Alpha-1 UK Support Group Newsletter

Issue 16
Autumn 2017

Welcome....

...to our Autumn 2017 Newsletter

A massive thank you to all our members for their loyalty and support over the years and a very special welcome to all our new members - we hope you enjoy being part of our group!

We would like to thank everyone who has contributed to this issue of our annual Newsletter. We hope you find this Newsletter informative and are inspired by the members stories' and the examples of fundraising for our group. This Newsletter also pays tribute to our dear friends who passed away in the last year, and who we remember with deep affection and respect.

In This Issue

20 years Alpha-1 UK Support Group - some key facts about the charity

by the Trustees

This year is our group's 20th anniversary! This is a cause for reflection, celebration and gratitude.

We have been on a long, at times challenging, but always interesting and deeply rewarding journey since the Alpha-1 UK Support Group was first established.

The group, initially called "Alpha-1 UK", was started in 1997 by five founding members as a discussion and self-help platform, true to the motto "from patients for patients". Our remit and objectives have since evolved, and the article on the next page provides a brief summary of how the charity works towards its objectives.

The group renamed itself "Alpha-1 UK Support Group" in 2008 and registered as a charity two years later. Our membership has steadily grown since, and today we have almost 600 registered members and many more supporters.

Thanks to the founding members' enthusiasm for digital technology, in 1997 we were one of the very first patient groups to launch our own website. The group also embraced social media platforms - we have been on Facebook since 2008 and started a Twitter account in 2010. Today, our closed Facebook group has over 2,700 members from around the world and is one of the main communication platforms for patients, families and supporters.

We are lucky to always have had keen fundraisers amongst our members and supporters - on average, more than 30 fundraising events are being held for our charity's benefit every year. Additional income is generated by generous donations from members and supporters and, occasionally, grants from pharmaceutical companies. Another valuable gift we receive is the time that members and supporters commit to the charity, without which we would not be able to operate.

THANK YOU for your continuing support!

Two decades of patient support - onwards and upwards!

by Dr Sandra Nestler-Parr, Trustee

Since the Alpha-1 UK Support Group started 20 years ago, it has provided a much-needed platform for patients with alpha-1 antitrypsin deficiency (Alpha-1), their families and friends, for communication, education about the condition, advice on how to gain access to relevant clinical expertise, and emotional and practical support.

All new group members are sent a Welcome Pack containing information leaflets, the latest Newsletter and other materials, such as posters and branded merchandise, to help them raise awareness or fundraise.

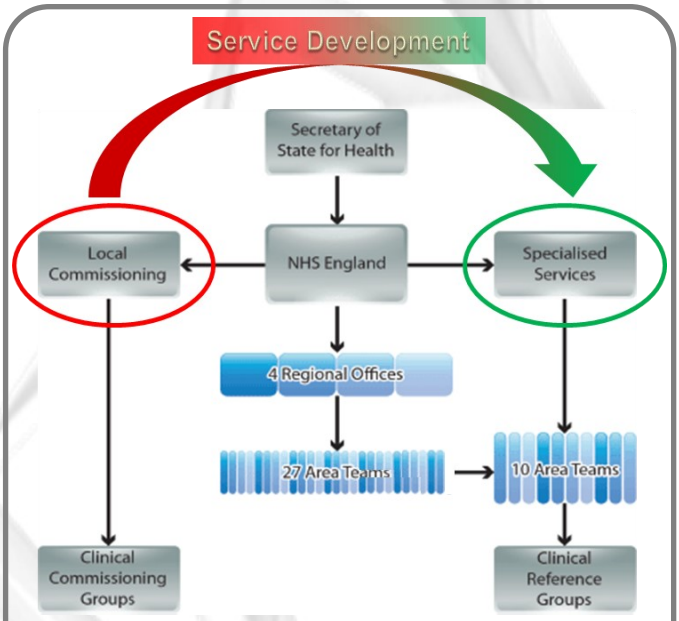
Every year, we hold a national meeting and regional meetings where our members, their families and friends can share their experiences of living with the condition, and provide advice and support for each other. These meetings are usually also attended by external guests and speakers, including expert Alpha-1 clinicians or representatives from pharmaceutical companies working in Alpha-1 who keep us updated on the latest developments in research, disease management and new treatments.

In addition, the charity owns a range of equipment that is available to members on a long-term lease basis to assist them in maintaining their independence for longer and improving their quality of life. This includes nebulisers, portable oxygen concentrators, portable air-conditioning units, mobility scooters and an electric wheelchair.

All of these activities are at the core of why the Alpha-1 UK Support Group was established. However, since its inception 20 years ago, the focus of the charity's work has significantly broadened to keep pace with the challenges faced by all rare disease patient communities, particularly in view of ever-increasing pressures on limited healthcare budgets and, with the rise of treatment for rare disease, the growing competition for healthcare resources .

Over the last ten years our group has had to evolve, building a stronger profile and voice in the UK and within the global Alpha-1 community. In addition to providing support for our members, we have adopted a patient advocacy role and we actively engage with clinicians, pharmaceutical and biotech companies with an interest in Alpha-1, NHS bodies and health technology appraisal agencies (such as NICE), to improve access to care and effective treatments for AATD.

It is generally accepted that, in order to optimise the prognosis of Alpha-1 patients, they require highly specialised management by a multidisciplinary team of clinicians trained in the multi-organ aspects of Alpha-1.



Simplified schematic of NHS service commissioning in England. *All currently operating specialist NHS Alpha-1 services/clinics are funded locally by hospitals or CCGs (red circle). Once the Specialised Service for Alpha-1, recently approved by NHS England, is operational in a few formally selected hospitals across the country, the service will be funded nationally by NHS England (green circle). During the Service Development process (red-green arrow), NHS England works closely with expert clinicians, patients, commissioners and other stakeholders to agree the "Service Specifications". These determine which Alpha-1 patients are eligible for the service, what tests, treatments and procedures the Service provides, the level of expertise required by the healthcare professionals providing the Service, how patients get referred to the Service, which elements of care the Service shares with the patients' local specialists and GPs, etc. The Service Specifications ensure that all centres that will be chosen to provide the Specialised Service work to the same expertise and care to the same, agreed standards. Once the Service Specifications and the number of centres necessary to ensure equality of access to the Service for all patients across England have been agreed, centres can apply to become providers of the Service and will be chosen based on patient need and centres' ability to meet the Service Specifications.*

Two decades of patient support - onwards and upwards! *(continued)*

However, the national patient survey we conducted a few years ago clearly highlighted the lack of access to the necessary clinical expertise and coordinated health services. About three-quarters of respondents stated that they were given little or no information about AATD, and over half rated their healthcare professional's knowledge of the disease as "poor". (The results of the patient survey in England and Scotland are available to download on our website as policy reports at www.alpha1.org.uk/index.php/alpha-1-patient-survey.)

Our efforts have started to pay off - after launching our national patient survey and policy recommendations in the English and Scottish Parliaments in 2014, after three years of campaigning at national level, after several expert centres in England setting up locally funded NHS Alpha-1 services (please see our 2016 Newsletter for details at www.alpha1.org.uk/index.php/newsletters), and after we submitted two applications for the establishment of a nationally commissioned (i.e. funded), Specialised NHS Service for Alpha-1, we are delighted to report that, earlier this year, the Prescribed Specialised Services Advisory Group has *finally* recommended that such a service be commissioned by NHS England (see www.gov.uk/government/uploads/system/uploads/attachment_data/file/598394/PSSAG_2017_report.pdf).

