CYSTIC FIBROSIS NZ

A guide to cystic fibrosis
for family, whānau and friends

cfnz.org.nz

About this guide

This guide is for New Zealanders who have had a friend, whānau or family member's child diagnosed with cystic fibrosis (CF). It aims to give you a better understanding of what CF is, how it impacts on the child and their family and how you can offer the best support.

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Dear family, whānau and friends

Finding out your child has a severe condition like cystic fibrosis is a life-changing event and one that comes with massive changes. Parents adjusting to a new diagnosis are under a huge amount of stress in addition to the usual stress of being a parent. Most need significant extra support to get through this difficult period following diagnosis and all that it entails.

Thank you for your interest in supporting your family, whānau or friends by taking the time to read this guide – I hope it answers some common questions you have about children with CF and their parents.

It is vital to remember everyone will deal with the diagnosis of CF in their child differently. Some people will be happy to discuss and share while others will be too upset to talk about it. But all will need some support.

The other important thing to know is that while CF is still a scary and life-limiting condition, things are continuously getting better. By reliably sticking to good chest physiotherapy, medications, exercise and nutrition, people with CF are living longer and doing more every year. Until there is a cure, adjusting to CF and treating CF will always be hard on people, but it is important to know things have improved even over the last five years and information on Google, which often terrifies, is often out of date.

Thanks again for your support of your loved one with a new CF diagnosis!

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

About Cystic Fibrosis New Zealand

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.

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If you would like to know more, please get in touch.

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Understanding cystic fibrosis

Cystic fibrosis (CF) is a genetically inherited condition some babies are born with that mainly affects their lungs and digestive system. It is caused by a faulty gene that has been passed down from both their mum and dad and is usually diagnosed soon after birth.

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

Every person has two copies of their genes – one from their father and one from their mother, and for a child to be born with CF, both parents must carry an abnormal Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene. About 1 in 25 people in New Zealand carry one copy of the CFTR gene (often called the 'CF gene') but you need two copies to be born with CF. Having one copy does not affect your health.

The CFTR gene

The CFTR gene carries instructions to make a protein (proteins are the building blocks of our bodies), called cystic fibrosis transmembrane conductance regulator protein, that controls the movement of salt and fluid in and out of our cells in different parts of our bodies. When the gene doesn't work properly, or is not made at all, salt can't get in and out of our cells as it is meant to.

When we don't have CF, our bodies make mucus that is thin and slippery and works as a lubricant to help protect us from infections and to keep the insides of our bodies working well.

If we have CF, our mucus becomes thick and sticky and blocks the tiny tubes of many of our organs.

In the lungs, the sticky secretions are difficult to cough up and bacteria can get trapped, causing infections. Children with CF have a much higher risk of developing serious complications from certain viruses, bacteria and fungi that are generally harmless to others. To help keep the child's lungs healthy they will need daily chest physiotherapy which the parents will be taught how to do and sometimes they will need antibiotics or other medications.

The thick secretions also block the flow of digestive enzymes from their pancreas to their small intestines where they are needed to help digest breastmilk, formula and solid food. Without these enzymes, fats and vitamins can't be absorbed by their bodies so it can be much harder for children with CF to put on weight. Most children will need to take pancreatic enzymes with every feed.

Receiving the diagnosis

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

If this test showed the baby had a high level of an enzyme called immunoreactive trypsinogen, 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. If this result is positive the baby is referred for further testing.

Further tests to confirm the diagnosis

Sweat test

A sweat test measures the amount of salt (sodium and chloride) in the baby's sweat and is usually done when they weigh over 3kg. 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 the baby is older.

Stool (poo) test

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

Blood test

The baby will have another blood test to ensure a correct diagnosis and to identify their CF gene mutation.

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

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 it.

When the parents learn their baby has CF, either soon after birth or a few weeks later, the news usually comes as a shock followed by a wide range of emotions including anxiety, grief, guilt and sadness. For many parents, the length of time waiting for confirmation of a diagnosis feels like a void which adds to the complexity of emotions.

