Issue 15
cysticfibrosis.org.uk

Cystic Fibrosis Trust

CFLife

Uniting for a life unlimited

Genetic therapies
Understanding what genetic therapy trials involve

Fly on the wall
Including everyone in clinical trials

Your stories
Kieron talks about his involvement in our awareness campaign

What's inside

Issue 15 – August 2023

If you'd like to give us feedback on this issue, or have ideas for what you'd like to see in the magazine, email us at **magazine@cysticfibrosis.org.uk**

On the cover: Kieron, who features on p14
This page: Jack, who features on p24

Social

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Useful contacts

Donations
020 3795 2177
supportercare@cysticfibrosis.org.uk

Events and fundraising enquiries
020 3795 2176
events@cysticfibrosis.org.uk

Cystic Fibrosis Trust Helpline
0300 373 1000
helpline@cysticfibrosis.org.uk

Our confidential Helpline offers general advice, support and information on any aspect of cystic fibrosis, including help with financial support.

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Welcome to CF Life

In this issue of CF Life, we're excited to share our brand new genetic therapy resources, explaining what they are and how they may be used to treat CF.

While more people with CF live longer and fuller lives, we are mindful of wider issues arising and ensuring we support people through them. Our feature on bowel cancer and CF is a stark reminder of how far we still have to go to achieve a life unlimited for everyone with CF.

Following the launch of our latest awareness campaign, 'You don't see CF', we speak to our amazing supporter Kieron about his involvement in the campaign. Plus, we talk with CF paediatrician Dr Maya Desai to learn about the importance of diversity in clinical trials.

In our What's on your mind? column, Matthew from our Helpline team answers some of your pressing questions. This month he covers CF Connect, our new peer support service for parents and carers with a child with CF.

Elsewhere in this edition, we take a break with BBC Interior Design Masters contestant, Jack Kinsey, who has CF, to hear about his work and time on the show.

We hope you enjoy this issue, and please get in touch with your ideas and feedback. We always love to hear from all of our wonderful supporters.

The CF Life team

Uniting for a life *unlimited*

In case you missed it

Fundraising

We wanted to give a big thank you to Carol from Reading, who attends an incredible amount of local school fetes, village fayres and elderly people's homes, selling items and donating all the profits to Cystic Fibrosis Trust.

Over the many years Carol has been fundraising for us, she has raised over **£14,500**, proving that every single donation, no matter how big or small, can add up to one magnificent total.

Thank you to Carol for all of her efforts and to everyone that helps her out along the way.

Carol

Research

In May, we announced a new co-funding partnership with the medical charity LifeArc to create a Translational Innovation Hub Network. **"This marks an exciting and innovative investment in understanding and fast-tracking treatment of lung infections for people with CF. By working with LifeArc and combining our expertise, we are able to deliver the best possible outcome for people with cystic fibrosis,"** said Dr Lucy Allen, the Trust's Director of Research and Healthcare Data. This new Network will build on the success of the Trust's existing Innovation Hub at the University of Cambridge, funded by AJN Steelstock, the Robert Luff Foundation and the Garfield Weston Foundation, along with a number of donors, trusts and companies.

Support

We've made a new factsheet all about non-tuberculous mycobacteria (NTM), which includes *Mycobacterium abscessus*. You'll find information on NTM lung infection diagnosis, how it can be treated, ongoing research and more.

You'll also see plenty of quotes from people who have experience with NTM. We had help writing this factsheet from Professor Andrew Jones, Consultant Physician and Director of the Manchester Adult Cystic Fibrosis Centre, and it was reviewed by CF professionals and people in the CF community with lived experience of NTM. We would like to thank everyone involved for their hard work and patience!

You can read the factsheet at **cysticfibrosis.org.uk/lung-infections**

Twins, Claudia (top) and George (who has CF)

Young people

Twins George and Claudia are seven and have been coming to the Trust's youth programme events for almost a year. George has CF, and Claudia doesn't. Here, they're showing off their delicious chocolate Easter egg brownies that they baked during the spring baking workshop led by a young woman with CF. George and Claudia were joined on the call by other children with CF and their siblings, and were encouraged to play fun ice-breaker games and carefully talk through the recipe. Look at how proud they are of their brownies!

If you want to know more about how to join the youth programme for children ages 6–18 with CF and their siblings, email **cfyouth@cysticfibrosis.org.uk**

Campaigning

Over the past two years, Cystic Fibrosis Trust has been working to highlight the experiences of those living with cystic fibrosis. In the summer of 2022, we published the *Cost of CF* report, highlighting the worries and lived experiences of people with CF and their loved ones.

The University of Bristol followed up this report with an academic piece of research to put a monetary figure on the average cost of living with CF. In June, the Trust published its latest report, *Your Life and CF*. This new report expands on previous research and updates on how the community is experiencing the current climate.

We will continue to raise awareness of the financial challenges facing people with CF, campaign for greater support from the UK Governments and strengthen the direct support that we provide to those in our community who need it most. Find out more at **cysticfibrosis.org.uk/cost-of-cf**

Understanding bowel cancer and CF

Significant improvements in treatment, standards of care and understanding of cystic fibrosis mean there are now more adults than children with the condition. We believe that we can reach the day when everyone born with cystic fibrosis can live a life unlimited by their condition.

As people with CF live longer into adulthood, they have exciting new opportunities but can also face different challenges and issues. At the Trust, we are mindful of these wider issues and are here to support you and your family through them.

In this article, Hannah, 44, a former NHS doctor, shares her journey of growing older with CF, including her diagnosis and treatment for bowel cancer. Hannah shares her own personal story, along with some information and advice based on her experiences and working in the medical field.

