SACFA • SOUTH AFRICAN CYSTIC FIBROSIS ASSOCIATION

FINDING OUT

A GUIDE FOR PARENTS AND CARE-GIVERS OF CHILDREN NEWLY DIAGNOSED WITH CYSTIC FIBROSIS

ACKNOWLEDGEMENT

This booklet has been based on the factsheet “Finding out about cystic fibrosis - a guide for parents”, published by the Cystic Fibrosis Trust in the UK in March 2013 (used with permission).

This initial document has been expanded and modified so as to be applicable and value adding to parents and caregivers within a South African context.

ADDITIONAL SOURCES

The South African Cystic Fibrosis Consensus Document 2012 (4th ed).

Cystic Fibrosis: A guide for patient and family. 2012 (4th ed) - David M. Orenstein Jonathan E. Spahr & Daniel J. Weiner

Nutrition for life. 2012 – Dr. Tuschka Reynders, Registered Dietician and Nutritional Consultant, Pretoria.

My thoughts about living with Cystic Fibrosis. 2006 – Prof. Warren J. Warwick MD, Professor of Paediatrics, Biomedical Science and Medical Physics, as well as Professor for Cystic Fibrosis Patient Care, University of Minnesota Medical School.

Eating well with Cystic Fibrosis: A guide for Children and Parents. 2006 – Carolyn Patchell, Dietetic Department, Birmingham Children’s Hospital.

Infant Reflux and Paediatric GERD 2005 – Roni Maclean, Infantrefluxdisease.com

This booklet has been compiled to:

- Help you get a better understanding of Cystic Fibrosis (CF) – but not intended to replace any advice you may receive from your CF clinic and / or CF specialist.
- Address most of the frequently asked questions (FAQ's) that parents new to CF may have.
- Offer helpful and practical advice from parents who have walked the road themselves.
- Enable and support you in giving your child the best possible care in all aspects of his / her life.
- Introduce you to the South African Cystic Fibrosis Association.
- The impact of hearing the news that your child has been diagnosed with a serious medical condition can be very challenging. A section of this document is therefore dedicated to addressing the emotional impact of coming to terms with the news that your child has Cystic Fibrosis.

The South African Cystic Fibrosis Association

The South African Cystic Fibrosis Association (also known as S.A.C.F.A) is the national governing body of the three regional associations based respectively in the Cape, Gauteng and KwaZulu-Natal.

The Association aims to:

- Increase both awareness and knowledge of Cystic Fibrosis.
- Provide access to sound advice and support to people diagnosed with CF and their respective families.
- Facilitate the maintenance of proper treatment facilities in state hospitals.

You are more than welcome to contact your regional SACFA Association for support, or enquire about membership and the registration of a Cystic Fibrosis patient / family on the "National Cystic Fibrosis Database".

The Association's website: http://www.sacfa.org.za

Any words appearing in ***bold italic type*** in this document can be found and are explained in the Glossary at the back of this document.

Contents

What is cystic fibrosis?

Cystic Fibrosis (CF) is an inherited disorder that affects the way in which salt and water move into and out of the body's cells - affecting mainly the lungs and digestion.

Normally, the liquids and ***mucus*** that line many of our organs are clear and lubricating, and help to protect them from infection. In CF there is a default in a ***gene*** (the 'CF gene') that controls the amount and composition of fluid lining the airways and other organs.

The fluid lining the airways in the lungs is reduced, resulting in an excess of sticky secretions (mucus) that are prone to infection and difficult to cough up. In the pancreas, the sticky secretions may block the flow of digestive juices into the gut leading to impaired digestion and poor absorption of food. The sweat glands are also affected, in that they excrete a much saltier sweat than normal.

Not all children are affected in the same way or to exactly the same degree – some are affected more and some less.

As a parent of a child with CF you will meet many people involved in the care of children with Cystic Fibrosis - but you will be asked to carry out some of the treatment yourself. To do this effectively, you will need to understand as much as you can about the condition.

This guide attempts to answer most of the questions that you may have regarding CF – covering the FAQ's (Frequently Asked Questions) that other parents, who have been trying to come to terms with the news that their child has Cystic Fibrosis, have asked over the years.

How do you feel?

Parents, on receiving news of their child's diagnosis, often experience overwhelming emotions and may experience the news as traumatic.

Even though you have not lost a child, it may feel like a time of intense grief as you come to terms with the loss of a "healthy" child and learn to deal with the realities of caring for a child with CF.

This is most likely an incredibly difficult time for you.

Parents may experience a lot of difficult feelings when faced with the news and the reality of the diagnosis. It most probably came as a considerable shock. Many parents in your situation feel in a state of panic or numb. You might feel angry, frightened, disbelieving and overwhelmed by the news.

Please be reassured - these feelings and reactions are VERY NORMAL.

For some of you, along with the intense emotion, there may also be a sense of relief at having a diagnosis - particularly if you "knew" something was not right but couldn't get your fears taken seriously, or couldn't get your questions answered.

This may be a time when you don't want to hear any more information, or it may be a time when you want to learn absolutely everything there is to know about CF. No matter how you are feeling, please know that it may be a little hard for you to take in a lot of new information right now.

You may however find it helpful to find people with whom you can talk about how you are feeling, as well as about CF.

You are more than welcome to contact SACFA for such support. If you are interested, we can connect you with other families who have been through what you are going through now.

Please make sure you talk to people who have up to date knowledge of CF, as treatment has improved considerably over recent years, and out of date information could give you an entirely wrong impression.

The following paragraphs will look at some of the confusing and conflicting feelings you may be experiencing at this time, and offer some suggestions on how you can deal with them.

Disbelief

Parents who receive the news of their child's diagnosis may find that they experience times when they cannot think of anything else, whereas at other times they may struggle to think about CF at all – finding it difficult to believe that the diagnosis is correct.

You just can't fathom that your child has a chronic condition which will always affect his/her life as well as your own – it feels so incredibly unreal.

You may feel paralyzed with shock or blanketed with numbness. You may feel that everything becomes meaningless and overwhelming – that nothing makes sense. You may wonder how you can go on, if you can go on, why you should go on. You may try to find a way to simply get through each day.

You may begin to question the how and the why, in trying to understand what has happened to you and your child.

Anger, guilt and blame

You may be angry, you may feel guilty or you may want to blame somebody. You may feel anger because this was not supposed to happen.

You may be angry at this unexpected, undeserved and unwanted situation that you find yourself in.

You may even feel anger towards yourself that you couldn't stop Cystic Fibrosis from happening to your child.

You may also be experiencing guilt – for "giving" CF to your child. It is very common for parents to feel guilty when their child is diagnosed with CF - even though the truth is that you did not GIVE your child Cystic Fibrosis.

It was not something that you chose to do or had any control over. If you could have changed things – of course you would have. BUT YOU CAN'T. You are not to blame.

You may, however, feel that you **want to blame something or someone** – be it yourself, your partner, your child - for having CF, Cystic Fibrosis itself, or God.

Self Pity

You may experience feelings of self-pity.

You may ask: "Why us? Why me? What did I do to deserve this? Having children is not supposed to be this way! This is not the plan I signed up for!"

You may compare your circumstances with how you "assumed" things would be: "I assumed my child would be healthy."

You may compare your circumstances with how it "should" have been: "I should have had a healthy child!"

