

Lung health

Looking after your lungs

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Supporting families affected
by primary and secondary
immunodeficiency

About this booklet

This booklet provides information on lung health in immunodeficiency. It has been produced by Professor John Hurst, Professor of Respiratory Medicine at University College London, and the Immunodeficiency UK Medical Advisory Panel and Patient Representative Panel, to help answer the questions patients and their families may have. It should not, however, replace advice from a clinical immunologist or a respiratory physician.

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Introduction

When people with a primary or secondary immunodeficiency disease have an infection, it can make them more susceptible to developing long-term problems with their lungs. This booklet gives a range of helpful information on this topic.

If you have an immunodeficiency and develop any respiratory problems, it is important to get in touch with your GP. Early diagnosis of problems and prompt treatment will help to ensure that your lungs are kept as healthy as possible.

What do our lungs do?

Our lungs help us breathe in oxygen and get rid of carbon dioxide. We use our lungs a lot. Over the course of a day we each take 17,000–30,000 breaths and breathe in around 11,000 litres of air. Lungs also act as a defence against infection and harmful environmental factors, such as dust, smoke and pollen.

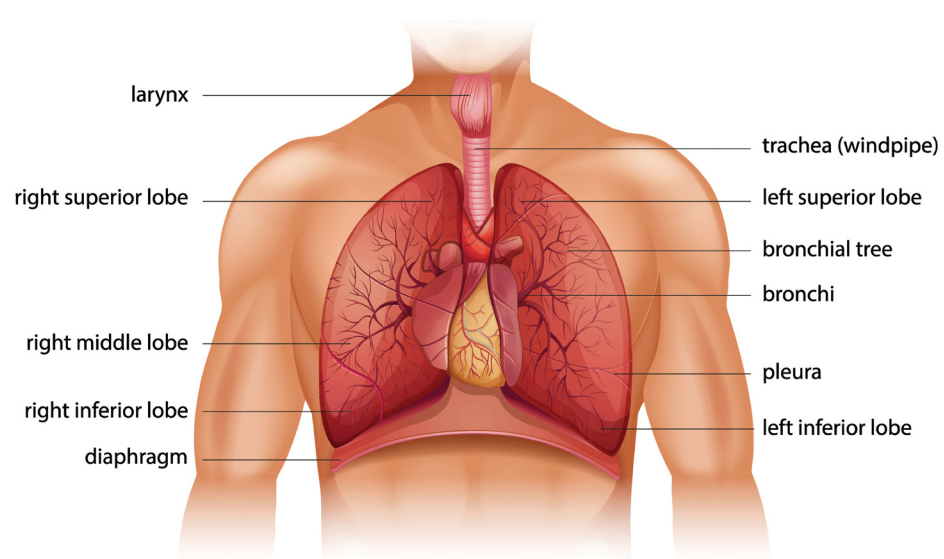
Lungs are designed to get oxygen into the bloodstream. Oxygen is necessary to generate energy, which keeps body tissues and cells healthy. Sometimes when people's lungs are not working properly they can have low blood oxygen levels, and individuals are given oxygen to help resolve this.

Our noses act as the first defence against harmful materials that we inhale, such as smoke, pollution and bacteria, and our lungs act as the second. Lungs have sticky mucus lining the surface that traps microbes and helps their protective white blood cells (phagocytes) to swallow and kill bacteria, and destroy harmful materials. Lungs also have tiny, brush-like fibres, known as cilia, to keep our airways clear. The cilia waft mucus up to the throat, where it is swallowed.

What can happen to my lungs if I keep getting infections?

If you get lots of infections, which is associated with inflammation, this can permanently enlarge the airways. This is known as bronchiectasis (pronounced 'bron-key-ect-a-sis'). This causes the airways to become baggy, inflamed and easily collapsible. Also, the cilia don't work as well, so you can't clear away microbes (bacteria, fungi and viruses) as efficiently. Bronchiectasis can be diagnosed using a CT scan (X-rays that build up a layer by layer picture of tissues in your body). Sometimes scar tissue (also called 'pulmonary fibrosis') can build up in the lungs, and this reduces how effective they are at transporting oxygen to red blood cells.

