Pearson Edexcel International Advanced Level

Biology

Advanced

Unit 5: Energy, Exercise and Coordination

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Scientific Article for use with Question 7

Paper Reference

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Scientific article for use with Question 7

COPD: chronic obstructive pulmonary disease

1. Chronic obstructive pulmonary disease (COPD) is the name for a collection of lung diseases including chronic bronchitis, emphysema and chronic obstructive airways disease. People with COPD have difficulties breathing, primarily due to the narrowing of their airways; this is called airflow obstruction.

2. Typical symptoms of COPD include increasing breathlessness when active, a persistent cough with phlegm and frequent chest infections, particularly in winter. The symptoms of chronic obstructive pulmonary disease (COPD) usually develop over a number of years, so you may not be aware you have the condition. If you have these symptoms, you should see your doctor to ensure you are diagnosed and treated as soon as possible.

3. If you have COPD, the airways of the lungs become infla[illegible] and n[illegible]ed. [illegible]e air sacs get permanently damaged, it will become increasingly diffic[illegible] breath[illegible]ou[illegible]er signs of COPD can include weight loss, tiredness and fatigue and swollen [illegible] chest p[illegible]n a[illegible]oughing up blood (haemoptysis) are not common symptoms of CO[illegible] they ar[illegible]ly [illegible] by other conditions, such as lung cancer.

4. Symptoms of COPD are often worse in winter, a[illegible]on[illegible]y to have two or more flare-ups a year. A flare-up (also known as an exac[illegible]bation) is [illegible] your [illegible]ptoms are particularly bad. This is one of the most common reasons f[illegible]e being [illegible]o hospital in the UK.

5. While there is currently no cure for CO[illegible] th[illegible] the c[illegible]ition is diagnosed and appropriate treatment begins, the less cha[illegible] there [illegible]vere[illegible] damage.

6. The main cause of COP[illegible] smo[illegible]h[illegible]od of developing COPD increases the more you smoke and the longe[illegible]u[illegible]king. This is because smoking irritates and inflames the airways in the lungs, wh[illegible]n[illegible]ring. Over many years, the inflammation leads to permanent chan[illegible]lu[illegible] walls of the airways thicken and more mucus is produced. Damage to th[illegible]elicate w[illegible]s in the lungs causes emphysema and the lungs lose their normal elastic[illegible]The sm[illegible]air[illegible]also become scarred and narrowed. These changes cause the symptoms of bre[illegible]ough and phlegm associated with COPD.

7. Some cases of [illegible]sed by fumes, dust, air pollution and genetic disorders, but these are rarer.

8. There are sev[illegible]s that may increase your risk of developing chronic obstructive pulmonary disease (COPD), many of which can be avoided. At least four out of five people who develop the disease are, or have been, smokers. Around 10–25% of smokers develop COPD. Exposure to other people's smoke is called passive smoking and this also increases the risk of COPD.

9. Exposure to certain types of dust and chemicals at work, including grains, isocyanates, cadmium and coal, has been linked to the development of COPD, even in people who do not smoke. The risk of COPD is even higher if you breathe in dust or fumes in the workplace and you smoke. According to some research, air pollution may be an additional risk factor for COPD. However, at the moment it is not conclusive and research is continuing.

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10. There are a few factors for COPD that you cannot avoid. A research study has shown that smokers who have brothers and sisters with severe COPD are at greater risk of developing the condition than smokers who do not. There is a rare genetic tendency to develop COPD called alpha-1-antitrypsin (A1AT) deficiency. This causes COPD in a small number of people (about 1%). A1AT is a protein that protects your lungs. Without it, the lungs can be damaged by other enzymes that occur naturally in the body. People who have an A1AT deficiency usually develop COPD at a younger age, often under 35.

11. A1AT is a glycoprotein which is mainly produced in the liver. It is a serine protease inhibitor. Its main function is to balance the action of neutrophil-protease enzymes in the lungs: for example, neutrophil elastase produced by neutrophils in the presence of inflammation, infection or smoking. If there is a deficiency of A1AT then elastase can break down elastin unchecked. In the lungs this can lead to the destruction of alveolar walls and worsening of emphysema.

12. A1AT deficiency is one of the most common inherited disorders among the people of North-western Europeans are most likely to carry a mutant A1AT gene. The UK has estimated that 1 person in 3,000–5,000 has A1AT deficiency. It is a condition [illegible] marked [illegible] diagnosed, which probably relates to the fact that even some people with very low levels of the protein may not exhibit problems. Furthermore, manifestation of the disease is [illegible] genetic predisposition and environmental factors. For example, a person who is heterozygous may simply have a predisposition to chronic obstructive pulmonary disease (COPD) if they smoke. Between 1–3% of people diagnosed with COPD are thought to have A1AT deficiency.

