ISSUE 1 JANUARY 2014

CF NEWSLETTER

NHS Lothian

SCOTTISH ADULT CYSTIC FIBROSIS SERVICE - EDINBURGH

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

Please send any information or feedback to:

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Anne Ferguson Building,
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Crewe Road South
Edinburgh
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INSIDE THIS ISSUE:

PROFESSOR GREENING RETIRES

Professor Andy Greening
Professor of Pulmonary Diseases

Time catches up with us all! It seems like only yesterday that I started work as a Consultant Physician in Lothian but the reality is that it is 29 years ago.

Shortly after I took up appointment, Sandy Raeburn, who worked in the human genetics unit, asked me if I would join him in helping to run the newly emerging service for Adult Cystic Fibrosis patients. This I did, but relatively shortly afterwards Sandy moved to a chair of genetics in Nottingham. Over the next three years I was fortunate enough to obtain support from the Cystic Fibrosis Trust and we developed the service with one CF liaison sister and one CF clinical fellow, until in 1992, we were able to persuade the Scottish Office that Cystic Fibrosis was changing and rapidly becoming an adult predominant clinical issue, and we got the go-ahead to set up the National Adult Cystic Fibrosis Service.

We persuaded the then Health minister, Michael Forsyth, that this should actually be based in two centres, one in Edinburgh and one in Glasgow and shortly after that a smaller service in Aberdeen, for geographical reasons. With this new national-based service, we were able to obtain funding that allowed proper development of the service with increased members of staff to cover medical, nursing, physiotherapy, dietetic, and pharmacy support, and at a later date psychology support. There was further evolution of the service with the formation of an outreach service in Dundee.

It still surprises me how the overall service has grown. In 1992 I think we had about 55 patients attending the service and currently there are over 180 patients linked to the Edinburgh service, with an additional 45 patients in the Dundee outreach clinic. A few of you will still remember those very early days but now there are many more and new members of the service and, indeed, I suspect I have yet to meet all the people who have very recently transitioned from the paediatric service.

One of my joys has been the increase in the superb team that now operates the service. I have had excellent colleagues clinically with Dr Alastair Innes and then Dr Helen Rodgers, and more recently Dr Robert Gray, who is based at the University. I will be replaced by Dr Maeve Smith and I am most pleased that this will leave the service in very good medical hands. I have to emphasise, however, that the service is a team and it would be hard for me to imagine a better group of specialist nurses, physiotherapists, dietitians, pharmacists and psychologists than those we have had working with the service. It has been an enormous privilege to work with you in our adult service, which is a truly National Service, since there are patients from all four corners of Scotland. Some attend the clinic in Edinburgh and others attend outreach clinics in Inverness.

Thank you for allowing me to work with you over the past 20+ years. I shall miss you and the CF clinics and the CF team. However, I will keep a distant watching brief on the service and I wish you all the very best for the future years.

Yours sincerely,

ANDREW P GREENING
Professor of Pulmonary Diseases

NEW CYSTIC FIBROSIS TEAM MEMBERS

CF CONSULTANT– Dr Maeve Smith

I am delighted to be joining the CF team as a Consultant. Some of you will know me from my years on the ward as a Specialist Registrar. I had really enjoyed my time as a Specialist Registrar in the CF team, and it is great to be back permanently in the CF team. Looking forward to meeting you all.

CF SPECIALITY DOCTOR– Dr Antonia (Toni) Tasiou

I studied at the University of Athens in Greece and have trained in both Respiratory Medicine and Intensive Care Medicine. My work in the past has brought me in contact with Cystic Fibrosis patients and I look forward to my new commitment in the Cystic Fibrosis team.

CF CLINICAL PSYCHOLOGIST– Dr Audrey Matthews

Having fallen in love with Edinburgh and the Fringe Festival on a trip to Scotland in 1997, I moved over from Ireland in 1999 to gain work experience in Clinical Psychology. I went on to train at the University of Edinburgh, specialising in physical health conditions and I have worked mainly in chronic pain, drug addiction, HIV and sexual health since qualifying. In June 2013, I was delighted to have the opportunity to come and work one day weekly with the Cystic Fibrosis Team, which has allowed me to meet many of you already. Having greatly enjoyed this secondment last year, I am now delighted to be a fully-fledged and more permanent member of the CF Team! I look forward to working with you all.