FIGURE 1: Structure of the lungs



The lungs are located within the upper part of the body called the thorax. They are surrounded by the ribcage (which protects them), and in between the ribs are intercostal muscles, which play a role in ventilating the lungs.

Beneath the lungs is a muscular sheet called the diaphragm. The diaphragm separates the lungs from the abdomen and it too plays a role in ventilating the lungs.

Within the lungs is a network of tubes through which air is able to pass. Air is firstly warmed, moistened and filtered as it travels through the mouth and nasal passages. It then passes through the trachea and down one of the two bronchi and into one of the lungs.

After travelling into the bronchial tree, which is made up of bronchioles, air finally passes through some of the millions of tiny sacs called alveoli, which have the specialised surfaces for gas exchange. Lungs have about 300 million alveoli with a total surface area of about 75 square metres.

Why can't I breathe properly when I have pneumonia or a chest infection?

When you have pneumonia or a chest infection it is harder for your lungs to absorb oxygen from the air you breathe. Also, more of the blood gets diverted to the parts of the lungs which are affected by the infection. Both of these factors make it harder to breathe properly.

Why is it important to keep my lungs as healthy as possible when I have an immunodeficiency?

We often don't consider the important role our lungs play in keeping us strong and well. It's not until we experience problems breathing that we take notice. But the truth is, like the rest of our body, our lungs need looking after. Breathing feeds oxygen to every cell in the body. Without sufficient oxygen, people are more prone to health problems, including respiratory illnesses.

Helping to keep your lungs as healthy as possible prevents them from deteriorating, by limiting the damage caused by infection and inflammation.

The causes of some lung disease in antibody deficiency (known as GLILD or granulomatous-lymphocytic interstitial lung disease) are unknown, but some may be driven by infection.

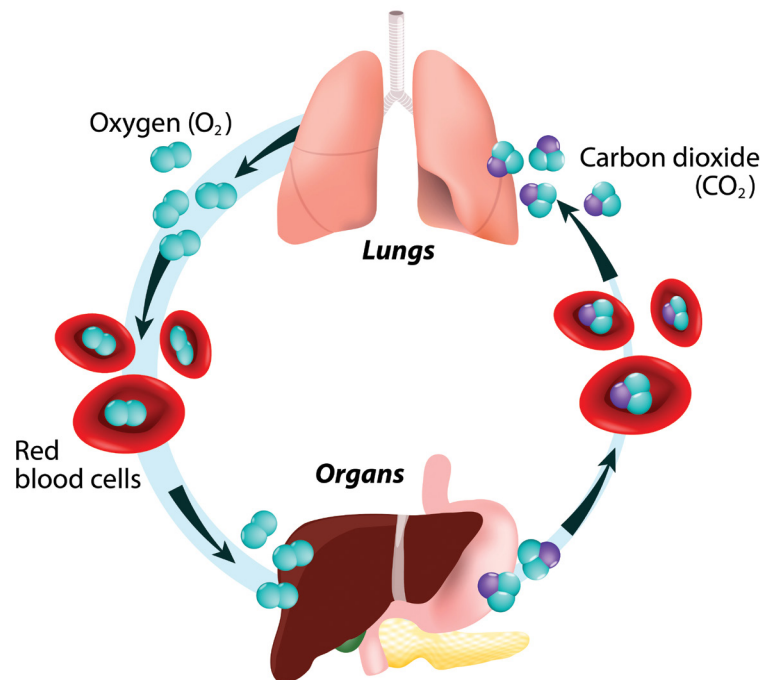
Asthma and chronic obstructive pulmonary disease are common in otherwise healthy individuals, and can also occur in immunodeficiency but are not usually caused by it. These underlying lung problems will independently increase the likelihood of infection, so require careful management to maximise lung health.

How can I keep my lungs healthy?

Here are some key things that will help:

- Don't smoke or vape and avoid breathing in second-hand smoke (see page 13).
- Help prevent infection by keeping up to date with vaccinations, including the seasonal flu vaccination.
- Take the medicines given by your doctor regularly and as recommended.
- Know the signs of a chest infection (see page 7).
- Take regular exercise (see page 13).
- Always get advice if you are worried; ask your doctor if you should see a chest specialist.
- Discuss with your healthcare professional whether you should have a test of your lung function (see page 9).

