scar, ensure that you look at the legs for evidence of venous harvesting scars. To not do this would be foolish; you can do it from the end of the bed before you have even touched the patient. You can tell that they have ischaemic heart disease and have had a coronary artery bypass graft.



# Station 4: Ethics and Communication Skills

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## HINTS FOR THE ETHICS AND COMMUNICATION SKILLS STATION

I've learnt that people will forget what you said, people will forget what you did, but people will never forget how you made them feel.

**Maya Angelou**

- Always remember the above quote in all your interactions in practising medicine and especially when dealing with difficult and sensitive situations.
- In this station, your communication skills are being thoroughly evaluated.
- It is not just your ability to speak that is being assessed, but also your ability to listen to the concerns that your patient presents, and how you empathize with them.
- Read the scenario you are given carefully. Look at what is being asked. The range of tasks in this station is wide, from breaking bad news to explaining a new diagnosis and addressing the related issues.
- Make good use of the time before entering the station to think through how you are going to tackle the scenario. Make a list of the issues to be addressed so that you do not forget them under the time pressure.
- Start with an open question. Give the patient as much time as they need to talk and allow them to finish before speaking.
- If this is an angry relative/patient that you are dealing with, always take on board their frustrations and apologise for any wrongdoing (regardless of what your assessment is of the complaint). Never disregard their concerns, as the matter will be of significant importance to them.
- Always try to empathise with the patient's situation.
- As the consultation progresses, use more closed questions to help guide the consultation.
- Summarise the main points discussed and addressed as you are going along, to act as a prompt to both you and the other individual for any other issues yet to be discussed.
- Remember that your ability to give accurate factual information and the way in which you relay this are both being assessed.
- Be clear and concise with your answers to any questions. Do not use jargon.
- During your discussion, remember the four main ethical principles and use them to help guide your approach to any ethical dilemma:
  - Autonomy: Respect for the individual and the choices they make.
  - Justice: Equality in the distribution of healthcare resources.
  - Beneficence: To act in the best interest of the patient.
  - Non-maleficence: Actions should not harm the patient (based on the principle of *primum non nocere*: first do no harm).
- Remember the issue of confidentiality in this station. Confidentiality may be breached in certain situations, such as child protection, notifiable diseases, fitness to drive and serious crimes.





## BREAKING BAD NEWS

### BACKGROUND INFORMATION

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You are asked to see a 64-year-old lady in the respiratory outpatient clinic with the results of her staging CT. The patient was referred with a history of a persistent cough unresponsive to several courses of antibiotics. A CXR requested by the general practitioner (GP) prior to the referral showed a probable mass in the right upper lobe. The CT confirmed the mass in the right upper lobe, with mediastinal and subcarinal lymph nodes, a small pleural effusion and multiple liver metastases. The current radiological staging is T3 N2 M1b, making this cancer inoperable.

### KEY POINTS FOR THE PATIENT

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- You have been seeing your GP over the past 2 months with a persistent cough.
- You have come to clinic to get to the bottom of this and don't want anyone else to be here with you.
- You have been given several courses of antibiotics which have not cleared the cough.
- You have been getting progressively more breathless over the past 1 month.
- Your children have noticed a change in your voice and you have lost a stone in weight recently.
- You smoked 10 cigarettes a day for 40 years but stopped 2 years ago.
- You have had pain in your right arm and difficulty raising it.
- The GP recently started you on blue and purple inhalers and a course of steroids.
- You are annoyed that your GP has not taken you more seriously.
- You have a strong suspicion that you have cancer, and this is keeping you awake at night.
- The main concern you have is that your father died of lung cancer and was in a lot of pain towards the end.
- You want to be offered all possible treatments available and are angry and upset when you are told that the treatment will be palliative and not curative.
- Other than smoking, you have always been very healthy, and so you understand that treatment will only be palliative.
- You worry how you will break the news to your family.
- You ask if the cancer may have been treatable if you had been referred earlier.
- You want to lodge a formal complaint against your GP, as things may have been different if you had been referred sooner.

### SUGGESTIONS FOR THE CANDIDATE

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- Introduce yourself to the patient and ask the patient their understanding of why they have been referred to the clinic.
- Ask them if they have come with anyone today and if they would like anyone else present.
- Ask the patient about the symptoms they have been having, what prompted them to see their GP and about any treatments they have had.

- Ask about their main concerns and what they think might be the underlying cause.
- Recap the history to the point where investigations began and tell the patient that you have all the results.
- Explain that you have some bad news and ask again if the patient would like anyone present with her.
- The main reason for investigation was to exclude a malignancy.
- Unfortunately, the results show that there is a tumour in the lung involving lymph nodes and lesions in the liver; the diagnosis is likely to be cancer.
- Pause for the patient to take the information in and give them as much time as they need.
- Ask the patient if they have understood what you have told them before proceeding.
- Explain that the case has been discussed at the lung cancer multidisciplinary team (MDT) meeting.
- Explain that because of the spread, the cancer cannot be surgically resected and that treatment options will be limited to disease control and not cure.
- Ask the patient what their main concerns are and acknowledge them.
- Try to acknowledge each concern and offer help.
- Gauge how much information the patient would like to be given.
- Reiterate that although it is not possible to treat the underlying cause, there are other services and support that can help the patient to cope with the diagnosis and the disease.
- Acknowledge the patient's anger and frustration with the GP, and that they ought to contact the GP to discuss the issues.
- Advise that it would be difficult to say if the cancer could have been caught earlier, as you don't know how aggressive it is.
- Tell the patient that there is a lung cancer specialist nurse that will see them today before they go, and that they will be a regular point of contact.
- Offer the patient to help break the news to the family.
- Recap the information given and ask if the patient has any questions.
- Explain that you will see them in clinic next week and ask if they would like to bring any family or friends along with them to discuss the diagnosis again.

## THEMES EXPLORED

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### BREAKING BAD NEWS FOR AN INCURABLE CONDITION

- This is often difficult due to the emotions involved and the range of patient responses that one may encounter. Remain calm yourself and make sure that you listen to the patient.
- Ensure that you ask the patient about their understanding of the current situation before proceeding to give them the news.
- Do not use any euphemisms or jargon and make sure that you are being clear with the diagnosis.
- If the patient has a diagnosis of cancer, use the word *cancer*.
- Make sure that you have all the correct/relevant information to hand.
- Appear empathic and ensure that the patient has time to digest the information given.



- Remember to summarise the key points at the end of the consultation and offer to see them again in clinic.
- Offer the relevant support: specialist nurses, palliative care teams, information leaflets and contacts for support groups.

## DEALING WITH AN ANGRY PATIENT

- Acknowledge that the patient is angry or upset and let them know that you are aware and that it is a normal response.
- Allow them to voice their anger and do not react to the situation.
- Ask them what specific things are making them angry and offer them potential solutions to help overcome these issues.

## CARE IN THE DETERIORATING PATIENT

You are the registrar covering the medical wards for the evening. You have received a call from one of the nurses on a respiratory ward to speak to a patient's son. The patient is a gentleman with bowel cancer with liver and lung metastases. He was admitted to hospital with dyspnoea and has been treated for a lower respiratory tract infection with intravenous antibiotics. Despite this, he has deteriorated in the 5 days since admission. The patient himself recognised this on the registrar ward round today, and following a discussion, antibiotics were stopped with a decision being made to commence morphine for dyspnoea and refer the patient to the palliative care team. The ward team were asked by the patient about his prognosis today, and thought this would likely be measured in 'days to weeks'. The family were not on the ward for afternoon visiting, and the nurse had called the son, asking him to come in for the consultant ward round tomorrow. Following the call, he has visited this evening and is angry that his father is 'being left to die'. You have gained consent from the patient to speak to his son.

## KEY POINTS FOR THE SON

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- You have come to the hospital to see your father, following a call from the nursing team. You are the patient's only child.
- Your father has had bowel cancer for a few years. During that time, he has had surgery and a few courses of chemotherapy.
- During your father's last trip to the oncologist, they felt he was not fit for more chemotherapy. You still hold out hope that he will be offered further treatment.
- You know that your father was admitted to hospital a few days ago and has been treated for a 'chest infection' with antibiotics.
- You were hopeful that he would improve while in and be discharged back to his house where he lives alone. You are aware that he's been bedbound since admission.
- On receiving a call from the nursing team, you came straight to hospital. On hearing antibiotics have been stopped, you feel like the hospital is 'giving up' on your father.

- You have also been told by the nurse that your father has been started on morphine. You have been told this was started to help his breathing, although you think it is being used for 'euthanasia'.
- You have not spoken to your father about this since you came in, but you are worried that he's 'lost his fight'.
- You find it hard to accept that your father is deteriorating. On direct questioning, you admit that you are lacking support (you don't have any other family). You don't want your father to die, as he is 'all you have'.
- If all the above issues are covered, you will feel supported, be accepting of the plan and come to the ward round tomorrow to meet the consultant and discuss future plans.
- If you are not given appropriate information or feel you are not listened to, you will get angrier and leave the conversation.

## SUGGESTIONS FOR THE CANDIDATE

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- Note that you are dealing with an angry relative.
- Allow the son to voice his concerns, providing adequate time for him to do so.
- Do not interrupt him.
- Acknowledge the son's distress.
- Talk through his understanding of his father's condition. This could include his father's likely prognosis.
- Explain that adequate treatment has been given for the lower respiratory tract infection.
- Explain that much of his father's dyspnoea could be related to lung metastases, and the general fatigue related to advanced cancer.
- Explain that oncological treatment will not be possible, but there is much that can be done to improve his father's symptoms.
- Explain that morphine is now used regularly to treat dyspnoea in advanced illness. This will be started at a low dose and prescribed safely. It is usually very well tolerated.
- Explain that a referral to the palliative care team has been made to optimise symptom control measures, but also to help with future plans (rapid discharge home may be possible).
- Reassure the son that the plan was discussed with his father, who was in agreement.
- Explain to the son that the medical and nursing teams on the ward, along with the palliative care team, will try to support him psychologically while his father is in hospital. Further support for the son could be offered from the hospital chaplaincy team if he were in agreement.
- Reassure the son that if he has further questions after this meeting, these could be discussed on the consultant ward round tomorrow.

## THEMES EXPLORED

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### SUPPORTING AN ANGRY RELATIVE

- It is important to both acknowledge the relatives' distress and find out why they are angry.
- Talk through their concerns one by one, first by listening and then by offering whatever information you can.



- Offer follow-up to the conversation, including a further opportunity to ask questions after this discussion.
- Recognise anticipatory grief (grief occurring before a patient has died) as a cause of distress in the families of unwell patients.

## RECOGNITION OF A DETERIORATING PATIENT

- Recognition of a deteriorating patient is important for many reasons. First, it allows an appropriate treatment plan to be constructed. Additionally, it allows clinical teams to keep patients and families updated, and to help them plan for the future (such as the patient's preferred place of death).
- Signs that a patient is in the last weeks of life include fatigue, decreased mobility, loss of independence in activities of daily living (ADLs), decreased appetite and impaired cognition.
- Signs that a patient is in the last days of life include decreased fluid intake, impaired swallowing, decreased conscious level and changes in breathing pattern.
- While this case did not cover this specifically, it is important to have an understanding of the 'priorities of care for the dying person'.
- It is also important to be aware of recent developments in decisions about cardiopulmonary resuscitation.

## REFERENCES

Leadership Alliance for Care of Dying People. (2014) One chance to get it right. Available from <https://www.gov.uk/government/publications/liverpool-care-pathway-review-response-to-recommendations>. Accessed 4 August 2016.

Resuscitation Council (UK). (2014) *Decisions Relating to Cardiopulmonary Resuscitation*. 3rd ed. Available from <https://www.resus.org.uk/dnacpr/decisions-relating-to-cpr/>. Accessed 4 August 2016.

## DRIVING REGULATIONS

Mr David Clegg has come to see you in the sleep clinic. He is a 55-year-old gentleman who has been complaining of symptoms of tiredness over the past year. He has been investigated by his GP, and recent thyroid function tests (TFTs) and a full blood count (FBC) have been normal. The patient is complaining that his symptoms are interfering with his work and he is falling asleep inappropriately. The GP is concerned that the patient may have obstructive sleep apnoea/obesity hypoventilation syndrome. The patient weighs 144 kg and is 1.65 m tall. Discuss the diagnosis and related information with the patient.

## KEY POINTS FOR THE PATIENT

- You have been referred to the clinic by your GP to investigate the cause of your sleepiness.
- All your blood tests have been normal.
- Your GP thinks that the tiredness may be related to the fact that you are overweight.
- The tiredness is having a significant impact on your life.

- You have fallen asleep in several important meetings at work, which was rather embarrassing.
- You are concerned as you have found yourself falling asleep at the wheel of your car. You nearly crashed into a barrier the other day.
- Your wife sleeps in a separate bedroom, and this is affecting your sex life as well.
- You have tried losing weight, but you are not getting very far.
- You drink up to three pints of beer most nights.
- You wake up most mornings complaining of a headache.
- You feel as though you have not had a good night's sleep for months.
- You are beginning to feel depressed about this now.
- You want to know what can be done.

## SUGGESTIONS FOR THE CANDIDATE

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- Start by asking the patient if he knows why he has been referred to the sleep clinic.
- Ask him specifically how the tiredness is impacting on the various aspects of his life. Ensure that you ask about work and home.
- Ask about a bed partner and if they have reported the patient snoring at night or having periods where they stop breathing.
- Ask the patient what they understand about the terms *obstructive sleep apnoea* and *obesity*, and whether the GP has explained anything about the potential diagnosis.
- Go on to explain the potential diagnosis.
- Explain to the patient that further tests will have to be carried out before confirming this.
- Initially, the patient will be asked to complete an Epworth Sleep Score and then further tests will include blood tests (FBC, urea and electrolytes [U&Es], liver function test [LFTs], TFTs and ferritin levels), overnight oximetry and possibly limited/full polysomnography.
- Advise the patient on lifestyle factors: cut down on alcohol intake and attempt to lose weight.
- One of the main issues that will need to be covered here is that the patient is falling asleep at the wheel of his car.
- You must advise him that he has a condition that is affecting his ability to drive, and he will need to stop driving and has a legal duty inform the Driver and Vehicle Licensing Agency (DVLA). The regulation for group 1 drivers is that they need to stop driving until satisfactory control of symptoms has been attained.
- Ask the patient if they have any questions.
- Summarise the above information for the patient before ending the consultation and offer to give them some information leaflets prior to leaving.

