Station 1
Abdominal and Respiratory

Clinical mark sheet

Clinical skill	Satisfactory	Unsatisfactory
Physical examination	Correct, thorough, fluent, systematic, professional	Incorrect technique, omits, unsystematic, hesitant
Identifying physical signs	Identifies correct signs Does not find signs that are not present	Misses important signs Finds signs that are not present
Differential diagnosis	Constructs sensible differential diagnosis	Poor differential, fails to consider the correct diagnosis
Clinical judgement	Sensible and appropriate management plan	Inappropriate management Unfamiliar with management
Maintaining patient welfare	Respectful, sensitive Ensures comfort, safety and dignity	Causes physical or emotional discomfort Jeopardises patient safety

Cases for PACES, Third Edition. Stephen Hoole, Andrew Fry and Rachel Davies.

Chronic liver disease and hepatomegaly

This man complains of weight loss and abdominal discomfort. His GP has referred him to you for a further opinion. Please examine his abdomen.

Clinical signs

SIGNS OF CHRONIC LIVER DISEASE

- **General:** cachexia, icterus (also in acute), excoriation and bruising
- **Hands:** leuconychia, clubbing, Dupuytren's contractures and palmar erythema
- **Face:** xanthelasma, parotid swelling and fetor hepaticus
- **Chest and abdomen:** spider naevi and caput medusa, reduced body hair, gynaecomastia and testicular atrophy (in males)

SIGNS OF HEPATOMEGALY

- Palpation and percussion:
  - Mass in the right upper quadrant that moves with respiration, that you are not able to get above and is dull to percussion
  - Estimate size (finger breadths below the diaphragm)
  - Smooth or craggy/nodular (malignancy/cirrhosis)
  - Pulsatile (TR in CCF)
- Auscultation
  - Bruit over liver (hepatocellular carcinoma)

EVIDENCE OF AN UNDERLYING CAUSE OF HEPATOMEGALY

• Tattoos and needle marks	Infectious hepatitis
• Slate-grey pigmentation	Haemochromatosis
• Cachexia	Malignancy
• Mid-line sternotomy scar	CCF

EVIDENCE OF TREATMENT

- Ascitic drain/tap sites
- Surgical scars

EVIDENCE OF DECOMPENSATION

- **A**scites: shifting dullness
- **A**sterixis: 'liver flap'
- **A**ltered consciousness: encephalopathy

Discussion

CAUSES OF HEPATOMEGALY

The **big three:**
Cirrhosis (alcoholic)
Carcinoma (secondaries)
Congestive cardiac failure
Plus: **I**nfectious (HBV and HCV)
Immune (PBC, PSC and AIH)
Infiltrative (amyloid and myeloproliferative disorders)

INVESTIGATIONS

- Bloods: FBC, clotting, U&E, LFT and glucose
- Ultrasound scan of abdomen
- Tap ascites (if present)

IF CIRRHOTIC

- Liver screen bloods:
  - Autoantibodies and immunoglobulins (PBC, PSC and AIH)
  - Hepatitis B and C serology
  - Ferritin (haemochromatosis)
  - Caeruloplasmin (Wilson's disease)
  - α-1 antitrypsin
  - Autoantibodies and immunoglobulins (PBC)
  - AFP (hepatocellular carcinoma)
- Hepatic synthetic function: INR (acute) and albumin (chronic)
- Liver biopsy (diagnosis and staging)
- ERCP (diagnose/exclude PSC)

IF MALIGNANCY

- Imaging: CXR and CT abdomen/chest
- Colonoscopy/gastroscopy
- Biopsy

COMPLICATIONS OF CIRRHOSIS

- Variceal haemorrhage due to portal hypertension
- Hepatic encephalopathy
- Spontaneous bacterial peritonitis

CHILD-PUGH CLASSIFICATION OF CIRRHOSIS

Prognostic score based on bilirubin/albumin/INR/ascites/encephalopathy

	Score	1 year survival
A:	5–6	100%
B:	7–9	81%
C:	10–15	45%

CAUSES OF ASCITES

- **C**irrhosis (80%)
- **C**arcinomatosis
- **C**CF

TREATMENT OF ASCITES IN CIRRHOTICS

- Abstinence from alcohol
- Salt restriction
- Diuretics (aim: 1 kg weight loss/day)
- Liver transplantation

CAUSES OF PALMAR ERYTHEMA

- Cirrhosis
- Hyperthyroidism
- Rheumatoid arthritis
- Pregnancy
- Polycythaemia

CAUSES OF GYNAECOMASTIA

- Physiological: puberty and senility
- Kleinfelter's syndrome
- Cirrhosis
- Drugs, e.g. spironolactone and digoxin
- Testicular tumour/orchidectomy
- Endocrinopathy, e.g. hyper/hypothyroidism and Addison's

AUTOANTIBODIES IN LIVER DISEASE

- Primary biliary cirrhosis (PBC): antimitochondrial antibody (M2 subtype) in 98%, increased IgM
- Primary sclerosing cholangitis (PSC): ANA, anti-smooth muscle may be positive
- Autoimmune hepatitis (AIH): anti-smooth muscle, anti-liver/kidney microsomal type 1(LKM1) and occasionally ANA may be positive (pattern helps classify)

Haemochromatosis

This 52-year-old man was referred after a diagnosis of diabetes mellitus was made by his GP. Please examine him and discuss further investigations.

