

Newsletter

Alpha1 Awareness

Vol 1
Issue 4
October 2009

A Word from the Chairman

The second Information Day of 2009 was held at the Concorde Club Conference Centre in Eastleigh near Southampton on the 5th of September. This was the second regional InfoDay of 2009 arranged by Alpha1 Awareness.

The format was the same as the Bristol InfoDay, that is to say, two expert medical speakers talking about the condition and two speakers from other fields of interest to most Alphas; respiratory rehabilitation and genetics.

Doctor Thomas Köhnlein from the Medical School in Hanover gave the introductory presentation on the causes and development of genetic emphysema and liver failure. Later in the day Doctor Bibek Gooptu, who works for the Wellcome Trust at Birkbeck College gave insights into the work being done to understand and treat the underlying mechanisms of Alpha1 Antitrypsin Deficiency. Alpha1 Antitrypsin Deficiency is a complicated condition and it produces a number of medical problems. Researchers and clinicians are still looking into new aspects of the causes and progress of AATD and it is a great tribute to the two doctors that they explained their work in a clear and understandable way.

Jack Cherry spoke about the rehabilitation services in the Portsmouth / Southampton area and the clear benefits that these give to Alphas with breathing difficulties. Jack is a young athletic man and when given a running shirt with the name of the charity on it he promised to run for us in a marathon.

Steve Nutt came from the Genetic Interest Group and told us about their work. As we have said before, AATD is a genetic condition and the work of GIG is important in promoting the interests of all similar conditions which often are rare and do not attract public attention and political support.

The event can claim to have the youngest Alpha attendee of any of our gatherings. Maya, who came along with her parents, was only four weeks old. She had been diagnosed as PiZZ only one day before the InfoDay.

Following a break for tea my wife, Dawn, spoke about augmentation therapy, its obvious benefits and our hope that it will become available in the UK.. Then Neil Jackson explained how the internet forum works and showed us how to register and start contributing on the various topics.

A number of AlphaKits kits were given out to those people who were either uncertain of their own genotype or were concerned about their close family relatives.

Overall the day was a great success but the numbers were slightly reduced because many people have fears of contracting Swine Flu. In total, 38 Alphas, carriers and carers came to the event.

I must thank Lin Daniels and Dawn for organising the Southampton Info Day.

My thanks also go to Meryl Darkins for taking on the role of Membership Secretary, giving Lin, who was doing this job alongside that of Secretary, one less job to do. Meryl has had a busy time of late as our membership is increasing rapidly. Thanks to Neil Jackson who has set up and helps administer our Forum on the internet, this is a useful tool for news items and chatting to members, who apply to join the Forum, Neil is also a big asset with his technical help and "surfing" the internet for the latest news items. More on the Forum later in our Newsletter.

The start of October saw the first Alpha Lunch. See Lin's report on the regional meeting for the Cornish Cluster in Bodmin.

Foreign Adventures

On the same day as the event in Southampton the Swedish Alpha association was holding an Information Day in Stockholm. The treatment of AATD emphysema is authorised but there are problems in paying for expensive augmentation therapy. In part this is because the incidence of AATD in Sweden is higher than in the UK – the condition is sometimes called "The Viking Disease". I sent my good wishes to the chairlady of the Swedish association and apologised for not going there since the dates clashed with the UK InfoDay.

On the world stage there are a number of changes which, in time, may have important consequences for UK Alphas. There are four main pharmaceutical companies which extract alpha1 antitrypsin (AAT) from human blood plasma, purify it and test it as being suitable to replace the missing AAT in PiZZ Alphas. The names of these companies are Baxter, CSL Behring, Talecris and Kamada. Earlier this year CSL Behring attempted to take over Talecris but this was blocked by the USA regulatory authorities as being anti-competitive. More recently Talecris has become a quoted company on the New York Stock Exchange. Meanwhile, Kamada is promoting its new inhaled AAT offering. European colleagues speculate on what will be the outcome of all this activity. The optimistic view is that increased competition will reduce the cost of their augmentation therapy. If this is so then the struggle to have augmentation therapy passed by NICE will be easier. But before that we have the stumbling block that such no augmentation therapy is authorised in the UK*. Our battle starts with the MHRA - Medicines and Healthcare products Regulatory Agency.

Alan Heywood-Jones

* AAT infusions are sometimes given on a Named Patient Basis.

