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Notes & Notes for MRCP
By Dr. Yousif Abdallah Hamad

• Hamman's sign (or 'crunch') is a clicking sound synchronous with the heart-beat , heard over the sternal edge in mediastinal emphysema or Left-sided pneumothoraces. Risk factors • Young adult males, often tall and slim, are frequently affected by spontaneous pneumothorax. • Patients with Marfan syndrome are prone to recurrent pneumothoraces. Investigations • Chest x ray : 1st step to confirm the diagnosis Questions sometimes discuss the size of the pneumothorax in percentage terms rather than giving the interpleural distance. A 30% pneumothorax ≈ 2 cm A 50% pneumothorax is likely to have a rim of > 3cm. • CT chest The next step after chest x-ray to investigate the underlying cause of recurrent pneumothorax • Video assisted thoracoscopy If CT not help in pointing to underlying cause of recurrent pneumothorax Differential diagnosis • Large bullae in COPD can mimic a pneumothorax: the most appropriate management option →CT chest to confirm place a needle or chest drain would be disastrous → shrinkage of the lung Management • Primary pneumothorax Definition: Spontaneous primary pneumothorax is defined as: Age less than 50-years-old No significant smoking history, minimal smoking history would still be considered as primary pneumothorax No evidence of underlying lung disease. Caused by the rupture of apical pleural blebs. Management If the rim of air is < 2cm and the patient is not short of breath then discharge should be considered If the rim of air is ≥2cm or the patient is breathless → Needle aspiration If following aspiration the rim of air is < 2cm and the breathing has improved then discharge should be considered with outpatient review. If needle aspiration fails (defined as > 2 cm or still short of breath) → chest drain should be inserted If a patient with a pneumothorax requires oxygen, this should be given at 10 L/min.

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Pulmonology • Secondary pneumothorax Definition: the patient is ≥50 years old, or has significant smoking history or evidence of underlying lung disease. Management: If the rim of air is ˂ 2cm → aspiration If the rim of air is ≥ 2cm → chest drain → Insert a small-bore chest drain (8-14 FG) and attach to an underwater seal drain If aspiration fails (i.e. pneumothorax is still ˃1cm) → a chest drain should be inserted. if the patient is very dyspneoic a drain should be inserted even though the pneumothorax is small (< 2cm). All patients should be admitted for at least 24 hours High flow oxygen should be given in all cases of pneumothorax, as it facilitates re-absorption of the pleural air, which is predominantly composed of nitrogen. Asthmatics should

be treated as a secondary pneumothorax Tension pneumothorax • should be suspected in people on mechanical ventilators or nasal non-invasive ventilation who suddenly deteriorate, and is frequently missed in the intensive care unit setting. • Treatment → needle thoracocentesis use a 3-6-cm-long cannula to perform needle thoracocentesis. the cannula should be left in place until bubbling is confirmed in the underwaterseal system to confirm proper function of the intercostal tube. If the history and examination are suggestive of a pneumothorax and the patient being relatively stable (tension pneumothorax are not suggested), the most appropriate first step would be → confirmation with chest x ray rather than place a needle or chest drain.

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Chest drains for pneumothorax • Point of insertion→in the 'safe triangle', in the mid-axillary line, above a rib margin • Chest drain situations When the patient coughs, nothing happens. When he breathes in and out, the fluid in the tube moves up and down that means →Air is no longer draining from the pleural space, but the drain is still working. Air is not bubbling out of the drain when the patient coughs because the air has stopped draining from the pleural space and the lung has re-inflated. If a drain does not bubble or swing, then it is blocked or kinked and is not working. • Next step after failure of chest drain Negative suction is necessary if the drain is still bubbling but the lung has not fully re-inflated on the chest X-ray. After chest drain if pneumothorax fails to re-expand or if there is a persistent air leak (bubbling present) after 48 hours, then you should → refer the patient to a respiratory specialist because negative suction might be required using a high-volume/low-pressure suction system. Cardiothoracic surgical referral → Video assisted thoracoscopic surgery indications: persistent pneumothorax despite low-pressure, large-volume suction, and the chest drain in position and is bubbling (may be have a bronchopleural fistula) Persistent air leak (more than five to seven days of drainage) Second ipsilateral pneumothorax for bullectomy and pleurectomy. Bilateral spontaneous pneumothorax Certain occupations, for example, pilots or divers. Chemical pleurodesis through the chest drain: used in older patients or frail individuals with recurrent pneumothorax, where surgery would be high risk. Fitness to fly • Pneumothorax is an absolute contraindication to air travel as trapped air may expand and result in a tension pneumothorax. • In general, it should be safe to travel approximately 1- 2 weeks after successful drainage of a pneumothorax with full expansion of the lung. Diving • The British Thoracic Society (BTS) guidelines state: 'Diving should be permanently avoided unless the patient has undergone bilateral surgical pleurectomy and has normal lung function and chest CT scan postoperatively.'