Clinical signs

- Increased skin pigmentation (slate-grey colour)
- Stigmata of chronic liver disease
- Hepatomegaly

SCARS

- Venesection
- Liver biopsy
- Joint replacement
- Abdominal rooftop incision (hemihepatectomy for hepatocellular carcinoma)

EVIDENCE OF COMPLICATIONS

- **Endocrine:** 'bronze diabetes' (e.g. injection sites), hypogonadism and testicular atrophy
- **Cardiac:** congestive cardiac failure
- **Joints:** arthropathy (pseudo-gout)

Discussion

INHERITANCE

- Autosomal recessive on chromosome 6
- **HFE** gene mutation: regulator of gut iron absorption
- Homozygous prevalence 1:300, carrier rate 1:10
- Males affected at an earlier age than females – protected by menstrual iron losses

PRESENTATION

- Fatigue and arthritis
- Chronic liver disease
- Incidental diagnosis or family screening

INVESTIGATION

- ↑ Serum ferritin
- ↑ Transferrin saturation
- Liver biopsy (diagnosis + staging)
- Genotyping

And consider:

• Blood glucose	Diabetes
• ECG, CXR, ECHO	Cardiac failure
• Liver ultrasound, α-fetoprotein	Hepatocellular carcinoma (HCC)

TREATMENT

- Regular venesection (1 unit/week) until iron deficient, then venesect 1 unit, 3–4 times/year
- Avoid alcohol
- Surveillance for HCC

FAMILY SCREENING (1ST DEGREE RELATIVES AGED > 20 YEARS)

- Iron studies (ferritin and TSAT)

If positive:

- Liver biopsy
- Genotype analysis

PROGNOSIS

- 200 × increased risk of HCC if cirrhotic
- Reduced life expectancy if cirrhotic
- Normal life expectancy without cirrhosis + effective treatment

Splenomegaly

This man presents with tiredness and lethargy. Please examine his abdominal system and discuss your diagnosis.

Clinical signs

GENERAL

- Anaemia
- Lymphadenopathy (axillae, cervical and inguinal areas)
- Purpura

ABDOMINAL

- Left upper quadrant mass that moves inferomedially with respiration, has a notch, is dull to percussion and you cannot get above nor ballot
- Estimate size
- Check for hepatomegaly

UNDERLYING CAUSE

Lymphadenopathy	Haematological and infective
Stigmata of chronic liver disease	Cirrhosis with portal hypertension
Splinter haemorrhages, murmur, etc.	Bacterial endocarditis
Rheumatoid hands	Felty's syndrome

Discussion

CAUSES

- Massive splenomegaly (>8 cm):
  - Myeloproliferative disorders (**CML** and **myelofibrosis**)
  - Tropical infections (**malaria**, visceral leishmaniasis: kala-azar)
- Moderate (4–8 cm):
  - Myelo/lymphoproliferative disorders
  - Infiltration (Gaucher's and amyloidosis)
- Tip (<4 cm):
  - Myelo/lymphoproliferative disorders
  - Portal hypertension
  - Infections (EBV, infective endocarditis and infective hepatitis)
  - Haemolytic anaemia

INVESTIGATIONS

- Ultrasound abdomen

Then if:

- **Haematological:**
  - FBC and film
  - CT chest and abdomen
  - Bone marrow aspirate and trephine
  - Lymph node biopsy
- **Infectious:**
  - Thick and thin films (malaria)
  - Viral serology

INDICATIONS FOR SPLENECTOMY

- Rupture (trauma)
- Haematological (ITP and hereditary spherocytosis)

SPLENECTOMY WORK-UP

- Vaccination (ideally 2/52 prior to protect against encapsulated bacteria):
  - Pneumococcus
  - Meningococcus
  - *Haemophilus influenzae* (Hib)
- Prophylactic penicillin: (lifelong)
- Medic alert bracelet

Renal enlargement

This woman has been referred by her GP for investigation of hypertension. Please examine her abdomen.

