

Newsletter

Alpha1 Awareness

Vol 2
Issue 2
April 2010

A Word from the Chairman



Plans are well underway for the Fifth European Alpha1 Congress which will be held in London on the 9th and 10th of July.

All members of Alpha1 Awareness UK, along with their family and carers, are invited to come to the congress to meet other Alphas and to learn more about the condition and the provision of health support.

Accommodation in London and meals during the Congress will be paid for. We can also help with travel expenses. We hope that many of you will be able to take this opportunity to enjoy the company of other Alphas and to talk about issues important to you.

This event is also the highlight of the year for the Alfa Europe Federation.



It is an occasion for representatives from European Alpha1 associations to meet, discuss common problems and to find out how AATD is researched, diagnosed and treated in the host country. The Congress is preceded by the General Assembly of the member associations at which the annual reports are presented, plans for the coming year are discussed and the Federation officials are elected. The Federation business is conducted in English.

London 2010

Alan Heywood-Jones

Details of the Congress and how to book to attend can be found at the end of this Newsletter

Note: The spelling Alfa is not a mistake – the official name of the Federation uses this spelling; it is more understandable by the majority of European Alpha associations.

Drug Trial for ADAPT at Queen Elizabeth Hospital, Birmingham.

Before I begin writing about taking part in the drug trial, I thought you may like to know a little about me. My name is John Morton; I'm 55 and live in Bristol. I'm a PiZZ Alpha. Some of you may have known my late brother Paul Morton. I was always a "sickly" child and was first in hospital, with Pneumonia aged five. Since then I've been in hospital every three or four years, always with chest infections. Most recently I was in Southmead Hospital in December 2009 for a fortnight having IV antibiotics three times a day for a Pseudomonas chest infection.

The trial I'm taking part in is the Kamada AAT trial for nebulised Alpha-1 Antitrypsin for patients with Emphysema. The trial involves 10 hospitals in 6 countries. Birmingham is looking for 40 patients to take part. It is hoped that the drug will reduce the frequency and severity of any chest infections.

The trial last for 59 weeks and involves about 10 visits to Birmingham. During the first visit the procedures involved and any risks are fully explained, if you agree then the screening process can begin. The whole process takes about 3 hours. The screening involves a full medical including, blood pressure, heart rate, breathing rate, temperature, height and weight measurement. Some lung function tests, an ECG, a urine sample and some 4 blood samples.

One of the blood samples is used to check for HIV and Hepatitis. This check is because the drug is made from human blood plasma. The risk of contracting HIV is very remote, as it is with all blood products. In this case because the drug is inhaled and does not enter the blood stream the risk is even further reduced.

Two weeks later is the training visit. I was shown the battery operated nebuliser being used in the trial. It's very small and fits in the palm of you hand and its SILENT!!! Not at all like the noisy table top monster I use for my nebulised antibiotics. I was also shown how to use the electronic daily diary. This asks the same questions as the booklets we're all used to filling in for ADAPT. You fill it in each morning for a fortnight as part of the training; it then transmits your answers to ADAPT daily.

After 2 weeks of using the diary in training I'm off to Birmingham again this Friday (7th June). If they are happy with everything I'll have another medical examination, an ECG and some more blood taken. I'll then be given the nebuliser and the first dose of the trial drug and given a further two weeks supply to take home. It will be a 50/50 chance of being the real drug or saline solution, as the trial is a double blind trial. After this the return visits to Birmingham get less frequent. ADAPT monitor you via the daily electronic diary entries and fortnightly 'phone calls.

Why have I agreed to take part? Whilst I realise the trial may come to nothing my answer is, if no one agrees to take part in drug trials then no future cures will be found and no progress is made in better treatments of diseases. This is also the reason I always agree to medical students practicing on me while I'm in hospital, they need to learn from real patients not just text books.

There is also a small trial taking place for Mannitol/Bronchitol via an inhaler. This is a natural sugar obtained from fruit & vegetables and is currently used to treat Cystic Fibrosis patients. It is being trialed on a few Alphas who have Bronchiectasis to see if it can help clear the "muck & gung" which can build up in the lungs.

I hope all this has help demystify the drug trials and encourages people to take part if asked by ADAPT.

I wish you all a fit and healthy summer.

John Morton

Some members report having difficulty persuading their GP to prescribe steroid tablets or antibiotics to keep at home.

Below is an extract from ***Chronic obstructive pulmonary disease - Understanding NICE guidance – information for people with chronic obstructive pulmonary disease, their families and carers, and the public***, published in February 2004. The clinical guidance clearly states that people with COPD, and that includes most lung-affected Alphas, should be given certain medication to be kept at home.

