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CF Life

Issue 3

Focus

Heelprick test: Now screening

Coughy Break

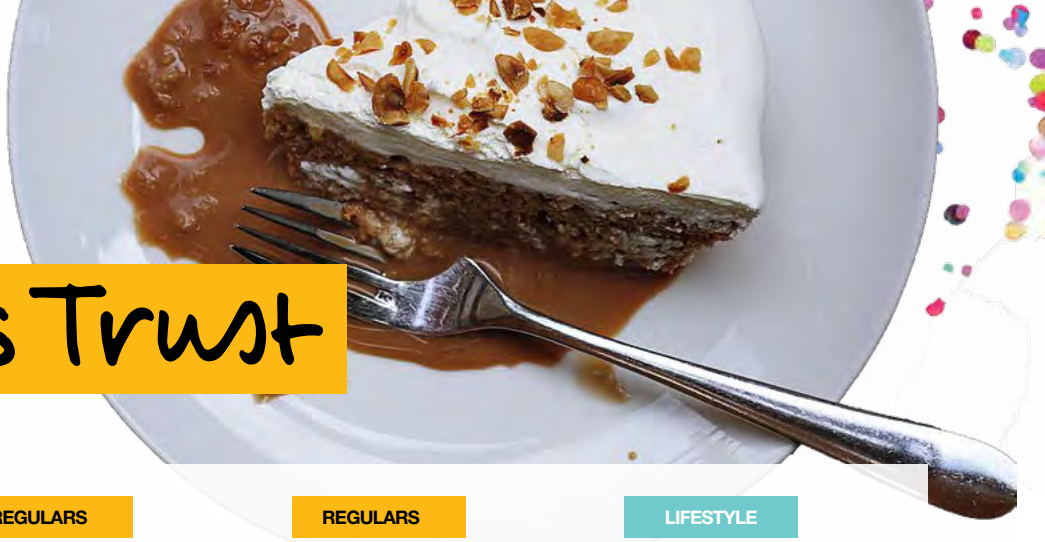
No woman is an island

Lifestyle

From motorbikes to MMA

Fighting for a Life Unlimited

Cystic Fibrosis Trust



What's inside

Issue 3 – September 2017

REGULARS

4 News
Cystic fibrosis in numbers - the UK CF Registry report.

5 News in pictures
Some of our highlights from the last six months.

11 Fly on the wall
Meet the people behind clinical trials.

14 Banding together
A novel way to fight cross-infection.

16 Spotlight
Six mums take Great Strides for the Trust.

REGULARS

10 Warrior not worrier
Lucy Taylor talks picking yourself up after bad news.

15 Fly on the wall
Meet Rachael Bass, our first ever Physiotherapy Fellow

18 Coughy break
Artist with CF, Kate, tells us about her latest project.

26 Cheering champions!
The marathon supporters who go the extra mile!

30 Day in the lives
Catch up with our Youth Advisory Group.

LIFESTYLE

20 Handy hobbies
Meet a motorsport engineer turned Bright Ideas Award winner.

28 Easy exercises
Warming up the Mixed Martial Artist way.

FEATURES: TWO TURNING TEN

6 Now screening
We explore the impact of the move to national newborn screening for cystic fibrosis.

22 The Registry: 10 years on
One decade, a lifetime of change.

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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

It's your birthday(ish)!

A year ago this month we launched CF Life, and want to say thank you for all your help in making it what it is. As you'll see on page 27, we are hoping to go a step further with our new editorial panel. Check it out and see how you can join the team.

But it's not about us, and in this issue we're looking at some bigger anniversaries. Our main feature marks the 10th anniversary of newborn screening for CF becoming compulsory across the UK. It took a long, hard-fought campaign, but the CF community pulled together to achieve one of the most significant advances in clinical care in the UK.

It's also 10 years since the Trust took over the running of the UK CF Registry, which has grown to be one of the most widely respected, comprehensive patient registries anywhere in the world. On page 22 we'll bring you up to date with some of the big moments in the story of the Registry so far, and how it supports breakthroughs in care and research. And you can check out some of the latest Registry data on page 4.

And that's not all. In this issue you'll meet the motorsport engineer with a dream, the intrepid mums who took Great Strides to make a difference, and we'll find out just who's who in clinical trials.

We hope you like this issue, and are as excited as we are about the future – get in touch and let us know at magazine@cysticfibrosis.org.uk.

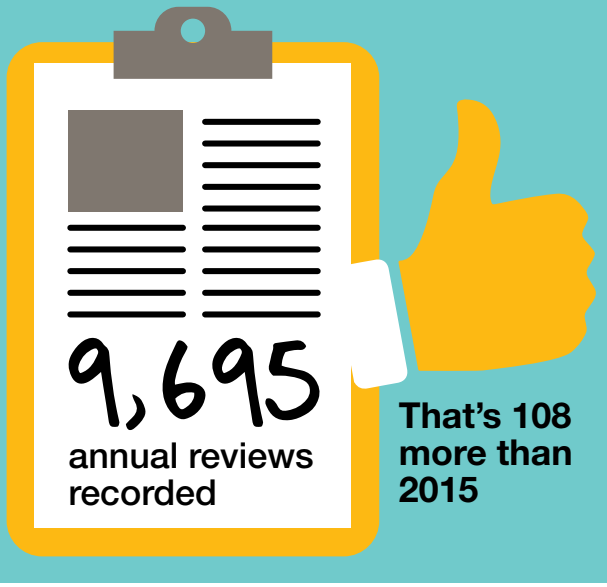
The CF Life Team

Cystic fibrosis in numbers: Registry report highlights

The UK Cystic Fibrosis Registry is a secure centralised database, sponsored and managed by the Cystic Fibrosis Trust. Registry data is used to improve the health of people with cystic fibrosis through research, to guide quality improvement at care centres and to monitor the safety of new drugs.

Our 'at a glance' version of the UK CF Registry Annual Data Report 2016 highlights the key information from the full report available at **cysticfibrosis.org.uk/registryreports**.

Annual Reviews



9,695
annual reviews recorded

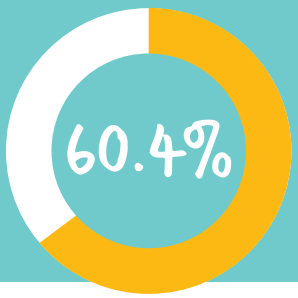
That's 108 more than 2015

Keep up to date

The latest news from the world of cystic fibrosis, keeping you up-to-date and in-the-know. Sign up to receive our newsletter 'CF News'.

cysticfibrosis.org.uk/newsletter-signup

Population age



of people on the Registry are aged 16 and over.

This is the first time it has been over 60%

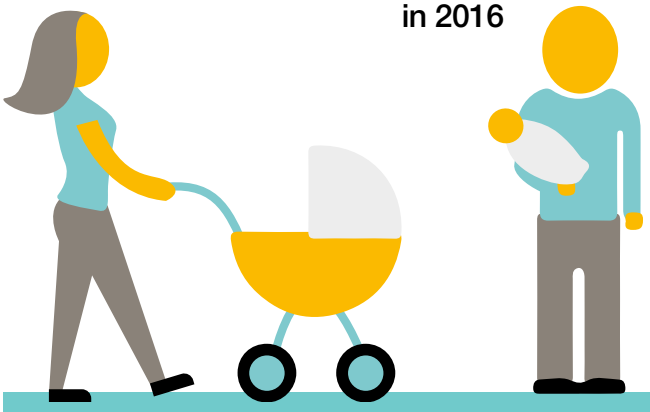
Physiotherapy

57%
of people with CF include exercise as part of their physiotherapy, up from 37% in 2015

Pregnancy

71 women
with cystic fibrosis had babies in 2016

48 men
with cystic fibrosis became fathers in 2016



221 people with CF were evaluated for transplant in 2016, and 96 were accepted onto a transplant waiting list

News in pictures

Mammoth marathon marriage proposal

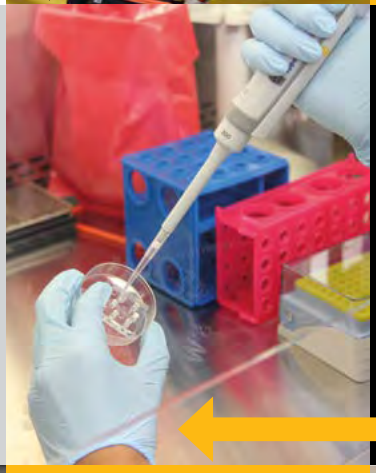
Over 130 runners took to the streets for the Virgin Money London Marathon, but Jon Higgins made it extra special by proposing to his girlfriend Amy before crossing the finishing line. The Facebook video was viewed over 500,000 times (spoiler – she said yes!).