Clinical signs

PERIPHERAL

- Blood pressure: **hypertension**
- Arteriovenous fistulae (thrill and bruit), tunnelled dialysis line
- Immunosuppressant 'stigmata', e.g. Cushingoid habitus due to steroids, gum hypertrophy with ciclosporin

ABDOMEN

- Palpable kidney: ballotable, can get above it and moves with respiration
- Polycystic kidneys: both may/should be palpable, and can be grossly enlarged (will feel 'cystic', or nodular)
- Iliac fossae: scar with (or without!) transplanted kidney
- Ask to dip the urine: proteinuria and haematuria
- Ask to examine the external genitalia (varicocele in males)

ASSOCIATED CONDITIONS

- Hepatomegaly: polycystic kidney disease
- Indwelling catheter: obstructive nephropathy with hydronephrosis
- Peritoneal dialysis catheter/scars

Discussion

CAUSES OF UNILATERAL ENLARGEMENT

- Polycystic kidney disease (other kidney not palpable or contralateral nephrectomy – flank scar)
- Renal cell carcinoma
- Simple cysts
- Hydronephrosis (due to ureteric obstruction)

CAUSES OF BILATERAL ENLARGEMENT

- Polycystic kidney disease
- Bilateral renal cell carcinoma (5%)
- Bilateral hydronephrosis
- Tuberous sclerosis (renal angiomyolipomata and cysts)
- Amyloidosis

INVESTIGATIONS

- U&E
- Urine cytology
- Ultrasound abdomen ± biopsy
- IVU
- CT if carcinoma is suspected
- Genetic studies (ADPKD)

Autosomal dominant polycystic kidney disease:

- Progressive replacement of normal kidney tissue by cysts leading to renal enlargement and renal failure (5% of end-stage renal failure in UK)
- Prevalence 1:1000
- Genetics: 85% *ADPKD1* chromosome 16; 15% *ADPKD2* chromosome 4

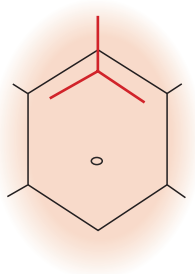
- Present with:
  - Hypertension
  - Recurrent UTIs
  - Abdominal pain (bleeding into cyst and cyst infection)
  - Haematuria
- End-stage renal failure by age 40–60 years (earlier in *ADPKD1* than *2*)
- Other organ involvement:
  - Hepatic cysts and hepatomegaly (rarely liver failure)
  - Intracranial Berry aneurysms (neurological sequelae/craniotomy scar?)
  - Mitral valve prolapse
- Genetic counselling of family and family screening; 10% represent new mutations
- Treatment: nephrectomy for recurrent bleeds/infection/size, dialysis and renal transplantation

The liver transplant patient

Please examine this man's abdomen.

Clinical signs

- Scars:



'Mercedes–Benz' or roof–top scar
Liver transplant

- Evidence of chronic liver disease

REASON FOR LIVER TRANSPLANTATION

- Slate-grey pigmentation — Haemochromatosis
- Other autoimmune disease — PBC
- Tattoos and needle marks — Hepatitis B, C

EVIDENCE OF IMMUNOSUPPRESSIVE MEDICATION

- Ciclosporin: gum hypertrophy and hypertension
- Steroids: Cushingoid appearance, thin skin, ecchymoses

Discussion

TOP THREE REASONS FOR LIVER TRANSPLANTATION

- Cirrhosis
- Acute hepatic failure (hepatitis A and B, paracetamol overdose)
- Hepatic malignancy (hepatocellular carcinoma)

SUCCESS OF LIVER TRANSPLANTATION

- 80% 1-year survival
- 70% 5-year survival

CAUSES OF GUM HYPERTROPHY

- Drugs: ciclosporin, phenytoin and nifedipine
- Scurvy
- Acute myelomonocytic leukaemia
- Pregnancy
- Familial

SKIN SIGNS IN (ANY) TRANSPLANT PATIENTS

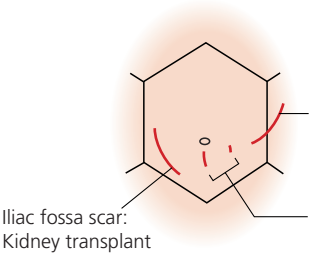
- **Malignancy**
  - Dysplastic change (actinic keratoses)
  - Squamous cell carcinoma (100 × increased risk and multiple lesions)
  - Basal cell carcinoma and malignant melanoma (10 × increased risk)
- **Infection:**
  - Viral warts
  - Cellulitis

The renal patient

Please examine this man's abdomen.