Chapter 2

Pulmonology Images Chest x ray reveals a 3.2 cm rim of air around the lung.

Pleural effusion Classification , pathophysiology and causes Exudate (> 30g/L protein) Transudate (< 30g/L protein) Pathophysiology ↑ Capillary hydrostatic pressure (increased capillary wedge pressure) ↓ Capillary oncotic pressure Causes • infection: pneumonia, TB, sub-phrenic abscess • connective tissue disease: RA, SLE • neoplasia: lung cancer, mesothelioma, metastases • pancreatitis • pulmonary embolism • Dressler's syndrome • yellow nail syndrome Notes & Notes

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↑ Capillary permeability (e.g., due to inflammation) • heart failure • hypoalbuminaemia liver disease, nephrotic syndrome, malabsorption • hypothyroidism • Meigs' syndrome

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Investigation • Chest x-rays should be performed in all patients • Ultrasound thorax: the next most appropriate step after chest x-ray Ultrasound is better for pleural imaging than CT. it increases the likelihood of successful pleural aspiration and is sensitive for detecting pleural fluid septations • Pleural aspiration ultrasound is recommended to reduce the complication rate a 21G needle and 50ml syringe should be used fluid should be sent for pH, protein, lactate dehydrogenase (LDH), cytology and microbiology • Thoracoscopy the investigation of choice in patients with cytology negative exudative effusions. • Video-assisted thoracoscopic surgery (VATS) A minimally invasive procedure, used if the diagnosis remains unclear Light's criteria • Developed to distinguish between a transudate and an exudate. • The BTS recommend using the criteria for borderline cases: exudates have a protein level of >30 g/L, transudates have a protein level of <30 g/L if the protein level is between 25-35 g/L, Light's criteria should be applied. Exudates Transudate Pleural fluid protein/serum protein ratio

> 0.5 ≤ 0.5 Pleural fluid LDH/serum LDH ratio 0.6 ≤ 0.6 Pleural fluid LDH ⅔the upper limit of normal serum LDH < ⅔the upper limit of normal serum LDH To differentiate exudates from transudates, remember that Exudates have Extra (think protein, LDH). Pleural infection • All patients with a pleural effusion in association with sepsis or a pneumonic illness require diagnostic pleural fluid sampling • Indications for chest tube insertion in patients with an infected pleural effusion are: Frankly purulent pleural fluid Pleural pH < 7.2 in the setting of an infected pleural effusion Presence of organisms on a Gram stain of the pleural fluid Loculated pleural effusions Poor clinical progress despite antibiotic treatment

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Pulmonology • What test can be performed to assess if the effusion is an empyema? Centrifugation of the pleural aspirate If the pleural fluid is turbid or milky it should be centrifuged. If the supernatant (liquid which lies above the sediment) is clear, the turbid fluid was due to cell debris and empyema is likely Characteristic pleural fluid findings • Low glucose Empyema Rheumatoid arthritis effusions (↓glucose, ↓pH < 7.2, ↑ LDH, ↑ cholesterol, ↑RF) Tuberculosis Malignancy Oesophageal rupture Lupus • Raised amylase Pancreatitis, Oesophageal perforation • Heavy blood-staining Mesothelioma, malignancy. Pulmonary embolism Tuberculosis Complications of plural fluid drainage • Re-expansion pulmonary oedema

 This is a potentially life-threatening condition which can occur when a large volume of fluid or air is rapidly drained, It is suggested by sudden onset of shortness of breath, cough and hypoxaemia following chest drain insertion. Exudate typically appears cloudy, has an increased cell count, and has high levels of protein, albumin, and LDH. Transudate is usually clear, has a decreased cell count, and has low levels of protein, albumin, and LDH. MEAT has low glucose: Malignancy, Empyema, Arthritis (rheumatoid pleurisy), and Tuberculosis are causes of pulmonary effusion associated with low glucose levels. Pleural fluid with a bloody appearance suggests a malignant etiology or haemothorax

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Diagnostic algorithm for the patient with a pleural effusion Oxford handbook of respiratory medicine. 3rd edition

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Pulmonology MRCPUK-part-2-march-2018: A patient admitted with severe pneumonia and pleural effusion, despite treatment with Tazocin®. Needle aspiration of 15 ml of pleural fluid reveals it to be pus-coloured, with a pH of 7.1 and a glucose level of 3.1 mmol/l. what is the most important intervention? Chest drain insertion

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Chylothorax Definition • Accumulation of chyle (a fatty lymphatic fluid with milky appearance) in the pleural space. • Chyle is a lymphatic fluid with a high content of triglycerides in the form of chylomicrons, which produce the milky appearance. Causes • Nontraumatic chylothorax : Malignancy (classically lymphoma) is the leading cause. • Traumatic chylothorax: surgical injury to the thoracic duct is the most common cause Features • Symptoms induced by the mechanical effects of a pleural effusion Diagnosis • Pleural fluid for triglyceride and cholesterol levels: elevated triglyceride strongly supports the diagnosis. Low cholesterol will differentiate chylothorax from cholesterol pleural effusion (Pseudochyle → low triglyceride , high cholesterol and empyema) Milky appearance is a classic sign of chylothorax Pleural fluid is classically exudative with a high lymphocyte count (>70 %), a normal glucose level, a low LDH, and a low cholesterol level). Detection of chylomicrons by lipoprotein electrophoresis is the definitive diagnostic test but not routinely performed

