A PATIENT'S JOURNEY

Cystic fibrosis

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As the Good Witch told Dorothy in the *Wizard of Oz*: it is always best to start at the beginning. Growing up, I always knew that I had cystic fibrosis, the same way I knew I had blue eyes and my cousins could all run faster than me.

I was lucky enough—though my parents did not think so at the time—to be diagnosed at two days old. The right information at diagnosis is crucial. After being told their daughter had a disease whose name they could not spell, and with no information from the hospital, my parents found out about it for themselves. Parents nowadays might use the internet; mine went to the library. The book they found, printed 15 years earlier, told them to abandon all hope and not become too attached to me.

Travelling alone

The road travelled with cystic fibrosis is often deserted, devoid of like minded companions. Growing up, some children picked on me because of my cystic fibrosis, but most of my classmates were too busy learning how to do chest physiotherapy or fighting over the honour of keeping me company when I was too unwell to go out at playtime. I decided that those who were mean were simply jealous of my "special treatment." If only they had known that I would gladly have swapped with them in an instant.

Today, I welcome and respect the practice of segregation to prevent cross infection. Technology has lessened the impact of infection, but the condition is still not easy to bear. I wish I could sit in a room of people who know what it is to live with it.

I know many people with cystic fibrosis, although none of them are close friends. This is a choice I have made. The close friends with cystic fibrosis that I had previously have all died. Having friends who have cystic fibrosis can become a burden. I know I have this disease and I live with it every day, but to have it staring back at me through the eyes of another can be daunting.

There are the bad days—the ones where I have to ask for a helping hand, when I cannot be independent, when people I love have to clear up vomit and faeces and change their plans around me. Although they probably do not see me as a burden, that is how I feel. It is a bizarre situation; having this disease has made me grow up faster, yet because I am ill I am still heavily dependent on others.

Companions on the journey

While still a teenager I told my parents, "I'm old enough to take care of myself." What rubbish. Nobody, whatever their age or health status, is past needing a bit of help. As I become sicker it will be my loved ones taking care of me once again. I know I will have to rely more on their support in the future. I am not sure who is looking forward to it the least.

Although I do not always want it to be, cystic fibrosis is a huge part of my life and of the lives of those close to me. It is important that healthcare professionals do not forget the people close to me; the relationship professionals have with their patients should extend to the people who care for them every day. My companions resent my illness more than I, because I have control over it while they look on helplessly, and some may need additional support.

It can be hard on my fiancé, as he feels responsible for my wellbeing and compliance with treatment. Moreover, he is still learning, as we all are. He has many questions, some of which seem a bit stupid, but it is important that members of the cystic fibrosis team do not make him feel the questions are unimportant.

What adults with cystic fibrosis need

A good relationship with healthcare professionals is essential, as they eventually become part of a patient's extended family. People with long term conditions need people with whom they can discuss their concerns, beyond the medical ones. I am lucky; I attend a specialist centre where every member of staff is dedicated to looking after people with cystic fibrosis.

It is important that health professionals should see the person with cystic fibrosis as part of the team, their views being as important as professionals' own. I become frustrated when clinicians seem unable to accept that I, having lived with cystic fibrosis for more than 20 years, might know more about my illness than they do. Health professionals may be experts in their field, but patients are experts in their lives. The best doctor-patient relationships are those in which both parties educate each other.

Patients need access to information about cystic fibrosis. If professionals do not provide the right information at the right time, patients will go and look for themselves. Plenty of good quality information is available, but there is just as much incorrect information,

USEFUL RESOURCES

The Cystic Fibrosis Trust (UK)—telephone: +44 (0) 20 8464 7211; www.cftrust.org.uk

The Cystic Fibrosis Foundation (USA)—toll free telephone: +1 800 FIGHT CF (344-4823); www.cff.org

The Breathing Room—A virtual community of adults with cystic fibrosis, sharing experiences with both informal and professional caregivers; www.thebreathingroom.org

too, especially on the internet. Patients need to be able to filter out misleading facts, or have access to people who can guide them.

