COPD and bronchiectasis

Objectives:

- Definition of COPD and Bronchiectasis.
- Clinical and radiological diagnosis.
- Differential diagnosis.
- General outline of management.
- Create a link to 341 clinical teaching.

[Color index : Important | Notes | Extra]

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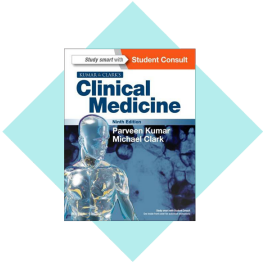
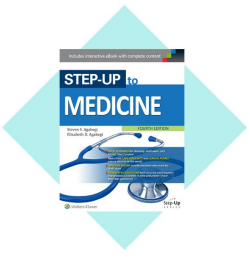
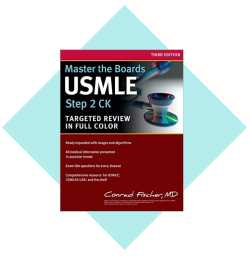
Resources:

- 435 slides, 434 & 432 teams, 435 respiratory block team.







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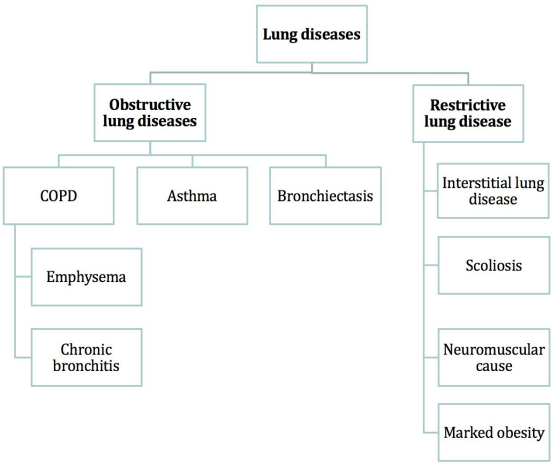
"Medicine is an art, nobody can deny it."

Definition of COPD

- **COPD:** 12 : 18 minutes
  - Is a disease state characterized by **persistent airflow limitation** that is **not fully** reversible (may improve slightly with salbutamol but doesn't normalize). The airflow limitation is usually both **progressive** and associated with an **abnormal inflammatory response** of the lungs to noxious particles (like smoking, occupational gasses, cooking gasses) or gases.
  - **COPD** is classified under obstructive pulmonary diseases along with other diseases such as asthma and bronchiectasis.
  - Related diagnoses include: **emphysema** (pink puffers) and **chronic bronchitis** (blue bloaters):
    - They are grouped together under COPD because they are both mainly caused by **smoking** (tobacco destroys elastin fibers)[1] and they **usually present together.** [2]
    - **Pure** emphysema or **pure** chronic bronchitis are **rare**.



- **Epidemiology and etiology:**
  - The prevalence of COPD is directly related to the prevalence of **tobacco smoking**[3] " developed country causes **90%** of the cases" and, in low- and middle- income countries " developing" , the use of **biomass fuels.**
  - COPD is the fourth leading cause of death in the US.
  - **by 2020**, it is forecast to represent the **third** most important cause of death worldwide.
- Usually after the age of 40, unlike asthma which can develop at any age.

- **Chronic bronchitis: " defined by clinical features"**

Chronic or recurrent expectoration **(cough up lots of sputum[4])** which is present on most days for a **minimum of 3 months a year for at least 2 successive years. (this is a warning)*.**

*(it is a clinical syndrome in which Ct scan can be normal , spirometry can be normal but there's cough because irritation of small bronchi " which is called simple chronic bronchitis " ,later on develop to COPD or sometimes become dyspneic without going through Chronic bronchitis (but the majority develop COPD) .

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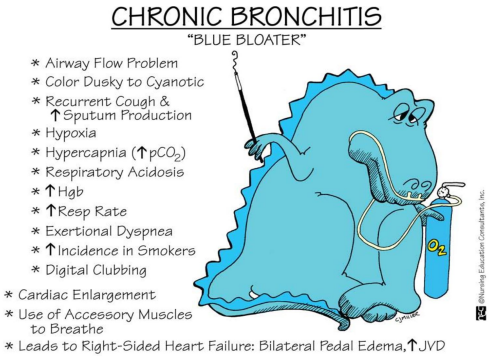
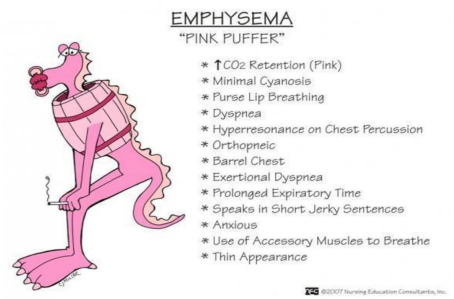


[1] Loss of elastic recoil of the lung leads to SOB..
[2] " In practice, these phenotypes often overlap"
[3] The development of COPD is proportional to the number of cigarettes smoked per day " 30 C/day → ↑ 20 time than non smoker". **But not all smokers develop the condition**, suggesting that individual susceptibility factors are important.
[4] **mucus plugs cause obstruction of bronchioles→ COPD**

- **Emphysema: "defined by structural changes"**

It is a pathologic diagnosis of **Permanent destructive enlargement** of airspaces distal to the terminal bronchioles **(ALVEOLI)** without obvious fibrosis.

- ***Secondary*** to inflammation destruction of alveolar walls.
- emphysema classified according to the site of damage:



10:07 minutes

centri-acinar emphysema	Pan-acinar emphysema	Irregular emphysema
extremely common: (in smoker) • Distension and damage of lung tissue is **concentrated around the respiratory bronchioles** (proximal acini) **,** whilst the more distal alveolar ducts and alveoli tend to be well preserved • Predilection for **upper lung zones .**	less common: • Distension and destruction appear to involve the **whole of the acinus** (both proximal and distal acini) •Predilection for lung **bases**. • inherited deficiency of alpha 1 antitrypsin [5]→ emphysema at a younger age	There is scarring and damage affecting the lung parenchyma **patchily** without particular regard for acinar structure.

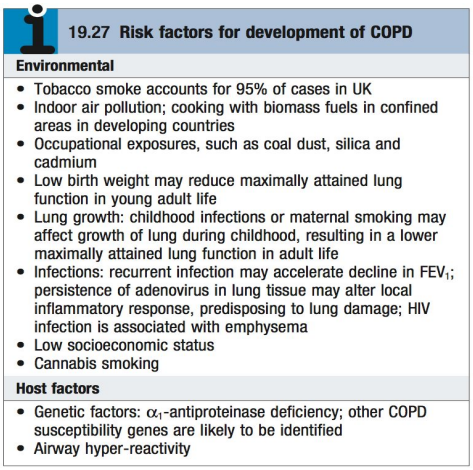
- **COPD systemic components:**

- Incorporates the extrapulmonary manifestations which contribute to pt morbidity or even mortality.
- Muscle weakness and Cachexia inflammatory markers "C reactive protein , TNF, IL.." → catabolic effect on both muscle and bone → look like cancer pt.
- Cardiac deconditioning
- Impaired salt and water excretion → peripheral edema
- Weight loss
- Osteoporosis poor mobility and inflammation (activate osteoclast & suppress osteoblast)
- Depression suffering from a self inflicted disease
- Social isolation cerebritis from inflammatory markers
- COPD leads to chronic respiratory acidosis with metabolic alkalosis as compensation.

