



# cf TODAY

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Sophie's genetic journey

Why bother to take vitamins?

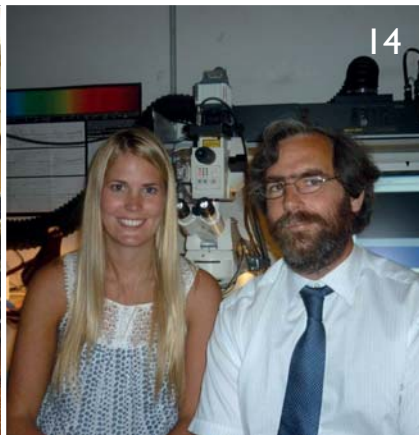
Welfare reforms

Research updates

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The primary purpose of *CF Today* is to provide for its readers a reliable source of medical, research and other information relevant to Cystic Fibrosis and to play a supportive role for CF families. Opinions expressed in articles do not necessarily express the official policy of the Cystic Fibrosis Trust. The editor reserves the right to edit and otherwise alter articles or letters submitted to the magazine for publication.

Some pictures used in this publication may be posed by models or taken from library images.

*Medical information included this publication is not intended to replace any advice you may receive from your doctor or CF multidisciplinary team and it is important that you seek medical advice whenever considering a change of treatment regimen.*

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Cover photo: Sophie Longton

Happy New Year. At the Cystic Fibrosis Trust, we are all looking forward to the challenges, and achievements, that we hope this year will bring.

As you may have read in the last issue of *CF Today*, our Chief Executive Matthew Reed joined the Cystic Fibrosis Trust in September last year. During the last few months, as well as learning as much as possible about Cystic Fibrosis, Matthew has been meeting CF Trust staff, stakeholders and the wider CF community, to find out more about our work and how we can achieve our organisational goals in 2011 and beyond. Over the coming months Matthew will be actively seeking your feedback on how you think we can best serve the needs of people with Cystic Fibrosis.

The past year has been one of continuing uncertainty with regard to the financial climate in the UK, and we are doing our utmost to ensure that the needs of people with Cystic Fibrosis are not compromised by spending cuts and reviews. On page seven, our Policy Advisor Jess Jones explains what the recent welfare reforms mean for people with Cystic Fibrosis.

The annual North American Cystic Fibrosis Conference was held in Baltimore in October. The Conference provides an invaluable opportunity for CF teams worldwide to converge and discuss best practice in the care of people with Cystic Fibrosis, and to hear about advances in medical research and new treatments, the most significant of which are summarised by our Chairman Dr Jim Littlewood on page 12.

Coughs and colds can be of particular concern to people with Cystic Fibrosis at this time of year. In *Ask the Expert* (page eight) our panel of clinicians advises one reader on how to counteract bugs in the office. We'll be very interested to see the results of a new clinical trial starting at Wythenshawe Hospital



Sophie Longton during filming of *Horizon*

in Manchester, which aims to find out exactly what the impact of respiratory viruses is on people with CF – something which is not yet clearly understood.

Many of you will have seen Sophie Longton (pictured above) on the BBC's *Horizon* programme in November. Learning about the progress made in gene therapy since the human genome was first mapped ten years ago gave Sophie, who has Cystic Fibrosis and is pictured on the cover of this issue, a lot of optimism for the future. Read about her experience of taking part in the documentary on page ten.

Finally, as always, we would welcome your feedback on anything you've read in *CF Today*, or would like to read about in the future. Email [cftoday@cftrust.org.uk](mailto:cftoday@cftrust.org.uk) with your comments or suggestions.

We hope you enjoy this issue.

Jacqueline Ali  
Editor, *CF Today*

## Allison is Pride of Britain

Congratulations to Allison John, who as many of you will have seen, received a Pride of Britain award at the annual ceremony held in November last year. 32-year-old Allison, who hails from Cardiff, has Cystic Fibrosis and has had all of her major organs transplanted. Despite the considerable challenges she has faced, Allison was determined to follow her dream to study medicine and last year qualified as a doctor. We hope you are as inspired by Allison as we are.

## New publications for people with Cystic Fibrosis and clinicians

The Cystic Fibrosis Trust has published a new factsheet on CF-related liver disease. As people with Cystic Fibrosis grow older, they may encounter liver problems, which can usually be effectively treated with a medicine called URSO. The factsheet describes how and why liver problems can occur in Cystic Fibrosis, what treatment is given and how liver problems should be managed.

We have also published a new consensus document to add to our series of guidelines for clinicians working in Cystic Fibrosis. Entitled Laboratory Standards for Processing Microbiological Samples from People with Cystic Fibrosis, this document is intended as a guideline for microbiologists and clinicians involved in CF microbiology. On page 11, Dr Miles Denton, Chair of the Laboratory Standards Working Group, explains why this latest consensus document will contribute to the improved clinical care of people with Cystic Fibrosis.

Finally if you'd like to find out about the Cystic Fibrosis Trust's activities during the last financial year, check out our annual review for 2010. The theme is *Living with Cystic Fibrosis* and you can read about how we help individuals and families living with CF and how they help us.

All of our publications can be downloaded online at [www.cftrust.org.uk](http://www.cftrust.org.uk).

## Cystic Fibrosis statistics

You may have heard mention or seen in some of our newer publications that there are now over 9,000 people with Cystic Fibrosis in the UK. The number of people living with CF has been increasing for a number of years, thanks to the progress that has been made in the treatment and understanding of the condition. In addition, the UK CF Registry – the national database of people with CF in the UK – is providing us with much more accurate data regarding the UK CF population, and in a much more timely manner, which we can then convey to the CF community.

## DVD for children with CF

You may have read in the last edition of *CF Today* that the Cystic Fibrosis Trust was producing a new, animated DVD for young children with Cystic Fibrosis. The film can be viewed online at [youtube.com/cftrust](http://youtube.com/cftrust). To order a DVD copy of *Getting Nosey about CF*, please use our online publications order form or email [publications@cftrust.org.uk](mailto:publications@cftrust.org.uk). The DVDs are available for loan and we ask that they are returned to the CF Trust when no longer needed. We wanted to make the DVDs available for loan as opposed to purchase so that they could be distributed as widely as possible to all who need them. However for those who wish to make a donation towards the costs of producing the DVD, we have set up a special online giving page; visit [www.cftrust.org.uk/oliandnush](http://www.cftrust.org.uk/oliandnush) for details.



