

**A guide for parents and caregivers**  
of children diagnosed with cystic fibrosis





## About this guide

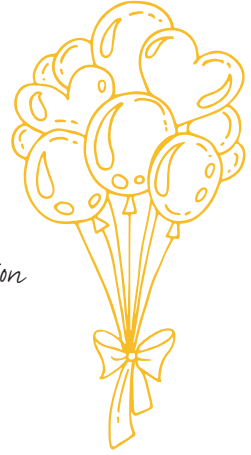
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This guide is for parents and caregivers whose child has been diagnosed with cystic fibrosis (CF), to help you and your family understand CF and learn how you can help your child grow and thrive.

Most of the information focuses on supporting you through your child's first year following diagnosis as a newborn and throughout the booklet, children and their parents share their stories about living with CF and tips for daily life.

Although there is no cure for CF, advances in our understanding of CF, improved treatments and new medications have significantly improved the quality of life and life-expectancy of people with CF. With ongoing research, there is every reason to be hopeful of ultimately finding a cure.

This guide has been developed together with 'A guide to cystic fibrosis for family, whānau and friends' that extended family, whānau members and friends can read to help support you and your family.



*Dear Parents*

*It's impossible to immediately absorb all the information in a guide like this when you've just received the life-changing news that your child has cystic fibrosis.*

*So, I would like to open it with a message of hope.*

*Every year, people with CF are living longer, getting jobs, going to university, playing sport, getting married and having kids of their own. Your child has a major illness, but there is a lot you as parents can do to slow down the disease and keep your child as healthy as possible.*

*There are great teams of health professionals around the country who will work to help your child too, and there is a great CF community (including CFNZ who put together this guide) who can provide support, with many of them having been through the same thing you are going through now.*

*As you look after your child and as you work your way through this guide, things can feel pretty overwhelming at times - remember to look after yourself too and take things one step at a time.*

*But most of all, know there is hope and there are things you can do to help your child.*

*Stephen*

*Father of a 6-year-old with CF.*

## Finding out your child has CF

**Receiving a diagnosis of cystic fibrosis for your baby has probably come as a shock to you and your family, and you may feel overwhelmed with how much there is to learn and the impact CF will have on your child's life.**

You are not alone. You may have already met some members of the healthcare team who will help care for your child, and as your child grows, you may find support from other families with children who have CF helpful.

As a parent, you are part of your child's healthcare team and an important partner and decision maker in deciding the best care for your child. With time and practice, you will develop the necessary skills to manage CF as part of everyday life.

## How are you feeling?

**Many parents describe the time following their child's diagnosis as a rollercoaster of emotions.**

Learning about CF and the impact it has on your child and your family often leaves little time to think about your own health and mental wellbeing.

But how you're feeling, and how you're coping with your child's diagnosis is important. Every parent deals with their child's diagnosis in their own way, and there is no right or wrong way to feel. Having a child with a chronic condition like CF is hard emotionally and 'staying positive' is not always easy to do. Parents, especially mothers, often feel if they're not doing everything, they're not doing it right. Seeing yourself as a key person in your child's healthcare team, but not it's only member, can be helpful.

*"In the early days, when CF was pretty new to us, we lived near a forest. We'd often throw the kids in the buggy and go for a long walk. The kids were out in the fresh air and we'd do a lot of talking. We used to do that three or four times a week. It was good to be out of the house and we used to talk a lot."*

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*"One of the things I wish I'd done earlier was to take my own time-out - to literally step away from the situation for a period of time on a regular basis. Just taking a break from everything for a day or two really helped put things into perspective. It gave me a chance to think about how I was managing everything and what I could do differently."*

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*"I think it's important to find something to do that's purely just for you - reading a book, going for a walk, going to the movies, or catching up with friends."*

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*"It's amazing how talking things over with someone can make a huge difference to how you feel, even if nothing actually changes. Having someone you feel you can unload to is really important."*

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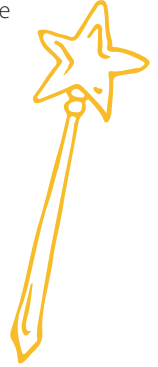
*"Talking to a professional has been really helpful. I can go into their office, dump all my emotions on them and they're paid to be quiet and listen. When I've finished I can turn around and walk out and I don't owe them anything like I do with friends."*

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It's important to acknowledge your feelings, and we hope with support from a range of people, including health professionals if needed, you look at your child's future with CF as one of hope and possibility.

Look after yourself. It's easy to put your own needs last when caring for other people, but it's important to find ways to take some regular time out, away from the demands of CF.

Some parents find support from their family and friends enough, while other parents need support from health professionals, including counsellors or psychologists. If you're finding it hard to cope, please reach out for advice and support. Talking with your CFNZ fieldworker, CF nurse or GP is a good place to start – they can make sure you're put in touch with the right people to help.



## Cystic Fibrosis New Zealand – here to help

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Established in 1968 as a volunteer support group for parents with a newly diagnosed child, we dedicate ourselves to shaping a brighter future for everyone with CF.

As people with CF live longer and have a better quality of life, there are many life transitions, milestones and challenges to manage along the way. We offer personalised support, whether it is emotional guidance, practical advice, or financial assistance, to individuals and their families throughout their changing journeys.