Treatment for exacerbations

Some people with COPD find that they have attacks when their symptoms become particularly bad. These are called ‘exacerbations’. The person may become much more breathless, may start to have a cough, or may notice that more sputum is being produced or that the sputum that’s coughed up is a different colour than normal. Your regular treatment may need to be adjusted and you may need additional treatment to help you recover from an exacerbation.

Helping yourself

If you and your doctor or nurse think that you are likely to have an exacerbation, you should be given advice about what to do at the first sign that one might be on its way. If you have a carer, he or she should also be given this advice. It should include advice about:

- starting steroid tablets if you think you are developing an exacerbation, that is if you notice you are coughing more or becoming more breathless and it’s interfering with your normal activities (as long as it’s OK for you to take steroid tablets, you should be given some to keep at home)
- starting antibiotics if you notice that your sputum has turned green or brown (again, you should be given some of these to keep at home)
- making changes to your bronchodilator medicine to help with your symptoms.

You should also be advised to contact your doctor or nurse if things don’t improve.

NEWS IN BRIEF

Donation

We were very, very pleased in March to receive, from our friends at the Alpha 1 Foundation in the USA, the promised matching donation, enabling us to give Dr Hadzic, from Kings College Hospital London, a further £1000 for his research into the effects of Alpha 1 on the liver of children.

It was a great pleasure to be able to send this second donation to Dr Hadzic and our thanks go to John Walsh, CEO and President of the Alpha 1 Foundation and his colleagues for making it possible.

Lin Daniels

Welsh Alpha Lunch

On Saturday 17th April, a buffet lunch was held at the Cophthorne Hotel in Cardiff. It was an opportunity for people in South Wales to meet and discuss issues associated with alpha 1 antitrypsin deficiency. Some of us had met previously, but we were able to meet two new members for the first time.

Interestingly two people had concerns about liver function, and it was good to exchange views on those problems. We also had very positive feedback from someone who had recently completed a pulmonary rehab course. It would have been good to have had greater numbers attending, but I felt that everyone had an opportunity to voice their concerns and a chance to speak personally to the Secretary, Lin Daniels with her wealth of experience caring for someone with the condition.

Meryl Darkins

Membership

It is now almost 12 months since I took on the task of recording membership. There has been quite a surge in membership every time that we have a national event like an information day. Therefore, I look forward to being very busy in the wake of the European Congress in July! However, in between events there is a steady trickle of new members

every week, and I am pleased to say that membership has trebled since I joined the charity in March 2009. For every member, there are family members who must have the genotype PiMZ at least. It is our hope that by giving information we are helping people to access the diagnosis and treatment they need to limit the effects of the condition.

If you know of someone who has A1AD, get them to join. If you would like to help spread the word, please email me to get flyers or posters to display at your chest clinic or your liver clinic.

Meryl Darkins (Membership Secretary)
meryl@alpha1awareness.org.uk

Forum

The online forum continues to go from strength to strength, with new Alphas, carriers and carers joining us every few days.

Remember, you'll find us at

<http://techno.demon.co.uk/a1aforum> for the time being (until we move to a site with a more 'catchy' web-address!) Do come along and join us - if you're sitting on the fence, wondering what these internet forums are all about, just remember you'll be safe with us! And you're more than welcome to join up and 'lurk' for as long as it takes until you feel comfortable to start chatting.

See you there! That link again:

<http://techno.demon.co.uk/a1aforum>

Neil Jackson
Forum Administrator

Have you changed your address or email

If you have please remember to let us know so that we can keep in touch with you.

**email to secretary@alpha1awareness.org.uk
or write to Alpha1, PO Box 2866, Eastville,
Bristol BS5 5EE**

Can You Help Your Charity ?