Our Helpline is available to anyone looking for information or support with any aspect of cystic fibrosis, a listening ear, or just to talk things through. **You can contact our team Monday – Friday, 10am – 4pm on 0300 373 1000.**

Born in 1979, I was diagnosed with CF at six weeks old due to significant weight loss – in the era before genetic testing and newborn screening. The average life expectancy at the time was mid-teens.

I was fortunate to have a brilliant paediatrician and proactive parents, so despite having a double F508 mutation, I was relatively well in my childhood and early teens but still had recurrent chest infections, issues with pancreatic insufficiency and constipation.

At the age of 15 years, I was diagnosed with severe liver disease. There were several discussions with the transplant team about the possibility of a liver transplant, but my liver deterioration was never severe enough to progress to this.

After several years of glucose intolerance, I was diagnosed with CF diabetes in my mid-twenties.

In my late twenties and early thirties, my chest infections became more frequent and worse. Despite various suggestions from my brilliant CF team to rest and be admitted for IVs, I continued to refuse!

I avoided IV antibiotics until I reached 30; in hindsight, that stubbornness was a mistake, forced in large part by my working culture.

By the time I'd reached my thirties, I had developed a range of CF-related issues: lung problems, pancreatic insufficiency, CF-related liver disease, CF diabetes, osteoporosis, vitamin B12 deficiency, hypothyroidism, antibiotic-induced hearing loss, gastroparesis (where the stomach empties too slowly), chronic neutropenia (low white cell count, which makes it harder to fight infection) and thrombocytopenia (low platelets, meaning my blood doesn't clot properly).

Over the past two years, my health has been much more stable, probably a combination of stepping back from working in the NHS, prolonged shielding and reduced socialising (unexpected but timely benefits of the COVID-19 pandemic), limiting opportunities for collecting germs, as well as the introduction of Kaftrio.

However, at the age of 40, a new diagnosis was added to the list… bowel cancer.

I was diagnosed with bowel cancer in 2019. I didn't have any early symptoms and wasn't part of any surveillance programme, as is the case in some CF centres in the UK and USA.

I suddenly developed a bowel obstruction, and a CT scan showed a tumour at the beginning of my colon. Within a week, urgent surgery was undertaken to remove half of my colon and to create an ileostomy (an opening in the tummy called a 'stoma' for the bowel to exit).

Due to my complicated history, my postop recovery was a challenge. I was in hospital for a total of six weeks, including 11 days in intensive care.

Fortunately, despite a large tumour and late diagnosis, the bowel cancer had only spread into the nearby blood vessels and had not gone to the lymph nodes or to the liver.

I found navigating cancer and a stoma whilst shielding in a pandemic challenging!

My stoma frequently misbehaved and was difficult to manage. I was actually relieved to be shielding at home rather than navigating work, social life, travelling etc. Significant complications with my stoma nine months after my original cancer surgery led to a stoma reversal to 're-join' my bowel, which was such a positive outcome for me both physically and emotionally.

Following my diagnosis three years ago, I am now on a surveillance programme via the colorectal surgeons with annual colonoscopies and CT scans to monitor for signs of recurrence of the initial tumour and to detect any potential new cancers as early as possible.

"It is important that if you develop any symptoms that you talk to your GP and explain the increased risks of cancer for adults with CF."

Hannah

Bowel cancer in CF

The CF population have an increased risk of bowel cancer compared to the wider population. The average age of diagnosis of bowel cancer is 52 years old for those with CF, with a quarter of cases arising before the age of 40 years, compared to 73 years in the general population.

This is why some CF centres in the UK and most in the USA conduct routine bowel cancer surveillance for CF patients from the age of 40 years (and from 30 years if you have had a transplant).

Monitoring is used to detect cancer early and treat it before it spreads further afield.

Symptoms of bowel cancer:

- Bleeding from your bottom and/or blood in your poo
- Change in bowel habits (becoming more constipated, having looser stools etc.)
- Unexplained weight loss
- Extreme tiredness for no obvious reason
- Pain or a lump in your tummy

Not everyone will experience these symptoms (as in my case), which is why surveillance is crucial. It is important that if you develop any of the above symptoms that you talk to your GP and explain the increased risks of cancer for adults with CF.

Some of these symptoms may be disregarded by medical teams as they may consider us **"too young to have cancer"** or, wrongly, attribute these symptoms to CF rather than cancer without looking into the matter further.

Therefore, it is really important for adults with CF to be aware of these symptoms and ask for appropriate investigations and surveillance.

We can also help to reduce our risks of developing bowel cancer by increasing physical activity and exercising regularly, maintaining a healthy weight, minimising alcohol consumption, not smoking and avoiding eating processed meat, limiting red meat and eating plenty of fibre from whole grains, fruit and vegetables.

> **"I am just so grateful to have lived 13 years beyond my original life-expectancy 'best before' date and to still be relatively well"**
>
> Hannah

My hopes for the future and living a life unlimited

Over the past 40 years, I have seen amazing improvements to life expectancy but also, and probably more importantly, advances in antibiotic treatments, physiotherapy, nebulised medications such as DNAse as well as CFTR modulators.

I see the fantastic educational work done by the Trust and the dedication and skill of our healthcare teams - learning not only how to deal with lung and digestive problems but rising to the challenge of CF being a multi-organ disease.

I am hopeful for the research projects that are currently underway, and those planned for the future that they will benefit the CF population on both a daily basis and also help individuals to live well with a chronic illness and to fulfil their potential.

I am just so grateful to have lived 13 years beyond my original life-expectancy 'best before' date and to still be relatively well and to be able to do a number of hobbies that give me joy whilst also contributing and being a positive influence in our community.

I would love for my health to remain stable enough for more travelling (insurance premiums and policies allowing!) and perhaps convince my husband to let us get more dogs... watch this space!

If you feel you have been affected by any of the topics covered in this article please contact our Helpline Monday – Friday, 10am – 4pm on 0300 373 1000.