INFLUENZA

What is it?

Influenza, commonly referred to as the flu, is an infectious disease that affects birds, humans and other mammals. The name influenza comes from an Italian word referring to the *influence* of bad or cold air which was at one time thought to be the cause of the disease.

Seasonal flu is the 'normal' flu that occurs every winter. In the United Kingdom the peak time for transmission of flu is November through to March. In the Southern Hemisphere the peak is six months later. In the Tropics flu is present all the year round.

Biologists categorise flu into three types: A, B, and C. Type A influenza causes epidemics or on a world-wide scale pandemics, while influenza viral types B and C are less important in causing human disease, mainly infecting animals. Unfortunately, influenza A strains are the most varied of the three types.

Symptoms

The most common symptoms of the disease are chills, fever, sore throat, muscle pains, severe headache, coughing, weakness and general discomfort. Fever and coughs are the most frequent symptoms. Although it is often confused with other influenza-like illnesses, especially the common cold, influenza is a much more severe disease than the common cold and is caused by a different type of virus.

The virus spreads from person to person in tiny droplets coughed or sneezed into the air by an infected person. These droplets can remain airborne for up to two hours and are easily inhaled by another person. Since the virus can also last for a short time on objects, you can become infected by touching something contaminated with the virus and then touching your mouth, nose or eyes.

Symptoms are usually at their worst after two or three days. Recovery typically takes five to eight days, though some people take three weeks or even longer to feel completely well again.

The time it takes from first contact with the virus until symptoms appear is between 1 and 4 days – typically 2 days. It is usually possible to pass on the infection for as long as 24 hours before the first symptoms appear and this is one reason why the infection spreads so easily.

People who contract flu should get plenty of rest, eat well and drink lots of liquids. Children should never be given aspirin because of the possible complication of Reye's syndrome.

Jabs

Some people are more likely to become infected and may also be more vulnerable to serious complications. Alphas are clearly an 'at risk' group as are the elderly, people who are diabetic, asthmatic, have kidney problems or reduced immunity. These people are given priority for free flu vaccination under the NHS. Close family members should also be vaccinated against the flu.

Seasonal flu vaccines have a long track record of safe and effective use. The vaccine is normally given as early as possible in the autumn, and protection lasts through the winter and beyond, though because of new strains of flu, new vaccines are generally required each year.

The effectiveness of these vaccines depends on there being a good match between the strains used to produce the vaccine, and the strains that ultimately cause infection. Seasonal flu vaccines cut the risk of infection by at least 70-90 per cent. The World Health Organisation makes global surveys to help predict which strain will arrive each year. The accuracy of these predictions is constantly increasing.

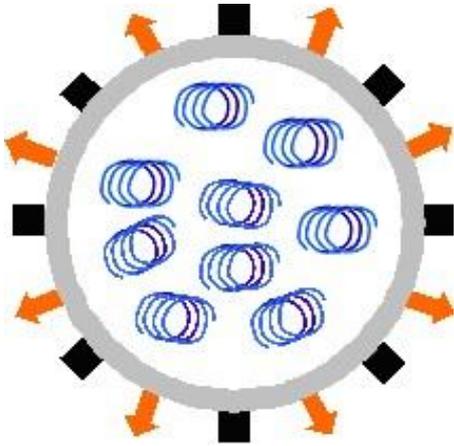
Published research data show that seasonal flu vaccination:

- Cuts the risk of confirmed flu infection by at least 70-90%
- Cuts total numbers of upper respiratory infections by at least 25-34%
- Cuts work loss from upper respiratory infections by 32-43%
- Cuts visits to the doctor by 42-44%

A possible complication is pneumonia which in effect is a bacterial infection taking advantage of a weakened host. A vaccine against *Streptococcus pneumoniae* is available for adults and is currently recommended for all healthy individuals over 65 and any adult with emphysema, congestive heart failure, diabetes mellitus, cirrhosis of the liver or who do not have a spleen. A repeat vaccination is recommended after five or ten years.

The Flu Virus

Viruses are little more than short sequences of genetic material in a small protective shell – they are not normal cells which are very much larger. A virus makes new copies of itself by invading a cell and tricking the cell into making new viruses. The new viruses break out of the cell and invade other cells.



The flu virus is roughly spherical. On the outside there are a copies of a protein called hemagglutinin – HA for short. These can attach to the type of cell in the nose, throat and lungs of humans, other mammals and in the intestines of birds. HA is key to breaking into the victim cell.