Nationally commissioned Specialised and Highly Specialised NHS Services support people with a range of rare and complex conditions that require coordinated multidisciplinary care by clinicians with hands-on expertise in managing the relevant condition, which is usually not available at community and local hospital level.

Although the biggest hurdle has been taken by receiving agreement in principle to create a Specialised NHS Service for Alpha-1, we are not yet on the home straight. It could be a long and rocky journey until the Service is conceptualised in detail, developed, established and up and running.

The schematic on page 2 illustrates and explains the basic process of how care arrangements and funding for Alpha-1 patients need to be transitioned from currently locally commissioned care to nationally commissioned specialised care that will eventually be provided under a Specialised Service agreement by centres, formally selected based on objective and transparent criteria. The development of these criteria, or Service Specifications, is a complex, consultative, transparent process, led by a committee of NHS officials, expert clinicians, patient group representatives and other stakeholders. We have very recently started to engage with NHS England, so you can rest assured that the charity will represent the patient community's interest with a strong voice.

We will keep you posted on our progress with these discussions!

A brief update on novel treatment approaches for Alpha-1

by Dr Sandra Nestler-Parr, Trustee

The last year has again seen exciting advancements in the development of novel therapeutic approaches for Alpha-1 with the potential to address its underlying cause, representing a real step-change from AAT augmentation therapy. The latest research news was presented at the 3rd International Research Conference on Alpha-1, held in April in Lisbon, and included reports on the advances of innovative technology platforms, such as stem cell and gene therapies and their potential application in Alpha-1. Another growing area of interest discussed at the meeting is the utilisation of the beneficial effects of augmentation therapy in other target diseases, such as diabetes or transplant organ rejection.

A summary of the Alpha-1 Research Conference can also be found on page 9, and some of the presentations are available as videos at the Alpha-1 Global website at http://alpha-1global.org/en/alpha-1-global-event-archive/3rd-international-research-conference-on-alpha-1-antitrypsin-and-6th-alpha-1-global-patient-congress/.

Since the detailed update in our 2016 Newsletter on the status of access to AAT augmentation therapy in the UK, there is, unfortunately, little news to report. Respreeza® received negative reimbursement recommendations by the Scottish and Welsh health technology assessment agencies and is therefore not currently available on the NHS in these nations. In England, a decision by NICE on which process to pursue for the assessment of Respreeza® is still pending. The decision of Ireland's Health Service Executive not to reimburse Respreeza® has resulted in a considerable level of public protest and

A brief update on novel treatment approaches for Alpha-1 *(continued)*

media attention. Both the healthcare decision makers in Ireland and Respreeza®'s manufacturer, CSL Behring, have come under attack by the Irish Alpha-1 community for their failure to negotiate a compromise - decision makers are not convinced that the drug offers value for money, and the manufacturer has not offered a price discount that the health care payers are willing to accept.

There have been a few other setbacks in the last year… Kamada's inhaled AAT augmentation therapy product, which was also trialled in the UK in a Phase 3 clinical study, has not obtained regulatory approval by the European Medicines Agency, following the company's recent withdrawal of its marketing authorisation application. However, encouragingly, Kamada announced its plan to commence a new Phase 3 pivotal study of inhaled AAT for the treatment of Alpha-1, thereby demonstrating continuing commitment to bring their inhaled AAT product to market.

Shortly after our 2016 Newsletter report about the promising progress in the clinical development of effective RNAi therapies for Alpha-1 liver disease by U.S. biotech firms Arrowhead Pharmaceuticals and Alnylam Phamaceuticals, both companies announced in short succession the discontinuation of the clinical trial programmes for their respective drug candidates following unexpected toxicology findings.

Despite these initial setbacks in the development of what could be the first specific therapy for Alpha-1 related liver disease, both companies stressed that they remain committed to developing RNAi therapeutics for Alpha-1 liver disease with new drug candidates.

Our research update concludes with the promising notion that there may soon be light at the end of the tunnel: Apic Bio Inc., a U.S. based pre-clinical stage biotech company that develops gene therapies for a range of diseases, announced less than a month ago that it has achieved a pre-clinical proof of concept for its lead gene therapy product for the treatment of Alpha-1. Having demonstrated efficacy *in vitro* and *in vivo*, the company's Alpha-1 drug candidate is currently undergoing further pre-clinical studies in non-human primates. Apic Bio is hopeful that it can test this potentially curative gene therapy in humans in the near future.

By the way - did you know? Research, partly conducted at an archaeological excavation site of Viking latrine pits in Denmark, has resulted in a new theory as to why the gene defect that causes Alpha-1 has been able to survive. According to a 2016 publication by UK researchers in the journal *Nature Scientific Report*s, deviant forms of the Alpha-1 protein are thought to have protected the Vikings from massive worm infestations by binding to an antibody that evolved to protect people vital organs from diseases caused by worms, which presented a survival advantage at the time. So, what once protected the Vikings from one health problem, now puts Alphas at risk from entirely different health conditions.

A reminder on the guide to genetic testing

by Professor David Parr and Dr Beatriz Lara, University Hospital Coventry

When people get diagnosed with alpha-1 antitrypsin deficiency (AATD) they naturally have a lot of questions about the condition and, when they see us in our AATD clinic, it is not uncommon for them to ask questions like "How did I get it?", "What do the results of my genetics test mean?" and "How is my health likely to be affected by this?". This kind of question doesn't come as a surprise because AATD is a complicated condition and it is understandable that patients may need to have information repeated or may want to hear it from more than one doctor, since most doctors have slightly different ways of explaining things. However, hearing this kind of question from patients attending a specialist clinic is occasionally a cause for concern when it becomes clear that the test for AATD may have been performed without their prior knowledge or without an explanation of why the test was being done and what the answer may mean. Of course, these issues are important for any clinical test but they are even more important with testing for AATD, because it is a genetic test.

AATD is an inherited condition, which means that it may be passed on in families through the genes; each person has two copies of the AAT gene, one inherited from their father and the other from their mother. There are approximately 100 variations of the alpha-1 antitrypsin (AAT) gene and it is generally the case that both copies of the gene have to be an abnormal variant in order to

A reminder on the guide to genetic testing *(continued)*

cause a significant risk to health. When both AAT genes are abnormal, there are many potential health effects that can lead to various lung and liver problems (as well as other rarer conditions). On top of this, the influence of the environment and personal habits, such as smoking and drinking, play major roles in determining whether and to what extent an individual person experiences ill-health, if at all.

All of these influences taken together create a complicated and wide range of potential scenarios! It takes time and specialist expertise to be able to provide patients with an understandable and accurate explanation of the commonest scenarios, but the time to do this is *before* the genetic test for AATD is performed; this is called 'pre-genetic test counselling'. Once the test results come back, it is possible to give more precise information about what the test result means, and this explanation may need to include the findings of other tests such as breathing tests and scans; this is called 'post-genetic test counselling'.

The terms 'pre-genetic test counselling' and 'post-genetic test counselling' apply to genetic testing in general, not just to AATD testing. They are just part of the best practice standards that are applied to the use of genetic testing and which should be followed by all doctors, whether they are GPs or hospital doctors, and by companies that provide what is known as 'Direct to Consumer' (DTC) testing.

In the UK, there are several organisations responsible for providing guidance to health care professionals and the public on the use of genetic tests. The guidance on best practice for doctors and advice for users of health services provide a good framework and lists the principles that should be adhered to when undertaking genetic testing. Some of this information is quite complex and the details differ according to whether the genetic testing is being done for someone with symptoms of 'disease' or for someone who is well but is related to an affected patient. However, there are some common themes and principles that should be followed in order to avoid confusion and worry.