## THEMES EXPLORED

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- The main points to cover in this case are to give the patient information regarding the potential diagnosis and also about what investigations may be required.
- The patient has specifically mentioned that he is falling asleep while he is driving. This needs to be taken seriously, and it is the responsibility of the medical practitioner to advise the patient to stop driving and inform the DVLA immediately. This must also be documented in the patient's notes.



- There are many medical conditions about which the patient needs to inform the DVLA and may also need to stop driving.
- Once you have discussed this with the patient, it is their responsibility to inform the DVLA. If the patient continues to drive against medical advice, you will need to urgently contact the DVLA and disclose the relevant medical information in confidence. Remember that you must inform the patient that you are going to do this and you should confirm this in writing to the patient and their GP once you have informed the DVLA.

#### Driving Regulations for Selected Medical Conditions

Condition	Group 1 regulations	Group 2 regulations
First unprovoked/isolated seizure	6 months off driving from date of first seizure (or factors to suggest increased risk of further seizures, then cannot drive for 12 months)	5 years off driving from first seizure and must be on any treatment for epilepsy in that time
Epilepsy or multiple unprovoked seizures	Review licence may be issued if no seizures for 5 years (with medication if required)	Must remain seizure-free for 10 years with antiepileptic medication
Stroke/transient ischaemic attack (TIA)	No need to notify DVLA (but stop for 1 month) unless residual neurological defect beyond 1 month (especially if visual field defect or cognitive defect and limb function impairment)	Licence revoked for 1 year If recurrent TIAs/stroke, must undergo functional cardiac testing first
Acute coronary syndrome	Cannot drive for 1 week after successful coronary angioplasty	Cannot drive for 6 weeks until exercise test requirements fulfilled
Pacemaker insertion or box change	Do not drive for 1 week	Do not drive for 6 weeks
Implantable cardioverter defibrillator (ICD) implantation (prophylactic)	Cannot drive for 6 months from the date of insertion	Permanently barred

Source: Adapted from Driver and Vehicle Licensing Agency, Assessing fitness to drive – A guide for health professionals' document, DVLA, Swansea, 2016. Available from [https://www.gov.uk/government/uploads/system/uploads/attachment\\_data/file/517268/Fitness\\_to\\_drive.pdf](https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/517268/Fitness_to_drive.pdf).

Note: Group 1 includes cars and motorcycles. Licences are valid until 70 (unless restrictions applied for medical conditions), after which renewal is every 3 years.  
Group 2 includes large lorries (Category C) and buses (Category D). Licence valid for a maximum of 5 years only and must be renewed every 5 years (or at 45 years if issued before 19 January 2013) until aged 65 years, following which there is an annual review.  
General Medical Council (GMC) guidance. The driver is legally responsible for informing the DVLA about any condition or treatment that impairs the patient's fitness to drive.

## REFERENCE

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Driver and Vehicle Licensing Agency. (2016) Assessing fitness to drive – A guide for health professionals' document. Available from [https://www.gov.uk/government/uploads/system/uploads/attachment\\_data/file/517268/Fitness\\_to\\_drive.pdf](https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/517268/Fitness_to_drive.pdf). Accessed 20 April 2016.

## INITIATING A NEW THERAPY

You are the medical registrar in a rheumatology outpatient clinic. Mrs Patel is a 52-year-old lady who has a history of severe rheumatoid arthritis. She has tried several disease-modifying drugs (including methotrexate and anti-tumour necrosis factor [TNF] therapy) without significant reduction in disease activity. She has come to see you in clinic today as you wish to start therapy with rituximab.

## KEY POINTS FOR THE PATIENT

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- You have suffered with severe rheumatoid arthritis for many years now.
- You have active disease which is proving difficult to control and is affecting your quality of life and ability to work as a shop assistant.
- Steroids have caused significant thinning of your bones, so you are reluctant to have the dosage increased.
- You have tried several disease-modifying antirheumatoid drugs (DMARDs), including methotrexate, which caused a problem with your liver tests and so had to be withdrawn. You have also been treated with anti-TNF therapy with infliximab, but your disease remains active.
- You are frustrated with repeated trials of medications which either do not work or cause more problems due to side effects, and have read about MabThera (rituximab) on the Internet and insist that you are treated with this.
- You understand this is an expensive treatment but insist that this is the treatment you want.
- You want to know more about how this drug works. Will it cure you?
- You want to know what the potential effects of the drug are. Importantly, will it damage your liver like methotrexate?
- You are afraid of the immunosuppressive effects of the drug, as you had a tuberculosis (TB) scare about 20 years ago, back in India, when a lump was found in one of your neck glands not long after your mother was diagnosed with TB.
- You do not recall whether you received any treatment at the time, only that you had a biopsy taken.
- You are concerned that the history of TB will affect your eligibility for treatment with rituximab and want to know what can be done to ensure that initiation of treatment is not delayed.

## SUGGESTIONS FOR THE CANDIDATE

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- You have a very knowledgeable patient in clinic with you, who is very well read about her condition.
- You ask how she is managing at present with her condition, and how it is affecting her life.





- Listen to and empathise with the patient's concerns regarding the effects of the condition and the impact it has on her.
- You note that the patient has tried several DMARDs and anti-TNF therapy in the past without much benefit (and with lots of complications).
- You explain that abnormalities with liver function tests are a well-documented effect of treatment with methotrexate, and unfortunately, osteoporosis is one of the long-term effects of treatment with steroids (as well as other effects, such as hypertension, diabetes, fluid retention, stomach ulcers and eye problems).
- You enquire where the patient read about MabThera – an official evidence-based website or from a general Internet search.
- You explain that rituximab is a treatment that works on the B-cells of the immune system to reduce inflammation and improve your symptoms, and that there are strict criteria for eligibility; however, she appears to have met the criteria, having failed DMARDs, including methotrexate and at least one anti-TNF agent.
- Explain that the medication works by reducing inflammation but cannot cure the disease.
- Give the patient time to take this in and ask any questions.
- Explain that the treatment is given as an infusion in hospital (usually as a day case procedure) and can take up to 6 hours to complete.
- Occasionally, people can feel unwell during the infusion and develop a fever and/or wheeziness or feel dizzy/light-headed (due to a drop in blood pressure), but usually measures can be taken to overcome these, such as slowing the rate of the infusion (but in severe cases, it may have to be stopped).
- The main risk of the treatment is of infections, as rituximab works by dampening down the immune system, so if you develop a sore throat, fever or other signs of infection, then they must contact a doctor immediately. Rituximab can also be associated with serious infections, such as TB. Enquire about past TB exposure.
- Before initiating therapy, TB needs to be excluded with a chest radiograph. If it is positive, and as there is uncertainty whether the TB was adequately treated previously, she will need chemoprophylaxis before treatment with rituximab, but this should have been addressed prior to treatment with anti-TNF therapy. In addition, she will need to have an influenza vaccination and ensure that she is up to date with pneumococcus vaccination.
- Reassure her this is usual practice, that it is safer to delay starting rituximab for a short time to prevent potential life-threatening infection with TB, and to ensure that she is well-vaccinated to reduce risk of infections, and that this does not mean she will be precluded from receiving treatment.
- Make sure you listen to and address the patient's concerns. Summarise and ask if there are any questions.
- Offer an information leaflet about rituximab and schedule another appointment to discuss any concerns and initiate the treatment pathway.

## THEMES EXPLORED

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- *Autonomy:* The main theme explored in this case is how to counsel a patient effectively regarding a new treatment with potentially serious side effects, without coercion, to enable them to make an informed decision as to whether to proceed with the treatment.

- *Beneficence*: With novel/biological therapies, there is a risk–benefit balance between potential adverse effects of the drug versus the desired benefits, which is often difficult when the long-term effects of such medications are often not fully known.
- *Justice*: National Institute for Health and Care Excellence (NICE) guidelines suggest that patients with active rheumatoid arthritis who had an inadequate response to, or are intolerant of, other DMARDs, including at least one TNF inhibitor, should be considered for rituximab (and methotrexate). This would justify the need to give such potentially harmful treatments.
- *Do no harm*: Prior to treatment with anti-TNF therapy, patients should be screened for TB, and active TB must be adequately treated, as therapy carries an increased susceptibility to developing TB. They should also be up to date with other vaccines, such as pneumococcus and receive influenza vaccine.
  - Patients with a past history of extra-pulmonary TB or abnormal CXRs require close monitoring on treatment.
  - Patients with previously inadequately treated TB require chemoprophylaxis before commencing treatment.
  - Patients require close monitoring for symptoms of TB while receiving biological therapy and for 6 months after stopping.
  - If patients develop symptoms suggestive of TB on biological therapy, they will require full treatment with chemotherapy.

## REFERENCES

- Buch MH, et al. (2011) Updated consensus statement on the use of rituximab in patients with rheumatoid arthritis. *Ann Rheum Dis* 70(6): 909–20.
- Bukhari M, et al. (2011) BSR and BHPR guidelines on the use of rituximab in rheumatoid arthritis. *Rheumatology* 50(12): 2311–3.
- National Institute for Health and Care Excellence. (2010) TA195. Adalimumab, etanercept, infliximab, rituximab and abatacept for the treatment of rheumatoid arthritis after the failure of a TNF inhibitor. Available from <https://www.nice.org.uk/guidance/ta195>. Accessed 24 April 2016.

## LONG-TERM CONDITION

You are the registrar in the renal outpatient clinic. You are seeing a 32-year-old female patient who has recently been given a diagnosis of autosomal dominant polycystic kidney disease (ADPKD). She has come back to clinic today wanting more information regarding her diagnosis and the implications it may have for her future. Her mother also has the condition and may be starting dialysis in the near future. She has a young daughter and is planning to expand her family.

## KEY POINTS FOR THE PATIENT

- You were recently seen in the renal clinic when you were given a diagnosis of polycystic kidney disease.
- You have had some time to digest the information given to you previously and you now have several concerns that you would like to discuss with the doctor today.



- You are extremely anxious regarding the diagnosis.
- You have a three-year-old daughter and had been planning to expand your family.
- You are concerned regarding the prognosis of the condition and associated complications.
- You enquire if there are any alternative treatments or surgical options available to help prevent any complications and decline in renal function.
- Your mother also has the disease, and her doctors are considering starting her on dialysis in the near future.
- You wish to know whether this might be the case for you in the future and, if so, when might this be and whether you require a kidney transplant at some stage.
- You wish to know if there is anything that can be done to prevent yourself from getting to the stage your mother is currently at, i.e. requiring long-term renal replacement therapy (RRT).
- As your family is not yet complete, you wish to know if this diagnosis would prevent you from having any more children and what are the risks involved.
- If you were to become pregnant, would pregnancy affect the progress of the disease?
- Since both you and your mother have the condition, you are extremely concerned about the inheritance of the condition and whether you could have passed on the disease to your daughter.
- You want to know if your daughter can be tested and if there is anything that can be done to prevent her from developing the condition.

## SUGGESTIONS FOR THE CANDIDATE

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- It is important first to establish the amount of information the patient received during the previous consultation and how much she has retained.
- Summarise the key points and offer to discuss the diagnosis again, and clarify any points that she may not have fully understood.
- Ascertain the patient's main concerns by asking if she has any specific questions in mind that she would like answered.
- Attempt to alleviate some of the patient's anxieties by reassuring her that you and the team are there to help and provide whatever support she requires.
- Explain again to the patient that ADPKD is an inherited condition (autosomal dominant) and that the inheritance pattern of the condition means that there is a 50% chance that she has passed it on to her daughter.
- Explain that as it is a cystic condition, the cysts could develop in other organs (liver, pancreas), as well as the kidneys. Problems she might experience include recurrent urinary tract infections, infection or bleeding within a cyst, pain due to the size of the cysts and high blood pressure, all of which are treatable.
- Explain that end-stage renal disease is a potential complication that may occur some years after diagnosis. If her renal function were to decline over the course of time, then she would be prepared for dialysis in advance of needing it. An alternative possibility would be transplantation when a kidney became available, ideally before she needed to start dialysis; however, if this were not possible, then she may require dialysis initially prior to transplantation.
- Explain that the mainstay of treatment is trying to preserve kidney function for as long as possible by reducing complications and treating them aggressively, i.e. good blood pressure control, treating infections, etc.

- Explain to the patient that she will be closely monitored over the coming years with regard to her renal function.
- Explain that newer treatments are being developed, such as tolvaptan, which has recently been approved by NICE for rapidly progressive PKD, but the treatment, as with any treatment, is associated with some side effects, such as polyuria and risk of liver injury. However, she would not currently be eligible for this.
- Reassure the patient that this disease will not prevent her from having more children, but that in future pregnancies, she will be monitored more closely and may have an obstetrician-led pregnancy with close monitoring of her renal function.
- Explain that this may mean more frequent prenatal visits to the hospital and scans to ensure that she and her baby are both doing well. In addition, she may be more closely monitored during and after delivery.
- Explain that pregnancy per se will not affect disease progression currently.
- Explain that the main way of screening for this disease is using ultrasound imaging to look for the presence of cysts within the kidney. This is usually done when the patient is in their 20s.
- When asked about genetic testing, explain that there are many genetic mutations that can give rise to the disease, so it may not be possible to identify the specific one causing her disease. Assure her that her daughter can be screened when she is older. You can offer for her to be referred to a geneticist if she is keen to do so.
- Summarise that although PKD is a chronic/lifelong condition, it will not prevent her from leading a full and active life. She will be closely monitored throughout with specialist services when needed. It will not prevent her from completing her family, and should she progress to needing dialysis, she will be thoroughly supported and helped through this.

## THEMES EXPLORED

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- When discussing a new diagnosis of a chronic/life-limiting condition, it is important to ascertain the level of information the patient has received beforehand, and their understanding.
- Respecting the patient's autonomy is crucial, and it is important to gauge the level of information the patient would like to receive. Be open and honest. Try not to use jargon and use language that the patient can understand.
- Offer the patient details of support groups where they can obtain further advice in dealing with the diagnosis, and provide information leaflets that explain the condition.
- Explain that having a chronic disease does not mean that she cannot complete her family.
- She must be informed that 50% of her offspring may also be affected with the condition due to the pattern of inheritance. She will not be expecting (or wanting) to hear this, so be cautious in your approach.
- The importance of reducing complications must be emphasised.
- If renal decline is progressive, then development of end-stage renal disease may be unavoidable, although there are options of renal replacement therapy and the need for RRT will not be imminent. Reassure her that new therapies are always emerging.
- Offer a further consultation to discuss any points and referral to an obstetrician, if desired, for future pregnancy planning and a geneticist for further genetic counselling if she so desires.