"Hannah very vividly talks about her life with CF, and through her personal experience, raises the very important issue of the increased risk people with CF have of developing bowel cancer at a younger age than the general population. Screening for bowel cancer in people with CF is especially important because it is more likely to occur at a younger age. Some studies suggest it is more likely to occur higher in the colon, resulting in delayed symptoms, as people with CF often already have bowel symptoms due to their CF. If you have concerns, please do discuss them with your clinical team."

Dr Keith Brownlee, Director of Medical Affairs at the Trust

Dr Keith Brownlee

Empowering everyone to take part in clinical trials

To make sure any new CF treatments can benefit everyone, we're keen to ensure that as wide a group of the CF community as possible has an opportunity to help design and participate in clinical trials.

Tonia from our Involvement group spoke to Dr Maya Desai, CF paediatrician at Birmingham Children's Hospital and co-leader of a project to increase access to CF research and clinical trials.

Dr Maya Desai

> "It's important to spread the word that opportunities to get involved in and take part in research and clinical trials are open to everyone"
>
> Dr Maya Desai

Tonia: Tell us more about your project to increase diversity in research.

Maya: In the past, everyone with CF got the same treatment. Now there are medicines like Kaftrio. However, there are a group of people who can't benefit from them, so we need to be working even harder to find solutions and better treatments.

From the people we see in the clinic in Birmingham, it's clear that there's a difference in ethnic diversity between people who are able to take modulators and those that are not. The UK CF Registry's 2021 Annual Data Report found that nearly 6% of people with CF were from Black, Asian and Minority Ethnic backgrounds.

It made me think are we involving people from these backgrounds enough in clinical trials and research? It's really important we do because we want to ensure that they can benefit from the treatments being tested if they become medicines.

Tonia: What's the first step?

Maya: It's important to spread the word that opportunities to get involved in and take part in research and clinical trials are open to everyone. We want to make people feel as welcome as possible when they do.

Parents I've spoken to on a one-to-one basis say **"we'd love to get involved"**, but they haven't asked about clinical trials. I think we all need to be brave and speak to each other about research and trials.

Tonia

Tonia: Is there evidence to suggest that people from minority ethnic backgrounds have a more severe CF diagnosis?

Maya: It's a great question, and it's a complicated answer! It is true that there are some factors that affect some groups disproportionately. There are so many things that determine the course of your CF, including your genetics, your symptoms and your socioeconomic background. Because there is a disadvantage in some ways, we need to do even more to give equity of access.

Find out more

If you would like to take part in the project to increase access to clinical trials, please get in touch with our Clinical Trials team to find out more **clinicaltrials@cysticfibrosis.org.uk**.

“Be proud of your CF – don’t try to hide it.”

Kieron Smith, 26, was the face of our latest #CFTruths awareness campaign, ‘You don’t see CF’, that launched across the UK in April this year. As part of this, his face appeared on billboards and adverts to raise awareness of the realities of cystic fibrosis. We caught up with Kieron to hear more about his involvement in the campaign, his life with CF and the importance of raising awareness.

Kieron

Hi Kieron! Thank you for chatting to us. Please tell us a little bit about yourself.

Hello! I'm Kieron. I'm 26 years old, have CF, and am a qualified nutritionist and fitness instructor. My passion for nutrition translates into a love of cooking and discovering new recipes. I am also a huge Manchester United supporter and spend a lot of time in the gym and training in jiu-jitsu.

What was your childhood like growing up with cystic fibrosis?

I didn't get diagnosed with CF until I was six due to a missed test, so the early years of my life were very tough. Growing up, I wanted to fit in and be like everyone else, but it wasn't easy. I was noticeably smaller than my peers, and I struggled with confidence. At school, I always tried to hide my CF as I didn't want to appear different, but this often meant I skipped certain medications, like Creon®, out of fear of being judged.

As I got to my teenage years, I had a few chest infections requiring IV antibiotics, which made it even harder to hide my CF and led to a lot of anxiety.

> **"I wanted to fit in and be like everyone else, but it wasn't easy. At school, I always tried to hide my CF as I didn't want to appear different."**
>
> Kieron

How did you get into fitness?

My friends and I started going to the gym when we were 16 years old, and this is where I realised I had minimal muscle and was quite underweight. The gym empowered me and showed me that I have complete control of certain aspects of my life.

How does CF impact your day-to-day life now?

The biggest thing for me is keeping up with my medication and physio regime to prevent any deterioration in my health, but this can be time-consuming. The other impact on my day-to-day life is the mental health side of CF. Having CF forces me to make the most of every day, so I try to keep a positive attitude as much as possible.

What do you find most challenging about living with an invisible condition?

The most challenging thing is having to explain your condition to new people. It's so easy to hide cystic fibrosis (when I'm well), but this can leave me isolated and make me feel like no one truly understands me or my condition.

What do you think are the biggest misconceptions about CF?

The biggest misconception is that all people with CF are very ill, coughing constantly and need oxygen 24/7. Although people with CF struggle with these symptoms, CF can affect us in many other ways. The invisible side of CF – the mental struggles, digestive issues and the amount of medication we have to take often gets ignored.

Why did you decide to get involved in the awareness campaign?

I wanted to get involved because I had a story to share. And if sharing my experience would benefit the CF community or inspire even one young person with CF, then it was worth doing.

Why is raising awareness of CF important?

Without awareness, people won't understand the condition, making it more difficult for people with CF to feel understood in society. Raising awareness can also increase donations to the Trust, which can fund future trials and improve access to new, game-changing medications!

If you had one bit of advice for a young person with CF today, what would it be?

Follow your passion and be proud of your CF. Don't try to hide it or be worried about judgement.

Looking to the future, what are your hopes for the CF community?

I hope that children with CF and their parents can look to the community for support and realise that although CF is a challenging condition, there are so many positive things currently happening for people with CF.

Thank you so much for your involvement, Kieron, and for sharing your story with us.