Our team of fieldworkers visit and support families, provide information packs, cover the costs of essential medical equipment, hospital allowances, welfare assistance and other means of support. We also fund CF research and advocate on behalf of the community for better access to services and care.

As well as our national office, based near Starship Hospital in Auckland, we have regional branches that are run by CF families for CF families. The branches provide invaluable friendship, advice and a caring support network.



## Support from health professionals

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A multidisciplinary healthcare team, including doctors, nurses, physiotherapists, social workers and dietitians, work together to provide the best care for your child. CFNZ also has fieldworkers working to support you and your family.

Knowing what role each person has, and who to call for advice, can be overwhelming when your child is first diagnosed. But as you meet the team you will understand their roles and how they can help you care for your child.

## Key health professionals involved in your child's care



Your child's CF nurse specialist or clinic nurse is:

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Your child's CF consultant is:

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Your child's CF physiotherapist is:

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Your child's CF dietitian is:

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Your child's CF fieldworker is:

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Your local hospital is:

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Your local CFNZ branch is:

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If I am worried about my child I can call:

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their number is:

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## What is cystic fibrosis?

**Cystic fibrosis (CF) is a genetically inherited condition some babies are born with that can affect many of their organs, but mainly their lungs and digestive system.**

**It is caused by a faulty gene passed down from both parents and is usually diagnosed soon after birth.**

Without CF, our lungs and digestive system make mucus that is thin and slippery and works as a lubricant to keep the insides of our bodies working well and helps protect us from infections.

If we have CF, the fluid lining our lungs and digestive system is reduced, causing our mucus to become thick and sticky and block the tiny tubes of many of our organs.

In the lungs, the sticky mucus is difficult to cough up and certain bacteria can become trapped under the mucus, causing inflammation and infection.

In our digestive system, the pancreas is affected the most. Without CF, our pancreas makes enzymes that help digest food in our small intestine, by breaking down fat, carbohydrates and protein and turning it into the energy we need.

If we have CF, the thick, sticky mucus within our pancreas can block the tiny tubes and prevent the flow of enzymes into our small intestine. Without these enzymes, nutrients can't be absorbed from food (in babies, breastmilk or formula), making it harder for babies with CF to gain weight.

CF does not affect your child's developing brain and so does not affect your child's intelligence.

## Why does my child have CF?

**All people have two copies of a gene called CFTR in their cells, one copy inherited from their mother and one copy from their father. CF is caused by a fault in both copies of this gene. It is a condition that occurred when your baby was conceived, when they received a set of genes from both parents, with each set carrying an abnormal CFTR gene.**

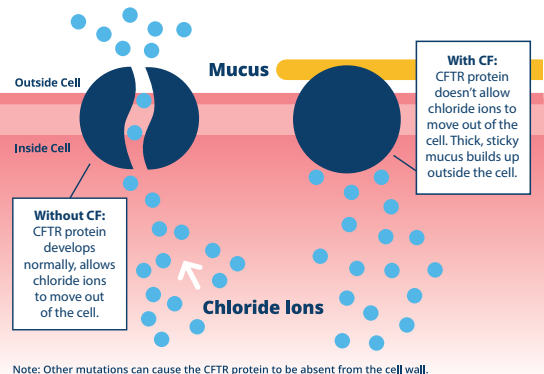
Having just one abnormal gene (which is the case for parents of children with CF) means you do not have CF or any CF-related symptoms, but because your child has two, they have CF. This is called autosomal recessive inheritance.

Genes are what makes us who we are – they affect our eye colour, hair colour, how tall we are and many of the things that make us individuals.

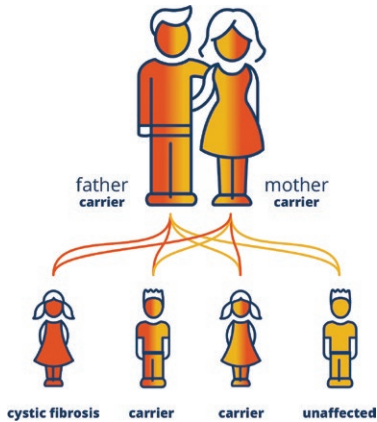
Genes also affect how our body work on the inside, and changes in the cystic fibrosis transmembrane conductance regulator (CFTR) gene cause CF.

## The CFTR gene

CF is caused by a mutation in the gene that produces the CFTR protein. Proteins are the building blocks of our body, and the CFTR protein controls the flow of salt



and fluid in and out of our cells in different parts of our body. Mutations in the CFTR gene cause the CFTR protein to not work properly, or not be made at all, which causes mucus to become thick and sticky.



Each time two people who carry the CFTR gene mutation have a child together there is a:

- **25% (1 in 4) chance**  
their child will have CF
- **50% (1 in 2) chance**  
their child will be a carrier but not have CF
- **25% (1 in 4) chance**  
their child will not be a carrier and will not have CF.

About 2000 mutations of the CF gene have been identified, each with its own name and numbers and most children with CF in New Zealand can have their mutations identified.

As a parent of a child with CF, you will be offered an appointment with genetic services and counsellors, who can discuss, and answer questions, about future pregnancies. They can also organise CF screening for extended family members if wanted.