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Haemothorax Definition • Bleeding into the pleural space Causes of nontraumatic haemothorax • Most common: spontaneous pneumothorax • Less common Vascular disease Malignancy Meigs syndrome: A triad of ascites, right pleural effusion, and benign ovarian tumor

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 Coagulation disorders Necrotizing pneumonia Diagnosis • Pleural fluid analysis Bloody appearance RBC count > 5,000 cells/ml haematocrit that is more than half that of peripheral

blood. (Haematocrit > 0.5 × peripheral hematocrit). This distinguishes it from a blood-stained effusion. Management • The treatment of choice is to insert a large intercostal drain (28-32 F). If this reveals continued bleeding, a thoracotomy might be required. A hemothorax, however small, must always be drained because blood in the pleural cavity will clot if not evacuated, resulting in a trapped lung or an empyema.

Eosinophilic Pulmonary Diseases Definition • Eosinophilic pulmonary diseases are a heterogeneous group of disorders characterized by the accumulation of eosinophils in alveolar spaces, the interstitium, or both. Causes of pulmonary eosinophilia • Known etiology Allergic bronchopulmonary aspergillosis (ABPA) Helminth infections Drug-induced pneumonitis (eg, antibiotics, phenytoin, or L-tryptophan) Eosinophilic granulomatosis with polyangiitis (previously referred to as ChurgStrauss syndrome) Loffler's syndrome Tropical pulmonary eosinophilia • Unknown etiology: The two primary eosinophilic pulmonary diseases of unknown etiology are Acute eosinophilic pneumonia Chronic eosinophilic pneumonia Diagnosis based on:

1. Demonstration of opacities on chest imaging and
2. Identification of eosinophilia in peripheral blood, bronchoalveolar lavage fluid, or lung biopsy tissue

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Pulmonology Acute eosinophilic pneumonia Definition • Chronic eosinophilic pneumonia is an idiopathic acute disorder characterized by an abnormal accumulation of eosinophils in the interstitial and alveolar spaces of the lung. Features • Acute eosinophilic pneumonia is an acute febrile illness of less than four weeks duration (often less than seven days), a nonproductive cough, and progressively worsening dyspnea. • malaise, myalgias, night sweats, and pleuritic chest pain. Association • new onset or resumption of cigarette smoking. Diagnosis based on: • Acute febrile illness of short duration (one month or less), • hypoxemic respiratory failure, • diffuse pulmonary opacities on chest radiograph, and • bronchoalveolar lavage eosinophilia (>25 %), after • exclusion of infection, vasculitis, or other known precipitants (eg, drugs, irradiation) Treatment • In severe hypoxemia or respiratory failure requiring mechanical ventilation → methylprednisolone • Mild to moderate (eg, spo2 >92 %) →oral prednisone The classic presentation of idiopathic acute eosinophilic pneumonia is the rapid onset of acute respiratory failure in a previously healthy patient. diffuse radiographic opacities, and bronchoalveolar lavage with ≥25 % eosinophils, and absence of infection or other known precipitant.

Chronic eosinophilic pneumonia Definition • Chronic eosinophilic pneumonia is an idiopathic chronic disorder characterized by an abnormal accumulation of eosinophils in the interstitial and alveolar spaces of the lung. Feature • dyspnea, cough, fever, and wheezing over 4 weeks to several months. Diagnosis • Chest imaging shows predominantly peripheral or pleural-based opacities described as the "photographic negative" of pulmonary edema, are virtually pathognomonic • Bronchoalveolar lavage (BAL)
 To look for eosinophilia →cell count showing eosinophilia (≥25 %). To exclude infection. • Infections and drug-induced pulmonary eosinophilia need to be excluded. Treatment • Prednisolone

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Tropical pulmonary eosinophilia Definition • Tropical pulmonary eosinophilia is an immune hyper-responsiveness to microfilariae that become trapped in the lungs. It is a clinical manifestation of lymphatic filariasis, a parasitic infection caused by nematodes (roundworms) such as Wuchereria bancrofti. Epidemiology • Seen in endemic areas of lymphatic filariasis (mainly India and South East Asia) • Occurs more frequently in males than in females Features • Dry cough that is frequently paroxysmal and nocturnal. • Asthma-like attacks → wheezing • fatigue, malaise, and weight loss, Diagnosis • ↑blood eosinophils • ↑serum immunoglobulin E. • ↑ filarial antibody titers (confirmatory test) Differential diagnosis • Tropical pulmonary eosinophilia is distinguished from Loeffler's syndrome by A. the severe and protracted course, B. measurable antibodies against filarial antigens, and C. the therapeutic response to diethylcarbamazine. D. If treated late or left untreated, it can lead to pulmonary fibrosis with chronic respiratory failure. Treatment • Diethylcarbamazine for 12 to 21 days. • Bronchospasm can be managed with bronchodilators and short-term corticosteroids. The diagnostic criteria for tropical pulmonary eosinophilia include:

1. history of residence or travel to a filarial endemic region,
2. paroxysmal nocturnal cough with dyspnoea,
3. leucocytosis with peripheral blood eosinophilia >3,000/microL,
4. elevated serum IgE and an antifilarial antibodies (IgG and IgE) levels,
5. pulmonary infiltrations in chest x-ray, and
6. clinical improvement with DEC (diethylcarbamazine).

Loffler's syndrome Definition • Löffler syndrome is a form of eosinophilic pulmonary disease characterized by absent or mild respiratory symptoms (most often dry cough), transient CXR shadowing and blood eosinophilia. thought to be due to parasites such as Ascaris lumbricoides (the most common parasite) causing an alveolar reaction. Features • Fever, cough and night sweats which often last for less than 2 weeks. Diagnosis based on • Characteristic and often transient respiratory symptoms • Chest x-ray findings → fleeting migratory pulmonary opacities • Peripheral blood eosinophilia. • Exclusion of other types of eosinophilic lung disease (e.g. acute eosinophilic pneumonia → severe hypoxemia, and typically a lack of increased blood eosinophils at the onset of disease). Treatment • Symptomatic and may consist of corticosteroids. • Generally, a self-limiting disease, usually resolves within 1 month.

Cryptogenic organising pneumonia (COP) Definition • A rare type of inflammatory interstitial lung disease, characterised by a buds of granulation tissue in the alveoli and bronchioles on histopathology • other names: Bronchiolitis obliterans organising pneumonia (BOOP) Causes • Idiopathic: most common 'cryptogenic means unknown cause'. • Secondary organising pneumonia:

connective tissue disease, malignancy, infection, drugs and toxins Epidemiology • Typically, age of onset is 50 to 60 years • Men and women affected equally. Feature • Mimic community-acquired pneumonia (eg, cough, dyspnea with exertion, weight loss). Investigations • Chest X-ray: bilateral patchy infiltrates • Chest CT: multiple ground-glass or consolidative opacities that tend to be at the lung periphery

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 Reversed halo sign, better known as an atoll sign (a region of ground-glass opacity surrounded by crescentic or annular denser tissue) . • Pulmonary function tests (PFTs) →restrictive pattern Diagnosis • Exclusion of any possible cause (e.g. COVID-19 → do PCR) • For definitive diagnosis → lung biopsy → excessive proliferation or "plugs" of granulation tissue within alveolar ducts and alveoli (Masson bodies). Granulation tissue extends uniformly into the alveolar ducts and does not distort pulmonary architecture, unlike usual interstitial pneumonia. If clinical presentation, radiographic appearance and bronchoscopy with bronchoalveolar lavage is consistent with COP, surgical lung biopsy is not required for diagnosis. Treatment • 1st line: Prednisolone (usually effective) • 2nd line: cyclophosphamide or azathioprine Prognosis • Relapse is common Cryptogenic organizing pneumonia (COP) • High-resolution CT of the chest → bilateral ground-glass opacities • Exclude other possible causes • Persistent pulmonary opacities despite antibiotic treatment. • Lung biopsy → granulation tissue plugs in small airways Cryptogenic Organizing Pneumonia/https/ncbi.nlm.nih.gov/

Pulmonary hypertension (PH) Definition • Sustained elevation in mean pulmonary arterial pressure of greater than 25 mmHg at rest or 30 mmHg after exercise, pulmonary artery wedge pressure ≤15 mmHg and pulmonary vascular resistance > 3 Wood units. Epidemiology • more commonly affects female Pathophysiology : Increased pulmonary vascular resistance • Occlusive vasculopathy (e.g., PE, connective tissue diseases) • Hypoxic pulmonary vasoconstriction: chronic hypoxic pulmonary vasoconstriction → airway smooth muscle hypertrophy and pulmonary vascular bed destruction → ↑ pulmonary vascular resistance • Inflammation (e.g., COPD) → ↑ inflammatory cell infiltration of intima → thickened endothelial wall → intimal fibrosis • ↑ Increased pulmonary vessel pressure: due to left heart dysfunction • ↑ endothelin and ↓ vasodilators (e.g., NO, prostacyclin) → vasoconstriction