Butterfly Trust

The Butterfly Trust provides a variety of 'support services in the community for people affected by Cystic Fibrosis.

They provide advice on the following:

- ◊ **Information and advice**
- ◊ **Benefits and DLA**
- ◊ **Housing**
- ◊ **Employment**
- ◊ **Access to financial support**
- ◊ **Other specialist services**

www.butterflytrust.org.uk

E-mail – info@butterflytrust.org.uk

THE CYSTIC FIBROSIS GENE THERAPY TRIAL

The UK CF Gene Therapy Consortium began a large multicentre trial investigating gene therapy for CF lung disease in June 2012. This follows on from the successful 'Run-In Study' over the last few years, which was carried out to select the appropriate patients and tests required for this large multidose trial.

The CF Gene Therapy Multidose trial is taking place at the Western General Hospital in Edinburgh and the Royal Brompton Hospital in London, although patients have been recruited from all over the UK.

Patients attend on a monthly basis to receive either nebulised gene therapy, or a placebo. The trial is 'double-blinded' so neither the patients nor the staff are aware as to which group a patient has been randomly assigned. After a total of 12 monthly dosing visits, patients attend for 2 follow-up visits to assess any changes in their lung function and general health.

We have successfully recruited the 120 patients required between the 2 centres, and approximately half of these patients have now completed the trial. The trial will finish when the last patient completes their follow-up, which is expected to be in June 2014. There will then follow a period of analysis as we try to identify any difference between the 2 groups. The results will be reported in the medical literature.

A trial of this nature would not be possible without the commitment and determination of all the patients in the trial and their families, many of whom have to travel considerable distances each month in order to attend their appointments.

All members of the UK CF Gene Therapy Consortium would like to express their huge gratitude to everyone who has participated in the trial. We would also like to thank the local CF teams of all our patients for their continuing help with our research.

Further CF Gene Therapy information is available at: www.cfgenetherapy. org.uk

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To keep up-to-date with how the trial is progressing, please see our website: **http://www.cfgenetherapy.org.uk**

CHANGES TO MEDICAL COVER IN WARD 54

Emergency reviews only on Saturday and Sunday.

All other reviews Monday – Friday

WEEKEND COVER

- The medical cover at weekends has changed and it is important for you to know that there is not a Respiratory Consultant on every weekend although there is always a Medical Consultant available.
- If you are unwell and need to be seen, it is essential that you contact Ward 54 by 10am in the morning, if possible, to arrange to be seen.

We appreciate that it is not always possible to predict when you need our service. Of course we would always arrange to see you and admit you to Ward 54 if you are acutely unwell. We would appreciate your help in contacting and arranging a review on the ward early in the day if possible. We appreciate your help making the CF service at the Western General Hospital run efficiently and continue to do so.
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ANNUAL REVIEW FEEDBACK LETTERS

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 produce a plan for the following year.

ANNUAL REVIEW INVESTIGATIONS
We have recently started to send annual review letters, which update you on your recent results. This began in May 2013 and you will receive this letter around the month of your birthday.

The annual review letter includes sections on lung function, nutrition, results on sputum, blood test, vitamin levels, bone scan and ultrasound scan. We also list your current medications, give advice regarding your treatments and have a general comments section.

The annual review letter is discussed with the full Cystic Fibrosis Team and written by one of the Cystic Fibrosis Consultants.

The annual review letter will be discussed with you at your next clinic appointment. You are encouraged to bring the letter with you so we can discuss the contents, answer any questions and agree a treatment plan for the following year.

We enclose a short questionnaire with the annual review letter, which we ask you to complete and bring with you to clinic.

Your feedback is valuable to us.
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**We would be very grateful if you would complete a feedback questionnaire when you receive one.**

**ANNUAL REVIEW INVESTIGATIONS**

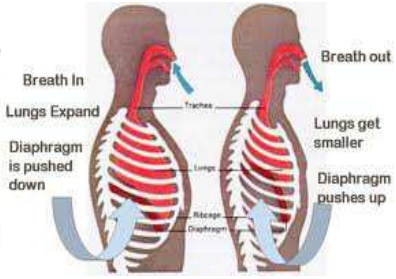
| Investigation | Reason | Frequency |
|---|---|---|
| Abdominal Ultrasound Scan | To screen for CF liver disease. | 1-3 yearly |
| DEXA Bone Scan | To screen for CF-related bone disease. | 3 yearly |
| Chest x-ray | To check the health of your lungs. | Annually |
| Oral glucose tolerance test | To screen for CF-related diabetes. | Annually |
| Exercise assessment | Lung function assessment during exercise. | Annually |
| Sputum culture samples | To check what is growing in your lungs. | Every visit |