## How was my child diagnosed with CF?

**Most babies in New Zealand are tested for CF through the Newborn Metabolic Screening Programme, often referred to as the Guthrie Heel Prick test. This test, usually carried out about 48 hours after birth, uses a blood sample taken from your baby's heel and screens it for rare disorders, such as CF.**

If this test shows your baby has a high level of an enzyme called immunoreactive trypsinogen (IRT), which is elevated in most babies with CF, a second test is done using the same heel prick sample to look for common mutations of the CF gene in New Zealand. If this test shows a CF gene, this is called a positive screen, meaning a baby is either a carrier (has one CF gene) or has CF (has two CF genes).

Your lead maternity carer is notified of the results of the Guthrie Heel Prick test and will refer your baby to a local paediatrician who will organise further tests and an appointment to see you and your baby.

## Further tests to confirm your baby's diagnosis

### Sweat test

A sweat test measures the amount of salt (sodium and chloride) in your baby's sweat and is usually done when they weigh over 3 kilograms. An abnormally high chloride result indicates CF. Occasionally, results are inconclusive, or not enough sweat is collected, so the test needs to be repeated when your baby is heavier or older.

### Stool (poo) test

Your baby's poo will be tested for an enzyme called elastase – a low level is common in babies with CF.



## Blood test

Your baby will have another blood test to ensure a correct diagnosis and to identify their CF gene mutation. Sometimes the gene mutations are not found immediately so the diagnosis can take a bit longer.

Both parents will also need to have a blood test.

Usually a local paediatrician will review the results from all your child's tests and confirm their diagnosis of CF.

The order of which the further tests are done varies around New Zealand.

For a small number of babies, the first symptom of CF is a blocked intestine at birth, called meconium ileus. Meconium is the normal black sticky secretion from a baby's bowel, usually passed in the first 24 hours after birth. In some babies with CF, the meconium is too thick to pass and blocks the intestine, needing bowel washouts or surgery to remove the blockage.

Occasionally, other factors such as siblings with CF or early symptoms such as breathing difficulties, malabsorption or poor weight gain may have led your baby to be tested for CF.

Children born in a country without newborn screening may not be diagnosed until they are older.



*"Our daughter was extremely slow to put on weight and was dropping through the lines on the growth chart despite huge amounts of feeding. It was a massive shock when we got the Guthrie test results and worse when we got the final diagnosis, but now I'm so grateful we received the diagnosis early and could start treatments which have made her into the healthy girl she is today."*

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*"When my baby was two weeks old, my midwife told me he'd had an abnormal result for CF on the newborn screening test. Initially, I focussed on what immediately needed to happen - how to get to Starship, what a sweat test was, etc."*

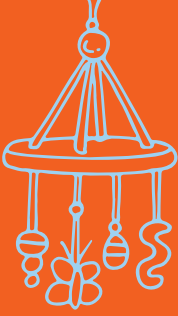
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*"I felt indignation that my baby would have to go through a battery of tests because the heel prick test was wrong (my midwife said my husband and I would know if we were CF carriers). Slowly, as I learnt more information (CF testing didn't start in NZ until 1981 so both my husband and I didn't know we were carriers as we were never tested), I felt scared, overwhelmed and full of hope my son wouldn't have CF and full of fear he would."*

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*"The stool sample came back with good news - his pancreas worked. The blood tests came back next with bad news - my son had two mutated CFTR genes and my husband and I were both carriers of CF... There was still an outside chance he didn't have CF as one of the gene mutations was rare and there was some uncertainty around it, or at least that's what we were told and hung onto."*

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"The doctor said he was 99.9% sure my son had CF but somehow we heard he was only 90% sure and we held onto that misguided 10% of hope for a few weeks. After an unsuccessful sweat test at two weeks of age, they finally got enough sweat for the sweat test when he was eight weeks old, which conclusively confirmed CF. We spent six weeks in limbo on the diagnosis and I oscillated between hope, fear and despair."

"I felt angry at people who commented 'you wouldn't know he's got CF, he looks so well'. Going to the children's hospital each week with my baby had not been my plan - I wanted to go to coffee groups and Rhythm & Rhyme each week, not see doctors. I felt a sadness about what was, and what it meant for the future - both for my son but also for me, and what it meant for my life as a mother."

"I felt disconnected and jealous of other mothers from my ante-natal class whose biggest concerns seemed to be whether their baby had reflux or not."

"I found I was stronger than I thought I would have been before I had my son about such news. I felt overwhelmed at all the care that needed to happen and unable to imagine how the diagnosis and the treatments would become 'normal'."

"I didn't even know what cystic fibrosis was when my midwife told me my son had returned an abnormal result for it on the heel prick test when he was two weeks. It was also the first day my husband went back to work!"

"I still wanted her to be a child. I don't want CF to define us. Once the diagnosis came through we were like, 'she's a child that has a condition, but she's not bound by that condition.'"

"For a while, I wanted my son exactly as he was, just without the dodgy CF genes. But it was the combination of genes that made him as awesome as he is - and I wouldn't change that for the world. I'd rather have him with CF, than not have him."

"Having a child with CF has made me look at people in a different light - everyone has a story, you just can't always see it when you look at someone."





