Cystic Fibrosis our focus

Inhaled therapy for people with cystic fibrosis

Factsheet – May 2016

Fighting for a Life Unlimited

Inhaled therapy for people with cystic fibrosis

Introduction

This factsheet contains information about inhaled treatments for people with cystic fibrosis (CF). The information covers different types of nebulisers and inhalers, types of medication that can be inhaled, how to look after your nebuliser and what to expect from your CF centre/network clinic.

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What are nebulisers?

Nebuliser systems change liquid medication into a mist so they can be breathed into the lungs. There are different types of nebuliser system, which this factsheet will describe.

Nebulised medications are used because:

- the medications go straight to where they are needed (in the lungs) without having to go around the body, reducing side-effects;
- some medication is only available as a nebulised medication, eg DNase;
- large doses of medication can be given compared with some types of inhaler; and,
- some inhalers can be difficult to use correctly – a nebuliser can ensure that more medication gets into the lungs.

What types of nebuliser systems are available?

There is a range of conventional and travel nebuliser systems available, some are described below. Your CF team will select the system that is most appropriate for you.

Conventional nebuliser systems

Conventional (jet) nebuliser systems are made up of a nebuliser (the pot that holds liquid medication) and an air compressor that pumps air through the liquid medication and changes it into a mist. The nebulised medication can be breathed in using a mouthpiece or facemask. Conventional nebuliser systems deliver the mist of medication constantly and need to be switched off when the nebuliser has finished, which is indicated by the sound of the nebuliser changing and 'spluttering'.

The way the medication is delivered can depend on the nebuliser system used. Some may be faster or get more medication to the lungs, but they may also cause side effects such as chest tightness. Different nebuliser systems may be needed for different medications, so you may be given more than one. **It is important that you use the nebuliser system/ systems given to you by your cystic fibrosis team as different nebuliser systems may affect the amount of medication that reaches the lungs.** If you are not sure if you have the best system to suit you and your medication, talk to your CF team.

Examples of conventional nebuliser systems:

- Philips Respironics Innospire delux® air compressor with SideStream® nebuliser.
- Vios® PRO Aerosol Delivery System with PARI LC Plus®/star® nebuliser.

Portable air compressors that may make travel/holidays easier are available. They are lighter than conventional nebuliser systems and may require a battery or a 12-volt car socket. Portable air compressors can be used with a normal nebuliser, but may take longer to use and may not be suitable with all medications.

Examples of portable air compressors:

- Innospire mini®.
- Trek S portable aerosol system®.

Ultrasonic nebuliser systems

Ultrasonic nebuliser systems use a crystal to vibrate the liquid medication at a high-frequency to change it into a mist. They deliver the mist of medication constantly. Portable versions can be smaller, quieter and quicker than conventional nebulisers. Heat produced by the vibrations may mean that they can't be used with some medications (eg DNase).

It is unusual to be given an ultrasonic nebuliser system to use at home. This is because fewer of the medications used in people with CF have been tested with these systems, they aren't suitable for all medications and other types of nebuliser systems are generally more suitable.

Vibrating Mesh Technology nebuliser systems

Vibrating Mesh Technology (VMT) nebuliser systems are made up of an electronic unit and a nebuliser handset joined by a connection cord. The electronic unit powers and controls the system. The nebuliser handset is a plastic pot that holds the liquid medication and contains a metal mesh.

VMT systems vibrate the mesh, which moves the liquid medication through the tiny holes in it. This changes the liquid into a mist with each drop a similar size. VMT systems deliver the mist of medication constantly and stop automatically when the nebuliser is finished. The advantages of VMT systems are:

- nebulised medication is given faster;
- the small similar-sized droplets get into airways better;
- they are silent; and,
- they are smaller than conventional nebuliser systems and can run on batteries.

In the UK, the VMT system most often used is the e-Flow rapid® with or without the eBase® controller. The eBase® controller displays information about what is happening while you nebulise.