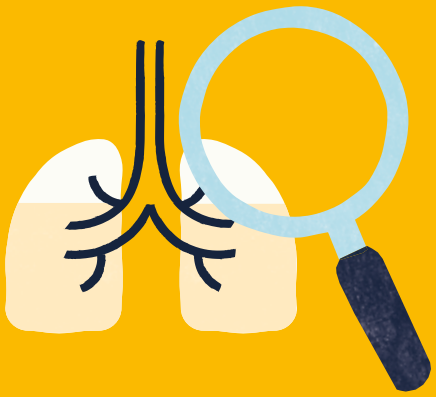
If you'd like to find out more about how you can raise awareness about cystic fibrosis, please email **stories@cysticfibrosis.org.uk**

Knowing me, knowing you

Dr Tim Lee is the Lead Consultant for the Leeds Children's Cystic Fibrosis Centre. We caught up with him to learn more about how he started his career in CF, his interest in genetic therapies and his useful barbecue tip!

Was there a moment when you knew you wanted to specialise in CF?
As a junior children's doctor, I worked on the CF ward at St James's Hospital in Leeds. There were 8 to 10 young people with CF on the ward at any one time and often for long admissions. I was inspired by their resilience and my mentor Dr Jim Littlewood's enthusiastic commitment to improving care and long-term outcomes for everyone living with CF.

How did you get involved in CF genetic therapy clinical trials?
I did a lab-based PhD in gene therapy. This led me to understand just how difficult and complex it can be to get gene therapy to work well within the lungs. I now support and review clinical trial protocols for genetic therapies for trials in America and Europe.



What would you say to someone with CF who asked you about CF genetic therapy clinical trials?
For people who don't have access to CFTR modulator drugs, the genetic therapy clinical trials now starting are potentially significant game-changers. Safety is very closely monitored, and we think there is a good chance of developing effective treatment to stabilise and improve their lung health. So please consider giving the time to join one of the trials.

Tell us something your patients or colleagues may not know about you!
Before studying medicine, I spent a year working for a barbecue company – I learnt it is worth paying the extra to get some good quality briquettes!

To find out more about CF genetic therapy trials, visit **cysticfibrosis.org.uk/genetictherapies**, or get in touch with our Trials team **clinicaltrials@cysticfibrosis.org.uk**

Easy exercise

We hear from Senior Physiotherapist Daniel Bowey, from the Respiratory Medicine (Adult Cystic Fibrosis) team at the Newcastle upon Tyne Hospital Foundation Trust, about the best ways to improve fitness levels over time.

How do we improve our fitness?

Whether you aim to increase your fitness, your general strength or a mixture of both, it's important to understand how your body responds and adapts to exercise.

When you're exercising, you're exposing your body to stress in the form of the exercise you're doing – this could be lifting weights, running or hiking. And it results in what is known as training stress.

The training stresses accumulated over a series of exercise sessions produce fitness adaptations, such as an increase in muscular strength and size, improved endurance, reduced resting blood pressure, improved insulin sensitivity and positive psychological changes.

Although exercise produces a lot of great fitness adaptations and health improvements, it also produces fatigue, muscular soreness, tiredness and the feeling of being 'run down'.

It is important to regulate your levels of fatigue so that you don't overstress your body.

How can you manage this?

Autoregulation tools. Rate of Perceived Exertion (RPE) is excellent for tracking and giving yourself feedback about how your body feels while exercising. Avoiding too much high-rated RPE exercise over a long period of time will help manage fatigue levels.

Over time your body will adapt to the stress, making the previously difficult stressors easier to overcome. This is how your body adapts to exercise.

Proper fatigue management is the basis for successful long-term exercise programming and will continue to help progress you towards your goals.

RPE Scale (Rate of Percieved Exertion)	
1	**Very light activity** (Anything other than light rest)
2–3	**Light activity** (Feels like you can maintain for hours, easy to breathe and carry on a conversation)
4–5	**Moderate activity** (Feels like you can exercise for long periods of time, able to talk and hold short conversations)
6–7	**Vigorous activity** (On the verge of becoming uncomfortable, short of breath, can speak a sentence)
8–9	**Very hard activity** (Difficult to maintain exercise intensity, hard to speak more than a single word)
10	**Max effort** (Feels impossible to continue, completely out of breath, unable to talk)

Cystic Fibrosis Trust

Laura

When Brenda and her siblings lost their sister Laura in 2012, they always wanted to keep her memory alive. Raising funds for Cystic Fibrosis Trust in Laura's name has helped them do this.

Brenda said: **"Although we only had Laura for 28 years, we will love her for a lifetime. Fundraising means we can keep Laura's memory close to us and know that we are supporting other people affected by cystic fibrosis and their families."**

If you'd like to find out more about how you can remember someone special please email our In Memory & Legacy Officer at **susan.jackson@cysticfibrosis.org.uk**