In any case, patients should always speak to their GP or their hospital doctor about the possibility of genetic testing if they think they may need it. Best practice includes obtaining a referral from a GP, or specialist doctor if applicable, for genetic testing to be carried out. If the doctor thinks genetic testing may be appropriate, they should usually refer their patient for genetic counselling as well.

It is also important to consider all implications of having a test for a genetic disease: some people may, for example, experience difficulties in finding or renewing insurance cover (for travel, private health or other insurances) after having had a genetic test, irrespective of the outcome.

The use of postal test kits for genetic conditions, where testing kits may be sent through the post to family members of known patients (often called 'pre-symptomatic testing'), should also follow the best practice guidelines for genetic testing. However, the logistics of using postal test kits clearly present an obstacle to being able to adhere to best practice standards, particularly the ability to provide direct face-to-face genetic counselling. Leaving the counselling to a family member with AATD, even if they have a good understanding of how the condition affects them personally, or searching for information about the disease on the internet, is not a substitute for professional genetic counselling. The use of postal test kits for genetic testing is therefore not recommended by the NHS.

We encourage anyone wishing to learn more about genetic testing to read the information available on the websites below, using the links provided. The specialist NHS clinics for AATD patients that are now running in Birmingham, Cambridge, Coventry, and London should also be able to provide advice and guidance by telephone or email. However, since there is no substitute for face-to-face discussion in a clinic appointment with an expert, some patients may prefer to be referred by their GP for an outpatient consultation. If you would like to do this, you simply have to ask your GP to make a referral through 'Choose and Book' to the expert clinic of your choice – often it takes only one appointment to put minds at rest.

Suggested links:

NHS UK Genetic Testing Network: http://ukgtn.nhs.uk/

NHS Choices – Genetic testing and counselling: http://www.nhs.uk/Conditions/Genetics/Pages/genetic-testing-and-counselling.aspx

Genetic Alliance – Benefits and risks of genetic testing: http://www.geneticalliance.org.uk/information/services-and-testing/benefits-and-risks-of-genetic-testing/

15th Annual Social and Information Day, Cheltenham 2016

Cheltenham, 24th September 2016

Our 15th annual gathering was held at the Jury's Inn Hotel in Cheltenham, in the beautiful Cotswolds, on Saturday, 24th September 2016.

The event is one of the largest annual gatherings of the UK Alpha-1 patient community where Alphas and their families can meet and socialise with fellow patients and be updated on news about research, treatment options, care options and other relevant topics by leading experts in the field of Alpha-1 Antitrypsin Deficiency.

This year, at the request of our members, we reduced the number of formal presentations to provide more time for our members to socialise and to have the opportunity of one-to-one conversations with our external guests.

Joanna Shakespeare, Head Respiratory Physiologist, University Hospitals Coventry & Warwickshire, delivered an excellent practical session entitled "Can you improve your inhaler technique?". She demonstrated the many different inhalers that are now available and the varying techniques required to ensure that inhalers are used correctly and patients get the best possible benefit from their inhaler.

Dr Sandra Nestler-Parr, one of our Trustees, gave an update on the latest research news, the status of access to augmentation therapy in the UK and the specialist Alpha-1 NHS clinics. More details on all of these topics can be found in last year's newsletter which can be downloaded on http://alpha1.org.uk/attachments/article/51/A1UK_NEWSLETTER%20ISSUE%2015%20AUTUMN%202016.pdf.pdf.

The panel members of our interactive "Meet the Experts" session were **Professor David Parr** and **Dr Beatriz Lara** from the Alpha-1 specialist centre at University Hospitals Coventry & Warwickshire, **Joanna Shakespeare** along with industry representatives **Eddie Owens,** General Manager at CSL Behring UK ,and **Peter Leone,** Vice President of Arrowhead Pharmaceuticals. Peter had travelled from the U.S. specially to be able to join our meeting! The panel answered many questions from our members in the audience.

L-R: Eddie Owens, Peter Leone, Joanna Shakespeare, Beatriz Lara, David Parr

This was followed by a break-out session where everyone had an opportunity to speak with the industry representatives and health care professionals.

Many members had dinner together and stayed until Sunday morning.

2nd Scottish Gathering, Kirkcaldy 2017

Kirkcaldy, 3rd June 2017

The 2nd Scottish Meeting of our charity was organised by Fay Whittaker and Graham Dalton and was held at Cluny Clays Activities in Kirkcaldy Fife, on Saturday, 3rd June, between 11 and 5 pm.

The event was intended to be a social occasion where Alphas, and their families and friends could meet and chat and socialize. We had the pleasurable company of Mr Andrew Deans, Respiratory Nurse at the Royal Infirmary of Edinburgh, who has been working in the field of Alpha-1 for many years and was happy to answer our questions and give advice.

Anyone who felt energetic enough had the opportunity to take part in any of the many leisure activities available at the venue. After a buffet lunch, we held a quiz for the adults - we were told that it was rather challenging, to say the least!

A treasure hunt was organised for the children. Clues were hidden throughout the adventure playground, and everyone thoroughly enjoyed running around trying to find them. Many of the children also took advantage of the golf driving range during the day.

We received very positive feedback from the meeting - everyone who attended told us that they had a lovely time and found the opportunity to talk with other Alphas, and to exchange experiences and advice, of great benefit.

Graham was our official photographer for the day and, considering that he was fighting a chest infection at the time, he did a marvelous job which was greatly appreciated.

6th Alpha-1 Global Patient Congress
Lisbon, Portugal, 7-8th April 2017

Friday, 7th April

The Congress program started with a summary of the presentations given during the Research Conference, which preceded the event. Research and therapeutic updates were presented by a variety of notable Alpha-1 researchers and physicians both from Europe and the USA. Topics included: Evidence for Efficacy of Intravenous Augmentation Therapy, Alpha-1 – Beyond Emphysema, Inhaled Therapy, Testing for Alpha-1, Liver Testing and Modifier Genes of Liver Damage.

The morning concluded with a panel discussion on streamlining global efforts of Alpha-1 researchers and clinicians within physician education, patient registries and funding for research. Attendees were able to engage in a question and answer session.

The afternoon started with a presentation by our Trustee, Dr Sandra Nestler-Parr, who discussed strategies to overcome barriers to augmentation therapy access (available at www.youtube.com/watch?v=dvceMgPLzJ4), followed by sessions on the development of the EU Policy Recommendations for Alpha-1 Antitrypsin, policy activities undertaken at the European Parliament level and specifically aimed at rare disease issues, and current concerted efforts underway within clinical practice advancement.

Dr Sandra Nestler-Parr
Trustee, Alpha-1 UK Support Group

Sessions on sleep related issues for Alphas and procedures for pre- and post-lung transplant patients were followed by a panel discussion on lung and liver transplantation. Lung and liver transplanted patients, including children, brought these sessions to life by sharing their personal experiences and concerns.

Saturday, 8th April

The topics of the second day of the Patient Congress included exercise, digital healthcare apps and web-based disease management care programs, and what they mean to the future of the patient community.

Two Global Steering Committee members presented proposed steps for countries currently without access to care to effectively work on progress on a national level, and for new country representatives to set concrete goals and to network with partnering organisations to strengthen their positioning and capabilities to achieve their objectives. Afterwards, three Alpha-1 physicians from Central and Eastern Europe shared their experiences on spearheading close patient-physician collaborations in their countries.

