ISSUE 2 AUGUST 2014

CF NEWSLETTER

NHS Lothian

SCOTTISH ADULT CYSTIC FIBROSIS SERVICE - EDINBURGH

Welcome to the second edition of our CF Newsletter, which aims to update everyone on CF service changes, news, research and publications.

We would like to include a patient section in your newsletter. If you would like to contribute please contact us in the CF Office on:
Tel– 0131 537 1762

INSIDE THIS ISSUE:

PEER REVIEW

The Scottish Adult CF service is undergoing Peer review in November 2014. This is a review of the service carried out by the CF Trust in partnership with the British Thoracic Society (BTS). We have previously been through these processes in 2007. The CF Trust usually reviews every service in the UK every 5 years.

The reviewing team look at all aspects of the service from protocols and quality outcomes to patient feedback. The visiting team consists of CF Doctor, nurse, and a number of other team members including Physiotherapist, Dietitian, Pharmacist or Psychologist, as well as members from the CF Trust.

You may have received a “patient feedback questionnaire” in May 2014 asking about your views on the service. Thank you to those who have completed the patient survey. We will receive the results of this survey in November, when the team visit us.

Over the last few months we have been collecting lots of information on our protocols and processes, so that the visiting team have information that they require to make a full assessment of our service. In November the team will visit the Unit and look at clinic and ward areas and interviews members of the CF team, having read all the information we have provided in advance.

The final outcomes of the CF Peer review report will be posted on the CF Trust website in December 214. We will let you know the outcome in the Winter CF newsletter.

Helen Rodgers

Director Scottish Adult CF Service

* VOLUNTEERS WANTED *

My name is Kath MacDonald and I am a lecturer in Nursing at Queen Margaret University. I have just completed a doctorate in Health and Social Science. As a previous CF nurse I was keen to do research in a subject I was familiar with. Thus I was delighted to return to the CF unit to explore what partnership meant to young people with CF and their CF team.

I chose this topic for several reasons. I believe that the nature of the relationships people with CF have with their team is a unique one. Many of you have been in those relationships for a long time and you know each other very well. Another difference is that many of you know so much about your disease, that you can teach us a thing or two. The literature describes people with long term conditions like CF who are very knowledgeable about their disease as "expert patients" and I wanted to explore this a bit further in my study. Finally I wanted to look at what helps and what hinders partnership between you and the team.

Some of you may remember me being in attendance during your clinic visits last year and interviewing you afterwards to discuss your views on partnership with the CF team and your thoughts on "expert patients."

Firstly I want to thank you all for giving of your time. It was a great experience for me and I saw some really good examples of young people and the CF team working together, sharing knowledge, making decisions together and meeting in the middle.

Here are the main findings of the study:

- Most of you and the CF team believed that the relationship they have is a partnership.
- The relationship was viewed as unique by the CF team, because of its long length, the fact that you are young (mostly) and the team see you through many different stages of life.
- Partnerships were not always seen as equal, and sometimes the team thought that you were more powerful and sometimes it was the other way round.
- CF nurses were seen as the link people in the partnership; they are usually the first people you call, and know lots about you as a person, because they see you at home and in hospital.
- Sometimes the team have different priorities to you: theirs is often about making sure you get the best treatment, whilst yours is about managing other things, not just CF.
- The CF team work hard to build relationships with you in order to find ways to negotiate treatments.
- Trusting each other is important so that you can both be honest with each other.
- Most of you don't like the term "expert patient" but prefer to use the word "experienced" to describe yourselves.
- This experience involves experimenting, (e.g drug times, doses), monitoring (side-effects), organising and managing many staff, appointments, supplies, having knowledge of self and of the systems in the NHS.
- Sometimes you come to clinic with an issue (e.g. needing IV's) but don't always come out and say it. Instead you give clues and wait for the doctor to suggest it.
- The clinic is long, repetitive and most of you want to get in and out as quickly as possible.
- Weekend care (inpatient and outpatient) is not as good as during the week, so most of you prefer to hang on till Monday before contacting the team.

What next?

I have talked to the team and they are very keen that we take some of these findings forward to look at ways in which we can work together to help develop partnerships and standards of care.