Adaptive aerosol delivery (AAD) nebuliser systems

The only adaptive aerosol delivery (AAD) system currently available is the I-Neb AAD nebuliser system®. AAD systems use vibrating mesh technology (described above) to change liquid medication into a mist. They also monitor your breathing pattern and deliver the mist of medication at the best time to ensure the medication will get to where it needs to be. AAD systems are small, quiet and have a rechargeable battery inside them.

The AAD system will only deliver medication when correctly set up and held flat and face down, so you always know that you are using it correctly. It has two ways of working (tidal breathing mode and target inhalation mode) which are controlled by the type of mouthpiece you are given. Tidal inhalation mode has a small block on the side of the mouthpiece. **You should always make sure that you are using the type of mouthpiece advised by your CF team.**

AAD systems give nebulised medication with little waste, so you will need to use a different amount of medication than with conventional, VMT or ultrasonic systems. Even though you put less medication into an AAD system, you will still get the same, or more, medication to your lungs than using a conventional system. You should not save any left-over medication to use later. **Your CF team will tell you how much medication to use in AAD systems.** The AAD system comes with different chambers, which are colour coded. You should use the correct chamber for each nebulised medication. If you use the wrong chamber you may get the wrong amount of medication. **Your CF team will tell you which chamber to use for each medication.**

The AAD system stores data about how you use it such as date and time, how long each dose of medication took and whether it was finished. The data can be downloaded by you and/or by your CF team using computer software (Insight®). This will help to spot any problems, such as long treatment times or difficulty taking the treatments often enough, and help you to work with your CF team to create a routine that suits you. The I-Neb® comes with a patient support programme run by Philips (the company that makes the I-Neb®). They will contact you or you can contact them about the upkeep of your AAD system and they will arrange new parts and servicing of the system.

Looking after your nebuliser system

You should check the information that comes with your nebuliser system and talk to your CF team about looking after the equipment. They will advise you about when nebulisers, chambers or handsets should be serviced or replaced. All nebulisers, chambers or handsets should be cleaned and dried as soon as possible after every use; leaving them damp can encourage bacteria and fungi to grow. Leftover medication can also damage or block the nebuliser or mesh, which can mean that it takes longer to work and/or the system may stop working altogether. Your nebuliser should be sterilised according to the manufacturer's guidance.

Travelling with your nebuliser system

When travelling by air you should take a letter from your CF team that lists all of your equipment and medications; this can help with security at the airport. The Cystic Fibrosis Trust helpline can also provide a travelling medications letter and can translate the letter into different languages if needed. Helpline details are on the last page of this factsheet. It is important to check with the airline about whether you can carry your nebuliser in hand luggage or use it on the flight (should you need to). Airlines may ask for information about the flight safety of the system; you can get this from the manufacturer.

You should check the following information as it may vary between airlines.

Most airlines allow you to carry powder/solid medicines with you in hand luggage. This is best in case your checked/hold luggage goes missing or gets damaged. Most airlines only allow you to carry enough liquid, aerosol or gel medication plus any cold pack needed (eg for DNase) in your hand luggage to last for the flight and to allow for delays. The rest should be packed in hold baggage. You will need to speak to the airline about hold carriage of medications that need to be kept cold/at steady temperature (eg DNase).

If you're travelling abroad, you may need an adapter plug as the type of plug socket may vary. You will also need to make sure that your nebuliser system works with the voltage available in the country you're visiting (for example the UK power supply is 230 volts, whereas in the US is it 120 volts). Some nebuliser systems (I-Neb, eFlow rapid, most travel compressors) are multi/universal voltage, which means that they can be used anywhere, but you may still need an adapter for the plug to fit. If you are unsure whether your nebuliser system will work in the country that you are visiting, check with your CF team before you book your trip.

General nebulised medication advice

- It is usually best to use a mouthpiece with your nebuliser, do not change to a mask without discussing it with your CF team.
- Nebulised medication should be at room temperature before being nebulised. Cold medication can cause chest tightness and may not run correctly through some systems.
- Most nebuliser systems leave some medication in the chamber even when the dose is finished. This is called residual volume and is normal. You should not shake or tip your nebuliser as the residual volume may be nebulised. This may change the amount of medication that gets to your lungs. You should empty the residual volume out of the nebuliser system before cleaning; do not try to reuse 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 should not mix nebulised medications unless your CF team asks you to. Mixing medications may mean that they don't work in the same way as when taken separately.