Registered with FUNDRAISING REGULATOR

Uniting for a life unlimited

Work Forwards overview

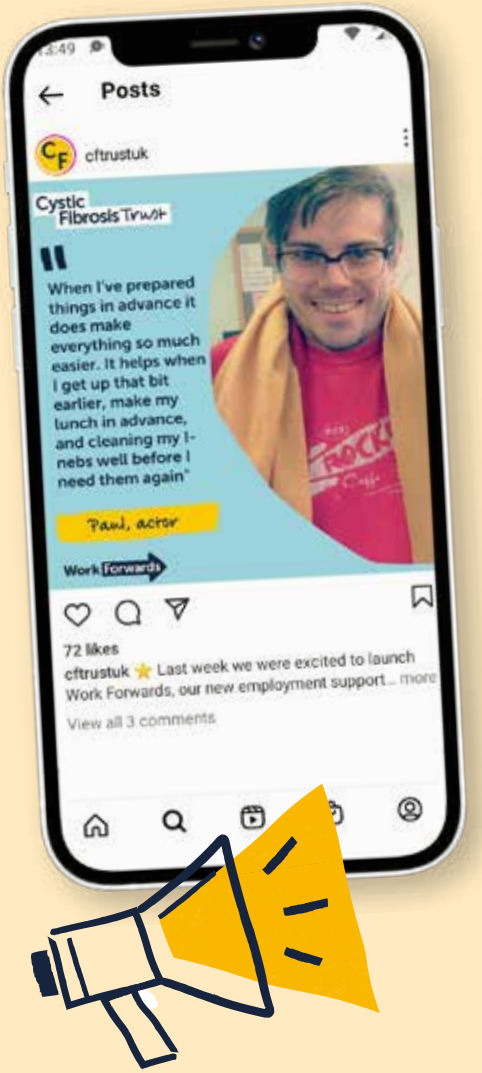
Our Work Forwards programme officially launched in February this year. Set up to respond to the evolving employment needs of the CF community, Work Forwards offers support with various work-related topics, from updating your CV to telling your employer that you have CF. Support is available to people with CF and those who care for someone with CF. Paul Warren, Employment Adviser at the Trust, reflects on our Work Forwards programme.

So far, the programme has helped a variety of people in a range of different jobs. Work Forwards has helped people with CF to apply for jobs, write CVs and cover letters, practice interview techniques and secure work.

In addition, support has been provided with challenging discrimination, negotiating reasonable adjustments, asking for flexible working and disclosing CF to an employer. The programme also allows clients to celebrate their achievements to date, talk about their goals, and discuss what isn't going well. People with CF can discuss their employment goals and issues with people who understand CF in a safe and confidential space where they won't be judged.

We have been collecting and sharing stories about people with CF in different types of work, such as 24-year-old actor Jack Norris, best known for his role in the 2021 film *Requiem*, where he starred alongside Bella Ramsey.

If you think the Work Forwards programme can support you, get in touch with us at **workforwards@cysticfibrosis.org.uk** or via the Work Forwards webpages at **cysticfibrosis.org.uk/work-forwards**. If you'd like to get involved further, you can also share your story of CF and work with us by email at **stories@cysticfibrosis.org.uk**



Jack Norris

"I need to keep physically and mentally fit, so no matter what happens or how chaotic my job is, there is a list of things for my CF that I have to do. I am very disciplined about it."

Jack

Work Forwards

Jamie

Becoming Ironman

Jamie Fox, from Hawkinge in Kent, was diagnosed with cystic fibrosis at six months old and then diagnosed with CF diabetes at 20. He has always been one for taking on challenges that scare him, but his latest challenge is his biggest yet.

After completing the London Marathon in 2022, Jamie decided in 2023, aged 37, that now was the right time to sign up and do a half Ironman.

"I hope that by doing the half Ironman, I can show others that cystic fibrosis is about ability, not disability. We all have our challenges in life and in goals we wish to reach. But it's not the size of the challenge; it's the drive to achieve it. CF is my fuel to build the fire that pushes me on to defy the odds," says Jamie.

> "I hope that by doing the half Ironman, I can show others that cystic fibrosis is about ability, not disability"
>
> Jamie

> **"When I'm out exercising, it helps me forget the challenges I'm facing with CF and allows me to feel free and enjoy the moment"**
> Jamie

Jamie's Ironman training is going well so far, and he has kept a positive attitude through it all, despite some setbacks. He has had a few injuries on the way, alongside two chest infections and a few lung bleeds, which have made training difficult.

The biggest challenge for Jamie regarding training has been finding the energy to fit everything in.

In terms of balancing his CF treatments and training, he wrote a plan early on to try and help him manage everything he needed to get done each day.

"I do have struggles some days, but I try not to panic as you can't get back a lost day. I'm not the fastest or strongest in any way, shape or form, but when I'm out exercising, it helps me forget the challenges I'm facing with CF and allows me to feel free and enjoy the moment," Jamie explains.

In the future, Jamie plans to take on the London Marathon again, as well as the London to Paris bike ride!

Jamie says that a challenge has to feel impossible for it to excite him, giving him the focus he needs to keep fighting for his health.

Thank you to Jamie and everyone who joins Team CF. We appreciate all you do to raise awareness and funds for Cystic Fibrosis Trust. If you're already taking part in an event on behalf of the Trust, or want to organise your own, get in touch with us at **events@cysticfibrosis.org.uk** or visit **cysticfibrosis.org.uk/teamcf**

Ready, set, DESIGN!

Jack Kinsey, an interior designer from Norfolk, appeared in series four of BBC Interior Design Masters, where he was awarded second place. Here he talks more about his passion for design, his time on TV, and his life with CF.

I'm Jack, I'm 27 years old, and I live in a converted 1909 chapel in Hingham, Norfolk, with my boyfriend, Alex.

I was diagnosed with cystic fibrosis a little later in life than most, when I was 16. My younger brother and I both suffered from frequent chest infections growing up and always felt quite unwell. But, it wasn't until my brother got very ill, aged 13, that we finally both got diagnosed with CF.

It was quite a shock for us and a big change, suddenly having to incorporate nebulisers and antibiotics into our lives.

It was also quite hard for me mentally. I suddenly felt immense pressure that I needed to achieve everything I wanted to in life while I was still young, especially because, at the time, we didn't have the medical breakthroughs that we have now.

Jack

> "I suddenly felt immense pressure that I needed to achieve everything I wanted to in life while I was still young"
>
> Jack

So I started to graft really hard, and that's why in 2018, with the help of my partner, I undertook my first renovation. We came across this derelict chapel which had been left unloved for so long. It had ivy growing up and leaves everywhere. But I just knew that we were destined for it and that we would bring it back to life. So that's what we did.