Clinical signs

- Stigmata:
  - Arms: arteriovenous fistula(e) – currently working (thrill), being used (thrill and dressings), or failed
  - Neck: tunneled dialysis line (or previous lines; scars in the root of the neck and over the chest wall)
  - Abdomen:



- Fluid status (leg oedema)

THREE THINGS TO CONSIDER IN ALL RENAL PATIENTS

1. **Underlying reason for renal failure**
   - Polycystic kidneys: ADPKD
   - Visual impairment, fingerprick marks, injection sites/pump, etc.: diabetes
   - Sclerodactyly, typical facies: systemic sclerosis
   - Rheumatoid hands, nodules: rheumatoid arthritis
   - (Hepato)splenomegaly: amyloidosis
   - Other organ transplantation (liver/heart/lungs): calcineurin inhibitor nephrotoxicity
   - Ungual fibromata, adenoma sebaceum, polycystic kidneys: tuberous sclerosis
2. **Current treatment modality**
   - Haemodialysis: working fistula, tunneled neck lines, arteriovenous grafts
   - Peritoneal dialysis: abdominal catheter
   - Functioning transplant: no evidence of other current dialysis access (in use)
3. **Complications of past/current treatment**
   - Side effects of treatment for the underlying disease: Cushingoid appearance from steroids (glomerulonephritis)
   - Side effects of immunosuppressive treatment in transplant patients:
     - Fine tremor (tacrolimus)
     - Steroid side effects
     - Gum hypertrophy (ciclosporin)
     - Hypertension (ciclosporin, tacrolimus)
     - Skin damage and malignancy (especially ciclosporin and azathioprine)
   - Scars from previous access for dialysis, failed transplant(s)

KIDNEY-PANCREAS TRANSPLANTATION

Consider if:

- Lower midline abdominal incision, with a palpable kidney in an iliac fossa (but no overlying scar)

- Evidence of previous diabetes (e.g. visual impairment)
- Patients are often younger (most commonly transplanted in 30s–40s)

Discussion

TOP THREE CAUSES FOR RENAL TRANSPLANTATION

- Glomerulonephritis
- Diabetic nephropathy
- Polycystic kidney disease (ADPKD)

PROBLEMS FOLLOWING TRANSPLANTATION

- **Rejection:** acute or chronic
- **Infection secondary to immunosuppression:**
  - *Pneumocystis carinii*
  - CMV
- **Increased risk of other pathology:**
  - Skin malignancy
  - Post-transplant lymphoproliferative disease
  - Hypertension and hyperlipidaemia causing cardiovascular disease
- **Immunosuppressant drug side effects/toxicity:**
  - Ciclosporin nephrotoxicity
- **Recurrence of original disease**
- **Chronic graft dysfunction**

SUCCESS OF RENAL TRANSPLANTATION

- 90% 1-year graft survival
- 50% 10-year graft survival (better with live-related donor grafts)

Pulmonary fibrosis

Examine this patient's respiratory system, she has been complaining of progressive shortness of breath.

Clinical signs

- Clubbing, central cyanosis and tachypnoea
- Fine end-inspiratory crackles (like Velcro® which do not change with coughing)
- Signs of associated autoimmune diseases, e.g. rheumatoid arthritis (hands), SLE and systemic sclerosis (face and hands)
- Signs of treatment, e.g. Cushingoid from steroids
- Discoloured skin (grey) – amiodarone
- Unless there are any associated features then describe your findings as pulmonary fibrosis, which is a clinical description pending further differentiation following investigations

Discussion

INVESTIGATION

- **Bloods:** ESR, rheumatoid factor and ANA
- **CXR:** reticulonodular changes; loss of definition of either heart border; small lungs
- **ABG:** type I respiratory failure
- **Lung function tests:**
  - FEV_1/FVC > 0.8 (restrictive)
  - Low TLC (small lungs)
- Reduced TLco and K_{CO}
- **Bronchoalveolar lavage:** main indication is to exclude any infection prior to immunosuppressants plus if lymphocytes > neutrophils indicate a better response to steroids and a better prognosis (sarcoidosis)
- **High-resolution CT scan:** distribution helps with diagnosis; bibasal subpleural honeycombing typical of UIP; widespread ground glass shadowing more likely to be non-specific interstitial pneumonia often associated with autoimmune disease; if apical in distribution then think of sarcoidosis, ABPA, old TB, hypersensitivity pneumonitis, Langerhan's cell histiocytosis.
- Lung biopsy (associated morbidity ~7%)

TREATMENT

- Immunosuppression if likely to be inflammatory; i.e. non-specific interstitial pneumonia e.g. steroids: combination of steroids and azathioprine no longer used following results of PANTHER trial which showed increased morbidity on this combination
- Pirfenidone (an antifibrotic agent) - for UIP when FEV_1 50–80% predicted (NICE recommended)
- N-acetyl cycsteine – free radical scavenger
- Single lung transplant
- NB: Beware single lung transplantation patient – unilateral fine crackles and contralateral thoracotomy scar with normal breath sounds

PROGNOSIS

- Very variable: depends on aetiology
- Highly cellular with ground glass infiltrate – responds to immunosuppression: 80% 5-year survival

- Honeycombing on CT – no response to immunosuppression: 80% 5-year mortality
- There is an increased risk of bronchogenic carcinoma

CAUSES OF BASAL FIBROSIS

- Usual interstitial pneumonia (UIP)
- Asbestosis
- Connective tissue diseases
- Aspiration

Bronchiectasis

This 60-year-old woman presents to your clinic with a persistent cough. Please examine her and discuss your findings.