Should I buy my own nebuliser system?

Your CF team must provide you with a nebuliser system to take any medications that they ask you to use. If you decide that you want to buy a different type of nebuliser to the one that you are given, it is important that you discuss this with your CF team, because:

- your CF team may be able to provide a different nebuliser system free of charge;
- the nebuliser system that you want to buy may not be suitable for you or for your medications; and,
- you may need a letter from your CF team to claim VAT exemption on the cost of the nebuliser system you want to buy.

What are inhalers?

Inhalers are devices that are used to deliver medication to the lungs either as a dry powder or as a spray. They are smaller and usually quicker to use than nebulisers but can be more difficult to use correctly and may not be available for certain medications. Some types of inhaler that deliver smaller amounts of medication, usually bronchodilators or corticosteroids, have been around for many years (for example metered dose inhalers, turbohalers or accuhalers). These are called aerosol inhalers and traditional dry powder inhalers. Newer types of dry powder inhalers have also been developed, which deliver much greater amounts of medication to the lung and so are suitable for medications such as antibiotics and osmotics (explained below). They are called dry powder inhalers.

Inhaled medications are used because:

- the medications go straight to where they are needed (the lungs) without having to go round the body, which can reduce side-effects;
- some medication is only available with an inhaler, eg Tiotropium Bromide; and,
- inhalers are usually smaller to carry, quicker to use and don't need time-consuming cleaning routines.

What types of inhalers are available?

Aerosol inhalers

Aerosol inhalers are usually one of two types; metered dose inhalers or breath actuated inhalers. Metered dose inhalers give the medication in a spray form as the canister is pressed into the inhaler. Breath actuated inhalers, eg 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. There are a number of different types of traditional dry powder inhalers that are usually used to deliver bronchodilators (explained below), such as Salbutamol or Terbutaline, or corticosteroids, such as Budesonide or Fluticosone. The most common types used include the turbohaler, accuhaler and handihaler.

Dry powder inhalers

Some medications are now available as capsules to use with an inhaler; these are Colistin, Tobramycin and Mannitol. Colistin is inhaled as one capsule twice a day through a turbospin inhaler, Tobramycin as four capsules twice a day through a podhaler inhaler for 28 days before a 28-day break, and Mannitol as ten capsules twice a day. **It is important to always use the capsules in the inhaler that comes with the medication and to replace the inhaler as often as the patient information and your CF team ask you to.** Advantages of dry powder inhalers are that they are small, quiet, may be quicker than nebulisers and don't need time consuming cleaning. They aren't suitable for everyone and some people will have side-effects, such as cough.

General inhaler advice

- You should use the inhaler as you have been shown, such as using a spacer if instructed to do so. Using the inhaler in a different way may mean that you get a different amount of medication to your lungs.
- You should always use the capsules or medication canister in the inhaler that comes with the medication, as using the capsules or medication canister in a different inhaler may mean a different amount of medication reaches your lungs.
- You should have your technique checked regularly by your CF team to make sure that you are getting the best effect from your inhalers. There are also some good videos to remind you about the best technique to use for each inhaler, which can be found on the Asthma UK website.

Which medications can be inhaled?

You should use your inhaled medications as instructed by your CF team. You should also read the patient information leaflet that comes with your medication, and contact your CF team if you have any queries.

Bronchodilators

Bronchodilators relax the muscles around your airways and are used to ease chest tightness. They can prepare your chest for chest physiotherapy or exercise by opening your airways and, if your CF team asks you to, they should be taken before chest physiotherapy and exercise. Many people with CF can use an inhaler rather than a nebuliser for their bronchodilator. Examples of bronchodilators are Salbutamol (Ventolin®), Terbutaline Sulphate (Bricanyl®) and Ipratropium Bromide (Atrovent®).