Nush and Oli approve the script with producer Dan at a recording session

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## Reforms to Disability Living Allowance

As this issue of *CF Today* was going to press, further details emerged about the Government's proposed reforms to Disability Living Allowance (DLA) (you can read more about the welfare reforms on page seven). It is planned that DLA will be replaced by the Personal Independence Payment (PIP), which will have two components: a 'mobility' component, based on a claimant's ability to get around, and a 'daily living' component, based on their ability to carry out 'other key activities necessary to participate in everyday life'. Each of these components will have two rates, and eligibility will be determined by assessment.

A Department of Health consultation on the proposed changes is open until **14 February** at [www.dwp.gov.uk/consultations/2010/dla-reform.shtml](http://www.dwp.gov.uk/consultations/2010/dla-reform.shtml).

For more information search for 'welfare reform' on the CF Trust website.

## Cystic Fibrosis Week 2011

We have a jam-packed Cystic Fibrosis Week lined up this year and would like to encourage you all to get involved. CF Week 2011 will take place from 8-14 May and there will be a range of events for you to take part in – or you can organise your own! Keep an eye on our website in the coming weeks, as we'll be posting all the details there.

## Parents Conference 2011

On 19 March, we will be holding a conference for parents and carers of children with Cystic Fibrosis aged up to 12 years. As with our previous parents conferences, the day will include talks from clinicians and the Cystic Fibrosis Trust on hot topics in the care and treatment of children with Cystic Fibrosis, including research updates, psychological support for parents and carers and general management of Cystic Fibrosis.

The conference will be held in Manchester; parents who have children with CF aged 12 and under will be receiving their invitations shortly. For further details search for conferences on the CF Trust website.



Oli Lewington and his dad Keith at the 2008 conference

*“A very worthwhile and interesting day.”* Quote from attendee at 2008 parents conference.

## Psychosocial support survey

Following our dietetics survey which we carried out in 2009/10, we have launched a survey to find out the psychosocial care people with Cystic Fibrosis are receiving at CF Centres and Clinics across the UK. We've had a great response so far – if you've received your survey but haven't returned it yet, please do so as soon as you can and before the end of January. Your feedback will help us campaign for improved psycho-social services for people with Cystic Fibrosis.

## Call for contributors

We're looking for people with Cystic Fibrosis, and family members, who would be interested in sharing their stories in future editions of *CF Today*. If you would like to contribute or have any suggestions for articles you'd like to read, please drop us a line at [cftoday@cftrust.org.uk](mailto:cftoday@cftrust.org.uk).

## Blue badge developments in Scotland

**Yvonne Hughes, our Expert Patient Adviser for Scotland, Northern Ireland and the North East of England, reports on blue badge developments north of the border.**



Yvonne Hughes

Recently I participated in a Blue Badge Scheme reform consultation for Transport Scotland, which is a branch of the Scottish Government.

The Blue Badge Scheme is an important service for people with severe mobility problems, which enables badge holders to park close to where they need to go. It operates throughout the UK, and is administered by local authorities who deal with applications and issue badges.

The aim of the consultation was to make it easier for local authorities to administer the badge and reduce fraudulent claims. However the Cystic Fibrosis Trust was concerned about the idea of introducing an independent medical assessment for those who do not have an automatic entitlement to the badge, i.e. those who receive the Higher Rate Mobility Component of the Disability Living Allowance (HRMC-DLA). In this event the CF Trust has stipulated that assessors must have specialist knowledge or be willing to consult with Cystic Fibrosis specialists to ensure the correct outcome.

Another concern is the intention to change the wording on the blue badge application form to bring it in line with applications for HRMC-DLA. At present the wording on the Scottish form states “has considerable difficulty walking”, which in some circumstances may be relevant for people with Cystic Fibrosis. However, in assessing applicants for DLA, the Department for Work and Pensions requires that the applicant is “unable to walk” or is “virtually unable to walk”. The Cystic Fibrosis Trust has stated its opposition to changing the application wording on the grounds of the variability of each person's condition at any given time.

These proposals are part of a number of changes that will affect the issue of blue badges. The report was due to be published by the end of 2010 and the Cystic Fibrosis Trust will be working with Transport Scotland to ensure the needs of people with Cystic Fibrosis are not overlooked.

The Blue Badge Scheme for England and Wales was amended in 2008.



# Why bother to take vitamins?

On top of the daily medication and physiotherapy regimen that Cystic Fibrosis involves, having to take vitamin tablets as well can be a bit hard to swallow. But as **Alison Morton**, Specialist CF Dietitian at St James's University Hospital in Leeds explains, ensuring an adequate amount of vitamins in the diet is essential for people with Cystic Fibrosis.

The fat-soluble vitamins are vitamin A, D, E and K. Deficiencies of these vitamins occur early in life in people with Cystic Fibrosis who are pancreatic insufficient (those who need to take digestive enzymes). This is because despite taking digestive enzymes they still cannot fully absorb fat-soluble vitamins properly. There are other reasons why deficiencies can occur however, for example some patients may have increased requirements for the antioxidant vitamins A and E. Those patients who are pancreatic sufficient (don't need enzymes) will need individual assessment and may not need vitamin supplements, though most will still need to take vitamin D supplements as dietary intake of vitamin D is quite low, and though we can make vitamin D by the action of sunlight on the skin this can only occur at certain times of the year so many people have low blood levels.

We know there are many reasons why some people with Cystic Fibrosis find it difficult to take their vitamins regularly. One reason is that if someone does not take their vitamins nothing seems to happen – there is no obvious benefit to taking them. Some people may forget, some do not think they are as important as their other medication and some may feel they are already taking too many tablets!

So... why bother to take vitamins? If vitamins are needed and not taken, over a period of time overt deficiencies will occur and deficiency symptoms will

develop. However, in the shorter term sub-clinical deficiencies (low levels with no visible signs) may occur, which may be equally important.

## Vitamin A

Vitamin A deficiency results in a condition called night blindness. This is when people can not see properly in the dark, and the problem is often noticed when driving at night. In severe deficiency the conjunctiva (the mucus membrane that lines the eye) becomes dry, thick and wrinkled, ulceration may also occur and at its worst it can cause blindness. Importantly for people with Cystic Fibrosis, sub-clinical vitamin A deficiency may have a damaging effect on lung function by altering the membranes lining the lungs, making mucus more difficult to clear and weakening the immune system, making people more prone to infection. Some studies suggest that people with CF with lower vitamin A levels have more respiratory exacerbations and poorer lung function.