[5] **In emphysema We have defense mechanisms to fight these inflammatory mediators "elastases" that is (alpha1 antitrypsin). However, the amount of inflammatory mediators exceeds our ability to counteract them**

- ## Risk factors:

1. Tobacco smoke (in 90% of COPD cases)
2. α 1-antitrypsin deficiency (risk is worse in combination with smoking)
3. Environmental factors e.g. second hand smoking
4. Chronic asthma[6].
5. See the other risk factors " davidson's p 647 ".



19.27 Risk factors for development of COPD

Environmental

- Tobacco smoke accounts for 95% of cases in UK
- Indoor air pollution; cooking with biomass fuels in confined areas in developing countries
- Occupational exposures, such as coal dust, silica and cadmium
- Low birth weight may reduce maximally attained lung function in young adult life
- Lung growth: childhood infections or maternal smoking may affect growth of lung during childhood, resulting in a lower maximally attained lung function in adult life
- Infections: recurrent infection may accelerate decline in FEV_1; persistence of adenovirus in lung tissue may alter local inflammatory response, predisposing to lung damage; HIV infection is associated with emphysema
- Low socioeconomic status
- Cannabis smoking

Host factors

- Genetic factors: α_1-antiproteinase deficiency; other COPD susceptibility genes are likely to be identified
- Airway hyper-reactivity

- ## Pathophysiology:

- In COPD there is an **airflow limitation** in the **small airways** caused by three mechanisms:
  - **Loss of elasticity** and alveolar attachments of airways due to *emphysema*. This reduces the elastic recoil and the **airways collapse during expiration.**
  - **Inflammation and scarring** cause the small airways to narrow.
  - **Mucus secretion** which blocks the airways.

  Each mechanism narrows the small airways and causes air trapping leading to **hyperinflation of the lungs and breathlessness**.
  - See davidson's p 673-674, kumar p 812-813.
  - **Very helpful Summary** here.

- ## Clinical features:

symptoms	signs[7]
- cough[8] with white or clear sputum[9] **- wheeze[10] and breathlessness[11]** here - Tachypnea and tachycardia - Cyanosis - Prolonged expiratory time - Use of accessory respiratory muscles - Signs of cor pulmonale. - oedema[12] and morning headaches[13]	**1. On auscultation:** a. End expiratory wheezes b. Decreased breath sounds "typically quiet " c. +/- Inspiratory crackles[14] **2. On percussion:** a. **Hyperresonance** (as lungs are hyperinflated with air) **3. Non specific signs and symptoms:** **a. Clubbing of fingers is not associated with COPD** b. Must exclude lung cancer or fibrosis if clubbing is present.

Note: Finger clubbing is not a feature of COPD and should trigger further investigation for lung cancer, bronchiectasis or fibrosis.

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[6] Asthma cause obstruction but doesn't destroy the alveoli no reduction in Dlco

[7] are nonspecific, correlate poorly with lung function, and are seldom obvious until the disease is advanced davidson's p 647

[8] Cough with or without hemoptysis.

[9] **If infective exacerbations occur, giving purulent sputum.**

[10] indicates obstructive lung disease(bilateral bronchiectasis _asthma _COPD)

[11] Some patients have sedentary lifestyle but few complaints. The may avoid exertional dyspnea, which is the **most common early symptom of COPD** + **patients who become insensitive to C02 are often edematous and cyanosed but not particularly breathlessness.**
(Differential: chronic asthma, TB, bronchiectasis & CHF).

[12] usually relates to failure of salt and water excretion by the hypoxic hypercapnic kidney

[13] **May suggest hypercapnia**

[14] may accompany infection but, if persistent, raise the possibility of bronchiectasis

How to differentiate between:

chronic bronchitis, emphysema, and COPD

1. **Chronic bronchitis**:
 is a clinical diagnosis of increased sputum over a period of time,
2. **Emphysema**;
is a pathological diagnosis which is defined by loss of alveolar capillaries
3. **COPD:**
 is a clinical syndrome which requires a history of smoking, wheezing, and spirometry (decreased FEV1/FVC ratio)

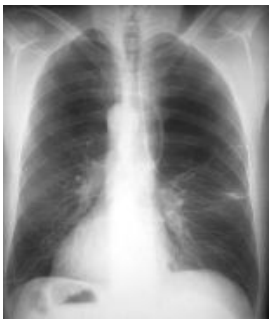
Asthma and COPD

1. Age of onset
2. History of atopy: present with asthma.
3. Eosinophilia and IgE: present with atopic asthma.
4. Bullae with COPD not with asthma
5. Low FVC (with both asthma & COPD)
6. Chronic respiratory failure: chronic obstruction of the lung (with COPD) may lead to respiratory failure. While asthma doesn't cause respiratory failure (yes asthma cause dyspnea during the attack but not causing respiratory failure or damage to alveoli)
7. Diffusing capacity: in asthma is normal bc the alveoli is not involved
8. Trial of inhaled corticosteroids: in atopic asthma if u give corticosteroid it will abolished the air way obstruction. While COPD has poor response to corticosteroid.

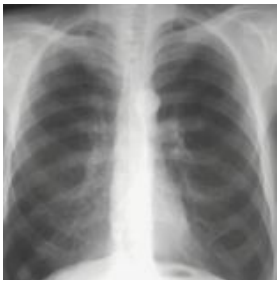
Clinical and radiological diagnosis

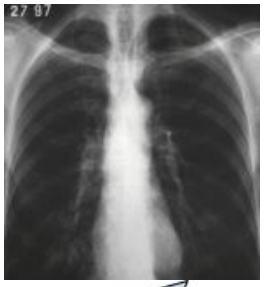
- **Clinical diagnosis:**
  - Use GOLD criteria. here
  - There is a history of breathlessness and sputum production in a **lifetime smoker.**
  - In the **absence** of a history of cigarette **smoking** an initial working diagnosis of asthma is usual unless there is a family history of COPD suggesting **α_1-antitrypsin deficiency.**

- **Investigations:**
  1. **Chest X-ray**[15]:Although there are no reliable radiographic signs that correlate with the severity of airflow limitation, a chest X-ray is essential to identify alternative diagnoses (**lung cancer, CHF, and bullae**)[16]
     a. Low sensitivity for diagnosing COPD; only severe advanced **emphysema** will show changes.
     b. Useful in acute exacerbations to rule out complications such as **pneumonia** and **pneumothorax.**