Antibiotics

Antibiotics are used to treat, prevent and control infection. They should be taken after bronchodilators and chest physiotherapy. When nebulising antibiotics using conventional or VMT systems, a filter should be used to stop the antibiotic being breathed in by other people or covering surfaces in your home. The filter is a case containing a pad that should be changed after every treatment. Your CF team will explain how to get replacement nebuliser parts such as filters.

If you are nebulising Cayston® (nebulised Aztreonam), you should make sure that you always use an e-Flow® with the Altera® handset which comes with your Cayston®. You should also make sure that you do not use the Altera® handset for any other nebulised medication. This is because the Altera® delivers a different amount of medication compared to a normal handset.

Mucolytics

DNase (Pulmozyme®)

DNase is used to make sputum thinner and easier to clear. It is normally prescribed once a day. You and your team should decide when it would be best for you to take the DNase, as how long it takes to work is different for different people. It is important to keep your DNase in a fridge and discuss any situations where this may not be possible (for example traveling) with your CF team. A separate nebuliser, handset or chamber should be used for DNase as it should not be mixed with other medications. Ultrasonic nebuliser systems should never be used to nebulise DNase as they may stop it working and/or not deliver DNase to the lung correctly.

Osmotics

Hypertonic sodium chloride 3, 6 and 7%

Hypertonic sodium chloride comes in different strengths; 3, 6 or 7%. You may also know it as Nebusal® or Mucoclear®. It draws more water into the airways and more water into the sputum in the lungs. It is used either to get a sputum sample (in people who do not cough up sputum easily) or to help to clear sputum better as part of chest physiotherapy. It should be nebulised after any bronchodilators you take and immediately before or during chest physiotherapy. Hypertonic sodium chloride may cause damage to some electrical equipment if you nebulise it while sitting close to the equipment (for example using a computer while nebulising). You should use it in a well-ventilated room away from electrical equipment or use filters with conventional or VMT nebuliser systems to avoid covering surfaces in your home with waste hypertonic sodium chloride.

Mannitol

Mannitol is a dry powder inhaler rather than a nebuliser (see page 8 for more information about dry powder inhalers). It acts in the same way as hypertonic saline and should be inhaled before chest physiotherapy. You should always take your bronchodilator before inhaling Mannitol.

Corticosteroids

It is not common for people with cystic fibrosis to need nebulised corticosteroids, however they are used by some. Examples of corticosteroids are beclometasone dipropionate (eg QVAR®), budesonide (eg Pulmicort®) and fluticasone propionate (Flixotide®). They are used to reduce inflammation in the airways and are usually taken twice a day. It is important that you do not miss a dose or stop taking the medication without discussing it with your CF team. You should rinse your mouth out after nebulising corticosteroids and you should also wash your face, if using a facemask. Ultrasonic nebulisers should not be used to nebulise corticosteroids.

Antifungals

Antifungals reduce the amount of fungal growth in the lungs. It is not common to nebulise antifungals but your CF team may ask you to do this in certain circumstances. An example of an antifungal is amphotericin B (Ambisome®). When nebulising antifungals using conventional or VMT nebuliser systems, a filter should be used to stop the waste antifungal being breathed in by other people and/or covering surfaces in your home.

Children and babies

Your CF team will choose a system suitable for your child's age and ability. Conventional and ultrasonic nebuliser systems are suitable for children of any age. VMT and AAD 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.

Inhaled medication should be delivered through a mouthpiece when possible, although babies and younger children may need a mask. When using a mask, it is important that 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. Your CF team should advise you about this and give you suitable equipment.

If using a facemask, you should wash your child's face after the nebuliser so that the medication doesn't stay on their skin. If using an e-Flow® or other PARI nebuliser mouthpiece with a blue flap valve, turning this to face down will help to stop the mist getting into the eyes.

It is important that your child is encouraged to get used to the nebuliser, and this may take some time and patience. Any medication that is taken should be seen as an achievement. It is important that nebulisers are accepted in both the short and long term, so praise, cuddles and small rewards can help. Letting your child play with the nebuliser system by pretending to give their teddies or dolls nebulisers and explaining what the nebuliser is and does can also help. Ideally your child should be awake and calm when using the nebuliser but, if this isn't possible, giving the nebuliser when asleep is usually better than giving it when crying. Your CF team should give you a plan for the best time and way to give nebulisers depending on your child's needs and the medications they take.