Clinical signs

General: Cachexia and tachypnoea
Hands: Clubbing
Chest: Mixed character crackles that alter with coughing. Occasional squeaks and wheeze. Sputum + + + (look in the pot!)

- **Cor pulmonale:** SOA, raised JVP, RV heave, loud P_2
- **Yellow nail syndrome:** yellow nails and lymphoedema

Discussion

INVESTIGATION

- Sputum culture and cytology
- CXR: tramlines and ring shadows
- **High-resolution CT thorax:** `signet ring' sign (thickened, dilated bronchi larger than the adjacent vascular bundle)

FOR A SPECIFIC CAUSE

- **Immunoglobulins:** hypogammaglobulinaemia (especially IgG_2 and IgA)
- ***Aspergillus* RAST or skin prick testing:** ABPA (upper lobe)
- **Rheumatoid serology**
- **Saccharine ciliary motility test** (nares to taste buds in 30 minutes): Kartagener's
- **Genetic screening:** cystic fibrosis
- History of inflammatory bowel disease

CAUSES OF BRONCHIECTASIS

- **Congenital:** Kartagener's and cystic fibrosis
- **Childhood infection:** measles and TB
- **Immune OVER activity:** allergic bronchopulmonary aspergillosis (ABPA); inflammatory bowel disease associated
- **Immune UNDER activity:** hypogammaglobulinaemia; CVID
- **Aspiration:** chronic alcoholics and GORD; localized to right lower lobe

TREATMENT

- Physiotherapy – active cycle breathing
- Prompt antibiotic therapy for exacerbations
- Long-term treatment with low dose azithromycin three times per week
- Bronchodilators/inhaled corticosteroids if there is any airflow obstruction
- Surgery is occasionally used for localized disease

COMPLICATIONS OF BRONCHIECTASIS

- Cor pulmonale
- (Secondary) amyloidosis (Dip urine for protein)
- Massive haemoptysis (mycotic aneurysm)

Old tuberculosis

Please examine this man's respiratory system.

Clinical signs

- Chest deformity and absent ribs; thoracoplasty scar
- Tracheal deviation towards the side of the fibrosis (traction)
- Reduced expansion
- Dull percussion but present tactile vocal fremitus
- Crackles and bronchial breathing

Discussion

HISTORICAL TECHNIQUES

- **Plombage:** insertion of polystyrene balls into the thoracic cavity
- **Phrenic nerve crush:** diaphragm paralysis
- **Thoracoplasty:** rib removal; lung not resected
- Apical lobectomy
- Recurrent medical pneumothoraces
- Streptomycin was introduced in the 1950s. It was the first drug shown to be beneficial in a randomized controlled trial

SERIOUS SIDE EFFECTS OF TB DRUGS

- **Isoniazid** Peripheral neuropathy (Rx Pyridoxine) and hepatitis
- **Rifampicin** Hepatitis and increased contraceptive pill metabolism
- **Ethambutol** Retro-bulbar neuritis and hepatitis
- **Pyrazinamide** Hepatitis

Prior to treating TB, check baseline liver function tests and visual acuity. Tell the patient the following:

1. Look at the whites of your eyes every morning. If yellow, stop the tablets and ring the TB nurse that morning.
2. Notice colours – if red becomes less bright than you expect ring the TB nurse that day.
3. You may develop tingling in your toes – continue with the tablets but tell the doctor at your next clinic visit.
4. Your secretions will turn orange/red. This is because of a dye in one of the tablets. If you wear contact lenses they will become permanently stained and should not be worn.
5. If you are on the OCP, it may fail. Use barrier contraception.

CAUSES OF APICAL FIBROSIS: 'TRASHE'

- **T**B
- **R**adiation
- **A**nkylosing spondylitis/ABPA
- **S**arcoidosis
- **H**istoplasmosis/Histiocytosis X
- **E**xtrinsic allergic alveolitis (now referred to as hypersensitivity pneumonitis)

Surgical respiratory cases

Please examine this man who initially presented to doctors with a cough and weight loss.