Plain chest radiograph in severe cases of emphysema might show "*barrel chest*"

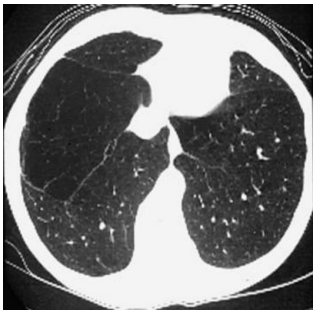




Advanced emphysema shows: *Hyperinflated*, flattened diaphragm, *enlarged retrosternal* space and *diminished vascular markings.*

[15] **X ray is the best initial test while PFT is the most accurate**
[16] A bulla is defined as an air space in the lung measuring more than one centimeter in diameter in the distended state

2. **High resolution CT scans:** used, particularly to show **emphysematous bullae.**



   a. Barrel chest due to hyperinflated lung(if u percuss him, the liver dullness will be in the 6th or 7th intercostal space)
   b. No subcutaneous fat (**cachexia**),
   c. Most serious: there's bilateral destruction of the alveoli (**bullae**),
   d. No blood vessels, alveoli, interstitium in the upper part of right lung "disappeared"
      EMPHYSEMA
   e. **No** obvious fibrosis

3. **Lung function tests**[17]**:** (Spirometry) > gives FEV1/FVC.
   a. Decreased both FEV1 and FVC →Decreased FEV1/FVC ratio. here
      - Which could be decreased and do not improve with bronchodilators (vs. asthma).
   b. **Lung volumes** might be measured provides an assessment of hyperinflation.
      - Increased total lung capacity (TLC)[18], functional reserve capacity (FRC) and residual volume.
      - Decreased vital capacity.
   c. **Gas transfer value (DLco):**
      ◆ carbon monoxide diffusing capacity(DLco)[19]:

19.4 How to interpret respiratory function abnormalities

	Asthma	Chronic bronchitis	Emphysema	Pulmonary fibrosis
FEV_1	↓↓	↓↓	↓↓	↓
VC	↓	↓	↓	↓↓
FEV_1/VC	↓	↓	↓	→/↑
TL_{CO}	→	→	↓↓	↓↓
K_{CO}	→/↑	→	↓	→/↓
TLC	→/↑	↑	↑↑	↓
RV	→/↑	↑	↑↑	↓

(RV = residual volume; see text for other abbreviations)

- To measure the efficiency of gas transfer across the alveolar-capillary membrane carbon monoxide is used as a surrogate, since its diffusion rate is similar to oxygen.

- Measure the number & structural integrity of the alveoli.

- The test make pt. to inhale small amount of carbon monoxide → then pt. hold breath for 10 sec→ the rate of CO absorption calculated.

★ **CO crosses the alveolar capillaries→ goes into RBC → attaches to hemoglobin:**

   a. In normal lungs the transfer of CO reflects the **diffusing capacity of the alveoli for oxygen.**
   b. In case of alveolar destruction ***(emphysema)*** → **↓ DLCO → decreased gas exchange**.
   c. In case of increased blood in the lungs **(pulmonary hemorrhage, Goodpasture syndrome)** there will be **more Hb available for CO** to bind to → **↑ DLCO → increased gas exchange**.
   d. **Kco reduced**[20]

4. **Pulse oximetry:**

Less than 93% patient might need oxygen therapy

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[17] Definitive diagnostic test.
[18] This is generally performed by using the helium dilution technique.
[19] Asthma cause obstruction but doesn't destroy the alveoli no reduction in Dlco
[20] **What does that mean if pt has disease involving the alveoli in addition to obstructive lung disease ?** carbon monoxide transfer coefficient (Kco) Will reduced

5. **Measure alpha 1 antitrypsin levels:**

- especially in patients with premature emphysema **(<50 years)** especially if the patient has coexisting hepatic disease (cirrhosis), or non-smokers

    a. **Alpha 1 antitrypsin** inhibits **trypsin** that damage the parenchyma therefore **emphysema** develops **early** in case of deficiency.
    b. The liver problems arise because there is a problem in the synthesis of alpha 1 antitrypsin (which is normally done in the liver) which leads to its **accumulation in the liver leading to raised LFTs**.

6. **Arterial blood gas (ABG):**[21]

    a. Low PaO2 Normal PaCO2

- "Because of emphysema they become hypoxic (because of obstruction of alveoli)"

    b. Low PaO2 High PaCO 2

- "Initially PaCO2 normal but later on will be hypercapnia(because of bronchitis no enough air getting out)".
- "there're some pt with PURE emphysema die from hypoxemia ONLY & never retain CO2 in the lung (no hypercapnia)"

    c. pH acidic or low normal

- "Because of retention of CO2® develop respiratory ACIDOSIS (↑ CO2 & low PH)"

    d. HCO 3 raised (compensatory metabolic alkalosis)

- "why HCO 3 raised? Result from a Compensation of acidosis but PH still acidic it not compensate fully

7. CBC: May have an increase in hematocrit from chronic hypoxia.

8. EKG:
    a. Right atrial hypertrophy and right ventricular hypertrophy
    b. Atrial fibrillation or multifocal atrial tachycardia
    c. In advanced cor pulmonale the P wave is taller (P pulmonale) and there may be right bundle branch block (RSR' complex) and the changes of right ventricular hypertrophy.

.

9. Echocardiography:
    a. Right atrial and right ventricular hypertrophy
    b. Pulmonary hypertension

[21] Some pt will progress to death due to hypercapnic respiratory failure & other due to normocapnic respiratory failure

- **Management:**

○ **Improve mortality and delays progression of disease:**	**1. Smoking cessation**	**prolongs** the survival rate but **does not reduce** it to the level of someone who has never smoked[22].
	2. Long term oxygen therapy (LTOT) here	a. **Improve survival and quality of life** in patients, might be continuous or intermittent only during sleep or exertion. b. Only in patients with hypoxia/cor pulmonale
	3. Vaccinations	a. **Influenza vaccine** yearly for all patients. Prevent winter exacerbations b. **Strep pneumoniae vaccine** every 5-6 years should be offered to patients with **COPD over 65 years** old, or **under 65** who have **severe** disease.
○ **Improves symptoms (But does not decrease disease progression or mortality):**	**4. Bronchodilators[23] (Inhaled)**	a. Short acting bronchodilators for **mild** disease - **Inhaled Beta 2 agonists:** Salbutamol, Terbutaline b. Long acting bronchodilators for **moderate** to **severe** disease - **Inhaled Beta 2 agonists**: Salmeterol, Formoterol, Indacaterol. Use **long-acting** agents for patients requiring frequent use. - **Inhaled Anticholinergics[24]** (muscarinic antagonists) are more appropriate for patients with **moderate** to **severe** disease as: Tiotropium bromide, Ipratropium bromide. *This is because it is believed that beta agonists might cause CVS side effects (tachycardia. etc.) They also have slower onset of action than beta blockers but last longer. - Combination of beta agonist Salbutamol "albuterol" with ipratropium bromide is more efficacious than either agent alone in bronchodilation. Also help in adherence to therapy "both medications in one inhaler"
		can be given to patients who ***cannot*** inhale efficiently

