

Medicines for children recently diagnosed with cystic fibrosis (CF)

Information for parents



leeds children's
hospital

caring about children

Routine Cystic Fibrosis (CF) medication, alongside good nutrition, physiotherapy and exercise, makes an enormous difference to the health and life expectancy of your child.

We understand it can feel odd to give your child, who appears well, these routine medications, but given as part of the daily routine they will make a massive difference to keeping your child well and able to enjoy life.

CF, although very responsive to preventative treatment, remains a serious disease and life-shortening if it is under-treated. The children and adults with the best health are those who have a good routine with all their regular treatments, diet, physiotherapy and exercise, and we all want the best possible health and happiness for your child.

Introduction

This information is to help you understand the medication that your child has been prescribed. It also provides additional information including how to give (administer) the medicines and how to get further supplies.

Topics covered are:

- List of long-term medications, with some information about them
- General advice about medicines
- How to store the medicines
- Obtaining medicines from your GP and community pharmacy.

You will also be given

- a leaflet about how to give liquid medicine
- a leaflet about Unlicensed Medicines
- a leaflet containing information about CF to pass on to your community pharmacist.

Medication

Children with CF are started on long-term medication shortly after diagnosis. Additional antibiotics may be prescribed for your child if he/she becomes unwell and needs a 'short' course of antibiotic, perhaps for one or two weeks, to overcome that short illness.

The common medicines that children are started on at diagnosis are listed on the following pages.

Vitamins

Vitamins A, D, E and K are known as fat soluble vitamins. In CF, low levels of these vitamins can often be found, due to the difficulty many people with CF have absorbing fat, and therefore fat soluble vitamins. There are a number of other factors that can affect vitamin levels, so it is not uncommon for pancreatic sufficient patients to also have reduced levels.

Your child's vitamin levels will be measured once a year, by measuring the amount of vitamins in the blood. The dose of the vitamin supplements may be altered depending on the results.

Why are these vitamins important?

Vitamin A: Helps keep the skin, teeth, skeletal and soft tissues, mucus membranes and cells in the eyes healthy. Low vitamin A levels can cause problems with eye sight.

Vitamin D: Can be produced by skin being exposed to the sun (only between April and October in the UK) and is also obtained from some foods. Reduced vitamin D can lead to poor bone development, an increased risk of developing osteoporosis (thinning of the bones), as well as increasing the risk of developing infections.

Colecalciferol is the full medical name for vitamin D.

Preparations containing vitamins A & D include Abidec® and Dalivit® liquids. When your child is able to take tablets and capsules, they can be prescribed Vitamin A & D capsules.

Vitamin E: Acts as an antioxidant, so it protects cells in the body from damage. It may play a role in protecting the lungs and therefore can help reduce the progression of lung damage.

Alpha-tocopheryl acetate/alpha tocopherol are the drug names for vitamin E that may appear on the medicine bottle.

Your child may be given vitamin E liquid (or Paravit-CF® see below). The manufacturers advise that once the bottle has been opened, it should be discarded after one month. It is therefore important that you request a new bottle each month.

When your child is older, they will be able to take vitamin E capsules.

Vitamin K: Is not given to babies, although it is in Paravit-CF®. It is usually started when the child is around five years of age and able to swallow capsules/tablets.

Paravit-CF® contains all the fat soluble vitamins (A, D, E & K) and may be prescribed as an alternative to Abidec and Vitamin E liquids. The dietitian will discuss the options available to you.

How to give vitamins

To keep vitamin levels within normal ranges it is important to give the vitamins each day. They should be given at a mealtime when pancreatic enzymes (if required) will also be taken, to improve the amount of vitamins absorbed.

Pancreatic enzymes e.g. Creon Micro®

Creon Micro® is provided with a tiny scoop to measure the correct dose for your child. Take care of the scoop - they are easily lost!

What are pancreatic enzymes?

The majority of babies and children with CF will need to take pancreatic enzymes to help control their symptoms of fat malabsorption (pale, frequent, fatty stools (poo)).

Enzymes are needed to digest food, breaking it down into proteins, carbohydrates, fats, vitamins, minerals, fibre and water. In CF, the enzyme lipase is the most important; lipase is mainly needed to digest fat. The higher the fat content of the meal, snack or drink, the more enzymes need to be taken.