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Pulmonology Causes according to WHO Classification • Group 1: Pulmonary arterial hypertension (PAH), Idiopathic, familial collagen vascular disease, HIV, sickle cell disease drugs and toxins → e.g. amphetamines, cocaine (but not heroin). • Group 2: Pulmonary hypertension with left heart disease • Group 3: Pulmonary hypertension secondary to lung disease/hypoxia COPD, interstitial lung disease, sleep apnoea, high altitude • Group 4: Pulmonary hypertension due to thromboembolic disease • Group 5: Miscellaneous conditions: lymphangiomatosis e.g. secondary to carcinomatosis or sarcoidosis Features • Exertional dyspnoea is the most frequent symptom • Symptoms of right ventricular (RV) failure (eg, exertional chest pain or syncope, loud P2, elevated

jugular venous pressure, right-sided murmurs, edema, right upper quadrant pain, ascites, and pleural effusion) Investigations • Transthoracic echocardiography is the initial test of choice If left heart disease (LHD) explain the PH →RHC is not indicated. If no LHD explain the PH → investigate for pulmonary causes Chest CT Pulmonary function testing (PFTs) ventilation-perfusion (V/Q) scanning → chronic thromboembolic disease Obstructive sleep apnoea Autoimmune serologies HIV serology If pulmonary dysfunction explain the PH → RHC is not indicated. If pulmonary investigations did not explain the PH → Do RHC • Right heart catheterization (LHC) is the best investigation for diagnosing pulmonary hypertension mostly for patients with no cardiac or respiratory causes explaining the PH → to evaluate for PAH. The diagnosis of primary pulmonary hypertension (PAH) requires RHC that demonstrates mean pulmonary artery pressure (mPAP) >20 mmHg at rest, pulmonary vascular resistance (PVR) ≥3 Wood units, and a mean pulmonary capillary wedge pressure (PCWP) <15 mmHg. a mPAP ≥20 mmHg, PCWP ≥15 mmHg, and a normal or reduced cardiac output is consistent with left heart disease- pulmonary hypertension (LHD-PH). • Pulmonary angiography is the definitive diagnostic test. Management • First step: Treat any underlying conditions, for example with anticoagulants or oxygen. • Second step: perform acute vasodilator testing to decide which patients show a significant fall in pulmonary arterial pressure following the administration of vasodilators such as intravenous epoprostenol or inhaled nitric oxide If there is a positive response to acute vasodilator testing → oral calcium channel blockers (nifedipine or extended-release diltiazem) If there is a negative response to acute vasodilator testing: endothelin receptor antagonists: bosentan

 phosphodiesterase inhibitors: sildenafil prostacyclin analogues: treprostinil, iloprost Complication • Cor pulmonale (right ventricular failure). Prognosis • Pregnant with pulmonary hypertension have a high mortality of 30% - 50% - highest immediately after delivery.

Sarcoidosis Definition • Sarcoidosis is a multisystem disorder of unknown aetiology characterised by non-caseating granulomas. Epidemiology • More common in black people (African descent) and subjects of Caribbean origin in Europe, sarcoid is commonest amongst Caucasians and has a significantly higher incidence in the Irish. • More common among females than males ♀ > ♂ (2:1) • Typically affects young adults. • More common in non-smokers Pathology • Noncaseating granulomas in the organ involved. the characteristic pathological feature of sarcoidosis. may occur anywhere The central area of the granuloma will occasionally contain a Schaumann body, formed of crystallised material (calcium phosphate). These granulomas have the capacity to produce 1,25 vitamin D explaining the associated hypercalcaemia. Features • Often asymptomatic in the early stages (≈ 50%) → incidental chest x ray finding. • Enlargement of lymph nodes The most common physical exam finding • Pulmonary (most common) Dry cough Dyspnoea (Pulmonary fibrosis) • Extrapulmonary Sarcoidosis CXR • 1 = BHL • 2 = BHL + infiltrates • 3 = infiltrates • 4 = fibrosis

Pulmonology Skin lesions: seen in 25 %, often an early finding. Erythema nodosum: tender erythematous nodules on the lower extremities and is a predictor of a good prognosis. lupus pernio: indurated plaques with discoloration of the nose, cheeks, lips, and ears. It is a predictor of a poor prognosis. Arthralgia: typically targets the ankle joint. Uveitis (25% of cases): red, painful eyes and blurred vision Neurologic: (5% of cases) → Cranial nerves (e.g. facial nerve or Bell palsy). Parotid swelling: (5% of cases) Löfgren syndrome (LS): a combination of erythema nodosum (EN), hilar adenopathy, migratory polyarthralgia, and fever → has 95 % specificity for sarcoidosis. Hypercalcaemia: (10% of cases) → nephrocalcinosis and nephrolithiasis. Systemic symptoms Fever weight loss Investigations Sarcoidosis is a diagnosis of exclusion of granulomatous lung diseases, including tuberculosis and histoplasmosis. Occupational history should be taken to exclude both berylliosis and silicosis which can present in a similar manner to sarcoidosis. • Chest x ray Best initial test abnormal in 85% of lung sarcoid may show: bilateral hilar lymphadenopathy. Lung fibrosis typically affects the upper zones • CT scan: If they have typical findings on a radiograph with a typical clinical presentation (eg, in the context of Lofgren's disease) then a CT scan may not be necessary It is the best next step after chest x ray in atypical presentation. demonstrate the degree of fibrosis, micronodules in a subpleural or bronchoalveolar distribution, fissure nodularity and bronchial distortion.
 Irregular linear opacities and ground-glass shadowing may also be seen.
 If the CT scan is diagnostic, then mediastinoscopy, bronchoscopy or biopsies can often be avoided. • Tissue biopsy →Non-caseating granulomas The gold standard test
 If the history and radiology is typical, the biopsy is not necessary. With less characteristic presentations, positive biopsies are needed. If you are asked to specify the investigation most likely to confirm the diagnosis, only transbronchial biopsy will determine whether non-caseating granulomas are present or not →Transbronchial lung biopsy is therefore the diagnostic investigation of choice. Skin biopsy for skin lesions • Routine blood tests CBC: Leukopenia in 5-10% of patients ESR → Elevated