13. A1AT deficiency is an inherited condition. In those with A1AT deficiency, the protein is still produced but the genetic defect means that the A1AT molecule configuration is changed. As a result, it cannot pass out of the liver into the blood stream and so cannot pass to the lungs and the rest of the body.

14. There is a mutation in the SERPINA1 gene on chromosome 14. Over 100 different allelic variants of the alpha-1 antitrypsin gene have been described. Everyone inherits two copies of chromosome 14 and the normal form is designated Pi MM. Pi stands for protease inhibitor. The two most important abnormal alleles are called S and Z. Both result from mutations of the alpha-1 antitrypsin gene. Someone who is homozygous Pi MM, will produce normal amounts of A1AT.

15. Individuals may have two abnormal alleles labelled Pi SS or Pi ZZ, or one of each Pi SZ. This is called being heterozygous. Or they may have one abnormal and one normal allele, for example, Pi MS or Pi MZ. The heterozygous state makes you a carrier of the disease.

16. Alpha-1 antitrypsin alleles are co-dominant, so each allele of the pair should make 50 per cent of the alpha-1 antitrypsin produced. But an abnormal Pi Z allele only makes about 10 per cent of the alpha-1 antitrypsin produced by a normal Pi M allele. Pi ZZ people have only 15–20 per cent of normal blood alpha-1 antitrypsin levels, which is linked to severe disease. People with one Pi Z allele and one Pi M allele, Pi MZ, have alpha-1 antitrypsin levels around 60 per cent of normal (50 per cent from M and 10 per cent from Z). This is usually enough to prevent disease.

17. People with both Pi S alleles (Pi SS) are less severely affected, with alpha-1 antitrypsin levels 60 to 70 per cent of normal. Common genotypes for people with A1AT deficiency are SS, ZZ, MS, MZ, SZ. However, not everyone with A1AT deficiency develops clinically significant disease. The different genotypes will lead to different serum levels of A1AT. It is the serum level of A1AT that will determine the likelihood of developing clinically significant disease. Most patients with clinical disease are homozygous SS or ZZ. They have the lowest serum levels of A1AT.

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18. A heterozygote MS or MZ will be a carrier of the disease. They produce lower than normal levels of A1AT (around 35% of normal) but do not develop clinically significant disease.

19. In 1991, scientists produced the first ever transgenic sheep called Tracey. The human DNA she contained helped her to produce and secrete A1AT in her milk. The aim was to extract the A1AT and use it for the treatment of people with A1AT deficiency.

20. A1AT deficiency is an under-recognised disease which should be considered in any young patient presenting with COPD or in any patient with severe, aggressive COPD.

21. It is thought there are over 3 million people living with the COPD in the UK, of which only about 900,000 have been diagnosed. This is because many people who develop symptoms of COPD do not get medical help because they often dismiss their symptoms as a 'smoker's cough'.

22. COPD affects more men than women, although rates in women are increasing. It is important that COPD is diagnosed as early as possible so treatment can be used to try to slow down the deterioration of your lungs. You should see your doctor if you have any of the symptoms mentioned above.

23. COPD is usually diagnosed after a consultation with your doctor, which may be followed by breathing tests. Being diagnosed early means you will receive appropriate treatment, advice and help to stop or slow the progression of COPD.

24. At a consultation, your doctor will ask about your symptoms and how long you have had them, and whether you smoke, or used to smoke. They will examine you and listen to your chest using a stethoscope. You may also be weighed and measured in order to calculate your body mass index (BMI).

25. Your doctor will also check how well your lungs are working with a lung function test that involves breathing into a machine called a spirometer. The spirometer takes two measurements: the volume of air you can breathe out in one second (called the forced expiratory volume in one second or FEV_1) and the total amount you can breathe out (called the forced vital capacity or FVC).

26. You may be asked to breathe out several times to get a consistent reading. The readings are compared with normal measurements for your age, which can show if your airways are obstructed.

27. You may have other tests as well as spirometry. Often, these other tests will help the doctor rule out other conditions that cause similar symptoms. For example, a chest X-ray will show whether you have another lung condition which may be causing symptoms, such as lung cancer. A blood test will show whether your symptoms could be due to anaemia, as this can also cause breathlessness.

28. Some people may need more tests. The tests may confirm the diagnosis or indicate the severity of your COPD. This will help you and your doctor plan your treatment.

29. An electrocardiogram (ECG) or echocardiogram may be used to check the condition of your heart. An ECG involves attaching electrodes (sticky metal patches) to your arms, legs and chest to pick up the electrical signals from your heart. An echocardiogram uses sound waves to build a detailed picture of your heart. This is similar to an ultrasound scan.

30. To confirm you have COPD and not asthma, your doctor might ask you to take regular measurements of your breathing using a peak flow meter, at different times over several days. The peak flow meter measures how fast you can breathe out.