Triple trial therapy

Drug developer Vertex announced encouraging results for their next generation of precision medicines. While it's still early days, if larger trials prove successful this treatment could benefit around 90% of the CF population. Phase III trials should begin in 2018.

What do we want?

Our incredible community set up camp at locations across the UK to shout loud for access to Orkambi. Supporter Sadie Dann handed a petition into the drug's manufacturer, Vertex, while Trust CEO David Ramsden sent letters to Vertex and the NHS in all four UK nations.

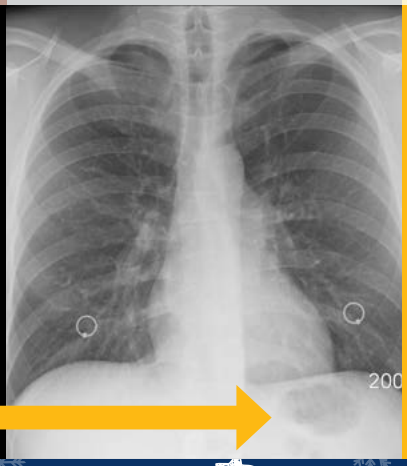


Three new SRCs

We announced three new Strategic Research Centres. These £750,000 projects include finding new ways to tackle *Mycobacterium abscessus*, working to prevent blockages in the gut, and looking at the link between gut bacteria and lung inflammation.

Transplant victory

Since 2014 over 10,000 of you have lent your voices to our Hope for More campaign to call for fairer access to transplant organs, and it was announced in June that the current regional allocation of donor organs will change to a fairer national system for the most urgent cases.



Hello yellow!

This CF Week you joined the most successful Wear Yellow Day yet by setting up fundraising events and sharing their #CFYelfies! We had loads of celebrity support, including Yelfies from Jenny Agutter, Bill Bryson and Evanna Lynch.

Our Christmas cards and gifts make a real difference

Buy yours today!

cysticfibrosis.org.uk/cardshop

Nearly **£25,000** was raised last year to make a difference to children and adults living with cystic fibrosis.

Look out for cards designed by the winners and runners up of our Children's Card Competition! (Open now!)

Fighting for a Life Unlimited

Now screening...

On the 10th anniversary of the newborn screening programme for cystic fibrosis (CF) being implemented nationwide, we look back at the way things were, and what earlier diagnosis means for people with CF and their families.

Today most babies are tested for CF and other rare conditions at around five days.

Ouch! You're five days old and some horrible person has just put something sharp in your foot, and made you bleed. You don't know it, but of all the people you will meet, this blurry shape with the spiky thing may have had one of the most fundamental influences on your life.

In the UK, newborn babies have a blood sample taken by pricking the heel, and this sample is then screened for signs of nine rare but serious health conditions. Since 2007 CF has been one of them, and a positive diagnosis means treatment can begin straight away, before major respiratory and nutritional problems can start.

"The introduction of national neonatal CF screening was undoubtedly one of the major clinical advances, if not the major advance, of the decade in the UK."

Dr Jim Littlewood, OBE, President of the Cystic Fibrosis Trust, dedicated his career to CF care. A key player in the campaign to make screening for CF compulsory, Dr Littlewood believes that alongside the introduction of CF centre care, newborn screening is one of the major factors for continuing improvement in health and survival in people with cystic fibrosis.

"The introduction of national neonatal CF screening was undoubtedly one of the major clinical advances, if not the major advance, of the decade in the UK."

– Dr Jim Littlewood, OBE

Testing testing

Let's go back in time, when newborn screening was in its, ahem, infancy. In East Leeds where Dr Littlewood worked, neonatal screening began in 1975 using a precursor to the heelprick test, called the Boehringer-Mannheim (BM) test. The BM test looks for a protein called albumin in meconium, the first stool a baby produces, made up of materials ingested in the womb.

As with most new procedures, introducing the test was a learning curve itself. Dr Littlewood recalls: "After repeatedly finding small pots of meconium on ward window sills, I soon realised that overworked midwives were not the people to do this test! The laboratory kindly offered to do the tests with proper laboratory standard control so meconium specimens were sent to them in small bottles."

In 1979, Crossley and Elliott designed a superior test that looks for elevated levels of IRT, a pancreatic enzyme, in dried spots of blood from the heel-prick. However, while neonatal CF screening enthusiasts embraced the new diagnostic tool, they faced opposition from the majority of public health specialists. It would take 22 years of research and campaigning before CF would be added to the conditions routinely looked for in the heel-prick test, and a further six to fully implement the change across the UK.

"Parents often had a miserable time before the diagnosis was finally established, with feelings of inadequacy and guilt at a time when they should be enjoying their new baby."

– Dr Jim Littlewood OBE

Rt Hon Norman Irons, Lord Provost of Edinburgh giving his support for newborn screening to be available to every family in Scotland.

Screaming for screening

Joining the Trust as Chief Executive in 1996, former MP Rosie Barnes worked closely with Dr Littlewood to win hearts and minds for neonatal CF screening. The campaign was an uphill battle, with ambiguous clinical evidence at the time, despite what CF clinicians and families across the UK knew to be true.

Rosie recounts: "I started work on this initiative soon after taking up my post, by writing to the Prime Minister, Tony Blair, who referred me to the Minister for Health, Yvette Cooper, who happened to be pregnant herself at the time Jim and I met her.

"The implementation of the programme was almost certainly considerably delayed by Public Health Specialists, who were strongly opposed to the introduction of the heel prick test for cystic fibrosis."

In an article on the history of newborn screening on cfmedicine.com, Dr Littlewood wrote: "Previously the Child Health Subgroup of the UK National Screening Committee had considered the evidence of long term benefit was insufficient to recommend national neonatal CF screening, including a paper from Wisconsin which was considered by Professor NJ Wald, to "provide no evidence of any benefits of screening."

Fortunately, the Wisconsin team ran a follow-up trial published in 2001, demonstrating significantly better long-term growth and nutritional health in the screened group. The trial helped win over the UK Government, bringing a happy end to what Dr Littlewood describes as "a seven-year, at times traumatic and heated campaign."

Why does it matter?

A delay in diagnosis can mean increasing malnutrition, with both short and long-term implications for the child's resistance to infection and growth. A chest infection is almost inevitable, which can quickly become irreversible if left untreated, leading to progressive lung damage. This marks the start of a downward spiral to what clinicians term 'the point of no return', and an early death.

Mum Katie Gallagher recalls the impact that treatment had on her son Lewis, when he was diagnosed: "Within days of getting him on all the medications, he filled out, he was perkier, he could hold his head up, he could look round, he was bright... He was as a baby should have been, it was amazing."