FIGURE 2: Exchange of oxygen and carbon dioxide in the body



The exchange of oxygen (O₂) and carbon dioxide (CO₂) in the body is the most important role of the respiratory system and occurs when we breathe in and out. Oxygen breathed in by the lungs passes through the alveoli into the blood and is then transported around the body by red blood cells, feeding the body's tissues and organs and keeping them healthy. The waste product CO₂ is then carried back by red blood cells or dissolved in the plasma (water component of blood) to the lung's alveoli and breathed out.



Don't smoke

People with an immunodeficiency are strongly advised not to smoke and to avoid breathing in second-hand smoke in order to prevent damage to their lungs.

The recommendation is to avoid vaping too.

What are the symptoms of a lung infection?

Symptoms include:

- a persistent cough
- breathlessness
- the production of sputum or phlegm (mucus that you might cough up from the airways)
- fever.

The best sign of a lung infection is a **change** in symptoms from your usual level. So, for example, if you usually produce some phlegm and if that phlegm becomes darker in colour, or there is more of it, then that may signify infection.

Always seek help when you have any of the above symptoms. Getting early treatment could reduce damage to your lungs.

Sputum and coughing

Sputum, the excessive sticky mucus present in your lungs, and coughing are common problems in people with lung damage.

Sputum has to be cleared from the chest so that you can breathe more easily. It becomes a problem when it is:

- thick
- sticky and difficult to cough up
- infected
- abundant.

Thick, sticky sputum can cause long, tiring and painful bouts of coughing, particularly in the early morning.

In many people with bronchiectasis the sputum can be transparent with a white or greyish tinge when it is free from infection. Other people with bronchiectasis may have yellow or green sputum all the time. When you have an infection though, it causes the sputum to thicken and turn yellow, creamy or green.

When you have an infection/bronchitis it is best treated as early as possible with an antibiotic prescribed by a clinician. The clinician may ask you to send a sputum sample to your GP or hospital. In the case of a child, a cough swab may be easier to obtain.

There are exercises to help clear sputum from your lungs - see page 10.

What should I do if I think I have a chest infection?

Your lung health should be checked as part of your regular check-up routine, but if you develop the symptoms of a chest infection during the interim period, contact your GP as soon as possible. Remember to tell him/her you have an immunodeficiency. Some patients have emergency antibiotics to have at home. If you have these, it might be appropriate to start taking the course – follow the guidance you have been given and ask if you are not sure.

What medicines are available to reduce lung infections and inflammation?

Antibiotics can be used to treat and prevent infections. The most common antibiotics used to treat chest infections would include penicillin drugs, such as amoxicillin and co-amoxiclav, or different types called macrolides (e.g. clarithromycin) or tetracyclines (such as doxycycline). If you are allergic to penicillin then alternatives will be chosen.

Sometimes antibiotics are used in a different way – not to treat but to prevent infections. This might be suggested to you if you are having frequent or severe chest infections. The most common antibiotic used in this way is called azithromycin, and it is often taken just three times each week. Using an antibiotic in this way – called a ‘prophylactic’ – is a decision that has to be considered carefully. Some patients will be put on nebulised (inhaled) antibiotics for the same reasons. Using an antibiotic long term has the risk of side effects or of the bugs becoming resistant to the antibiotic. These risks have to be balanced against the benefits of having fewer infections.

Your doctor may give you a broad range antibiotic but sometimes he/she will want to identify the microbe causing your infection, if it doesn’t respond to treatment. This will involve you giving a sample of sputum (mucus that you might cough up from the airways). In practice, this isn’t so easy because the sputum test results take a few days to come back and it is not usually wise to delay starting treatment. This means that your doctor will usually recommend treatment with an antibiotic that is known to work against a range of lung infections, or be guided by the results of previous sputum tests. People with established lung disease, such as bronchiectasis, may sometimes need higher doses of antibiotics, or longer courses, to stop the infection.

Other medicines that may be used include inhaled steroids, bronchodilators (which help to relax the airways), allergy treatments and decongestants to shrink swollen mucous membranes.

What is a sputum test?

