

Cystic **Fibrosis** *our focus*

Nebuliser therapy in cystic fibrosis

Factsheet – July 2013

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Introduction

This factsheet contains information about nebuliser therapy in cystic fibrosis (CF), including the different types of nebulisers available, the types of medication that can be nebulised and nebuliser maintenance.

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What are nebulisers?

The word 'nebuliser' is used to describe a pot that holds liquid medication and changes it into a mist to be breathed in. The machine that is attached to the pot and blows air through it is called an air compressor. 'Nebuliser' can also describe other machines that hold liquid medication and change it into a mist to be breathed in (for example vibrating mesh technology system, adaptive aerosol delivery system, ultrasonic system). Finally, the word 'nebuliser' can describe a medication that is breathed in as a mist. Throughout this leaflet we use the word 'nebuliser' to mean a pot that holds liquid medication and changes it into a mist, 'nebuliser system' to mean a nebuliser chamber and air compressor or another type of machine that changes a liquid medication into a mist to be breathed in and 'nebulised medication' to mean a medication that is breathed in as a mist.

Nebulised medications are used because:

- The medications go straight to where they need to work (in the lung) without having to go round the body. This can reduce side-effects.
- Some medication is only available as a nebulised medication, for example DNase.
- Large doses of medication can be given compared with some types of inhaler.
- It can be difficult to use some inhalers correctly. Using a nebuliser can mean that more of the medication gets into the lung.

What types of nebuliser systems are available?

Conventional nebuliser systems

Conventional nebuliser systems are made up of a nebuliser, which holds liquid medication, and an air compressor, which pumps air into a tube attached to the compressor and to the nebuliser. The air runs through the nebuliser and changes the liquid medication into a mist. The nebulised medication can be breathed in using a mouthpiece or facemask.

Different air compressors and nebulisers change how the medication is delivered. The time that nebulised medication takes to finish is less with compressors that pump out air faster and with nebulisers that change the medication into a finer mist. Faster compressors can get more medication to the lung but may give more side effects such as chest tightness. Some types of nebulisers can also get more medication into the lung. Conventional systems deliver the mist of medication constantly and will need to be switched off when the nebuliser has finished; this is when the sound of the nebuliser changes and starts 'spluttering'. It is important that you use the compressor and nebuliser given to you by your cystic fibrosis team and, if you are given different nebuliser systems to use with different medications, that you do this. If you are not sure whether you have the best system to suit you and the nebulised medication you are taking, talk to your CF team about the options.

Examples of conventional nebuliser systems:

- Philips Respironics Porta-neb® air compressor with SideStream® nebuliser
- PARI BOY S® / PARI BOY SX® air compressor with PARI BOY LC Plus®/star® nebuliser

There are portable air compressors available, which may make travel/holidays easier. They are lighter than normal conventional nebuliser systems and may work on a battery or from a 12 volt car socket. Portable air compressors are used with your normal nebuliser but it may take longer than normal for your medication to finish nebulising.

Examples of portable air compressors:

- Freeway Lite®
- PARI BOY Mobile S®

Ultrasonic nebuliser systems

Ultrasonic nebuliser systems use a crystal to vibrate the liquid medication at a high-frequency to change the liquid medication into a mist. They deliver the mist of medication constantly. Portable versions of ultrasonic nebuliser systems can be smaller, quieter and quicker than conventional nebulisers. Heat generated by the vibrations may mean that they can't be used to nebulise some medications. 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 cystic fibrosis have been tested with these systems than with other types of nebuliser system and because they aren't suitable for all medications.

Vibrating Mesh Technology (VMT) nebuliser systems

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 tiny holes in the mesh. 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 that nebulised medication is given faster and the small similar-sized droplets get into airways better. They are silent and smaller than conventional nebuliser systems and can be run on batteries. They can however break down or get damaged more than conventional systems. In the UK the VMT system most often used is the e-Flow rapid® with or without the eBase® controller. The eBase® controller gives information using pictures about what is happening while you nebulise.

The easycare cleaning aid is available for the e-Flow rapid®. This is a system to clean the mesh and can help to keep the time the nebuliser takes to finish as short as possible. It does this by running liquid through the mesh the opposite way to normal (backwashing). This helps to unblock the tiny holes in the mesh. It can be used as needed but no more than twice a week after normal cleaning but before sterilising. You may not automatically get an easycare cleaning aid with your eFlow rapid®. If you think that your nebulised medication is slow to finish and you are following other cleaning/sterilising instructions then you should discuss the easycare with your CF team.