*“Vitamin A deficiency can have a damaging effect on lung function.”*

## Vitamin E

In vitamin E deficiency red blood cells are more fragile and have a shorter survival time so are more prone to breaking down, and this can lead to anaemia. Though unusual these days, severe vitamin E deficiency also results in nerve damage.

In Cystic Fibrosis, vitamins A and E may also play important roles as antioxidants (substances that may protect cells from the damage caused by unstable molecules known as free radicals). Antioxidants stabilise free radicals and help to minimise damage to membranes. Infection increases oxidative stress in



Vitamin deficiencies can occur early in life in people with Cystic Fibrosis

patients with Cystic Fibrosis. Improved antioxidant status is associated with improved lung function.

## Vitamin D

Severe vitamin D deficiency is rare in Cystic Fibrosis but moderate deficiency is common and contributes to reduced bone mineral density (thin bones or osteopenia/osteoporosis). Recent studies have shown that higher vitamin D levels in people with CF are associated with better lung function.

## Vitamin K

It has not always been routine practice to supplement with vitamin K but it is becoming more common. In vitamin K deficiency the blood does not clot effectively and severe bleeding can occur. In addition, vitamin K deficiency can contribute to reduced bone mineral density. More recently it has been found that vitamin K may play a role in reducing inflammation and therefore may help to prevent or reduce lung damage in Cystic Fibrosis.

A word of caution – excessive intakes or high blood levels of the fat-soluble vitamins can also contribute to problems so it is important that levels are monitored regularly and the dose is adjusted to maintain levels within a healthy range. For this reason there is no standard dose of vitamin supplementation for people with Cystic Fibrosis. As always, if you have any concerns discuss them with your specialist CF team.



Vitamins can be viewed as an additional treatment burden

# Welfare reforms

Our Policy Adviser **Jess Jones** summarises the recent Government Spending Review – and the key points people with Cystic Fibrosis need to be aware of.



Jess Jones

I watched the October Government Spending Review with figures swimming in the back of my head, conscious of the fear and insecurity change can bring to someone already in poor health. Roughly half of all over 16s with Cystic Fibrosis in the UK are in paid employment, leaving a significant proportion reliant on help and support from the benefit and care systems. Changes to the welfare system were much talked about prior to the announcements, and touted as a major money saving route for a country with an enormous financial deficit. Changes were made to the current system, some of which can only be viewed positively by those of us in a position of genuine need; others are a little more concerning depending on how they are enforced.

## Employment and Support Allowance (formerly Incapacity Benefit)

Employment and Support Allowance (ESA) terms have been changed, and a new 12-month time limit will be enforced on claimants in the Work Related Activity Group. After that they will be forced to find work or face sanctions. This move to work will not apply to those in the Support Group of ESA, who are categorised as unfit to work.

Work and Pensions Secretary Ian Duncan Smith has stated that these sanctions will not be brought in until proper support to help people back into work has been created – an essential factor, particularly in the case of Cystic Fibrosis where the burden of treatment required on a daily basis is ever increasing, and the individual may only be available for part-time work or to work for an employer who shows considerable flexibility in enabling them to take the necessary time for hospital appointments and inpatient episodes. Those with CF who are able to work should of course be encouraged to, but with the correct help and support when appropriate. There can be a thin dividing line between the psychological benefit in terms of boost to self esteem from being in work, and the work actually having a negative impact on the health of those with Cystic Fibrosis. This is something we have heavily emphasised to the Government, and will fight to ensure it is taken note of.

## New medical assessment for Disability Living Allowance

Following on from the shake up of Employment and Support Allowance, a new medical assessment for Disability Living Allowance is due to be introduced in 2013. It is to introduce ‘expert assessors’ for new claimants, which will slowly be rolled out to those already in receipt of the benefit. With just over 9,000 people with Cystic Fibrosis in the UK it is unlikely each assessor will have the required knowledge of this complex condition. It is essential therefore, that the assessors take on board the information given to them by the person’s specialist medical team, who will have a good understanding of what capacity or ability to work the person with CF has, and the treatment burden they endure.

## Housing Benefit

Concerns regarding the increase in the age threshold for Shared Housing Rate of Housing Benefit have been raised. The Government claims that moving the age from 25 up to 35 “will

ensure that Housing Benefit rules reflect the housing expectations of people of a similar age not on benefits.” Our issue is that people with Cystic Fibrosis are often not a reflection of other people of a similar age. Although those who claim Severe Disability Premium are exempt from the ruling, not all with CF will be eligible for this exemption. A heavy medication regime means people with CF often require extra room to store their medication, as well as a clean living environment.

## Other developments

We are pleased to report that the £26,000 benefit cap will not apply to those in receipt of Disability Living Allowance, reflecting the higher cost of living for disabled people. Last year’s temporary increase in Cold Weather Payments from £8.50 to £25 has been made permanent – a welcome addition to those who are on ESA (Income-based).

Funding for the NHS has in theory been ‘ring-fenced’, but as we reported in the last edition of *CF Today*, there are reports that some essential frontline services are suffering at the hands of the economy, most specifically specialist nursing posts. We hope that through full implementation of the Payment by Results system which has been developed by the Cystic Fibrosis Trust, funding for Cystic Fibrosis patients will be more secure in the current climate. This system would enable fair and equal funding for individual patients and would remove the ‘Postcode lottery’ element from medication prescribing.

The Labour Government’s promise to remove prescription charges for all those with long-term conditions will unfortunately not be taken forward by the coalition Government. People with Cystic Fibrosis are still able to obtain annual pre-payment certificates (PPCs), which are more cost effective than paying for individual prescriptions, and the Cystic Fibrosis Trust offers a welfare grant to help pay for the first year of payments. We recognise that this is an important issue for many people with CF and we will continue to make the case for free prescriptions for people with Cystic Fibrosis where possible, but due to the economic climate we do not expect the situation to change in the foreseeable future.

The full impact of the Spending Review is naturally going to take time to filter down, but there are a number of issues it is important we are pro-active over to help ensure the Cystic Fibrosis community is not unfairly targeted. We are working with a number of other disability charities to provide a strong voice opposing the way ESA eligibility is currently assessed, and to help iron out any issues with the upcoming DLA assessment. The Cystic Fibrosis Trust continues to respond to all relevant consultations and initiatives the Government introduces, alongside our other work.