You may even find that you compare your child to other "normal" children.

You may cry and even rage because you did not get the healthy child that you wanted.

You may feel incredible pain and emotional distress at the mere thought that your child has been diagnosed with CF.

You may. And it is OK. It is OK.

Strained relationships

This is a very difficult time for you and your whole family – everyone trying, in their own way, to come to terms with the diagnosis of Cystic Fibrosis.

You may experience that relationships with some family members and friends become strained.

You may feel and think that some people don't care because their actions / lack thereof do not make you feel cared for, supported, or understood.

Most people, however, just DON'T KNOW HOW to appropriately respond to the news of a CF diagnosis - or for that matter – to YOU - who is experiencing and grieving the loss of a healthy child.

It is important; however, that during this time, you try to find at least ONE relationship, where you can openly express and share your emotions, concerns, fears, worries and your daily walk with CF.

Bewilderment

You may be given a huge amount of information and advice from various sources - some of it may even seems to conflict. Cystic Fibrosis is a complicated condition and each child is affected differently, so everyone's experience differs somewhat.

You cannot expect to know everything about the condition immediately - and NO-ONE EXPECTS YOU TO.

It may be a long time before you understand CF in general - and only experience, over time, will tell you exactly how it is affecting your child.

Please ask as many questions as you feel you need to, until you understand sufficiently whatever it may be that you need to understand.

Many parents find it helpful to start a "List of Questions", to which they add anything that they wonder about or may be unsure about, as and when it is thought of. This "updated" list with questions may then be discussed with the CF doctor / clinic team during scheduled appointments. Your CF team will expect and will encourage you to ask questions, as learning about CF is very important and will only happen over time.

It is also really helpful to make short notes regarding the answers that you receive, for future reference, as one just cannot remember everything!

If something is worrying you or stressing you out – please contact someone who can help. Phone your clinic or e-mail your CF doctor. Never wait for things to get worse.

Positive approach

It can be difficult to know how to engage with a child who has a chronic condition / illness. Parents may try to over-protect their child because they want to keep them safe and healthy. However, one of the most valuable gifts that you can give your child with CF (and yourself) is to treat him or her as a normal child who happens to have CF, and not as an ill or sickly child who should not be expected to do normal things. There is no reason to treat them differently in relation to behaviour, education, extracurricular activities and even physical activities. If you do treat them differently, you will not only be doing them a disservice - but may also be creating problems for yourselves, as parents, in the long run.

However, children should know that they have CF, and that having CF means that for them, certain things are different from other children – but they should also know that they have the same rights, privileges and responsibilities as anyone else in the family and at school.

Children with CF should thus not be allowed or encouraged to use CF as an excuse for getting out of unappealing chores, activities or responsibilities.

Treated normally, they will think of themselves as normal and subsequently will be normal - which they are – except that they have CF.

The life expectancy for children with CF has improved significantly over the years due to improved treatment methods and, with the latest advances in research and medicine, there is every chance that new therapies will be developed to more effectively limit the harm from the disease.

Many people with CF live well into adulthood leading normal lives – so it is of utmost importance that your child with CF follows the normal developmental milestones of their peers to prepare them for independence later in life. Independence is something you can speak to your CF doctor / clinic team about as your child grows and matures. It is, however, to be introduced and encouraged from a very young age.

It is, however, also necessary to acknowledge and emphasize that it can sometimes be really difficult to follow a positive approach.

The wish to protect your child is very powerful and understandable and can make it really hard to treat children with CF normally and encourage them to become independent.

It is furthermore important not to forget the impact there may be on any brothers and sisters of your child with Cystic Fibrosis. They are just as likely to be upset and anxious and may even feel either guilty – because they are well – or left out because of all the attention the child with CF is receiving.

Getting help

As with all things, life is easier if there is someone or somewhere to turn to for practical advice, as well as emotional support.

Your CF doctor, CF clinic team and SACFA are sources of information and experience, which parents can draw on regarding any aspect of life with Cystic Fibrosis.

It is necessary to remember that Cystic Fibrosis affects the whole family including brothers, sisters, aunts, uncles, grandparents and other relatives. It is thus important for them to be included in the learning process as well.

May you, at your own pace, and in your own way, come to terms with the news that your child has Cystic Fibrosis.

And may you come to experience, over time, the truth of Si Kahn's lyrics: "It's not what you've been given, it's what you do with what you've got."

Why does my child have CF?

CF is a ***genetic*** condition. A baby may be born with CF only if **BOTH** parents are carriers of the faulty / ***mutated*** Cystic Fibrosis ***gene*** (the CFTR gene on chromosome 7).

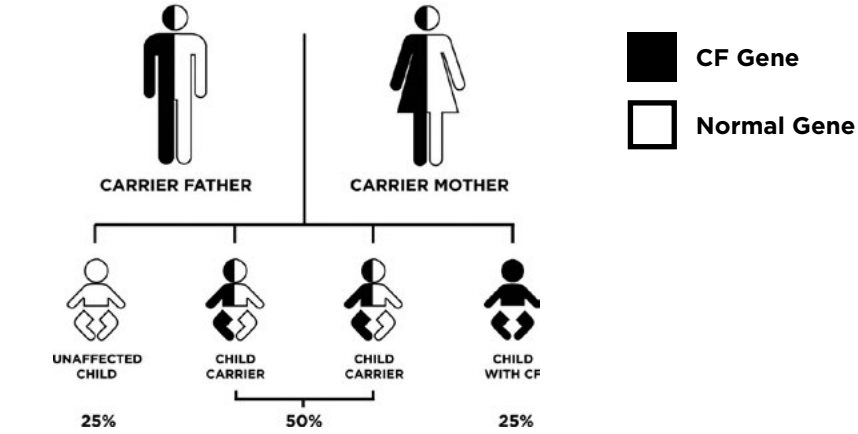
It does however not automatically mean that because both parents have the faulty / mutated CF gene, that every baby they give birth to will have Cystic Fibrosis.

If both parents are carriers, a child has a:
- One in four (25%) chance of being born with CF.
- Two in four (50%) chance of being a carrier – but not having the disease.
- One in four (25%) chance of being completely free of it, i.e. not having CF nor being a carrier of the faulty CF gene.

Most carriers of the faulty gene have no idea that they are carriers, because they are completely healthy. Most people find out only when:
- they have a child with CF, or
- a close relative is affected and they are tested.

In South Africa one person in every 20 in the white population and one person in every 55 in the population of mixed ancestry, is estimated to be a carrier of the faulty CF gene. The current carrier rate in the Black African population is not clear with the carrier rate being reported between one in 14 and one in 91 depending on the language group in Southern Africa. It is thus approximated that statistically one in every 2 000 white babies and one in every 12 000 babies of mixed ancestry will be born with CF. For the black African population it would vary between one in every 1 000 and one in every 33 000, depending on the language group.

The diagram below shows that if both parents are carriers there is a 1 in 4 (25%) chance of having a child with CF, and a 3 in 4 (75%) chance of having an unaffected child (child without CF, even though he / she may be a carrier).



However, as each baby is conceived separately (apart from identical twins), the risks are exactly the same each time and therefore what happened in the last pregnancy doesn't increase, or decrease, the risk next time. The chances are exactly the same for each pregnancy.