*"We were devastated by the news our daughter had CF. It's still hard to comprehend some days. But the hospital team sprang into action and started her first medication and within days she'd started really gaining weight and doing well. It was such a relief to know there were things we could do that had an impact on her health."*

## How will CF affect my child?

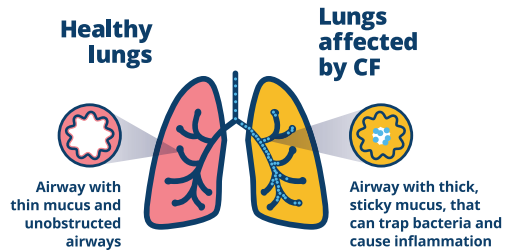
**Cystic fibrosis affects many of your child's organs but mostly their lungs and digestive system.**

### Lungs

When we breathe, air travels along our main breathing tube, called the trachea, into our left and right lungs via smaller tubes called the bronchi. These bronchi become even smaller tubes called bronchioles that end at tiny grape shaped air sacs, called alveoli.

In the alveoli, oxygen we have breathed in enters our blood stream and is used by our body, in exchange for carbon dioxide, which we breathe out.

With CF, mucus produced to keep your lungs working well becomes thick and sticky. This thick mucus can block the tiny tubes in your lungs, causing an obstruction, or trap bacteria on the airway lining. Mucus and bacteria can cause inflammation, infection and lung damage, however, this damage can be greatly reduced with good treatment.



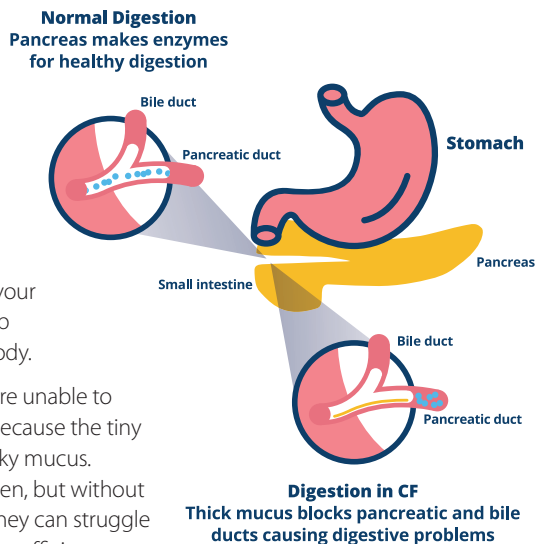
### Digestive system

Our digestive system is made up of many organs working together to digest and absorb food. The pancreas is an important organ of this system, and is usually affected by CF.

Your pancreas is found behind your stomach, and two of its jobs are to make hormones (chemicals that control many of your body's functions) and make enzymes to help break down food to be absorbed by your body.

With CF, usually these pancreatic enzymes are unable to flow out of the pancreas into the intestine because the tiny pancreatic tubes are blocked with thick, sticky mucus. A baby may be feeding well and feeding often, but without enzymes to absorb breast milk or formula, they can struggle to gain weight. This is known as pancreatic insufficiency.

If your baby is found to have pancreatic insufficiency they will need to take pancreatic enzymes with their feeds, and as they grow, with most of their food. A small number of babies with CF produce pancreatic enzymes, called pancreatic sufficiency. These babies don't need to take pancreatic enzymes but will be monitored closely as often their pancreas can stop producing enzymes.



# Taking care of your baby

Most CF treatments focus on keeping your baby's lungs free of inflammation and infections and helping their digestive system absorb nutrients from their feeds.

*"We were feeding every three hours for the first four weeks and our baby was only just gaining weight. The midwife didn't believe us that we were following her instructions on feeding. Starting on enzymes made an amazing difference - she suddenly stopped getting skinnier and started getting chubbier."*



## Healthy lungs

### Chest physiotherapy/airway clearance

Chest physiotherapy/airway clearance helps to keep your baby's lungs clear of thick, sticky mucus. A physiotherapist will teach you how to do chest physio and watch as you learn so you can be confident doing it at home.

At most clinic appointments while you are learning about CF, your child will be reviewed by a physiotherapist who will check your technique and answer any questions.

It's recommended all children with CF have chest physio/airway clearance twice a day for the rest of their lives, usually starting as soon as your child has been diagnosed.



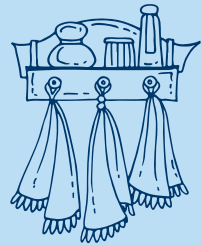
*"It took us, and our baby, a while to get used to doing chest physio. We found reading to our baby, distracting them with noisy toys or toys with flashing lights, listening to music or watching TV (Baby Einstein, the Wiggles, etc.) helpful."*

### Preventing infections

Chest infections are caused by 'bugs', including viruses and bacteria, that are usually harmless to people without CF, but can cause damage to your baby's lungs. Children with CF can also become more unwell from viruses and bacteria that only mildly affect a person without CF.

### General ways to help prevent infections

- Wash your hands regularly or use hand sanitiser.
- Ask family, friends and visitors to wash their hands before holding your baby.
- Ask visitors to only visit if they are feeling well, no coughs and colds (this applies to people visiting all babies, not just babies with CF!).
- Avoid smoking or exposing your child to second-hand smoke. Even if you smoke outside, studies have shown this still increases the risk of your child getting a chest infection.
- Have your baby immunised on time.
- Attend regular appointments with your CF team.
- Good nutrition is key. A dietitian is a part of your healthcare team and will work with you to make sure your baby is growing well.