Gonny Gutierrez,
Alpha-1 Global Director

An interactive panel session with Global Steering Committee Members closed the morning session by addressing questions and comments from the audience. Alpha-1 Global Director, Gonny Gutierrez, then provided a strategic overview for 2017-2019 and Alpha-1 Foundation President & CEO Henry Moehringer closed the Congress.

3rd International Research Conference on Alpha-1 Antitrypsin Deficiency

Lisbon, Portugal, 5-6th April 2017

The Global Patient Congress was preceded by the International Research Conference, at which leading researchers and physicians discussed novel treatment paradigms for Alpha-1 and other conditions that have become targets for Alpha-1 augmentation therapy. The meeting was attended by 112 participants from 17 countries.

Attendees heard about new findings from research in the liver cell model, studies of potential new therapies using stem cells, the role of white blood cells for gene delivery, and the investigation of liver therapies in the experimental Alpha-1 Z mouse model. Presentations focused not only on new treatments for the lung and liver diseases of Alpha-1, but also on new disease targets for augmentation therapy, including diabetes, organ rejection, autoimmune disorders and viral infections.

Memorial Service

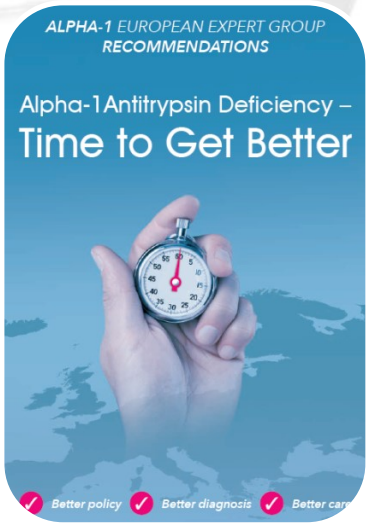
After the Research Conference closed, Alpha-1 patients, scientists, clinicians, industry partners and other members of the Alpha-1 community came together to express their gratitude for the contributions of Alpha-1 Foundation co-founder John W. Walsh, who passed away in March following more than 20 years of outstanding service to the Alpha-1 community.

During the tribute, Walsh was recognised as an inspirational leader whose dedication to finding a cure for Alpha-1 and collaboration on a global level set the stage for Alpha-1 Global. Walsh's colleagues and friends from the international Alpha-1 community shared their personal experiences with him, and used the opportunity to strengthen their resolve that they would honour his legacy by advancing the work he started.

2017 Highlights

Rhode Public Policy Group

In 2012 the first "Alpha-1 EU Expert Recommendations" document was published. The EU Commission and the EU Parliament have gone through many changes since then and it was therefore thought necessary to ensure continuity and current visibility of Alpha-1 Antitrypsin Deficiency within these EU and national institutions.



In collaboration with Rhode Public Policy Group in Brussels, an expert committee of European Alpha-1 physicians, researchers and patients developed updated Policy Recommendations. They were officially launched in the EU Parliament on 22nd March 2017. Translation of the Policy Recommendations into different languages and country specific distribution strategies will commence this autumn.

Global Alpha-1 Investigator and Physician Forum

Dublin, Ireland, 3-4th November 2016

Investigators and physicians from across four continents met in Dublin, Ireland, at the end of last year to discuss ways of advancing Alpha-1 research and the quality of care for Alphas around the world. The purpose of the meeting was to assemble expert investigators and physician educators from different countries to start an ongoing dialogue about important Alpha-1 issues, to identify areas of mutual interest, and to propose feasible programs to improve treatments for Alphas.

AT the end of the meeting, the following next steps were agreed:

- **The Global Guidelines Initiative**, spearheaded by Dr. Robert Sandhaus from the U.S., to compare the rapid accumulation of national and local guidelines on the detection, management, and treatment of Alpha-1 Antitrypsin Deficiency. A summary manuscript will be prepared that presents the similarities and differences between the various guidelines along with an outline of potential methods for reconciling differences and consolidating similarities.
- **A Research Registries Collaboration**, spearheaded by Dr. Charlie Strange from the U.S. The aim is to integrate data collection efforts from Alpha-1 patients globally, to understand data gaps and to promote establishment of new country registries where necessary.
- **A Physician & Researcher eNewsletter**. This bi-monthly publication is now published with specific information to benefit the international Alpha-1 physician and research communities. An archive of past e-Newsletter issues can be found on the Alpha-1 Global website.

Alpha-1 Global 2017-2020 Strategic Plan

In support of Alpha-1 Global's mission statement, our 2017-2020 strategic focus will be in the following areas:

- Facilitate effective stakeholder collaboration
- Raise Alpha-1 awareness globally
- Support the launch and strengthening of Alpha-1 organiations
- Promote early diagnosis and optimal access to care

www.alpha-1global.org

Participants of the Global Alpha-1 Investigator and Physician Forum in Dublin

A debt of gratitude to Sheila Mugford

by the Board of Trustees

The Trustees and Committee would like to take this opportunity to express our sincere thanks and appreciation for the selfless dedication, help and support Sheila Mugford has given to us and members of the Alpha-1 UK Support Group, especially during her late husband John's term as Chair and Vice Chair.

Due to Sheila's continued efforts, new members promptly received their Information Packs; Christmas cards were posted, which often meant queuing in the Post Office at the busiest time of the year, sometimes on a daily basis, parcels were packed with t-shirts, information leaflets and posters, then posted to members around the country to support our fundraisers. Sheila also ensured that literature and merchandise was available at our annual meetings, loading and unloading it and sacrificing their garage as a charity storing facility.

It was only due to Sheila, who became the extension of John in his role as the group's Chair, that John was able to continue to be the vital and active member of the charity that he had always been and to work tirelessly for the benefit of the Alpha-1 community until the day he passed away.

Sheila, the words "thank you" do not seem enough to fully express our gratitude for all you have done in support of John and our members, and for sharing John with us at times when this must have been very hard. We sincerely thank you from the bottom of our hearts.

Member Stories - Living with the enemy

by Kate Bartlett

Some may say living in hell, I say living the dream. You may wonder how a wife/carer of a young man with Alpha-1 can say that!

My husband Mick is 40 years old, he was a healthy kid apart from having ulcerative colitis for five years. He started to notice symptoms in late 2011, work had become much harder for him. Exercise was difficult and his breathing just "wasn't right". A few tests later he was diagnosed with Alpha-1 Antitrypsin Deficiency in January 2012, with the phenotype ZZ and severely obstructed airways.

I remember sitting in the consultants office when he gave us the news. It almost sounded slow motion, I felt dream-like and detached from reality. We both left hand in hand, silent, confused and scared.

A few sleepless nights later everything slipped back to normality. Both of us working full-time, bringing up our two sons, and I guess we swept the diagnosis under the carpet for the next few years.

The monotony and routine of life again took hold and, as many do, we ended up wishing away mid-week so the weekend would hurry up.

On one of our many walks with the dog, in October 2016, I looked at Mick's face as his breathing was so loud and, to my surprise, he was blue. Within a month or so he had a 6-minute walk test done at Coventry Hospital. He failed miserably after just 1 minute, so he was prescribed 4 litres of oxygen for use when he's up-and-about (ambulatory oxygen). Unfortunately, that led to the end of his long career due to the environment he worked in. So, in December 2016, Mick was medically retired.

From that day, life really changed for us as a family, financially things are hard. That said, suddenly life became beautiful. We as a family learnt to love harder than ever before, to appreciate every second we have together and soon realised it's the little things in life that actually are the big things.

It's easy to take life for granted and for materialistic things to dominate our existence, but Mick's Alpha-1 diagnosis has certainly opened our eyes to live life in a very much different way.

