Cystic Fibrosis Trust

Inhaled therapy for people with cystic fibrosis

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Trusted Information Creator

Patient Information Forum

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Contents

This factsheet covers:
- Different types of nebuliser systems and inhalers.
- Types of medication that can be inhaled.
- How to look after your nebuliser system.
- What to expect from your cystic fibrosis (CF) centre.

Keeping up with treatments is understandably hard and a burden on people with CF and their families. It's very common to struggle with taking inhaled medications. If you're struggling, talk to your CF team.

Nebuliser systems

What are nebuliser systems?

Nebuliser systems change liquid medication into a mist that can be breathed into the lungs. You may know them as just a 'nebuliser'.

In general, a nebuliser system is made up of:

- a nebuliser handset or pot – including a mouthpiece and a nebuliser pot that holds liquid medication
- and an air compressor or electronic part.

Nebuliser systems are used to get the medication straight to the lungs without having to go around other parts of the body like with tablets or injected medications. This can reduce side-effects.

A nebuliser system might be used instead of an inhaler because:

- you can have larger doses of some medication with a nebuliser.
- some inhalers can be difficult to use correctly – nebulising a medication can help to get more medication to your lungs.
- some medication is only available as a nebulised medication, for example DNase.

General nebulised medication advice

- **It is usually best to use a mouthpiece with your nebuliser system.** More medication can get to your lungs and some nebuliser systems don't work well with a mask. You shouldn't change to a facemask without discussing it with your CF team.
- **Your medication should be at room temperature before being nebulised.** Cold medication can make your chest tight and might run slowly through some systems. Ask your CF team how long you can leave your medication at room temperature.
- **Many nebuliser systems have some medication left over in the pot after nebulising has finished.** This is called 'residual volume' and is normal. You should not shake or tip your nebuliser system as the residual volume may be nebulised which can change the amount of medication that gets to your lungs. You should empty the residual volume out of the nebuliser system before cleaning. You shouldn't re-use this medication.
- **If your CF team asks you to add liquid to a medication, always add the correct amount.** Changing the amount of liquid may change the amount of medication that gets to your lungs.
- **You shouldn't mix nebulised medications unless your CF team asks you to.** Mixing medications may mean that they won't work in the way they're meant to.

What different types of nebuliser system are there?

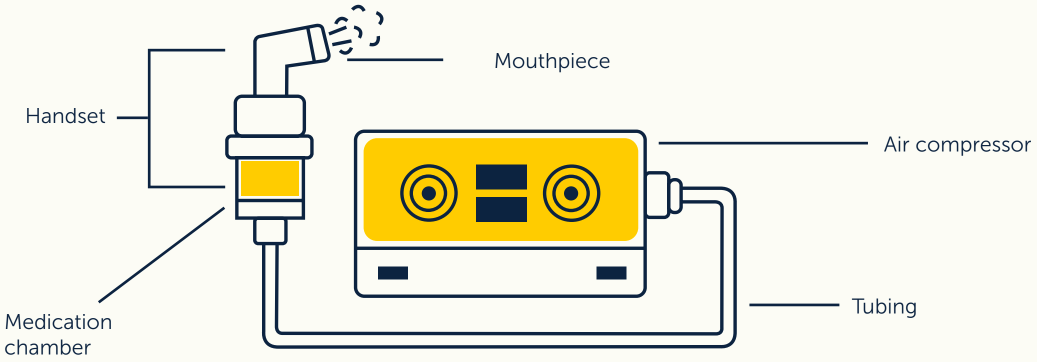
It is important that you use the nebuliser system given to you by your CF team as different nebulisers can change the amount of medication that reaches the lungs.

The medications you are taking may each need different types of nebuliser system or different handsets, so you may be given more than one.

If you are not sure if you have the best nebuliser system to suit you and your medication, talk to your CF team.