## REFERENCE

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National Institute for Health and Care Excellence. (2015) Guidance TA358. Tolvaptan for treating autosomal dominant polycystic kidney disease. Available from <https://www.nice.org.uk/guidance/ta358/resources/tolvaptan-for-treating-autosomal-dominant-polycystic-kidney-disease-82602675026629>. Accessed 24 April 2016.

## MEDICAL ERROR

You are the registrar covering the medical wards for the weekend. You have received a call from one of the nurses on the elderly care ward to speak to a patient's daughter. The patient's daughter is an ITU nurse and is very concerned about the fact that her mother had appeared quite drowsy on her arrival. She alerted the nurses, and they checked her finger-prick blood glucose level, as she is on insulin, and found her to be hypoglycaemic. It further transpired that the dose of insulin that was prescribed on the chart was incorrect, and as a result, she was given a much higher dose of subcutaneous insulin than normal for her.

## KEY POINTS FOR THE DAUGHTER

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- You have come to the hospital to see your mother, who was admitted with an acute coronary syndrome.
- You are very distressed, as when you arrived, she appeared to be drowsy and not responding to you.
- You alerted the nursing staff and asked them to check her blood glucose level, as she has diabetes treated with insulin, and to check the rest of her observations.
- Your mother was found to have a very low blood glucose level, and this was the likely cause of her drowsiness. She was given glucogel, and this improved her symptoms.
- The nursing staff had informed you that your mother had recently had a sliding scale of insulin stopped and was given her regular dose of insulin. The nurses tell you that she was prescribed and given 30 units of Novomix 30. You tell them that her regular dose is only 10 units of Novomix 30 BD.
- You are very angry and upset that this has happened and want to see the doctor on call.
- You ask the doctor why the insulin was prescribed incorrectly when your mother's list of medication was brought in with her.
- You want to know why this was not double-checked by the pharmacist.
- You feel that it is completely unacceptable that this has happened when there should be procedures in place to prevent prescribing errors such as this. You are relieved that your mother has recovered, but still disappointed in the lack of care.
- You want to speak with the consultant in charge of your mother's care and make an official complaint about the incident, as you are concerned that it was a case of bad practice.
- You also insist that an incident form is completed regarding the event to ensure the matter and the individuals involved receive feedback and training.

## SUGGESTIONS FOR THE CANDIDATE

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- Note that you are dealing with an angry relative who is a fellow health professional.
- Allow the relative to voice all their concerns and provide adequate time for them to do so.
- Do not interrupt.
- Acknowledge the daughter's concerns.
- Acknowledge what has happened and apologise for the error.
- Address each of the concerns that the patient's relative expresses.
- Be open and honest and acknowledge the fact that a prescribing error has been made resulting in harm to her mother.
- Assure the daughter that all junior doctors are given training in insulin prescribing and that you will ensure that it is correctly prescribed on the drug chart now.
- Assure the daughter that you will regularly review her mother to ensure that her blood sugars remain stable.
- Offer to arrange a time for the daughter to speak with her mother's consultant.
- Reassure her that you will fill out an incident report regarding the matter, and this will be escalated to the ward manager and the consultant in charge of the patient.
- Advise the daughter that if she or her mother would like to take the matter any further, they can contact the patient liaison office and write a formal complaint, which would be addressed in accordance with hospital policies.
- Once again, apologise for any distress caused to the patient and assure the daughter that you will relay her concerns to the various parties involved.

## THEMES EXPLORED

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- The main ethical principle addressed in this case is non-maleficence (first do no harm). The other theme explored is the duty of candour.
- The patient has directly suffered harm as the result of a wrong prescription of insulin.
- The duty of candour is a legal duty which entails all healthcare providers to inform patients (or their representative) and to apologise to them if there has been a mistake made in their care which has resulted in harm. This ensures that patients are given accurate and honest information.
- When addressing any form of complaint, it is important that you have on hand as much information as possible to give to the patient or their relatives.
- You should acknowledge any error that has been made, and you must apologise for this.
- Offer to answer any questions that the patient or relative may have in relation to their concerns to help alleviate the situation.
- Do not try to conceal any information which may later come to light.
- If they are not happy with your explanation, offer to arrange for them to speak with the consultant in charge of the patient's care.
- Offer them other sources of support regarding patient welfare, such as patient advice services, which are available in all hospitals.
- Reassure them that the matter will be taken seriously and dealt with appropriately.
- All hospitals will have processes in place to report adverse incidents that have taken place and to learn from the outcome of these reports.



## REFERENCES

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- Care Quality Commission. (2015) Duty of candour: Information for all providers: NHS bodies, adult social care, primary medical and dental care, and independent healthcare. Available from [www.cqc.org.uk/sites/default/files/20150327\\_duty\\_of\\_candour\\_guidance\\_final.pdf](http://www.cqc.org.uk/sites/default/files/20150327_duty_of_candour_guidance_final.pdf). Accessed 4 August 2016.
- National Health Service. (n.d.) Report a patient safety incident. Available from [www.nrls.npsa.nhs.uk/report-a-patient-safety-incident/](http://www.nrls.npsa.nhs.uk/report-a-patient-safety-incident/). Accessed 4 August 2016.

## MENTAL CAPACITY

You are the medical registrar on call covering the weekend. You have been asked to speak to a relative on one of the medical wards, who wants her husband discharged against medical advice. The patient has a background history of Parkinson's disease, epilepsy and vascular dementia. The patient was admitted on this occasion with a lower respiratory tract infection and a UTI. He is currently being treated with intravenous fluids and antibiotics and has systemic signs of sepsis. The patient's wife is adamant that she wants him discharged from hospital and sent home. You try to speak to the patient, but he is too confused to communicate.

## KEY POINTS FOR THE PATIENT'S WIFE

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- You are the patient's main carer and have looked after him for many years now.
- You are adamant that he should be discharged, albeit against medical advice.
- You feel that his condition is worsening, as he is not in a familiar environment.
- You also feel that there is no point of him being in hospital, as he is not getting the one-to-one care which he would be getting at home.
- You question the doctor as to why the patient cannot just have antibiotics at home.
- You know that the patient has signed an advance directive stating that he should not get treatment that would unnecessarily prolong his life.
- You feel that the patient is not sleeping at night as it is too noisy on the ward.
- You know that you do not hold lasting power of attorney status, but you know the patient best and this is what he would have wanted.
- You are angry at the doctor because you think nothing is being done as it is the weekend.
- You question the doctor as to what he can do to stop you from taking your husband off the ward.
- You feel that keeping him in hospital is not in his best interests.
- You can give him antibiotics orally and make sure he drinks plenty of fluids at home.
- You are frustrated, as you feel that your views are not being taken into consideration.

## SUGGESTIONS FOR THE CANDIDATE

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- You are approached by a very angry and frustrated relative.
- Make sure you listen and address the relative's concerns. The best way to counter this discussion is to approach the relative with empathy and kindness to diffuse the situation.
- Ask her what is worrying her regarding the patient's stay in hospital.

- Reiterate that the patient is suffering from a urinary tract infection and a chest infection, and is currently having intravenous antibiotics and fluids. Explain that the patient is not well enough for treatment with oral antibiotics as yet, as evidenced by his ongoing signs of sepsis.
- Explain that you are part of a team and that you are all acting in the patient's best interest.
- When the relative tells you about the advance directive, you can tell her that this infection is a treatable condition and you are actively treating her husband and are not preparing for end-of-life care currently.
- Explain to the relative that you will try to ensure that everything possible will be done to make the patient more comfortable, such as moving him to a side room or a quieter area of the ward where he should be able to get more rest.
- When the patient's relative asks what you can do to stop her, tell her that you would hope to resolve the situation before this happened, but if the need arose, you would have to involve hospital security.
- Explain that you have tried to assess the patient's mental capacity, but he is too confused to understand what you are saying.
- Ask the relative if she has lasting power of attorney.
- Explain that as she does not have this status, as the healthcare professional, you are acting in the patient's best interest by keeping the patient in hospital.
- You would be going against your duty of care by allowing the patient to leave.
- Summarise what you have discussed and ensure that you convey an empathic tone throughout to the relative and acknowledge her concerns.
- Suggest that the relative could speak to the consultant in charge of the patient's care, after the weekend.
- Reiterate that you are doing your best to treat the patient.

## THEMES EXPLORED

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- The main theme explored in this case is the issue of mental capacity. The Mental Capacity Act came into force in 2007 in England and Wales, and it helps provide a framework to empower and make decisions for people who are unable to do so for themselves.
- Every adult has the right to make their decisions and must be assumed to have capacity unless proven otherwise. People should be supported to make their own decisions, unless deemed to lack capacity, and any individuals with capacity who make seemingly unwise decisions should be respected.
- To assess whether a patient has capacity, they must be able to understand and retain the information presented to them, weigh up that information and communicate the decision back to you.
- If a patient does not have capacity, then as the healthcare worker responsible for the patient, you can take decisions regarding the patient's medical care. It is also important to involve the family and carers and seek their views and opinions as to what the patient may have wanted.
- Any intervention made should be the least restrictive intervention and should always be in the patient's best interest.
- Patients can sign advance directives refusing specific treatments that they would not want should the situation arise. These are legally binding documents that need to





be signed in the presence of a witness. They should also contain a statement that the decision should be enforced even if life is at risk.

- If there is any issue regarding the validity of an advance directive in an emergency situation, the appropriate treatment can be given until the validity of the document is verified.
- The patient can also appoint a lasting power of attorney who can make medical (and other) decisions on behalf of the patient who lacks capacity. This document needs to be registered with the Office of the Public Guardian for it to be valid.
- In a situation where a patient lacks capacity and has no representative to discuss their medical treatment, an independent mental capacity advocate (IMCA) can be appointed to act as an advocate for the patient in the decision-making process. An IMCA is not needed if it is an emergency treatment or if the patient is detained and being treated for a mental illness under the Mental Health Act.

## REFERENCE

Office of the Public Guardian. (2014) Mental Capacity Act: Making decisions. Available from <https://www.gov.uk/government/collections/mental-capacity-act-making-decisions>. Accessed 5 August 2016.

## NEW DIAGNOSIS

You are the registrar in the gastroenterology outpatient clinic. Your next patient is a 19-year-old man who has recently presented with several episodes of bloody diarrhoea. The patient has had a recent colonoscopy which confirms the diagnosis of ulcerative colitis (UC). Discuss the diagnosis with the patient and the implications of the disease.

## KEY POINTS FOR THE PATIENT

- You have come to clinic today to find out the results of a colonoscopy that was undertaken to investigate the cause of the bloody diarrhoea you have been having.
- Previously, you have been fit and well with no other health problems. Over the past 6 weeks, you have had multiple episodes of bloody diarrhoea daily.
- When you were previously seen in clinic, the doctor mentioned that they were looking to see if there was any inflammation of your bowels that could account for your symptoms. However, you did not really understand the explanation given.
- When you are given the diagnosis of ulcerative colitis, you ask the doctor to fully explain this and any problems that are associated with the disease.
- You are worried, as you have just finished your gap year and are due to start university soon and wonder how this will affect your social life.
- You want to know what medication you will need to take.
- You want to know if you will still be able to drink alcohol and go out with your friends and lead a 'normal' life. You hope to go on to university – will this still be possible?
- You want to know how often you will need to see a doctor.

- You read an information leaflet in the waiting room and you want to know if you will need to have an operation and, more importantly, if you will end up with a stoma.
- When pressed by the doctor, you tell them that you are very concerned about this problem, as your dad died at a young age from bowel cancer and you wonder if this is something that you should be worried about, now that you have this diagnosis.

## SUGGESTIONS FOR THE CANDIDATE

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- Recap the history for the patient and initially gauge their understanding of why they have come to clinic and why they have had the procedure done.
- Ascertain the amount of information previously given and their level of understanding.
- Summarise the previous information.
- Explain that colonoscopy is the usual investigation to look for any active inflammation of the bowel that could be causing his symptoms.
- Explain that biopsies were also taken at the same time to help clarify the underlying disease/inflammatory process.
- Explain that all the results are now available and are consistent with a diagnosis of ulcerative colitis, which is an inflammatory condition affecting the bowel.
- Give them time to absorb the information that they have been given and then wait to see if they have any questions.
- Ask if they have heard of UC or any other inflammatory bowel condition before and, if so, what is their understanding of this.
- Offer to give them an overview of the disease. Do not use jargon.
- Explain that UC causes inflammation of the bowel wall. It is restricted to the large bowel and therefore results in bloody diarrhoea, so his presentation was not unusual or atypical.
- Although the disease mainly affects the bowel, there can be other manifestations outside the bowel with symptoms such as joint aches, rashes, mouth ulcers and sore eyes, but bowel symptoms are most common.
- Empathise with the patient and reassure him that with treatment, there is no reason why he cannot lead a normal life and pursue his studies, and that the condition could be looked after by a specialist in his university town if needed, or he could continue to come and see you and his appointments could be worked around his studies.
- Treatment aims are to control active disease, improve quality of life and maintain remission.
- Treatment decisions will always take into account the patient's individual needs and preferences.
- As he has only bowel symptoms, treatment would initially be local therapy with oral aminosalicylic acids (ASAs) and/or steroids if symptoms persist.
- If symptoms are still not well controlled, stronger therapies to dampen the immune system could be added at that point.
- Explain that the amount of alcohol he can drink will depend on the treatment he is taking; if on stronger immunosuppression, it would not be recommended, but he should only drink in moderation (and not exceed the current weekly maximum recommendation of 14 units per week spread over 3 or more days)



- Explain that initially he will need a more regular review (every couple of months or so) until the disease is under control, but thereafter, the intervals between clinic reviews would be increased.
- Reassure him that in between visits, there is always a specialist nurse available who he can contact for further advice or earlier review if required.
- Regarding surgery, reassure him that this is undertaken only in severe cases of colitis when the inflammation cannot be controlled with medical therapy, or in emergency circumstances. However, there are now many new treatments available (biological) that can help avoid surgical intervention altogether, even in circumstances where it might have been unavoidable in the past.
- Ask the patient if they have any other specific concerns or if there is anything at all they are worried about. Tell them it is quite natural to be frightened and worried about the new diagnosis, and that you are here to help/address any concerns they may have.
- When bowel cancer is mentioned, be honest and tell him that in patients with UC, there is a slightly increased risk of bowel cancer after having the condition for many years. The risk, however, is still small. Emphasise that all patients are closely monitored and there are surveillance measures with colonoscopy (as recommended by NICE) to pick up any disease early.
- Summarise the main points and emphasise that there is plenty of support available to help him cope with the condition, and the aim of treatment is to help him lead as normal a life as possible.