[22] (This initial inflammation of the small airways is reversible and accounts for the improvement in airway function if smoking is stopped early.)
[23] minimal effect. Improve FEV_1 by 5-10%
[24] Inhaled anticholinergic agents are most effective in COPD

cont.. **Improves symptoms**	**Cont…** **Bronchodilators (Oral)**	❖ **Theophylline:** - not commonly used because it has a lot of side effects and drug interactions + Narrow therapeutic index, so serum levels must be monitored. - Improves **mucociliary clearance and central respiratory drive.** - Occasionally used to patients with refractory COPD.
	5. Corticosteroids[25]	a. **Inhaled corticosteroids** (anti-inflammatory) b. Usually given in combination with **long acting beta agonists.**[26] c. **Oral corticosteroids** are useful in acute exacerbations (discussed later) "systemic glucocorticoids are only used for acute exacerbations and **should not** be used for long-term treatment, even for pts with severe COPD" - Remember steroids have many side effects such as osteoporosis and Cushing's syndrome.
	6. Pulmonary rehabilitation (grade 3-5 SOB) here	- patients with dyspnea grade 3-5 will benefit from rehabilitation (treadmill exercise) a. Education, physiotherapy encourage patients to exercise patients reassured that breathlessness, whilst distressing, is not dangerous. Improves walking distance by 25%-40% **without** improving the FEV1 and dyspnea due to conditioning of the heart and skeletal muscles.
	7. Surgery:	may be beneficial in selected patients: a. **Bullectomy: Very rare** i. indications? to remove a large bullae that compresses the lung in young patients + **minimal** airflow limitation + **no** generalized emphysema + **FEV1 > 40%** b. **Lung volume reduction surgery (LVRS):** i. Indications? Patients with **predominantly upper lobe emphysema** with preserved gas transference and **no** evidence of pulmonary HTN + FVE1 and DLCO above 20% c. Lung resection. d. Lung transplantation. e. When all medical therapy is insufficient, the answer is "refer for transplantation"

[25] In symptomatic patients with moderate/severe COPD, a trial of corticosteroids is always indicated.
[26] Combination of a corticosteroid with a long-acting p2 agonist has been shown in a trial to protect against a decline in lung function but there was no reduction in overall mortality

Treatment guidelines	
Mild to **moderate** disease	**Severe** disease
1. **Bronchodilator** in an inhaler (use spacer to improve delivery), anticholinergic drugs and/or beta agonists are first-line agents. 2. Inhaled **corticosteroids** may be used as well. Use lowest dose possible 3. Theophylline may be considered if the above do not adequately control symptoms.	1. Medications as mild-moderate. 2. Continuous **oxygen** therapy if patient is hypoxemic **3. Pulmonary rehabilitation.** 4. **Triple inhaler therapy** (long acting beta agonist + long acting anticholinergic + an inhaled glucocorticoid)

- **Complications of COPD[27]:**
  - Respiratory failure " COPD is the most common cause"
  - Pulmonary HTN constriction of pulmonary arterioles due to hypoxia
  - Cor pulmonale Hypoxia induces constriction of pulmonary arterioles →increase pulmonary resistance → result in pulmonary hypertension and right ventricular hypertrophy with or without right sided heart failure
  - Bacterial colonisation
  - Secondary polycythemia
  - Hemoptysis
  - Pneumothorax not very common "occasionally" (hyperinflation → rupture)
  - **Acute exacerbations** (mostly due to infections or noncompliance).
  - Extrapulmonary "like cachexia and osteoporosis"

- **Acute exacerbation of COPD[28] = worsening :**
  - **Definition:**
  - Any worsen of symptoms that requires a change of medication or a doctor visit.
  - characterised by an **increase in symptoms** "dyspnea, sputum production and/or cough." and **deterioration in lung function** and **health status,** necessitating a **change in management**
  - Patients might be managed at **home** by increasing the doses of their medications[29]
  - If patient has cyanosis, edema, or altered level of consciousness > must go to **hospital**
  - Can lead to acute respiratory failure requiring hospitalization, and possibly mechanical ventilation; potentially fatal.

[27] Predictors of a poor prognosis are increasing age and worsening airflow limitation.
[28] They may be accompanied by the development of respiratory failure and/ or fluid retention.
[29] **increased bronchodilator therapy, a short course of oral corticosteroids and, if appropriate, antibiotics.**

- **Causes:**

1. **Viral infection** (common cold) followed by **bacterial activity** (gram negative). a. One third associated with virus **(rhinovirus or influenza)** b. Bacterial colonization (20%-30% due to remissions. 30%-50% during exacerbation) i. **H.influenza** and parainfluenza ii. Strep. Pneumoniae **Uncommon** iii. Branhamella catarrhalis.	2. **Bronchospasm** **The most serious** (can be treated with bronchodilators however somebody with infection with COPD the majority will become worse) a. Pollution or occupational	3. **Minor causes** uncommon a. Pneumonia b. Left or right cardiac failure c. Pneumothorax

- **Life threatening exacerbations:**
  - Deterioration of consciousness
  - Marked distress
  - Paradoxical thoracoabdominal movement
  - Worsening ABGs in spite of oxygen and bronchodilators (50 - 70 - 7.3) you don't need to remember the level of gases
  - Other comorbidities heart failure or liver failure
  - Social support.

- **Management:** similar to treatment of acute asthma exacerbations

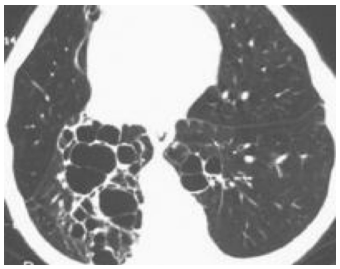
1. O_2 24%[30] why we don't give these patients too much O_2? They go comatous? Why? Because they depend on hypoxemia for respiratory drive and high oxygen concentration will slow down breathing and CO_2 rises, pH go down and they go into coma.
   - **Don't** give 100% O_2 because it might cause respiratory depression and worsens the patient's ventilation and acidosis. (but WHY? Here if ur interested)
2. Give nebulized **short acting beta 2 agonists** and an **anticholinergic agent** (salbutamol and ipratropium) alone or in combination are **first-line therapy.**
3. **Oral corticosteroids**, do **NOT** use inhaled corticosteroids in acute exacerbation.
4. Systemic corticosteroids are used for patients requiring hospitalization (IV methylprednisolone is a common choice)
5. **Antibiotics**
   a. (aminopenicillin or macrolide or azithromycin, levofloxacin, doxycycline, etc) "no antibiotic superior to another" if there is increase in sputum color, volume, purulence, or breathlessness (step up)
   b. amoxycillin/Clavulanate, cephalosporin (e.g. cefuroxime), quinolone (ciprofloxacin, levofloxacin, moxifloxacin) If worsening 2 out of 3 of the following: in slides
      i. **Shortness of breath**
      ii. **Amount of sputum**
      iii. **Purulency of sputum**
   c. Because bacterial infections might be involved in causing exacerbations of COPD