Katie shared her experiences in the support pack the Trust developed for parents when they receive a diagnosis for their newborn. It is important to recognise that newborn screening isn't just a benefit for the child. In the past families faced agonising periods of stress not knowing why their child failed to thrive. Today they can access support straight away.

Gloucester MP Tess Kingham celebrates the success of her campaign to test all newborn babies for cystic fibrosis.

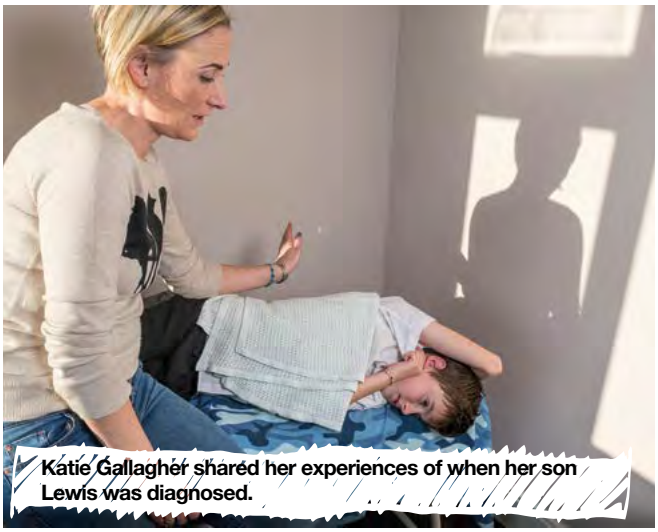
Dr Littlewood sums it up: “The parents often had a miserable time before the diagnosis was finally established, with feelings of inadequacy and guilt at a time when they should be enjoying their new baby. Sadly they had often been repeatedly reassured by their family doctors that there was no serious problem and even occasionally by a paediatrician.

“Seeing so many children and their parents where there had been late diagnosis, accounts for my strong uncompromising support for neonatal CF screening and CF centre care.”

The Gale family from Essex took part in the Trust’s campaign after a five-year battle to diagnose their son James. During that time, James underwent two unnecessary operations, one of which resulted in losing the hearing in one ear.

James’s dad Stephen says: “James first became very ill with an ear infection. He ended up having a mastoidectomy [a procedure to remove diseased air cells from the mastoid bone, part of the skull behind the ear]. A year later he was very poorly, his weight wasn’t good and we were referred to a paediatrician who, after a series of tests, including a sweat test, diagnosed James with cystic fibrosis.”

Some years later doctors would question this diagnosis because of James’ high level of health. It is possible James has a rarer and less severe CF-causing genotype, but Stephen hopes that families now will be spared the uncertainty they experienced: “For today’s parents, I hope there is a more definitive diagnosis.”



Katie Gallagher shared her experiences of when her son Lewis was diagnosed.

> “Seeing so many children and their parents where there had been late diagnosis, accounts for my strong uncompromising support for neonatal CF screening and CF centre care.”
>
> **– Dr Jim Littlewood OBE**

Knowledge is power

A further result of the newborn screening programme is families finding out about CF much earlier. One in 25 of us carries the faulty gene that causes CF, but most don’t even realise it. If two gene carriers have a child, there is a one in four chance that their baby will have CF, and studies show that the introduction of screening can result in incidences of CF falling.

It may be knowing why your baby is unwell, and how to care for them. It may be reaching out to people who have walked the road ahead, or knowing your options before starting a family. Whatever it is, newborn screening gives people power, and hope for a brighter future.

For more about the campaign for newborn screening, visit cysticfibrosis.org.uk/screeningcampaign

Visit Professor Daniel Peckham’s Leeds website www.cfmedicine.com for a dedicated history of cystic fibrosis by Dr Jim Littlewood.

How to be a warrior not a worrier

Life is full of ups and downs. But sometimes when you have CF it can feel like there are more downs than ups. What about hearing your lungs aren't doing as well as you thought? Here Lucy Taylor talks about coming back from clinic, and searching for the "head held high moment".

When it's time to make the journey to clinic, it's always a nerve-wracking feeling as I'm entering the hospital.

But when you're feeling well in yourself, those nerves seem to vanish from within and you walk into clinic with a little confidence and with your head held high for once.

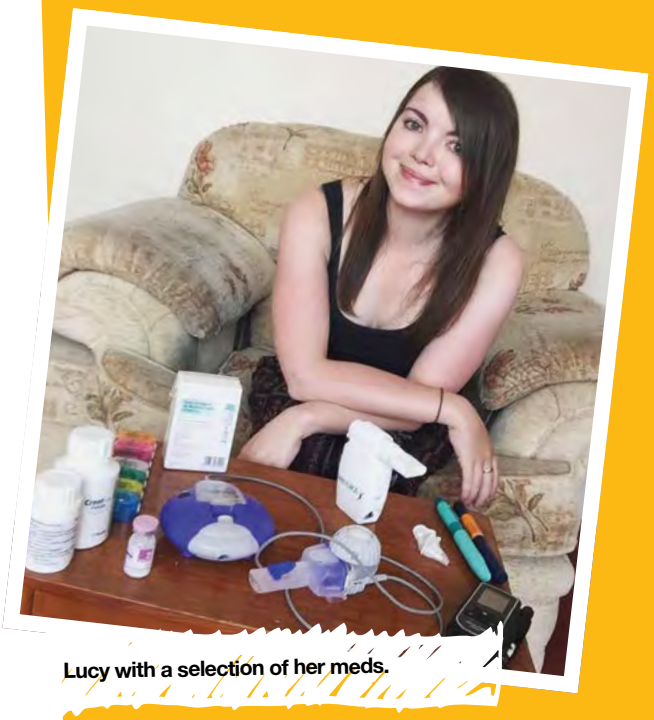
Doing a lung function test is always a very nervy time during clinic; you don't know when CF will decide to be a pain in the bottom and not let your lungs blow the best lung function that reflects how great you have been feeling lately.

So, when you do the lung function test and you look at the numbers afterwards and they're not how you think they should be, it is like 'a crash down to earth with a bump' moment. Truly frustrating and even saddening. You start to think that all the hard work with the much-needed vigorous sessions in the gym and the 20 minutes of physiotherapy blowing through the Aerobika/Acapella every day, or whatever you use, was just a complete waste of time. I start to question if it really is all worth it, after all CF is a progressive illness, it won't get better for us.

But after a day or two of sulking and moping about the lung function result, you've got to pick yourself back up and become the permanent warrior you were born to be and use that fighting spirit that's deep inside you to get your lung function back to where it should be. I always think that it's just a blip and that I'll keep working hard because I don't ever want cystic fibrosis to beat me!

Aim for the 'leaving clinic skipping and with your head held high' moment!

Share your own ideas on how you pick yourself back up in our Forum at cysticfibrosis.org.uk/forum



Lucy with a selection of her meds.

"I always think that it's just a blip and that I'll keep working hard because I don't ever want cystic fibrosis to beat me!"

– Lucy

Guess Who?

In recent issues we've talked about clinical trials and the Clinical Trials Accelerator Platform, part of our commitment to stopping the clock on cystic fibrosis. There are lots of key people who make trials happen - but what do they all do?

We invited five people involved in various aspects of clinical trials to tell us a little bit about what they do, and what they like most about their job.

So who is…

The Research Nurse?

The Trial Coordinator?

The Chief Investigator?

The Data Manager & Information Analyst?

The Co-lead?

A

"I provide support to my clinical colleagues to get new projects started quickly. I also help to streamline processes to ensure we collect high quality data whilst minimising the burden of additional paperwork on patients and my colleagues."

B

"I play a central role in facilitating the annual review process and data reporting onto the CF Registry, and assisted with data reporting for the CF Gene Therapy Trial."

C

"I work as a consultant at one of the lead centres, coordinating the trials at that particular centre, making sure the trials are set up and run according to best practice."

