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“Everyone with cystic fibrosis is unique and will have their own relationship with CF and their own range of symptoms. It’s hard to paint a picture that illustrates 10,000 different stories,” says the Cystic Fibrosis Trust. Here’s one very personal story.

Ruth van den Broek

Ruth is a 28 year old Oxfordshire woman who has late stage CF. Her lungs are badly damaged and she wears oxygen 24 hours a day: the result of thick sticky mucus clogging up her lungs (caused by a faulty gene), leading to frequent infections that damage the lungs.

Like all of us, Ruth needs to take about 25,000 breaths a day, and each breath can be a struggle when you’ve got CF. Daily physiotherapy can help clear the lungs; strong antibiotics can fight the lung infections. In Ruth’s case, however, the lung bacteria have become resistant to many antibiotics.

She says, “When lung function becomes very low, a double-lung transplant is an option for most. Unfortunately, there aren’t enough organs available and more than a quarter of people waiting for lungs won’t get them. The people who do receive lungs can go on to have a much improved quality of life.”

Enlisting celebrity support

Ruth revealed that more than half of people with CF in the UK will live for more than 47 years. She explains that this number keeps rising as new treatments are being developed and becoming available. But there isn’t a cure yet and life with Cystic Fibrosis remains extremely challenging.

Aiming to raise awareness of CF and funds for vital research, people living with CF have worked alongside celebrities such as James Corden and Lorraine Kelly to record themselves simply breathing. The resulting charity single [5] envisions a day when those with CF can lead ‘a life unlimited’ (the name of the track), but there’s still a long way to go.

Damaged digestion

The thick sticky mucus of CF doesn’t only affect breathing. Ruth explains how mucus in the pancreas [6] stops enzymes which digest food from reaching the intestine. As a result, people with CF often find it hard to maintain a healthy weight and a high fat, high calorie diet is necessary; supplementary enzymes and vitamin capsules can also be taken. Sometimes a feeding tube is required to help with weight gain.

Currently Ruth is trying to increase her weight to make her eligible for the double lung transplant waiting list.

“Other complications of CF can include CF related diabetes, osteoporosis [7] (due to poor absorption of vitamin D), arthritis, male infertility [8], liver damage, sinusitis, reflux disease, and kidney and hearing damage from antibiotic toxicity. People with CF often struggle with mental health issues as it can be difficult to live with the disease and its implications.”

What are current treatments?

“Diet and exercise are important for people with CF. Exercise [9] has been shown to slow lung function decline.

In the past few years, genetic treatments have been, and are continuing to be developed. There are, though, hundreds of different gene mutations, making it extremely difficult to treat CF. The most successful of these medications targets a mutation carried by 4% of people with CF [10] and has transformed the lives of many.”

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