*"I never wanted to be that parent who was concerned about bugs and worried about what my baby touched, etc. Before my son was born, I had totally bought into the theory that germs were good for immunity and we used too much disinfectant these days. I've had to adjust my thinking, which has been hard."*

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*"We have put a sign on our door asking people to leave their shoes outside, use hand sanitiser and not come in if they're sick - to save us having to remind people every time they come around!"*

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*"We've planted our pot plants outside and ironically they're thriving outside more than they ever did inside... we have hand sanitiser everywhere... every room, in the car, handbags, etc."*

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*"We had our young man at home for the first 12 months but wanted him to socialise and be more than just his CF, so did home-based childcare and eventually went into school. It's a balance. In winter, we hang out at parks and the zoo, versus visiting friends or inside places. It's a hard call as a mum, but I always worked on the fact I could bring home bugs from work, or we could get sneezed on in the supermarket."*

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Infection control, or infection risks, are terms used by health professionals when they talk about ways to minimise the chances of your child being exposed to potentially harmful bacteria and viruses. Infection risks specific to your child, and how you choose to manage them, is an area where advice and guidance from your CF team is best.

When your baby is first diagnosed, you might read or hear a lot about 'bugs' and the consequences for your baby's long-term lung health and a lot of it can sound quite scary. In time, you will find a balance between letting your child explore their environment and minimising infections risks.

Although it's not possible to avoid all infections, there are some ways you can help protect your baby.

### **Practical ways at home to help prevent infections:**

- Keep your home warm and dry and make sure it has good insulation.
- Use disposable cloths for cleaning.
- Throw away rotting onions – the black spots can be harmful to your baby's lungs.
- Only use bath toys which don't take in water, clean them regularly and dry them after use.
- Outside, think about areas where bugs may live – get rid of stagnant water, clean up drains and cover rotting vegetation.
- Keep your exploring baby away from pot plants, especially if dirt is warm and damp.

Bugs thrive in wet, warm places so a general rule of thumb around the home is to try and reduce damp areas, or areas that retain moisture. Stagnant water, compost, dust and dirt are prime places for bacteria to thrive in.



### Swimming

*Many children in New Zealand start swimming lessons from a young age, and children with CF should be encouraged to learn how to swim. There's no right or wrong age to start introducing your child to water but there is an increased infection risk with stagnant water around swimming pools and changing rooms to think about if you're keen to take your baby into a pool.*

*Domestic pools are not checked for adequate chlorination or salination in the way commercial pools are. You may want to discuss this with family or friends before your child uses their pool.*

*Swimming in the ocean is a good way to get your baby used to water but we recommend talking with your CF team for specific advice about public and private pools.*

Everything is a balance when you have a child with CF. They need to experience everyday life like other children and should be encouraged to try all new experiences. You can't place them in a 'bubble' and over-protect them, but you can be aware of their vulnerable lungs and help minimise the risk of exposure to bacteria and other harmful 'bugs'.

### Playgrounds

*If you've got older children, you probably know the benefits of an afternoon spent at a playground – for both kids and adults alike to be out in the fresh air.*

*Once your child is more mobile and keen to explore their surroundings, playing at a playground is encouraged, but there is an increased infection risk at playgrounds with wet or dry bark, stagnant water or decaying leaves.*

### Other children and adults with CF

Everyone with CF – including babies, children and adults – carry different 'bugs', or bacteria, in their lungs, that can potentially be passed on to other people with CF. We recommend your baby doesn't have any contact with other people who have CF.

### How do I know my baby has a chest infection?

As your baby grows, you will get to know what are normal baby breathing noises and what is abnormal or a sign they could be developing a chest infection. Starting treatments early, such as more chest physio and/or antibiotics can help prevent permanent lung damage.

With practice, you will be able to recognise the signs and symptoms of an infection early, such as:

- coughing – your baby may start coughing during the day, start coughing at night or their cough may sound different than normal
- increased mucus – it also may be a different colour, e.g. green instead of clear
- tiredness
- feeding less
- weight loss or failing to gain weight
- irritable
- fever
- changes in their breathing, such as breathing faster or harder.

### Who to call:

Your CF nurse specialist or clinic nurse at the hospital is the best person to call if you are concerned your baby is unwell. After hours or on the weekend, follow your GP clinic's after-hours procedures.



"I was pretty scared the whole first year and did keep our son in cotton wool, but you do eventually learn to let go bit by bit. I wiped toys from libraries down, didn't go to playgroups until he was about two years old. Now he's 15 and swims in rivers, has played in mud plenty of times, goes tramping and is planning to do all kinds of crazy things in life. I thought the first few years were important to try and get through without too much sickness and we did achieve that mostly. My thought was, the less infections as a kid, the better his lungs would be as he grew up. You have to do what feels right for you."

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"There is no 'right' answer and it is something you will have to work out for yourself. I find there are three groups of parents - newbies who are wading through all the information and are still in shock over the diagnosis and who want to stay as protective as possible. Then you have seasoned parents who are super relaxed and let their child do anything because they want their child to actually live their lives. And then you have folks like me with a foot in both camps. What one parent or caregiver is comfortable with might fire your anxiety and vice versa. There is so much to learn and accept on this journey, just do the best you can."