A sputum test is a test used to detect or identify bacteria, viruses or fungi that may be causing chest problems. It involves giving a sample of sputum. Some people don’t produce sputum. If this is the case, and it is really important to know which infection is in the lung, sputum production can be encouraged using a cold salt-water mist (saline nebuliser). Alternatively, an examination with a telescope, called a bronchoscopy, might be carried out to collect samples from deep inside the lung. The sputum sample is placed in a sterile container and sent to the laboratory to see if it grows a certain type of bacteria or fungi. Viruses can be detected too. From these cultures, doctors are able to identify the microbes involved and give better-targeted drugs to clear the infection.

What other tests will doctors use to assess my lung health?

Doctors use lung function tests, also called pulmonary (pronounced ‘pull-mon-ary’) function tests, to diagnose and monitor your lung health and to see if there is any damage. These tests may also be used to check how well treatments are working. Lung function tests are usually painless and rarely cause side effects. The technician will ask you to blow as hard as you can to get the best results, and the tests are repeated a few times, which can make people feel tired.

Lung function tests measure:

- how much air you can blow in and out of your lungs
- how much air you can blow out of your lungs and how fast you can do it. This amount is compared with that of other people your age, height and sex. This allows your doctor to see whether your lungs are working normally
- how well your lungs deliver oxygen to your blood
- the strength of your breathing muscles.

What lung function tests are used?

The breathing tests most often used are:

- Spirometry (pronounced ‘spi-rom-eh-tre’). This test measures how much air you can breathe out. It also measures how fast you can blow air out. This is the simplest and most important test. It requires you to blow out as hard as you can to get the best results.
- Lung volume measurement. This test, taken along with spirometry, measures how much air remains in your lungs after you breathe out fully.
- Lung diffusion capacity. This test assesses how well oxygen passes from your lungs to your bloodstream.

Some tests measure the oxygen level in your blood. These are called blood oxygen tests. There are two tests:

- Pulse oximetry, which measures your blood oxygen level using a light attached to a finger probe. The test is painless and only if the results indicate a problem would your clinician normally suggest a blood oxygen test.
- Blood oxygen test, when the doctor inserts a needle into an artery, usually in your wrist, and takes a sample of blood. The oxygen level of the blood sample is measured. This test is more accurate than pulse oximetry. Some units are now able to do this test from the ear lobe too. You may feel some discomfort during an arterial blood oxygen test – when the needle is inserted into the artery.

However, these tests may not show what is causing breathing problems. This has to be assessed separately and may involve giving a sample of sputum.

Physiotherapy and lung health

Physiotherapists have a key role in maintaining the health of your lungs. Perhaps the most important role is teaching people techniques to clear sputum effectively. Being very breathless can be frightening, and physiotherapists are also trained to teach people specific breathing techniques that help with these symptoms. They also run exercise and education classes called ‘pulmonary rehabilitation’ that can be very effective for people with more severely affected lungs.

A respiratory physiotherapist and respiratory physiotherapy classes are accessed via your respiratory clinic and by recommendation from your specialist. This usually happens as part of a combined lung-immunodeficiency clinic appointment.

Exercises to improve your breathing and to help clear sputum

Although it isn’t usually possible to stop the production of excessive sputum, you can improve the way it is cleared from your chest.

The techniques used to clear the chest are:

- relaxed breathing control, to avoid tightness of the chest and exhaustion
- deep breathing exercises, to loosen the sputum
- huffing.

If you produce only about two or three tablespoonfuls of sputum a day, you can do the exercises either sitting or lying down, first on one side and then on the other.

However, if you have bronchiectasis and produce more than one cupful of sputum a day, you will need to lie in positions in which gravity helps to drain the affected areas of the lungs. This is often known as postural drainage. A physiotherapist must advise you on the postural drainage positions that are most appropriate for you.

Even if you can’t manage to take active exercise, breathing exercises are essential to keep your lungs clear and free of infection. Practising breathing exercises can help you to:

- breathe more efficiently and make better use of your lungs
- control the unpleasant, panicky feeling of breathlessness
- remove sputum from your chest.

Your physiotherapist can show you positions to help reduce your breathlessness when it is bad. However, there are simple exercises that will help your general breathing. These are relaxed breathing control, huffing and deep breathing.

Make sure you discuss these with your doctor or physiotherapist.

Relaxed breathing control

This exercise will help you to control breathlessness, particularly after coughing, huffing or taking active exercise.