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31. The level of oxygen in your blood can also be measured using a pulse oximeter, which looks like a peg and is attached to the finger. If you have low levels of oxygen, you may need an assessment to see whether extra oxygen would help you. If the condition runs in your family or you developed the symptoms of COPD under the age of 35 and have never smoked, you will probably have a blood test to see if you are alpha-1-antitrypsin deficient.

32. Some people may need a CT scan. This provides more information than an X- ray and can be useful in diagnosing other lung diseases or assessing changes to your lungs due to COPD. The doctor may also take a sample of phlegm to check whether it has been infected.

33. Although the damage from COPD that has already occurred to your lungs cannot be reversed, you can slow down the progression of the disease. Stopping smoking is particularly effective at doing this.

34. Treatments for COPD usually involve relieving the symptoms with medication, for example by using an inhaler to make breathing easier.

35. There is no cure for chronic obstructive pulmonary disease (COPD), but treatment can help slow the progression of the condition and reduce the symptoms. The best way to prevent COPD from getting quickly worse is to stop smoking and avoid further damage to your lungs. There is support available to help you quit. Stopping smoking may help for people with COPD to help them feel better and is the only proven way to reduce the rate of decline in lung function in people with COPD.

36. Stopping smoking at an early stage can make a huge difference. Any damage already done to the airways cannot be reversed, but stopping smoking can slow the rate at which the condition worsens.

37. If COPD is in the early stages and symptoms are mild, no other treatments may be needed. However, it is never too late to stop smoking. People with fairly advanced COPD are likely to benefit from quitting, which may prevent further damage to the airways.

38. Research has shown you are sometimes more likely to give up smoking successfully if you use healthcare support alongside stop smoking medicines such as tablets, patches or gum.

39. There are also medicines that can help relieve the symptoms of COPD. The type of medicine you take will depend on where your COPD is and what symptoms you have. You may have to try different medicines to find which suits you best. If an inhaler is prescribed for you, your doctor, practice nurse or pharmacist can explain how to use it. They will check you are using it properly. Most people can use an inhaler successfully, but if you are having problems, a spacer or a different type of inhaler device may help you take your medicines correctly. A spacer is a device that increases the amount of medication that reaches the lungs.

40. Short-acting bronchodilator inhalers deliver a small dose of medicine directly to your lungs, causing the muscles in your airways to relax and the airways to open up. There are two types of short-acting bronchodilator inhaler: beta-2 agonist inhalers, such as salbutamol and terbutaline, and antimuscarinic inhalers, such as ipratropium. The inhaler should be used when you feel breathless and this should relieve the symptoms.

41. If a short-acting bronchodilator inhaler does not help relieve your symptoms, your doctor may recommend a long-acting bronchodilator inhaler. This works in a similar way to a short-acting bronchodilator, but each dose lasts for at least 12 hours. There are two types of long-acting bronchodilator inhalers: beta-2 agonist inhalers, such as salmeterol and formoterol, and antimuscarinic inhalers, such as tiotropium.

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42. Steroid inhalers, also called corticosteroid inhalers, work by reducing the inflammation in your airways. If you are still getting breathless or having flare-ups (when symptoms are particularly bad) even when taking long-acting bronchodilator inhalers, your doctor may suggest including a steroid inhaler as part of your treatment. Most people with COPD will be prescribed a steroid inhaler as part of a combination inhaler.

43. Often, people with COPD have to take a combination of medicines. In addition, many people keep different medicines available in case they have a flare-up. Your doctor will discuss the best options with you. If you are getting breathless or having flare-ups when using a combination of inhalers, your doctor may prescribe theophylline tablets. Theophylline also causes the muscles of your airways to relax and open up.

44. When you have been taking theophylline tablets regularly, you may need to give a blood sample to measure the amount of theophylline in your blood and help your doctor prescribe the appropriate dose of tablet. This will allow you to get the correct dose of theophylline while reducing the likelihood of side effects. Due to the risk of potential side effects, such as increased heart rate and headaches, other medicines, such as a bronchodilator inhaler, are usually tried before theophylline.

45. Taking mucolytic tablets or capsules can also help relieve symptoms. Mucolytics, such as carbocisteine, make the mucus and phlegm in your throat thinner and easier to cough up. They are particularly beneficial for people with a persistent cough and lots of thick phlegm or who have frequent or bad flare-ups.

46. If you have a chest infection, your doctor may prescribe a course of antibiotics. Steroid tablets may also be prescribed as a short course if you have a bad flare-up. They work best if they are taken as the flare-up starts, so your doctor may give you a supply to keep at home. Occasionally, you may have to take a longer course of steroid tablets. Your doctor will give you the lowest effective dose and monitor you for side effects. It is not usual if steroid tablets are given for less than three weeks.