Adaptive aerosol delivery (AAD) nebuliser systems

The only AAD system 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 only during the part of your breath in when 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 [TBM] and target inhalation mode [TIM]) which are controlled by the type of mouthpiece you are given. TIM 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.** Your AAD system stores data about how you use it such as date and time you used it, 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 and help you to work with your CF team to get a nebulised 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 you have.

They will advise you about when nebulisers, chambers or handsets should be 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. Left-over medication can also damage or block the nebuliser or mesh, which can mean that it takes longer to nebulise medication and/or the nebuliser system may stop working at all.

Travelling with your nebuliser system

When travelling by air, you should take a letter from your CF team which lists all of your equipment and medications; this can help with security at the airport. It is important to check with the airline about whether you can carry your nebuliser in hand luggage and whether you can use it on board a 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 with the airline you are traveling with as it may vary. Most airlines allow you to carry powder/solid medicines with you in hand luggage. This is best in case your checked luggage goes missing or gets damaged. Most airlines only allow you to carry enough liquid, aerosol or gel medication plus any cold pack needed (e.g. 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 (e.g. DNase).

If you are 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 at the country you're visiting (for example the UK power supply is 230volts whereas the US is 120volts). 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 to allow 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 well before you plan to go.

Which medications can be nebulised?

You should use your nebulised medications in the way that your CF team has asked you to. You should also read the patient information leaflet that is in the box with your medication and contact your CF team for advice if you have any queries.

Bronchodilators

Bronchodilators relax the muscle 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 use them for this, they should be taken before chest physiotherapy and exercise. Many people with CF can use an inhaler rather than a nebuliser to give 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 and covering surfaces in your home. The filter is a case into which fits a pad that should be changed after every treatment. You can order filter cases/pads from your pharmacist when you are prescribed some antibiotics. Your CF team should make you aware of 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.

Some antibiotics are available as a dry powder inhaler as well as a nebulised medication (see the section about dry powder inhalers on page 10).

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%,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 sputum in the lung. 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 that 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 playing games on your 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 10 for more information about dry powder inhalers). Mannitol draws more water into the airways and sputum in the lung and is used to help to clear sputum better during chest physiotherapy so it should be inhaled immediately 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. Examples of corticosteroids are beclometasone dipropionate (for example QVAR®), budesonide (for example 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 antibiotic being breathed in by other people and/or covering surfaces in your home.

General nebulised medication advice

- 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.

Children and babies

Your CF team will choose a system suitable for your child's age and what they can do. 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.

A mouthpiece may be used by children of any age and many find it less upsetting than a facemask. If a child is crying, most of the medication won't be breathed in and it won't work as well. In those who can't use a mouthpiece, a facemask can be used to give the medication. 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 with a blue flap valve, turning this to face down will reduce 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 and facemask by pretending to give their teddies or dolls nebulisers and explaining what the nebuliser is and does can also help. 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.

Should I buy my own nebuliser system?

Your CF team should 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.
- You may need a letter from your CF team to claim VAT exemption on the cost of the nebuliser system you want to buy.

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 for 28 days before a 28 day break through a podhaler inhaler, 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 to use, may be quicker than nebulisers and don't need time consuming cleaning. They aren't suitable for everyone and some people will have more side-effects such as cough.

What should I expect from my CF centre?

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 to and you shouldn't choose to share your nebuliser system.
- 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 use the quickest and simplest nebuliser system possible for each medication.

England and Wales

- 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 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.

Scotland

- Mannitol is not recommended for use at the moment.
- Tobramycin by dry powder inhaler can be used but different areas/units may have different guidelines for when to use it.
- There is no guidance about whether you can have Colistin by dry powder inhaler.

Northern Ireland

See the England and Wales guidance, however in Northern Ireland NICE guidance may have a local review before being used. There may so be 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 information about nebulisers?

- By asking your CF team, usually your physiotherapist
- The CF Trust Standards of Care
www.cftrust.org.uk/media/82070/CD_Standards_of_Care_Dec_11.pdf
www.cftrust.org.uk/media/82076/CD_Standards_of_Care_Physio_Jun_11.pdf
- The electronic medicines compendium www.medicines.org.uk/emc
- The Cochrane library www.thecochranelibrary.com
- NICE www.nice.org.uk
- Youtube www.youtube.com
- Travelling www.heathrowairport.com/heathrow-airport-guide/heathrow-security/faqs#carriageofmedicines

Further information

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

Most of our publications can be downloaded from our website ordered using our online publications order form.

Visit cysticfibrosis.org.uk/publications.

Alternatively, to order hard copies of our publications you can telephone the CF Trust on 020 8464 7211.

If you would like further information about cystic fibrosis please contact:

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The information included in this publication is not intended to replace any advice you may receive from your doctor or CF multidisciplinary team and it is important that you seek medical advice whenever considering a change of treatment.

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