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"For me, it's putting things in categories; small, medium and big risk. Big risk really isn't worth it if there's no big gain. Medium risk - if he's going to get something out of it that will have a positive outcome on his life, go for it but be as careful as you can. Small risk is go for it with precautions at the ready."

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"We bought bottles of hand sanitiser and kept them by the door so everyone who came in cleaned their hands. Some people look at you as if you have two heads when you ask them, but it's so important when they're so little."

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"If you have a damp house it's worth getting a dehumidifier, and air purifiers are good too. We keep an air purifier with HEPA filter in our daughter's bedroom and if she's coughed quite a bit overnight, I'll change her bed linen. I usually buy the asthma approved pillows too and replace them once a year."

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"If I've been given flowers I do accept them (somewhat begrudgingly!) and I'll make sure to change the water every day. As soon as they begin to wilt I get rid of them."

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## Healthy digestive system

**Good nutrition is especially important for people with CF, for good growth, good health and to help fight infections.**

### Pancreatic enzymes

About 85–90% of people with CF are pancreatic insufficient, meaning they need to take synthetic pancreatic enzymes with food. The small percentage of people whose pancreases still function, called pancreatic sufficient, don't need to take synthetic pancreatic enzymes.

In New Zealand, Creon is a brand name for pancreatic enzymes that is commonly available and prescribed.

### Giving your baby enzymes

Pancreatic enzyme capsules contain granules that help your baby digest and absorb their feeds and gain weight. The special coating means the enzymes aren't dissolved in their stomach but are absorbed in their small intestines.

The granules in the capsules are measured out onto a teaspoon, mixed with an acidic fruit puree such as apple or pear and given with each breast or formula feed.



After giving your baby enzymes, check to make sure there are no granules left in their mouth as these can cause irritation to their mouth and chin.

Pancreatic enzymes are prescribed by your doctor or a CF dietitian. Your dietitian or CF nurse will talk with you about how many enzymes your baby needs and show you how to open the capsules and separate the granules.

Giving your baby the right number of enzymes is initially a bit of trial and error, but over time you will become confident.

If your baby has any of the following signs it may mean they aren't having enough enzymes with their feeds:

- tummy pain
- changes in their poo e.g. increase in volume, frequency or smell
- feeding more
- difficult to settle
- increase in wind
- losing weight or slow to gain weight.

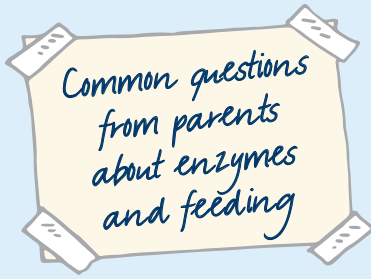
### Who to call:

Call your CF nurse specialist, clinic nurse or dietitian about any changes to the number of enzymes you're giving or if you're having any issues with feeds or with giving enzymes.

### Vitamin supplements

Babies with CF don't absorb some vitamins (vitamin A, D, E and K) very well, so your baby may need daily vitamin drops. These vitamin drops are different than children's multivitamins you find in a chemist and are prescribed by your doctor or CF dietitian.

*"Once we were ready to start leaving the house for something other than CF appointments (which took a long time!) we just got used to taking apple puree, a spoon and enzymes wherever we went."*



*Common questions  
from parents  
about enzymes  
and feeding*

? *Should my baby be breast or formula fed?*

Just like babies without CF, breast milk or standard infant formula is perfect for your growing baby.

? *What do I do if my baby isn't gaining weight?*

Your baby will be weighed regularly at CF clinic appointments and your CF dietitian will offer advice if your baby is having trouble feeding or they're slow to gain weight. Occasionally, babies with CF need a special formula or special energy supplement if they're struggling to gain weight.

? *Do I give my baby more pancreatic enzymes if they have a top-up feed soon after a full feed?*

Pancreatic enzymes work for about 30 minutes, so you should feed your baby as soon as they are given. If your baby takes a while to feed, try giving them half their enzymes at the start of their feed and the rest half way through.

If your baby has a top-up feed 1–2 hours after their usual feed, they will need more enzymes.

Your dietitian will give you advice if you baby is feeding frequently and you're unsure when you should give more enzymes. It's important not to exceed your dietitian's recommendations regarding the maximum number of enzymes your baby needs per day.

? *What if my baby doesn't feed after I've given them their enzymes?*

Occasionally it is not harmful to your baby if they are given enzymes and then don't feed.

? *What if I forget to give my baby their enzymes before I feed them?*

If enzymes are missed at the start of a feed, you can give them part way through the feed, or at the end. Forgetting a single dose isn't harmful, although your child may have tummy pains or loose poos afterwards. If enzymes are regularly missed, their growth will be affected.

? *Can I only use apple puree to give enzymes?*

Pancreatic enzymes need to be sprinkled on something acidic, so apple or pear puree is recommended.

? *It doesn't feel right giving my baby puree from such a young age. Is it really okay?*

Giving enzymes is important for babies with pancreatic insufficiency to gain weight and thrive. They shouldn't be given in breastmilk or formula so think of apple puree as a carrier for the enzymes to be absorbed in the right place in the intestines. Babies soon get used to taking the puree.