After the cell has made new copies of the virus the progeny viruses need to break out of the cell. Another set of proteins, also on the outside of the protective shell is used to do this. This protein is called neuraminidase – NA for short.

There are different varieties of HA and NA proteins and each one has been given a number. Because these proteins are on the outside of the virus it is relatively easy to identify the varieties of HA and NA present. This

- HA hemagglutinin
– breaks into cells
- NA neuraminidase
– breaks out of cells

is why influenza virus type A (the one that can affect humans) is named H1N1 and so on.

Subtypes H1N1, H2N2 and H3N2 are common in man while pigs have these as well as H5N1 and H4N6. H5N1 is not as infectious as other subtypes but it does attack more than just the upper respiratory tract and is therefore more dangerous. Most documented cases are in humans who live in close proximity with pigs and domestic fowl. Very few cases have been reported for professional workers with these animals.

The genetic material within the virus is capable of recombining with itself and producing new variants. Another possibility is that two different subtypes attack a cell at the same time and then an altogether new strain may be produced.

Influenza Pandemics

There is good evidence for flu pandemics dating from 1580. The last century saw the Spanish Flu pandemic of 1918 (H1N1), the Asian Flu pandemic of 1957 (H2N2) and the Hong Kong Flu pandemic of 1968 (H3N2).

The current pandemic, first identified in April 2009, is subtype H1N1 but this does not mean that it is the same as the pandemic of 1918. The official name is "new H1N1" but it is commonly called "Swine flu". It is thought to be a mutation (reassortment) of four known strains of Influenza type A virus subtype H1N1: one generally present in human populations, one present in birds, and two present in pigs.

News in Brief

Alpha 1 Awareness Forum

In mid-July, by popular demand, we set up our new internet web-forum at <http://techno.demon.co.uk/a1aforum>. The idea of the forum is that it gives all us Alpha1 Awareness charity members and our families an opportunity to ask questions, lend support, and talk generally about all aspects of Alpha1 Antitrypsin Deficiency.

So far, nearly 40 of our bravest members have signed up and are posting messages, and the forum is already becoming a busy place. We have discussions going on about all sorts of Alpha1 queries: everything from people comparing and learning about the various inhalers that they have been given, to idea-sharing about the best kinds of diet that help look after your liver. There are also ongoing discussions about the various UK-based drug trials that are coming up, as well as a look across the pond at some of the startling but exciting gene therapy experiments going on in the USA. Some of these discussions give us hope; others give us insight - and all of them give us plenty of scope for talking together and putting information into our own, personal perspectives. Which is exactly what a support forum like ours is for!

We hope that you will come along and join us too. There's a wealth of information and discussion already - and it is bound to keep growing and improving, the more members that use it! Joining is really simple - just surf along in your browser using the link above, and choose the 'Register' option. Choose a username for yourself (don't worry - if you decide later that you want to be known as something else, it's easy to change what people see you as), choose a password, read and accept the user agreement, and press the Register button. Your application will be whisked over to us automatically, and all being well, should be approved and activated within a day or two, and you can begin posting.

If you prefer to 'lurk' for a while, reading what others are discussing, then that's fine too! But remember, almost all the posts and topics are hidden until you register; unregistered Guests can only see News articles. Everything else - the detailed discussions, chats and conversations are all safely tucked away from general view, so that you have the freedom and privacy

to discuss what really matters to you. There are no 'tourists' on our forum - everyone is either a fellow Alpha sufferer, or a carer or family member connected with an Alpha, or someone in the medical profession dealing with Alphas. So you can speak your mind or ask difficult or even embarrassing questions without worrying whether Google will dig up your words years later, or whether some someone on the forum has different agenda.

We hope to see you there soon, and to hear from you about whatever aspect of the Alpha experience you think needs to be aired! It's your forum, and it's there to serve you - so please, make the most of it, and get chatting! The link again?

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

Neil Jackson

Forum Administrator

Membership

During 2009, membership of Alpha 1 Awareness UK has trebled. Undoubtedly, the website has made a tremendous difference, and recently, most members have joined online. We have more new members joining us every week. However, this still only represents a small proportion of those with the condition. Therefore, we have a great capacity for growth.