We cannot do this without you. Thus we hope to be able to work with some of you in a new project. As long as you have access to the internet, we hope that you will be able to do it from the comfort of your own home.

We are looking for about 15 people to join in some activities via a downloadable App, (e.g. online chat, surveys,) to consider what is important to you in the service and how we can work together to develop it.

If you are interested, please let one of the CF nurses know or contact me directly by text, e mail, or phone.

Thank you,

Kathy MacDonald
kmacdonald@qmu.ac.uk
Mobile: 07988 958 549

ANNUAL REVIEW FEEDBACK

Annual review is a detailed assessment of every aspect of your condition and current therapies. This allows us to assess changes over the last year, identify where treatments can be improved and help you with a plan for the following year.

From May 2013 we have been writing to you with these results, which you receive once a year. The Cystic Fibrosis Team have over the last year asked for your feedback on these letters.

We have collated all the feedback you have given us and wish to share it with you.

Annual Review Feedback – Results

41% of feedback questionnaires were returned.

- 94% understood the lung function section
- 100% understood the nutrition section
- 100% understood the test results section
- 100% understood the treatment section
- 92% agreed with medication list.

We intend to review our annual review feedback letter and are currently working to improve this. Your feedback is valuable to us; please remember to bring your annual review letter to clinic for discussion with the Consultant.

Many thanks to all patients who completed an annual review feedback questionnaire.

As part of the annual review process, the Pharmacist will access your GP medication list. This ensures that we have an accurate list of your medication, including medication for conditions that may not be related to your CF.

We require this to ensure that all your medication is up to date and compatible. The Pharmacist will contact you and your GP if there are any important issues. Anything else can be discussed at your next appointment.

Please let us know if you do not wish us to access your GP medication list.

Patient quotes

"I feel this is a good system as it saves me texting/calling the CF nurses to find out results."

"Dates would be useful; possibly a list of previous results to compare to."

"Seeing the range of healthy weights was interesting and reassuring."

"The annual review can be worrying but I think this letter is a good idea as sometimes you never felt you got much feedback from all the tests."

"It was good to end on some encouraging advice as mentally this can be very important."

TRAVEL HEALTH PLANNING

Now that the holiday season is here and summer has arrived in Scotland, it is a good time to mention the importance of fluid replacement and the risk of blockage in the bowel. In hot weather, especially if you are out sunbathing or exercising, it is very important to ensure you are well hydrated.

A rough guide is to replace 2-3 litres of water per day as a baseline. If you are exercising or out in the hot sun you may require even more than this. It is very important to remember that you also need to replace salt with any large volume of water you are drinking to avoid serious problems.

You can do this either with "Salt tablets" or oral rehydration powders such as Dioralyte®. These provide sodium and other salts which help your body to absorb water.

Slow sodium® tablets: 4-8 tablets daily with water are usually enough. Take them throughout the day, when you are in hot temperatures or exercising.

Dioralyte® - drink up to 5-10 sachets per day. Dissolve each sachet in 200ml water. Water or other fluids should also be taken to ensure adequate replacement throughout the day.

You should also ensure that you have travel insurance and the EC111 card for travel within Europe, in case anything happens. The CF Trust has a link regarding holiday insurance.

See the link: http://www.cysticfibrosis.org.uk/get-involved/ways-to-donate/online-shopping/insurance-products

TRANSITION TO ADULT CF SERVICE

Transition is a process of planning and preparing a move from Children's to Adult Services. It is important that this process happens gradually to allow time for the patient and their families /carers to get ready for this move and to plan, together with their new Adult Team, their future healthcare in adulthood. Our aim is always to put the young person at the *centre* of the process.

We understand that transition can be very stressful for patients and/or their families, and that everybody needs time to understand and develop trust in the Adult Team and their model of care. We have therefore been trying, over the last few years, to update and improve the transition process for all concerned.

We have spent time looking at the transition process for cystic fibrosis patients and for patients with other long-term conditions elsewhere in the UK, Europe and further afield. In 2005, we carried out a questionnaire of the patient/parent experience of transition and you may have completed one for us. We also introduced a *'Getting To Know You'* questionnaire in 2010. In addition, we have been working closely with our colleagues at the Royal Hospital for Sick Children in Edinburgh to reflect upon and try to modify the process of transition between our services. This has resulted in the introduction of new meetings in 2010 to share all important information before we meet our new patients.