After finishing the chapel, Alex kept encouraging me to sign up for Interior Design Masters. I kept putting it off but eventually caved in, applied and after a few stages, I was shortlisted and got on the show.

Taking part in the show was amazing, and I feel so lucky to have come in second place.

Inside the beach chalet Jack redesigned for A&E staff

Coughy break

My favourite moment was during episode five, where I was asked to redesign a beach chalet that Eastbourne's A&E department rents for their staff to share.

The NHS has saved my life several times, and I'm very thankful for everything they do for me, so designing that beach hut felt like such a special moment where I could give back to them and communicate my appreciation for the NHS through design.

Interior Design Masters has definitely taught me that I can do anything in life that I'm passionate about.

Do you have a creative hobby or passion you'd like to share in the magazine? If so get in touch with us at **stories@cysticfibrosis.org.uk**

Understanding genetic therapies

As we prepare for the first genetic therapy clinical trials for CF to open in the UK, we've created a new information resource to help explain what they are and what trials might involve. Genetic therapies are very different to other treatments we've seen before for CF, and there can be a lot of misconceptions about them. Here we speak to people across the CF community to find out what these trials could mean and answer some of your questions about genetic therapies.

After decades of behind-the-scenes research, we'll see genetic therapies being tested in clinical trials this year. Genetic therapies work very differently from current treatments for CF. There are many different types of genetic therapy, but they all aim to make working copies of the CFTR protein. While there are no guarantees with any new trial, they have the potential to revolutionise CF treatment if they're successful.

We have already seen dramatic changes in CF treatments over the last few years with the development of CFTR modulator medicines like Kaftrio. However, nearly 10% of people with CF are unable to take them. Genetic therapies have the potential to benefit anyone with CF, including those unable to take modulators.

What does this mean for the community?

We spoke to people across the CF community to find out what these developments in genetic therapy research mean to them.

Dr Maya Desai, CF paediatrician

Dr Maya Desai works with children and young people in the Midlands and is co-leader of a project to increase access to CF research and clinical trials (see Maya's interview on page 12.)

"As professionals involved in caring for people with CF, we strive to provide equal care to enable everyone to reach their full potential. It is very important that there are alternative, highly effective treatments available to all people with CF.

It is exciting to know that there is already a lot of research work continuing in this area. Before the advent of modulators, we were able to speak positively to families of newly diagnosed babies and children about future advances. We did not know when this would happen, but that time has come. In the same way, we can now provide hope to those families whose children are not already on a highly effective treatment."

> **"It is very important that there are alternative, highly effective treatments available to all people with CF."**
> Dr Maya Desai, CF paediatrician

Rachel (right) and daughter Anna

Rachel, whose daughter has CF

Rachel's 14-year-old daughter is unable to benefit from modulators.

"It's very exciting to hear about potential clinical trials for genetic therapies. I try to stay positive that something will come along for my daughter that could be a game changer, but there are times when I can't help but feel despondent. This gives me real hope that a 'wonder' drug could help her and give her more of a 'normal' life with no limitations. Clinical trials mean that that is one step closer. "

Michelle, who has CF

Michelle, one of our trustees, has cystic fibrosis and is unable to benefit from modulators.

"I have a progressive illness that I have no control over. That is an underlying fear that never leaves me. While my CF team do a fantastic job of treating exacerbations as they arise, the underlying cause cannot be treated. The knowledge that medical science is catching up for those who cannot benefit from modulators is so uplifting.

I'm currently weighing up the risks and level of commitment associated with early-phase trials and whether my involvement is possible. Ultimately, I want to be involved, and I am keeping a close eye on progression."

Michelle

> "I have a progressive illness that I have no control over... The knowledge that medical science is catching up for those who cannot benefit from modulators is so uplifting"
>
> Michelle

Genetic therapy myth-busting

Genetic therapies are a very new type of treatment, and there can be a lot of misconceptions about them. Here are just a few of the questions we hear. You can find more FAQs at **cysticfibrosis.org.uk/genetictherapies**

Will this eventually lead to a cure for CF?

This depends on what you call a cure! Researchers hope that some types of genetic therapy (such as gene therapy or gene editing), will permanently correct the CFTR gene in 'basal cells' or 'stem cells' in the lungs. Stem cells are basic cells that make up all of the other types of cells in the lungs.

If the CF gene in stem cells is corrected, then all the cells they make will have the corrected CF gene too. This means that people would have one course of genetic therapy treatment and wouldn't need to receive it on a regular basis.

Who could they benefit? Will they only benefit people with rare variants?

Genetic therapies offer the potential of being able to treat everyone with CF, regardless of their variant. Dr Jamie Duckers, Research Lead for the All Wales Adult Centre, explains:

"There are people we know who don't have the 'right' genes to qualify for the modulators, who haven't seen a huge benefit, or who can't tolerate them. They have the potential to really benefit from these types of therapies. Beyond that, there's the hope we can improve outcomes and reach the entire CF population."

Who can take part in trials?

This will vary by trial. Some trials will target or prioritise people who are currently unable to benefit from CFTR modulators such as Kaftrio. Certain genetic therapy treatments may be specifically designed for people with certain CF mutations, whereas other treatments may be investigated across a range of CF mutations.

In terms of age groups, Jamie says:
"Usually, these therapies are started in adults and then come down the age brackets. What you'll see is it'll go down in the age groups until you get down to children."

More questions? Take a look at our new online information:

Genetic therapies glossary
Our explanations of some of the scientific words used to explain genetic therapies.

General FAQs
Find simple explanations of how genes and genetic therapies work.

Q&A videos with experts
Hear genetic therapy experts answer your questions.

FAQs about genetic therapy trials
Find out if you can take part and what might be involved.

Training resources for CF professionals
Training and information resources for research and healthcare professionals.