Our ever increasing numbers are important, because if we are to lobby for improved research and treatment for the condition, there is strength in numbers. Therefore: if you know of anyone who might have the condition; if you are in contact with your local chest/liver clinic; if you attend a pulmonary rehab course, please tell them about us and encourage others to join us.

We have leaflets which explain our role, and which are suitable to leave in surgeries and clinics. Please contact us if you feel you can help distribute information.

Finally, I hope that in the next newsletter I will be able to report a doubling of our membership.

Meryl Darkins

Membership Secretary

Fund Raising

Our fundraising has received several big boosts this month, you may recall that Gary Stone ran in the London Marathon, raising money in memory of his Dad, Ian. Ian Stone owned and ran a Social Club at a holiday camp in North Wales, the club supports a Charity each year throughout the summer months and this year they chose to support Alpha 1 Awareness UK. At the beginning of October Dawn & Alan, attended an event as representatives of the Charity at Stones Social Club, where they were presented with a cheque for £2720.00, it was a huge surprise to receive such a huge sum and we are extremely grateful to everyone at Stones, the family, staff and customers for their help in raising it. We will be deciding what the money will be used for over the coming weeks.

A substantial sum was also received in memory of a gentleman, Mr Peter Parsons, from Kent, who sadly died in August from Alpha related liver problems. The family chose AAW to receive donations in his memory, for that we are extremely grateful.

Following on from the news in our last Newsletter about our runners in the London and Paris Marathons, I am pleased to say that we have had further runners raising money for us, Antonia Limbuwala, whose mother suffers from Alpha 1, ran in the Guernsey Marathon at the end of August and has so far raised approx £570. Sheila Anderson, whose sister, Val Harris is an Alpha, ran in the York 10K, raising £300, we also had 2 young ladies running in the Adidas 10K run, but we do not know yet what they have raised.

We are also delighted that Jack Cherry, a Senior Physiotherapist on the Portsmouth City PCT Pulmonary Rehab Team, who kindly came to give a presentation at our Information Day in September, will be raising money for Alpha 1 Awareness UK and The National Autistic Society, when he runs in the Great South Run and Gosport Half-marathon in November, we wish Jack luck with his runs, anyone who wishes to sponsor him just go to www.justgiving.com/jackcherryalpha1.

All this is very good news for the Charity and we send our heartfelt thanks to all who have donated or raised such large sums of money for the work of Alpha 1 Awareness UK.

Christmas Cards

Can I remind you that we do have our Christmas cards on sale, I have attached an order form for them to this Newsletter, they are the same style as last year, but we have some new designs this year.

Don't forget if you are shopping online...postal problems allowing

Shop Online and Raise Funds

Don't forget when shopping online to register with easyfundraising.org.uk and it won't cost you a penny more to shop and raise funds in this way. In fact you could even SAVE MONEY as many retailers give exclusive discounts, special offers and even 'e-vouchers' when you shop through the easyfundraising site.

Easyfundraising is a shopping directory featuring over 600 trusted online stores, including:

Asda, Tesco, Argos, Amazon, the Body Shop, NEXT, Debenhams, John Lewis, Toys'R'Us, HMV, Virgin, iTunes, CD WOW, Marks and Spencer, Currys, Dixons, Staples, PLAY.COM, Pets at Home, Choices Direct, WH Smith, The AA, RAC, Direct Line, Churchill, The Carphone Warehouse, Ticketmaster and over 600 others...

Register for **Alpha 1 Awareness** and just by doing your shopping online you will be helping raise funds for the Charity.

Please share with us any plans you have for raising funds, be it a special birthday or anniversary, a coffee morning with your friends, a Quiz Night, getting the children to do a sponsored silence, we would love to hear. Thank you

Other News

Cornish Lunch

We have mentioned that we are getting a large number of people contacting us and I noticed that we had quite a little cluster of Alphas in Cornwall, who found that travelling to our two Info Days this year would be too far. Dawn and I decided that although it would be too costly to run another Info Day in Cornwall, we could host a lunch, this was organised for Saturday 9th October in Bodmin.

Initially we were disappointed that 3 people failed to attend, but then a young couple turned up on speck having seen about the lunch on the website, they were young parents whose, now 18 month old son, was diagnosed at birth with Alpha 1, he was on medication at first but like a lot of children he has been taken off these and is just being monitored by the liver specialist at Birmingham Children's Hospital, who they told me is very good and knows a lot about Alpha 1, as does their neonatal specialist at Treliske Hospital in Truro, which in itself was good to hear.