Children should always be supervised for safety while using nebuliser systems due to small parts and cables. Most children will also need supervision using nebuliser systems to check that they are using it correctly.

What to expect from my CF centre/network clinic

NHS England, the Cystic Fibrosis Trust, the National Institute for Clinical Excellence (NICE) and the Scottish Medicines Consortium (SMC) have produced guidelines for CF units about the service they should offer. **There may be reasons why your CF unit offers something different to the guidelines and you should discuss any differences with them.** The following is a summary of what these guidelines say that you should expect:

- You should have your own equipment; you should not be expected or choose to share your nebuliser system.
- You should be provided with suitable equipment by your CF centre/clinic.
- Before starting a nebulised medication, you should have a test dose where the CF team checks if the medication is suitable for you. For some medications, such as bronchodilators, this should be rechecked regularly.
- You should be taught how to use your nebuliser system and nebulised medications and you should have support over time as you need it.
- You should be asked to use expiratory filters with some nebulised medications through conventional and VMT nebuliser systems so that other people don't breathe in the medication and to avoid the medication damaging property.
- A mouthpiece should be used when possible.
- Your CF team should offer the quickest and simplest nebuliser system possible for each medication and suitable for you.

England and Wales

- You should be able to have Colistimethate sodium (brands Colomycin, Promixin, Colobreathe), Tobramycin (brands Tobi, Tobi Podhaler, Bramitob), Aztreonam lysine (brand Cayston), Dornase alpha (brand Pulmozyme), Mannitol (brand Bronchitol) as needed.
- You should be able to have Mannitol if you cannot use DNase or other osmotic medications and your lung function is quickly getting worse.
- You should be able to have Colistin by dry powder inhaler if your health would benefit from using Colistin but you can't take it using a nebuliser.
- You should be able to have Tobramycin by dry powder inhaler if you can take nebulised Tobramycin but cannot take Colistin or it isn't working well for you.
- You should be able to alternate inhaled antibiotics (for example one month of Colistin followed by one month of Tobramycin or one month of Tobramycin followed by one month of Aztreonam lysine) if needed.

Scotland

See the England and Wales guidance, however there may be variation in which nebulised antibiotic you are offered. If you have questions about what you are being offered, you should discuss it with your CF team.

Northern Ireland

See the England and Wales guidance, however, in Northern Ireland, NICE guidance may have a local review before being used. There may be some differences between the guidelines for each area in Northern Ireland compared to England and Wales. If you have questions about what you are being offered, you should discuss it with your CF team.

Where can I find out more?

- By asking your CF team, usually your physiotherapist
- The CF Trust Standards of Care

  The CF Trust Standards of Physio Care
- The electronic medicines compendium
- **The Cochrane library**
  Nebuliser systems for drug delivery in cystic fibrosis.
- NHS England Clinical Commissioning Policy: Inhaled Therapy for Adults and Children with Cystic Fibrosis
- NICE
- Inhaler technique
- Youtube
- Travelling

Further information

The Cystic Fibrosis Trust provides information about cystic fibrosis through our factsheets, leaflets and other publications.

Most of our publications are available through our helpline and can be downloaded from our website or ordered using our online publications order form. Visit cysticfibrosis.org.uk/publications.

The Cystic Fibrosis Trust helpline can help you with a range of issues, no matter how big or small. Our trained staff can provide a listening ear, practical advice, welfare/benefits information or direct you to other sources of support. The helpline can be contacted on 0300 373 1000 or helpline@cysticfibrosis.org.uk and is open Monday to Friday, 9am – 5pm.

Calls to 0300 numbers cost no more than 5p per minute from a standard BT residential landline. Charges from other landlines and mobile networks may vary, but will be no more than a standard geographic call and are included in all inclusive minutes and discount schemes. If you are worried about the cost of the call please let us know and we'll call you back.

You can also find more information at our website cysticfibrosis.org.uk.

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More factsheets available at:
cysticfibrosis.org.uk/publications