CYSTIC FIBROSIS NZ

Still Living in Hope

– written by the parent of a person with CF

NB: To protect my "children's" identity and privacy I have used gender-neutral pronouns and not named them.

The first time I ever heard the words cystic fibrosis was from my GP's nurse shortly after the birth of my second child in 1989, when she rang with the heel prick results. The results were inconclusive, so sweat tests were required for both my baby and my 17-month-old first born.

When I asked what cystic fibrosis (CF) was, the nurse's response was direct and brutal: ***"It's a genetic disease of the lungs and digestive system. Babies do not thrive and die at a young age."***

She then tried to reassure me saying the heel prick often resulted in a false positive and she was sure my baby would be fine. But when she described the symptoms – runny nose and poos, constant desire for feeds with poor weight gain – I knew the outcome of these tests would not be good.

What was good news, we were told at the hospital, was that the CF gene had been identified only a month earlier. Because of this, we were told we could expect a cure to be discovered within five years. This filled us with hope.

From the moment of diagnosis, our lives went off on an unplanned trajectory – learning how to navigate and self-advocate within the health system and hospitals, understanding genetics, dealing with grief, putting expansion of our family on hold.

I found myself contemplating the impact of a possible second child with CF when, almost exactly one year later, I discovered I was expecting. Because of the CF camps, I knew of other families with more than one child with CF and their support helped me through that difficult time.

We were placed under the paediatric CF clinical team in our area. Unfortunately, they had never dealt with a newborn before, and at the time CFNZ did not have social workers to guide families through the diagnosis stage - something I am glad our families can access now.

There are so many things I know now that I wish I'd known back then - from how to give enzymes while breastfeeding to help my baby digest food, to how to perform chest percussion effectively. Much of it I learned the hard way. It wasn't until later that I discovered apple purée on a spoon was the recommended way to administer enzymes - who knew you could give solids to a newborn?

With the physio, I performed chest percussion for an hour every morning and night. This involved sitting with the baby lying headfirst down my legs, patting them gently but firmly with cupped hands across the lungs to clear their airways. As my husband was a shift worker, I did every session on my own whilst also caring for my two other pre-schoolers. My voice was the only tool I had to keep them safe.

It was during those hours each day that I felt the full impact of the diagnosis. Pinned to the sofa with nothing but my three beautiful babies and my thoughts of what the future would hold.

Long after my children had left home, I found out that an hour each session was far more than required – but I survived it.

In the end, I worked things out myself. I learned some parenting strategies (good ones, or not) and like to think I gave my child the best start. I stayed active within our local community to ensure those who came after me had access to support, or just a listening ear.

Another reason for becoming involved with the CF Association (CFNZ) was to keep abreast of the post gene discovery groundbreaking research. The support from our local branch had been tremendous but limited. Together we formed a committee and launched into our first major awareness activity.

Shares in Life was a national initiative to raise funds for research towards a cure, and the awareness and donations raised in our area were amazing.

Since then, our branch has actively raised funds for equipment, welfare needs, support during hospital stays, family get-togethers and conferences. I eagerly attended organisation-wide meetings and conferences every year, always hopeful for news of THE CURE. Sadly over 30 years on, while there have been important clinical breakthroughs, a cure is still on our wish list.

We also funded and hosted the very last CF family camp in 1996.

Those camps were so important to me as a young mother of 3 preschoolers – a place to learn, have fun, share experiences, and where the whole family was supported. Sadly, in the early 1990's evidence indicated people with CF were cross-infecting one another with life-threatening bugs by being in close proximity. Despite all the benefits, the camps had to stop.

From that time on, people with CF were isolated from one another and opportunities for families to connect, learn and support each other were compromised.

Those first 10 years of our journey were a balance – keeping up with research and hoping for a cure, while becoming part of the CF community and learning from shared stories. This was tempered by the reality that many of the people with CF that I was getting to know were more vulnerable having been born before CF was fully understood and before treatments and clinical practices had evolved.

There was a period in the 1990's when, seven years in a row, I attended funerals for people with CF – none of whom had reached the age of 33. I remember every single one of these brave people from our local community and think of them often.

My "baby" is now an independent adult in their late 30's. I am grateful that they have survived – but this is all that it is: survival.

New therapies have given the gift of easy breathing, and their lungs are infected less often. But joy is not a given with this new chance at life. My child is one of the few who is affected by the mental health side-effects of these medicines. Combined with a diagnosis of CF-related PTSD, every day remains a struggle.

As a mother, I am still waiting for that cure.

I still live in hope.

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