The initial embarrassment of the "nose hose" (oxygen tubes) soon wore off and he now struts around like he used to. I, as his wife, don't even notice it anymore, he is

Member Stories - Living with the enemy *(continued)*

as handsome now as when I first met him 16 years ago.

Our boys, Harry 15 and Stanley 13, are MZ carriers, both healthy happy kids. They appreciate family and enjoy life and all it offers.

Mick has more bad days now than ever before, although you will never hear him moan. We still laugh together and enjoy our days even if it is just a cuddle on the sofa as his bad days restrict his mobility.

It's these moments we share together that I almost thank Alpha-1 for, as it's given us the opportunity to retire young and enjoy the time we have. I can't be angry, sad or depressed about it. I am so grateful for this special time we are having as a family.

Mick has recently undergone the three-day assessment for transplantation, as he needs a double lung transplant. We were both so very shocked to hear that he is currently in that window of transplant opportunity, but they are happy to assess him every 8 weeks as long as he stays healthy and does not get poorly with infections.

The assessment was hard, draining and mentally consuming. So much information to take in, there's no sugar coating from the team at Papworth, I must say though that I much prefer that approach. Again, we walked away hand in hand, deflated, exhausted but by no means beaten. So we brush ourselves off, suck it up and again enjoy each and every moment of our lives.

I often find myself stuck in a daze starring at him whilst he's reading his books or having a nap. I smile to myself as I see the strongest, bravest man I have ever met. He wears his oxygen, he struggles to breathe, he can't work, he faces a huge operation, yet he smiles and laughs his way through each day. How can I not smile at that courage he shows?!

I can't tell you what it's like to live with Alpha-1, I can only tell you what it's like to live with an Alpha sufferer. For me, it's the hardest thing to watch, yet it has opened my eyes to appreciate life in a completely different way, I can only take that as a positive. No amount of tears, doom and gloom thoughts or the "poor old me" attitude will ever change him, so what's the point of feeling that way?

So, find that smile, wipe away those tears and grab hold of the hand that may need a pull sometimes! Trust me when I say you will soon see life with sunshine rather than a big black cloud it would be so easy to sink under.

Alpha-1, you are a tough battle to beat, but believe me, if anyone can, my Mick can.

To all you Alpha's and carers… don't stay under the cloud because the sunshine behind it is shining so bright ☺ xx

Member Stories - Alpha-1 associated panniculitis: My story

by Nicola Lynch

My world, as I knew it, was shattered in May 2015. Who knew that panniculitis is a rare condition associated with Alpha-1 Antitrypsin Deficiency? I certainly didn't. It is rarely mentioned in most information about Alpha-1.

I was originally diagnosed with Alpha-1 by chance (perhaps an extra box mistakenly ticked on a blood sample form - it's a bit of a mystery) as I did not have any Alpha-1 symptoms at the time, back in 2006. It was still a devastating diagnosis to be given.

Forward then to 2015. Since my diagnosis I had been continuing with what I'd call a 'normal' life. The only differences were that I had changed what cleaning and other products we as a family used, and I was careful to avoid smoky environments and other pollutants that might affect my lungs. I thought I was being careful, keeping healthy, rarely needing to see my GP. Indeed, the fact I had Alpha-1 had become something I rarely thought about.

That changed in the last few days of a holiday in Portugal in May 2015 when, out of the blue, large, raised red patches started appearing on my body. Overnight, and the following day, more and more of them appeared. I can't put into words just how agonising and painful they were - I couldn't bear any pressure on them. The local doctor thought it was perhaps an allergic reaction and gave me an antihistamine injection and steroids. But I was worried. It wasn't like anything I had ever come across before.

Member Stories - Alpha-1 associated panniculitis: My story *(continued)*

New raised, red lesions (also often referred to as plaques or nodules) continued to appear. These were on my lower back, buttocks, legs and abdomen at first. The pain was immense. I could feel my body swelling in these areas as my skin felt tight and stretched. We flew home the next day and I still can't believe how I managed that journey and managed to walk out of the airport. I just wanted to get home but I felt so ill; paracetamol and ibuprofen went nowhere near to reducing the pain. So I asked my husband to drive me straight to our local hospital where, after being bounced from A&E triage to the out-of-hours doctor then back to A&E, I was admitted into hospital.

You could say I was lucky! Fortunately, I already knew I had Alpha-1 Antitrypsin Deficiency when asked about my general medical history at my hospital intake. It was, and continues to be, a recurring theme of surprise amongst medical staff that I am an Alpha whilst I currently have none of the classic lung or liver symptoms. After various tests and other potential causes having been ruled out, I had a deep tissue skin biopsy of one of the lesions. It is only from this (and a blood test confirming I was indeed AAT-deficient) that a diagnosis of Alpha-1 related panniculitis was made. (It should be noted that there are other causes of panniculitis, although I cannot speak as to whether the symptoms and progression are the same or not.)

At first, I was relieved. I had a diagnosis, so help would surely be given and these painful lesions would go away and my body would stop swelling? But none of my doctors had ever seen a case (I've seen it written that there are less than 100 known cases worldwide), and they were at a loss as to what to do. I was already frightened but now I became very anxious. I was put under the care of four different medical teams and departments. It felt like no-one wished to take responsibility for my case, as they did not know what to do.

More and more lesions continued to appear and my body continued to swell. I went up three dress sizes in less than a fortnight! By this time it was incredibly difficult to walk properly and the words that come to mind to describe how I felt are "fear", "uncertainty", "agonising pain", "anxiety", "despair" and "frustration". I was given a variety of pain killers to try to control the pain and the doctors were trying to see if steroids would control the panniculitis. They did not. It was then decided to refer me to one of the top dermatology centres in the UK at St Thomas' Hospital, London. However, at this point, for a while, things took a turn for the worse!

My transfer to St Thomas' did not go smoothly. I found myself being transferred in a transportation ambulance on a narrow hard seat (I insisted on taking a hospital pillow to sit on, though it didn't help much in cushioning me from the pain or the lack of suspension of the ambulance on our journey). Even now, as I write this, my chest becomes tight and anxiety starts to grip me, as the events at the time were extremely traumatic and, two years later, still affect me.

It was now two weeks since the first lesions had appeared and I had multiple lesions all over my body except for my chest, neck, face, hands and feet. Some lesions had come and gone, changing straight from a red lesion to a yellow bruise before disappearing though the structure beneath the skin was much altered. Others seemed to be going nowhere, and some had merged into

Member Stories - Alpha-1 associated panniculitis: My story *(continued)*

each other, forming large hard red lesions, all the time stretching my skin even tighter. I remember feeling at times like I was going to burst. Worst of all was that some of the lesions seemed to be forming fluid filled blisters, and when I arrived at St Thomas', I found that the journey had not only left me in extreme pain and distress, but that I was now leaking. I could feel liquid slowly running down my legs. This was liquefied subcutaneous fat!

The subcutaneous fat that lies beneath our skin, the essential layer that helps regulate our temperature, protects our body, houses the nerves and blood vessels that feed our skin and senses, was being attacked by my immune system, liquefying the fat and breaking it down, unopposed, due to my lack of Alpha-1 antitrypsin. It seems that Alpha-1 antitrypsin also plays a large role in the skin, which is less understood than its role in the lungs.

Originally, it had been organised for me to be a day patient at the dermatology department, and to stay in their family hospital accommodation. However, when I was brought in by the ambulance team, the resident doctor took one look at me and decided I was too ill to be a day patient and needed to be admitted to a ward immediately. In fact, the team at St Thomas' was surprised that my local hospital had even considered it a possibility that I would be a day case. To say that it was a relief to be admitted to Professor Catherine Smith and her team is an understatement.