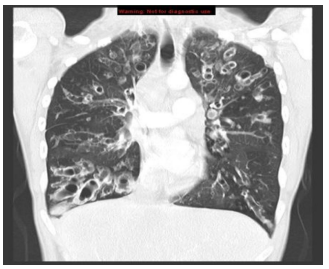
6. Non-invasive ventilatory support
   a. May decrease the likelihood of respiratory failure.

[30] used with the aim of maintaining a *Pa*O2 above 8 kPa (60 mmHg) (or an *Sa*O2 between 88% and 92%) without worsening acidosis.

- What do we see in CT scan? These patients have holes in the lung like emphysema but they're different. They have thick fibrous wall which mean that they are cysts NOT bullae (which has no wall) means there's inflammation.
- Dilated bronchi (Normally the artery is bigger than bronchus , but in this pt the bronchi is bigger).

  * **DX: dilated bronchiectasis**





Definition of bronchiectasis

- Bronchiectasis:
  - Abnormal **permanent** dilation and irreversible damage of bronchi of small and medium sized bronchi. (Master the boards: chronic dilation of large bronchi),
    - **Pathogenesis:**

      There is **permanent**, abnormal dilation and destruction of bronchial walls with **chronic inflammation**, airway collapse, and **ciliary loss/dysfunction** leading to **impaired clearance** of secretion[31],
  - Bronchiectasis is uncommon because of better control of infections of the lung which lead to the weakening of the bronchial walls.
    - **Causes:**

1. Any condition that causes chronic inflammation, destruction, and scarring:	a. **Cystic fibrosis[32]:** autosomal recessive disease that causes thick mucus secretion. Most common cause, accounts for half of all cases. b. **Mucus plugs** increase the likelihood of infections→ bronchiectasis
2. Hypogammaglobulinemia:	(caused by common variable immunodeficiency[33] (CVID)).
3. **Ciliary dysfunction[34] (immotile cilia syndrome)**	a. Cilia not working properly→ increased mucus plugs and infections→ bronchiectasis. b. Also causes sinusitis and male infertility. c. About 50% of patients with primary ciliary dyskinesia will have situs inversus and sinusitis (kartagener syndrome)
4. Foreign body or tumors:	a. Obstruction→ inflammation→ destruction→ bronchiectasis.
5. Infections:	a. Fungal (allergic bronchopulmonary aspergillosis), b. measles, whooping cough, mycobacterium etc. c. **H.influenzae,** K.pneumoniae. S.pneumoniae, d. staph. aureus (cystic fibrosis) e. **p.aeruginosa** (associated with rapid decline of FEV1).

[31] The bronchiectatic cavities may be lined by granulation tissue, squamous epithelium or normal ciliated epithelium. There may also be inflammatory changes in the deeper layers of the bronchial wall and hypertrophy of the bronchial arteries.

[32] is heredity disease not common in arab

[33] (is a disorder that impairs the immune system. Pt are highly susceptible to infection from foreign invaders such as bacteria, or more rarely, viruses and often develop recurrent infections, particularly in the lungs).

[34] the common cause of ciliary dyskinesia in saudi? post tb & childi infection .

Others:	6. Agammaglobulinemia and immunodeficiency Immunocompromised → infections → bronchiectasis 7. Collagen-vascular disease e.g. rheumatoid arthritis 8. Autoimmune disease: Rheumatoid arthritis, systemic lupus erythematosus, crohn's disease, etc.) 9. Humoral immunodeficiency (abnormal lung defense) Airway obstruction.

- Note that bronchiectasis due to **cystic fibrosis** will be *different* than **foreign body** in areas of lung involved..
  - **Systemic diseases** such as cystic fibrosis or ciliary dyskinesia would cause bronchiectasis in **both lungs**
  - A foreign body or pus accumulation will cause **localized bronchiectasis** (**distal** to the obstructed bronchus)
    - Agammaglobulinemia will cause bronchiectasis in **lung bases**.

○ Clinical features:

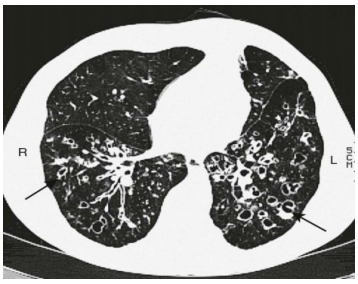
- Mainly similar to COPD. May have increased likelihood for infections **commonly pneumonias** "recurrent or persistent pneumonia" (because the blocked area is an excellent place for bacteria to grow)
- **Chronic cough** with **large amount of mucopurulent copious foul smelling sputum**[35] leading to **halitosis**. Due to infection
- **Hemoptysis**[36] due to rupture of blood vessels near bronchial wall surface. May be life threatening
- **Dyspnea, Weight loss and Fever**.

19.37 Symptoms of bronchiectasis

- **Cough**: chronic, daily, persistent
- **Sputum**: copious, continuously purulent
- **Pleuritic pain**: when infection spreads to involve pleura, or with segmental collapse due to retained secretions
- **Haemoptysis**:
  - Streaks of blood common, larger volumes with exacerbations of infection
  - Massive haemoptysis requiring bronchial artery embolisation sometimes occurs
- **Infective exacerbation**: increased sputum volume with fever, malaise, anorexia
- **Halitosis**: frequently accompanies purulent sputum
- **General debility**: difficulty maintaining weight, anorexia, exertional breathlessness

- On auscultation: **Wheezes**[37] similar to asthma and COPD or **crackles**[38]
- On examination: Clubbing is uncommon used to be common but now we have better control of infection and it never occurs in COPD and asthma. Seen in cancers.

○ Investigations:



1. Should **culture** patient's sputum because they often have special infections (Pseudomonas aeruginosa) and we should know their antibiotic sensitivity by culture in order to properly treat it
2. **CT scan**[39] (the best non-invasive test) shows thickened dilated bronchi **"most accurate test, study of choice."**
3. Chest x-ray (CXR): It might be **normal** BUT in advanced cases it may show **dilated bronchi** with **thickened bronchial walls** and sometimes multiple **cysts containing fluid** and crowding of bonchi (tram tracking) **best initial test.**
4. Can also look for diseases that cause this condition For example: screen for ciliary dysfunction, CF, etc.
5. PFTs reveals an obstructive pattern.
6. Bronchoscopy applies in certain cases.