We are all aware that now is a time of financial hardship, but we maintain our stance that the most vulnerable in society should not be hit hardest.

# Ask the expert

Ask the Expert is the Cystic Fibrosis Trust's expert advice service for people with Cystic Fibrosis and their families. Questions are answered by a panel of clinicians who specialise in different areas. Although your query and the response may appear on the Cystic Fibrosis Trust website or in this magazine, your name and personal details will always remain confidential and will not be published.

## Floristry

**Q** My daughter has Cystic Fibrosis and at 16 has decided that she wants to study floristry. Can you advise if this is a good move? My concern is that working in a potentially damp environment may not be a good thing.

**A** We presume your daughter wishes to work in a florist shop? There is a great deal of published work on the presence of bacteria in the water of cut flowers to the extent that they have been banned from some hospital wards. The bacteria are those of a type that include *Pseudomonas*.

So presumably handling flowers that had been in water would carry an infection risk. Also there would be possible exposure to *Aspergillus* in container soil and compost. However, we are not aware of anyone with CF acquiring infection from the water in containers used for cut flowers – nor can we find a reference to such an occurrence.

So working from the knowledge that the water in flower containers commonly contains potentially harmful bacteria, it would be better for your daughter to choose some other form of occupation. However, if her mind was made up we have no definite proof that being a florist would be harmful.

An expert CF microbiologist agrees with our answer to your question.

## Financial support

**Q** Having Cystic Fibrosis myself means I regularly receive *CF Today*, which mentions there is financial support available for people with Cystic Fibrosis. As well as having to pay for my own prescriptions I am also living away from home at university which means money is quite an issue for me.

Is there any advice/help you could offer me?

**A** If you have not already done so, check out our Higher Education factsheet on the website – [www.cftrust.org.uk/aboutcf/publications](http://www.cftrust.org.uk/aboutcf/publications) – there are organisations such as SKILL who can advise on grants and benefits. Applications to the Joseph Levy Memorial fund for instance are in March for the following academic year.

If you are no longer eligible for free prescriptions you may qualify for a grant to cover the first annual pre-payment certificate. Check out our Welfare Grants helpline (details on back page) – there is an application form available.



Potentially harmful bacteria can be found in the water of cut flowers

## What is a pulmonary exacerbation?

**Q** I've heard the term 'pulmonary exacerbation' used in relation to Cystic Fibrosis. I have an idea what it is but I'm not quite sure – please could you clarify?

**A** Defining a pulmonary exacerbation is quite tricky in Cystic Fibrosis. People with CF generally know when they are having a pulmonary exacerbation and the combination of symptoms and signs allow their doctors and CF teams to also identify them, but arriving at a single unified definition of these events is difficult.

The term pulmonary exacerbation describes what happens when there is worsening of respiratory symptoms, usually with increased cough, more sputum and possibility also of sputum becoming darker. There is sometimes a fall in measurements of lung function (FEV1) and in some cases a reduction in appetite, weight loss and energy. Sometimes exacerbations are caused by new infections such as *Staphylococcus aureus*, *Haemophilus influenzae* or a first infection with *Pseudomonas aeruginosa*. Viruses also play an important part in causing them.

In general, exacerbations are treated with a combination of intensifying airways clearance and antibiotics, which particularly with *Pseudomonas* infection may need to be given intravenously. Optimisation of other therapies such as DNase, hypertonic saline etc, increased energy intake and exercise are also important. Sometimes admission to hospital for treatment

is necessary if the symptoms are very marked or if airways clearance is difficult or perhaps extra oxygen is required because saturations are low. Two weeks of antibiotic therapy usually improves lung function and reduces symptoms and puts things back on an even keel. During the two-week treatment, cough and sputum get less and lung function improves.

Avoiding exacerbations, where possible, is important in Cystic Fibrosis. Some of the drugs for long-term treatment such as inhaled antibiotics, DNase, hypertonic saline and Azithromycin have been shown in clinical trials to reduce exacerbations. Exacerbations are very annoying and have a negative impact on quality of life but also are associated with more progressive reductions in lung function. Taking long-term treatment which prevents these is therefore really important.

### Incidence of Cystic Fibrosis

**Q** I noticed on the CF Trust website that Cystic Fibrosis is now described as 'one of' the UK's most common life-threatening inherited diseases, as opposed to 'the most common'. Could you tell me why this fact has now changed?

**A** Cystic Fibrosis was for a very long time the most common life-threatening inherited disease in the UK; however we now know that sickle cell anaemia now affects more people in the UK than Cystic Fibrosis. Cystic Fibrosis remains the most common life-threatening inherited disease in white people, whereas sickle cell anaemia predominantly affects people of Afro-Caribbean descent. We felt it was important to amend this fact to accurately represent the situation as it currently stands in the UK.

### Avoiding colds and flu

**Q** I am a 32-year-old with Cystic Fibrosis and I will shortly be moving to a new office at work, which is quite small and has several employees. The room is air conditioned and does not have any opening windows. I am concerned that an air conditioned room will recirculate any coughs and colds that people will inevitably develop over the winter months and I will be frequently unwell. Last winter, when I shared a normally ventilated room with just two other people, I required a course of intravenous antibiotics following exacerbations which developed after my colleague came into work with a bad cough and cold for about a week.

I'd be grateful if you could let me know whether the air conditioned environment poses any more risks.

My manager is aware of my CF and has arranged for an alternative desk in an office shared with just one other person for me to use occasionally as necessary if my colleagues turn up to work with flu symptoms or similar. I seem to remember that 'close contact' in the context of developing swine flu was defined as being in close proximity to (e.g. sitting in at a desk opposite) a symptomatic person for greater than one hour. Does this also apply to other types of flu and cold viruses? I would be grateful for any advice on how to minimise the risks of picking up bugs, in



Winter coughs and colds are a concern for people with CF

terms of whether I should use the alternative office accommodation if colleagues come in with bad colds, and whether this should be for the duration that they are symptomatic or whether this isn't necessary. Hopefully if I have a better understanding, I can reduce the chances of being unwell and requiring time off work.

**A** We have discussed your question with a CF microbiologist. There is no definite evidence that an air conditioned environment poses any more risks of viral infection than one without air conditioning.