D

"I have overall responsibility for the set-up and running of the trial at our centre, ensuring that we reach out to all eligible patients and families under our care throughout the network."

E

"I am responsible for the day-to-day coordination of clinical trials. This involves setting up the trial, identifying and recruiting patients and supporting them throughout their time on the trial."

Guess who: how many did you get right?

Hannah

A The Trial Coordinator

Hannah Baber,
University Hospitals Bristol NHS Foundation Trust

I have worked as a Trial Manager for over a decade and I hope to use my experience in my new role as Trial Coordinator to ensure that new studies are set up quickly to minimise delays in patients benefitting from the treatment regimens being offered.

The best part of my job: I get to hear about exciting research developments and make a difference to people's lives by helping them access treatments that may not routinely be available.

B The Data Manager & Information Analyst

Dawn Wilson,
Scottish Adult Cystic Fibrosis Service

I am delighted that our Edinburgh CF Centre has been selected for the Clinical Trials Accelerator Programme. This will provide greater access to CF trials for our large geographical service in Scotland.

The best part of my job: Doing all our publications for the service, like leaflets and newsletters, and we have a brand new CF Data Registry which I enjoy keeping updated.

Dawn

Visit cysticfibrosis.org.uk/trialsaccelerator to find out more about our Clinical Trials Accelerator Platform.

Kevin

C The Chief Investigator

Professor Kevin Southern, University of Liverpool

I am the Chief Investigator on CF START. The team are a tremendous group of talented people, ranging from parents and patients through to scientists and clinicians.

The best part of my job: The enthusiasm of families affected by CF to contribute and make a difference.

D The Co-lead

Maya Desai, co-lead of the Birmingham Trials Accelerator Centre

I have been the Principal Investigator for the TORPEDO study, and the CF START study which we are just embarking on. The Trials Accelerator will enable us to run more trials, enabling patients and families in the West Midlands who are interested in advancing our understanding of new treatments to take part.

The best part of my job: Having the ability to bring about studies which patients can participate in safely to help us answer important questions, to advance the health of the patients we care for. It is amazing how children and families are so keen to help.

Maya

Kim

E The Research Nurse

Kim Doolan, Alder Hey

My role is very varied, when I think of all of the steps involved! I also work closely with the consultants to ensure that the trial is carried out safely.

The best part of my job: The determination and dedication of children and their families to participate in often lengthy, time-consuming trials – it is often difficult to say goodbye to them when they have completed their time on the trial!

Now you've been introduced – get to know our experts better at cysticfibrosis.org.uk/trialteam.

Banding together to battle cross-infection

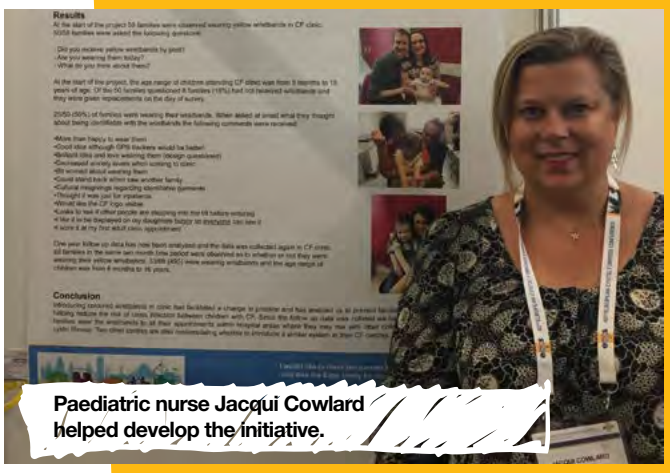
You know you can trust your specialist CF clinic to have proper cross-infection controls, but what about the rest of the hospital? Jacqui Cowlard, a paediatric nurse at the Royal London Hospital, explains how a wristband initiative she introduced is making people safer.

Cross-infection is an issue in CF care that we know a lot about, and we're careful to reduce the risks during clinic visits. However, families have expressed concerns over the risk of cross-infection in non-clinical areas like outpatient clinics, play spaces and eating areas.

We issued families of children with CF yellow silicone wristbands to wear, making it easier for families to identify each other and reduce the risk of cross-infection. The feedback has been positive and a lot of families are still wearing their wristbands two years later!

"Our daughter Erin was born premature and underweight, and was three weeks old when we first took her to the clinic. Being so young Erin was at risk from cross-infection from any older child, so we were very paranoid about her coming across anyone else. The wristbands gave us a discreet way of identifying each other that really put our minds at rest at a very stressful time."
- Gillian and Adrian

"Our CF team at the Royal London Hospital are scrupulous about cross-infection, but there's always a chance that kids with CF might accidentally meet. Now, if you see a family wearing the yellow wristbands, you can choose to avoid each other. It is a sensitive subject - some people can feel uncomfortable being visually identifiable. Ultimately though, we feel this is about working together as a community to help all of our children stay healthier for longer." - Hannah and Tom.



Paediatric nurse Jacqui Cowlard helped develop the initiative.


Gillian, Adrian and baby Erin


Griffin and his parents Hannah and Tom, who helped fund the wristband initiative.

Meet... Rachael Bass

Earlier this year we launched our Physiotherapy Fellowship Programme to help improve opportunities for physiotherapists to undertake careers in CF care, and enable them to lead ground-breaking CF services in the future. We spoke to our first ever Physiotherapy Fellow, Rachael Bass, to get her answers to some of your common physio questions.

Rachael Bass

"Aerobic exercise can be used alongside airway clearance techniques, but may not be enough to clear the airways on its own, and cough and huff techniques should be used alongside it."

Why is the Vest not widely used in the UK?

"We don't recommend that high-frequency chest wall oscillation devices like the Vest are used as the sole method of airway clearance. The Vest has been shown to be as effective or, in some cases, less effective in comparison to other airway clearance techniques such as Positive Expiration Pressure therapy (PEP).

"In recent research, more frequent exacerbations were noted with the Vest when compared to PEP over a sustained period of time. While we recognise the Vest as an option, given the associated high financial burden, with no proven additional benefits it is not something we recommend."

What airway clearance techniques are available?

"There are many different options available, and you should take time to discuss these with your physiotherapist to find out which is most appropriate for you. Treatments include: PEP, Aerobika, Acapella, Flutter, Active Cycle of Breathing, Autogenic Drainage, Postural Drainage, Intermittent Positive Pressure Breathing, Cough Assist and Non-Invasive Ventilation."

Which airway clearance techniques are the most effective?

"There has been a lot of research into this question, but there is little to suggest that one technique is better than another. As physiotherapists, we try to ensure we are providing the best evidenced-based treatments for our patients, and often that comes down to how these treatments suit the individual.

"We get the best results from our patients by fostering good relationships and creating treatment plans together."

Can exercise replace other airway clearance techniques?

"Aerobic exercise can be used alongside airway clearance techniques, but may not be enough to clear the airways on its own, and cough and huff techniques should be used alongside it. Some key points to remember are:

- Exercise daily for at least 20 minutes.
- Ensure you are exercising at the right intensity, working to a level where you are out of breath."

Keep an eye out for an update from Rachael on her fellowship!

Taking Great Strides together

This summer six mums joined forces to face one of the hardest fundraising challenges in the Trust's calendar – despite never having met face to face!

Together for Their Tomorrows

"We all met on social media," says Esther Valentine, mum to two-year-old Joshua, "and just hit it off from there – since then we have talked every day."

Esther lives in Oxfordshire, a long way from her Together for Their Tomorrows teammates: Elizabeth Brennan (Bristol), Kath Barclay (Kent), Claire O' Grady (Essex), Emma Corr (Newcastle) and Emma Hughes (Telford).