In 2011, we decided to audit the transition process in a little more depth and this work was lead by Grainne O'Brien, Trainee Clinical Psychologist at the time. Grainne followed our transition patients over a one-year period to try to gain a better understanding of how they and their parents/ carers felt throughout this process. **This is what she found:**

Parents: What is the transition experience like?

"Recognise that child is ready for transition. But apprehensive that parents not going to be involved."

"Biggest thing is responsibility. For 16 years, I have been in charge and suddenly goes from me doing everything."

"Was right time for him. Not just a magical age, maturity age more important."

"Nobody wanted to talk to me any more about my son with CF. Felt completely shut off."

Young people: What is the transition experience like?

"Easier than expected. Expected people to be 'more serious, more formal.'"

"Feeling a lot more independent ……taking more control of medications at home."

"Been a breeze."

"Don't like it as been more unwell this year and 'more challenging with team don't know.'"

"Getting to know a new team annoying. Prefer if a year or two later as might have been easier. Expected to be adult in this avenue in life whereas at home still seen as child."

TRANSITION TO ADULT CF SERVICE

As a result of Grainne's audit findings and feedback from parents/ carers at clinic, it was decided to Improve the support provided to parents/ carers throughout the transition process. A new, informal evening for parents/ carers was arranged on 18 September 2013 to answer any questions and to alleviate any anxieties that they had about their child coming across to the Adult Cystic Fibrosis Service. A ward tour was also offered.
A small group of parents attended and you can see from the positive feedback below that the night seemed to be a success.

Would you recommend the Parents' Transition Open Evening to other parents whose child is about to transfer to the Adult Cystic Fibrosis Service? (100% 'Yes')

"It is important to see the new environment that your child is entering. Also to meet all the staff and if possible other parents of CF children."

"Nice to meet staff, see where ward is and how easy it is to get to hospital."

"Good opportunity to meet staff/allay fears/have questions answered/meet other parents."

What next?

We want to make the transition process as smooth and carefree as possible for all our patients and their families, so we will continue to involve you all to help us to get it right.

Just now, we are working on:

- Developing a booklet: *'Introduction to Adult CF Services*'.
- Updating the leaflet *'Transition to Adult CF Services'.*
- Updating our *'Getting To Know You'* questionnaire.
- Liaising further with our colleagues at the Royal Hospital for Sick Children.
- Continuing to evaluate the process.

In addition, we have planned our next Transition Open Evening for Parents / Carers of those transitioning later this year on Wednesday 27 August 2014.

If you would like to find out more or suggest any improvements to the transition process, please get in touch with:
Audrey Matthews or Catriona McMullan on (0131) 537 1762.

The Butterfly Trust

Supporting People with Cystic Fibrosis in Scotland

PERSONAL INDEPENDENCE PAYMENT

We know that having Cystic Fibrosis can restrict your life choices and we want to help.

The main issues adults with Cystic Fibrosis have been bringing to us have been about finance, employment and housing and we have been able to help on many levels. Some people are having difficulties in their workplace and don't know their rights, want to be independent but struggle to find a suitable property, need to increase their income but don't know what help they're entitled to or need a break but cannot afford a holiday.

- If you are struggling with the rising cost of everyday living expenses then give us a call. We will discuss your circumstances in confidence and look at ways that you can increase your income or find funding for something you need and cannot afford.
- We have helped people acquire their own house and buy what they need to live independently. So if you have been on the housing list for ages with no progress or you want some support to cope independently we may be able to help.
- We have helped many people with employment issues and often that help is to negotiate adjustments in their workplace, which has allowed them to stay in work. So if you are having difficulties in work do give us a call before making any life changing decisions.

The changes in the benefits system that can affect adults with Cystic Fibrosis are proving hard to understand but we can help you through that process to ensure you get a fair result and the best possible outcome for your circumstances. In particular, we know many people are having their **Employment and Support Allowance (ESA)** reviewed sooner than expected. The form must be completed and returned or the benefit will be stopped. The questions need careful consideration and appeals can be complicated and prolonged. So do ask us for advice and support before submitting your application form to ensure you retain your ESA with the minimum of effort.