Whilst I received and continue to receive, great care and help from Professor Smith and her team at St Thomas' and Guys Hospital, the events and illness were very traumatic. No-one who hasn't experienced it themselves can comprehend exactly how intense, agonising, and really horrific the pain and damage caused by panniculitis can be. With the majority of my lesions having been across my buttocks, hips and thighs (the most common areas it appears), sitting was near impossible, yet I was asked/expected to do so often – during wheelchair transport from the ward to other departments for scans, tests, and a further deep tissue skin biopsy. I was also transferred across to Guys Hospital to be seen at a meeting of senior doctors and consultants from various other hospitals for further advice and input into treatment suggestions.

During this time, I was taken off steroids and put onto a medication called Dapsone which is often tried in cases of panniculitis, although with limited success. It did nothing to control my panniculitis. I was also given stronger pain medication and intravenous antibiotics, as I developed an infection after a second biopsy was taken.

Then the holes appeared! I cannot describe how horrifying it is to see deep holes opening up on your body, where once there was a blister. When would it stop? Would it ever stop? What would be next?

As great as my doctors at St Thomas were, no-one had answers as they had never seen such a case before. I think we were all searching the internet for information, medical papers or published case studies. But even the few reports we found provided very little information on

Member Stories - Alpha-1 associated panniculitis: My story *(continued)*

what happened to that patient after the initial treatment? Did panniculitis return? Did they survive?

What causes panniculitis and why mine started all of a sudden is not understood. There seems to be no common cause, although physical trauma has been linked to it in some cases. The gold standard of treatment for Alpha-1 associated panniculitis is to receive infusions of Alpha-1 antitrypsin enzyme, which is purified from pooled human blood plasma ("augmentation therapy"). Receiving these infusions has been reported to halt progression of panniculitis as it allows the body to start healing. Regular infusions are required until the panniculitis stops; in some patients, panniculitis never stops entirely (relapsing) and infusions need to be continued regularly.

The impact of receiving Alpha-1 antitrypsin infusions for me was immediate. No new lesions appeared, and the existing ones finally started to heal. I was very lucky to have found a medical team willing to take on my case, research it and its treatment options and to obtain access to augmentation therapy, which I believe has saved my life. In the UK, where currently no augmentation therapy product is available on the NHS, special permission must be sought and frequently re-sought, as these infusions are only granted in exceptional circumstances.

During the nearly five weeks I was in hospital, the anxiety attacks that started in my local hospital continued, and I felt very alone and isolated. My family and friends did their best to visit me and send me messages but it was not easy being so far away from home. My husband spent a lot of time visiting most days before or after work but both he and the children were under a lot of stress themselves, trying to deal with my illness, and the fact that I was not at home. How my husband managed juggling work, visits, the kids, keeping food in the cupboard, planning meals, sorting out the dog, bills and everything else that comes with the running of a household, I will never know. But he did, and I love him for it, for standing by me and never giving up on me. I am also very proud of how my girls who stepped up to help out at home and how the eldest two took responsibility of looking after their young sister whilst going through their GCSE's and A levels.

We are lucky enough to have some of the most amazing friends and neighbours who stepped in to help with dog walking, making evening meals, giving lifts to the children, distracting and looking after my youngest daughter still in junior school at the time, coming up to London to visit me and so much more. We will be forever grateful.

The result of many weeks of untreated panniculitis has meant that I have been left with much tissue damage, many scars both physical and mental, peripheral neuropathy and living in chronic pain from the nerves that were damaged. I struggle with temperature regulation in my legs (dysesthesia), post-traumatic stress disorder, anxiety and mobility issues. Today I am still learning to live with my condition. My days are far from easy, as I live with extremely high pain levels, where even some of the strongest pain medications only just keep it at levels I can manage and still put a smile on my face. Just trying to do small household chores or walk short distances is difficult. I still struggle to stand for any length of time, or walk for any distance and I often need my wheelchair as the pain and fatigue is becoming too much. Sitting upright puts intense pressure on the areas of damage and is extremely uncomfortable as my pain levels rocket, so most of the day is broken up lying down or reclined in a chair to rest and ease the pain.

I am still struggling to be able to cook an evening meal without being left in tears and in intense pain for hours. Family life has had to change and adapt. I still wrestle with the fact that I am unable to do the things I wish to do for my family, that my husband still has to pull both his weight and mine, and that I cannot do the things with my younger daughter that I did with her older sisters.

I grieve at the loss of my life as I knew it, my loss of independence and acceptance of my need to use a

Member Stories - Alpha-1 associated panniculitis: My story *(continued)*

wheelchair, at not being able to work. Most importantly, I'm still coming to terms with the changes and effects it has had on the family and my own role as a wife and a mother. The fear of the lesions returning and me not getting treatment is a constant threat. For a long time, my mind kept replaying traumatic events that happened during my hospitalisation. I was extremely fearful when leaving the house or being in busy crowded places. I withdrew a lot from family and friends as everything was overwhelming and exhausting. It usually left me in tears and I struggled having to explain what happened and how I am. I have struggled with depression and the limitations of life now, but although I still have difficult days, above all I am grateful.

I am grateful that I am still here, that I can hear the birds singing, go outside and watch the wind move through the trees. Over a month in hospital with no outside view increases your appreciation of the simple things in life.

I am extremely grateful to everyone who has enabled me to receive Alpha-1 antitrypsin infusions. It is hard to write this knowing how many other Alphas out there would benefit also from receiving augmentation therapy, but currently do not. It is my hope that all of us will one day soon be able to have access to this essential therapy.

There are still many challenges, but I engage once more with the world, although not in the same ways as before. I'm kinder to myself, allowing time to rest and recover or doing things in small doses. I have also realised that there is still a long way to go in making the world truly accessible for wheelchairs (but that's another story!), and how we often need as much help and loving care with emotional and mental challenges as we do with physical impairment.

I don't know what the future holds, but then who does?! Continued infusions of Alpha-1 antitrypsin are not guaranteed and I don't receive them weekly, so I am regularly seeing signs of panniculitis starting to reoccur before receiving another set of infusions. My life is full of stress and uncertainties, as for any Alpha, but I have learnt to live day by day and enjoy what each day brings, regardless of its ups and downs. It is my family that keeps me going, and I take it one step at a time. One day at a time.

Support group for patients with Alpha-1 associated panniculitis

Every patient is different, but I have found that there is a lot of inaccurate and misinformation about Alpha-1 associated panniculitis out there. However, my own experience and that of others who I have since managed to get in touch with, is far removed from many descriptions of the condition. While there may be less severe cases, Alpha-1 associated panniculitis can make people very ill and have a huge detrimental impact on their lives ranging from leaving them with scars, trauma, tissue loss, skin depressions and other changes, living in chronic and severe pain, and more.

We may be very few but we are part of the greater Alpha-1 community and many of us are also affected by other, more common symptoms of Alpha-1. I hope that you will help raise the awareness of Alpha-1 associated panniculitis in your own patient community to help share more accurate information about this awful condition.

It is my desire to bring together other Alphas with panniculitis, to support each other and to pool our collective knowledge of our disease, and to create a more detailed bank of information and hopefully help to further research on this aspect of Alpha-1. One of the many good things that have come out of my own experience is that I have created a Facebook group specifically for those who have Alpha 1 panniculitis (Alpha 1 Panniculitis Support Group, at https://www.facebook.com/groups/1748644008780254/, as I know how lonely and isolated I felt when first diagnosed.