Follow these steps:

- Relax your head, neck and shoulders. Your physiotherapist can show you some exercises to achieve this.
- With your shoulders dropped, place your hands across your stomach, just below your ribs.
- Breathe out gently through your mouth.
- After a long breath out, breathe in gently through your mouth or nose, keeping your shoulders relaxed.
- Repeat this exercise four to six times.

This exercise should not tire you. If it does, ask your physiotherapist for advice, as you might not be doing it correctly.

Huffing

This is a forced breath out of an open mouth that helps to move the secretions upwards; for example, as if creating a mist on a window.

There are two types:

- Take a medium-sized breath in and try and ‘squeeze’ the breath out.
- Take a big breath in and do a shorter, sharper breath out.

You should start with the first type of huff and as the secretions move upwards then try the second type.

Always remember to allow for an essential short period for relaxation and gentle breathing control after every one or two huffs. Between 15 and 30 seconds should be enough. This will prevent the feeling of tightness in the airways and chest, and allow you to relax and recover your breath comfortably. Remember, relaxed breathing control is also helpful after a bout of coughing.

Deep breathing

This exercise helps you make better use of the lower parts of your lungs.

Follow these steps:

- Relax your head, neck and shoulders.
- Place your hands across your stomach, just below your ribs.
- Breathe out gently.
- Keeping your shoulders relaxed, take a deep breath in through your mouth or nose, feeling your ribs rising under your hands.
- Hold the deep breath for a second, then breathe out gently through your mouth.
- Repeat the exercise three or four times; then rest.

Children can be encouraged to take deep breaths through the use of blowing games, for example, blowing tissues, or even by playing a musical instrument.

Clearing sputum in children

Babies and small children will swallow their sputum, but as soon as a child is old enough he or she should be encouraged to cough it up and spit it out (expectorate).

Postural drainage for small children is best performed across the parent's knee, before a feed. As the child grows older, postural drainage can be performed over a wedge of foam or pillows. It is often difficult to persuade a toddler to remain in one position for any length of time. However, running or jumping up and down between short sessions of postural drainage can actually help, as these activities often stimulate a cough.

Seek advice from your physiotherapist before practising postural drainage on your child.

Lung health when living with an immunodeficiency

Smoking

Smoking is harmful even if you are healthy, but if you have an immunodeficiency it's even more important not to do it. When you smoke, the brush-like fibres in your lungs are inactivated or destroyed. This allows thick mucus to accumulate in the lungs and reduces your defence against infection. It's also best to avoid smoky places and breathing in second-hand smoke.

Cigarette smoking also increases the risk of lung cancer and disease in other parts of the body, especially heart attacks and strokes. There is lots of help available these days to help you stop smoking – speak to your doctor, nurse or local pharmacist.

Smoking marijuana has been associated with an increased risk of infection in the lungs of patients who have poor immunity. This is in addition to all the many other harmful effects of smoking 'ordinary' tobacco and is due to contamination with *Aspergillus*, a type of fungus.

Whilst vaping is likely to be safer than smoking, we have no long-term data. Therefore, the recommendation for people living with immunodeficiency has to be avoid vaping too.

Air pollution

Hot weather conditions tend to come with a significant rise in pollution levels. For people with an immunodeficiency living in cities and with pre-existing lung disease it is worth taking extra precautions. If you have a wheezy condition, please contact your doctor and see if you should be on more treatment at this time.

It may be impossible to avoid travel into high pollution environments, owing to work or education commitments, but if possible travel at non-peak times and avoid being out in the middle of the day, when heat and pollution levels are at their highest. The general advice to keep cool when possible and ensure adequate hydration with water applies to everyone.

Getting active and lung health

The effect of lung infections in immunodeficiency can lead to unpleasant feelings of breathlessness, even after short bursts of activity. As a result, it can be tempting to take the easy option and give up trying to lead an active life when you are well and free from infections. Don't! This in itself can pose problems: to your physical well-being, to your mood and to the way you feel about yourself. Isolating yourself and being confined to the home is not much fun for anyone.

Strange as it may seem, the solution lies not in stopping all exercise but in ensuring that you get enough exercise. Just a few simple exercises performed for a few minutes a day can make all the difference to your physical and mental well-being. Remember though, when you do have an infection, it may not be wise to exercise.