Conventional (jet)

Philips Respironics InnoSpire Delux® air compressor with SideStream® nebuliser, PARI Boy® compressor with PARI LC Plus®/Star® nebulisers



Conventional nebuliser systems, sometimes called jet nebulisers, are made up of a nebuliser pot and an air compressor. The compressor pumps air through the liquid medication and turns it into a mist. The nebulised medication is breathed in using a mouthpiece or facemask. Mouthpieces are recommended wherever possible as more of the medication gets to the lungs this way.

Conventional nebuliser systems deliver medication constantly and need to be switched off when the liquid medication has run out – the sound the nebuliser system makes will change to a ‘spluttering’ sound.

Portable air compressors are also available. They work in the same way but are smaller and lighter which may make traveling easier. They may have a chargeable battery or be able to use a 12-volt car socket for power. They might take longer to use and cannot be used with all medications. Ask your CF team if you are unsure.

Examples of portable air compressors:

- Phillips InnoSpire Mini®.
- Trek S Portable Aerosol System®.

Vibrating Mesh Technology

e-Flow® rapid, eTrack®

Vibrating Mesh Technology nebuliser systems are made up of a nebuliser handset and an electronic unit joined by a connection cord. The nebuliser handset has a metal mesh which vibrates. This moves the liquid medication through the tiny holes in the mesh. This turns the liquid into a mist of similar-sized droplets. Vibrating mesh systems deliver the mist of medication constantly and stop automatically when the liquid medication has been fully nebulised.

The advantages of Vibrating Mesh Technology systems are that:

- the medication is given faster
- the small similar-sized droplets are better at getting into your lungs
- they are almost silent
- they are smaller and more portable than conventional nebulisers
- they can run on batteries.

Your CF centre may offer you an eTrack® system as part of **CFHealthHub**. CFHealthHub is a programme which supports people with CF and CF teams to help to build habits to increase treatment taking and decrease the burden of treatment. An eTrack® records when you do your treatment on the CFHealthHub app. You can choose to share this with your CF team. Ask your CF team if your CF centre is part of CFHealthHub.

Adaptive aerosol delivery (AAD)

I-neb® AAD System

AAD nebuliser systems use Vibrating Mesh Technology (see above) to turn liquid medication into a mist. AAD nebuliser handsets come with colour-coded medication pots. You should use the correct pot for each medication. If you use the wrong pot, you may get the wrong amount of medication. Your CF team will tell you how much medication to use and which pot to put it in. You should not save any left-over medication to use later.

AAD systems need less medication than some other systems. They can keep track of your breathing pattern and work out the best time to deliver the medication on each breath in.

AAD nebuliser systems have two ways of working:

1. Tidal breathing mode (TBM) – breathing in slow, long breaths without guidance from the system
2. Target inhalation mode (TIM) – guided deep and slow breathing, with vibration reminders of when to breathe in and out

The type of mouthpiece you are given controls which mode you use – the mouthpiece for tidal inhalation mode has a small block on the side. Make sure you are using the one your CF team has asked you to use.

The advantages of AAD nebuliser systems are that:

- the medication is delivered quickly
- they are small and portable, with a built-in rechargeable battery
- they are quiet
- they will only deliver medication when correctly set up and held flat and face down, so you always know that you are using it correctly
- there is very little medication waste.

The I-neb® is the only AAD nebuliser system available in the UK. The I-neb® comes with a patient support programme which will contact you (or you can contact them) about new parts and servicing.

The I-neb® also tracks when you use it, how long each dose of medication took and whether it was finished. The data can be downloaded by you and your CF team with Insight® computer software. This will help to spot any problems, such as the nebuliser system taking too long or if you are having difficulty taking the medication often enough. You can work with your CF team to create a routine that suits you.

The I-neb® data can also be used in the **CFHealthHub programme**. See the 'Vibrating Mesh Technology nebuliser systems' section for more details.