The biggest benefit change to have a direct impact on adults with Cystic Fibrosis is the introduction of **Personal Independent Payment (PIP)**. This benefit is replacing Disability Living Allowance and has already been introduced in areas with the post codes EH, TD, ML. Other post codes will be included from October 2014. It is important to be aware of the following changes

PIP has two rates of the daily living component (equivalent of DLA care component) instead of three. These are Standard rate, which is the equivalent of Middle rate in DLA and Enhanced rate, which is the equivalent of High rate in DLA. The amounts paid are also similar to those of DLA.

The criteria to qualify for the mobility part are determined by ability to walk instead of ability to walk without discomfort and for pre-determined distances. This is a much more rigid approach and the distances are shorter than those preciously considered.

The application is registered by phone on: Tel- 0800 917 2222.

Then the form is sent out to be completed. This is where we can *help*.

A points system is used to determine the level of benefit awarded from answers to specific questions. We are now familiar with those and can help you to describe how your daily requirements meet the criteria used to decide the rate of award. Applicants are likely to be called for a medical assessment when applying for PIP. We can provide support at this assessment to ensure the assessor understands the full impact of the daily regime where adults often understate the time and effort required.

If your circumstances have changed and you are considering asking for your award to be looked at again you will be transferred to PIP and so will be considered under the new criteria. You then cannot return to DLA if unsuccessful. So do call us for guidance early on so we can advise you on the potential level of risk to your current award before making the request.

The referral to an external agency for the medical assessment and then returned to DWP for decision making is making the process more lengthy, stressful and we believe less efficient with an increased likelihood of an unfair outcome. Our aim is to offer help that frees up your time and energy, increases your choices and where possible maximises your income.

Of course we can also provide a massage when you are in hospital or at clinic.

Ask your CF team or call us on **0131 445 5590 or email us at info@butterflytrust.org.uk**

DIET AND EXERCISE

Food for energy

Carbohydrate is the most important source of energy for an active individual.

Starchy and **sugary foods** are excellent sources of carbohydrate.

Starchy foods should be included at all mealtimes, e.g. breakfast cereals, bread, potatoes, pasta, rice, noodles.

Sugary foods should be taken in addition to starchy foods to meet the energy demands of exercise, e.g. soft drinks, sweets, jam, honey, cakes, cereal bars, muffins, pancakes, biscuits and puddings.

Fruit and **fruit juices** also provide carbohydrate and are a good source of vitamins, minerals and fibre.

Your carbohydrate requirements will depend on the intensity and duration of the exercise. For individual advice, please discuss with the dietitian.

What about protein?

It is important to eat good sources of **protein** daily, e.g. meat, poultry, fish, cheese, eggs, pulses, nuts, Quorn, tofu, soya mince.

Your usual **protein** intake should be adequate to meet the demands of training and exercise, provided you are eating a balanced diet with sufficient carbohydrate and energy.

Note:

- Strength/resistance training is responsible for changes in muscle mass, not the amount of protein consumed.
- Protein supplements are usually unnecessary.
- At present, there is no evidence that creatinine supplementation will improve sporting performance and should therefore be avoided.

Time it right!

- Eat a well balanced meal 2-3 hours before exercising.
- Have a light carbohydrate snack one hour before exercising (e.g. fruit, cereal bar, biscuits, pancake, muffin, crisps, bagel)
- Start refuelling with high carbohydrate snacks as soon as you have finished exercising.

CF-related diabetes and exercise

Exercise will usually **lower** your blood glucose levels.

This depends on:

- Duration / type of exercise.
- Time of last meal and insulin injection.
- If extra snacks are taken before or during exercise.

Adjusting your insulin will be discussed with you on an individual basis.

IMPORTANT

Check your blood glucose level before, during and after exercise, especially if it is a new activity.

Fluids and hydration in exercise

Evidence suggests that individuals with CF who exercise lose more salt and water as they sweat.

Most people need 2-3 litres of fluid per day to remain fully hydrated. It is possible to lose 0.5-1.5 litres per hour of sweat during exercise. Sweat losses may increase further in hot/humid weather.