Due to illness, we have lost our Treasurer, this is an important and necessary role. At this moment in time I am doing the role alongside that of Secretary, I am also working full time in a paid job. I am not a figures person and I would like to make an appeal to all of you, I feel sure that somewhere among our members is someone, not necessarily you, but a relative, spouse or friend, who would be able to take over this role. Please think about it, if you would like to offer help, please email me at secretary@alpha1awareness.org.uk or write to me at Alpha 1, PO Box 2866, Eastville, Bristol BS5 5EE
I look forward to hearing from you. Thank you

Lin Daniels
Secretary, Alpha 1 Awareness UK

Adidas Women's 5K Challenge Sunday September 5th 2010 Hyde Park London

Last year we had 2 runners enter the Adidas 5K Challenge run. Do you have a daughter, sister, wife or family friend, who runs for fun and would like to enter the run this year and raise money for Alpha 1 Awareness UK, it costs £15 to enter and £5 of that goes to your chosen Charity (us we hope) then the person running can get sponsors, (we have our own sponsor forms we can send out to you) to raise more money

If you know anyone who would like to enter, you can go online to www.womenschallenge.co.uk and your place will be confirmed immediately, or you can fill out the entry form, I have copies if anyone would like one, just send me an email at secretary@alpha1awareness.org.uk or drop me a line at Alpha1, PO Box 2866, Eastville, Bristol BS5 5EE and I will send a copy to you. Entries must be in by the 6th August 2010, however this will be earlier if the entry limit is reached.

Lin Daniels

We reported in our last Newsletter about Rare Diseases Day in February, the following is a report I received from Buddug Williams, from Rare Diseases UK. A similar event was held in Scotland, if anyone who attended that event would like to send in a report we will happily include it in our next Newsletter.



A group of one hundred families, clinicians, researchers and politicians in Wales came together to mark Rare Disease Day 2010 with a reception at the National Assembly for Wales on Wednesday 24th February. Rare Disease Day is part of a global initiative to raise awareness of rare diseases and improve and coordinate services for people living with rare conditions and their families. The event was organised by Rare Disease UK and hosted by Helen Mary Jones AM for Llanelli.

Several families from Alpha 1 Awareness UK attended the event including Jeff & Meryl Darkins from Blaenau Gwent and also Angela & Ray Miller from Maesteg.

Rare Disease Day 2010 was on Sunday the 28th of February and the theme was 'Patients & Researchers: Partners for Life' and raised awareness of the need for research into rare conditions and development of treatment and support for people living with them.

A rare disease is one that affects about one in 2,000 people. There are more than 6,000 different rare conditions. Rare diseases are surprisingly common – 3.5 million in the UK (175,000 people in Wales) - live

with a rare condition at some point in their lives. Rare Diseases are diverse in severity and symptoms presented by patient. People living with these conditions, however, report similar issues around diagnosis and treatment. It can take patients years to achieve a diagnosis, leading to a deterioration of the condition, inappropriate treatment and medication and significant distress for them.

Rare Disease UK is campaigning for a strategy in Wales and across the UK for integrated service delivery for rare diseases to aid speedy diagnosis and effective treatment and care for people living with rare conditions.

Alastair Kent, Chair of Rare Disease UK, said: "There are tens of thousands of people in Wales and millions of people in the UK living with a rare condition. For many, there is little knowledge on how to treat and manage their condition and scientific research is lacking. Raising awareness of rare diseases and the need for a co-ordinated approach for the diagnosis, treatment and research of rare diseases is hugely important

Rare Disease Facts:

- 75% of rare diseases affect children and 30% of rare disease patients will die before their fifth birthday
- 1 in 17 people will develop a rare condition at some point in their life
- Rare diseases are often chronic, progressive, degenerative, and often life-threatening
- Rare diseases are disabling: the quality of life of patients is often compromised by the lack or loss of autonomy
- Rare diseases are characterised by a broad diversity of disorders and symptoms that vary not only from disease to disease but also from patient to patient suffering from the same disease. Relatively common symptoms can hide underlying rare diseases, leading to misdiagnosis.

- Rare disease patients face common problems:
 - ▶ Lack of access to correct diagnosis
 - ▶ Delay in diagnosis
 - ▶ Lack of quality information on the disease
 - ▶ Lack of scientific knowledge of the disease
 - ▶ Heavy social consequences for patients
 - ▶ Lack of appropriate quality healthcare
 - ▶ Inequities and difficulties in access to treatment and care

Alpha 1 Awareness UK

Raising Awareness of Alpha 1 Antitrypsin Deficiency in the UK

Contact us at : Alpha 1
PO Box 2866
Eastville
Bristol BS5 5EE

email : secretary@alpha1awareness.org.uk
visit our website : www.alpha1awareness.org.uk

5th Alfa Europe Federation Congress
Friday 9th- Saturday 10th July 2010

As you are all aware, from reading our last Newsletter, Alpha 1 Awareness UK are very pleased to be hosting the 5th Alfa Europe Federation Congress, organised by the Alpha Europe Foundation on the 9th and 10th July in London.

You as a member of Alpha 1 Awareness UK are invited to attend the Congress, along with your spouse, partner, carer or family members.