Lobectomy

CLINICAL SIGNS

- Reduced expansion and chest wall deformity
- Thoracotomy scar: same for either upper or lower lobe
- Trachea is central
- Lower lobectomy: dull percussion note over lower zone with absent breath sounds
- Upper lobectomy: may have normal examination or may have a hyper-resonant percussion note over upper zone with a dull percussion note at base where the hemidiaphragm is elevated slightly

INVESTIGATION

- CXR: maybe no overt abnormality apparent other than slight raised hemidiaphragm; remember that the right hemidiaphragm should be higher than the left in health
- CT chest: loss of a lobe with associated truncation of bronchus or pulmonary vessels

Pneumonectomy

CLINICAL SIGNS

- Thoracotomy scar (indistinguishable from thoracotomy scar performed for a lobectomy)
- Reduced expansion on side of the pneumonectomy
- Trachea deviated towards the side of the pneumonectomy
- Dull percussion note throughout the hemithorax
- Absent tactile vocal fremitus beneath the thoracotomy scar
- Bronchial breathing in the upper zone with reduced breath sound throughout remainder of hemithorax (bronchial breathing is due to transmitted sound from major airways)

DISCUSSION

- CXR: complete white out on side of pneumonectomy
- Pneumonectomy space fills with gelatinous material within a few weeks of the operation

Lung transplantation

Signal lung transplant

- Clinical signs: thoracotomy scar; normal exam on side of scar; may have clinical signs on opposite hemithorax
- Indications for 'dry lung' condititions: COPD; pulmonary fibrosis

Double lung transplant

- Clinical signs: clamshell incision – from the one axilla along the line of the lower ribs, up to the xiphisternum to the other axilla
- Indications: 'wet lung' conditions: CF, bronchiectasis or pulmonary hypertension

Chronic obstructive airways disease

Please examine this patient's chest; he has a chronic chest condition.

Clinical signs

- Inspection: nebulizer/inhalers/sputum pot, dyspnoea, central cyanosis and pursed lips
- CO_2 retention flap, bounding pulse and tar-stained fingers
- Hyper-expanded
- Percussion note resonant with loss of cardiac dullness
- Expiratory polyphonic wheeze (crackles if consolidation too) and reduced breath sounds at apices
- Cor pulmonale: raised JVP, ankle oedema, RV heave; loud P_2 with pansystolic murmur of tricuspid reurgitation
- COPD does not cause clubbing: therefore, if present consider bronchial carcinoma or bronchiectasis

Discussion

- Spectrum of disease with airway obstruction (with or without sputum production); can be low FEV_1 at one end and emphysema with low O_2 sats but normal spirometry at the other
- Degree of overlap with chronic asthma, although in COPD there is less reversibility (<15% change in FEV_1 post-bronchodilators)

CAUSES

- Environmental: smoking and industrial dust exposure (apical disease)
- Genetic: α_1-antitrypsin deficiency (basal disease)

INVESTIGATIONS

- **CXR:** hyper-expanded and/or pneumothorax
- **ABG:** type II respiratory failure (low PaO_2 high $PaCO_2$)
- **Bloods:** high WCC (infection), low α_1-antitrypsin (younger patients/FH+), low albumin (severity)
- **Spirometry:** low FEV_1, $FEV_1/FVC < 0.7$ (obstructive)
- **Gas transfer:** low T_LCO

TREATMENT

- **Medical** – depends on severity **(GOLD classification):**
  - **Smoking cessation** is the single most beneficial management strategy
  - Cessation clinics and nicotine replacement therapy
  - Long-term oxygen therapy (LTOT)
  - Pulmonary rehabilitation
  - Mild (FEV_1 >80) – beta-agonists
  - Moderate (FEV_1 <60%) – tiotropium plus beta-agonists
  - Severe (FEV_1 <40%) or frequent exacerbations – above plus inhaled corticosteroids; although avoid if patient has ever had an episode of pneumonia (TORCH trial)
  - Exercise
  - Nutrition (often malnourished)
  - Vaccinations - pneumoccoal and influenza

- **Surgical** (careful patient selection is important)
  - Bullectomy (if bullae >1 L and compresses surrounding lung)
  - Endobronchial valve placement
  - Lung reduction surgery: only suitable for a few patient with heterogeneous distribution of emphysema
  - Single lung transplant

LONG-TERM OXYGEN THERAPY (LTOT)

- **Inclusion criteria:**
  - Non-smoker
  - PaO_2 <7.3 kPa on air
  - $PaCO_2$ that does not rise excessively on O_2
  - If evidence of cor pulmonale, PaO_2 <8 kPa
- 2–4 L/min via nasal prongs for at least 15 hours a day
- Improves average survival by 9 months

TREATMENT OF AN ACUTE EXACERBATION

- Controlled O_2 via Venturi mask monitored closely
- Bronchodilators
- Antibiotics
- Steroids 7 days

PROGNOSIS

COPD patients with an acute exacerbation have 15% in-hospital mortality

DIFFERENTIAL OF A WHEEZY CHEST

- Granulomatous polyarteritis (previously Wegner's): saddle nose; obliterative bronchiolitis
- Rheumatoid arthritis: wheeze secondary to obliterative bronchiolitis
- Post-lung transplant: obliterative bronchiolitis as part of chronic rejection spectrum

Pleural effusion

This patient has been breathless for 2 weeks. Examine his respiratory system to elucidate the cause.