The sensational six all have children with CF, and are used to supporting each other through CF's challenges. As Elizabeth says, receiving that first diagnosis can be a shattering experience:

"When you find out that your newborn baby has cystic fibrosis, your world comes crashing down. How long will they live? Will they live in pain? Will they go to school? Everything becomes uncertain."

And that's when the strongest friendships are formed.

"As a group we are truly a rock and inspiration for each other," says Esther. "We support each other through the tough days, celebrate our children's achievements, share ideas on physio and discuss treatments and research, as well as fundraising and awareness ideas."

Prompted by Emma Corr, and their desire to make a difference for their children and thousands across the UK, the group decided to take on a challenge. (Sadly Emma C was unable to do the walk in the end, due to injury.)

"As a group we are truly a rock and inspiration for each other. We support each other through the tough days, celebrate our children's achievements, share ideas on physio and discuss treatments and research, as well as fundraising and awareness ideas." – Esther

Great Strides, great group!

Emma Hughes was the brains behind these fab leggings, which became a must-have fundraising item.

#WalkingWins!

Great Strides 65 is an endurance event, with teams setting out to walk (or even run!) a 65km course in 18 hours. Established in Surrey, there are now Great Strides events in the West, North and East too.

The ladies set about training individually, and organising all manner of fundraisers, from bake offs to book sales. They also made a series of Facebook videos to raise awareness of CF – one of which attracted more than 6,000 views alone!

It all paid off in June when the team crossed the finish line, raising over £20,000, with Gift Aid. And how did they feel? "Wow, we did it!"

An endurance walk has both literal and emotional ups and downs. How was it for Together for Their Tomorrows?

Highlights:

- Meeting for the first time! The time flew by because we had so much to talk about.
- Reaching the top of Martha's Hill.
- Liz running "because it felt better than walking!"
- Donations coming in throughout the walk with amazing messages of support.

And the lows?

- Climbing Martha's Hill!
- So many blisters – ouch!
- Walking in the dark wondering if we would get there (but we did!)

So what's next?

"Potentially another Great Strides event next year…?"

Coughy break, shining a spotlight on the talented, creative side of the cystic fibrosis community.

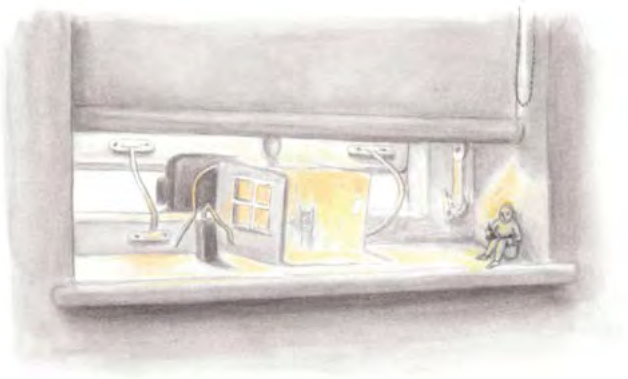
No woman is an island

Kate Hughes, 32, has cystic fibrosis and has been working as an artist for almost 10 years. For her project, 'Island Collaborations', she got in contact with people with CF staying in the Royal Brompton Hospital, who sent her photos and recordings from their hospital rooms. She used these to create drawings and animations that explore the experience of spending time alone in a CF ward.

Kate says: "I was very interested in the idea of parallel experience; where you don't meet your neighbours because of the cross-infection risks, but you're going through something very similar. The participants' comments were quite touching. A lot of people said that it was nice to feel like there was someone else there, in the same situation."

'Sleeping Visitor - March 2016'
Ink on paper

"Debbie's photo of her husband asleep next to her seemed so similar to a photo I had sneakily taken of my dad in 2014. The echo fitted so well with the theme of this project – spaces inhabited by people with so much in common, who never meet each other."

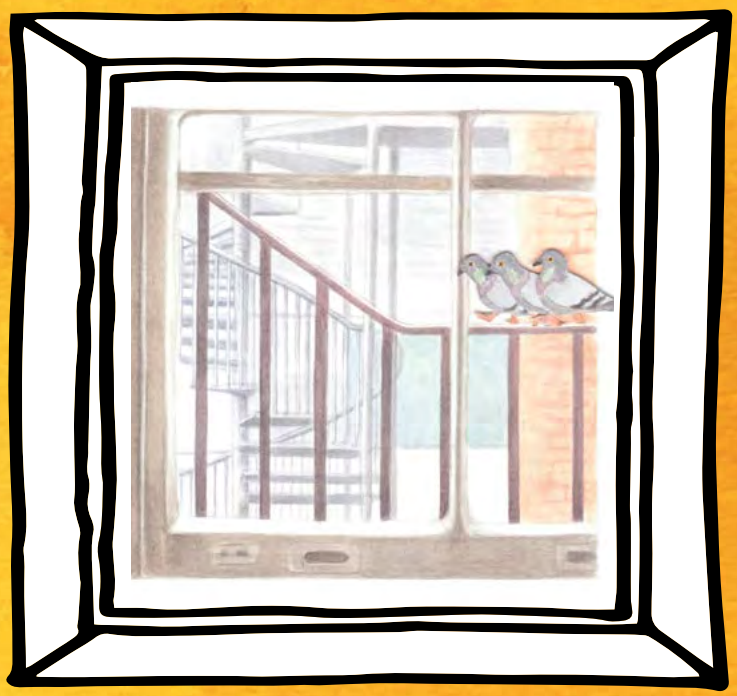


Still A from 'Smaller Worlds'
Drawn film, acrylic and cut paper
Watch the video: goo.gl/1nQHwM

"Claire sent me pictures of the doll's house she made and put in her window during her stay. I imagined it as a parallel for the individual rooms patients inhabit with no contact with their neighbours due to the cross-infection rules that mean they communicate only by virtual means."

Still B from 'High Windows'
Drawn film, acrylic on paper
Watch the video: goo.gl/YqgMBT

"I asked participants to take photos of the view from their bed. Sometimes you can look out of your window and wonder how many hundreds of times the previous and subsequent inhabitant of your room saw or will see the same subtly differing view."



Still from 'Debbie's Visitor'
Drawn film, acrylic and cut paper
Watch the video: goo.gl/TZ9pAf

"I made a short video from a photo Debbie took of a curious pigeon outside her window – a minor event that, if you were getting on with your daily life, would go unnoticed."

Tell me more

www.katehughesart.com
www.islandcollaborations.wordpress.com
facebook.com/katehughesart

See more of Kate's work at the Poetry in Aldeburgh festival in November.

Kate's project was facilitated by rb&hArts with the kind permission of the staff of Foulis Ward. It was made possible thanks to support from rb&hArts, The Royal Borough of Kensington & Chelsea Arts Grant Scheme, InTRANSIT Festival, and the Cystic Fibrosis Trust's Helen Barrett Bright Ideas Awards.

Gearing up

Chris Frappell is a motorsport engineer and the founder of Tru-Tension, a company that manufactures motorcycle tools and care products. He was also one of the lucky recipients of our Helen Barrett Bright Ideas Awards, which he used to fund a trip to the United States to promote his tools on the American market.

We spoke to Chris about his incredible journey and what the future holds for Tru-Tension! Chris' incredible journey to where he is now has been down to more than just luck!

I started showing an interest in engineering around the age of five. At eight I started racing karts and dismantling anything mechanical that I could find – most impressively my grandparents' lawnmower engine, which didn't go down too well when they wanted to mow their lawn and I couldn't work out how to put it back together!

When I was a bit older I became interested in kart racing, and by 16 I had raised enough money through three part-time jobs to enter myself into a national kart racing championship. I was seeded 18th in the country, which was a huge achievement as I'd worked so hard to get there with no financial backing.