As you have already experienced, airborne transmission of respiratory viruses is common but the precise risk varies. So it is better to avoid any contact with infected people as far as possible. As a broad guide the risk of infection is usually taken as seven days after the start of the illness.

It would be very wise to use the alternative accommodation provided if colleagues have colds and continue to use it for at least a week after the start of the colleague's illness.

### Our experts for this issue were:

**Dr Miles Denton, Consultant Microbiologist,  
Leeds Teaching Hospitals NHS Trust.**

**Professor Stuart Elborn, Adult Physician,  
Queen's University Belfast**

**Dr Jim Littlewood, Chairman, Cystic Fibrosis Trust**

You can view an archive of past Ask the Expert questions at [www.cftrust.org.uk/aboutcf/asktheexpert](http://www.cftrust.org.uk/aboutcf/asktheexpert).

Please note if you have any queries or concerns about any aspect of Cystic Fibrosis you should contact your CF team in the first instance.

Submit your question to [asktheexpert@cftrust.org.uk](mailto:asktheexpert@cftrust.org.uk)

# My experience of training as a specialist CF consultant

The Cystic Fibrosis Trust's training grant scheme started in 2005. The aim was to provide training for doctors to specialise in Cystic Fibrosis, ensuring good clinical care will be available in the future for the increasing adult Cystic Fibrosis population. Training fellow **Dr Alex Horsley** shares his experience of participating in the scheme.



Dr Alex Horsley

I was appointed to a Senior Training Fellow post at the Manchester Adult CF Centre at the start of 2010. As I am now coming to the end of the post's 12-month tenure, it is an ideal time to reflect on the aims of the post, what it has achieved and what it has meant for me.

The Cystic Fibrosis Trust has rightly recognised the need for more training in Cystic Fibrosis, in adult CF

particularly. Burgeoning numbers of patients in recent years, and the increased complexity of these patients, are testament to the success of medical progress but have somewhat left the standard CF training behind. Typically an adult respiratory trainee will spend only three months working on Cystic Fibrosis and, since care is rightly concentrated in highly specialised units, will only rarely encounter the condition outside of this period. In order to aid training, the CF Trust Senior Training Fellowships have been awarded annually to doctors at the end of their general respiratory training (i.e. as an alternative to taking up a consultant post) in order to acquire further experience in the condition and its myriad complications. The post lasts for a year and, though based at a single unit, also includes time spent in other adult units, paediatric units, related Specialist CF Centres and the CF Trust HQ. The intent is that it allows the doctor to gain a much wider perspective and knowledge of the process and politics of CF care.

Personally, I have greatly enjoyed my time as Senior Training Fellow. Before I started the post I had completed a research PhD post with the UK CF Gene Therapy Consortium in Edinburgh, and had worked at the Scottish Adult CF Centre, so already had some knowledge of Cystic Fibrosis care. Working at such a large and well-established unit as the Manchester Adult CF Centre however has allowed me to interact with a much greater number of patients, and to experience a much wider range of different challenges. As is often the

nature with Cystic Fibrosis, not all of these have been purely medical. Importantly however, there has always been someone on hand to discuss problems with and I have learned much not just from the other medics but from all the members of the team. One of many the strengths of the Manchester unit is the emphasis placed upon social work and psychology input. Professor Webb is an excellent and entertaining mentor, and a better guide through NHS politics would be hard to find. The importance of this at such challenging times is hard to underestimate.

I have also enjoyed my visits to other units, and the wider medical profession has much to learn from Cystic Fibrosis. Cross-fertilisation of ideas and skills is difficult when most medics work only in one hospital or region. The process of peer review informs not only the reviewed unit but also the reviewer and is a model that other specialist bodies would do well to duplicate. I am grateful to the units I visited for hosting me, and hopefully one day can return the favour.

*“The Cystic Fibrosis Trust has recognised the need for more training in Cystic Fibrosis.”*

I hope that I have also contributed to the workings of the Manchester unit. My aspiration is to continue as a CF specialist, and also to contribute to further research. This opportunity has been an important stepping stone – one that both I, and hopefully my patients, have benefited from enormously.

## Share your experiences

The Health Experiences Research Group, University of Oxford, is developing a new resource on Experiences of Clinical Trials for the [www.youthhealthtalk.org](http://www.youthhealthtalk.org) and [healthtalkonline.org](http://healthtalkonline.org) websites. They are looking to talk to **young people ages 12-25 years and parents of children all ages (from birth upwards)** about their experiences of being invited to take part in a clinical trial, even if they declined, withdrew, are currently going through or have completed a trial. The sites will include video, audio and written clips from the interviews, and questions and answers about clinical trials involving children and young people. Please contact Lesley Powell, Researcher: [lesley.powell@dphpc.ox.ac.uk](mailto:lesley.powell@dphpc.ox.ac.uk) or call 01865 617768.

# Peer reviews prove their worth

As of the end of last year, the Cystic Fibrosis Trust's peer review programme had resulted in £18 million in additional NHS funding being directed to the Specialist CF Centres and Networked Clinics visited. For every £1 the CF Trust spends administering the peer review programme – the core aim of which is to help CF teams improve the level of clinical care they are able to offer

their patients – £20 of NHS funding is directed to CF services. We are extremely encouraged by the tremendous impact the peer reviews are having and with several visits already arranged for this year, the programme will be continuing apace in 2011, reflecting our core aim to ensure appropriate clinical care for everyone with Cystic Fibrosis in the UK.

## New guidelines to improve identification of CF infections

*Laboratory Standards for Processing Microbiological Samples from People with Cystic Fibrosis* is the latest addition to the Cystic Fibrosis Trust's range of consensus documents. **Dr Miles Denton**, chair of the working group that produced the document, explains why it is important.

The Laboratory Standards consensus document is aimed at those who work in the laboratories processing sputum and cough swab samples from people with Cystic Fibrosis. It has been developed as a means to improve current lab practices.

A number of publications have highlighted problems with the correct isolation and identification of bacteria from respiratory samples, particularly *Burkholderia cepacia* complex. It is essential that this is done correctly to ensure that the most suitable antibiotics are administered, the appropriate level of segregation is in place and other issues, such as transplant status, are properly managed.