The right sort of exercise does you no harm at all and can do you a great deal of good. It allows the body to use oxygen more efficiently and can help your lungs work better. Only exercise until you feel out of breath. Be careful when you are not feeling well; exercise at your own pace and don't do too much. Ask to discuss this with a clinician if you are not sure.

How much physical activity you need to keep fit and healthy depends on your age. The NHS website (www.nhs.uk/Livewell/fitness/Pages/physical-activity-guidelines-for-adults.aspx) gives useful recommendations for different age groups. For example, the guideline for adults aged 19–64 years old is to complete 150 minutes of weekly physical activity, incorporating exercise into your routine for 30 minutes, five days a week.

Even when it causes breathlessness, exercising can:

- increase the fitness of your muscles
- reduce the fear of breathlessness
- reduce the feeling of breathlessness
- increase your ability to exercise
- improve your well-being and self-confidence
- break the vicious circle of inactivity.

Discuss what exercises might be suitable for you with your clinician.

Other useful information

The booklets 'Primary immunodeficiency and respiratory disorders' and 'A guide for pulmonologists' are available from the Immunodeficiency UK website - www.immunodeficiencyuk.org.

The British Lung Foundation has a range of information about different lung conditions, including bronchiectasis and chronic obstructive pulmonary disease (COPD) - www.blf.org.uk/support-for-you.

An information leaflet on granulomatous-lymphocytic interstitial lung disease (GLILD) - <https://bit.ly/31euNIS>.

National Institute for Health and Care Excellence (NICE) guidelines on antibiotic prescribing in bronchiectasis - www.nice.org.uk/guidance/ng117.

Glossary

allergy - an exaggerated sensitivity resulting from a heightened or altered reactivity of the immune system to an external substance.

antibody - a type of protein (immunoglobulin) that is produced by certain types of white blood cells (plasma cells - a type of B-cell). The role of antibodies is to fight bacteria, viruses, toxins and other substances foreign to the body.

antibody deficiency - covers a range of disorders resulting from the failure of the immune system to produce sufficient antibodies in the bloodstream to fight infections.

asthma - a common long-term condition that can cause coughing, wheezing, chest tightness and breathlessness; it is usually driven by allergy.

bronchiectasis - a widening of the tubes (bronchi) that lead to the air sacs of the lung; this can happen because of repeated bouts of infections.

bronchodilators - medicines that make breathing easier by relaxing the muscles in the lungs and widening the airways.

CAT scan - also known as a CT scan. A specialised X-ray test that gives pictures of the inside of the body.

chronic obstructive pulmonary disease - a collection of smoking-related lung diseases, including chronic bronchitis and emphysema, associated with narrowing of the airtubes.

decongestants - medicines used to relieve swollen mucous membranes. They are available as tablets or as a nasal spray.

expectorate - to cough up fluid from the lungs or to spit.

mucus - a protective layer in the airways, eyes and nose.

phagocyte - a type of white blood cell that protects the body by eating (phagocytosing) dirt, bacteria and dead or dying cells.

phlegm - mucus from the lungs.

pneumonia - a swelling (inflammation) of the tissue in one or both of your lungs. It is usually caused by an infection.

prophylactic/prophylaxis - something that works to defend or protect against disease.

sputum - mucus that is coughed up from the lower airways.

steroids - also known as corticosteroids. These are medicines used to reduce inflammation and that affect the way the immune system works.

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About Immunodeficiency UK

Immunodeficiency UK is a national organisation supporting individuals and families affected by primary and secondary immunodeficiency.

We are the UK national member of IPOPI, an association of national patient organisations dedicated to improving awareness, access to early diagnosis and optimal treatments for PID patients worldwide.

Our website has useful information on a range of conditions and topics, and explains the work we do to ensure the voice of patients with primary and secondary immunodeficiency is heard. If we can be of any help, please email us or call on the number above, where you can leave a message.

Support us by becoming a member of Immunodeficiency UK. It's free and easy to do via our website. Members get monthly bulletins.

Immunodeficiency UK is reliant on voluntary donations. To make a donation, please go to **www.immunodeficiencyuk.org/donate**



Supporting families affected
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immunodeficiency