 Creatinine → elevated in renal involvement Electrolytes → Hypercalcaemia (Seen only in 10% if patients) produced by macrophages within the granulomas ↑1-alpha-hydroxylase → activates vitamin D → ↑Ca Hypergammaglobulinaemia (↑ Immunoglobulins) in 30-80%. • Exclusion of granulomatous lung diseases TB should be excluded by sending sputum or BAL washings for AFB A positive tuberculin test in a patient with chronic sarcoidosis is suggestive of active tuberculosis Occupational history to exclude both berylliosis and silicosis • Lung parenchyma involvement → Spirometry usually shows a restrictive defect (Decreased gas-transfer factor (Tlco) with decreased gas-transfer coefficient (Kco) • Cardiac involvement ECG: cardiac sarcoidosis (e.g. heart block →prolonged PR interval) Abnormalities in ECG or echocardiogram which suggest cardiac sarcoidosis should be confirmed with cardiac magnetic resonance imaging (CMR) or positron emission tomography (PET). • CNS involvement → CSF: Intrathecal oligoclonal band production, elevated protein and lymphocytosis • Broncho-alveolar lavage typically shows a lymphocytosis increased CD4+/CD8+ ratio • ACE levels have a sensitivity of 60% and specificity of 70% and are therefore not reliable in the diagnosis of sarcoidosis although they may have a role in monitoring disease activity. • Kveim test (where part of the spleen from a patient with known sarcoidosis is injected under the skin) is no longer performed due to concerns about

cross-infection

Chapter 2

Pulmonology Staging of chronic sarcoidosis Stage Finding Likelihood of spontaneous resolution

Normal chest radiograph

> 90% I Bilateral hilar lymphadenopathy (BHL) II BHL plus pulmonary infiltrates 40-60% III Pulmonary infiltrates (no BHL) 10-20% IV Pulmonary fibrosis (+/- bullae) <20% Notes & Notes for MRCP
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Chest x-ray and CT scan showing stage 2 sarcoidosis with both bilateral hilar lymphadenopathy + interstitial infiltrates. The reticulonodular opacities are particularly noted in the upper zones. Remember that pulmonary fibrosis (which this case has not yet progressed to) may be divided into conditions which predominately affect the upper zones and those which predominately affect the lower zones - sarcoidosis is one of the former. The CT of the chest demonstrates diffuse areas of nodularity predominantly in a peribronchial distribution with patchy areas of consolidation particularly in the upper lobes. There is some surrounding ground glass opacities. No gross reticular changes to suggest fibrosis. 60-90%

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Differential diagnosis of bilateral hilar lymphadenopathy • Sarcoidosis • Tuberculosis • Malignancy including lymphoma • Cystic fibrosis • Churg Strauss disease • HIV • Extrinsic allergic alveolitis • Phenytoin • Pneumoconiosis, especially berylliosis. Exposure to beryllium is seen in the nuclear power, telecommunications, semi-conductor and electronics industries. Management The majority of patients with sarcoidosis get better without treatment • Mild disease (Patients with asymptomatic and stable stage 2 or 3 disease who have only mildly abnormal lung function) →NO treatment Sarcoidosis remits without treatment in approximately two-thirds of people • Moderate to severe disease First-line → Prednisolone. Indications for steroids: patients with chest x-ray stage 2 or 3 disease who have moderate to severe or progressive symptoms. Systemic involvement: hypercalcaemia, eye, heart or neuro involvement Second-line: Methotrexate is the first choice of second-line agent. Third-line: Infliximab given in combination with methotrexate or azathioprine Lung transplantation should be considered in all patients with advanced pulmonary fibrosis and associated pulmonary hypertension. Prognosis Erythema nodosum is associated with a good prognosis in sarcoidosis. • Factors associated with a good prognosis HLA B8 Lofgren's syndrome (bilateral hilar lymphadenopathy, erythema nodosum, polyarthritis and fever). • Factors associated with poor prognosis insidious onset, symptoms > 6 months (chronic pulmonary involvement) absence of erythema nodosum extrapulmonary manifestations: e.g. lupus pernio: a chronic raised indurated (hardened) lesion of the skin, often purplish in colour, and is associated with sarcoid. Splenomegaly Cardiac involvement: Cardiac sarcoidosis is rare but

can manifest as a prolonged PR interval. Chronic hypercalcaemia Nasal mucosal involvement Neurosarcoidosis CXR: stage III-IV features black people (Afro-Caribbean or Afro-American race) Age of onset >40 years