With this in mind the Cystic Fibrosis Trust convened a Working Party to review all available evidence in relation to

best practice for processing CF respiratory samples in the microbiology laboratory. This group included a number of microbiologists working in clinical and research aspects of CF microbiology, CF clinicians, and representatives of the CF Trust. In all the Working Party reviewed over 200 publications before producing its recommendations. These cover areas such as which samples to take, how they should be transported to the lab, how the lab processes them and identifies any micro-organisms they isolate and how the labs should best work with the doctors, nurses and physiotherapists looking after those with Cystic Fibrosis.

The document has now been sent to every microbiology laboratory providing support for CF Centres and Clinics. The Cystic Fibrosis Trust has also asked these



*Burkholderia cepacia*

Image: Janice Haney Carr

*“It is essential that bacteria are isolated and identified correctly.”*

laboratories to participate in an online audit of current practices which will be repeated to find out if these have changed

as a result of the new recommendations. The preliminary results of the audit will hopefully be presented at this year's European Cystic Fibrosis Conference in Hamburg.

The hope is that each and every sample will now be processed to the same standard whichever laboratory receives it.

*Dr Miles Denton*

*Consultant Medical Microbiologist, Leeds Teaching Hospitals Chair, CF Trust Laboratory Standards Working Group*

### Cystic Fibrosis Trust consensus documents

Our consensus documents are detailed guidelines aimed at clinicians and other professionals involved in the clinical care and treatment of people with Cystic Fibrosis.

All of our consensus documents can be downloaded from the publications area on our website [www.cftrust.org.uk](http://www.cftrust.org.uk) or hard copies obtained by emailing [enquiries@cftrust.org.uk](mailto:enquiries@cftrust.org.uk) or by phoning 020 8464 7211.



The new Laboratory Standards document

# Highlights of the North American Cystic Fibrosis Conference 2010

Cystic Fibrosis Trust Chairman **Dr Jim Littlewood** summarises some of the advances in clinical care and treatment in Cystic Fibrosis reported at the 2010 North American Cystic Fibrosis Conference.

Nearly 4,000 delegates attended the 24th North American Cystic Fibrosis Conference held last October in Baltimore. The three main plenary sessions are recommended, and can be viewed on the CF Foundation website ([cff.org](http://cff.org)) in the Research section.

1. *Pipeline: airway surface modulation.* A review of how abnormality of the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene causes the problems in the airways.
2. *Animal models.* An excellent summary of the advances in understanding that have resulted from work on animal models, in particular the recently created CF pig.
3. *Transforming CF healthcare: Partnerships for life.* Dealing with quality improvement initiatives in the USA. This was less relevant for UK delegates but the principles of good care are international.

## General topics of interest

The expansion of newborn Cystic Fibrosis screening has been impressive in both North America and Europe. There are separate European and North American guidelines on the early management of the screened CF infants. On the whole the two statements are consistent, except for anti-staphylococcal prophylaxis (with long term flucloxacillin) being recommended for all infants in the UK but not in the USA – even though some 60% of children there are infected by *Staphylococcus aureus*. Both guidelines recommend breastfeeding which, even for a month, was associated with fewer *Pseudomonas aeruginosa* infections than if formula-fed. For older children, a seven-year study of physical activity demonstrated a slower decline in respiratory function in those children who undertook more activity – more evidence to support our attempts to keep children with CF active.

“A seven-year study demonstrated slower decline in respiratory function in children who undertook more activity.”

## Some new treatments for infection

- *Aztreonam for inhalation (AZLI).* Clinical studies are now completed on aztreonam for inhalation. The results suggest that AZLI has a positive impact on respiratory function and quality of life and it will be available in the clinic.
- *Liposomal amikacin (Arikace™).* Studies have demonstrated clinically relevant improvements in respiratory endpoints with inhalation of this preparation where amikacin is enclosed in fatty liposome droplets. The once-daily dosing schedule makes it attractive to patients and the early results are promising.
- *Levofloxacin for inhalation (Aeroquin™).* Studies with a preparation of this oral antibiotic designed for inhalation have demonstrated impressive results. This agent may have a role

in more resistant infections.

- *Tobramycin inhalation powder (TIP).* The preparation appears to be equivalent to TOBI solution and is naturally preferred by patients for convenience. Hopefully this preparation will soon be available for patients.
- *Fosfomycin/Tobramycin for inhalation (FTI).* A US study demonstrated positive results for this new combination. Fosfomycin and Tobramycin work well together and the combination provides a broad spectrum of activity against gram-negative (including PA), gram-positive (including MRSA) and anaerobic bacteria.
- *Anti-pseudomonal IgY.* IgY is an immunoglobulin extracted from the egg yolk of hens previously inoculated with *Pseudomonas*. The immunoglobulins are proteins that provide protection against bacteria and viruses. Patients will use the IgY solution as a gargle at night time; early trials have shown a reduction in new positive cultures. A multi-centre clinical trial is planned in Europe.

## Other new treatments

- *VX770.* This is a “potentiator” (improves channel function) and improves CFTR function in patients with the G551D mutation – sweat tests and nasal potential differences approached normal and improved respiratory function was seen. More results will be available this year.
- *VX809.* A “corrector” (targeting the production of faulty F508del-CFTR) improves the trafficking defect associated with the F508del mutation. With treatment there was a significant reduction in sweat chloride, but not to the extent as seen with VX770. It is planned to explore the possible additive effect of a combination of VX809 (to correct the F508del defect) and VX770 (to potentiate the function of that protein once it reaches the cell membrane). If this combination works, these orally available drugs should have a profound impact on treatment of the majority of people with Cystic Fibrosis.
- *Ataluren™.* This compound (PTC-124) causes read-through of stopcodon mutations (such as G542X and others ending in an X), potentially correcting that genetic defect. Already clinical studies suggest adequate efficacy to move to a Phase III study and the results should be available next year.
- *Denfospol.* This inhaled agent stimulates chloride transport through non-CFTR pathways and so improves airway hydration. There are modest benefits but a good safety profile suggesting future use as an early intervention treatment.
- *Mannitol (Bronchitol™).* Data from three studies provides some good evidence of benefit in respiratory function from this osmotic agent that attracts water into the airways and improves mucus clearance. Pending licensing issues, this compound should be available within the next year.

### The UK Gene Therapy Consortium

There were six presentations from members of our UK CF Gene Therapy Consortium including a review by Professor Eric Alton describing the run-in study and selection of patients for the forthcoming multi-dose trial of liposome-based gene therapy starting in the UK this year.