If the incorrect amount of enzymes are taken, it will affect the stools and may also affect the growth of the child. Your dietitian will give you individual advice on the dose, timing of when the enzymes are given, and how to adjust the amount, depending on the food and the appearance of the stools.

Flucloxacillin

Flucloxacillin is a penicillin antibiotic that is currently prescribed to all children with CF. However, at present the CF Start Clinical Trial is ongoing to determine if regular daily flucloxacillin is still the best treatment from diagnosis; you may be invited to participate in this trial if it is appropriate and relevant to your child.

If your child is allergic to penicillin you should let your doctor and nurse know. In our CF Unit, it is our practice to give flucloxacillin every day, life-long. Antibiotics that are given long-term to prevent infection are called 'prophylactic' antibiotics. It has been shown that taking this antibiotic can help to prevent lung infection, caused by the organism ('bug') *Staphylococcus aureus*.

Flucloxacillin has a bitter taste and some parents find that their child prefers one particular manufacturer of flucloxacillin over another. Flucloxacillin liquid is available either as a sugar-free liquid or one that contains some sugar, which helps to mask the bitter taste.

Flucloxacillin liquid is available in two strengths - 125mg/5mL and 250mg/5mL. If your child is having difficulty swallowing the lower strength, it may be helpful to give the stronger strength, which will reduce the volume to be given. Discuss with your nurse, doctor or pharmacist if necessary.

Flucloxacillin liquid is manufactured as a 'dry powder' which needs making up with water to prepare a liquid. The pharmacy will usually make this liquid for you, but once it had been reconstituted (made into a liquid), it has an **expiry of seven days**. This may mean that you will need to collect a fresh bottle each week from your pharmacy.

Some parents prefer to ask the pharmacist to leave it as a powder and make it up themselves at home. The pharmacist will make it up into a liquid unless you ask them not to. It may be helpful if the GP writes 'Supply as dry powder' on the prescription.

Directions for making flucloxacillin into a liquid

Before making it up, the dry powder can be stored at room temperature.

1. Wash your hands.
2. Boil enough water to reconstitute the liquid medication. Allow water to cool to room temperature. **Do NOT** use boiling water to make up the medication.
3. The amount of water you will need to add will be stated on the manufacturer's label. Use an appropriately sized syringe or measuring cylinder to measure the correct volume of cooled, boiled water.
4. Shake or tap the dry powder in the bottle to loosen it.

5. Add the measured water to the medicine bottle, replace the lid and shake thoroughly until all the powder has dissolved.
6. Write on the bottle the date you made up the liquid. Once your medication has been reconstituted it has **seven days expiry**. Please discard any remaining liquid after this time.
7. Store your reconstituted medication in a fridge.

Please note the volume of water needed to make up your medication may change if the brand of your medication changes.

If you have any questions or problems, please ask your pharmacist.

Flucloxacillin is available in capsules for older children.

How to give Flucloxacillin

Flucloxacillin should be given in divided doses, evenly spaced throughout the day. The dose will depend on the weight of your child and the doctor will increase the dose as your child grows. The dose is usually given between two to four times a day.

If possible, flucloxacillin should be given when your child has an 'empty stomach' i.e. one hour before feeds. This helps to ensure that the maximum amount of antibiotic is absorbed, but we understand that with babies this can be very difficult. The most important thing is that you remember to give it as often as directed on the label, every day.

Sodium chloride oral solution

Some babies are prescribed sodium chloride supplements. Babies with CF can be slightly deficient in sodium, causing a delay in growth. The need for supplementation is determined by checking the baby's urine. Sodium chloride solution will

be prescribed until the baby is growing normally, has started to eat sodium containing weaning foods and has a normal level of sodium in their urine. The dose is based on the baby's weight and directions will be given on the label.

Preparation:

Sodium chloride 1mmol/mL solution (292.5mg/5mL).

This medicine is not routinely stocked by community pharmacies and the pharmacy will need the prescription in advance so that they can order it. Please ensure that you obtain a prescription and give the pharmacy adequate notice. Always check that the correct concentration of sodium chloride solution has been given (1mmol/mL).