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Pulmonology

Lofgren's syndrome Lofgren's syndrome: a variant of sarcoidosis with acute clinical presentation with tetrad of:

1. Migratory polyarthritis (acute arthritis), most commonly involves ankles (>90 %).
2. Erythema nodosum.
3. Bilateral hilar lymphadenopathy.
4. Fever. Overview • Seen in less than 5 -10 % of sarcoidosis • Typically, more common in Scandinavian patients and less common in Afro-Caribbean patients • Typically occurs in young females • Carries an excellent prognosis • Usually self-limiting Other sarcoidosis variants Heerfordt syndrome : a variant of sarcoidosis with chronic clinical presentation with tetrad of:
5. Parotitis
6. Uveitis
7. Facial palsy
8. Fever

Yellow nail syndrome Definition • Yellow nail syndrome is an uncommon disorder characterized by the triad of pulmonary disease, lymphedema, and yellow nails Features • Nails are yellow, thickened, curved, with loss of the lunula and cuticle. and may become detached from the nail bed. • Congenital lymphoedema • Pulmonary disease (bronchiectasis, pleural effusions) • Chronic sinusitis

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Hepatopulmonary syndrome (HPS) Definition • oxygenation defect induced by pulmonary vascular dilatation in patients with liver cirrhosis or portal hypertension. Mechanism • The vascular dilatation is thought to be induced by increased pulmonary levels of nitric oxide. Prevalence • It is seen in 15-30% of patients with cirrhosis. Features • Dyspnoea • Platypnoea (dyspnoea whilst standing) and Orthodeoxia (hypoxaemia exacerbated by being upright) are characteristic Due to the predominance of vascular dilatation in the lung bases. Blood flow to these areas is increased in the upright position. Hepatic disease →intrapulmonary vasodilatation mainly in the lower Lobes →rightto-left shunting (similar to pulmonary arteriovenous malformations) →increased blood flow through the lower lobes when the patient moves from the supine to the erect position →blood from the lower lobes, which is more poorly oxygenated, entering the left side of the heart → oxygen

desaturation in the erect position. Investigations • The diagnosis of HPS can only be made in a patient who has liver disease, impaired oxygenation, and intrapulmonary shunt when other etiologies have been excluded. • Contrast-enhanced transthoracic echocardiography is the best test to demonstrate intrapulmonary vascular dilatation. It can also exclude intracardiac shunting which may result in similar signs and symptoms to hepatopulmonary syndrome.

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Pulmonology Method performed by injecting agitated saline intravenously during transthoracic echocardiography. Interpretation In a normal subject microbubbles are visualised in the right ventricle within seconds, which are then absorbed in the alveoli. Immediate visualization in the left ventricle (within three cardiac cycles) indicates intracardiac shunting. Delayed visualisation in the left ventricle (3-6 cardiac cycles) is diagnostic of intrapulmonary shunting. • Impaired oxygenation is confirmed when an arterial blood gas analysis demonstrates an alveolar-arterial (A-a) oxygen gradient ≥15 mmHg or an arterial oxygen tension (PaO2) <80 mmHg (10.7 kPa) • Chest imaging and pulmonary function testing are often normal Treatment • Liver transplantation is the only proven beneficial available treatment, with 85% of patients showing resolution or significant improvement in gas exchange postoperatively. Prognosis • It is a poor prognostic indicator.

Pulmonary alveolar microlithiasis (PAM) Definition • Pulmonary alveolar microlithiasis (PAM) is a rare, autosomal recessive disorder, characterized by widespread deposition of calcium phosphate microliths throughout the lungs. Epidemiology • PAM has the highest prevalence in Turkey, Japan, and Italy Pathophysiology • It occurs in the absence of disorders of calcium metabolism. • SLC34A2 gene mutations →↓ activity of the type IIb sodium-phosphate cotransporter (which located mainly in alveolar type II cells) →accumulation of phosphate in the alveoli →formation of microliths • SLC34A2 gene is responsible for the uptake of phosphate released from phospholipids in outdated surfactant. Feature • Most patients are asymptomatic despite striking radiological abnormalities. often found incidentally during imaging studies for another reason. • Symptoms included dyspnea, nonproductive cough, chest pain

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Diagnosis • Chest x-ray: 'sandstorm-appearing' is a typical diagnostic finding (diffuse scattered micronodules, often obscuring the contours of the heart and diaphragm) • HRCT: micronodular calcifications, diffuse ground glass opacities • Bronchoalveolar lavage (BAL) and transbronchial biopsy can be useful if the diagnosis is uncertain. BAL and biopsy show the characteristic calciospherocitis (microliths) in the alveoli (deposition of calcium and phosphate crystals). Treatment • There is no established therapy for PAM. • lung transplantation is the only effective therapy.