Two carrier parents might have several affected children, or only unaffected children or a mixture of both affected and unaffected children. **It is impossible to predict what will happen for any particular pregnancy.**

So, if carrier parents already have a child with CF, this does not change the risk in the next pregnancy. Each pregnancy has exactly the same chance as the one before: 25% to have a child with CF, and 75% to have a child without CF. Both boys and girls have an equal chance of being affected.

Genetics is a complicated subject – keep asking questions until you are sure that you understand. You can ask to be referred to a genetic counsellor if you have any more questions in relation to the risks of conceiving a child, or another child, with CF.

How is CF diagnosed?

Sweat test
In the 1950's it was recognised that children with Cystic Fibrosis have more salt in their sweat than children who do not have CF. Some parents comment that their child tastes salty when they kiss him or her. However, children with CF do not sweat more than children without CF.

The ***sweat test*** measures the amount of salt in the sweat. There are a number of methods for collecting sweat, none of which is painful or dangerous.

First the skin, usually on an arm or leg, is cleaned and two discs of a special jelly are placed on the skin a few inches apart. The discs of jelly are connected to a battery, which passes a tiny electric current between them – this does not hurt. After about 5 minutes the skin underneath one of the discs should be sweating nicely. The discs are removed and the skin is dried.

A paper disc or special device (Macroduct) is put over the place that was sweating, and the new sweat produced will flow into the tubing. Collecting enough sweat for the laboratory to be able to measure the salt level takes anything from 10 to 30 minutes. Occasionally not enough sweat is produced and the test has to be repeated.

If the salt level is abnormally high, your child will be diagnosed as having Cystic Fibrosis. Sometimes the result will be border-line and the test may need to be repeated to be sure of the result.

A sweat test is done in the following situations:

- When symptoms that may indicate CF, are present:
  - a troublesome cough
  - repeated chest infections,
  - prolonged diarrhoea / abnormal stools
  - poor weight gain
  - failure to thrive

- If your baby is born with a blocked bowel, CF may be suspected. Sometimes a baby born with CF may be very ill in the first days of life with an obstruction of the bowel. There are several types of obstruction but the most common in CF is ***meconium ileus***. All new-born babies have ***meconium*** in their bowel – this is the thick, black material they pass the first time their bowels open. In CF the ***meconium*** can be so thick and sticky that it blocks the bowel. Babies with meconium ileus often need an urgent operation to relieve and bypass the blockage to allow the bowel to recover. When the baby has recovered from this, a sweat test will be done to see if the blockage was due to Cystic Fibrosis.

- If you have a child with CF, other children in your family with similar symptoms may need to undergo a sweat test.

Genetic testing

In cases where the result of the sweat test is border-line, a sample of blood is taken and this specimen is used to look for the CF gene. This is also useful for identifying the specific gene mutation, which may be useful in future for prescribing specialised CF medications targeting specific ***genetic mutations.*** More than 1800 different mutations of the CF gene have thus far been discovered.

Currently the most common mutation in the white and mixed ancestry ("coloured") population in South Africa is the ΔF508 mutation. The most common in the black South African population is the 3120+1G→A mutation.

It is furthermore useful to know which mutation/s is/are present in a family with CF, especially when a sibling or other family members are planning their own families. If any individual family member tests positive for CF carrier status, their partner should also be tested to assess their risk of having a child with CF.

It is recommended that CF-affected individuals and their families see a ***genetic*** counsellor in order to accurately calculate risks with regard to their family members.

How does CYSTIC FIBROSIS affect the body?

Because Cystic Fibrosis results in the production of thick, sticky mucus, the organs that are particularly affected are those where the mucus has an important job to do, particularly the digestive system and the lungs.

How does CF affect the digestion?

The ***pancreas*** is a gland in the abdomen. One of the pancreas' functions is to produce digestive juices, or ***enzymes*** and bicarbonate, which pass into the intestine, where they help break down the fat, protein and carbohydrate in food, so that the nutrients can be absorbed into the body.

In someone who has CF, the small channels down which the digestive juices flow become blocked with sticky ***mucus***. The enzymes then build up in the pancreas, which becomes inflamed. This causes the formation of ***cysts*** and ***fibrosis*** = CYSTIC FIBROSIS.

As a result, the enzymes produced by the pancreas that are needed to digest the food do not reach the food. Food will then pass through the system undigested. This is often referred to as pancreatic insufficiency and will require enzyme replacement therapy.

The effect of CF on the pancreas and digestion varies from person to person. Some children with CF produce enough pancreatic enzymes to digest their food. Normally, however, the majority do not. As such, most new-born babies with CF cannot digest milk and without enzyme replacement therapy, fail to gain weight and have very loose stools.

How can I help my child's digestion?

It is now possible to replace most of the missing ***enzymes*** with ***pancreatin*** – this is a general name given to all pancreatic enzyme medicines. They come in the form of capsules and contain many ***enteric-coated microspheres*** or granules that contain the enzymes.

The outer capsule dissolves in the stomach, releasing all the microspheres. They then pass into the upper part of the small intestine, mixing with the food, allowing digestion and absorption to take place. Because the microspheres are protected from the acid in the stomach by a special coating, the microspheres are not released in the stomach but in the upper small intestine where they are needed to do their job.

The most commonly prescribed pancreatic enzyme for people with CF in South Africa is Creon.

Do all babies with CF need enzymes?

A few babies do not need enzymes at first but may need them later on. It is important to watch and see whether their bowel function and growth remains normal.

Most babies with CF need them from birth / time of diagnosis and will need to take them for the rest of their lives. This is normally not a problem. It becomes routine and children soon learn to take enzymes by themselves.

How should pancreatic enzymes be taken?

Most young babies and toddlers will take the microspheres removed from the capsules. The capsules are to be opened and the contents are to be mixed with a little fruit puree or applesauce and given to your baby / young child right at the start of the feed or meal from a teaspoon. This is to be taken in one or two swallows. Fruit puree / applesauce is recommended as it is an acidic food and as such does not damage the special coating on the microsperes.

As soon as your child is able to swallow tablets, the capsule it is to be taken whole. This is normally around 5-6 years of age, although some children may learn earlier.

Do not mix the enzyme with hot food, or with the meal itself, as this will reduce the effectiveness of the enzymes.

Try to give the enzymes at the beginning and part way through a meal, as the enzymes work best for 25-30 minutes after having been taken.

If your child is a slow eater, it may be helpful to split the enzyme dose for the meal in two and take ½ at the beginning and ½ in the middle of the meal.

However, if you forget to give the enzymes at the start, give them as soon as you remember, as long as it is within 5 -10 minutes or so after your baby or child has finished eating. Although enzymes work best when taken at the beginning or during a meal, it's better to take them shortly after the meal / snack than to miss them altogether.

Do not crush the granules or allow your child to chew them, as this will render them ineffective.

Make sure no granules remain in your baby or child's mouth, as these can sometimes cause mouth sores.

How many enzymes will my child need?

The dosage of enzyme needed varies from one child to another and is very much a matter of trial and error. **You will be advised on the amount to give your baby / child by your CF doctor and dietician.** You are advised not to adjust the recommend dose without discussion with your CF doctor / dietician.