## THEMES EXPLORED

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- When giving the diagnosis of a new/chronic/life-limiting condition, the patient has the right to know the full facts regarding the diagnosis and all treatment options available.
- The patient's autonomy regarding their treatment decisions must be respected if they are of sound mind, especially any refusal of treatment.
- The individual must be allowed to use their own judgement in weighing the pros and cons of available options. They should never be coerced into making any choices.
- It is crucial for the information giver to not allow any preformed judgements about the patient to cloud the information given to the patient.
- If there are any errors or delays in getting to the underlying diagnosis, these should not be hidden and any apologies should be made to the patient and their family.
- In this scenario, it is a young patient who is clearly frightened as to what the future holds with this condition (regarding treatments including surgery, the possibility of cancer and whether he will still be able to lead a normal life). Reassure the patient constantly and give them plenty of sources of further information. Allow them time to digest the information and address their concerns as they arise.
- Remember his concern about the risk of cancer and be aware of the family history and with this, he is at increased risk and so should be offered ongoing annual colonoscopic surveillance. The benefits of such would be prevention of colorectal cancer with early detection, but the psychological impact of having this must also be considered with the patient's wishes.

- Remember to offer further ongoing support with use of the multidisciplinary team by offering their contact details, and arrange another follow-up appointment where the patient can go through the diagnosis and rediscuss any concerns they have or anything they did not understand.

## REFERENCES

- National Institute for Health and Care Excellence. (2011) Guidance CG118. Colonoscopic surveillance for preventing colorectal cancer in adults with ulcerative colitis, Crohn's disease or adenomas. Available from <https://www.nice.org.uk/guidance/cg118/resources/colonoscopic-surveillance-for-preventing-colorectal-cancer-in-adults-with-ulcerative-colitis-crohns-disease-or-adenomas-35109396155077>. Accessed 24 April 2016.
- National Institute for Health and Care Excellence. (2013) Guidance CG166. Ulcerative colitis: Management. Available from <https://www.nice.org.uk/guidance/cg166/resources/ulcerative-colitis-management-35109695126725>. Accessed 24 April 2016.

## ORGAN TRANSPLANTATION

You are a doctor working on the intensive care unit. You have been asked to discuss the possibility of organ donation with the family of a patient on ITU who has been pronounced brainstem dead. The patient is a young, fit and healthy 19-year-old girl who was involved in a road traffic accident. The patient's father is her next of kin, and you approach him to discuss the possibility of donation.

## KEY POINTS FOR THE FATHER

- Your daughter has been involved in a road traffic accident, in which she was hit by a car.
- She suffered extensive head injuries and also had a ruptured spleen.
- You were informed by the consultant in charge of her care that her injuries were very severe and that she may not survive. You were naturally extremely upset, as your daughter was young, fit and healthy.
- You were informed that if she deteriorated clinically, tests would be carried out to assess the condition of her brainstem.
- The nature of the tests was fully explained to you, and subsequently, after two rounds of tests, your daughter was pronounced brainstem dead.
- As a family, you are all in shock, as the last time you saw your daughter before the accident she had been excited about her plans for her gap year.
- When asked by the doctor about the possibility of organ donation, you are initially taken aback that this has been raised at your time of loss.
- After some time to think, you ask what this would involve.
- You recall that your daughter had previously mentioned that she had ticked on her driving licence application that she would wish to be an organ donor.
- You want to know what the process of donation would involve and how long it would take.
- You ask which organs are likely to be taken.



- You ask who would receive the organs and whether you would be able to contact them.
- Your main concern is whether your daughter would experience any pain when the organs are removed.
- You are also worried about the appearance of her body after the organs have been taken.
- You ask if there is anyone else who can give you any further information about the process.
- You answer on the family's behalf that despite your loss, you are willing to give permission for your daughter to be an organ donor, as this was her wish.

## SUGGESTIONS FOR THE CANDIDATE

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- Initially offer your condolences to the family and empathise with their loss.
- When you bring up the subject of organ donation, acknowledge what a difficult time this must be for them, but explain that the possibility of organ donation is regularly addressed on the intensive care unit (ICU).
- Ask the family if they are aware of any wishes their daughter may have had concerning organ donation. Specifically ask about an organ donation card, or whether it was recorded on her driver's licence.
- Explain the process ahead. You will contact the organ transplantation team, who will coordinate the process of organ removal. There will be various teams from around the country who will come to obtain the organs, as this hospital is not a transplant centre.
- Advise that you will need to seek the permission of the coroner before organ retrieval can take place, as the patient was involved in an accident
- The organs will go to individuals on a transplant waiting list, who have been identified as being suitable and most urgently in need of an organ transplant.
- As the patient was previously healthy, all the major organs could be used, as well as other body parts, such as corneas, tendons and possibly bones.
- Reassure the family that the patient will not feel any pain, as she is brainstem dead and therefore unable to feel pain, but that she will also be given opiates prior to the procedure to ensure this.
- The surgeons will carefully close the body after removing the organs so the patient's appearance will not be significantly disfigured.
- The family will not be able to contact the recipients of the organs, but the transplant coordinator would be able to tell them some general information on who the organs have gone to.
- The transplant team/coordinator will also be available to answer any further questions that the family may have.
- Empathise with the family again regarding their loss and thank them all for considering this decision at such a difficult time.

## THEMES EXPLORED

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- Organ transplantation is an area which is often very difficult to address, as there is so much emotion involved when broaching the subject with relatives who have just lost a loved one.

- Any patient who meets the following criteria should be considered for organ donation, and their family or carers should be offered this option if
  - The patient has suffered major and irreversible neurological damage leading to brainstem death or, alternatively, whose condition is such that continuing critical care is considered futile and withdrawal of treatment is being considered.
  - The patient is HIV negative.
  - The patient is not known or suspected to have Creutzfeldt–Jakob disease.
- If the next of kin disagrees with donation, despite the patient being on the organ donation register, it is current practice not to proceed with the donation regardless of patient autonomy.
- The legal aspects to remember are that the coroner’s permission must be sought (if required) before going ahead with retrieval.
- In the case of determination of death by brainstem testing, medical practitioners must follow the code of practice issued by the Department of Health in 1998.
- Prior to brainstem death testing, three preconditions must exist:
  - The patient’s condition is due to irreversible brain damage of known aetiology.
  - The patient is in unresponsive coma (potential reversible causes have been excluded).
  - The patient is apnoeic and mechanically ventilated.
- If these conditions are met, the standard tests are carried out by two qualified doctors and are repeated after a short amount of time.

## REFERENCES

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- Academy of Medical Royal Colleges. (2008) A code of practice for the diagnosis and confirmation of death. Available from <http://www.bts.org.uk/Documents/A%20CODE%20OF%20PRACTICE%20FOR%20THE%20DIAGNOSIS%20AND%20CONFIRMATION%20OF%20DEATH.pdf>. Accessed 19 August 2016.
- Department of Health. (1998) A code of practice for the diagnosis of brain stem death. Available from [http://webarchive.nationalarchives.gov.uk/20130107105354/http://www.dh.gov.uk/prod\\_consum\\_dh/groups/dh\\_digitalassets/@dh/@en/documents/digitalasset/dh\\_4035462.pdf](http://webarchive.nationalarchives.gov.uk/20130107105354/http://www.dh.gov.uk/prod_consum_dh/groups/dh_digitalassets/@dh/@en/documents/digitalasset/dh_4035462.pdf). Accessed 19 August 2016.
- Department of Health. (2003) Saving lives, valuing donors: A transplant framework for England. Available from <http://webarchive.nationalarchives.gov.uk/20031220221856/doh.gov.uk/transplantframework/>. Accessed 4 August 2016.



## ETHICS AND COMMUNICATION SKILLS STATION SUMMARY

- Thoroughly prepare for the range of themes commonly encountered in this station, and this will stand you in good stead.
- Remember that your communication skills are being challenged, and inadequate preparation will be evident to the examiners.
- Use the time before entering the station to plan a structure for your discussion and the topics to be addressed. This will help you organise your thoughts and think through the information to be gathered and given to the patient.
- Always bear the four main ethical principles (autonomy, beneficence, non-maleficence and justice) in mind when approaching the task.
- Remember that there are other issues that may be the theme of discussion, such as
  - Confidentiality
  - Driving regulations/DVLA medical guidelines
  - Breaking bad news
  - Relaying a new diagnosis
  - Organ transplantation
  - Cardiac resuscitation orders
  - Suitability for ITU transfer/care
- Allow plenty of time for pauses to give the individual enough time to take the information on board and ask any questions they might have.
- Summarise the key points of discussion as you go along.
- Always ensure that you are listening to the patient, empathise and remain calm at all times.



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# 7

## Station 5: Brief Clinical Encounters

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## HINTS FOR THE BRIEF CLINICAL ENCOUNTERS STATION

- Station 5 is worth many marks in the exam, and it is crucial to dedicate a substantial proportion of revision time to it.
- Carefully read the instructions to candidates before entering the station, and brainstorm the specific questions you need to ask to help get the diagnosis.
- Formulate a structure for your focused clinical assessment.
- On entering the room, look closely for any clues to the diagnosis; 'spot diagnoses' are common in this station.
- Generally, begin with an open question, but quickly become more focused after this. Establish a likely diagnosis (and differential) and then find out about specific complications.
- Examination generally involves looking for specific signs to clarify a diagnosis rather than following a set pattern – remember that the examination should be focused.
- Be sure to examine for evidence of complications/manifestations of the primary disorder.
- When presenting the case, give the likely diagnosis first, but have a differential available in case you are asked for this.
- Always ask about any specific concerns that the patient has, and provide a solution or explanation as appropriate.
- Addressing the patient's welfare and concerns is vital in this station.

## ANKYLOSING SPONDYLITIS

This patient, a 25-year-old man, has been experiencing back pain (including pain at night), stiffness and fatigue. Please ask any relevant questions and proceed as appropriate.

### FOCUSED HISTORY

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1. Duration, timing (often have pain at night) and nature of backache; location of pain
2. Age and gender of patient (onset before 40 years and males affected more than females)
3. Morning stiffness: Duration (improves with exercise and worse with rest)
4. Falls, trauma and injuries to spine
5. Spinal deformity
6. Other joints affected: Sacroiliac joints, hips, knees, ribs
7. Family history of back problems
8. Neurological symptoms: Ensure no bladder/bowel disturbance (all histories of back pain), paraesthesia/numbness/limb weakness
9. Other features: Chest pain, breathlessness, eye symptoms (pain, redness, floaters), enthesitis, fatigue
10. Patient welfare/concerns: Deformity, genetic link, mobility aids



## FOCUSED EXAMINATION

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- Spine
  - Ask the patient to stand up, back and front fully exposed (ensure that you preserve dignity).
  - Kyphotic spine, compensatory hyperextension of the neck ('question mark' posture).
  - Reduced spinal movements: Rigid, immobile spine.
  - Increased anteroposterior (AP) diameter of chest wall.
- Cardiac
  - Listen to aortic area and left sternal edge for early diastolic murmur of aortic regurgitation.
- Chest
  - Fine apical fibrotic crepitations
- Eyes
  - Iritis, visual acuity check
- Gait
  - Likely antalgic, will make the spinal deformity more obvious

## QUESTIONS

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1. What are the immunological associations with ankylosing spondylitis?
  - Seronegative spondyloarthropathy
  - HLA-B27 positive in >90% of individuals
  - Tumour necrosis factor (TNF)- $\alpha$  and interleukin (IL)-1 also implicated in disease activity
2. How is the diagnosis of ankylosing spondylitis made?
  - Mainly clinical from history and examination with supporting radiological evidence
  - Young patients (<40), possible family history
  - Plain radiograph – erosions and fibrosis/sclerosis of the sacroiliac joints, squaring of the vertebra ('bamboo spine') due to ossification of the anterior longitudinal ligament and intervertebral spaces
  - Blood tests: Raised erythrocyte sedimentation rate (ESR)/C-reactive protein (CRP) during active inflammation, normocytic anaemia
  - Genetic testing: HLA-B27
3. What is the treatment for ankylosing spondylitis?
  - No known cure: Mainly symptomatic
  - Physiotherapy: Encourage increased exercise, physiotherapy and exercises for maintaining good posture
  - Analgesia: Nonsteroidal anti-inflammatory drugs (NSAIDs), paracetamol and weak opioids
  - Corticosteroid injections: For sacroiliitis and enthesitis

- Biological agents: TNF- antagonists such as etanercept, adalimumab, infliximab, golimumab and certolizumab pegol (help slow disease progression)
  - Treat complications such as iritis
  - Treat osteoporosis with bisphosphonates to prevent spinal fractures
4. What are the extra-articular manifestations of ankylosing spondylitis?
- Respiratory: Restrictive lung defect/reduced lung capacity due to restricted chest wall movement and apical lung fibrosis.
  - Cardiac: Chronic aortitis leading to aortic regurgitation, conduction defects and cardiomyopathy.
  - Neurological: Atlantoaxial instability/dislocation and cauda equina syndrome.
  - Eyes: Iritis and cataracts.
  - Amyloidosis (secondary/amyloid A [AA]): Multisystem involvement, including hepatic and renal involvement.
  - Coexistent inflammatory bowel disease is common.

## PATIENT WELFARE AND CONCERNS

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1. Adequate management of pain.
2. Concern regarding the impact upon activities of daily living – will this stop the patient working?
3. Cosmetic concerns regarding possible spinal deformity.
4. Is this inherited, and could I pass this onto my children doctor?
5. Is there a cure?

