



**DIAGNOSING AND TREATING
NON-CYSTIC FIBROSIS BRONCHIECTASIS
IN CHILDREN AND YOUNG PEOPLE**
understanding the professional guidelines



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1. Introduction

Who is this document for and what is it about?

This document explains the recommendations in the European Respiratory Society clinical guideline for the management of children and young people with bronchiectasis. It is aimed at parents or carers of children with bronchiectasis, or young people with bronchiectasis.

This document is aimed at people with an experience of bronchiectasis that exists separately to cystic fibrosis.

What is a clinical guideline?

Clinical guidelines are produced after a scientific process used to gather and evaluate the latest evidence in the field. Guidelines also consider the opinions of leading experts and the priorities of patients and carers who have experience of a condition. Clinical guidelines are aimed at healthcare professionals. They use them as a 'best practice' document in how to diagnose, manage and treat specific conditions.

What does this document include?

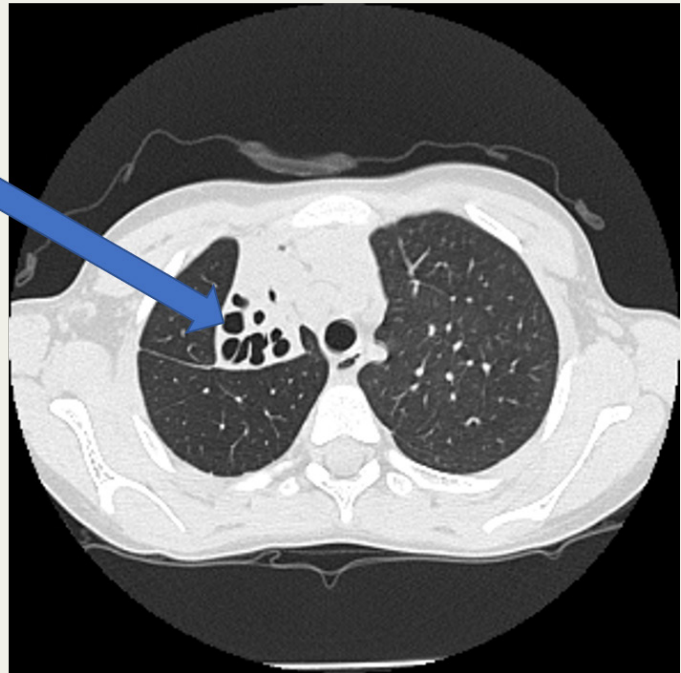
This document summarises the key points from the clinical guideline and explains them in a way that is more easily understood by people who do not work in a medical field.

It will cover what bronchiectasis is, how it is diagnosed and what treatments are used to help manage the condition. By providing this information in an accessible way, this document aims to help parents/carers and young people with bronchiectasis understand more about the condition and feel informed when making decisions about care.

2. What is childhood bronchiectasis?

Chest CT scan showing abnormally widened airways. In this part of the lungs, that is diagnostic of bronchiectasis

The rest of the lungs in this child are normal.



Bronchiectasis is a long-term lung condition where the airways, or tubes, in a person's lungs are damaged and become abnormally widened. The airways in the lung are lined with a layer of mucus that works to trap bacteria and help keep the airways clean. This mucus is constantly being moved upwards through the airways so that it can be coughed or swallowed.

When a person has bronchiectasis, the airway tubes in the lungs are less effective at clearing the mucus. As it gathers, it can become thicker, allowing bacteria to grow more easily. This can make a person more at risk of becoming ill with an infection.

Infections can cause the lungs to become inflamed (swollen), which can further damage or block parts of the lung. This can lead to symptoms like coughing up mucus, frequent chest infections, breathlessness, chest pain, wheeze and tiredness.



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Bronchiectasis can affect one, or several parts of the lung. It can be quite mild, where there is not a lot of mucus, or can be more severe where people might cough up more mucus.

Although the symptoms of bronchiectasis are similar between children and adults, there are differences in the way the condition is managed.

It is important to interrupt the cycle of infections at a young age, to prevent further damage or even reverse the damage that exists in the airways. This is achieved with diagnosing bronchiectasis as early as possible and finding the best treatment approaches. This will improve quality of life for children and young people living with the condition and aims to keep the lungs working well into adult life.

The guideline identifies 5 objectives for managing bronchiectasis in children and young people:

1. Help the young lungs to grow and work as they should.
2. Improve the quality of life for people living with the condition.
3. Reduce the number of infections and flare ups of symptoms.
4. Prevent any additional complications.
5. If possible, reverse the damage in the airways.