In general, it helps to remember the following:

- Pancreatic enzymes are to be taken with all foods containing fat, protein or starch.
- Food containing sugar (glucose and fructose) and little protein or fat, **do not need enzymes**. These "free snacks" includes fresh or tinned fruit, jelly, fruit juice, fizzy drinks, boiled sweets and jelly sweets.
- Fatty meals will need more enzymes than low fat meals.
- Although most snacks will require fewer enzymes than a meal, some snacks are very fatty and may need as much, or even more enzymes.
- Infants are normally started on a ⅓ or ½ capsule (i.e. 3000 to 5000 units) and one or two capsules per meal in older children. Your CF doctor will increase the dosage gradually until your child's bowel symptoms are controlled.
- On average 1 capsule of Creon, which contains 10 000 units, is required for every 5 gram of fat consumed.

How will I know if the amount of enzyme given is correct?

There are several signs, which may indicate the dose of enzymes given is insufficient:

- Pale, floating stools, which are difficult to flush away.
- Oil floating on the water.
- Stool is foul-smelling.
- Increased stool frequency.
- Abdominal pain, bloating, flatulence or wind.
- Poor weight gain.
- Failure to grow and thrive.

If you child is suffering from any of the above-mentioned symptoms, please contact your CF doctor and dietician for advise – please do not change the pancreatic enzyme dose without checking with your CF team.

What if I forget to give the enzymes?

Forgetting a single dose is not likely to have an adverse effect, although your child may have looser stools and some stomach cramps afterwards. However, if enzymes are missed regularly, digestion and absorption will be poor and your child may fail to gain weight and his / her growth may suffer as a result.

What if my child does not eat a meal after having the enzymes?

Remember that your child is no different from any other. All toddlers / young children refuse to eat at some time - some more than others - and your child will be no different. However, your child has CF and may not feel very hungry when they have an infection.

Your child will come to no harm if occasionally nothing is eaten after a full doze of enzymes. They may, however, become a little constipated as a result.

If this happens frequently then it may be helpful to give half the dose at the beginning of the meal and the remainder halfway through. It also helps to have some chocolate on hand for those times when they have not eaten enough food for the enzymes already given.

Nutrition is very important in CF, but as with all children, you do neither yourself nor your child a favour in the long run if each mealtime ends in a battle because of untouched or unfinished meals.

If mealtimes are becoming difficult, please talk to your CF doctor and dietician for support and some useful ideas and suggestion that may help. Do remember however, that most children will go through periods when they do not seem to eat much.

Breast or bottle?

Most babies with CF thrive on breast milk. If you are able to breastfeed this is generally the best idea – but formula milk is fine as well.

Whichever way your baby is fed, his/her weight gain will be monitored during visits to your CF doctor. If your baby fails to pick up weight or grow normally, a change of milk or addition of an extra-calorie supplement may be advised. Your dietician will be able to assist and advise you in this regard.

What about weaning?

This is the same as with any baby. If a baby appears exceptionally hungry, weaning a little earlier than usual at approximately three months may be advised. You may be asked to start introducing solids from an earlier age for your child with CF. Remember that children with CF may need more calories than other children to grow at the same rate. Your CF doctor and dietician will advise and support you in this regard.

What should a child with CF eat?

In general, children with CF should eat whatever the rest of the family is having.

However, even with pancreatic enzyme replacement a child with CF may not absorb all the nutrients and nourishment needed to grow normally, and may therefore need more calories.

Children with CF are also encouraged to have small high calorie snacks in between meals – not to replace meals but to supplement them.
Some suggestions for adding nutrients and calories to your child's food:

Eggs

- Mash a boiled egg yolk in with porridge.
- Mix raw eggs in with mashed potatoes.
- Mix egg with some mayonnaise and use on sandwiches.
- Make scrambled eggs or omelettes with cream and add some grated cheese.
- Add boiled eggs to salads.
- Add raw eggs to mince, stews or sauces when cooking.

Cheese

- Filling on a sandwich.
- Add cheese powder to food.
- Add full cream cheeses (cottage, cream, gouda, cheddar and feta) to meals or give as snack in-between meals.
- Grated over vegetables.
- Grated and mixed in with mashed potatoes or mac-and-cheese.
- Extra cheese on pizza.

Full cream milk

- Drink after meals or in-between meals as a snack.
- Prepare porridge with full cream milk instead of water.
- Prepare soup or gravy with full cream milk instead of water.
- Make enriched milk by adding 4 teaspoons full cream milk powder to each cup of fresh milk, Milo, Nesquick, Hot chocolate or Horlics.
- Add full cream milk powder to yoghurt or mashed potato.
- Make jelly with milk instead of water.
- Add ice cream to cold drinks or milk to make a milkshake.

Sugar, honey and jam

- Add to porridge and vegetables.
- Add 2 teaspoons to a glass of fruit juice or yoghurt.
- Add extra to porridge, custard and milkshakes.
- Prepare instant puddings with cream and extra sugar as dessert.
- Spread on bread together with peanut butter or cheese.
- Please note not to give any honey to children younger than 1 year of age.

Peanut butter

- Mix with custard and a little bit of milk to make a delicious dessert / protein snack.
- Add to porridge.
- Spread it thick on bread.
- Mix in with spinach.
- Eat it by the spoonful.

Fruit

- Serve with custard.
- Mash a banana and mix in porridge.
- Mash banana or other soft fruit and liquidise with milk, ice cream and yoghurt.
- Drink fruit juice in between meals.
- Puree fruit and mix into yoghurt.
- Give as a snack between meals.

Dried fruit

- Give as snack in-between meals.
- Boil in a small amount of water and give as desert together with custard.
- Boil in small amount of water, mash and mix in with porridge or yoghurt.

Nuts

- Give as snack in-between meals.
- Blend very fine and mix in with porridge or yoghurt.
- Please note not to give any nuts to children younger than 1 year of age.

Butter and cream

- Add to porridge, mashed potato and vegetable puree or vegetables.
- Add to cold drinks to make milkshakes.
- Spread thick on bread or toast.

Yoghurt

- Add to porridge.
- Eat full cream yoghurt as a snack between meals.
- Spoon over fruit or fruit salad.
- Add some cream to low fat yoghurt if full fat yoghurt is difficult to come by.

Avocado pear

- Mashed and spread on bread.
- Just as is.
- In a salad.

Custard

- Prepare with full cream milk and give as a dessert or a snack.

A booklet by Prof. Tony Westwood, of the Red Cross War Memorial Children's Hospital in Cape Town furthermore provides useful recipes and advice on CF nutrition.

"Eating, enzymes and exercise – recipes and nutritional ideas for people with Cystic Fibrosis in South Africa" is available both through SACFA, and the dietician at your CF clinic.

Will my child need special dietary supplements?
If your child's weight or appetite is poor, your CF doctor or dietician may recommend a special supplement to boost nutrient intake. These will only be used if ideal growth rates cannot be achieved using normal foods.

There are a wide range of dietary supplements available on the market – your doctor or dietician will advise you on which ones may the most suitable to use, taking into consideration the age of your child as well as his / her particular needs.

Does my child need extra vitamins?
Children with CF normally do not absorb vitamins well, especially those vitamins which are dissolved in fat (vitamins A, D, E and K). Your child will usually need additional supplements of these vitamins in the form of drops or capsules each day or at regular intervals.

These vitamin supplements will not be the same as the normal over-the counter vitamin supplements given to other babies and young children. Your CF doctor will advise which preparation is the best to use, as well as the dose of each supplement.