Everyone who attended were all very appreciative of the day, of having the chance to talk about Alpha and how it affects them and hear from others that the way they feel sometimes is just how others feel, always a great reassurance, everyone one there was very positive and said they had gained a great deal from attending the day. It was really good to meet with everyone who attended and put faces to names, thank you to all of you who came to the day, it made it a worthwhile trip for Dawn and myself

Both Dawn and I arrived back in Bristol very tired, but also felt quite buoyant that it had been the right thing to do, to hold this lunch and both agree it will be good to do it again in another part of the country, so maybe next year we will be able to hold one or two Alpha Lunches, near you.

Lin Daniel, Secretary

Info Day Southampton



AAW Info Day 5th er 2009 in Southampton (see the Chairman's letter for a report)

Information Booklets

As you will be aware we have produced two Information Booklets this year, "*Newly Diagnosed*" and "*Your Childs Liver*", these have been very popular, but I am pleased to say we now have a new booklet available, it is called "*Living with Alpha 1*".

If you would like copies of our new booklets or any of our other titles, please do not hesitate to get in touch with me by email or letter and I will be happy to send you some. They are useful to have when you are going to hospital appointments to pass on to medical staff.

Our fourth and final booklet will be aimed at medical staff and we hope to have this available in the New Year.

Test Kits

Thanks to the generous sponsorship of Talecris Biotherapeutics, we are able to offer AlphaKits® to people who have family members who need to be tested for Alpha 1. There is no charge to the patient or the GP for these Test Kits, they have full instructions for the GP as to how to carry out the test and we even put a stamp on the envelope to send the kit to the testing laboratory. If anyone one would like a Test Kit for their family members, please either email to secretary@alpha1awareness.org.uk or drop a line to the PO Box address at the end of this Newsletter.

AlphaKits® is a registered mark of Talecris Biotherapeutics GmbH

We would love to have items from members in our Newsletters, if you have any Alpha related topics you feel you could write a small piece about, we would love to see it and consider using it in a future issue. Also if there is a topic you would like us to cover please email or write to the addresses at the bottom of this Newsletter.

Remember this Newsletter is your newsletter and we want to hear your views and opinions and possibly have some of your contributions.

Thank you all for your support during 2009. We look forward to continuing to grow and help you more in 2010

We would like to send all Members of Alpha 1 Awareness UK
And their Families
Very best wishes for a Very Happy Christmas
And
A Healthy and Happy 2010
from
Everyone on the AAW Board



Alpha 1 Awareness UK

Raising Awareness of Alpha 1 Antitrypsin Deficiency in the UK

Contact us at : Alpha 1
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email : secretary@alpha1awareness.org.uk

visit our website : www.alpha1awareness.org.uk

Alpha 1 Awareness UK

Registered Charity Number: 1125467

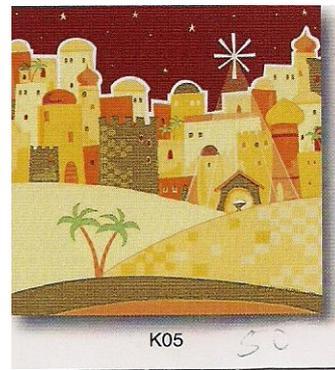
CHRISTMAS CARD DESIGNS 2009



ADORATION



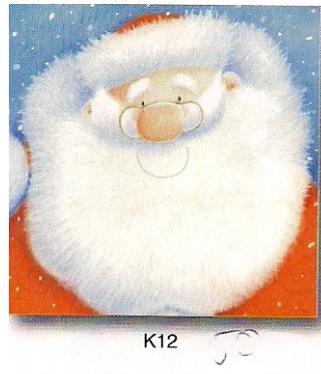
CHRISTMAS



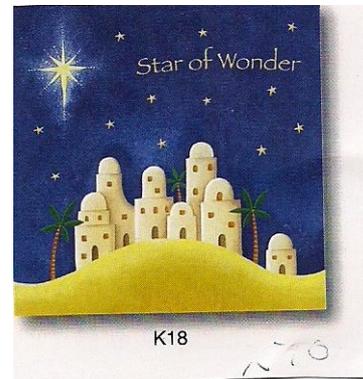
HOLY CITY



STARRY NIGHT



SANTA



STAR OF WONDER