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3. How should childhood bronchiectasis be diagnosed?

A type of scan, called a multidetector chest computed tomography (MDCT) scan with high resolution (HRCT), is recommended to confirm a diagnosis of bronchiectasis in children and young people. This is a kind of X-ray that can build up a very detailed picture of the lungs. A radiologist or respiratory specialist can look at the detailed picture of the airways to find any damage or widening of the tubes.

Will any other tests be performed?

Alongside an MDCT scan with HRCT, other tests will also be carried out to help understand how severe the bronchiectasis is and what may have caused it. This information can help to inform decisions about treatments. All children should have the following tests:

- A sweat test – a small sample of sweat is collected from the arm. Doctors look at the salt levels in sweat to understand whether bronchiectasis is caused by cystic fibrosis.
- A breathing test such as spirometry, which measure lung volumes. This can help to show how much damage there is in the airways. This test will be done if the child is old enough to do the test.
- A blood test to take a full blood count and check general health.
- A blood test looking at how the immune system is working, which could help understand what has caused bronchiectasis.
- Sputum (mucus) samples – a sample of mucus is coughed into a pot and can be tested to look at bacteria in the lungs. This can help decide which treatments might work best.



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Experts also suggest that in places where tuberculosis (TB) or human immunodeficiency virus (HIV) are common, or a person has come into contact with someone with TB, tests to find these conditions should also be carried out as they may have an impact on bronchiectasis. Some children might need additional tests such as in-depth tests for how the immune system is working a bronchoscopy condition, called primary ciliary dyskinesia.

What is an exacerbation?

When symptoms get worse over the course of three days or more, it is called an exacerbation. It could include:

- Coughing more than normal.
- Feeling that more mucus is being coughed up, or the mucus is thicker and harder to move.
- Feeling more tired.
- Changes in behaviour or appetite.
- Some children also have additional symptoms such as wheeze and other chest sounds, and blood in the sputum.

This could be caused by a chest infection, but not all exacerbations come from infections.

Managing exacerbations is a key part of care for children and young people living with bronchiectasis and it is important to recognise the signs that an exacerbation is starting. These will be different from person to person. Exacerbations may require additional treatments to help manage any worsening in symptoms.

For children or young people who become very short of breath or have low oxygen levels, the exacerbation will be considered severe, regardless of the number of the days it has lasted for.

4. How to manage bronchiectasis in children?

Airway clearance

Clearing the sticky mucus from the lungs is an important treatment that can be practiced regularly at home. A respiratory physiotherapist will teach you the best exercises, depending on a child's age and how severe their symptoms are. This can include different breathing exercises and sitting in different positions to do them. As a person gets older, the techniques and frequency of the exercises usually change and can be incorporated into an active lifestyle which may include trampolining, swimming, running, cycling and playing wind instruments. A healthcare professional should review this at least twice a year to check the exercises are still helping.

Mucoactive treatment

Mucoactive agents are medicines that help people to clear mucus from their lungs. These medicines are currently recommended for use in adults with bronchiectasis who experience a very poor quality of life and struggle to remove mucus from their lungs.

The guideline for children and young people does not recommend that these medicines are used routinely. This is because there is no evidence from research to show that these medicines work in children. For 2 types of mucoactive medicines (bromhexine and rhDNAse), there is also a risk of side effects, such as more exacerbations and worsening lung function over time.

In certain cases, two specific types of mucoactive treatments, could be used: inhaled mannitol or hypertonic saline. These should only be used for young people who are old enough to manage taking the medicines and they should be shown by a



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healthcare professional how to do this. They should also only be used after taking an asthma medication called a short-acting beta2-agonist (SABA), and known as 'rescue medication'. This could help improve the effectiveness of the mucoactive medicines.

Antibiotics

Antibiotics are medicines that are used to fight bacterial infections. Bacteria are tiny germs that can make us sick. The guideline recommends that an antibiotic should be used for 14 days to treat exacerbations. The type of antibiotic used will be different, depending on how a person has reacted to antibiotics before and on the type of bacteria found in their airway samples. If the exacerbation does not improve with antibiotics, an injection directly into the veins, known as intravenous antibiotics, may be needed.

If a child or young person experiences repeated exacerbations, a long-term antibiotic (known as a macrolide antibiotic) may be recommended for at least 6-months, with regular check-ups to see if it is still working well. Macrolides are safe and given in a very low dose, often 3 times a week, just over during the winter period. The guideline suggests this should be given for people who have had three or more exacerbations in the last year, or a severe exacerbation that needed hospital treatment in the last year.