To ensure maximum absorption of these vitamin supplements, it is advised to give them just after you have given your child a dose of pancreatic enzyme, before he / she starts eating their meal. If your child refuses to take the vitamins, it may help to give it to them disguised in a spoonful of yogurt.

Extra minerals, such as iron, are not usually needed.

Does my child need extra salt?
Please note that it may be dangerous to give a baby extra salt without medical advice.

Your CF doctor and dietician will advise you on whether, and how much, salt should be supplemented, as well as when and how to give them to your baby or child. Children with CF are however encouraged to use the saltshaker liberally and choose more salty foods and snacks during warm and hot weather, as children with CF lose more salt through sweating.

It is recommended that your child increase their fluid intake if they increase their intake of dietary salt.

Why is a child with CF weighed and measured more often?
It is important to ensure that a child with CF is growing well, so your CF doctor will plot your child's measurement on a growth chart to carefully assess his / her growth on a regular basis. This enables them to support you with the necessary advice to ensure your child is reaching their full growth potential.

Does CF affect the teeth?
Cystic Fibrosis does not affect the teeth directly, although poor nutrition may affect the teeth's growth. Most antibiotic medicines are now sugar-free but a lot of foods that are full of calories are very sweet and therefore, although very helpful for CF, may not be so good for their teeth.

To help keep teeth healthy, it is recommended that:
- Your child should brush his/her teeth thoroughly every morning and night with a small amount of fluoride toothpaste.
- Children under 8 years are supervised while brushing their teeth.
- They rinse their mouths with water after sugary snacks and drinks.
- You encourage your child to eat sugary foods at the end of a meal rather than in-between meals.
- Your child visits the dentist every 4 months for a check-up.
- You explain the CF diet to your dentist and ask for additional advice on oral hygiene.
- You ask your dentist if any fluoride supplements are necessary.

Other problems in the digestive system or the gut

There are several other ways in which CF can affect the gut:

Tummy aches

Many children without CF have tummy aches or cramps for no apparent reason. They usually go as mysteriously as they come, and children with CF will occasionally have these types of tummy aches too.

Children with CF may however complain of tummy aches after a bout of coughing. If this is happening regularly then it is advised that you seek advice from your CF doctor who may arrange for further investigation.

If you are worried about your child's tummy ache, please speak to your CF doctor.

Extremely loose stools or mild constipation are frequent in CF and may be the cause of stomach cramps. If this persists, speak to your CF doctor and dietician and possibly review the pancreatic enzyme replacement therapy.

If your child has severe acute abdominal ache, **please seek medical advice immediately.**

Constipation

With simple constipation, the stools are infrequent and usually hard, dry, small and difficult to pass (straining). Passing a tiny amount of stool with straining several times every day still represents simple constipation.

Some triggers for constipation in children with CF may include: inadequate hydration (especially low fluid intake during hot weather), a low fibre diet, inactivity, or even an excessive enzyme dosage.

Passing a very wide diameter stool suggests chronic constipation as the rectum is stretched out by lots of stool being present all the time. A wide stool is painful to pass and can potentially cause a fissure or tear in the anus.

Treating chronic constipation is different from treating simple constipation. If you think that your child may be either constipated or chronically constipated, please consult your CF doctor, for proper diagnosis and prescription of applicable and effective treatment.

Distal Intestinal Obstruction Syndrome (DIOS) or Meconium Ileus Equivalent (MIE)

This has nothing to do with the meconium ileus seen in new-born babies, but occurs in older children and adults.

When it comes to CF, it is important to differentiate between constipation and ***distal intestinal obstruction syndrome (DIOS).*** Because the intestinal secretions in CF are thicker than normal and some malabsorption may be present, the CF stool can be sticky and bulkier than normal. This sticky stool may not pass easily through the colon.
DIOS occurs when more and more stool "gets stuck" in the colon.

This is usually accompanied by cramping abdominal pain on the right side. Parents often report a decrease in the amount of stool passed by their child. The pain gets more intense as more and more stool accumulates. It can lead to a decrease in appetite, fullness and vomiting. DIOS can, however, be treated without hospitalisation with special medicines if it is recognised early enough.

Although it is good to be informed, knowledge about potential problems such as DIOS can sometimes cause unnecessary worry.

Remember it is not unusual for a child with CF to have occasional stomach aches or brief changes in bowel pattern. This can be due to an unusually high fat meal, a GI "bug" or even a change in routine.

Contact your CF doctor if the following signs of DIOS is present in your child:
- Increasing abdominal pain (especially on the right side).
- A decrease in the normal amount of stool passed.
- Decreased appetite, fullness and vomiting.

Gastro-Oesophageal Reflux Disease (GORD)

The incidence of GORD, or acid reflux, is increased not only in children with CF, but also adults with CF.

Potential symptoms of GORD:
- Heartburn.
- Spitting up or vomiting.
- Wet burps or frequent hiccups.
- Refusing food or only accepting a few bites despite being hungry.
- Poor sleep habits – typically with frequent waking.
- Chronic hoarse voice.
- Frequent red, sore throat without infection present.
- Gagging themselves with their fingers or fist.
- Respiratory problems – pneumonia, bronchitis, wheezing, asthma, night-time dry coughing, aspiration.
- Poor weight gain, weight loss or a failure to thrive.

As the signs and symptoms of GORD are perfectly mimicked by the traditional symptoms of CF, making a definitive diagnosis may be difficult.

GORD will normally be considered in any CF patient who, despite compliance with their CF therapy, continues to have problems with their lungs, vomiting, abdominal pain or a failure to thrive.

Your CF doctor / clinic team will do some tests, if necessary, to determine if GORD may be present and whether medical intervention, normally in the form of acid suppression medicines, is necessary.

How does CF affect the chest?

In the lungs there are tiny tubes, called ***bronchi***. Air passes down these tubes to reach the specialised parts of the lung, the ***alveoli***, where oxygen enters the bloodstream and carbon dioxide leaves the bloodstream, to be breathed out of the body.

We all have liquid and mucus in our lungs, which help them to function, but in children with CF the mucus produced is abnormally thick due to there being too little liquid. This thick mucus can block some of the smaller airways and this can lead to infection. If not controlled, infections can lead to damage to the lungs.

In the early years infections are usually caused by viruses and certain bacteria e.g. ***Haemophilus influenza*** and ***Staphylococcus aureus***. Later on, infections may be caused by other bacteria as well, including ***Pseudomonas aeruginosa.***

Every infection may potentially cause small, but significant, lung damage that is irreversible. Over time, the accumulated effect of repeated infections therefore may result in permanent lung infection and lung damage.

However, paying meticulous attention to keeping the lungs healthy in early childhood can minimise much of the damage that these infections can cause to the lungs.

Proper treatment of the lungs, as described below, is therefore the key to minimising the chance of severe lung damage in adulthood.

How is the chest treated in CF?

The aim is to keep the lungs as clear of mucus and infection as possible.

There are two main ways in which this is done, both of which are important.

- Prevention and treatment of chest infections, usually with antibiotics.
- Clearing the sticky mucus from the lungs, by physiotherapy and regular physical exercise.

Prevention and treatment of chest infections

Should my child be kept away from other children?
It is impossible to prevent your child picking up infections from other children and adults – so there is no point in isolating him or her for that reason. It is sensible, though, to try and reduce your child's exposure to friends and relatives who have just started a "cold", as this is when they are at their most infectious.