Clinical signs

- Asymmetrically reduced expansion
- Trachea or mediastinum displaced away from side of effusion
- **Stony** dull percussion note
- Absent tactile vocal fremitus
- Reduced breath sounds
- Bronchial breathing above (aegophony)

SIGNS THAT MAY INDICATE THE CAUSE

- **Cancer:** clubbing; lymphadenopathy; mastectomy (breast cancer being a very common cause of pleural effusion)
- **Congestive cardiac failure:** raised JVP; peripheral oedema
- **Chronic liver disease:** leuconychia, spider naevi gynaecomastia
- **Chronic renal failure:** arteriovenous fistula
- **Connective tissue disease:** rheumatoid hands; butterfly rash of SLE

CAUSES OF A DULL LUNG BASE

- **Consolidation:** bronchial breathing and crackles
- **Collapse:** tracheal deviation towards the side of collapse and reduced breath sounds
- Previous lobectomy = reduced lung volume
- **Pleural thickening:** signs are similar to a pleural effusion but with normal tactile vocal fremitus; may have three scars suggestive of previous VATS pleuradesis
- **Raised hemidiaphragm** ± hepatomegaly

Discussion

CAUSES

Transudate (protein <30 g/L)	Exudate (protein >30 g/L)
Congestive cardiac failure	Neoplasm: 1° or 2°
Chronic renal failure	Infection
Chronic liver failure	Infarction
	Inflammation: RA and SLE

Pleural aspiration (exudate)

- **Protein:** effusion albumin/plasma albumin >0.5 (Light's criteria)
- **LDH:** effusion LDH/plasma LDH >0.6
- **Empyema:** an exudate with a low glucose and pH <7.2 is suggestive

Empyema

- A collection of pus within the pleural space
- Most frequent organisms: anaerobes, staphylococci and Gram-negative organisms
- Associated with bronchial obstruction, e.g. carcinoma, with recurrent aspiration; poor dentition; alcohol dependence

TREATMENT

- Pleural drainage and IV antibiotics intrapleural DNAse plus TPA (MIST 2 Trial)
- Surgical decortication

Lung cancer

Please examine this patient who has had a 3-month history of chronic cough, malaise and weight loss.

Clinical signs

- Cachectic
- Clubbing and tar-stained fingers
- Lymphadenopthy: cervical and axillary
- Tracheal deviation: towards (collapse) or away (effusion) from the lesion
- Reduced expansion
- Percussion note dull (collapse/consolidation) or stony dull (effusion)
- Absent tactile vocal fremitus (effusion); increased vocal resonance (collapse/consolidation)
- Auscultation:
  - Crackles and bronchial breathing (consolidation/collapse)
  - Reduced breath sounds; absent tactile fremitus (effusion)
- **Hepatomegaly or bony tenderness:** metastasis
- **Treatment:**
  - Lobectomy scar
  - **Radiotherapy:** square burn and **tattoo**
- **Complications:**
  - **Superior vena cava obstruction:** suffused and oedematous face and upper limbs, dilated superficial chest veins and stridor
  - **Recurrent laryngeal nerve palsy:** hoarse with a 'bovine' cough
  - **Horner's sign and wasted small muscles of the hand (T1):** Pancoast's tumour
  - **Endocrine:** gynaecomastia (ectopic βHCG)
  - **Neurological:** Lambert–Eaton myasthenia syndrome, peripheral neuropathy, proximal myopathy and paraneoplastic cerebellar degeneration
  - **Dermatological:** dermatomyositis (heliotrope rash on eye lids and purple papules on knuckles (Gottron's papules associated with a raised CK) and acanthosis nigricans

Discussion

TYPES

- Squamous 35%, small (oat) 24%, adeno 21%, large 19% and alveolar 1%

MANAGEMENT

1. Diagnosis of a mass:
   - **CXR:** collapse, mass and hilar lymphadenopathy
   - **Volume acquisition CT thorax** (so small tumours are not lost between slices) with contrast
2. Determine cell type:
   - **Induced sputum cytology**
   - **Biopsy** by **bronchoscopy** (central lesion and collapse) or **percutaneous needle** CT guided (peripheral lesion; FEV_1 >1 L))
3. Stage (**CT/bronchoscopy/endobronchial ultrasound guided biopsy/mediastinoscopy/thoracoscopy/PET**):
   - **Non-small cell carcinoma (NSCLC): TNM staging to assess operability**
   - Small cell carcinoma (SCLC): limited or extensive disease
4. Lung function tests for operability assessment:
   - Pneumonectomy contraindicated if FEV_1 < 1.2 L
5. Complications of the tumour:
   - Metastasis: ↑ LFTs, ↑ Ca^{++}, ↓ Hb
   - NSCLC: ↑ PTHrP → ↑ Ca^{++}
   - SCLC: ↑ ACTH, SIADH → Na^{+} ↓

TREATMENT

- **NSCLC:**
  - **Surgery:** lobectomy or pneumonectomy
  - **Radiotherapy:** single fractionation (weekly) versus hyper-fractionation (daily for 10 days)
  - **Chemotherapy:** benefit unknown; EGFR Positive – erlotinib
- **SCLC:**
  - **Chemotherapy:** benefit with six courses

Multidisciplinary approach

PALLIATIVE CARE

- Dexamethasone and radiotherapy for brain metastasis
- SVCO: dexamethasone plus radiotherapy or intravascular stent
- Radiotherapy for haemoptysis, bone pain and cough
- Chemical pleurodesis for effusion – talc; tetracycline no longer used
- Opiates for cough and pain

Cystic fibrosis

Please examine this young man's chest and comment on what you find.