Some people may be carrying a bacteria known as *P. aeruginosa* (also called *Pseudomonas*) and will need to be treated to get rid of it. Evidence has shown that getting rid of this bacteria has helped reduce exacerbations in adults. Antibiotics are taken for up to 3-months to get rid of the bug completely.

Asthma medicines

In the past, some healthcare professionals have suggested taking medicine that is regularly used for people with asthma to help manage bronchiectasis. These medicines are usually delivered by inhaler and include medicines that help relieve symptoms of breathlessness and medicines that help prevent a worsening of these symptoms over time.

The guideline suggests that these medications, other than short-acting beta2-agonist (SABA) are not routinely used for children and young adults with bronchiectasis. This is because the evidence suggests that they do not work well for this group of people and could also have harmful effects. As mentioned above, one specific asthma medication known SABA, may be beneficial to take before taking a medicine designed to clear the mucus.

Surgery

Surgery for bronchiectasis is now uncommon. The operation would involve taking away the parts of the lungs that are no longer working with the aim of stopping the illness from spreading to the rest of the lungs. It should only be considered when all other treatment options have been tried and have not worked and a child's life is being severely impacted by their condition.

The guideline suggests a number of factors should be considered before deciding to have surgery: a child's age, how severe the symptoms are, whether the bronchiectasis has been found only in a specific part of the lung, what is causing the bronchiectasis and whether surgery is likely to improve a child's day-to-day life.



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Self-care

There are other skills and techniques that can help children and young people manage their condition themselves, alongside the medication given from doctors. These include:

- Eating healthily and getting a balanced diet.
- Getting enough vitamin-D – either from sunlight, food sources or a dietary supplement (always consult your doctor before taking a dietary supplement).
- Keeping active through regular exercise.
- Keeping up to date with childhood vaccinations and annual vaccinations, such as the flu jab.
- Avoiding smoking.

It is also important for all other people in the household to practice regular hand washing to help reduce the spread of infections. It can also help to avoid spending time with people that you know have cold or flu symptoms.

5. What check-ups will be needed?

The guideline suggests that check-up visits with a healthcare provider take place every 3-6 months. These appointments will be to check how well the child or young person is, look for any complications and test how well the lungs are working with spirometry tests.

For children who are able to cough up mucus, samples should be taken every 6-12 months. This will help healthcare providers learn more about the bacteria in the lungs and decide which antibiotics may work best in the future.



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Chest X-rays are not needed on a routine basis, unless there is an underlying issue that has not been resolved through other tests.

If a child or young person's condition is gradually getting worse over time, tests looking for new infections or to find any other underlying conditions should be carried out.

6. Further reading

Full clinical guideline

You can access the full clinical guideline at:

www.ers-education.org/guidelines/all-ers-guidelines/

Bronchiectasis patient priorities

Information and support for people diagnosed with bronchiectasis and their caregivers, developed with the help of patients and healthcare professionals across Europe.

www.europeanlunginfo.org/bronchiectasis

Child-BEAR-Net (Children's Bronchiectasis Advocacy Research Network)

A Clinical Research Collaboration (CRC) supported by the European Respiratory Society (ERS) and the European Lung Foundation (ELF) that brings together different stakeholders including parents and patients to advance science and clinical research to improve respiratory health and medicine for people diagnosed with bronchiectasis as children.

www.improvebe.org/



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EMBARC

EMBARC is a pan-European network committed to promoting clinical research and education in bronchiectasis, through sharing of protocols, research ideas and expertise.

www.bronchiectasis.eu

About ERS

European Respiratory Society (ERS) is an international organisation that brings together physicians, healthcare professionals, scientists and other experts working in respiratory medicine. It is one of the leading medical organisations in the respiratory field, with a growing membership representing over 140 countries. The ERS mission is to promote lung health in order to alleviate suffering from disease and drive standards for respiratory medicine globally. Science, education and advocacy are at the core of everything it does. ERS is involved in promoting scientific research and providing access to high-quality educational resources. It also plays a key role in advocacy – raising awareness of lung disease amongst the public and politicians. www.ersnet.org

About ELF

European Lung Foundation (ELF) was founded by ERS to bring together patients and the public with professionals. ELF produces public versions of ERS guidelines to summarise the recommendations made to healthcare professionals in Europe, in a simple format for all to understand. These documents do not contain detailed information on each condition and should be used together with other patient information and discussions with your doctor. More information on lung conditions can be found on the ELF website: www.europeanlung.org



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