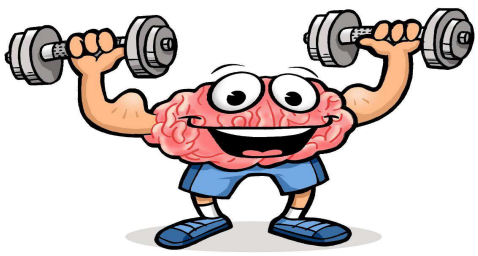
# PHYSIOTHERAPY UPDATE– REVIEWING IV'S

As advised by the national standards of care for CF, our physiotherapists would like to review you when starting IV antibiotics. The aim is to ensure you are doing the best treatment to help you through your infection.

This may include advice on:

- Airway clearance technique
- Timing of inhaled medications
- Exercise programme.



If you are being admitted, the physiotherapist will treat you on the ward as usual. If you are being reviewed as an outpatient, the physiotherapist will aim to see you during this visit to discuss your self management.

They hope to catch everyone but this will depend on staff availability.
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DIETETIC UPDATE– VITAMIN D

Role of vitamin D in health

Vitamin D is essential for normal bone health. This is because vitamin D promotes calcium absorption and contributes to calcium and phosphate balance in the blood. Recently there has been interest in the potential role of vitamin D in lowering the risk of several chronic conditions including cancer, cardiovascular disease, auto-immune dysfunction and multiple sclerosis.

Factors contributing to vitamin D deficiency

In the UK, vitamin D insufficiency can probably be explained by weather patterns, sun exposure, oily fish consumption and obesity.

The body can only make vitamin D from May to September, when UVB light is of the correct spectrum. However, time of day, cloud cover and pollution all reduce the impact of UVB light. Unfortunately, weather patterns in recent years have resulted in fewer hours of sunlight during the summer months and more rain. On top of this, concerns about risk of skin cancer have encouraged people to cover up or avoid the sun and to use sun cream.

Obesity can limit circulating levels of vitamin D, therefore high obesity rates seen in the UK over the past few decades have had the added effect of contributing to vitamin D insufficiency.

Low intakes of oily fish – the best natural source of vitamin D – limits the topping-up effect of diet, especially in winter. Studies show that only 25% of adults consume oily fish.

What can I do to improve my vitamin D status?

- Between May and September, expose your arms and face to the sun for 15 minutes a day before taking usual precautions against sunburn (darker skinned people will need more time)
- Take part in regular weight-bearing exercise, avoid smoking and excessive alcohol consumption
- Aim to eat a balanced diet, containing nutrients from a variety of food sources
- Eat oily fish once a week (140g portion)
- Try to have eggs and foods fortified with vitamin D on a regular basis
- Take your vitamin supplements every day as prescribed

Your vitamin D level will be checked as part of your annual review and will be discussed with you at clinic.

PHARMACY UPDATE

Ivacaftor

Ivacaftor is an exciting new treatment, taken as a tablet each day. It corrects the underlying defect in CFTR channels, which is the cause of symptoms in CF. It has been shown to have many beneficial effects to lung function and nutrition.

We do not yet know the long term effects or if this means people can stop taking their other treatments yet. It is only suitable for about 8% of our population.

All relevant people have been contacted and have either started treatment or are in discussions with the team.

"Two new inhalers have been approved."

New Inhaled Therapy

Two new inhalers have been approved:

- Colobreathe (Colomycin)
- Tobi Podhaler (Tobramycin)

These are both dry powder inhalers where you inhale the contents of a capsule twice a day and there is no need to nebulise.

You may be offered the option to switch to one of these new products at clinic. A test dose is required to check they agree with you. If you wish to discuss switching, then please mention it at your next appointment.

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

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Nicola Duncan	

CF DATA MANAGER

Dawn Wilson	Resp Office: (0131) 537 1108

CF SECRETARY

Myfanwy-Ann Bradley

Resp Office: (0131) 537 1783

THE CF TRUST

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

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