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[35] Yellow or green

[36] Treatment of the haemoptysis consists of bed rest and antibiotics, when most stop bleeding. Blood transfusion is given if required. Urgent fibreoptic bronchoscopy is occasionally necessary to detect the source of bleeding. If the haemoptysis does not settle rapidly, the treatment of choice is bronchial artery embolization. Surgical resection may be required if embolization fails.

[37] (when the disease become bilateral)

[38] When there are large amounts of sputum in the bronchiectatic spaces, coarse crackles may be heard over the affected areas.

[39] with a sensitivity of 97%.

- **Management:**

1. If the patient has airflow obstruction→ treat that same as COPD patients **(inhaled bronchodilators and corticosteroids)**
2. **Bronchial hygiene** is very important.
    a. Chest physiotherapy: (cupping and clapping) and postural drainage, chest percussion are essential for dislodging plugged-up bronchi + help to remove the mucus.
    b. hydration.
3. **Antibiotics**[40]:

- for 1 week. However if there's exacerbation give antibiotics for 2 weeks
    a. usually same as COPD (aminopenicillin or macrolide) but usually for **longer durations and higher doses.**
4. For pseudomonas colonization, **Nebulized antibiotic therapy** (Gentamicin or Tobramycin twice daily) in slides
5. If culture shows **Pseudomonas** or Staph aureus, antibiotic therapy might be more difficult and should use sensitivity to guide antibiotic therapy
6. Surgical resection of focal lesions may be indicated.

- **Prevention:**

Treat any cause of bronchial obstruction early to avoid the development of bronchiectasis (remove the foreign body early before it progresses to chronic inflammation and bronchiectasis).

- **Prognosis:**

The disease is progressive when associated with ciliary dysfunction and cystic fibrosis, and eventually causes respiratory failure. In other patients, the prognosis can be relatively good if physiotherapy is performed regularly and antibiotics are used aggressively.

TABLE 2-1 Pink Puffers Versus Blue Bloaters

Pink Puffers (Predominant Emphysema)	Blue Bloaters (Predominant Chronic Bronchitis)
• Patients tend to be thin due to increased energy expenditure during breathing. • When sitting, patients tend to lean forward. • Patients have a barrel chest (increased AP diameter of chest).	• Patients tend to be overweight and cyanotic (secondary to chronic hypercapnia and hypoxemia). • Chronic cough and sputum production are characteristic. • Signs of cor pulmonale may be present in severe or long-standing disease.
Tachypnea with prolonged expiration through pursed lips is present.	Respiratory rate is normal or slightly increased.
Patient is distressed and uses accessory muscles (especially strap muscles in neck).	Patient is in no apparent distress, and there is no apparent use of accessory muscles.

Table 14.11 Causes of bronchiectasis

Congenital
Deficiency of bronchial wall elements
Pulmonary sequestration

Mechanical bronchial obstruction
Intrinsic:
- Foreign body
- Inspissated mucus
- Post-tuberculous stenosis
- Tumour

Extrinsic:
- Lymph node
- Tumour

Postinfective bronchial damage
Bacterial and viral pneumonia, including pertussis, measles and aspiration pneumonia

Granuloma
Tuberculosis, sarcoidosis

Diffuse diseases of the lung parenchyma
e.g. idiopathic pulmonary fibrosis

Immunological over-response
Allergic bronchopulmonary aspergillosis
Post-lung transplant

Immune deficiency
Primary:
- Panhypogammaglobulinaemia
- Selective immunoglobulin deficiencies (IgA and IgG_2)

Secondary:
- HIV and malignancy

Mucociliary clearance defects
Genetic:
- Primary ciliary dyskinesia (Kartagener's syndrome with dextrocardia and situs inversus)
- Cystic fibrosis

Acquired:
- Young's syndrome – azoospermia, sinusitis

[40] The incidence of complications "Pneumonia, pneumothorax, empyema and meta- static cerebral abscess" has fallen with antibiotic therapy.

Questions:

- Ali 65 years old
  - C/O exercise intolerance for 2 years
  - History of occasional wheeze
  - Slight cough for a while 5 may be 10 years
  - Morning sputum most of the time
  - Smoked on and off for 40 years / 1.5 packs
  - No clubbing
  - Wheeze[41] and hyperinflation[42]
  - FEV1 / FVC < 70% (obstruction)
  - Diffusing capacity (DLco) Kco reduced (due to obstruction).

- Saleh 55 years[43]
  - smoked 60 since age 16
  - Barrel-shaped
  - Liver 6th space
  - Cough, expectoration, SOB 2 years
  - FEV1 44% FEF 19% RV%TLC 200% FEV1 / FVC 60%
  - Ventolin neb. 5mg----FEV1 INCREASED 140 ml (10%)
  - KCO 98% Allergic rhinitis and hypret. Turbinates (asthma)
  - SOB triggered strongly by dust and irritants
  - A trial of Symbicort for 3 weeks
  - FEV1 and FEF50 rose to 80%
  - FEV1 relapsed to 64% but recovered
  - D.D. with asthma: -Age of onset -History of atopy -Eosinophilia and IgE -Bullae -Chronic respiratory failure (asthma doesn't destroy lungs, patient won't develop chronic respiratory failure) -Diffusing capacity -Trial of inhaled corticosteroids (improvement in asthma)

[41] What is the significant of wheeze in this pt? indicates obstructive lung disease(bilateral bronchiectasis _asthma _COPD)

[42] **How to detect hyperinflation CLINICALLY?**

- by looking to the shape of chest would be barrel shape,
- by persecution the upper border of liver :normally the liver dullness in 5th intercostal space but in case of hyperinflation the liver will be pushed down so the dullness will be in 6 or 7th intercostal space.

[43] Carbon monoxide diffusing capacity is NORMAL , which mean his alveoli is normal , indicating PURE obstruction with no alveolar damage.
Also this pt has Hx of atopy (atopy refers to the genetic tendency to develop allergic diseases such as allergic rhinitis, asthma and eczema).

- level of exertion the patient can manage before stopping.

19.28 Modified MRC dyspnoea scale

Grade	Degree of breathlessness related to activities
0	No breathlessness, except with strenuous exercise
1	Breathlessness when hurrying on the level or walking up a slight hill
2	Walks slower than contemporaries on level ground because of breathlessness or has to stop for breath when walking at own pace
3	Stops for breath after walking about 100 m or after a few minutes on level ground
4	Too breathless to leave the house, or breathless when dressing or undressing

(MRC = Medical Research Council)

- Long term oxygen therapy:

19.32 Prescription of long-term oxygen therapy in COPD

Arterial blood gases measured in clinically stable patients on optimal medical therapy on at least two occasions 3 weeks apart:

- PaO_2 < 7.3 kPa (55 mmHg) irrespective of $PaCO_2$ and FEV_1 < 1.5 L
- PaO_2 7.3–8 kPa (55–60 mmHg) plus pulmonary hypertension, peripheral oedema or nocturnal hypoxaemia
- the patient has stopped smoking.