Chris Frappell

"My business plan for the future is to become a household motorcycle brand. It's going to be a very long journey but one I hope I can continue on for a long time."

– Chris Frappell

I designed my first product, Chain Monkey, after hearing my mechanic using some very choice words when trying to achieve the correct chain tension. I spent 18 months and a lot of my own money developing the same product for the motorcycle industry, and following the launch of it in 2015 the product went from strength to strength. I left my job to concentrate my time on my growing business, and in a year I've expanded the brand to nearly 14 products.

My dad and friends were keen that I applied for a Bright Ideas Awards so that I could fund my trip to the USA and help demonstrate what can be done even when coping with cystic fibrosis. The trip was a huge success, and I'm hoping to receive an order and secure my brand in the American market. All of this would not have been possible without the Bright Ideas Award, and I will always be grateful for the amazing opportunity.

My business plan for the future is to become a household motorcycle brand. It's going to be a very long journey but one I hope I can continue on for a long time. I sold everything motorsport-related I had to start my business, so hopefully in the future I can return to what I love doing: racing.

Find out more about Chris' company by visiting www.tru-tension.co.uk. Do you have cystic fibrosis and a hobby you'd like to share with us? Email magazine@cysticfibrosis.org.uk.

The Registry: 10 years on

A lot has changed in the 10 years since the Trust began managing the UK Cystic Fibrosis Registry, a database that records the health and wellbeing data of consenting people with CF across the UK.

Median predicted survival of people with CF has risen from 35 to 47 years old, median age of diagnosis has dropped from over five months to just 26 days, and chronic Pseudomonas infections have reduced by over 15% in people with CF aged 16–19*.

Registry data doesn't just show us how CF has changed, it also helps people with CF and their families take control of their lives by making information available that can help benefit their health and wellbeing. In a survey of almost 900 people in 2015 it was found that Registry results support people with CF and their families to choose treatments, understand their condition and have meaningful discussions with their CF teams.

So, without further ado, here are the big changes to the Registry and the lives of the people with cystic fibrosis that have been recorded within it over the last 10 years.

2007
The Trust begins hosting and maintaining the Registry.

2007

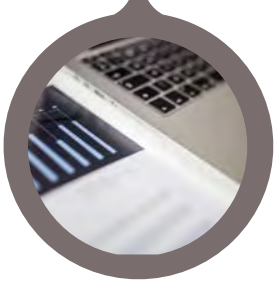
2007
The first Annual Data Report by the Trust calculates a median predicted survival age of 35.2 years, significantly higher than the quoted age at the time of 31 years.

Not only that, but in the decade since the Trust began managing the Registry, the data it produces has helped to inform research, assist CF centres and influence policy and standards of care, and has become self-funding through its support for the NHS and generation of drug safety reports.

*Based on 2015 Registry Report data.

2008
Over 70% of people with CF have their 'complete data' stored on the Registry…

2008



2009

2009
…which rises to 82%.

2010
Median predicted survival increases to 41.4 years, up by more than six years since 2007.

2010

2012
Registry data helps to introduce ivacaftor. The Registry supplies accurate figures on the number of people with CF with the G551D mutation and helps to calculate the impact the new therapy could have.

2013
Using Registry data, the Trust introduces the national tariff 'Payment by Results' system to ensure that the resources given to CF centres reflect the complex needs of the people they treat under the NHS in England.

2011 2012 2013 2014

2013
50 people with CF receive a double lung or lung-heart transplant, compared to 19 in 2009.

2014
97.7% of people with CF have their genotype recorded on the Registry.

"It is impossible to overstate how important the Registry has been to improvements in care and outcomes for people with cystic fibrosis throughout the UK. The Registry contains high quality demographic and clinical information and enables research, improvements in the quality of care of people with CF, development of CF services, future planning and funding of services."

– Dr Keith Brownlee, Director of Impact at the Trust.

Median predicated survival age: An estimate of the age beyond which we expect half of the population of individuals born today with CF in the UK to live. For example, if this age was 40, we would expect half of people born with CF in the UK today to live to 40 years old or older.

2015

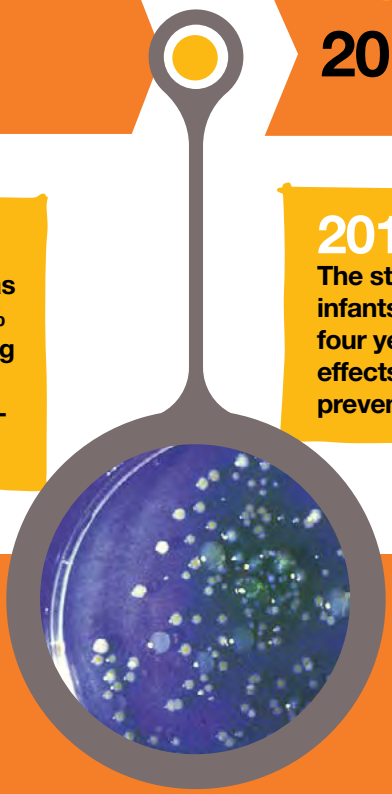
Data on pregnancy in people with CF is reported on for the first time.

2015

The first Registry report to use groups of three years' worth of data to predict survival is released, showing a 47-year median predicted survival age between 2013 and 2015.

2015

Chronic Pseudomonas has reduced by 15.2% since 2008, suggesting the effectiveness of treatments and cross-infection guidelines.



2016

The world's first CF Registry-based clinical trial is launched, called 'CF START'.

2016

The study will observe infants with CF over four years to study the effects of prescribing preventative antibiotics.

2017

The first adults with CF begin entering their own quality of life data directly onto the My CF Registry portal as part of the CF-EpiNet study.

Today over 99% of people with CF have consented to their data being submitted to the Registry.

Visit cysticfibrosis.org.uk/registryreports to find out more.

Cheering champions!

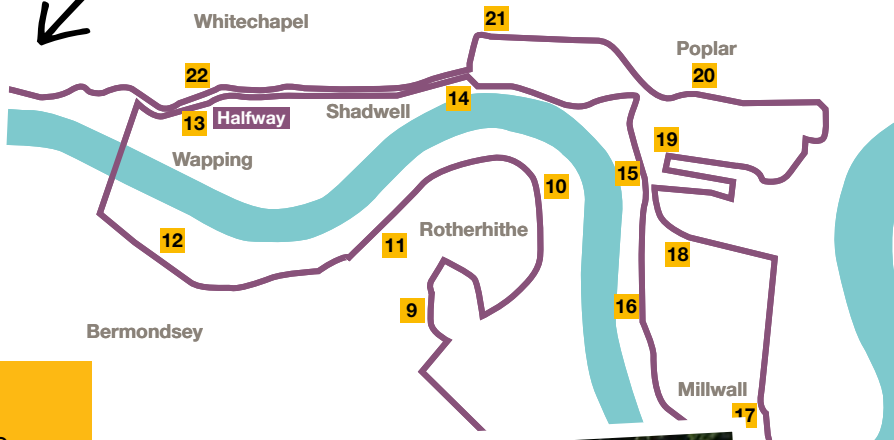
130+ runners, 26 miles and hundreds of thousands of pounds raised – find out what it's like to be part of our London Marathon cheer crews!

Mile 7

Volunteers and staff set up early at mile seven to make sure they were there to high-five everyone supporting the Trust.