## CANDIDATE EXPECTATIONS

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1. Recognition of an inflammatory arthropathy, and in particular ankylosing spondylitis, from a detailed history
2. Awareness of the need to rule out 'red-flag symptoms'
3. Understanding of the extra-articular manifestations of inflammatory arthropathies
4. Examine for features of ankylosing spondylitis, including extra-articular manifestations, e.g. auscultate for aortic regurgitation and lung fibrosis
5. Explanation that treatment aims are to maintain a good quality of life and prevent disease progression and complications

## REFERENCES

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- National Institute for Health and Care Excellence. (2013) Clinical knowledge summary: Ankylosing spondylitis. Available from <http://cks.nice.org.uk/ankylosing-spondylitis#!scenariorecommendation:4>. Accessed 5 July 2016.
- National Institute for Health and Care Excellence. (2016) TNF-alpha inhibitors for ankylosing spondylitis and non-radiographic axial spondyloarthritis. Available from <https://www.nice.org.uk/guidance/ta383/resources/tnfalpha-inhibitors-for-ankylosing-spondylitis-and-nonradiographic-axial-spondyloarthritis-82602848027077>. Accessed 5 July 2016.



## ANTICOAGULATION

This 65-year-old man has been having palpitations intermittently for the past 3 months. On one occasion, he attended the emergency department and was found to be in atrial fibrillation (AF) with rapid ventricular response. The team in the emergency department has given him some bisoprolol to slow the rate, and he attends your rapid access outpatient department to discuss anticoagulation.

### FOCUSED HISTORY

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1. Start by summarising the history and ensuring that the patient knows that the focus of the consultation will be to discuss anticoagulation.
2. Enquire about the risk factors for stroke, i.e. does this patient need anticoagulation?
3. Is the patient known to have hypertension?
4. Is the patient known to have diabetes?
5. Is the patient known to have heart failure?
6. Need to assess the patient's bleeding risk?
7. Prior history of gastrointestinal (GI) bleeding?
8. Prior history of liver disease?
9. Does the patient have any known renal problems?
10. Prior history of intracranial bleeding?
11. Does the patient have an active cancer?
12. Take an alcohol history.
13. Take a thorough drug history, including over-the-counter and herbal medications.
14. Social history is important. Contact sport should be avoided if the patient is to take anticoagulants.

### FOCUSED EXAMINATION

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- Observe for stigmata of bleeding/chronic liver disease.
- Check the patient's blood pressure.
- Look for clinical signs of cardiac failure.

### QUESTIONS

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1. Do you know of any risk stratification tools that can be used to assess bleeding risk for patients on warfarin?
  - The HASBLED score (see below).
  - This takes into account hypertension, renal and liver failure, age, drug use, alcohol consumption, labile international normalised ratios (INRs) and medication usage that would predispose to bleeding (NSAIDs and antiplatelets).

- A score out of 9 is given and can be used to calculate bleeding risk. If the patient scores 3 or more, caution with anticoagulation is advised and regular review must be in place if a decision is taken to anticoagulate.
2. Do you know of a tool one can use to predict stroke risk for patients with AF?
    - The CHA2DS2-VASc score (see below).
    - This score is out of 9 and takes into account other stroke risk factors, including age, diabetes, hypertension and heart failure.
    - The higher the score, the greater the risk of stroke.
    - Dependent on the score, a percentage stroke risk per year for a patient can be given.
  3. Under what circumstances would one use novel anticoagulants such as rivoroxaban as opposed to warfarin?
    - The National Institute for Health and Care Excellence (NICE) guidelines suggest that novel oral anticoagulants (NOACs) can be used where appropriate. They are guidelines, and use will be clinician dependent and dependent on local guidelines. An example may be if a patient has mild cognitive impairment and has difficulty managing warfarin.
    - Local guidelines may vary.
    - If the patient cannot get to and from hospital for blood tests.
    - If they have had a cardiovascular event while on warfarin with a subtherapeutic INR.
    - If they have a labile INR.
  4. List some disadvantages of novel anticoagulants?
    - If the patient misses one dose, then they are not receiving the benefit; therefore, likely compliance must be assessed and the importance of this stressed.
    - Not all are safe in renal failure, so this may need to be monitored if there are concerns. Dose will vary depending on estimated glomerular filtration rate (eGFR); consult the British National Formulary (BNF).

## PATIENT WELFARE AND CONCERNS

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1. Clear and concise explanation of the rationale behind the use of anticoagulation, i.e. risk versus benefit and, in the case of anticoagulation, why the benefit outweighs the risk
2. Need to counsel the patient on the risks of anticoagulation (drug–food interactions/noncontact sports/regular blood tests/dentists, etc.)
3. Appreciate that the risk may be unacceptable to a patient who has capacity.

## CANDIDATE EXPECTATIONS

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1. Ensure that you ask questions that will demonstrate that you are assessing the risk of stroke and the risk of bleeding.
2. Ensure that you thoroughly explain the risks surrounding anticoagulation, but remind the patient that these risks are outweighed by the benefits in stroke risk reduction.
3. Show an awareness of the patient's autonomy.



## HASBLED

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- H** – Hypertension uncontrolled (systolic BP <160)  
**A** – Abnormal renal function: Dialysis, transplant, Cr >2.26 mg/dL or >200 µmol/L  
 Abnormal liver function  
**S** – Stroke: Prior history of stroke  
**B** – Bleeding: Prior major bleeding or predisposition to bleeding  
**L** – Labile INR: (Unstable/high INRs), time in therapeutic range <60%  
**E** – Elderly: Age >65 years  
**D** – Prior alcohol or drug usage history (≥8 drinks/week)  
 Medication usage predisposing to bleeding: (Antiplatelet agents, NSAIDs)  
 Each point scores 1

## CHADSVASC

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- C** – Congestive heart failure (or left ventricular systolic dysfunction) scores 1  
**H** – Hypertension: Blood pressure consistently above 140/90 mmHg (or treated hypertension on medication) scores 1  
**A** – Age ≥75 years scores 2  
**D** – Diabetes mellitus scores 1  
**S** – Prior stroke or transient ischaemic attack (TIA) or thromboembolism scores 2  
**V** – Vascular disease (e.g. peripheral artery disease, myocardial infarction, aortic plaque) scores 1  
**A** – Age 65–74 years scores 1  
**Sex Category** – Females score 1

## REFERENCES

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- Lip GY, et al (2010). Refining clinical risk stratification for predicting stroke and thromboembolism in atrial fibrillation using a novel risk factor-based approach: The Euro Heart Survey on atrial fibrillation. *Chest* 137(2): 263–72.
- National Institute for Health and Care Excellence. (2014) Guidance CG180. Preventing stroke in people with atrial fibrillation. Available from <http://pathways.nice.org.uk/pathways/atrial-fibrillation#path=view%3A/pathways/atrial-fibrillation/preventing-stroke-in-people-with-atrial-fibrillation.xml&content=view-node%3Anodes-anticoagulation-treatment>. Accessed 24 August 2016.
- Pisters R, et al. (2010) A novel user-friendly score (HAS-BLED) to assess 1-year risk of major bleeding in patients with atrial fibrillation. *Chest* 138 (5): 1093–100.

## DIABETIC RETINOPATHY

This patient has noticed his vision has become blurred at times. Please ask any relevant questions and proceed as appropriate.

## FOCUSED HISTORY

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1. Duration of symptoms: Has there been a progressive change rather than sudden onset?
2. Has there been any visual loss?
3. Assess diabetic control.

4. Other microvascular complications: Nephropathy, neuropathy, autonomic dysfunction.
5. Macrovascular complications: Peripheral vascular disease, ischaemic heart disease, stroke.

## FOCUSED EXAMINATION

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- Eyes
  - Glasses
  - Visual acuity (ideally using a Snellen chart)
  - Pupillary reactions
  - Eye movements
- Fundoscopy
  - Cataracts, features of diabetic retinopathy (nonproliferative, proliferative, maculopathy, photocoagulation scars)
- Extras
  - Evidence of neuropathy
  - Ask for blood glucose measurement, urine dipstick for proteinuria and blood pressure

## QUESTIONS

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1. What are the features of nonproliferative diabetic retinopathy?
  - Microaneurysms (dot haemorrhages)
  - Blot haemorrhages
  - Hard exudates
  - Soft exudates (cotton wool spots)
2. What are the features of proliferative diabetic retinopathy?
  - Features of nonproliferative diabetic retinopathy, with evidence of new vessel formation.
  - Photocoagulation scars are evidence of treatment.
3. What are the features of diabetic maculopathy?
  - Any features of diabetic retinopathy at or near the macula.
  - Most commonly, there is circinate formation of hard exudates.
4. What effect does glycaemic control have on the risk of diabetic retinopathy?
  - This was assessed by the UK Prospective Diabetes Study (UKPDS) and the Diabetes Control and Complications Trial (DCCT).
  - These studies showed that better glycaemic control lowers the risk of retinopathy. For example, a 10% reduction in HbA1c is associated with a more than 40% reduction in the development of retinopathy.
5. What else can be done to lower the risk of retinopathy in type 2 diabetes?
  - Optimal treatment of hypertension





6. How is peripheral (nonmacular) diabetic retinopathy managed?
  - Background retinopathy does not need treatment but should be monitored.
  - If preproliferative retinopathy is severe, laser treatment may be considered.
  - Proliferative retinopathy requires laser treatment.
  - Severe proliferative retinopathy may not respond to laser treatment and may require vitrectomy.

## PATIENT WELFARE AND CONCERNS

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1. Vision: Concerns of visual loss?
2. Prevention: Prevention of further deterioration?
3. Treatment: Is treatment possible?

## CANDIDATE EXPECTATIONS

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1. Recognition that diabetic retinopathy can be asymptomatic or can present with visual disturbance
2. Recognition of the features of diabetic retinopathy
3. Understanding of the stages of diabetic retinopathy
4. Formulate the correct diagnoses and convey this to the patient
5. Generate a management plan, recognising when urgent intervention is necessary to save vision

## REFERENCES

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- Diabetes Control and Complications Trial Research Group. (1995) The relationship of glycemic exposure (HbA1c) to the risk of development and progression of retinopathy in the Diabetes Control and Complications Trial. *Diabetes* 44(8): 968–83.
- Royal College of Ophthalmologists. (2012) Diabetic retinopathy guidelines. Available from <https://www.rcophth.ac.uk/standards-publications-research/clinical-guidelines/>. Accessed 5 July 2016.
- Stratton IM, et al. (2001) UKPDS 50: Risk factors for incidence and progression of retinopathy in type II diabetes over 6 years from diagnosis. *Diabetologia* 44(2): 156–63.
- UK Prospective Diabetes Study Group. (1998a) Tight blood pressure control and risk of macrovascular and microvascular complications in type 2 diabetes: UKPDS 38. *BMJ*; 317(7160): 703–13.
- UK Prospective Diabetes Study Group. (1998b) Efficacy of atenolol and captopril in reducing risk of macrovascular and microvascular complications in type 2 diabetes: UKPDS 39. *BMJ* 317(7160): 713–20.

## FACIAL NERVE PALSY

This patient has been experiencing difficulty with closing their eyelids on one side. As a result, the eye is becoming dry and irritable. They are anxious to know the cause of the symptoms and what can be done. Please ask any relevant questions and proceed as appropriate.

## FOCUSED HISTORY

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1. Which eyelid is affected? What appears to be the problem? Does the eyelid shut at all, and if so, how much?
2. Duration of symptoms?

3. Are there any associated problems: speech, swallowing, taste or hearing impairment (tinnitus,\* hyperacusis or deafness\*)?
4. Any vertigo\*/nausea\*/vomiting or abnormality in gait\* noted?
5. Any associated facial droop or distortion of the angle of mouth, change in taste?
6. Any recent viral (herpes) infection, surgery to the neck (parotid gland), inner ear or mastoid?
7. Medical history/family history: Neurofibromatosis ± acoustic neuroma with or without previous surgery.

\*Be alert to these symptoms, as they may be indicative of acoustic neuroma.

## FOCUSED EXAMINATION

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- General
  - Examine the patient at rest to assess facial symmetry.
  - Any obvious drooping of the angle of the mouth, ptosis (unilateral). Ask patient to close their eyes. Assess ability to close fully. Is there a tarsorrhaphy scar?
- Movements
  - Assess muscles supplied by facial nerve (demonstrate the movements yourself).
  - Ask patient to squeeze their eyes tightly shut, raise their eyebrows, blow out their cheeks, smile and show you their teeth.
- Extras
  - Look behind the ear for a mastoid surgery scar. Examine for any cranial scars from acoustic neuroma removal.
  - Assess for any hearing disturbance (cranial nerve [CN] involvement).
  - Assess gait (cerebellar/ataxic).

## QUESTIONS

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1. What are the causes of unilateral CN VII palsy?
  - Upper motor neurone (UMN) lesion
    - Cerebellopontine angle lesion (CNs V, VI, VII, VIII and loss of taste on anterior two-thirds of the tongue): Acoustic neuroma, meningioma
    - Pontine lesion: Demyelination (multiple sclerosis [MS]), vascular lesion
  - Lower motor neurone (LMN) lesion
    - Bell's palsy: Most common cause; caused by herpes simplex type 1
    - Ramsay Hunt's syndrome: Caused by herpes zoster virus
    - Parotid gland tumour/surgery
    - Facial neuroma
    - Cholesteatoma
    - Mononeuritis multiplex (diabetes, systemic lupus erythematosus [SLE], polyarteritis nodosa [PAN], sarcoid, amyloid, Wegener's granulomatosis): Remember WARDS PLC
    - Trauma
2. What are the causes of bilateral LMN cranial nerve VII palsy?
  - Motor neurone disease (MND).
  - Guillain-Barré syndrome.
  - Bilateral Bell's palsy.



- Lyme disease.
  - Myasthenia gravis.
  - Sarcoidosis.
  - Moebius syndrome (inherited rare form due to underdevelopment of cranial nerves VI and VII).
  - Note that this question is another Practical Assessment of Clinical Examination Skills (PACES) 'classic'.
3. Which condition is associated with an acoustic neuroma?
    - Neurofibromatosis type 2: Defect on chromosome 22q12. This condition results in bilateral acoustic neuromas.
  4. What is the management of an acoustic neuroma?
    - Conservative: Observe the tumour size and growth.
    - Radiotherapy: To retard tumour growth.
    - Surgical resection.