Ultrasonic

Ultrasonic nebuliser systems use a crystal to vibrate the liquid medication at a high frequency to turn it into a mist. They deliver the mist of medication constantly. Portable versions can be smaller, quieter, and quicker than conventional nebuliser systems. Heat can be produced in ultrasonic nebulisers so they can't be used with some medications such as DNase.

It is unusual to be given an ultrasonic nebuliser system to use at home because fewer of the medications used in people with CF have been tested with ultrasonic nebulisers. Other types of nebuliser systems are generally more suitable.

Looking after your nebuliser system

You should check the manufacturer's information that comes with your nebuliser system and talk to your CF team about looking after it. They will tell you how often each part of the nebuliser system should be serviced or replaced.

All nebuliser handsets should be cleaned and dried as soon as possible after every use. Leaving them damp can encourage harmful bacteria and fungi to grow. Leftover medication can also damage or block parts of the nebuliser system. This could mean that it takes longer to work or stops working altogether.

Your nebuliser handset should be sterilised according to the manufacturer's information. This may mean boiling or steam cleaning the nebuliser handset.

Looking after your nebuliser system is important for two main reasons:

1. It reduces the risk of harmful bacteria and fungi being on your nebuliser system when you breathe from it.

2. It keeps the medication running through your nebuliser system as quickly as possible.

Travelling with your nebuliser system

Some nebuliser systems are more portable than others. Talk to your CF about getting the best nebuliser system for your travels.

If you are travelling by plane you should check where your medication and medical equipment can be carried as it may vary between airlines:

- You should be able to carry powder, solid and liquid essential medicines in hand luggage – this includes those over 100mL and inhalers. It is best to carry medication in your hand luggage in case your hold luggage goes missing or gets damaged.
- You should be able to carry medical equipment like nebuliser systems if it is essential to your journey, but check with the airline. Airlines may ask for information about the flight safety of the system – you can get this from the manufacturer.
- You will need to speak to the airline about hold carriage of medications that need to be kept cold or at steady temperature, such as DNase. Speak to your CF team about travelling with cold medicines.

You should also take a letter from your CF team or a prescription script that lists all your equipment and medications to help with security at the airport.

You may need to make sure that your nebuliser system works with the voltage in the country you are visiting – this should be stated somewhere on the nebuliser. Most are dual voltage, so will work anywhere. If you are unsure, check with your CF team. You may need an adapter for the plug.

Who will give me the nebuliser system?

In general, your CF team should give you:

- **your own equipment which is suitable for your needs** – your CF team should give you quickest and simplest nebuliser system possible for each medication. **You should not share your nebuliser system with anyone else.**
- **a test dose of any medication** – also known as a drug response assessment. Your CF team will check if the medication is suitable for you.
- **instructions on how to use your nebuliser system and nebulised medications** – you should have support with this as you need it.
- **filters** and filter pads – filters are needed for some nebulised medications through conventional and Vibrating Mesh Technology nebuliser systems. A filter is a case containing an absorbent filter pad that should be changed after every use. This is to stop other people breathing in the medication and to avoid the medication damaging property such as electronics.

Sometimes a homecare provider will deliver the medication that you take through your nebuliser system.

If your nebuliser system breaks, tell your CF team as soon as you can. They may be able to provide replacement parts or can tell you where to get them. If you use an I-neb®, the company's patient support programme will contact you (or you can contact them) about new nebuliser parts and servicing. Ask your CF team for more details.

The Cystic Fibrosis Trust, NHS England, the National Institute for Clinical Excellence (NICE) and the Scottish Medicines Consortium (SMC) make guidance for CF teams about what they should offer people with CF and their families. There may be reasons why your CF team does something different to this guidance. You can talk to them about this if you have any questions.

Should I buy my own nebuliser system?