There is currently no cure for CF and life expectancy is reduced to the mid to late 30s. This is shared with all parents and will have a huge effect on them as they come to terms with the news.

"My daughter was 18 days old and my midwife rang me at night, so I knew it wasn't a good sign. She said there were cystic fibrosis markers that came up in her heel prick test. When you don't know anything about CF you know it's bad because it's obviously screened for, but you don't know exactly what that means."

"The initial stages were obviously denial. I was in denial right up until we had the bloods done, until the genetics came back and confirmed the diagnosis."

"I didn't want people to feel sorry for us. My daughter was perfect to me – it meant a lot when people were still really happy for us and celebrated our new baby."

Living with CF day to day

CF affects every baby differently – some may need to be in hospital from an early age and have many issues impacting on their health, while other babies stay well and only need to visit the hospital for clinic appointments.

At home, parents will be juggling learning how to be a parent while also learning how to manage CF. Some babies with CF can have a lot of upset tummies, wind, more frequent and smelly poos and can be harder to settle.

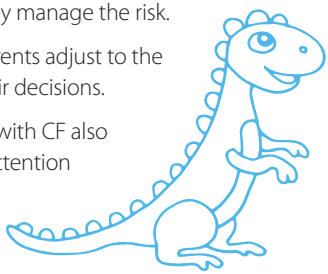
Some babies with CF manage to gain weight by just breastfeeding or having infant formula but some may need a special high-calorie formula. Most will also need to take pancreatic enzymes with every feed (this includes night-time as well!).

One of the biggest and most time-consuming treatments parents need to learn is how to do chest physiotherapy or airway clearance. Because the mucus in the baby's lungs is thick and sticky it can block some of the smaller airways and cause infections, so twice daily physio is a good way to help keep their lungs clear. Learning how to do physio is daunting to most parents, and so is the prospect of having to do it twice a day, every day.

Certain bacteria and viruses which are harmless to people without CF, can cause long-term damage to a child's lungs when they have CF, potentially impacting on their quality of life, lifespan or opportunities for future treatments. Parents with newly diagnosed children are taught early on some areas around the house and in the environment where their child is more at risk. Often parents find the risk of infection a huge worry, so it's important to listen to parents and support the decisions they make about how they manage the risk.

Establishing a routine with a new baby takes time, and as parents adjust to the additional care their baby needs it's important to respect their decisions.

Even though it takes a lot of work to keep them well, babies with CF also need what every baby needs – cuddles, playtime, love and attention and they will reward you with smiles (and crying!) just like other babies do too.



"After a short period of time it's amazing how quickly living with CF became our new normal. But it's still important people remember, even if we don't talk about CF all the time, we are still impacted by it on a daily basis."

"I still want her to just be a baby. 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."

What about you?

The diagnosis of CF also impacts on the wider family and whānau and, like the child's parents, you may be coping with a wide range of emotions.

Because CF is a genetic condition, many parents feel guilty following diagnosis that they have passed on the gene to their child and this guilt may be shared by extended family members.

It's important to remember there is nothing anyone could have done to prevent CF, but acknowledging your feelings and working towards acceptance is an important step in supporting the family and their baby.

How can you help?

Every family finds their own way to cope with the day-to-day challenges of living with CF. Some parents naturally find it easy to ask for help, while others find it hard and may not get enough good support from friends and family.

CF is a life-long condition and parents benefit from ongoing practical and emotional support.

Practical support

Even without a diagnosis of CF, caring for a newborn baby is hard work. Practical ways you can help include:

- *preparing meals*
- *picking up groceries*
- *looking after siblings – at home, after school or during hospital appointments*
- *picking up supplies and medications from the hospital and pharmacy*
- *learning about their treatments, such as how to give pancreatic enzymes or how to do chest physio-therapy, so parents can have a break*
- *helping with the housework*
- *visit skip.org.nz for more ideas.*

If you're unsure how you can help, ask!