Losing the path

Having cystic fibrosis is different for everyone. Even with specialist multidisciplinary care it is still possible to lose grip of the steering wheel and veer off course. For example, when I have an acute infection I would love to take my intravenous antibiotics at home. It is much better than sitting in hospital for two weeks, especially when it is too far for anyone to visit. During my childhood my local hospital sent me home, armed with 15 minutes' worth of training and one nervous mother, because they needed the bed. It was a disaster; we managed only three days, and the experience has stopped me from attempting intravenous treatment at home. I am probably capable, but as I live outside the hospital catchment area for homecare nursing I would have no support. With proper training, sufficient supplies, and adequate support I am sure I would feel differently.

I would have given anything to attend a transition clinic when I was 16. Instead, I received a letter stating that my next appointment would be in an adult clinic at another hospital. Now, people with an up to date atlas of knowledge patrol the rocky roads of Transition. Although this is far from perfect, it is good to know that certain things are improving.

The good news

Mostly, the future is bright. Gene therapy, while not exactly just round the corner, is looking extremely likely and is an incentive to adhere to treatments. The healthier patients' lungs are, the more likely they will be able to benefit from the therapy.

The treatments themselves have become less cumbersome. Carrying out my nebulised treatments used to take an hour of my day; now it takes around 30 minutes, including cleaning the equipment. The new nebulisers are compact enough not to need a separate suitcase for taking them on holidays. The physiotherapists now supply patients with a range of gadgets to clear the chest of mucus. It may not take less time to do, but it is much more pleasant than chest percussion—and I can do it by myself, so nobody has to find time to help me.

The best news is that people with cystic fibrosis are living their lives, and living them longer. We have jobs, families, and children. Most of us are going out and doing things our peers would never dream of doing.

The bad news

Of course, not everything is as it could be. Everyone with cystic fibrosis in the United Kingdom does not receive safe and appropriate care from a specialist multidisciplinary team. Because adults are not automatically entitled to free prescriptions, I spend a fortune paying for drugs—when I can get them in the first place. My general practitioner is fantastic, but sadly "postcode prescribing" is as prevalent as ever, and across the country, people living with cystic fibrosis are fighting battles with primary care trusts.

Journey's end

My biggest fear is the future; it is difficult to know what it will bring. It is difficult to plan for the time that my parents were told I would not have. It is hard to think about getting a mortgage, or starting a pension when you're not sure you'll live long enough to have a retirement.

But I do not sit worrying about when I'm going to die. I think about it, but do not walk around clutching my funeral arrangements.

So, I will worry about the future when it arrives. Until then, there are too many things to live for—my wedding, books to read, and bands I still have to discover.

A RESEARCHER'S PERSPECTIVE

Identification of the cystic fibrosis gene in 1989 has led to earlier, more accurate diagnosis, and neonatal screening is being rolled out throughout the UK. Better understanding of the condition has ensured progressively more effective, patient friendly, treatment and care, most of which is carried out at least daily by family members in the home.

Many young people around Emma's age decide against befriending others with cystic fibrosis, and this may result in a feeling of isolation. In 1996, the Cystic Fibrosis Trust began to fund expert patient advisers, contactable through the trust, whose role is to enable all those affected by cystic fibrosis to have a voice in service planning, delivery, and review.

Average survival age for those with cystic fibrosis, currently 31 years, is expected to reach 50 years for those born at the turn of the 21st century. For the first time in history, adults with cystic fibrosis in Britain will outnumber children living with the disease. As individuals age and their health declines, many conditions related to cystic fibrosis, such as diabetes, osteoporosis, and liver disease, become more likely.

Living into adulthood also presents those affected with new psychosocial challenges: taking over responsibility for their treatment and care; negotiating further education, employment, and finances; gaining greater independence from parents; managing personal relationships; and deciding whether to have families of their own. Transition clinics, staffed by a multidisciplinary team, have been established to enable a smoother journey between paediatric and adult care, although resources are not always available to provide the holistic care that these young adults need. Despite requiring daily, life sustaining treatment, most adults with cystic fibrosis continue to pay prescription charges.

The possibility of a lung or heart-lung transplant is a hope shared by many with end stage cystic fibrosis, yet fewer than half of those on the waiting list will live long enough to receive donor organs. Of those who do, fewer than half will survive for 10 years or more. End of life care for this population is variable; currently many young people die in their teens, without access to a dedicated supportive or palliative care service. Emma is typical of the scores of young people and families affected by cystic fibrosis that I have worked with in research projects over the past 10 years. Although they face adversity almost daily, an attitude of "life is short; live it to the full" prevails. Excellent progress has been made in helping these young people to fight this disease, but there is still much more to be done

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