The expiry of the product and how to store the sodium chloride will depend on the manufacturer. Please read the label on the product.

General advice about medicines

- **Make sure that you always have enough medicine. Order a new prescription at least two weeks before you will run out.**
- Read the information leaflet provided with each medicine. It contains information on how to take the medicine. It also lists possible side-effects and provides additional advice and information about the medicine. If your child experiences any side-effects from any of their medicines please inform the doctor or nurse.
- Try to give medicines at about the same time(s) each day so that this becomes part of your child's daily routine, which will help you to remember to give the doses.

- If you are not sure a medicine is working, contact your doctor but continue to give the medicine as usual in the meantime. Do not give extra doses as you may do harm.
- Make sure the medicines you have at home have not reached the 'best before' or 'use by' date on the packaging. Give old medicines to your pharmacist to dispose of.
- Medicines must not be mixed in the baby's milk, baby bottle or any other drink bottle:
 - The medicines may not work as well, if they mix in the milk
 - If the baby does not like the taste of the medicine, they may not drink the milk and could put them off milk
 - If the medicine is placed in the bottle and the baby does not drink it all, the child will not receive the full dose of medicine

Where should I keep the medicines?

Some medicines, particularly antibiotics once reconstituted, need to be stored in the fridge. Your doctor or pharmacist will tell you where to keep the medicine, for example, if it needs to be kept in a fridge.

- Make sure that children cannot see or reach the medicine.
- Keep the medicine in the container it came in.

Unlicensed medicines (Medicines for Children leaflet)

Many medicines used in the treatment of CF, particularly in children, are unlicensed. The enclosed leaflet about unlicensed medicines will help to explain what this means. If you wish to discuss this further, please speak to your doctor or pharmacist.

Obtaining medicines from your GP and pharmacy

Prescriptions for children are free of charge.

Most GP practices offer an online service, allowing you to request repeat prescriptions online, using a website or an app. Medicines can then be collected from your chosen pharmacy. Your GP practice will be able to advise you.

This link to the NHS website is very useful in explaining the process of obtaining medicines and the services offered by pharmacies, some of which may deliver to your home.

www.nhs.uk/nhs-services/prescriptions-and-pharmacies

You can request that the prescriptions are dispensed at the pharmacy of your choice and nominate that pharmacy to receive your electronic prescription.

You can change the pharmacy at any time and if you are not satisfied with the service you are getting, we advise you to find an alternative pharmacy that can offer the service you require.

If you find that the prescription that you get from your GP does not provide enough for a full month of treatment (e.g. not enough enzymes for a month), speak to the GP or Practice Pharmacist and request that the amount on the prescription is altered.

It is helpful to bring a copy of your child's repeat prescription to clinic appointments. Medication reviews in clinic may include looking at the records of prescriptions that the GP has prescribed for your child.

Oral syringes

Whenever your child is dispensed a liquid, you are entitled to an oral syringe to measure the liquid. If your pharmacist does not provide one, please ask. If you require additional oral syringes, you may be able to obtain them from the CF Unit.

The oral syringes should be cleaned and reused.

Cleaning the syringe

- Clean immediately after each administration using fresh, warm, soapy water. It is essential to draw the plunger in and out several times until all traces of medicine are removed from inside tip.
- Separate the barrel and plunger and wash both thoroughly in warm soapy water.
- Rinse under the cold tap and shake off excess water. Wipe dry with a clean paper towel.
- Store in a clean, dry container and reassemble when required.

The oral syringes can be sterilised between use if necessary (e.g. for young babies using sterilised bottles). They can be sterilised:

- in cold sterilising solution (but should not be left permanently in the sterilising solution between uses),
- in a steam steriliser,
- by immersion in boiling water for three minutes.

And finally

If you experience any problems in obtaining medicines, either you, your GP or pharmacist can telephone any of the contacts below for further advice.

Paediatric Cystic Fibrosis Unit: 0113 392 7125

Paediatric Cystic Fibrosis Pharmacist: Helen Cunliffe,
direct line: **0113 392 5250** or **0113 243 2799 bleep 80-2534**

CF Nurse Specialists: 0113 392 2582

If you contact the CF Unit it is helpful if you can have the contact details of your GP and pharmacy available so that we can contact them to help resolve any problems.



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