## PATIENT WELFARE AND CONCERNS

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1. Aesthetic: Facial asymmetry secondary to the nerve palsy due to medical/surgical cause.
2. Will the facial nerve palsy resolve?
3. Is there any treatment for this?
4. Will the deafness resolve or will it be permanent? Is there any treatment for this?

## CANDIDATE EXPECTATIONS

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1. Obtain a thorough history of symptoms of a CN VII lesion.
2. Be aware of the range of causes leading to CN VII palsy.
3. Examine carefully to differentiate between UMN and LMN CN VII palsy.
4. Provide the patient with a clear explanation of the problem and the likely effects; e.g. if it is a Bell's palsy, there is a good chance of recovery. However, if it is a surgical consequence, then that is unlikely to be the case.

## FALLS

This 80-year-old male has been falling regularly at home. He has limited medical history (he has hypertension and diabetes mellitus, managed by his general practitioner [GP]) and hasn't ever been to hospital with the exception of having his appendix removed when he was 28. Please ask any relevant questions and proceed as you feel appropriate.

## FOCUSED HISTORY

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1. Need to establish exactly what occurs when the patient falls over. Does it sound like syncope or presyncope? Do they lose consciousness? If so, how quickly do they recover? (Need to be sure to rule out seizures.)

2. Ask about injuries postfall? Head injuries tend to indicate a loss of consciousness.
3. What environment are they in when they fall?
4. What had they been doing before each fall?
5. What are the precedent and antecedent symptoms?
6. How often do they fall?
7. Important: Past medical history/drug history will lead to many clues around the fall. For example, in this case the patient may be overtreated with antihypertensives and have postural hypotension or may have a sensory neuropathy from their poorly controlled diabetes.
8. Ask about their health in general. How far do they walk normally? (If they have severe osteoarthritis [OA], they are likely to have a degree of disuse myopathy and fall because of this.) What is their vision like? When was it last checked?
9. Ask about continence? Are they falling because they are rushing to the toilet because they have overactive bladder syndrome that is undiagnosed?
10. Make an enquiry about their social circumstances.

## FOCUSED EXAMINATION

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- General
  - Comment on all/any injuries/bruises.
  - Ask the patient to walk to assess gait (Parkinsonian/Marche à petits pas in vascular dementia).
  - If possible, use your fundoscope to check for cataracts/look for signs of diabetic/hypertensive retinopathy.
  - Essential: Check postural blood pressure.
  - Perform (or at least ask for) a finger-prick blood glucose check (note that diabetes can lead to both iatrogenic hypoglycaemia and autonomic neuropathy, which may result in falls).
  - Look for proximal myopathy – common in older persons and often overlooked as a cause of falls.
- Cardiac
  - Check pulse for arrhythmias.
  - Listen for the murmur of aortic stenosis (syncope can be a symptom).
- Neurological
  - Check for focal weakness/cerebellar signs.
  - Examine for peripheral neuropathy that may be affecting sensory feedback.

## QUESTIONS

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1. How do you perform a lying and standing blood pressure?
  - Lay patient down for 15 minutes and then take blood pressure.
  - Stand patient.
  - Take blood pressure at 1 minute.
  - Take blood pressure at 3 minutes.



2. What constitutes a positive test?
  - Systolic drop of 20 mmHg
  - Diastolic drop of 10 mmHg
  - Symptomatic patient (presyncopal)
3. What is the cause of this patient's fall?
  - Falls are almost always multifactorial, particularly if there is no syncope; e.g. in this case, he could be falling because he has dyspraxia secondary to vascular cognitive changes, a proximal myositis because of the statin his GP has given him for primary prevention and possibly a postural drop and sensory changes associated with his diabetes mellitus (autonomic neuropathy).
  - You may not be expected to get the full diagnosis (it may be beyond the scope of the station), but it is important that you demonstrate a holistic approach to the case.
4. What is the best approach to managing a patient with falls?
  - Multidisciplinary team (MDT): The patient will need assessment by an occupational therapist (OT), physiotherapist, optometrist, pharmacist, nurse and geriatrician. A comprehensive geriatric assessment is often needed, and this will be performed by all of the above (and more). Of note, there is evidence that balance training and tai chi reduce the frequency of falls.

## PATIENT WELFARE AND CONCERNS

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1. Is the patient's home environment safe? They may well benefit from a home visit by the OT to review their environment.
2. Consider if it is safe for the patient to drive if syncope is the cause (see ethics case on driving regulations in Station 4).

## CANDIDATE EXPECTATIONS

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1. Being holistic is most important.
2. You are unlikely to cover every aspect, but you should demonstrate that you are thinking about multiple potential causes.
3. You need to demonstrate an awareness of common problems that older persons who fall face.
4. Aim to be proactive when presenting the case. If they are falling regularly and at risk of osteoporosis, have you considered bone protection?

## REFERENCE

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Gillespie LD, et al. (2009) Interventions for preventing falls in older people living in the community. *Cochrane Database Syst Rev* 15(2): CD007146.

## HEADACHE (IDIOPATHIC INTRACRANIAL HYPERTENSION)

This 30-year-old lady with a family history of diabetes mellitus had a glucose tolerance test 6 months ago and was found to have impaired fasting glycaemia. Since then, she has been complaining of headaches and visual disturbance, getting much worse in the last 2 weeks. She does not usually wear glasses and wants to know what can be done to improve the symptoms.

### FOCUSED HISTORY

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1. Nature of visual disturbance: Diplopia or blurred vision, floaters/flashing lights, visual loss
2. Duration of visual disturbance: Stable or deteriorating
3. Effect on visual acuity: Any recent eye tests
4. Headaches: Worse in morning, on bending/coughing/sneezing/vomiting, nausea
5. Associated symptoms: Paraesthesia, weakness, hearing disturbance
6. Relevant other history: Weight loss/gain, obstructive sleep apnoea, drug history (vitamin A derivatives, tetracyclines, oral contraceptive pill)

### FOCUSED EXAMINATION

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- General
  - Obesity, any obvious cranial nerve palsies
- Eyes
  - Enlarged blind spot, ophthalmoplegia (abducens nerve palsy)
  - Fundoscopy
  - Assess for papilloedema and optic atrophy
- Extras
  - Check for scars from ventriculoperitoneal (VP) shunts. Ask for blood glucose measurement and blood pressure.

### QUESTIONS

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1. What are the causes of papilloedema?
  - Space-occupying lesion (tumour/abscess): Idiopathic intracranial hypertension (IIH)
  - Hypertensive encephalopathy
  - Infection: Encephalitis, meningitis
  - Vascular: Intra-/extra-axial haemorrhage, venous sinus thrombosis
  - Drugs: Tetracyclines, vitamin A derivatives
2. What are the fundoscopic features of papilloedema?
  - Disc hyperaemia, blurred margins, absent venous pulsation
  - Elevation of the disc with obscured vessels at the disc margin

- Loss of the cup with obscured vessels in the disc
  - Bulging disc with all vessels obscured (this will eventually lead to optic atrophy)
3. What are the causes of optic atrophy?
- Congenital
    - Friedreich's ataxia, Leber's hereditary optic neuropathy
  - Acquired
    - Vascular: Ischaemic (including temporal arteritis)
    - Inflammatory: Multiple sclerosis, Devic's disease
    - Compression: Optic nerve tumour, Graves' ophthalmopathy, glaucoma
    - IIH (untreated)
    - Nutritional deficiencies: Vitamin B12, folate
    - Toxins: Tobacco, alcohol, ethambutol, ethylene glycol, lead, cyanide, carbon monoxide
    - Infective: Syphilis
4. What treatments can be used to treat IIH?
- Medical
    - Weight loss if obese
    - Stop any contributing medications
    - Diuretics, e.g. acetazolamide
    - Steroids: Beneficial in inflammatory conditions/causes
    - Repeated lumbar punctures
  - Surgical
    - Optic nerve sheath decompression and fenestration is performed if vision is severely affected or threatened.
    - Lumboperitoneal/ventriculoperitoneal shunt.

## PATIENT WELFARE AND CONCERNS

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1. Concerns regarding the visual disturbance: Could I lose my sight?
2. Concerns regarding the associated headache: Is this a brain tumour?
3. How can you be certain that this is a benign condition and that there is no sinister underlying pathology?
4. What are the treatment options? Would having a shunt inserted require brain surgery?

## CANDIDATE EXPECTATIONS

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1. Obtain a thorough history regarding the visual disturbances and headache.
2. Identify features suggestive of raised intracranial pressure and/or a space-occupying lesion, e.g. weakness/paralysis, seizures.
3. Carefully assess visual acuity, visual fields and ocular fundi.
4. Explain the likely diagnosis of IIH, but make the patient aware of the need to exclude a space-occupying lesion.
5. Explain that treatment strategies involve both medical and surgical options.

## HYPERTHYROIDISM

This patient has a tremor and weight loss. Please ask any relevant questions and proceed as appropriate.

### FOCUSED HISTORY

1. Is there any history of weight loss despite normal or increased appetite (caveat – occasionally some gain weight)?
2. Does the patient suffer from heat intolerance?
3. Do they sweat excessively?
4. Is there any history of GI upset, specifically diarrhoea?
5. Do they suffer from tachycardia or palpitations?
6. If they are female, ask about their menstrual cycle and specifically about oligomenorrhoea?
7. Do they suffer from anxiety and/or irritability?
8. Do they have a goitrous thyroid swelling?
9. Eye symptoms: Excessive watering, grittiness, redness, puffiness, change in field of vision? Eye pain (exophthalmos)? Ensure that you ask about all these even if no obvious eye signs are seen.
10. Ask the patient if they are on any medication: Beta-blockers – for anxiety symptoms and tachycardia; carbimazole/propylthiouracil – to block production of T4 and T3.

### FOCUSED EXAMINATION

- Initial impression: Wide staring anxious expression – possible proptosis. Anxious fidgety demeanour, a peripheral tremor and goitre point towards a diagnosis of hyperthyroidism.
- Hands and arms: Acropachy (clubbing – rarely found and must have Graves' ophthalmopathy), sweaty palms, tachycardia/arrhythmia, fine tremor, proximal myopathy.
- Eyes: Observe from the side and above for degree of proptosis. Observe from the front for lid retraction, oedema and tearing, and perform eye movements for lid lag and ophthalmoplegia.
- Eye signs from any cause of thyrotoxicosis: Lid retraction (see the white of the eye above and below the iris), lid lag.
- Graves' ophthalmopathy: Reddened eyes, excessive lacrimation, periorbital oedema, proptosis, conjunctival oedema and ophthalmoplegia
- Neck: Thyroidectomy scar, observe goitre while swallowing water, palpate goitre (diffuse or nodular) and palpate while swallowing water, palpate nodes, percuss the retrosternal extent of the thyroid. Auscultation for thyroid bruit.
- Legs: Pretibial myxoedema (rarely found and must have Graves' ophthalmopathy).
- Extras: Evidence of other autoimmune disease, especially vitiligo, as it is common and easily observed in PACES.





## QUESTIONS

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1. What are the causes of hyperthyroidism?
  - Graves' disease
  - Toxic multinodular goitre
  - Thyroid adenoma (toxic)
  - De Quervain's thyroiditis (painful, fever at onset)
  - Postpartum thyroiditis
  - Drugs (amiodarone, rarely lithium)
2. What are the causes of a goitre?
  - Nodular
    - Multinodular goitre (especially iodine-deficient areas)
    - Adenoma
    - Carcinoma
  - Diffuse
    - 'Simple'
    - Graves' disease
    - Hashimoto's thyroiditis
    - De Quervain's thyroiditis
    - Thyroid lymphoma
3. What signs of thyroid disease are specific to Graves' disease?
  - Graves' ophthalmopathy – See above
  - Thyroid acropachy
  - Pretibial myxedema
4. What are the 'hyperthyroid emergencies'?
  - Thyroid storm
  - Exophthalmos causing fixed gaze, diplopia or decreased acuity or loss of colour vision (may lead to optic nerve compression)
  - Cardiac failure (high output)

## PATIENT WELFARE AND CONCERNS

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- What treatment options are available?
- Is my condition curable?
- Is my condition life threatening?

## CANDIDATE EXPECTATIONS

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- Recognise early that the patient has thyroid disease.
- Assess the patient's thyroid status (hyper-/hypo-/euthyroid).
- Recognise signs specific to Graves' disease.
- Be able to discuss treatment options with the patient and address their concerns.

## NECK LUMP

This patient presents complaining of a neck lump. Please ask any relevant questions and proceed as you feel appropriate.

### FOCUSED HISTORY

---

1. Ask the patient what their most concerning problem is.
2. Where in the neck is the lump? (Thyroid: Midline, and just above the clavicle; other lumps may be nodes; make sure you can describe the surface anatomy.)
3. How long has it been there? (Rapidly progressing lumps can be lymphoma or anaplastic thyroid carcinoma.)
4. Is the lump painful?
5. Does the patient suffer from dysphagia or dysphonia (hoarseness)?
6. Are there any other symptoms of thyroid disease?
  - Over-/underactive
  - Weight loss/gain, appetite
  - Intolerance to heat/cold
  - Mood instability, irritability
  - Bowels: Diarrhoea/constipation
  - Palpitations
7. Is there any family history of thyroid disease or malignancy?
8. Has there been any history of radiation exposure?
9. Is there any history of autoimmune disease? Type 1 diabetes mellitus, rheumatoid arthritis (RA), adrenal insufficiency, vitiligo, coeliac disease, pernicious anaemia, myasthenia gravis and multiple sclerosis

### FOCUSED EXAMINATION

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- General
  - Evidence of hyperthyroid or euthyroid status, rarely hypothyroid
  - Then proceed as per the hyperthyroidism case (hands, eyes, neck, legs)
- Additional extras
  - Evidence of other autoimmune disease, especially vitiligo, as it is common and easily observed in PACES

### QUESTIONS

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1. What are the types of thyroid cancer?
  - Papillary: Most common
  - Follicular
  - Medullary: Arise from parafollicular calcitonin-secreting C-cells
  - Anaplastic: Most aggressive
  - Lymphoma
  - Metastases from other primaries (kidney most common)



2. What investigations are used to diagnose the cause of a thyroid mass?
  - TSH, T3 and T4 (rarely malignant if thyroid dysfunction)
  - Gold standard is ultrasound-guided fine needle aspiration (FNA)
  - CT useful to show the extent of a goitre causing compressive symptoms to trachea, or staging for malignancy

## PATIENT WELFARE AND CONCERNS

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1. What is the cause? Is this a cancerous lump, and how will this be differentiated from something that is noncancerous?
2. Cosmetic: Address concerns about how this lump sticks out and if it will be fully treated. Will it need surgery?
3. Associated features: Hoarse voice, swallowing difficulties and symptomatic thyroid disease. Are these all treatable?