It is also recommended that your child with CF (ideally) only start attending nursery school or crèche at around 3 years of age, if possible.

Although the risks of picking up CF related infections from other children with CF is low, it is advisable to avoid close contact with others with CF, where possible. CF clinic attendance is important though and necessary arrangements will have been made by your CF clinic to ensure that children with different infections are kept sufficiently far apart from each other. You may ask your CF doctor / clinic about their arrangements in this regard.

It is **VERY IMPORTANT** that children with CF avoid smokers. There is sufficient evidence that even "passive smoking" – inhaling the smoke coming from the end of the cigarette or the smoker's exhaled smoke - can harm the lungs, and those with Cystic Fibrosis are particularly vulnerable.

Pets are not a problem unless your child has an obvious allergy to them. This includes horses, although it is important
to avoid the stables, which are commonly contaminated with fungus spores. So – children with CF can ride horses, but should not be allowed to "muck out" the stables.

Can antibiotics prevent chest infections?
There is increasing evidence that early, frequent, and in some cases, continuous antibiotics can prevent or delay the lung damage in Cystic Fibrosis. Your CF doctor / clinic team will discuss the most suitable approach for your child.

Whereas the general population is advised to be cautious in the use of antibiotics, for those with CF antibiotics are taken on a regular basis, when necessary. The benefits are invaluable and life-saving.

What about immunisation?
Children with CF are particularly at risk from the common childhood diseases, especially those infections that may affect the lungs.

Some of these childhood infections, such as measles and whooping cough, may have lasting effects on the lungs of children with CF. The aim is to protect children with CF from these infections from an early age, even before they come into contact with other children at pre-school or school.

Normal childhood immunisations including pneumococcal vaccination are thus recommended, as well as immunisation against chicken pox and hepatitis A.

Children with CF respond in the same way to immunisation as other children, and are no more likely to have reactions to the injections. The normal immunisation schedule is appropriate for children who have CF and injections should be postponed only in very exceptional circumstances (i.e. when they have a fever) and after consultation with your CF doctor. Just having a cough or a cold is normally not enough reason to delay having an immunisation, as there are very few medical reasons to avoid immunisation.

It is furthermore recommended that an annual influenza vaccine - covering the expected strains for the coming winter season - is to be given as a routine in March /April, as soon as it becomes available - except when there is a known allergic reaction to egg.

Your CF doctor / clinic team will furthermore advise you on the benefits that an influenza vaccination for each member of your family, and not only your child with CF, offers your child, as well as your other family members.

Your CF doctor / clinic team will be familiar with the latest immunisation recommendations and will be able to give you advice on what is best for your child.

As such, please discuss any questions or concerns that you may have with regard to immunisation with them.

How will coughs and cold affect my child?
All children, whether they have CF or not, suffer numerous colds in their first few years.

In children with CF the symptoms often lasts longer because of the increase in lung secretions. Colds are caused by viruses and the only cure is time – but children with CF may need antibiotics to prevent an infection with bacteria following straight on (secondary infection).

Extra physiotherapy is often needed, and helpful, if there is a lot of extra mucus.

Are cough medicines useful?
Cough medicines supress the cough, and therefore reduce the ability to clear mucus, which is essential in children with CF. If a child with CF is coughing, it is important to find out what is causing it; there may be an infection, which requires antibiotic and physiotherapy treatment.

Cough medicines should thus not be given to children with CF without discussing it with your CF doctor / clinic team first.

How are chest infections treated?
- A child with a chest infection needs extra physiotherapy (longer and more often), to clear the mucus and infected material from the lungs.
- She / he also needs antibiotics. Your CF doctor will decide which antibiotics to use by knowing which bacteria are likely to be present and by taking a ***sputum*** sample or swab for verification. The antibiotics will usually be given by mouth as liquid medicine, tablets or capsules. Sometimes they are inhaled as a mist from a ***nebuliser*** or given by intravenous route, i.e. directly into a vein.

Are any special tests needed?
If possible a ***sputum*** sample, either from a throat or cough swab or nasopharyngeal aspirate ***(NPA)***, is taken to identify the bacteria (if any) causing the infection.

A chest X-ray and, occasionally, blood tests may be helpful.

Older children may be asked to blow into a tube or machine to check their lung function. This is particularly useful if it is done regularly between infections, because the levels often fall before the infection becomes obvious, enabling it to be caught and treated early.

Will my child have to go to hospital every time he / she has a chest infection?
Most chest infections can be treated at home, but if the infection is severe or persistent, your child may need intensive treatment, including ***intravenous*** antibiotics. If so, your child may need to be admitted to hospital. In some cases children are even able to finalize their intravenous antibiotics at home.

Physiotherapy

Regular physiotherapy and physical exercise help to clear the sticky mucus from the lungs of someone with CF.

What is chest physiotherapy?
Chest physiotherapy is a process of clearing the sticky, excess mucus from the lungs, which can be done passively by a second person (e.g. "clapping" or "percussions"); or independently using different types of breathing exercises.

There are different ways that chest physiotherapy can be given to your baby or child. It is important that you learn the correct technique and your CF physiotherapist will advise and teach you the best methods for your child.

Do not be hesitant or afraid to ask the physiotherapist to watch you doing it from time to time, to make sure that you are still doing it as effectively as possible for your child.

In some cases, the doctor may also recommend that your child nebulize with 2-3ml of either 0.9 % (isotonic) or 3-5% (hypertonic) sterile saline solution, before each physiotherapy session, so as to assist in the loosening, mobilisation and clearance of the sticky mucus from your child's lungs. Some children may be prescribed with an additional inhaled medication, Pulmozyme, which also loosens sticky mucus.

Your CF doctor and physiotherapist will be able to answer all the questions you may have regarding nebulizing. They will advise you with regard to which nebulisers and compressors are recommended, as well as teach you how and when to use, clean and sterilize your child's nebulizer.

When should I start to do physiotherapy?
You will be taught to do physiotherapy soon after your baby or child is diagnosed as having Cystic Fibrosis. Your physiotherapist will advise you on when and how to do it.

When should I do physiotherapy and for how long?
It is very important that you learn the correct form of physiotherapy soon after the diagnosis of Cystic Fibrosis. How well your child is at any given time will determine how often you need to give your child physiotherapy.

Physiotherapy is normally carried out routinely twice a day and more often when your child is unwell with a chest infection.

It is advisable to schedule and perform physiotherapy for about 10 -15 minutes at a time, **before** a feed or your child having a snack or meal.

Both the duration and the frequency of the physiotherapy sessions may be increased if your child is unwell. Your physiotherapist at your CF Clinic will be able to advise you in this regard.

Who should do the physiotherapy?
To begin with, those adults who care for your child on a daily basis should to the physiotherapy – usually it is the parents.

However, later on other relatives or friends can also learn how to do it, so that no single person becomes indispensable to your child, furthermore enabling the person who normally does the physiotherapy, to also have a break from time to time.

Breathing exercises can be introduced in the form of a game from the age of two and three, and as your child gets older, he / she will learn to do his or her own physiotherapy.

From about the age of nine, most children can start doing some physiotherapy themselves without help from other family members. Most teenagers become completely independent and only require help from time to time.