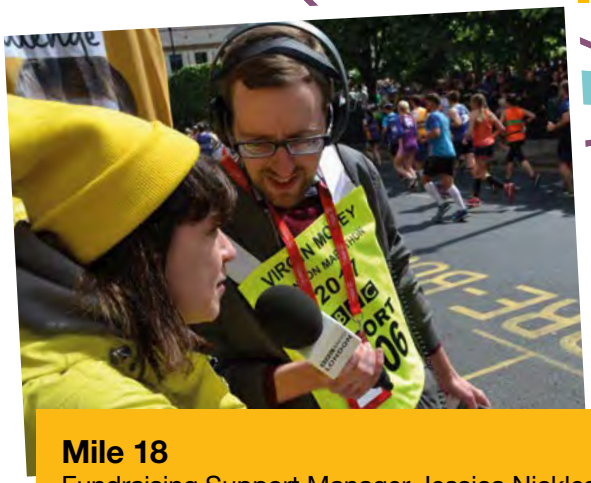
Mile 12

Team Mile 12 met one fantastic family who have run so many marathons they've raised over £27,000, and our balloon arch in the middle of Tower Bridge was so big and bright that the film crews could see it from the sky!



Mile 15

Team Mile 15 managed to get the attention of every Team CF runner that went past, except for Trust Chairman George Jenkins, who was so in the zone that he didn't hear them cheering his name. Surprising, as the marshals awarded Team Mile 15 'the most noise generated' in the area.



Mile 18

Fundraising Support Manager Jessica Nickless represented the Trust on BBC Radio 5 Live.

This year's marathon raised over £300,000 and counting, enough to fund the helpline for over four years.

Mile 23

Team Mile 23 ended up with sunburn, croaky voices and sick stomachs from eating too many sweets and cakes, but still managed to shout their hearts out.

Mile 25

Romance was in the air when runner Jon surprised his girlfriend Amy by getting down on one knee and proposing. The story was shared by the Metro, Sun, Telegraph, Huffington Post and Manchester Evening News, and our Facebook video of the proposal reached almost two million people!

Mile 26

One of our runners, Alfie, carried a fellow runner towards the finish line.

Back at Marathon HQ

Chief Executive David Ramsden was on hand to offer congratulations, and post-event hospitality including hot meals and sports massages was available for all our runners!

Information Manager Michelle Rostant-Bell waved and cheered so much that her FitBit thought she had completed one and a half hours of 'outdoor biking', burning 635 calories!

Choose CF Life…

Choose taking back control, choose a zero hours non-contract, choose your magazine, the way you want it, choose the media and the issues you want to read about, choose the stories that mean something to you, choose showing the world what the CF community can do… choose CF Life!

Join our readers panel and help make journalism great again! Email magazine@cysticfibrosis.org.uk.

cysticfibrosis.org.uk/magazine

Mixed Martial Arts Conditioning

This feature may be called easy exercise, but you'd be amazed what happens when you combine a couple of moves effectively! Aaron Aby is a professional mixed martial artist (MMA) with cystic fibrosis, and an instructor on Pactster.com, where you'll find a wide range of exercises you can try at home.

In this example, Aaron introduces a simple step and jab move to get the heart rate up and the blood pumping. **Remember to consult your CF team before attempting any new exercise.**

1

Put your marker down! Use a cone, jumper or any object to mark the position of your 'opponent'.

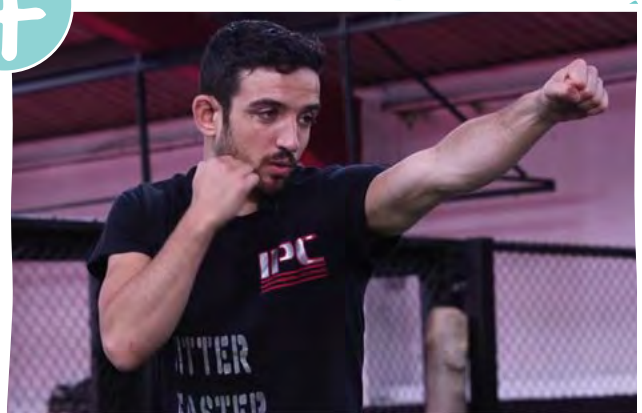
2

To move forward, I push off my right leg, and to move back I push off my left leg – then we're going to move in and out of range.

3

You can do for this for 30 seconds or a certain amount of repetitions. Both feet are moving in, both feet are moving out.

4



Then we throw in the jab! As we come forward, I extend my arm through and the shoulder protects my chin. As I move back, I pull my arm back.

"At a very early age I understood how health, nutrition and psychological mindset are all linked together in achieving your potential."

– Aaron Aby

A professional with the MMA Academy in Liverpool, Aaron also owns the IPC Wrexham gym, where he coaches students of different ages and abilities in strength and conditioning, and mixed martial arts.

Aaron has long been a firm believer in looking after mind and body.

"I have always been extremely passionate about health and nutrition due to being born with cystic fibrosis. I recognised this had a big impact on how I treated my body and mind. It was at a very early age I understood how health, nutrition and psychological mindset are all linked together in achieving your potential.

"Hopefully my videos can help you improve your life and inspire you to exercise. My videos are aimed at all levels and you can change the difficulty depending on your own individual ability and aims."

Aaron Aby

We have partnered with Pactster to offer free membership for people with cystic fibrosis and their families. You can enjoy a wide range of exercises from Aaron and other instructors with CF, including yoga, pilates and High-intensity Interval Training. – visit **www.pactster.com** and find out more.

A chinwag with YAG

"The Youth Advisory Group (YAG) is made up of young people 14-25 with cystic fibrosis who help to influence work at the Trust and ensure that the voices of young people with CF are heard.

"YAG members have had social media training, vlogged at Bestival, and even worked on an award-winning CF app. Want to give it a go? Head over to **cysticfibrosis.org.uk/YAG** to get involved.

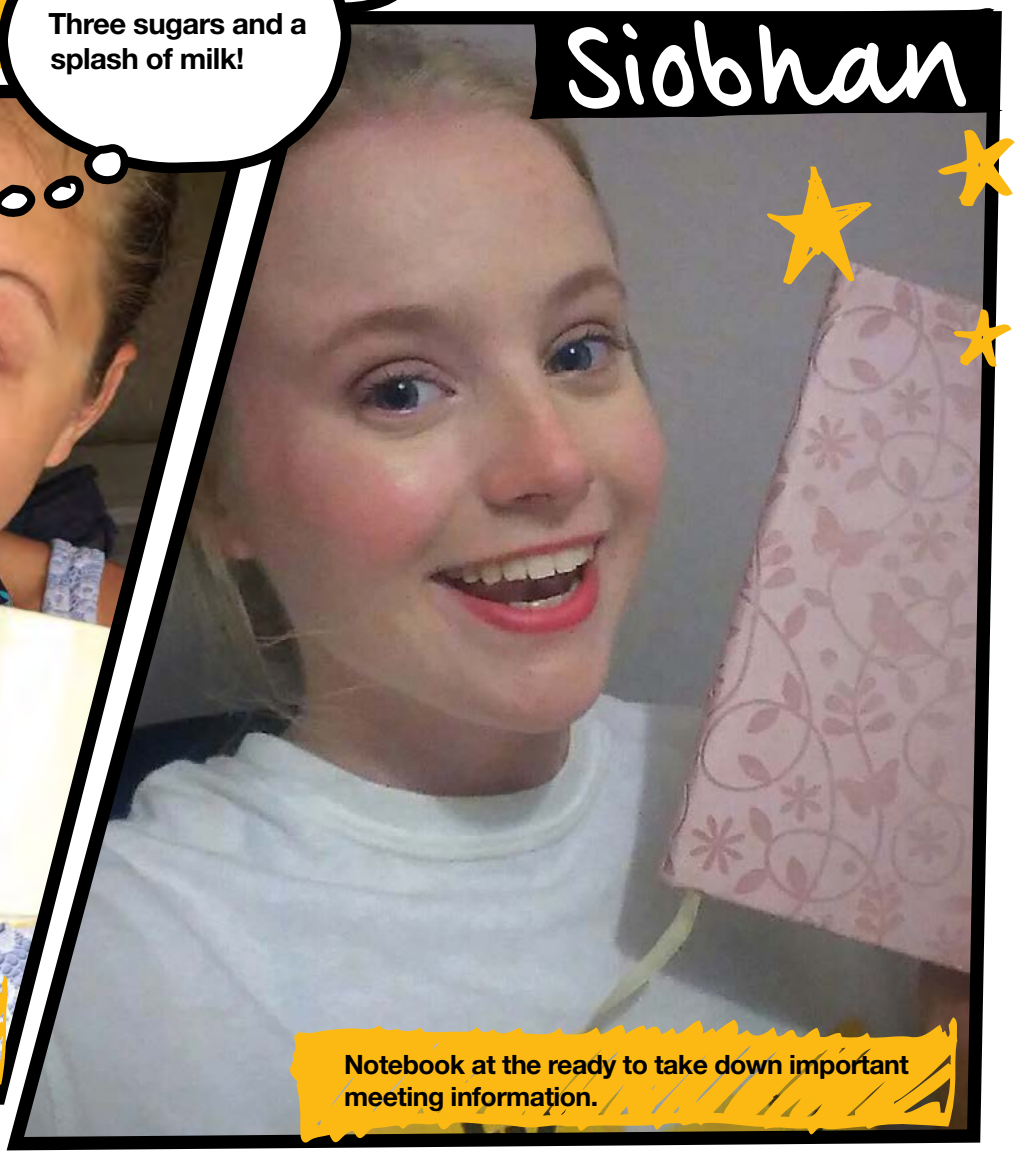
"In the meantime, take a look at what we get up to at our meetings!"