IMPORTANT

Start exercise fully hydrated, aiming to drink 500-600mls of fluid two hours before exercising, followed by an additional 150-350mls, depending on tolerance, before exercise commences.

What should I drink?

The following advice is only a guide; your individual hydration needs can be discussed in more detail with the dietitian.

Exercise ≤30 minutes	Exercising ≤30 minutes at high intensity and/or in warm weather	Exercise ≥30 minutes
Water or hypotonic drink (e.g. R2, Lucozade Sport Lite)	Water or isotonic drink (e.g. Powerade, Lucozade Sport, Isostar)	Isotonic drink

IMPORTANT

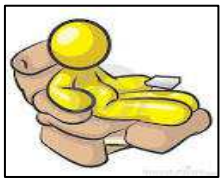
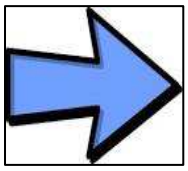
Avoid carbonated drinks for hydration during exercise.

- Aim to take 150-200mls fluid every 15-20 minutes during exercise.
- Start rehydration immediately after exercise.
- Consider additional salt supplementation during prolonged exercise and/ or in very warm weather. (refer to travel health planning article.)
- Avoid stimulant drinks– there is no evidence that they will improve sporting performance.

GET EXERCISING

GLASGOW 2014
XX COMMONWEALTH GAMES

Rather than watching the Commonwealth Games from the couch, how about setting your own sport's challenge?


Activities you could choose from include: athletics, swimming, cycling, badminton, weight lifting, table tennis – take your pick!

Here is one example of a beginner's guide to preparing for walking/jogging a 5 kilometre race.

RUN.com | 5K WALK/JOG TRAINING PROGRAM: NOVICE'S PLAN

WEEK	MONDAY	TUESDAY	WEDNESDAY	THURSDAY	FRIDAY	SATURDAY	SUNDAY	TOTAL
1	WALK 20 MIN.	WALK 20 MIN.	WALK 20 MIN.	REST DAY	WALK 20 MIN.	WALK 30 MIN.	WALK 30 MIN.	WALK 2:20
2	REST DAY	WALK/RUN 30 MIN. EVERY 4TH MIN. RUN FOR 1 MIN.	WALK/RUN 30 MIN. EVERY 4TH MIN. RUN FOR 1 MIN.	WALK 4 MIN. RUN 2 MIN. REPEAT 5X	REST DAY	WALK 4 MIN. RUN 2 MIN. REPEAT 5X	WALK 4 MIN. RUN 2 MIN. REPEAT 5X	WALK 1:46 RUN 44 MIN. TOTAL 2:30
3	WALK 4 MIN. RUN 2 MIN. REPEAT 5X	REST DAY	WALK 3 MIN. RUN 3 MIN. REPEAT 5X	WALK 3 MIN. RUN 3 MIN. REPEAT 5X	WALK 3 MIN. RUN 3 MIN. REPEAT 5X	REST DAY	WALK 3 MIN. RUN 3 MIN. REPEAT 5X	WALK 1:20 RUN 1:10 TOTAL 2:30
4	WALK 2 MIN. RUN 5 MIN. REPEAT 5X	WALK 2 MIN. RUN 5 MIN. REPEAT 5X	REST DAY	WALK 2 MIN. RUN 5 MIN. REPEAT 5X	WALK 2 MIN. RUN 5 MIN. REPEAT 5X	WALK 2 MIN. RUN 8 MIN. REPEAT 4X	REST DAY	WALK 48 MIN. RUN 2:12 TOTAL 3:00
5	WALK 2 MIN. RUN 8 MIN. REPEAT 4X	WALK 2 MIN. RUN 8 MIN. REPEAT 4X	WALK 2 MIN. RUN 8 MIN. REPEAT 4X	REST DAY	WALK 3 MIN. RUN 12 MIN. REPEAT 3X	WALK 3 MIN. RUN 12 MIN. REPEAT 3X	WALK 3 MIN. RUN 12 MIN. REPEAT 3X	WALK 51 MIN. RUN 3:24 TOTAL 4:15
6	REST DAY	RUN 18 MIN. WALK 2 MIN. REPEAT 2X	RUN 18 MIN. WALK 2 MIN. REPEAT 2X	RUN 18 MIN. WALK 2 MIN. REPEAT 2X	REST DAY	RUN 22 MIN. WALK 3 MIN. RUN 15 MIN.	RUN 22 MIN. WALK 3 MIN. RUN 15 MIN.	WALK 18 MIN. RUN 3:02 TOTAL 3:20
7	RUN 30 MIN.	REST DAY	RUN 30 MIN.	RUN 35 MIN.	RUN 30 MIN.	REST DAY	RUN 35 MIN.	RUN 2:40
8	RUN 40 MIN.	RUN 30 MIN.	REST DAY	RUN 35 MIN.	RUN 45 MIN.	RUN 30 MIN.	REST DAY	RUN 3:00
9	RUN 45 MIN.	RUN 30 MIN.	RUN 45 MIN.	REST DAY	RUN 10 MIN. EASY 6 X 30 SEC. AT A FASTER PACE (WALK A FEW METERS BETWEEN EACH) 5 MIN. EASY	RACE DAY! 5K RACE (OR REST DAY IF RACE IS ON SUNDAY)		RUN 2:20 + RACE TIME