Use at least 15 hrs/day at 2–4 L/min to achieve a PaO_2 > 8 kPa (60 mmHg) without unacceptable rise in $PaCO_2$.

- Gold criteria:

I : Mild	II : Moderate	III : Severe	IV : Very severe
• FEV_1/FVC < 0.70 • FEV_1 ≥ 80% predicted	• FEV_1/FVC < 0.70 • 50% ≤ FEV_1 < 80% predicted	• FEV_1/FVC < 0.70 • 30% ≤ FEV_1 < 50% predicted	• FEV_1/FVC < 0.70 • FEV_1 < 30% predicted *or* FEV_1 < 50% predicted *plus* chronic respiratory failure
Active reduction of risk factor(s); influenza vaccination **Add** short-acting bronchodilator (when needed)			
	Add regular treatment with one or more long-acting bronchodilators (when needed) **Add** rehabilitation		
		Add inhaled glucocorticosteroids if repeated exacerbations	
			Add long-term oxygen if chronic respiratory failure **Consider** surgical treatments

Fig. 19.28 Global Initiative for Chronic Obstructive Lung Disease (GOLD) guidelines for treatment of COPD. Post-bronchodilator FEV_1 is recommended for the diagnosis and assessment of severity of COPD. From www.goldcopd.com – see p. 732.

- spirometric classification of COPD

19.29 Spirometric classification of COPD severity based on post-bronchodilator FEV_1

Stage	Severity	FEV_1
I	Mild*	FEV_1/FVC < 0.70 FEV_1 ≥ 80% predicted
II	Moderate	FEV_1/FVC < 0.70 FEV_1 50–79% predicted
III	Severe	FEV_1/FVC < 0.70 FEV_1 30–49% predicted
IV	Very severe	FEV_1/FVC < 0.70 FEV_1 < 30% predicted or FEV_1 < 50% predicted if respiratory failure present

*Mild COPD should not be diagnosed on lung function alone if the patient is asymptomatic. Based on NICE guidelines 2010.

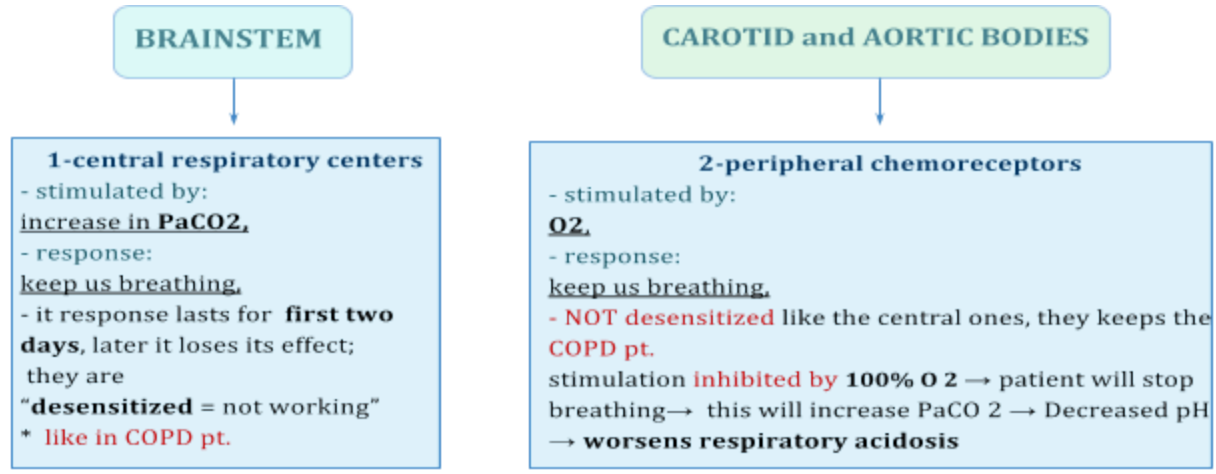
→

- FEV1: the volume exhaled in the first second
- FVC: the total volume exhaled
- In case of obstruction: the degree of reduction of FEV1 (↓ ↓ FEV1) is much more severe than FVC (↓ FVC).
- In obstruction there's difficulty in exhaling more than 70 % of air per second

- Long term rehabilitation:

LONG –TERM REHABILITATION 1	LONG –TERM REHABILITATION 2
• Benefit independent of age, no improvement of FEV_1, exercise capacity, PaO_2 • walk test 25-40% improve walking distance by 25-40% • 6 minutes walk + 60 metres • Only modest rise VO2peak • Well being .	• 2 supervised + 1 unsupervised session • As little as 6 weeks (Max. 12 weeks) 20-30 min • Anaerobic (cycle, brisk walking)?? Strength exercises • Lower limbs > upper limbs • Respiratory muscles: no effect • 60 – 85% peak performance • Benefit maintained 12-18 months without formal maintenance regimen.

Managing Acute exacerbation of COPD with O2 at 24%



Bottom line: don't give 100% O 2 to patients presenting with COPD exacerbations

Cases

1) Which of the following are the most likely physical examination findings in a patient with emphysema?

A. Diffuse expiratory wheezing
B. Clubbing of the fingers
C. Bibasilarinspiratorycrackleswithincreasedjugularvenouspressure (JVP)
D. Inspiratory stridor
E. Third heart sound

2) A 56-year-old woman admits to a 60-pack-year smoking history. She complains of fatigue and dyspnea with minimal exertion, and a cough that is productive each morning. Which of the following is the most likely finding in this patient?

A. Normal diffusing capacity of lung for carbon monoxide (DLCO)
B. Decreased residual volume
C. Normaltoslightlyincreasedforcedexpiratoryvolumeinfirstsecond (FEV1)
D. Decreased forced expiratory volume in first second/forced vital capacity (FEV1/FVC)
E. Decreased forced vital capacity(FVC)

3) Which of the following therapies is most likely to provide the greatest benefit to a patient with chronic stable emphysema and a resting oxygen saturation of 86%?

A. Inhaled tiotropium daily
B. Inhaled albuterol as needed
C. Oral prednisone daily
D. Supplemental oxygen used at night
E. Supplemental oxygen used continuously

4) You see a 68-year-old man in clinic, with a 40 (cigarette) pack year history, who has been experiencing breathlessness on exertion and a productive cough of white sputum over the last four months. You assess his spirometry results which reveal an FEV1/FVC of 51 per cent with minimal reversibility after a 2-week trial of oral steroids. Cardiological investigations are normal. Which of the following is the most likely diagnosis?