Your CF team will give you the nebuliser system you need. If you decide that you want to buy a different type of nebuliser system, it is important that you discuss this with your CF team, because:

- your CF team may be able to give you a nebuliser system free of charge
- the nebuliser system that you want to buy may not be suitable for you or for your medications
- you can sometimes get a letter from your CF team to claim VAT exemption on the cost of the nebuliser system you want to buy.

Inhalers

What are inhalers?

Inhalers are devices that deliver medication to the lungs during inhalation (breathing in), either as a dry powder or as a spray. They are smaller and usually quicker to use than nebuliser systems. They can be more difficult to use correctly and some medications cannot be used in an inhaler.

Inhaled medications are used because:

- the medications can go straight to your lungs without having to go around your body like with tablets or injected medications. This might reduce side-effects
- some medication is only available with an inhaler, for example tiotropium bromide
- inhalers are usually smaller to carry, quicker to use and are easier to clean.

General inhaler advice

Your CF team will show you how to use your inhaler, including how to use a spacer if needed. Using the inhaler in a different way may mean that there could be different amount of medication delivered to your lungs. Your CF team will check your inhaler technique regularly. You can also watch videos on the **Asthma UK** website to remind you about the best technique to use for each inhaler.

You should always use the capsules or medication canister with the inhaler that comes with your medication. Using the capsules or medication canister in a different inhaler may mean a different amount of medication reaches your lungs. Make sure you replace the inhaler as often as the patient information and your CF team ask you to. Contact your CF team if you have any queries.

What types of inhalers are there?

Aerosol inhalers

There are two types of aerosol inhalers: metered dose inhalers or breath-actuated inhalers. Metered dose inhalers release medication as a spray when the canister is pressed into the inhaler. Breath-actuated inhalers such as Easi-Breathe®, Airmax® or Autohaler® automatically release a spray of medication when you begin to breathe in.

Traditional dry powder inhalers

Traditional dry powder inhalers give medication as a dry powder instead of a spray. They are usually used to deliver bronchodilators such as salbutamol or terbutaline, or corticosteroids, such as budesonide or fluticasone (see ‘Which medications can be inhaled?’ below). These include the Turbohaler®, Accuhaler® and HandiHaler®.

Other dry powder inhalers

Colistin (Colobreathe®), tobramycin (TOBI®) and mannitol (Bronchitol®) can be used with newer dry powder inhalers.

Advantages of dry powder inhalers are that they are small, quiet, and may be quicker than nebuliser systems. They also don’t need time-consuming cleaning. They aren’t suitable for everyone and some people will have side-effects, such as cough.

General information

Which medications will I use an inhaler or nebuliser system for?

It is unlikely your CF team will tell you that you need to take all of the medications below. Feel free to only read about the medications you are taking. If you're unsure about whether you should be taking any medication, talk to your CF team.

Bronchodilators

Bronchodilators relax the muscles in your airways when your chest is feeling tight. They can be used to open your airways before chest physiotherapy or exercise.

People with CF usually use an inhaler to take bronchodilators rather than a nebuliser system. Examples of bronchodilators are salbutamol (Ventolin®), terbutaline sulphate (Bricanyl®) and ipratropium bromide (Atrovent®).

Mucolytics and osmotics (mucus thinners)

DNase (Pulmozyme®)

DNase, also known as dornase alfa, breaks down mucus in your lungs, making it thinner and easier to clear.

DNase is taken using a nebuliser system. You and your team should decide when it would be best for you to take DNase, as it doesn't always take the same amount of time to work for everyone.

DNase should be kept in a fridge – ask your CF team what to do if this is not possible, for example while travelling.

Ultrasonic nebuliser systems should never be used to nebulise DNase as they generate heat that may stop it working.

Hypertonic sodium chloride (also called hypertonic saline)

You may know hypertonic sodium chloride as hypertonic saline, Nebusal® or Mucoclear®. It draws more water into the mucus in your lungs. This may help you clear mucus during airway clearance, or sometimes to help you give a sputum sample if you find that difficult.