Clinical signs

- Inspection: small stature, **clubbed**, tachypnoeic, sputum pot (purulent++)
- Hyperinflated with reduced chest expansion
- **Coarse crackles** and wheeze (bronchiectatic)
- **Portex reservoir** (Portacath®) under the skin or **Hickman line/scars** for long-term antibiotics plus PEG for malabsorption

Discussion

GENETICS

- Incidence of 1/2500 live births
- Autosomal recessive chromosome 7q
- Gene encodes CFTR (Cl^-channel)
- Commonest and most severe mutation is the deletion Δ508/ Δ508 (70%)

PATHOPHYSIOLOGY

Secretions are thickened and block the lumens of various structures:

- Bronchioles → bronchiectasis
- Pancreatic ducts → loss of exocrine and endocrine function
- Gut → distal intestinal obstruction syndrome (DIOS) in adults
- Seminal vesicles → male infertility
- Fallopian tubes – reduced female fertility

INVESTIGATIONS

- Screened at birth: low immunoreactive trypsin (heel prick)
- Sweat test: Na^+ > 60 mmol/L (false-positive in hypothyroidism and Addison's)
- Genetic screening

TREATMENT

- **Physiotherapy:** postural drainage and active cycle breathing techniques
- Prompt antibiotics for intercurrent infections
- Pancrease® and fat-soluble vitamin supplements
- Mucolytics (DNAse)
- Immunizations
- Double lung transplant (50% survival at 5 years)
- Gene therapy is under development

PROGNOSIS

Median survival is 35 years but is rising. Poor prognosis if becomes infected with *Burkholderia cepacia*

Pneumonia

This patient has been acutely unwell for 3 days, with shortness of breath and a productive cough. Please examine his chest.

Clinical signs

- Tachypnoea, O_2 mask, sputum pot (rusty sputum associated with *pneumococcus*)
- Reduced expansion
- Dull percussion note
- Focal coarse crackles, increased vocal resonance and bronchial breathing
- Ask for the temperature chart
- If dull percussion note with absent tactile vocal fremitus, think parapneumonic effusion/empyema

Discussion

INVESTIGATION

- **CXR:** consolidation (air bronchogram), abscess and effusion
- **Bloods:** WCC, CRP, urea, atypical serology (on admission and at day 10) and immunoglobulins
- **Blood** (25% positive) and **sputum cultures**
- **Urine:**
  - *Legionella* antigen (in severe cases)
  - *Pneumococcal* antigen
  - Haemoglobinuria (*mycoplasma* causes cold agglutinins → haemolysis)

COMMUNITY ACQUIRED PNEUMONIA (CAP)

- Common organisms:
  - ***Streptococcus pneumoniae*** 50%
  - ***Mycoplasma pneumoniae*** 6%
  - *Haemophilus influenzae* (especially if COPD)
  - *Chlamydia pneumoniae*.
- Antibiotics:
  - 1st line: penicillin *or* cephalosporin + macrolide

SPECIAL CONSIDERATIONS

- **Immunosuppressed:**
  - Fungal — Rx Amphotericin
  - Multi-resistant mycobacteria
  - *Pneumocystis carinii* — Rx Cotrimoxazole/Pentamidine
  - CMV — Rx Ganciclovir
- **Aspiration** (commonly posterior segment of right lower lobe):
  - Anaerobes — Rx + Metronidazole
- **Post-influenza:**
  - *Staph. aureus* — Rx + Flucloxacillin

SEVERITY SCORE FOR PNEUMONIA: CURB-65 (2/5 IS SEVERE)

- **C**onfusion
- **U**rea >7
- **R**espiratory rate >30
- **B**P systolic <90 mm Hg or diastolic <60 mm Hg
- **Age >65**

Severe CAP should receive high-dose IV antibiotics initially plus level 2 care (HDU/ITU)

PREVENTION

Pneumovax II® to high-risk groups, e.g. chronic disease (especially nephrotic and asplenic patients) and the elderly

COMPLICATIONS

- Lung abscess (*Staph. aureus, Klebsiella*, anaerobes)
- Para-pneumonic effusion/empyema
- Haemoptysis
- Septic shock and multi-organ failure