## CANDIDATE EXPECTATIONS

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1. Look out for any red-flag symptoms of malignancy, such as weight loss and strong family history. Poor prognostic indicators for thyroid malignancy include: rapid enlargement, hoarse voice, age, sex (male) and type of cancer (papillary carcinoma has the best outcome, anaplastic has the worst).
2. Thoroughly assess a thyroid mass and examine for any associated features of thyroid disease.
3. Aim to reach a diagnosis and convey this to the patient, along with the management plan that will follow. Address each of the different investigations and what these will help to elicit.
4. Address the patient's concerns, ranging from the concern that they may think that it's cancer to any cosmetic concerns. Reassure the patient that thyroid malignancy is just one of many causes of a thyroid mass, and is not the most common cause. However, you need to rule this out if there is a nodule.

## OSTEOPOROSIS

This 82-year-old female has recently been diagnosed with polymyalgia rheumatica and started on prednisolone tablets. She has a past history of chronic lower back pain for which she takes several painkillers. She has presented today with worsening backache and wants to know what more can be done about it. Of note, she is known to be diabetic.

## FOCUSED HISTORY

---

1. Is the diabetes a red herring, or is it pertinent to the history?
2. Duration of current episode of back pain: Onset (acute/sudden or gradual worsening), severity (pain score), character, site and radiation. Has the pain ever been this severe in the past? Analgesic history.

3. Precipitating factors: Heavy lifting/fall/trauma.
4. Neurological symptoms: Paraesthesia/weakness or paralysis in lower limbs, bladder or bowel dysfunction.
5. Red-flag symptoms: Weight loss, nocturnal pain and fevers.
6. Relevant other history: Risk factors for osteoporosis (steroid use, immobility, family history), hypertension.

## FOCUSED EXAMINATION

---

- General
  - Examine patient standing: Obvious spinal deformity, skin/palmar pigmentation if pigmented appearance, central obesity, round face, thin skin, bruising
- Spine
  - Palpate vertebrae to localise symptoms; assess spinal movements as far as able.
- Limbs
  - Focused assessment of power in lower limbs, sensation and gait
- Extras
  - State you would examine perineum for perianal sensation and anal sphincter tone. Ask for blood glucose measurement and blood pressure reading.

## QUESTIONS

---

1. Assuming her back pain is caused by an osteoporotic fracture, should the patient in this case have medical management of osteoporosis?
  - Patients with fragility fractures over the age of 75 should have treatment regardless of dual-energy X-ray absorptiometry (DEXA) results. Review the NICE guidance on this.
2. What are the risk factors for developing osteoporosis?
  - Family history
  - Disease associations: Cushing's syndrome, malabsorption, hyperparathyroidism, chronic inflammatory arthropathy
  - Toxins: Excess alcohol or caffeine intake, smoking
  - Prolonged immobility or inactivity
  - Underweight
  - Early menopause, late menarche, postmenopause, bilateral oophorectomy, hypogonadism in males
  - Drugs: Prolonged steroid use (>7.5 mg prednisolone daily for 6 months), prolonged use of low-molecular-weight heparin
3. What strategies can be used in the treatment of osteoporosis?
  - Lifestyle changes: Nutrition, exercise
  - Prevent falls: Appropriate footwear, OT/physio assessments (home adjustments), avoid sedative medications
  - Drug therapies
    - Reduce bone resorption: Bisphosphonates (zoledronic acid, ibandronic acid, alendronic acid), raloxifene (selective oestrogen reuptake inhibitor), calcitonin



- Aid new bone formation: Calcium and vitamin D supplements
- Reduce bone resorption and increase new bone formation: Strontium ranelate, teriparatide (recombinant parathyroid hormone [PTH]), denosumab (receptor activator of nuclear factor kappa-B ligand [RANKL] inhibitor)

## PATIENT WELFARE AND CONCERNS

---

1. Concerns regarding the chronic back pain and the need for a diagnosis.
2. The impact of the back pain on daily life and ability to work.
3. Is there a treatment, medical or surgical, other than simple analgesia, for the back pain/osteoporosis?

## CANDIDATE EXPECTATIONS

---

1. Obtain a thorough history of the back pain and quickly rule out 'red-flag signs'.
2. Rule out spinal cord compression in all patients presenting with worsening back pain.
3. Search for causes of osteoporosis if the patient has had an osteoporotic compression fracture leading to the back pain (in this case, possible Cushing's).
4. Thoroughly examine any deformity of the spine and the range of spinal movements. Check for any neurological deficit and offer to check for perianal sensation and tone.
5. Explain the likely diagnosis.
6. Explain that bisphosphonates are first-line agents in the management of osteoporosis, for both treatment and prevention of further fracture.

## REFERENCE

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National Institute for Health and Care Excellence. (2014). Guidance CG146. Osteoporosis: Assessing the risk of fragility fracture. Available from <https://www.nice.org.uk/guidance/cg146>. Accessed 24 August 2016.

## PROXIMAL MYOPATHY

This lady has been recently seen by her doctor for generalised aches and pains present for several months. She is now complaining of weakness in her limbs. She is housebound due to severe osteoarthritis and often does not go out for weeks at a time. Please ask any relevant questions and proceed as you feel appropriate.

## FOCUSED HISTORY

---

1. What is the nature of the weakness? Is it symmetrical, and which limbs are affected? Is it proximal/distal/widespread?
2. How long has the weakness been present? How rapidly has it progressed?

3. Has there been any associated muscle wasting? Are the muscles tender?
4. Ask what impact the symptoms have on daily life to help decipher the pattern of weakness.  
Proximal myopathy: Difficulty getting up out of chairs, washing and dressing, combing hair  
Distal weakness: Difficulties with fine movements/tasks (writing, doing up buttons)
5. Which joints are involved? Any symptoms suggestive of synovitis?
6. Drug history: Prolonged steroid use, statin therapy, vitamin D supplement.
7. Sinister symptoms: Weight loss, fevers, speech/swallowing impairment, bladder/bowel dysfunction.

## FOCUSED EXAMINATION

---

- General
  - Gait should be normal.
  - There should not be any evidence of fasciculation on inspection.
  - Observe any areas of muscle wasting/disuse atrophy.
  - Evidence of trauma/deformity.
- Palpation
  - Check that the muscles are not tender to palpation.
  - Assess muscle power throughout each limb. Pay close attention for proximal muscle weakness; power will be reduced in affected areas, and the signs will be symmetrical.
  - If a chair is available, ask the patient to sit on it and get up off it without using their hands.
- Extras
  - Examine the spine for any obvious deformity.
  - Examine the hands: Rule out dermatomyositis, rheumatoid disease.
- The differential diagnosis here is wide. A detailed drug history is crucial to eliminate a drug cause, for example, statin-induced myopathy. In a housebound individual who has little sunlight exposure, never forget vitamin D deficiency as a cause of proximal myopathy.

## QUESTIONS

---

1. What are the manifestations of vitamin D deficiency?
  - Rickets: Impaired growth and deformity of long bones in children
  - Osteomalacia: Reduced bone mineralisation resulting in proximal muscle weakness
  - Osteoporosis: Reduced bone density and increased fracture risk
2. What are the risk factors for vitamin D deficiency?
  - Nutritional/poor dietary intake
  - Malabsorption syndromes
  - Reduced sunlight exposure
  - Darker skin pigmentation



- Lack of vitamin D in breast milk
  - Renal/liver impairment
  - Enzyme-inducing medication (e.g. anticonvulsant medications)
3. What are the biochemical features of vitamin D deficiency?
- Reduced serum vitamin D levels
  - Reduced serum calcium levels
  - Reduced serum phosphate levels
  - Raised alkaline phosphatase (ALP)
  - Raised PTH (secondary hyperparathyroidism)

## PATIENT WELFARE AND CONCERNS

---

1. What is the cause of the weakness? Is this reversible?
2. Will they lose their independent mobility?
3. Seriousness of the pathology: Is there an underlying sinister cause?
4. How long will the symptoms take to recover following initiation of treatment?

## CANDIDATE EXPECTATIONS

---

1. Be aware of the range of differential diagnoses contributing to proximal muscle weakness.
2. The key in this case is to take a focused history around the causes of a proximal muscle weakness to help rule in/out a diagnosis.
3. Examine the important areas of each system that help confirm the diagnosis.
4. Formulate the correct diagnosis and convey this to the patient.
5. Reassure the patient that there are many causes of a proximal myopathy, and most of them are readily treatable; however, the main part of the management will be rehabilitation.

## PSORIASIS

This patient has attended complaining of a significant change in the appearance of her nails. She is becoming increasingly distressed by the problem, which seems to be worsening despite several treatments by the GP. She has also noticed pain and swelling in her hands, which is impacting on her job as a seamstress.

## FOCUSED HISTORY

---

1. Duration of symptoms
2. Nails: Colour, change in appearance, brittleness, extent of the nail involved, nail-bed involvement, number of nails involved, history of trauma, treatments tried by GP
3. Skin changes: Plaques/scales, pustules, sites of skin involvement, itchy/dry skin

4. Joints: Sites of arthritis (hands, feet, spine, hips), swelling, tenderness, deformity
5. Impact of illness on lifestyle
6. Family history of psoriasis/arthritis

## FOCUSED EXAMINATION

---

- Hands
  - Nail changes: Discolouration, pitting, onycholysis, hyperkeratosis of nail bed
- Joints
  - Polyarthritis, distal interphalangeal joint arthritis, arthritis mutilans
- Skin
  - Inspect the back, abdomen, extensor and flexor surfaces, scalp and posterior aspect of ears
  - Plaque psoriasis: Shiny, scaly, white plaques
  - Guttate: Numerous widespread, erythematous small 'teardrop' lesions
  - Pustular: Widespread red pustules (often tender)
  - Flexural: Plaques/areas of inflammation at limited sites
- Other joints
  - Spine and sacroiliac joint involvement
  - Asymmetrical oligoarthritis (examine joints highlighted in history)

## QUESTIONS

---

1. What are the causes of psoriasis?
  - Possibly immune-mediated via T-cell-stimulated cytokine release causing excess proliferation and production of skin cells within the dermis
  - Genetic susceptibility: Major histocompatibility complex (MHC) antigen mediated
  - Poststreptococcal infection (especially guttate form)
  - Drug related
2. What treatments are used in the management of psoriasis?
  - Moisturising creams
  - Topical treatments: Coal tar, dithranol, steroids (usually short term), vitamin D analogues (calcipotriol) and calcineurin inhibitors (e.g. tacrolimus cream)
  - Phototherapy: ultraviolet B (UVB)
  - Photochemotherapy: Psoralen and ultraviolet A (PUVA)
  - Medications
    - Oral steroids
    - Disease-modifying antirheumatoid drug (DMARDs): Methotrexate and ciclosporin are most commonly used
    - Vitamin A analogue: Acitretin
    - Biological agents: Anti-TNF therapy, e.g. adalimumab, and monoclonal antibodies, e.g. infliximab





3. How is psoriatic arthritis managed?
  - In patients with psoriatic arthritis who also have severe skin/nail disease, collaboration should take place between rheumatology and dermatology, as systemic therapy may address both areas.
  - The following guidance is targeted at predominantly peripheral arthritis (axial disease is managed more similarly to ankylosing spondylitis):
    - NSAIDs
    - DMARDs: Methotrexate, sulfasalazine, leflunomide, ciclosporin
    - Biologics: Infliximab, etanercept, adalimumab, golimumab

## PATIENT WELFARE AND CONCERNS

---

1. Cosmetic/aesthetic concerns regarding plaques, nail changes and joint involvement
2. Impact of arthropathy on job
3. Curative or symptomatic control only
4. Duration of treatment (particularly immunosuppression) – short course versus lifelong
5. Number of medications needed to control/abate symptoms
6. Long-term effects and risks of newer biological agents – susceptibility to infection, risk of malignancy

## CANDIDATE EXPECTATIONS

---

1. Take a relevant history regarding the nails and skin lesions.
2. Obtain a detailed history for arthritis.
3. Be able to differentiate rapidly from the history whether this is likely to be an inflammatory or degenerative joint problem.
4. Examine the psoriatic plaques appropriately commenting on colour, location, size of area and distribution affected, using correct terminology.
5. Explain the diagnosis to the patient of psoriasis and the possible association between psoriasis and arthropathy.
6. Explain there are both local and systemic treatments that can be used to control symptoms.

## REFERENCES

---

- Coates LC, et al. (2013) The 2012 BSR and BHPR guideline for the treatment of psoriatic arthritis with biologics. *Rheumatology (Oxford)* 52(10): 1754–7.
- DermNetNZ. (2015) Treatment of psoriasis. Available from <http://www.dermnetnz.org/scaly/psoriasis-treatment.html>. Accessed 5 July 2016.
- National Institute for Health and Care Excellence. (2012) Guidance CG153. Psoriasis: Assessment and management. Available from <https://www.nice.org.uk/guidance/Cg153>. Accessed 5 July 2016.

## RHEUMATOID ARTHRITIS WITH CARPAL TUNNEL SYNDROME

This patient has been complaining of a recent history of dropping objects. Ask any relevant questions and proceed as you feel appropriate.

### FOCUSED HISTORY

---

1. Which hand is dominant?
2. One or both hands affected?
3. Duration of symptoms: Worsening or stable.
4. History of trauma or injury to hand/arm/shoulder.
5. Associated pain/numbness/paraesthesiae (including distribution).
6. Joint synovitis, muscle wasting, early morning stiffness.
7. Extent of weakness: Functional limitations, impact on quality of life (QOL)/ activities of daily living (ADLs).
8. Any other joint involvement?
9. Constitutional symptoms: Fever, weight loss.
10. Any other system involvement?
11. Drug history.
12. Family history of RA or autoimmune conditions.