Hypertonic sodium chloride is usually nebulised after you've taken any bronchodilators, if you have them, and immediately before or during airway clearance. You should use it in a well-ventilated room and use a filter with a conventional or Vibrating Mesh Technology nebuliser system to avoid getting hypertonic sodium chloride on surfaces and electrical equipment in your home. Don't nebulise hypertonic sodium chloride through other devices such as an Aerobika(R) unless your CF team have told you to.

Mannitol (Bronchitol®)

Mannitol, also called Bronchitol®, is used in a dry powder inhaler. It works in a similar way to hypertonic sodium chloride. If you use a bronchodilator, you should use it before inhaling mannitol. Mannitol should be inhaled before doing your airway clearance.

Antibiotics

Antibiotics are used to prevent, treat, and control bacterial lung infections in people with CF. You should take them after any bronchodilators and your airway clearance.

Some antibiotics come with their own nebuliser handsets, which you should always use for that antibiotic. You should also make sure that you do not use them for any other nebulised medication. This is because they deliver a different amount of medication compared to other nebuliser handsets.

If you use a conventional or vibrating mesh nebuliser system, you should use a filter to stop the antibiotic being breathed in by other people or covering surfaces in your home.

- Vantobra® (tobramycin) comes with the Tolero® handset to use with an e-Flow® rapid.
- Cayston® (aztreonam) come with the Altera® handset to use with an e-Flow® rapid.
- Quinsair® (levofloxacin) comes with the Zirela® handset to use with an e-Flow® rapid.

Antifungals

Antifungals are used to treat fungal infections in the lungs. An example of an antifungal is amphotericin B (Ambisome®). Antifungals may sometimes be taken with a nebuliser system.

If you use a conventional or vibrating mesh nebuliser system, you should use a filter to stop the waste antifungal being breathed in by other people or covering surfaces in your home.

Corticosteroids

Corticosteroids reduce inflammation in your airways.

People with CF occasionally take corticosteroids through a nebuliser system, but more often they are recommended as a tablet or an in an inhaler. Ultrasonic nebuliser systems should not be used to nebulise corticosteroids.

You should rinse your mouth out after nebulising corticosteroids. If you use a facemask instead of a mouthpiece you should also wash your face.

Examples of corticosteroid inhalers include:

- Beclometasone dipropionate (Becotide®, Clenil®, QVAR®).

- Budesonide (Pulmicort® and others).

- Fluticasone propionate (Flixotide®).

Sometimes, steroids are mixed in the same inhaler with a bronchodilator. Examples are:

- fluticasone plus salmeterol (Seretide®)
- budesonide plus formoterol (Symbicort®)

For more information on inhaled steroids, see our **Steroid treatment in cystic fibrosis** factsheet.

Which medication will I be given?

This will depend on several things, such as:

- The type of infection that is growing in your lungs
- Any allergies you have
- Where you live (see next page)

Before starting a medication, you should have a test dose (also known as a Drug Response Assessment) where your CF team will check if the medication is right for you. For some medications, such as bronchodilators, this should be rechecked regularly. It is important that you do not miss a dose or stop taking medication without talking to your CF team.

If you have any questions about your medication, talk to your CF team.

In England, Wales and Scotland

The following medications are available for people with CF to use in nebuliser systems and/or inhalers:

- Colistin or colistimethate sodium (also known as Colomycin®, Promixin®, Colobreathe®) –you should be able to take this as a dry powder inhaler if you can't take it using a nebuliser system.
- Tobramycin (also known as TOBI®, TOBI® Podhaler®, Bramitob, Vantobra® or Tymbrineb®). You should be able to take this as a dry powder inhaler if you can take nebulised tobramycin but cannot take colistin or it isn't working well for you.
- Aztreonam lysine (Cayston®).
- Levofloxacin (Quinsair®) – you should be offered alternate months Levofloxacin (either alone or alternating with Colistin or Tobramycin) if previous routines haven't worked well for you
- DNase, also known as Dornase alfa (Pulmozyme®).
- Mannitol (Bronchitol®) – you should be able to take this if you cannot use DNase or hypertonic sodium chloride and your lung function is getting worse.