There are many other programmes/ apps available to help you with your 'sport's challenge,' although they may not be suitable for everyone. Always exercise within your own limits. Why not ask your physiotherapist at your next clinic? Whatever your fitness level, we are always here for advice.

Just remember that regular exercise:

- Increases energy and fitness levels
- Improves strength and posture
- Helps keep your bones healthy
- Gives your self confidence a boost
- Improves sleep
- Gives you the feel good factor
- Can be fun!

THE CYSTIC FIBROSIS GENE THERAPY TRIAL

Gene Therapy Trial – Data Collection Complete

At the end of May we celebrated a landmark in this major research effort – the final patient visit to the Wellcome Clinical Research Facility. This trial has been many years in the planning, and I am pleased to say that thanks to the enthusiasm and commitment of many patients, a remarkable total of 46 patients contributed completed data to the trial at the Edinburgh research centre. An additional 34 patients made some trial visits but for a variety of reasons were not able to complete participation in the trial. Well over 700 patient visits were made to the Wellcome CRF during the 2 years that the trial was active, with individual patients contributing up to 13 visits. Many patients travelled repeatedly from distant parts of Scotland, and several from the Newcastle adult centre.

The Gene Therapy Research team want to say a huge "thank you!" to the patients and their families who gave so generously of their time and effort to help us complete this major undertaking. Your commitment and effort was fantastic given the large amount we asked of our trial patients. No matter how cunning the scientists are we can never make treatment progress without the support of our patients for clinical trials.

We are now in the final stages of collecting together all the data, and combining it with the data from the London research centre, then professional statisticians at Imperial College will begin the complex task of analysing the results. We hope to be able to announce the first results at the North American CF Conference in Atlanta in October, and will keep our trial participants fully informed of the outcome.

Thanks again from the team for all your support for this innovative work

Alastair Innes

Further CF Gene Therapy information is available at:

www.cfgenetherapy.org.uk

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Cystic Fibrosis our focus

The CF Trust is the only UK-wide charity making a daily difference to the lives of people with cystic fibrosis, and those who care for them.

Explore the website below to find out everything you need to know about the trust; from our core belief and why they are here.

www.cftrust.org.uk

CYSTIC FIBROSIS TEAM

Out-of-Hours: Ward 54: (0131) 537 1788

CF Consultants

Dr Helen Rodgers	Secretary: (0131) 537 1779
Dr Alastair Innes	Secretary: (0131) 537 1783
Dr Maeve Smith	Secretary: (0131) 537 1781

CF Fellow

Antonia Tasiou	CF Office: (0131) 537 1762

CF Nurses

Catriona McMullan	CF Office: (0131) 537 1762
Lynne McIntosh	CF Mobiles: 07770 326301/
Julie Robertson	07770 326302

CF Psychologist

Dr Audrey Matthews	Secretary: (0131) 537 1783

CF Pharmacist

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Lianne Robb	

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Nicola Duncan	

CF Data Manager

Dawn Wilson	Resp Office: (0131) 537 1108

CF Secretary

Myfanwy A. Bradley	Resp Office: (0131) 537 1783