### FOCUSED EXAMINATION

---

- General
  - Evidence of rheumatoid arthritis, acromegaly, thyroid disease or pregnancy
- Hands
  - Symmetrical arthropathy
  - Thenar eminence wasting
  - Deformity: Ulnar deviation, metacarpophalangeal joint (MCPJ) subluxation, swan neck, boutonnière, Z-thumb
  - Evidence of scars from previous tendon release or nerve decompression surgery
- Palpation
  - Hand, wrist and elbow joints: Check for evidence of active synovitis.
- Movements
  - Test specifically the lumbricals, opponens pollicis, abductor pollicis brevis and flexor pollicis brevis.
  - Assess function: Doing up a button, gripping a key and opening a door handle.
- Neurological
  - Phalen's and Tinel's tests to assess the median nerve



## QUESTIONS

1. What are the diagnostic criteria for rheumatoid arthritis?

### American College of Rheumatologists Criteria (2010)

#### Classification criteria for RA

(Score-based algorithm: Add score of Categories A–D. A score of  $\geq 6/10$  is needed for classification of a patient as having definite RA.)

A. Joint involvement	
1 large joint	0
2–10 large joints	1
1–3 small joints (with or without involvement of large joints)	2
4–10 small joints (with or without involvement of large joints)	3
>10 joints (at least one small joint)	5
B. Serology (at least one test result is needed for classification)	
Negative rheumatoid factor (RF) <i>and</i> negative anticitrullinated protein antibody (ACPA)	0
Low-positive RF <i>or</i> low-positive ACPA	2
High-positive RF <i>or</i> high-positive ACPA	3
C. Acute-phase reactants (at least one test result is needed for classification)	
Normal CRP <i>and</i> normal ESR 0	0
Abnormal CRP <i>or</i> normal ESR 1	1
D. Duration of symptoms	
<6 weeks	0
$\geq 6$ weeks	1

2. What are anticyclic citrullinated peptide (anti-CCP) antibodies?
  - Antibody markers used for the diagnosis of rheumatoid arthritis
  - Aid assessment of likely future progression to RA, in undifferentiated arthritis
  - More sensitive (~70%–75%) and specific (~95%–98%) than immunoglobulin (Ig) M rheumatoid factor antibodies
  - Prognostic value as marker of erosive disease
3. What biological therapies are available for the treatment of rheumatoid arthritis?
  - TNF- $\alpha$  blockers: Infliximab, adalimumab, etanercept
  - Monoclonal B-cell (anti-CD20) antibodies: Rituximab
  - IL-6 inhibitor: Tocilizumab
4. What are the serious side effects associated with the use of biological therapies?
  - Opportunistic infections: Fungal, bacterial, viral (e.g. severe infection with varicella-zoster virus).
  - Activation of latent TB  $\pm$  progression to miliary TB.
  - Anaphylaxis.
  - Increased risk of skin cancer.
  - Central nervous system demyelinating disorders (anti-TNF).
  - Anti-TNF and rituximab should not be used in Stage 3/4 NYHA heart failure.

## PATIENT WELFARE AND CONCERNS

---

1. Concerns regarding the impact of an inflammatory arthropathy on daily life and the possible cosmetic effects of deformities
2. Concerns regarding systemic extra-articular manifestations
3. Impact of DMARDs/biological agents and their side effects
4. Risks to offspring in terms of passing on the condition and their chances of developing rheumatoid arthritis in the future

## CANDIDATE EXPECTATIONS

---

1. Obtain a thorough history regarding the extent of joint involvement and duration of symptoms that clearly points to a diagnosis of inflammatory arthritis.
2. Ascertain the impact of symptoms on daily life. Can the patient still function as normal?
3. Thoroughly examine all affected joints. Assess active synovitis and identify any associated extra-articular manifestations.
4. Explain the likely diagnosis of rheumatoid arthritis.
5. Explain that management is a multidisciplinary team approach with physiotherapists, occupation therapists and nurse specialists involved, etc. Treatment strategies include anti-inflammatory agents, disease-modifying agents and biological agents. (There are NICE criteria for guidance on biologics use.)

## REFERENCE

---

Funovits J, et al. (2010) The 2010 American College of Rheumatology/European League Against Rheumatism classification criteria for rheumatoid arthritis: Methodological report phase I. *Ann Rheum Dis* 69: 1589–95.

## SYSTEMIC SCLEROSIS

This patient has been complaining of colour changes in her fingers and difficulty moving them. Please ask any relevant questions and proceed as appropriate.

## FOCUSED HISTORY

---

1. How long have these symptoms been going on for?
2. Have they noticed any changes in their skin over the fingers or elsewhere in the body?
3. Any discolouration of the skin anywhere on the body?
4. Any red patches (telangiectasiae)?
5. Any hard deposits anywhere, especially the fingers?
6. Any shortness of breath that is new or that is getting worse?
7. Any chest pain, swelling of legs or palpitations?
8. Any heartburn or difficulty swallowing?



9. Any joint pains or stiffness?
10. Any generalised symptoms, such as fatigue, difficulty sleeping, depression, anxiety, weight loss and malaise?

## FOCUSED EXAMINATION

---

- Hands
  - Sclerodactyly (thickened, tight skin), calcinosis, evidence of Raynaud's, digital ulcers, telangiectasia.
  - Assess the patient's hand function.
  - In someone with colour change in hands/digital ulcers, always assess the pulses.
- Face
  - Telangiectasiae, microstomia
- Lungs
  - Fine end inspiratory crackles in keeping with pulmonary fibrosis or evidence of pulmonary hypertension (loud P2, parasternal heave, raised JVP, tricuspid regurgitation and peripheral oedema)
- Extras
  - Ask for blood pressure and urine dip (renal failure)

## QUESTIONS

---

1. How is scleroderma classified?
  - Localised scleroderma
    - Localised skin involvement without internal organ involvement
  - Limited systemic sclerosis (SSc) (previously also known as CREST syndrome)
    - Calcinosis, Raynaud's phenomena, oesophageal dysmotility, sclerodactyly, telangiectasiae
    - Skin changes limited to hands, forearms, feet, neck and face
    - Risk of pulmonary hypertension
  - Diffuse systemic sclerosis
    - Skin changes proximal to the elbows or knees, face or neck
    - Can develop pulmonary fibrosis, scleroderma renal crisis or cardiac involvement
2. Which other organs are affected?
  - Kidneys: Scleroderma renal crisis (occurs in 5%–10% of SSc patients, who may present with an abrupt onset of hypertension, acute renal failure, headaches, fevers, malaise, hypertensive retinopathy, encephalopathy and pulmonary oedema)
  - Lungs: Pulmonary hypertension, pulmonary fibrosis
  - Cardiac: Pericardial effusions, myocardial fibrosis, conduction defects, congestive cardiac failure
3. What are the treatment options?
  - Raynaud's: Calcium channel blockers, e.g. nifedipine, ARB, e.g. losartan, intravenous prostacyclin (iloprost), bosentan (a dual endothelin-1 receptor antagonist).

- Reflux/oesophageal dysmotility: Proton pump inhibitors, H2 antagonists.
  - Hypertension/renal protection: ACE inhibitors.
  - Skin/joint/lung: Immune modulation – steroids, steroid-sparing agents, cyclophosphamide.
  - Surgical option includes digital sympathectomy.
  - Smoking cessation is key.
4. What immune tests can be used to help identify the cause?
- Diffuse systemic sclerosis: Anti-SCL-70 antibodies (against topoisomerase I)
  - Limited systemic sclerosis: Anticentromere antibodies
  - U1-RNP: Indicative of a mixed connective tissue disorder (systemic sclerosis, SLE, polymyositis)

## PATIENT WELFARE AND CONCERNS

---

1. Concerns regarding skin changes in hands
2. Is the reflux an unrelated separate issue to the Raynaud's/skin features?
3. What are the other complications of the condition, and are they treatable/curable?
4. What is the prognosis of the condition if there are cardiac, renal or pulmonary manifestations?
5. How will this condition affect life expectancy?

## CANDIDATE EXPECTATIONS

---

- Obtain a thorough history of the skin features.
- Recognise that the patient has Raynaud's and look for an underlying cause of this.
- Realise that the Raynaud's is associated with underlying systemic sclerosis.
- Examine the hands and face and other systems that can be affected by systemic sclerosis.
- Explain the likely diagnosis and what investigations will need to be carried out to determine this.
- Explain the range of treatment options, depending on the severity of the disease and the importance of preventing disease progression.

## THIRD NERVE PALSY OR PATIENT PRESENTING WITH DIPLOPIA

This 56-year-old male presents with diplopia, and his wife says his eye looks 'funny'. He has brittle diabetes and hypertension. Take a focused history and perform a focused examination to determine the diagnosis and formulate a management plan.

## FOCUSED HISTORY

---

1. When did this occur? (Note that it may be congenital.)
2. Describe the onset of diplopia (sudden vs. gradual).



3. Is there a specific direction of vision where the diplopia occurs?
4. Is there associated pain/headache?
5. Enquire about symptoms of raised intracranial pressure. Headache that is worse in the morning? Vomiting?
6. Need to review past medical history specifically focusing on diabetes/prior neurosurgery/multiple sclerosis/autoimmune disorders/hypertension.

## FOCUSED EXAMINATION

---

- Note: Do not waste your time performing a full cranial nerve examination.
- Observe for unilateral ptosis, dilated pupil and deviation of the eye laterally and downward (due to unopposed actions of lateral rectus and superior oblique).
- Observe for stigmata of diabetes, e.g. finger-prick blood glucose testing.
- Formally examine eye moments and check cranial nerves IV and VI at the same time.
- If you suspect a surgical third nerve palsy, then perform (or mention that you would like to perform) fundoscopy to observe for signs of raised intracranial pressure (i.e. papilloedema).
- Be sure to differentiate (via your examination) whether this is 'medical' third nerve palsy or a 'surgical' third nerve palsy.
- A medical third nerve palsy will usually be pupil sparing, and surgical third nerve palsy will not

## QUESTIONS

---

1. Explain the difference between a 'medical' and 'surgical' third nerve palsy?
  - Medical third nerve palsy tends to be pupil sparing, and surgical third nerve palsies lead to a dilated pupil. This is because surgical third nerve palsies are due to compressive effects on the sympathetic fibres that run along the outside of the nerve bundle that go on to supply the iris. As such, the pupil will be dilated early in a surgical palsy (note that in a medical palsy, the pupil *can* also become dilated).
2. What are the medical causes of a third nerve palsy?
  - Diabetes mellitus
  - Infection
  - Demyelinating disease
  - Autoimmune disease
  - Cavernous sinus thrombus
3. What are the surgical causes of a third nerve palsy?
  - Aneurysm or bleed from the posterior communicating artery
  - Postneurosurgery
  - Posttrauma
  - Congenital cause
4. Why does an oculomotor palsy lead to ptosis?
  - In addition to supplying the intrinsic muscle of the eye, the oculomotor nerve also supplies levator palpebrae superioris.

## **PATIENT WELFARE AND CONCERNS**

---

1. Be sympathetic if talking about prognosis. Medical causes may partially resolve with time. Prognosis with surgical causes is variable.
2. If talking about reconstructive eye lid surgery in chronic oculomotor palsy cases, remain empathetic regarding the appearance.

## **CANDIDATE EXPECTATIONS**

---

1. Demonstrate the signs of oculomotor palsy.
2. Differentiate between surgical and medical causes.

## **BRIEF CLINICAL ENCOUNTERS STATION SUMMARY**

- It is crucial to dedicate substantial revision time to Station 5.
- Be sure to look out early for 'spot diagnoses'.
- Any obvious signs may help guide towards a relevant history.
- In a case of back pain (such as ankylosing spondylitis), be sure to rule out spinal cord compression/cauda equine.
- There no set time to be spent on history and examination. This must be managed by the candidate on a case-by-case scenario. If there are likely to be many relevant signs, ensure that you demonstrate them.
- In cases of diabetes, always consider other micro- and macrovascular complications.
- Be able to assess a patient's thyroid status clinically.
- In a case of psoriasis, always look for evidence of arthritis.
- In any joint disorder, it is crucial to assess function.
- Take note of any patient concerns throughout and attempt to resolve them later in the consultation.





# Afterword: Hawks and Doves

As soon as you start revising for the Practical Assessment of Clinical Examination Skills (PACES), you should ask registrars and consultants to take you around to watch you examine patients, ask you questions and critique your performance. Very quickly you'll realise that some of these registrars and consultants are hawks and some are doves.

What is a hawk? What is hawkish behaviour? Hawks are tough. They believe in tough love, they give you a hard time, they expect high standards, they knock you off your perch and they keep you honest. Hawks are negative and pessimistic. They pick up on every omission or fault in your examinations, they ask you difficult questions and they critique you harshly.

Like yin and yang, doves are the opposite. Dove-like in behaviour, they are gentle. They encourage, they flatter, they appreciate your efforts, they build your confidence and they let you fly. Doves are positive and optimistic. They praise your thorough examinations, they ask sensible questions that you can answer and they critique you fairly.

These stereotypes are not absolute, but every registrar or consultant who takes you around will either have hawkish tendencies or be dove-like. But does this matter? Who cares? Does it change anything? The answer is yes. It really matters – I'll explain why.

During a typical PACES revision session in the first 2–3 weeks, you're learning your examinations, you're learning how to present and you're gaining in confidence, but it's all quite daunting. The last thing you need is a hawk to swoop down and tear you to shreds, destroying your confidence before you've even started. Go for the doves, to break you in gently.

After 2–3 weeks, your examinations become slicker, your presentations are smoother and you feel increasingly confident. You start to think you can do it, you can be good enough and you can make it. But PACES is difficult, examiners are not always nice; they can be tough. Now what you need are hawks to take you around, give you a kick up the backside, put the fear back into you and keep you honest and working hard, and not lazy or sloppy with your examinations and presentations.

Finally, the exam approaches, about a week away. The hawks and doves have worked well, building your confidence and keeping you honest. You start to peak, you're really good and you have every chance of passing. But the exam is daunting, you become nervous and you need a strong will and a positive mental attitude to perform on the day. The last thing you need is a hawk to tear you down before your exam, destroying you and making you think the whole PACES experience is all too much. Seek those reassuring doves again.

Good luck.

Ajay M Verma



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