Will physiotherapy hurt my child?
When physiotherapy is done correctly and effectively, it does not hurt - although small children may try to avoid it by complaining that “it hurts”.

Do we need special equipment?
There are lots of different physiotherapy techniques. Most do not require special equipment at all.

Your CF physiotherapist will advise you which is the best method of chest physiotherapy for your child and whether any special equipment (big exercise ball / trampoline / airway clearance devices like flutter or PEP valves) is needed or can be helpful.

Will my child be able to run, play swim and exercise like other children?
Regular exercise is an important part of care for children (and adults) with Cystic Fibrosis.

It helps slow down the deterioration of the lungs, improves the physical strength of your child, and is very good for keeping bones healthy.

Toddlers often like running, jumping and trampolining, all of which are very good for them.

When at school, children with CF should be encouraged to take part in PE, games and sports just like other children in their class.

It is advisable that you encourage them to do plenty of physical exercise outside of school as well - cycling, football, swimming, tennis, gymnastics and anything that they show an interest in.

Please note, however, that swimming in hot springs and heated swimming pools is generally advised against, as this may increase the likelihood of possible infection with ***Pseudomonas aeruginosa.***

It is often more fun to have company when exercising, therefore it helps to do some of these activities with the whole family and / or some friends.

Other problems in the chest

There are several other ways in which CF can affect the chest:

Asthma
About 40% of children with CF wheeze from time to time. This happens when the muscles surrounding the small airways contract and cause them to narrow slightly. They may find it more difficult to catch their breath and sometimes have a feeling of tightness in the chest.

Wheezing responds well to medicine used for children with asthma such as bronchodilators and steroids. They are usually inhaled from an inhaler device, which can be easily carried around (asthma pump).

Haemoptysis
Haemoptysis is the coughing up of blood and is very rare in children with Cystic Fibrosis. In adults with CF it is more common for streaks of blood to be in the sputum. It can be a sign of an infection and it is recommended that you see your CF doctor if this occurs.

If your child does have haemoptysis, you are advised to stop doing physiotherapy by means of clapping/percussions or using the flutter device/ PEP valves - until you have seen your CF doctor and/or CF physiotherapist. You can usually continue with gentle breathing exercises and “huffing” to clear secretions.

Can CF affect other parts of the body?

Yes, but it varies widely from person to person.

Ears, nose and sinuses
People with CF can be prone to ***sinusitis*** and hay fever, which may need to be treated with nasal sprays, sinus rinses, antihistamines and sometimes, antibiotics. Some older children and adults may develop nasal ***polyps,*** which if troublesome, may need to be removed by a small operation.

Liver
Some people with CF develop a kind of ***cirrhosis***. Older children and adults will have their liver function checked from time to time, as treatments is now available to prevent progression of any liver problems.

CF-related Diabetes Mellitus (CFRD)
This condition develops in about 30% of teenagers and adults with CF and results in an abnormally high level of sugar in the blood.

Early warning signs of diabetes in CF include unexplained weight loss, excessive thirst and increased passing of urine.

The recommended medical treatment for CFRD is insulin therapy.

Bones and Joints
Some older children develop a form of arthritis, usually in one or two of the large joints, such as the knee, ankle, shoulder, elbow or wrist. In most cases this improves with time and treatment.

Older adolescents and adults can also be prone to osteoporosis (thin, brittle bones).

As children born with CF are now treated more effectively and aggressively than in previous years, they are not expected to develop osteoporosis to the same degree as was previously the case.

Regular supplementation of Vitamin D and calcium may be recommended by your CF doctor / specialist so as to prevent the onset of osteoporosis.

Puberty
In a few children with CF, particularly those who are underweight, puberty may be later than usual, but they will still develop normally over time.

Fertility
Fertility, or the ability to have children, is usually normal or only slightly reduced in women, but most men with CF are usually infertile.

This means that their sexual function is entirely normal but they can seldom father children naturally. However, recent advances in in-vitro fertilisation and aspiration of sperm have allowed some men with CF to father children with clinical assistance.

Urinary Incontinence
Urinary incontinence is common in the female CF population and onset has been reported as young as 9-11 years. Some males also show mild symptoms of urinary incontinence.

Coughing, sneezing and laughing are the main causes of UI, and this may affect both airway clearance and daily life.

Your CF doctor / clinic team will teach pelvic floor exercises and controlled coughing to all pre-pubertal girls, and where necessary, to young boys as well.

Psychological issues
A life-long condition such as CF, which requires daily treatment, frequent clinic attendance and some hospitalizations, definitely has some emotional consequences – for both the child with CF, and his / her family.

Most patients and families cope reasonably well but may need help in anticipation of, and at crucial times - such as at diagnosis, admission to hospital, starting school, adolescence, and other times of heightened difficulty.

Most CF clinics should have specialised social workers or psychologists who can help to relieve stress and enable families to cope more effectively, by providing a range of services - including counselling and emotional support.

The healthcare team

When should I take my child to the doctor?
The answer is quite simple – whenever you are worried about your child's health.

Here are some useful pointers:
- Cold symptoms
- Fever
- Increased or frequent cough
- Increased sputum (phlegm)
- Change in colour of sputum
- Breathlessness
- Tummy aches
- Frequent or loose stools
- Vomiting
- Weight loss
- Decreased or poor appetite
- Increased fatigue
- Decreased ability or unwillingness to exercise

To which doctor can I take my child?
Children and adults with CF in South Africa have access to treatment at various CF clinics, either in specialised State Hospitals or Private Practices.

As Cystic Fibrosis is a complex condition, a team approach is followed at the CF clinics. Clinic visits, which are recommended every 1- 4 months, include regular consultations with doctors specialising in CF (e.g. ***paediatricians*** or ***pulmonologists***), physiotherapists, dieticians, clinic sisters, pharmacists, social workers, psychologists and also parent support groups.

Some CF patients will, however, be living far from a CF clinic or a CF Specialist, and will thus find it difficult to attend CF clinics. These children and adults can be cared for by their general practitioners (GP)'s.

Even though most GP's do not have much experience with CF, these GP's can align themselves with the nearest CF clinic and send their patients with CF at least yearly for assessment at the CF clinic ("shared care").

The CF clinic, on their side, can send a summary of the CF Clinic visit report to the GP – extending the team approach to include the local doctor - who in turn can phone the CF Clinic for advice and referral of his CF patient when necessary.

Where can I find or contact a CF Clinic?
A schedule of all the specialised state hospitals as well as the private practice clinics, with the respective doctors' names and updated contact information, is available on SACFA's website: http://www.sacfa.org.za

Will complementary medicine / alternative therapy help my child?
There is normally no major objection to using any alternative therapy - provided it **DOES NOT** counteract, contradict or distract from the regular, conventional accepted therapies that your CF doctor / clinic recommends.

It is essential for the future health of your child that the conventional treatments that your CF doctor / clinic recommend are adhered to in the prescribed way.

As long as the alternative therapies **DO NOT REPLACE** conventional therapies, the addition of complementary treatments should do no harm and some families have reported positive results.

Please consult your CF doctor / clinic team before starting any alternative therapy or complementary medicines.

Some other questions parents have asked

What should I tell other people about my child's CF?
Only you can decide, but you may wish to discuss it with your CF doctor / clinic team and with other people with relevant experience. You are also more than welcome to contact the South African Cystic Fibrosis Association (SACFA) in this regard.