A. Asthma
B. Chronic obstructive pulmonary disease (COPD)
C. Left ventricular failure
D. Chronic bronchitis
E. Lung fibrosis

5) The severity of COPD is assessed using post bronchodilator spirometery analysis. From the list below, select the values that you would expect to see in a patient with moderate COPD.

A. FEV1/FVC <0.7, FEV1 per cent predicted 30–49 percent
B. FEV1/FVC <0.7, FEV1 per cent predicted ≥80 percent
C. FEV1/FVC <0.7, FEV1 per cent predicted <30 percent
D. FEV1/FVC <0.7, FEV1 per cent predicted 50–79 percent
E. FEV1/FVC <0.7, FEV1 per cent predicted 60–70 percent

6) A 58-year-old man with known COPD, diagnosed eight months ago, attends your clinic with persistent shortness of breath despite stopping smoking and using his salbutamol inhaler (given to him at the time of diagnosis), which he finds he is using more frequently. You assess the patient's lung function tests that have been recorded just before he saw you in clinic on this occasion. His FEV1 = 65 per cent of the predicted value. Oxygen saturations are 95 per cent on room air, respiratory rate in 18, and his temperature is 37.1°C. From the list below, select the next most appropriate step in this patient's management.

A. 40 mg daily oral prednisolone for 5 days
B. Start long-term oxygen therapy
C. Start inhaled corticosteroid therapy
D. Add oral theophylline therapy
E. Add a long-acting β 2 agonist inhaler

Answers

1) **A**. COPD is characterized by chronic airway obstruction, with most airflow resistance occurring in small airways of the lower respiratory tract, producing expiratory wheezing. Inspiratory stridor would occur with upper airway, usually extrathoracic, obstruction. Clubbing is not generally a feature of COPD and should prompt investigation for another disease process such as a bronchogenic carcinoma. Crackles, elevated JVP, and an S3 are signs of congestive heart failure.

2) **D**. This patient likely has COPD, based on the smoking history and symptoms. A decrease in the forced expiratory volume in first second/ forced vital capacity ratio is the hallmark of airflow obstruction. The FEV1 is decreased in obstructive, as well as in restrictive, lung dis- ease. The diffusing capacity is typically deceased in COPD as well as intrinsic restrictive lung disease. The DLCO indicates the adequacy of the alveolar-capillary membrane; the residual volume is the volume of air remaining in the lungs after a maximal expiratory effort and is usually increased in COPD due to air trapping.

3) **E.** For patients with chronic hypoxemia, supplemental oxygen has a significant impact on mortality, with a greater benefit with continuous usage, rather than intermittent or nocturnal-only usage. Bronchodilators such as tiotropium and albuterol improve symptoms and improve FEV1, but offer no mortality benefit. Chronic use of oral corticosteroids should be avoided because of unfavorable side effects such as osteoporosis, glucose intolerance, and gastrointestinal (GI) side effects.

4) **B.** The patient's symptom history coupled with the spirometry results indicate that he has an obstructive defect. Spirometry is typically used to measure functional lung volumes. The ratio of the forced expiratory volume in one second (FEV1) to the forced vital capacity (FVC), provides a reliable approximation of severity of airflow obstruction; the normal being 80 per cent. An FEV1/FVC ratio of less than 80 per cent indicates an obstructive defect seen in COPD and asthma while a ratio of greater than 80 per cent is representative of a restrictive defect seen in lung fibrosis (E). The spirometry results coupled with minimal reversibility points the diagnosis to COPD (B) rather than asthma (A), where reversibility of the FEV1/FVC ratio is usually seen. Chronic bronchitis (D) can be defined as cough productive of sputum for three months of two successive years which does not corroborate with the onset of symptoms. Left ventricular failure (C) is obviously incorrect due to the fact that cardiological tests have been mentioned as normal.

5) **D.** With reference to the NICE guidelines 2010, COPD can be divided into mild, moderate, severe and very severe. The values are obtained with post bronchodilator spirometry and are as follow: Mild COPD: FEV1/FVC <0.7, FEV1 % predicted ≥80 per cent (B) Moderate COPD: FEV1/FVC <0.7, FEV1 % predicted 50–79 per cent (D) Severe COPD: FEV1/FVC <0.7, FEV1 % predicted 30–49 per cent (A) Very severe COPD: FEV1/FVC <0.7, FEV1 % predicted <30 per cent (C) Very severe COPD can also be seen in patients with FEV1 % predicted <50 per cent with respiratory failure.

6) E :

This patient was diagnosed with COPD eight months ago and was started on a short-acting β 2 agonist inhaler. With reference to the NICE guidelines in managing stable COPD, this patient can either have a long-acting β 2 agonist (LABA) inhaler (e.g. salmeterol) or a long-acting muscarinic antagonist (LAMA) inhaler (e.g. tiotropium). Therefore, from the list, E is the most appropriate answer.

Inhaled therapy is usually started when the patient is diagnosed with COPD and is breathless and/or has exercise limitation. Either a short-acting β 2 agonist (SABA) inhaler (e.g. salbutamol) or short-acting muscarinic antagonist (SAMA) inhaler (e.g. ipratropium) can be used initially.

If the patient experiences frequent exacerbations or persistent breathlessness, despite being on either a SABA or SAMA, two pathways can be taken:

- If the FEV1 ≥50 percent :
- either a LABA inhaler can be added (this can coexist with the SABA)
- or a LAMA inhaler can be used – the LAMA will replace the SAMA in this case
- If the FEV1< 50 percent :
- either the addition of a LABA + inhaled corticosteroid (ICS) in a combination inhaler (e.g. salmeterol + fluticasone).
- or the addition of a LAMA inhaler – again the SAMA inhaler should be stopped.
- If the patient remains breathless or experiences persistent exacerbations:

• ICS (C) therapy can be added as a combination inhaler if LABA inhaler was added before. If the patient remains breathless and experiences exacerbations, a LAMA inhaler can be offered.

• If a LAMA was added before, addition of a LABA + ICS can be considered

• A LAMA inhaler can be offered if the patient is already on a LABA + ICS.

Oral therapy:

• Oral corticosteroid (A) maintenance therapy is not advised in COPD, although patients with advanced COPD may require low-dose oral corticosteroid therapy to prevent exacerbations. Osteoporosis prophylaxis should be considered.

• Theophylline therapy (D) is offered to patients who cannot use inhalers or after trials of short- and long-acting bronchodilators. Theophylline can be used in combination with β 2 agonists and muscarinic antagonists.

• Mucolytic therapy (e.g. carbocysteine) can be considered in people with a chronic productive cough.

Oxygen (i.e. long-term oxygen therapy) (B) therapy should be assessed in patients who have any of the following:

• very severe airflow obstruction (e.g. FEV1 <30 per cent predicted);

• cyanosis;

• polycythaemia;

• peripheral edema;

• raised JVP;

• oxygen saturations less than or equal to 92 per cent on room air.