### A new animal model – the CF pig

One of the main scientific highlights of this conference was the progress with the CF pig (second plenary session) developed by researchers in Iowa and Missouri. Although at first most died early with bowel problems, recently a team of scientists reported results from five CF pigs that lived for two to six months; during this time, the animals developed some of the key features of human CF lung and pancreatic disease and also revealed that infection precedes inflammation. These animals will be of immense value in developing and testing new treatments and further understanding the role of CFTR.

In summary, the 24th NACFC was of the usual high standard. It is clear that many new therapies are being



Delegates at the North American CF Conference

evaluated for both the basic defect and to correct and control the symptoms in Cystic Fibrosis. It is a credit to all involved that so much has been achieved so quickly – undoubtedly the next five years will see many significant advances.

*Prepared in collaboration with  
Dr Kevin Southern, Reader in Paediatric Respiratory Medicine,  
Alder Hey Children's Hospital, Liverpool.*

## Research sheds more light on role of CFTR

Research funded by the Cystic Fibrosis Trust is steadily increasing our understanding of the basic defect in Cystic Fibrosis. One such example is a recently-concluded project under Dr Mehta at the University of Dundee, which has published some very interesting findings that may shape future treatments into inflammation in the CF lung. This project, a continuation of findings by Dr Mehta in previous work funded by the CF Trust, cost around £124,000 and ran for two years and nine months.

Dr Mehta and his team investigated how inflammation in the lung (that causes the progressive damage in Cystic Fibrosis) is controlled at a molecular level. Cystic Fibrosis is caused by the defective CF gene producing a mutated version of the protein Cystic Fibrosis Transmembrane Conductance Regulator (CFTR), which controls the movement of sodium and chloride through the linings of cell walls in the body. However, the very presence of a healthy (non-CF) CFTR may itself induce other effects. This type of CFTR is sometimes called the wild-type CFTR.

The researchers have found that the wild-type CFTR that is found in healthy people not only controls this movement, but that it is also key to suppressing inflammation in the body even when massive pro-inflammatory signals are present. In people who don't have Cystic Fibrosis, the correct CFTR protein

strongly keeps inflammation at bay. In people with Cystic Fibrosis therefore, not only does the lack of sodium and chloride movement cause the lungs to become clogged with thick mucus, attracting infection, which the body tries to fight off causing lung damaging inflammation, but also this inflammation is not reduced properly because the healthy protein form of CFTR is missing from most people with Cystic Fibrosis.

This failure of suppression idea resonates with independent work in the US at Johns Hopkins in Baltimore, which agrees with the idea behind Dr Mehta's findings. His findings could also explain recent data from Professor Elborn's group in Belfast that patients in whom the CFTR is present in the cell, but cannot move chloride nevertheless have less damaged lungs than those whose CFTR is almost completely destroyed. Dr Mehta and his team are now on track to work with international colleagues to determine the mechanism, and this new venture has already begun to bring together scientists who are working on the proteins that control the transport of sodium and chloride and those who work on those very same proteins in inflammation, as it appears as if they are more closely linked than previously believed. This work may be an important step in determining new treatments in the Cystic Fibrosis lung with view to creating novel ways of tackling CF lung inflammation.

*John Devlin, Communication & Policy Manager, Cystic Fibrosis Trust*

# New horizons

Last October, BBC2's *Horizon* looked at how gene therapy for a range of conditions has progressed since the human genome was mapped ten years ago. The programme featured 23-year-old **Sophie Longton**, who has Cystic Fibrosis and works as an academic mentor. As Sophie explains, the experience gave her renewed optimism for the future.

The whole *Horizon* experience was an amazing journey for me. Getting access to the science behind my condition and being able to share that with the public was an honour. Everything from interviewing Professor Eric Alton to filming with my students was interesting and of value.

The filming took place over five days, the first being at school, which was very exciting for the students who were not used to seeing 'Miss Longton' being followed around by cameramen. I was also filmed while out jogging, something I do every day to help clear my lungs. It was great fun but exhausting, as I had to keep running up and down the lane in order to get the shot right.

It was fascinating learning about foetal gene therapy as I didn't even know this research was taking place. To think that one day Cystic Fibrosis could be cured by a simple injection before a baby is born is mesmerising. *Horizon* also gave me the pleasure of meeting nine-year-old Rhys Evans, the 'boy in the bubble' and the first child in the UK to be successfully treated by gene therapy. Professor Adrian Thrasher, who treated Rhys and who is President of the British Society for Gene Therapy, also appeared in the documentary. Talking to him and Rhys' parents about their positive experience of gene therapy brought me a lot of informed optimism.

Interviewing Professor Eric Alton was most certainly the highlight of the filming. I have often read about the research he does along with the rest of the UK CF Gene Therapy Consortium, and to actually meet him and ask questions was a fantastic opportunity that I realised I was lucky to have. He really gave me courage to continue working hard at managing my condition and keeping my lungs as clear as possible. My standout moment was when we were talking about the future possibilities to treat CF, and he said, "*If anyone can make it happen... we will!*"



Sophie with Professor Alton during filming

*"I am more inspired than ever to stay as healthy as possible."*

Professor Alton also accompanied me to see the pharmacists preparing the gene therapy in a lab, who let me hold the vial which contained the product. This was a very significant moment for me; I almost wanted to take it home and inhale it through my nebuliser! I had so many questions to ask about the trials – importantly, why everything was taking so long. Professor Alton explained just how difficult it is to get a healthy copy of the gene into the right place in the cells lining the airways, showing me under a microscope the cilia cells in the lungs and where the gene has to get to in order to be effective. This made me realise the challenges the researchers are faced with.

I also asked about the positive results of the pilot study for CF gene therapy. I was aware that in some cases the new, normal CFTR protein produced in the nose and lungs following gene therapy was up to the level of that in people without Cystic Fibrosis. I wanted to know why this was only seen in a few cases, but this will only become clear with further, multi-dose trials, which are due to start later this year. In some ways however, this news was encouraging, as after the trial finishes in 2013 we should have the results, giving us a much clearer picture of where the research is at.

Learning first-hand about the work that the Gene Therapy Consortium does made me realise just how revolutionising CF gene therapy could be. The experience has really made me believe that one day it will become a reality and eventually change the lives of people living with Cystic Fibrosis. To think that my treatment load could be reduced from five nebulisers and two hours of physiotherapy a day to just one nebuliser a month is simply incredible.