Member Stories - Lee's lung transplantation story

by Cora Paterson

We found out about Alpha-1 Antitrypsin Deficiency just after my father-in-law died. It was listed as his cause of death, and he had only found out about it shortly before he died. He didn't tell me, as I was pregnant with my second child at the time. My son was 10 days old when my father-in-law passed away. His son, my husband Lee, then went for genetic testing and found out that he was phenotype ZZ. We didn't think anything of it for a while until he started to experience some breathing difficulties. Firstly this was ignored by Lee's GP, then Lee was fobbed off with "you are asthmatic" although the GP had the results from his genetic test!

With things not improving, Lee insisted on a hospital referral where he was diagnosed with Alpha-1 antitrypsin related emphysema. At first, the decline in Lee's health was gradual. He got lost in the hospital system and had not been seen in clinic for three or four years, so Lee chased for an appointment and finally got to see a new consultant. It was at that point that the possibility of a double lung transplant was discussed.

Lee was referred to Harefield Hospital for transplant assessment. This initially took two days, followed by five months of further tests. Lee asked what his likely prognosis was without transplant and, although he knew his health had declined, he was shocked when he was told that he was likely to have around three years left to live. Lee was finally placed on the transplant list and, as we were advised, we packed a bag and awaited the call from the hospital for a suitable transplant organ.

Lee attended pulmonary physiotherapy at this time, and was allowed to keep attending after his initial course had ended, in order to maintain his fitness level so that he could be in the best possible physical condition for his body to cope with, and heal, after a transplantation.

A couple of weeks after being added to the transplant list, we received a phone call at one o'clock in the morning. I can't say what Lee was feeling, but he was shaking, and I could hear my heart pounding in my ears, my throat and mouth were extremely dry and I too was shaking. We called Lee's mother to look after with our 5 children while we dashed to Harefield. After several hours of waiting, Lee had a bed but was too nervous/excited to sleep. I had the choice of a couple of very uncomfortable chairs, so I couldn't sleep either. After all this excitement, we were told that the operation couldn't go ahead, and we got our first "sorry, not this time".

It was initially hard to take in this news and the disappointment hits you hard. At the time it is difficult to see beyond your own disappointment to spare a thought for the donor family who have lost their loved one.

Afterwards, sleep at night became fragmented for Lee and myself, and even the cooling of the heating pipes would wake us and we'd think "is that another call?". Eventually, after a total of six night-time and three daytime calls (the latter taking me away from work) and nearly six months of being on the transplant list, we got the call that changed all of our lives.

We received the call at 3.15 am and did the usual sitting and waiting in the hospital, thinking it's going to be another "sorry, go home" when the co-ordinator came in and said "we'll be going to theatre in an hour". It hit us both like a bolt. This was the first time I had seen Lee cry

Member Stories - Lee's lung transplantation story *(continued)*

throughout the whole process. All the waiting had been extremely hard and stressful.

I went down to theatre with Lee and gave him a kiss before leaving him in the hands of the hospital staff. I was moved to the relative's room to await news, and two hours later the co-ordinator came to tell me that the operation was well underway and that I should have a cup of tea before driving home. I can't remember much about the drive home.

I went to Harefield the next day. Lee was in ITU, and he had a breathing tube, drain tubes, a monitor and leads and lines bleeping and buzzing everywhere. The staff did try and warn me, but seeing it yourself can be quite shocking and overwhelming. Lee's body was extremely swollen and, in need of some normality, I joked with a friend that he now had bigger breasts than me. The next day, one of our children was unwell so I couldn't go to the hospital to see Lee and he was moved from ITU to his own room on the ward.

Lee made good and steady progress. Without the support from Lee's mother, a few really good friends, Andy and Keith from the Alpha-1 UK Support Group visiting Lee, it would have been impossible for me to cope with that quite confusing time.

Lee did so well that, 13 days after his transplant and on his 41st birthday, he came home. He continued to recover well and pushed his exercising a little more each day. Lee was advised that it could take up to one year post-operation to recover, but as the UK benefits system was under reform, all of his disability benefits stopped after three months. Luckily, I was still working and had taken as little time off work as possible.

All of our lives have changed so much for the better. One Sunday morning Lee walked over five kilometers without oxygen - just over a year ago he had struggled to walk six meters from the sofa to the toilet without oxygen.

Lee has traded an existence for a life that now revolves around phone alarms to remind him of the medication that needs to be taken several times a day. There are other limitations to Lee's and other transplantee's lives, such as restrictions in diet. For example, eating out should be at 5* Hygiene rated premises. Finding a job that won't harm his health has been difficult: he can't go back to his job as a car valet because of the chemicals; no customer facing roles and no office environments are suitable due to infection risk; he should avoid dusty environments and gardening as the spores can cause problems.

Shortly before reaching the one-year milestone since transplant, Lee did a charity abseil on behalf of Harefield Hospital.

During this last year, Lee has changed from the dad who sat on the sofa just watching the children to a dad who got up and danced with our three youngest children on a recent holiday, something that we never thought possible over a year ago.

We have recently celebrated one year post-transplant. As we celebrate a year of good health and quality of life, our thoughts are with the donor and we spare a thought for their family and loved ones for their loss. We thank Lee's donor and all donors along with their families who let their loved ones' end be someone else's new beginning.

Member Stories - My transplant journey

by Tracy-Anne Lawrence (Gayatri)

I was pushed along the corridor of Rowan Ward at the world-renowned Harefield Hospital by a very fast transplant coordinator and one of the lovely nurses that seem to be everywhere at that hospital. Also walking serenely and very calmly was my boyfriend Simon, known to many as Burn. We made a very quick dash to the lift when the sharp-eyed coordinator noticed my pretty toes, with pink and blue varnish and silver toe rings. You would have thought she was rushing me to surgery with a critical wound considering the speed she ran back for nail varnish remover! They worked on my feet in transit, in the lift. Most of you will appreciate how the fumes from the varnish would be effecting compromised lungs.

Tuesday 16th May 2017, 7.30 am

I was wheeled into the small area just before anaesthetics and surgery. Simon and I were given a few minutes alone. We basically stared at each other in disbelief with massive big grins on our faces. After a few minutes I was pushed into the anesthetiser area and, after a very short chat, I woke up in ICU on Wednesday at about lunch time.

Waking up was quite surreal. Those few seconds between sleeping to waking, when you can be a little disoriented, I realised I was alive and that was the most amazing realisation.

In the next few hours they had me sitting out of my bed in my chair. I have vague memories of my loved ones visiting me but, to be honest, I was a bit woozy so don't recall much.

I was later taken to the step-up Ward and to room 11 where I spent the first part of my journey to recovery.

I first started to notice the effects of Alpha-1 Antitrypsin Deficiency when I was in my mid-twenties. I was a smoker, not a heavy smoker - maybe 20 a day. I had an awful smoker's cough along with a deeper voice, it was how I was known. It didn't really effect my fitness levels and I was very active. Looking back now, I realise I did get a bit more out of breath at exercise than I should have.

I lost a lot of lung function when I renovated my house after a fire, and also after the sad loss of my daughter who was born prematurely and still born. Then, two years later, I had another miscarriage of a daughter who I carried to late term.

So, by age 34, I was still working full-time as a restaurant manager but finding it quite challenging as the restaurant was on 3 levels.

By then, I had visited my GP who sent me for a chest X-ray, and they discovered a shadow on my lungs. I was not offered any support, or even inhalers, just told to stop smoking, which I had started again after the loss of the babies. I didn't heed that advice initially.