Head to **cysticfibrosis.org.uk/genetictherapies**

> **"We've recently extended our CTAP network to ensure we have the expertise and resources to deliver this type of trial safely"**
> Jess Longmate

Jess Longmate

How we're supporting genetic therapy trials

The CF community has told us their top 10 CF research priorities, and we know that research into genetic therapies and improving the options available to those unable to take modulators are priorities for the community.

The Trust will continue to fund early research into genetic therapies and support the delivery of trials through the Clinical Trials Accelerator Platform (CTAP).

Jess Longmate, Head of CTAP, says: "We've recently extended our CTAP network to ensure we have the expertise and resources to deliver this type of trial safely. We are also developing our Business Development Strategy to ensure we continue to have a strong pipeline of genetic therapy trials in the UK."

If you're interested in taking part:

We will list all trials on our Trials Tracker before they open at **cysticfibrosis.org.uk/trialstracker**.

You can speak to your clinical team and let them know that you would be interested in opportunities or contact one of our early phase CF trial co-ordinators.

You can also contact Cystic Fibrosis Trust clinical trials team on **clinicaltrials@cysticfibrosis.org.uk**

What's on your mind?

Our Helpline Manager Matthew answers some of your questions about life with cystic fibrosis.

I'm feeling very isolated and anxious since my son was diagnosed with CF a few months ago. He's 5 months old and doing well but I think I need some support. Can you help?

Matthew's answer: Thank you for your question. It's very normal to feel a little isolated as a new parent or carer with a child who has cystic fibrosis (CF). CF can be very complicated to explain to the people in our lives, and it can be hard to help them understand the challenges of having a child with CF. Sometimes it can feel easier to just get on with it or struggle alone.

Practically, it can also be very difficult to reach other parents or carers who have a child with CF. The risk of cross-infection means parents of children with CF cannot meet up as easily as they might like and often miss out on the chance to talk to someone who knows how they feel.

At the Trust, we have a peer support service called CF Connect, where you can speak to a trained volunteer who also has a child with cystic fibrosis. Whether your child has recently been diagnosed or you've got worries or questions further down the line, CF Connect can put you in touch with someone who understands.

Our CF Connect volunteers are all parents of children with CF, so as well as offering a listening ear, advice and support, and the chance to share experiences, they also understand the challenges you may be facing. We have volunteers across the country with children of different ages who are there to help.

If you'd like to find out more, or chat to one of our CF Connect volunteers, just contact our Helpline on **0300 373 1000**, email **helpline@cysticfibrosis.org.uk**, or WhatsApp us at **07361 582053**

Cystic fibrosis in school curriculums

Hi everyone, my name is Rosie, I am 15 years old and have CF. Today I want to talk to you about why information on CF needs to be updated across the school curriculum.

Schools across the UK often use outdated or incorrect information when teaching their students about cystic fibrosis. This is because treatments for CF are getting better all the time and the curriculum doesn't always keep up. My school was an example of this until I took action and had the content changed.

If the information is not correctly taught, it will lead to issues in knowledge about a sibling or friend of a child or teenager, and could cause them to be treated differently.

> **"If the information on CF is not correctly taught, it will lead to issues in knowledge about the condition"**
>
> Rosie

A prime example of this is airway clearance treatments. In my school and many other schools it is taught that the main airway clearance method for all people with CF is being patted on the chest and back (percussion). But in reality, there are many other forms of physio including PEP masks, the Acapella® and nebulisers.

I have also looked at my school's slideshow for these lessons and a main part of the misinformation is the effect and the results of enzyme tablets like Creon®. According to my school's lessons, enzyme tablets help convert carbohydrates into energy, when in reality, most enzyme tablets contain lipase and protease, which help break down fats and proteins. Insulin is a hormone that helps drives sugars from the blood into the cells of the body. The insulin is not taken in tablet form but is given through injections or pumps if a CF patient develops CF diabetes.

When people I know did lessons on cystic fibrosis they came out to me in tears, saying that the teachers had told them that the average life expectancy for people with cystic fibrosis was 32 years. However, thanks to new medications this is increasing all the time – some people with CF live 'til they are over 80!

They were also not taught about different variants (mutations) of CF and were taught it's all the same. They didn't go over how each variant can affect people with CF differently, so my friends thought everything that was said applied to me and every other person with CF. There are over 2,000 different cystic fibrosis genetic variants, and they are definitely not all the same.

Rosie

Need help supporting a student with CF in your class? Creating a lesson plan on CF and want to find out more about the condition? Check out our resources for teachers at **cysticfibrosis.org.uk/what-is-cystic-fibrosis/cf-for-teachers**

Through our annual schools campaign, we also provide information and guidance for teachers on how to support children with CF in their schools.

Day in the life

Bradley is 10 years old and has CF. He took some time out of his day to share his CF routine during the weekends in term time. Over to you, Bradley!

1

I start my day with physio and my nebuliser while watching one of my favourite programmes... Lego Ninjago!

2

Breakfast and my daily tablets. Creon for my food, antibiotics to keep me infection clear, vitamins and others to balance out my liver with my Kaftrio!

3

I'm very lucky that I qualify for Kaftrio!