- Holly-Rae Smith,
Youth Empowerment Officer

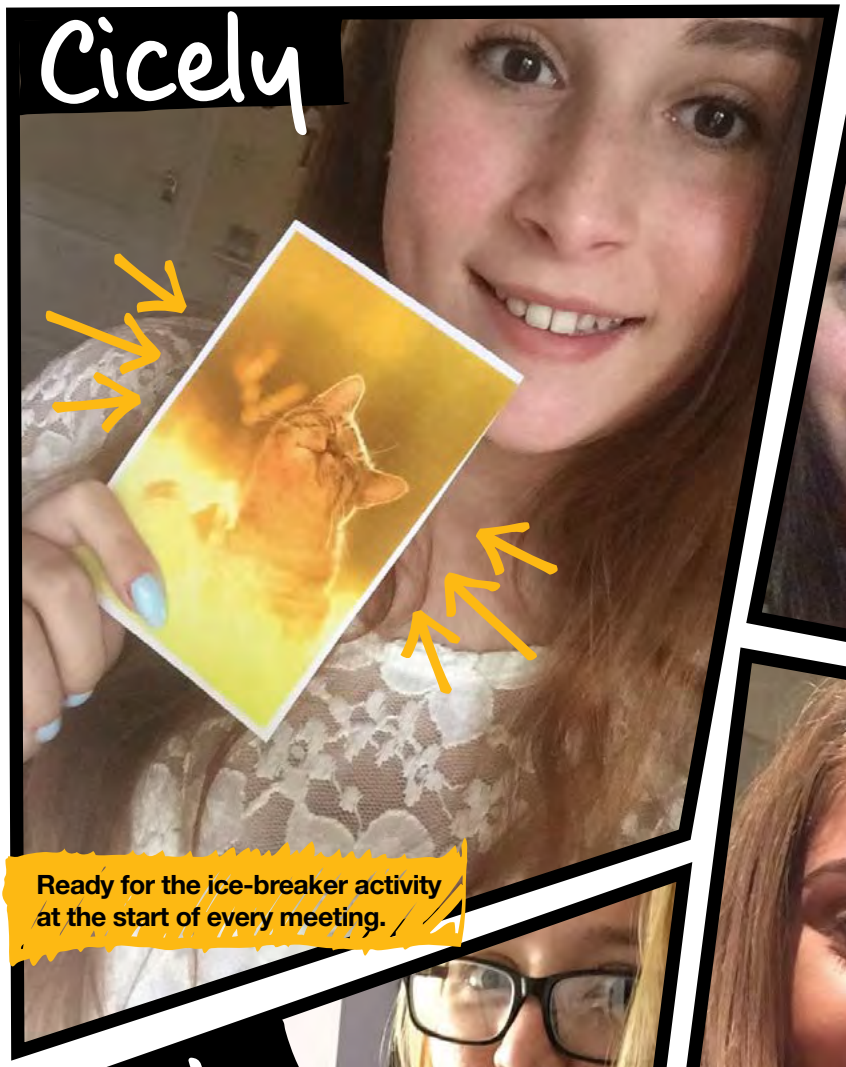
Jade

Niki

Siobhan

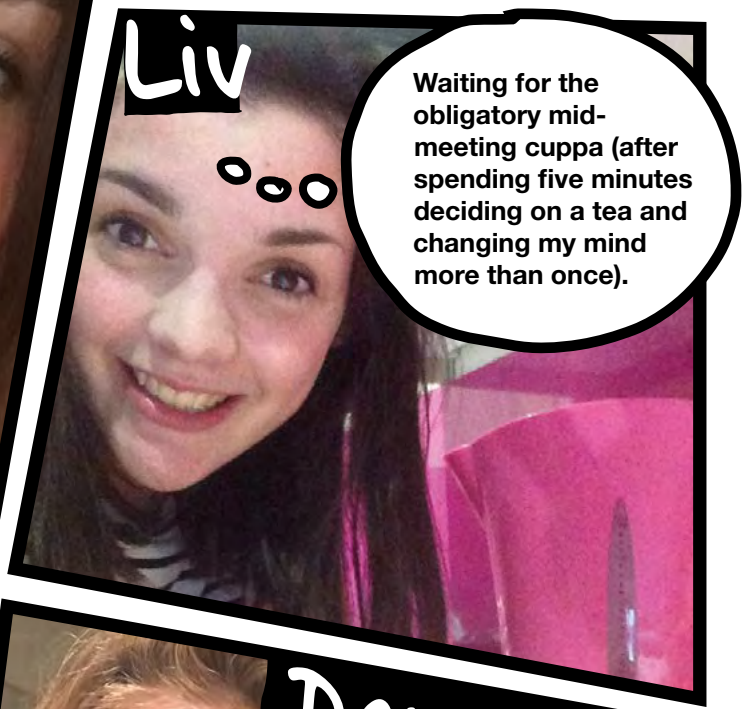


Cicely



Ready for the ice-breaker activity at the start of every meeting.

Liv



Waiting for the obligatory mid-meeting cuppa (after spending five minutes deciding on a tea and changing my mind more than once).

Dommie

Scavenging for Creon for late night chicken.

Beth

Needing my inhaler after all the laughing that happens during our meetings.

Are you aged 14-25 and living with cystic fibrosis? Find out more about taking part in our Youth Advisory Group by visiting cysticfibrosis.org.uk/YAG

Cystic Fibrosis making a difference

Have your say!

Have your say in who should benefit from your estate and consider including a gift in your will to help us to continue fighting for a life unlimited by cystic fibrosis.

This September we are offering a free will-writing service. You don't have to include a gift to the Trust in your will to take advantage of this fantastic offer, but we sincerely hope you will consider doing so. More information can be found at **cysticfibrosis.org.uk/freewills**.

We are also running Gifts in Wills focus groups during October in Dundee, York, and Abingdon. We want you to #HaveYourSay on our future marketing of this important means of raising income.

For more information please contact Michael Clark by Friday 6 October on 020 3795 2132 or giftsinwills@cysticfibrosis.org.uk.

#HaveYourSay

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