Pulmonary Alveolar Proteinosis (PAP) Definition: A rare diffuse lung disease characterized by the progressive accumulation of surfactant protein in the alveoli, that characteristically stain for periodic acid-Schiff (PAS) Epidemiology: Common in males (M: F = 4:1) , the typical age at

presentation is 40 to 50 years. Pathophysiology: ↓ alveolar macrophages → ↓ ability to remove surfactant → ↑surfactant accumulation in the alveoli. Causes • Autoimmune: due to granulocyte macrophage-colony stimulating factor (GM-CSF) antibodies, the most common • congenital • Secondary: chronic infections, immunosuppressants, organic dusts, malignancies. Feature • progressive dyspnea, cough, sputum production, fatigue, and weight loss Diagnosis • Chest x-ray: bilateral symmetric alveolar opacities located centrally in mid and lower lung zones, sometimes resulting in a "bat wing" distribution. • High resolution computed tomography (HRCT): ground-glass opacification that typically spares the periphery and may have a "crazy-paving" appearance due to thickening of the interlobular and intralobular septa. • Spirometry → shows a restrictive pattern (↓lung capacity, ↓CO diffusion) • Autoantibodies : ↑ autoantibody against GM-CSF in serum and BAL fluid • Flexible bronchoscopy with broncho-alveolar lavage (BAL) The standard diagnostic test PAS-positive stains Treatment • Whole lung lavage: excess surfactant is removed from the lungs via saline solution; may require repeated application • Treatment of the underlying condition

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Pulmonology

Carbon monoxide poisoning Confusion, pyrexia and pink mucosae are typical features of carbon monoxide poisoning

Epidemiology • Carbon monoxide is the commonest cause of poisoning-associated death in UK Causes • House fires • Wood-burning stoves • Furnaces in enclosed and poorly ventilated spaces. Often involves multiple individuals (e.g., family) during the winter • Fumes from cleaning fluids and paint removers that contain methylene chloride (dichloromethane) can also cause carbon monoxide poisoning. When breathed in, methylene chloride is converted into CO gas. Pathophysiology • The affinity of hemoglobin for CO is ~240 times stronger than for O2 → formation of COHb (carboxyhemoglobin)→ ↓ oxygen-carrying capacity of hemoglobin → tissue hypoxia • COHb →Shift the O2 dissociation curve to the left → ↑ affinity for O2 → ↓ release of O2 in tissue Questions may hint at badly maintained housing e.g. student houses Features of carbon monoxide toxicity • Nonspecific symptoms Headache: the most common symptom ≈ 90% of cases Dizziness Fatigue Nausea/vomiting • Neurotoxicity Altered mental status (e.g., agitation, confusion, somnolence, memory loss) Seizures Loss of consciousness/coma Cerebellar signs are the most reliable indicator of significant neurological toxicity • Cardiorespiratory toxicity Inhalation of hot smoke →upper airways burn →mucosal swelling → Bronchoscopy is the best tool to establish whether there is significant oedema or mucosal ulceration obstructing the airways. hypertension, tachycardia Shock • COHb levels have prognostic implications, which are summarised here:

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 < 30% cause only headache and dizziness 40–60% produces syncope, tachypnoea, tachycardia and fits 60% cause an increasing risk of cardiorespiratory failure and death. Suspect carbon monoxide poisoning when multiple people from the same confined household complain of the

headache and fatigue. Diagnosis • Arterial blood-gas analysis Typical carboxyhaemoglobin (COHb) levels: < 3% non-smokers < 10% smokers 10 - 30% symptomatic: headache, vomiting ˃30% severe toxicity PaO2: usually appears normal • Direct spectrophotometric measurement of Carboxyhaemoglobin (COHb) in a bloodgas analyser is the gold standard. A bedside HbCO oximeter is now available • ECG and cardiac monitor for all patients for 4 – 6 hours → signs of myocardial ischemia; arrhythmias Pulse oximeters cannot distinguish between COHb and HbO2. Pulse oximetry appears normal because carboxyhaemoglobin has similar absorption spectra to oxyhaemoglobin. Management • First-line: 100% oxygen → Give high-flow oxygen (12 l/min) via a tight-fitting mask without a re-breathing circuit • Second-line: hyperbaric oxygen shorten the washout of COHb, but access and transfer times to a hyperbaric chamber can make this not practical. Indications for hyperbaric oxygen CO level >25 % Loss of consciousness Severe metabolic acidosis (pH <7.1) Evidence of end-organ ischemia (eg, ECG changes, chest pain, altered mental status) pregnancy • In severe cases intubation and mechanical ventilation may be required

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