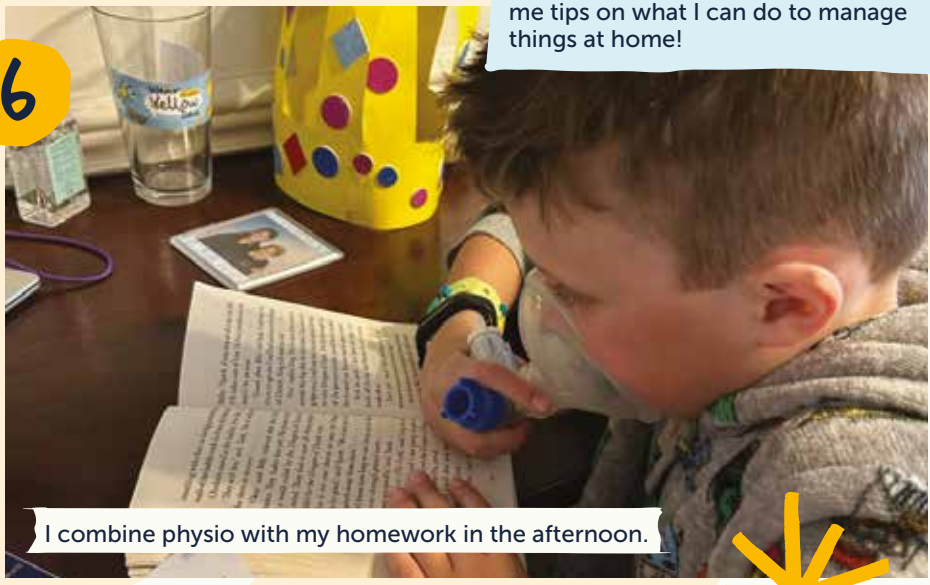
Exercise is a really good form of physio, so every other Saturday, my step-sister and I have football training with the Southampton Hospital Charity kids' team!

4

5

Having good mental health is really important, so when I'm feeling worried or anxious, I have a chat with my child psychologist Maria. We talk, colour and make pictures to help express how I'm feeling, and she gives me tips on what I can do to manage things at home!

6



I combine physio with my homework in the afternoon.

If you would like to share your story, please get in touch at **magazine@cysticfibrosis.org.uk**

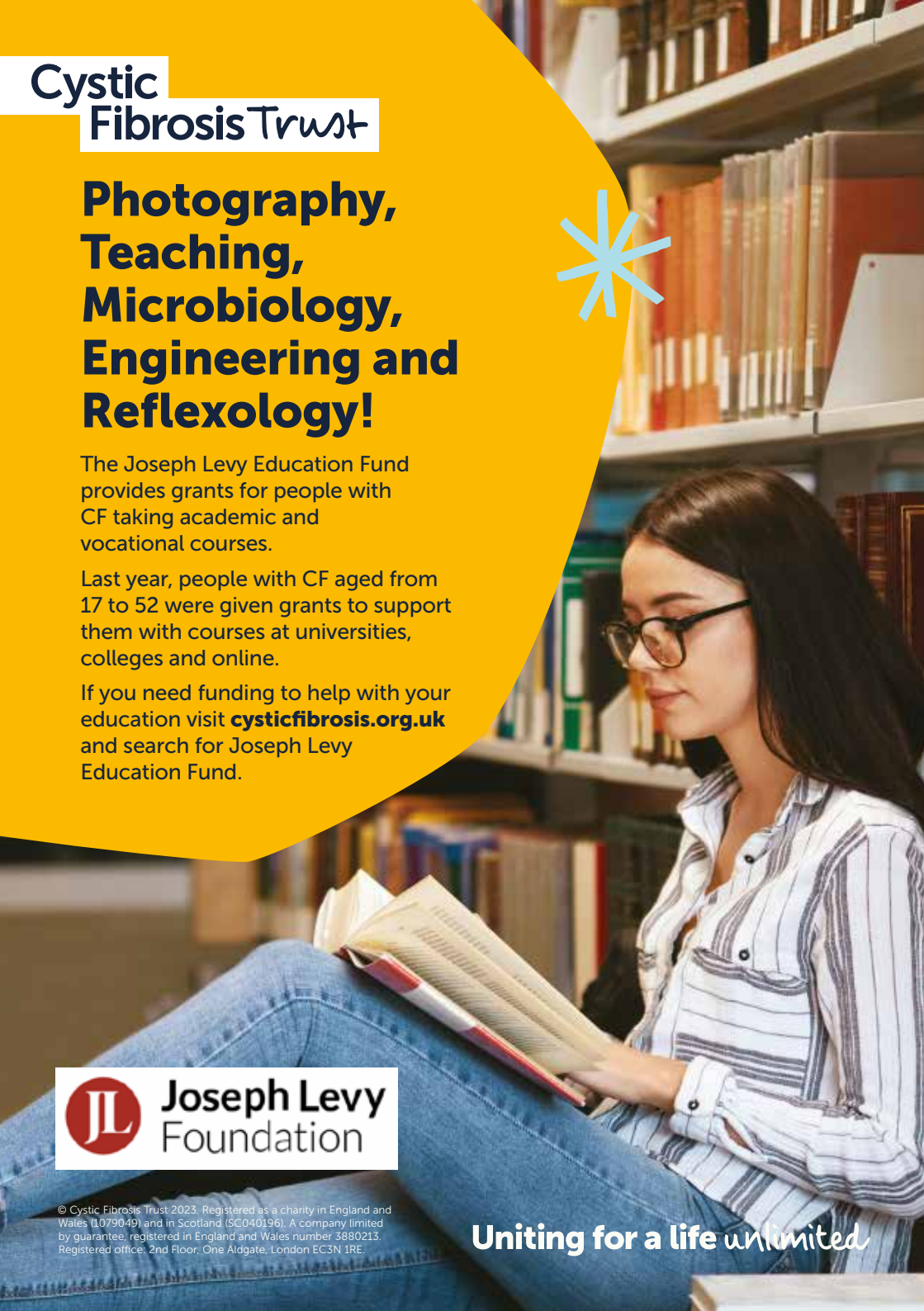
Cystic Fibrosis Trust

Photography, Teaching, Microbiology, Engineering and Reflexology!

The Joseph Levy Education Fund provides grants for people with CF taking academic and vocational courses.

Last year, people with CF aged from 17 to 52 were given grants to support them with courses at universities, colleges and online.

If you need funding to help with your education visit **cysticfibrosis.org.uk** and search for Joseph Levy Education Fund.



Joseph Levy Foundation

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