During the filming I met one lady who works for the Consortium and has been involved in Cystic Fibrosis research for over fifteen years. Speaking to her and meeting others out there working so hard to treat the disease I am fighting was very moving and gave me a lot of strength. I can say for sure, having met Professor Alton, that I am more inspired than ever to stay as healthy as possible, so that should gene therapy for Cystic Fibrosis become a reality I will be able to benefit from it.

The positive response I received from people about the documentary has been overwhelming. I have received messages from people with Cystic Fibrosis, parents of children with CF and strangers that didn't know what the condition was before they watched the programme. All the responses have been heartening and everyone I have spoken to said they learnt something from watching the documentary. I am truly grateful for the experience and I hope the documentary inspired others living with Cystic Fibrosis to try and keep as well as possible and remain positive about the future.

Last November, the Cystic Fibrosis Trust Scientific Advisory Committee (the body set up to monitor the work and progress of the UK CF Gene Therapy Consortium) met in London, and the Consortium presented the findings of their work and the latest results from the pilot study in gene therapy.

So far, 27 people with Cystic Fibrosis have received a single dose of the gene therapy in the pilot study. This was the number anticipated at the start of this work. As previously reported in *CF Today*, the pilot study is primarily aimed at testing the safety of gene therapy and to determine the dose to be taken through to the multi-dose trial.

As there had been a mild reaction to the 20ml dose of gene therapy (see *CF Today*

Winter 09/10), the team had continued to test two other doses, and to pre-treat people with a normal painkiller (ibuprofen). People in this second group were given either 10ml or 5ml doses of gene therapy to see if this would reduce the reaction.

The Consortium reported that the lower doses had indeed reduced this reaction, but had not removed it entirely. It is worth remembering that this reaction was noticed far more by the clinicians and scientists than by the people with CF who had taken part.

On the advice of the Scientific Advisory Committee, the Consortium is going to give a further 5ml dose to three more people with Cystic Fibrosis in the pilot study so they can decide which dose to

take through to the multi-dose trial. There is a need to balance the dose that will likely give the least reaction, with the need to ensure enough of the gene is transferred to the patient so any potential clinical benefit can be measured.

At the same time, the Consortium has been undertaking the multi-dose toxicology study in animal models to ensure that giving repeated doses of gene therapy will not be harmful. These trials are ongoing and their results, along with the outcome and decision on dosing, will be presented at the next meeting of the Advisory Committee in the early part of this year. If all of the questions are answered satisfactorily, the multi-dose clinical trial will begin in the latter part of 2011.

*John Devlin, Communication & Policy Manager, Cystic Fibrosis Trust*

## Where there's a will

As we mentioned in the last issue of *CF Today*, the Cystic Fibrosis Trust has launched a campaign called 'Rosie's Lasting Legacy' to encourage people to think about leaving a gift to the CF Trust in their wills. We all know we should make a will, but it is often one of those things we put off. For those of you who would like to know more about the process as a result of this campaign, we asked Wills and Probate Solicitor Mel Finlay of Sweeney Miller Solicitors to answer some of your questions about making and changing wills.



Mel Finlay

law your partner will have no entitlement to anything from your estate. Instead everything would be given to your blood relatives. Where there are no immediate blood relations it is possible that your estate could be depleted to pay for the expense of locating beneficiaries.

### Q Is it time to update my will?

A It is wise to review your will every two to five years and upon any significant changes in your family or financial circumstances, so that it remains up to date.

### Q Why is it important to have a will?

A A will is one of the most crucial documents a person creates. When you die, your property, financial and personal affairs must be dealt with. Making a will ensures that your specific wishes are carried out, allowing your property, money and other important personal possessions to be passed onto who you choose. If you have children under 18 years (or 16 in Scotland) then a will enables you to appoint their guardians.

### Q What happens if I don't have a will?

A If you die without a will then your estate will be distributed according to the law of intestacy, meaning it is possible that some or all of your estate could be given to someone whom you would not want to benefit. Unmarried couples are particularly vulnerable if they do not have wills – under the

### Q Is it expensive to make or update my will?

A On average it will cost £100-£200 to have a professionally drafted will and perhaps slightly less to update your will. In order to save money you may be tempted to make a will yourself on a form bought from a stationer or to employ the services of a cheaper will writing agency. However, there are hidden complexities to will making and preparing your will with the assistance of a solicitor ensures that it covers all aspects of your assets, responsibilities and circumstances.

*When your solicitor is writing or updating your will you can ask them to include a donation to your favourite cause. To find out more about including a gift to the CF Trust in your will or to order a copy of our legacy booklet, please contact Sue Whitehead at [swwhitehead@cftrust.org.uk](mailto:swwhitehead@cftrust.org.uk) or 020 8290 8051.*

# Founded in 1964, the Cystic Fibrosis Trust is the UK's only national charity dealing with all aspects of Cystic Fibrosis.

## Our objectives are to:

- Fund medical and scientific research to develop a cure and provide effective treatments for Cystic Fibrosis.
- Ensure appropriate clinical care for those with Cystic Fibrosis.
- Provide information, advice, support and, where appropriate, financial assistance to anyone affected by Cystic Fibrosis.

## Cystic Fibrosis Trust

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[www.cftrust.org.uk](http://www.cftrust.org.uk)

You can view *CF Today* online and download our extensive range of factsheets and booklets providing further information about Cystic Fibrosis at

[www.cftrust.org.uk/aboutcf/publications](http://www.cftrust.org.uk/aboutcf/publications)



## Cystic Fibrosis Trust Helplines

**Our Support Service has three Helplines offering the following services:**

For information and advice about benefits and how to apply for them.



**Benefits Advice**  
0300 373 1010

For a *confidential* service that enables anyone to obtain information, advice and support on any aspect of Cystic Fibrosis –



**CF Helpline**  
0300 373 1000

For information and advice on how to access small grants from the Cystic Fibrosis Trust and other organisations –



**Welfare Grants**  
0300 373 1020

Our Helplines operate from 9am – 5pm weekdays. An answer machine is available during busy periods and outside these hours.

You can also access our website [www.cftrust.org.uk](http://www.cftrust.org.uk) to find out more about CF Trust Helplines and to download various forms and factsheets relating to these services.