47. Nebulised medicines can be helpful for severe cases of COPD if other inhaler devices have not worked effectively. A nebuliser is a machine that administers nebulised medicine through a mouthpiece or face mask. The medicine is in a liquid form and is converted into a fine mist. This enables a large dose of medicine to be taken in one go. You can usually choose whether to use nebulised medication with a mouthpiece or a facemask. Your doctor will advise you on how to use the machine correctly.

48. If the oxygen level in your blood is low, you may be advised to have oxygen at home through nasal tubes, also called a nasal cannula, or through a mask. Oxygen is not a treatment for breathlessness, but it is helpful for some patients with persistently low oxygen levels in the blood.

49. You will probably be referred for more detailed assessment to see whether you might benefit from long-term oxygen therapy. If you are prescribed long term oxygen therapy, it must be taken for at least 15 hours a day in order to be effective. The tubes from the machine are long so you will be able to move around your home while you are connected. Portable oxygen tanks are available if you need to use oxygen away from home. The aim of long-term oxygen therapy is to extend your life.

50. Do not smoke when you are using oxygen. The increased level of oxygen produced is highly flammable, and a lit cigarette could trigger a fire or explosion.

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51. Non-invasive ventilation (NIV) helps a person breathe using a portable machine connected to a mask covering the nose or face. You may receive it if you are taken to hospital because of a flare-up. You may be referred to a specialist centre to see if home NIV could help you. NIV is used to improve the functioning of your lungs. Pulmonary rehabilitation is a programme of exercise and education designed to help people with chronic lung problems. It can increase your exercise capacity, mobility and self-confidence.

52. Pulmonary rehabilitation is based on a programme of physical exercise training tailored to your needs. It usually involves walking or cycling, and arm and strength-building exercises. It also includes education about your disease for you and your family, dietary assessment and advice, and psychological, social and behavioural changes designed to help you cope better. A rehabilitation programme is provided by a multidisciplinary team, which includes physiotherapists, respiratory nurse specialists and dietitians.

53. Pulmonary rehabilitation takes place in a group and the course usually lasts for about six weeks. During the course, you will learn more about your COPD and how to control your symptoms. Pulmonary rehabilitation can greatly improve your quality of life.

54. The National Institute for Health and Clinical Excellence (NICE) has produced guidance about the diagnosis and treatment of COPD. The Department of Health has been working with patients and healthcare providers to produce a strategy for COPD.

55. Surgery is only an option for a small number of people with COPD. Simple steps such as keeping healthy, being as active as possible, learning breathing techniques, and taking your medication can help you to reduce the symptoms of COPD.

56. Chronic obstructive pulmonary disease can have an impact on many aspects of your life. However, there are some simple tips and techniques to help reduce its impact.

57. Self-care involves taking responsibility for your own health and wellbeing with support from people involved in your care.

58. People living with long-term conditions can benefit enormously if they receive support for self-care. They can live longer, have less pain, anxiety, depression and fatigue, a better quality of life and are more active and independent. It is important to take your medication as prescribed, even if you start to feel better. Correct medication can help prevent flare-ups. If you have questions or concerns about the medication, taking or side effects, talk to your healthcare team.

59. It may also be helpful to read the information leaflet that comes with the medication about possible interactions with other drugs or supplements. Check with your healthcare team if you plan to take any over-the-counter remedies, such as painkillers, or any nutritional supplements. This is because these can sometimes interfere with your medication.

60. Because COPD is a long-term condition, you'll be in regular contact with your healthcare team. A good relationship with the team allows you to easily discuss your symptoms or concerns. The more the team knows, the more they can help you.

61. Regular meetings with a healthcare professional may also mean that any complications of COPD are spotted early. These include: cor pulmonale, a condition where there is raised pressure in the arteries of the lungs (the pulmonary arteries), fluid retention in the body, and depression.

62. Everyone with a long-term condition such as COPD is encouraged to get a yearly flu jab each autumn to protect against flu. They are also recommended to get an anti-pneumococcal vaccination, a one-off injection that protects against serious infection caused by pneumococcal bacteria.

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63. Check the forecast because the weather might have an effect on COPD symptoms. Cold spells lasting at least a week and periods of hot weather and humidity can cause breathing problems.

64. To reduce symptoms of COPD and chances of a flare-up, there are certain things that should be avoided if possible, including: dusty places, fumes, such as car exhausts, smoke, air freshener sprays or plug-ins, strong-smelling cleaning products, hairspray and perfume, unless there is plenty of ventilation. People with COPD who exercise or keep active regularly have improved breathing, have less severe symptoms and a better quality of life.

Acknowledgements

clinical trials http://www.nhs.uk/Conditions/Chronic-obstructive-pulmonary-dis[illegible]/Pages/clinical-trial.aspx

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