You might sometimes be offered a different nebulised antibiotic if you live in Scotland. If you have any questions about your medication, talk to your CF team.

These are high-cost medications that have policies on when they can be used. Hypertonic sodium chloride (hypertonic saline) is not on this list as it is not a high cost medication, so your CF team can prescribe it whenever they think it would be helpful.

In Northern Ireland

See the 'In England and Wales' section.

For some new medications or new uses for existing medications that are approved by NICE, there might be a delay because of extra local reviews. There may be some differences between the guidelines for each area in Northern Ireland compared to England and Wales. If you have any questions about your medication, you can talk to your CF team.

Young children and babies

Your child's CF team will help to choose a nebuliser system that your child can use.

- Conventional and ultrasonic nebuliser systems are suitable for children of any age.
- Vibrating mesh technology and adaptive aerosol delivery systems are suitable for children over the age of two, but some children will be older before they can manage to use these systems and in some cases your CF team may feel that they are suitable for younger children.

A mouthpiece should be used with nebuliser systems and inhalers when possible. If using an e-Flow® rapid or other PARI brand mouthpiece with a blue flap valve, turning this to face down to the floor will help to stop the mist getting into the eyes.

Babies and younger children may need a facemask. When using a facemask, your child's CF team will help you work towards using a sealed mask which is held firmly on the face to make sure as much medication gets into the lungs as possible. You should wash your child's face after treatment is finished so that the medication doesn't stay on their skin.

It may take some time and patience before your child gets used to the nebuliser system. Your child's CF team are there to help if you're struggling with this.

Some general tips include:

- Any medication that is taken is an achievement for you and your child
- Praise, cuddles and small rewards can help
- Let your child play with the nebuliser system by pretending to give their teddies or dolls
- Explain what the nebuliser system is and what is does

Ideally your child should be awake and calm when using the nebuliser system but if this isn't possible, doing treatment when they are asleep is usually better than when they are distressed. Your CF team can help you make a plan around your child's needs and the medications they take.

Your child should always be supervised while using a nebuliser system as they have small parts and cables. You might also need to check that they are using it correctly.

Where can I find out more?

If you have any questions about your inhaled medication or nebuliser system, you can talk to your CF team, usually your physiotherapist.

You could also look at:

- **How to use your inhaler** by Asthma & Lung UK
- Videos or information for your own nebuliser system online
- **Nebulisers for giving medication in cystic fibrosis** from Cochrane
- **Can I take my medicine abroad?** from the NHS

Further information

Find more information resources about living with cystic fibrosis at **cysticfibrosis.org.uk/information**.

Our Helpline is open 10am – 4pm Monday to Friday. It's available to anyone looking for information or support with any part of cystic fibrosis, a listening ear, or just to talk things through.

How to reach us:
- Call **0300 373 1000** or **020 3795 2184**
- Email **helpline@cysticfibrosis.org.uk**
- Chat with us on **Facebook**, **Twitter** or **Instagram**
- Message us on WhatsApp on **07361 582053**

Visit **cysticfibrosis.org.uk/helpline** for more information.

We welcome your feedback on our resources.

You can also ask for this resource in large print or as a text file. Email **infoteam@cysticfibrosis.org.uk**.

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The information in this resource does not replace any advice from your doctor or CF team. It is important that you seek your team's advice whenever you want to change your treatment.

Cystic Fibrosis Trust

Cystic Fibrosis Trust is the charity uniting people to stop cystic fibrosis. Our community will improve care, speak out, support each other and fund vital research as we race towards effective treatments for all.

We won’t stop until everyone can live without the limits of cystic fibrosis.

cysticfibrosis.org.uk

Uniting for a life unlimited