"My youngest was born with cystic fibrosis and I had two other healthy children to take care of. This was quite stressful when my daughter was really sick. I was so grateful to my mum and sister who came over regularly and helped take care of the other two kids. A few close friends would also come over to cook meals for me – they made sure I was taking care of myself while I focused on my daughter."

"As a new dad, getting my head around CF and being there for my family meant being away from work quite a bit – I was so grateful to my colleagues who covered for me when I needed time away from work."

"Practical help trumps gifts – the best thing anyone has done for me is cook me a meal or two."

Learn about CF

Learn more about CF – this can help you be empathetic to what the family is experiencing and it can help you explain to other people what CF is and how they can support the family.

One of the challenges many parents face is finding a balance between letting their baby explore the world and be a 'normal' baby and toddler and minimising the risk of picking up bugs from other people and the environment. Be guided by parents as to how you can help.

Chest infections in children with CF can have long-lasting consequences, including a permanent decrease in how well their lungs work. Viruses and bacteria that cause chest infections in children without CF may make children with CF much sicker because of their lung problems and thick mucus.

People who don't understand the consequences of catching certain bugs may think parents are being overprotective or the child is being 'put in a bubble'. Parents can find it hard to explain to others why they are being cautious, so if you understand the risks you can explain it to others.

> "Some of our friends were great with wiping down surfaces and toys before we came to visit, which made visiting much easier and I could relax more."

Be mindful that information about CF on the internet is not always accurate and different articles that give conflicting advice can add to a parent's anxiety and stress. We encourage you to visit the CFNZ website for accurate, up-to-date information about cystic fibrosis.

> "People sometimes look at us like we have extra heads when we ask about compost heaps and fish tanks when we visit – but several of our friends have come to understand and gotten rid of or moved the hazards. It is not like they're guaranteed to get sick from being close to these things, but why would you take the chance when you don't have to when the consequence might be a week in hospital and long-term lung damage?"

> "What can someone say or do to help? Educate yourself. It's hard to get advice from people who don't even know what's going on. Those who are not living this day in, day out. 'But he looks so healthy' or 'he'll grow out of it' drives me crazy! CF is a progressive and invisible disease and my son is going to look healthy as long as we stay on top of our daily routines, and he is only going to get worse, not better. That's why I have routines. It may mean missing coffee dates with friends, cancelling dinner at times we have planned or not calling you even though I promised three weeks ago, but often it can't be helped."

Emotional support

Knowing the right things to say to someone who is dealing with a life-limiting diagnosis can be hard, but just being there to listen is helpful. Having a child with CF is something parents don't expect and it can take time to process what the diagnosis means for them, their child and their family. Some days will be harder than others, but over time, living with CF becomes part of their everyday life.

"Sometimes just a shoulder to cry on or an ear to listen when you need to vent or have a moment when reality hits – though as time goes on it doesn't happen too often."

"It helped that our friends still treated us the same way, rather than isolating us because we had special requirements. They still invited us to things and gave us the opportunity to decide for ourselves if we would come."

"Being told 'you're doing a good job' or 'he's a fighter' is encouraging to hear. Because this is a marathon, there is no end to these treatments, so encouragement like this keeps us going."

Support for siblings

It is inevitable parents will be spending a lot of time following diagnosis learning about CF, so family, whānau and friends can play a significant role in helping brothers and sisters to not feel left out or forgotten about.

Make time for siblings – take them for an outing, play with them at home or offer to have them at your house. Ask the parents the best way you can support them or ask siblings what they want to do.

"One of the best ways friends have supported us is by taking my other three children to a park for a few hours. We are in the hospital a lot, so my other kids miss out on the extra fun stuff. I'm trying to learn balance for them too other than school and home life. I feel, at times, they are overlooked at times of stress. It's a constant thought for me."

"When my friend's daughter was in hospital, I used to go in during the evenings and sit with her, so her parents could go home and have dinner and spend time with their other children."

Cystic fibrosis is a life-long condition and support from family and friends will always be needed. Parents will need different support at different times as their baby grows, **so keep offering to help and stay connected.**

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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.

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