Unfortunately, some people may make unintentional, but hurtful, comments about your child's condition, usually out of ignorance, so it is best to be prepared. These ignorant comments may include: "He is quite small and skinny, isn't he?", "She hasn't grown very much, has she?", "Imagine taking out a child with a cough like that", "Imagine giving a child medicine (enzymes) like that in public."

Try to take such remarks in your stride and, if possible or necessary, be prepared with explanations. People may also be ill informed or years out of date in their understanding of Cystic Fibrosis. They are usually willing to understand and are, if anything, over-sympathetic when the situation is explained to them.

You may also be faced with the decision on how and when to tell your other children. This may be difficult for you to do. In general it is better not to be secretive. An honest, simple explanation is very helpful, as it will set the tone for how siblings will respond and react to your child with CF, for many years to come.

Is there something that Cystic Fibrosis is NOT:

Cystic Fibrosis is NOT contagious. Due to the frequent coughing by children with CF, it is often thought to be contagious. As it is an inherited disorder with which children are born, your child cannot "give" CF to anyone.

Cystic Fibrosis is NOT caused by anything the parents did. Parents often feel responsible for everything that happens to their children. Nothing that parents do before or during pregnancy can cause / not cause CF to happen. It really is "all in the genes."

Cystic Fibrosis does NOT impair intellectual ability. People sometimes confuse CF with cerebral palsy (CP). Cystic Fibrosis does not affect the brain or the nervous system at all, and as such does not lead to intellectual impairment.

Cystic Fibrosis is currently NOT curable. CF is however highly treatable and the prognosis is constantly improving.

School years

As Cystic Fibrosis does not affect intelligence, children with CF attend playgroups and other preschool in just the same way as other children. Most children with CF attend normal schools and join in all the normal activities.

It is important though, that the teachers and other staff in the school know that your child has been diagnosed with CF, as well as what the condition of Cystic Fibrosis entails.

The details of your child's particular treatment though will best come from yourself. You know your child, so talk to the school before term begins and keep them informed of new developments or changes in treatment.

Teenagers and leaving school

Adolescence is a challenging period for anyone. It is especially so for young people with CF, particularly if the condition causes them to mature later than their peers.

As children get older, most will also want to know more about their condition, and how it will affect them in future.

As the child with CF reaches adolescence, and then adulthood, important life decisions need to be made with regard to both the short and long term.

Nevertheless, the full range of further education and employment opportunities should be available to any young adult with CF, depending on their intellectual and physical abilities. No field should be totally closed to the patient, but consideration should be given to the amount and intensity of manual work, cleanliness in the work environment, exposure to extremes of temperature, dust, smoke or fumes, and the ability of the individual to cope with the lifestyle demanded of his/her chosen profession.

There are some occupations which are less suitable, of course, but these considerations are for the future and decisions can be made at the time with help from the CF doctor / clinic team.

The future

Only thirty years ago, the outlook for a baby born with CF was very poor. Today, the outlook for an individual with CF has improved dramatically – and the outlook continues to improve year by year.

Young adults with CF are now living into their thirties, forties and beyond - leading active and fulfilling lives.

There have been major and rapid advances in the treatment of CF in recent years. As such, many of the effects or consequences of Cystic Fibrosis, previously thought to be inevitable, can be prevented, delayed or improved by effective and intensive treatment.

It is likely that the quality and length of life for individuals with CF will continue to improve as a result of
patient compliance with prescribed and recommended therapies,
continuous new research, and
the development of new treatment, medicines and therapies.

The future is without a doubt an optimistic one, but – until a cure is found – we must aim to keep every child and adult with CF as fit and as well as possible by preventing and treating infections as well as controlling symptoms, so as to maximise both the quality and the length of their lives.

Glossary

Alveolus (alveoli)
The specialised part of the lung where oxygen enters the blood and carbon dioxide can leave the blood.

Bronchus (Bronchi)
The small airways in the lung.

Cirrhosis
A term that is used in a general sense to mean progressive fibrous tissue overgrowth in an organ.

Cyst
A fluid or air-filled space. In CF these are usually in the pancreas or the lung.

DIOS
Distal Intestinal Obstruction Syndrome. A blockage of the gut that occurs in older children and adult with CF.

Enteric Coated
Covered with a coating which protects against acid in the stomach. This is useful for pancreatin.

Enzyme
A chemical that effects change in some way. In CF this usually refers to digestives enzymes which digests food so that it can be absorbed and used by the body.

Fibrosis
Fibrous tissues are useless "gristle" which replaces normal tissues when they are damaged. In CF this occurs in the lungs and the pancreas.

Gene / Genetic
Every cell has thousands of genes that are made up of DNA and are passed on from parent to child. Genes are responsible for a person's overall health and functioning, as well as for individual characteristics such as eye colour and blood group. Faulty / mutated genes can cause certain genetic conditions such as Cystic Fibrosis.

Gene Mutation
A change in a gene or genes.

Genetics
The study of genes, heredity, and variation in living organisms.

Genetic Testing
The method of detecting certain genes, for example tests can determine when a person carries the gene for Cystic Fibrosis.

Haemophilus influenza
A virus which is a common cause of respiratory infection in Cystic Fibrosis.

Haemoptysis
The coughing up of blood.

Health care team
Doctors, nurses, physiotherapists, dieticians, social workers, psychologists and pharmacists.

Intravenous
Sometimes antibiotics or other medicines are given directly into the vein rather than by mouth. To make it easier, a small needle and plastic cannula (tube) can be left in the vein so the drug can be put through it rather than a fresh injection every time. A cream known as "emla" is used to numb the skin before the needle and cannula is put into the vein.

Meconium Ileus
An obstruction of the small intestine at birth.

MIE
Meconium Ileus Equivalent. See DIOS above.

Microspheres
Enzyme granules contained within a pancreatin capsule.

Mucoviscidosis
Another name for CF. It literally means that the mucus is thick and sticky or viscouos.

Mucus
An essential fluid secreted by mucus membranes. Mucus lubricates and protects parts of the body, particularly the lungs and digestive system.

Nebuliser (Neb for short)
A small machine which converts liquid medication to a fine mist that can be breathed in, so as to work directly in the lungs.

Paediatrician
A doctor who specialises in the treatment of children.

Pancreas
A gland which lies behind the stomach and makes digestive juices or enzymes, as well as insulin.

Pancreatin
An extract of animal pancreas, and the general name for all pancreatic enzymes.

Physiotherapy
Part of the treatment for Cystic Fibrosis, which entails clearing sticky mucus from the lungs.

PNA or NPA
Per Nasal Aspirate or Naso Pharyngeal Aspirate – a special way of getting a sample of sputum from a child too young to cough it up.

Polyps
A small growth of mucous membrane that can grow on the lining of the nose.

Pulmonologist
A doctor who specialises in the treatment of lungs.

Pseudomonas aeruginosa
A bacterial infection that affects the lungs.

Sinusitis
Inflammation of the membrane lining the facial sinuses (the air-filled cavities in the bones surrounding the nose).

Sputum
Mucus material produced by the cells lining the respiratory tract.

Staphylococcus aureus
A bacterial infection that affects the lungs.

Sweat Test
The test used to diagnose Cystic Fibrosis.