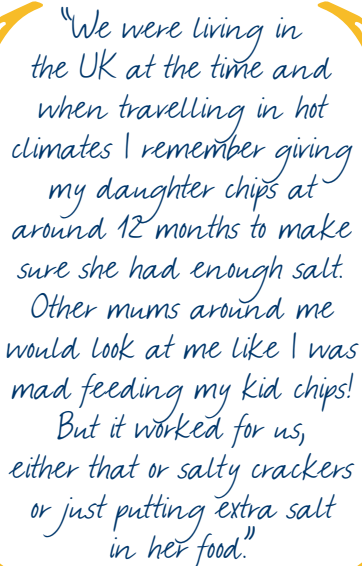


## Extra salt

Babies with CF lose more salt in their sweat. This means they can become dehydrated faster than babies without CF, especially on hot, humid days.

Most babies with CF need extra salt – if your baby needs to start on a salt supplement, your CF dietitian or nurse will show you what to do. Like pancreatic enzymes, salt is given with apple puree, usually with feeds.

Your baby will have their salt levels monitored periodically, which is easily done by collecting and testing a sample of your baby's urine (wee) and you will be advised if your baby needs extra salt and how to give it.



*"We were living in the UK at the time and when travelling in hot climates I remember giving my daughter chips at around 12 months to make sure she had enough salt. Other mums around me would look at me like I was mad feeding my kid chips! But it worked for us, either that or salty crackers or just putting extra salt in her food."*

## How do I know my baby is dehydrated?

Signs your baby is dehydrated include:

- *fewer wet nappies*
- *dry lips*
- *sunken eyes*
- *lethargic*
- *breathing fast.*

## Who to call

See your GP or take your baby to your local hospital if your child has any of the above symptoms.

Giving your baby pancreatic enzymes, vitamins and salt supplements may seem overwhelming at first and it's hard learning how to care for your baby on top of broken sleep and learning about cystic fibrosis.

With the support and advice from your CF team, you will soon become more confident in caring for your baby, but please remember, the team is there to help, so get in touch with them if you have any questions or if you need more support.

## Moving on to solids

When your baby is ready to move on to solids, usually between 4–6 months, the number of pancreatic enzymes they need will change. Your dietitian will work with you as you learn more about enzyme doses and foods containing fat.

Just like children without CF, your baby will progress from eating pureed to mashed to finger food and family foods. They will likely need more calories than other children to grow well and this is usually achieved with a balanced diet but including more high fat, high salt foods. Your dietitian and CF nurse will work closely with you to help you manage their enzymes and diet – you won't be alone in this exciting, but messy, milestone!

# Clinic appointments and hospital admissions

Initially, you and your baby will be seen frequently at the hospital clinic, often weekly or fortnightly.

Regular appointments help you and your family learn about CF and it's a great opportunity to have your questions answered. Your CF team will make sure your baby is gaining weight and thriving, help you learn chest physiotherapy/airway clearance, discuss signs and symptoms of a chest infection to look out for and make sure you're gaining confidence giving pancreatic enzymes and coping with the demands of caring for your child.

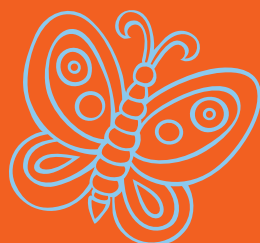
*"One of the cool things is so much of CF care is done at home, and it's great to be able to look after your baby and do physio and things that make such a big difference to their health. And even though sometimes it's really hard and you don't want to do it, you think of all the people who have to go to hospital for everything, and I can do it for her."*

It may be helpful to jot down questions as you think of them, and bring them with you to your next appointment.

After a few months, and if your baby stays well, clinic visits will become less frequent – your baby may need to see the CF team once a month, or sometimes every few months.

As your child grows they will usually have clinic appointments every three months.

Remember: even if you haven't seen your CF team for a while, they are always there if you need them. Your CF nurse is usually just a phone call away – please call them if you need to.





*"My son was admitted to hospital at three months old with a lung infection. I was very apprehensive about being admitted, and felt overwhelmed about having to go in. However, it wasn't nearly as bad as I imagined. I learnt a lot about CF during our week stay in hospital and how to better care for my son, I got to know members of my son's CF team better - there was more time for longer conversations than at clinic appointments, and my physio technique and confidence improved dramatically with daily assistance from the physios."*

## Hospital admissions

If your child becomes unwell, for e.g. develops a chest infection, they may need to be admitted to hospital for treatment. The length of time they need to stay in hospital varies.

Your child may also have a planned admission to hospital which is usually for two weeks.

Your CF nurse will talk with you about what happens when your child is admitted.

## Annual review

Once a year, your child will have a longer clinic appointment, called an annual review, where all members of your child's healthcare team – consultant, CF nurse specialist, dietitian and physiotherapist, will meet and discuss your child's care with you.

Your child will usually need to have a blood test, chest x-ray and as they get older, a sputum test and a lung function test.

You will be asked if you agree to have your child's information from the annual review entered into the New Zealand CF data registry, called PORT CF. This data is anonymised, which means no-one can tell which data comes from your child. The data is used to improve the health of people with CF through research and to guide improvements in care.

CF can also affect other parts of the body, such as your child's ears, nose and sinuses, liver function, bones and joints. Over time, some children can develop CF related diabetes, delayed puberty and problems with fertility. Your CF team will closely monitor for any signs of further complications as your child grows.