The Congress is being held at **Hilton London Kensington Hotel, 179 -199 Holland Park Ave, London W11 4UL. Tel: 0207 6033355**

Your hotel accommodation, dinner Friday evening, breakfast and lunch on Saturday will be paid for by the European Foundation.

If your return journey home on the Saturday evening after the Congress is more than 200 miles or 150 miles if you are on O2 or have children under the age of 6 years of age travelling with you, you will be entitled to stay over on the Saturday night, with Bed & Breakfast, at the hotel, again paid for by the European Foundation.

You will be able to claim a contribution towards your travelling expenses, we will send out more details with your booking confirmation.

Child Care, provided by a registered Child Care Agency, will be available all day on the Saturday, during the presentations, for any children attending with you.

A booking form and a brief resume of the programme is attached to this Newsletter. Please complete your booking form and return it as soon as possible, **all forms must be received by the 31st May**. If you wish to email your booking form to us, please go onto our website www.alpha1awareness.org.uk and following the links to the Congress, where you will find a booking form and other information. If you require any further information please email congress@alpha1awareness.org.uk or drop a line to Alpha 1, PO Box 2866, Eastville, Bristol BS5 5NN.

We at Alpha 1 Awareness are excited and honoured to be asked to host this Congress, we feel it is a wonderful opportunity for Alphas in the UK to hear some of the leading experts in the field of Alpha 1 and learn more about the condition and developments for the future. We hope you will want to share in this occasion and will be able to join us on the 9th and 10th July, we are certainly looking forward to meeting many of you for the first time and finally putting faces to names

The Hotel is wheelchair accessible. O2 will be available

5th Alfa Europe Federation Congress
Friday 9th- Saturday 10th July 2010

Programme:

Friday 9th July 2010

19:30

Dinner for all Alpha1 Awareness members, European delegates from Alfa Europe Federation and guest speakers

This is the opportunity to meet Alphas, their carers, family and friends.

Saturday 10th July 2010

09:30 – 09:40

Conference Opening & Introduction

09:40 – 10:00

Professor Sabina Janciauskiene

MHH Hannover, Germany

What is Alpha1 Antitrypsin Deficiency?

10:10 – 10:30

Doctor Nedim Hadzic

King's College Hospital, London

Liver Diseases of Children

10:40 – 11:00

Coffee Break

11:00 – 11:20

Professor David Lomas

University of Cambridge

Developing a Cure for Antitrypsin

11:30 – 12:00

Doctor Heinz Steveling

Ruhrlanklinik Essen, Germany

20 Years of Replacement Therapy

12:00 – 14:00

Lunch

14:00 – 14:20

Melissa Hillier

Genetic Alliance UK

Improving access to healthcare for families with rare genetic conditions

14:30 – 14:50

John W Walsh

Alpha-1 Foundation, USA

Patient Empowerment

14:50 – 15:00

A UK Alpha patient

A view of the position of Alphas in the UK

15:00 – 15:30

Tea/Coffee Break

An opportunity to say farewells to new found Alpha friends and ask any final questions

We look forward to meeting you and hope that you will enjoy and find the Congress useful and informative.

A booking form for the Congress is found on the next page

Alpha 1 Awareness UK

Registered Charity Number: 1125467

Scottish Charity Register Number: SC041100

Alfa Europe Congress, London, 9th & 10th July 2010 Booking

Form

Hilton London Kensington, 179 -199 Holland Park Ave,

London W11 4UL. Tel: 0207 6033355

NAME

ADDRESS.....

.....

.....POSTCODE.....

TEL.....

EMAIL.....

NAMES OF THOSE ATTENDING

.....

NUMBER OF NIGHTS

SPECIAL REQUIREMENTS Diet.....

If you require O2 name of supplier.....

liquid and/ or concentrator.....

Disabled room.....

Do you require Child care during Saturday 10th July ? Trained staff from an agency will be on hand to take care of your children during the presentations.

Number of Children..... will you require a cot in your room YES/NO

Name.....Age.....

Name.....Age.....

Name.....Age.....

Return to Alpha 1
 PO Box 2866
 Eastville
 BRISTOL BS5 5EE

If you wish email your booking go to www.alpha1awareness.org.uk and follow the link to Congress.

Please return this booking form by 31st May 2010 at the latest

Chairman: Alan Heywood-Jones, Secretary: Linda Daniels,
www.alpha1awareness.org.uk : email: info@alpha1awareness.org.uk

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Scottish Charity Register No. SC041100