CF affects everyone differently – some children remain well for a long time and don't need to spend any time in hospital, while others need more frequent admissions.

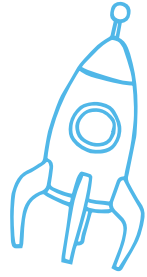
*"Clinics take a long time because of needing to see so many people - the nurse, doctor, physio and dietitian and sometimes others. But each one of them has been so important to our child's care, it's well worth the investment of time."*

*"We were pretty familiar with hospitals, but nothing really prepares you for that first admission! Make sure you take those little things that keep you sane - for us it's her favourite toys and books, snacks, iPad and charger and a coffee plunger with real coffee."*

*"My son was delighted at the constant attention from a stream of medical professionals through his room each day - far more entertaining for him than spending a week at home just with me! The CFNZ fieldworker visited a couple of times and managed to be both kind and empathetic and incredibly practical all at once."*

*"One of the difficulties I found with managing clinic appointments was partly just coping with the idea I needed to go to a hospital with my child on a regular basis - I had planned on going to playgroup with my son regularly, not a hospital. I found going with someone else was helpful as it meant I could answer and ask questions and the other person could entertain my son. It also meant there were two pairs of ears to get the information, which helped."*

## The future



"Our son is six years old now, and although he's been in hospital a few times, he's been fine for the last two and a half years - in fact more than fine. He's a healthy weight and doing so well at sport. He's a fast runner and received the most valuable player of the year award for his first year at Rippa."

"My son sometimes asks, 'Why did I have to be born with cystic fibrosis?' That's a really hard one to answer because there is no answer. He also asks why he needs to take tablets with each meal, and he's old enough now for me to explain that they are important for him to digest food properly and put weight on."

"Over time, cystic fibrosis became normal to us. I guess you just learn to accept it. I've recently read there are three stages of cystic fibrosis: one - you want to wrap your child in cotton wool, two - you become a bit more relaxed as they get older, and then three - you end up doing a bit of both - and I think that's where we are now."

"The first few weeks after we got the diagnosis of CF were a blur of feeding, feeding, feeding, appointments, medicines, physio, and no sleep. It was so impossible to see a future beyond the next five minutes. But things got better. A lot better than I ever could have imagined at the start. There's still lots of time spent on treatments, uncertainty about when the next infection will come, and concern about the future, but we haven't been admitted to hospital for over two years, our daughter's doing really well at school and we've managed to travel to the US and UK as a family. Life is different to what I thought it would be, but it's great."

"One of the reasons I'm so open about our son having cystic fibrosis is that if you try and hide it, it becomes more of an interest to people. We've always been very open at school and with friends and family that he has what he has. At first it was a big deal at school with kids asking, 'Why are you taking pills?' but after finding out they don't make a fuss or make him feel like he's different."





*"We've never held our son back from anything. We offer him any opportunity we can. Living where we are in Southland he can do anything - he can go biking, boating, motorbiking, hunting and fishing. We want him to be able to take advantage of any opportunity he wants. We think it's important to live your life as normally as possible and don't let CF hold you back. As soon as you start putting them in a bubble that's when things can change."*

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## More information and support

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As your baby grows – from a newborn, to a toddler, to a pre-schooler and then on to school, the information you want, and need, will change.

The CFNZ website, which we encourage you to visit, has up-to-date, New Zealand-based

information. You can find out about the services we offer, such as fieldworker support, regional branches, financial grants, information packs and learning opportunities, as well as recommended sources for further information.

We also offer two types of newsletters that are a great way to keep up-to-date with news about CF and what we are doing to support the CF community. The CF Panui is emailed monthly and the CF News magazine is published twice a year.

A private CFNZ Facebook group, a safe space to connect with other parents, is also available to join.

As your child grows we have awards and endowment funds that help to celebrate and support the amazing achievements of people with CF.

Cystic Fibrosis New Zealand is here for you, no matter what stage of the journey you are on. We understand a diagnosis of CF can be very difficult, and there is a lot for you and your family to think about.

*"CFNZ has been fantastic to us in lots of ways - but probably the most important way is just knowing there are people out there who are looking out for and caring for you even though you haven't met. It makes a hard road significantly easier."*



*We are here to help.  
When you are ready,  
find out more at  
[www.cfnz.org.nz](http://www.cfnz.org.nz)  
or call us on  
0800 651 122.*

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# Notes

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*A heartfelt thank you to the families who gave us valuable insight into their personal experiences with cystic fibrosis, through both words and photography. We could not have completed this project without your help.*



A special thank you to The Lion Foundation and SKIP for generously funding the writing and publication costs of this guide.

Lastly, thank you to the fieldworkers, clinicians and medical experts who shared their knowledge and expertise on cystic fibrosis, and took the time to review and fact check this guide.

*This publication has been written as a general guide for parents and caregivers of a child diagnosed with cystic fibrosis as a newborn in New Zealand.*

*While Cystic Fibrosis New Zealand (CFNZ) has consulted with healthcare professionals and families of children with cystic fibrosis to ensure the information is accurate, it does not substitute specialist advice from your medical team.*

*CFNZ will not be held liable for inaccuracies or omissions.*