By the time I was 38, I was severely underweight and I had left my job, sold my house and was on a spiritual quest.

I spent the good part of a year living in a spiritual yoga community in Wales before travelling, first to France to another Yoga ashram, and then a Zen Buddhist monastery.

I then stayed with a friend in Catalonia where I ended up in hospital with multiple chest infections which, apparently, had been eating away at my lungs for many years. After treatment there I was healthier than I had

Member Stories - My transplant journey *(continued)*

been in many years and with a new found vigour I travelled to Ireland where I house sat in the summer months.

I spent summers in South West Ireland and winters in South East Asia. I took a teacher training course in India with what, I now realise, must have been the maximum of 45% FEV1.

Thankfully, while In Ireland, I was sent by my GP to The Matter Hospital in Dublin to Professor McElvaney. He swiftly diagnosed me with Alpha-1 Antitrypsin Deficiency. This was the first time I had had spirometry tests, and my FEV1 was 36%. I was offered to go on a double-blind clinical trial for augmentation therapy. I declined the offer, as I found travelling to warm climates where I didn't get chest infections was good not only for my physical but also for my mental health.

About ten years later it was suggested that I was nearing the time where lung transplantation should be considered. I decided to come back to UK, as support for such a major operation was needed and my family was mainly in the UK.

I was referred to Harefield Hospital and, after testing, I was offered a place on the waiting list. I declined the offer initially, as I was still popping off in the winter months for my sun and fresh fruits and to live on the beach.

By this time, I was on ambulatory oxygen up to three litres for walking. I had a fabulous Sequel Eclipse portable oxygen concentrator. I promised myself, and my doctor Dr Anna Reed, that as soon as my oxygen needs changed the quality of my life, then I would get on the active transplant list. That happened 18 months ago. After three “dry runs”, I finally got my call on Monday 15th May 2017.

For the first 12 weeks after the operation, I had to be extra careful to avoid public transport and busy places. I spent this time at home doing all the little jobs that would normally take me days to complete. It has been amazing to be able to do simple tasks like hang the washing on the line or to be able to bend down to take the laundry out of the washing machine.

Today, three and a half months after the operation, I’m in Bournemouth for the August bank holiday, spending time with my eldest son Dan and his wife Mel. We walked three miles to the beach front and home again with a little detour via the shops.

How life has changed! I have spent the last ten years or so putting my life on hold and saying “when I’ve had my transplant...”. Well, that time is now! I’m not quite sure yet what direction I would like my life to go. I need to think about how I can use this second chance I have at life to combine earning a living and pursuing my interests?

I would like to start to train for the transplant games in Birmingham next year. This training will also strengthen me for doing the El Camino de Santiago de Compostela pilgrimage, which is my big aim. I will look for sponsors for this challenge, and would like to give the donations to the Alpha-1 UK Support Group and to Harefield Hospital. I have been given the okay from Harefield to start at the gym in September. I need to do a daily walk, gradually increasing the distance to get to the point where I’m able to walk 8 kilometres daily.

I also love to travel, I love yoga, meditation, healthy cooking. I have also started a shamanic journey to thank my organ donor and to enable them to cut ties with their gift to me. This has given me peace.

I am still experiencing some pain, especially since two of my pain killers have been stopped. I also seem to have some stomach issues that need to be addressed, I’m swollen and I have put on over 24 pounds in weight.

But I can breathe!!! I still feel overwhelmed and grateful.

Member Stories - Mike & Alison Dyke's Story

by Mark Acland

Mike was diagnosed with Alpha-1 at the age of 21. Local doctors carried out blood tests following recurrent water infections, which showed his white blood cell and platelet levels to be extremely low. Following this, Mike was admitted to hospital for further tests, including a lumbar puncture to test his bone marrow – they initially thought he may have leukaemia. Whilst in hospital he vomited blood, and an endoscopy revealed he had varices. A liver biopsy finally confirmed his diagnosis of Alpha-1 Antitrypsin Deficiency, with an SZ phenotype.

Mike was advised to stop smoking and drinking immediately – he was told that even one alcoholic drink could result in him losing his life. Mike reduced his drinking, but did not completely give up. He thought what many probably think: it's never going to happen to me! Along with his Alpha-1, Mike also suffered from an enlarged spleen, varices that burst and bled, and which he has had to have glued and banded a few times to save his life.

I met Mike when he was 24 years old. I knew nothing of Alpha-1, and as Mike didn't know much about it either, we didn't talk about it much. We had our son one year later in 2010. Life was normal, we both worked and enjoyed life as a family. In 2012 - I was eight months pregnant with our first daughter and we had just returned from a weekend away - Mike had a massive bleed. One of the varices had burst. He was given transfusions and platelets, but was told he was risking his life – he has not had an alcoholic drink since that day. He very nearly didn't meet our daughter.

Everything settled down again but, unfortunately in February 2014, Mike had another huge bleed and spent two weeks in hospital. Following this hospital admission, he returned to work for a short time, but then sadly had to give up work completely. Mike hated giving up the job he loved, and this led to him becoming very depressed and he was prescribed anti-depressants. This is when Mike's condition started to worsen.

In 2014 and 2015, Mike spent more time in hospital than at home. He was carrying a lot of fluid in his legs, and he had a few bleeds from the varices. Ascites was diagnosed, and his liver was failing. In early 2015, he had a transjugular intrahepatic portosystemic shunt (TIPS) procedure at Queen Elizabeth Hospital in Birmingham. The purpose of this shunt is to relieve the blood pressure in and around the liver, which reduces the risk of bleeds.

Doctors were very hopeful that this procedure would improve Mike's condition, however the results weren't as positive as we had hoped for. Mike suffered from confusion. Life was very hard, and I became his full-time carer. We decided to complete our family with another baby, and our daughter was born in March 2016. It was very difficult for Mike to be in hospital all the time, and for me and the children not to have him around. Luckily, we had amazing support from both our families. We then started to get more knowledgeable about AATD, and how it may affect our children's health. We had to fight for three years for our son to be genetically tested, but we now have confirmation that he is a carrier with an MS phenotype. Doctors don't want to test our daughters unless they become symptomatic.

Mike was placed on the liver transplant list in 2016. He had three phone calls for the operation, but none of them went ahead. We were never informed of the exact

Member Stories - Mike & Alison Dyke's Story *(continued)*

reasons why the transplantations were cancelled. In January 2017, it was discovered there was something wrong with Mike's heart - it wasn't pumping as it should be, more than likely as a consequence of the liver not working properly. In February 2017, we attended a routine appointment at our local hospital, only to be told that Mike was being taken off the transplant list due to the issues with his heart. At that point, Mike was given just 12 months to live, as there was no further treatment that he could be offered. Mike, myself and his mother sat in a room, just crying and holding each other. You never think you'll ever have to hear those words, especially at the age of 31.

We had to go home to our three children and put a brave face on. How do you tell a 6- and a 4-year old and a 11-month old that they've only got daddy for another year? You can't! So, we didn't. They knew that daddy was poorly as, by this time, he was on a feeding tube and going to the hospital for weekly check-ups.

Mike started to plan his own funeral. I just couldn't face it - the thought of losing him and raising our children alone was too much for me to deal with. In March 2017, doctors agreed to do some further tests including stress testing on his heart. A week after the tests we were informed that Mike was active on the transplant list again. I can't even begin to describe how much that meant to us!

Within two weeks, a suitable donor organ was found. As Mike went into theatre at Queen Elizabeth Hospital in Birmingham, Mike's mum and dad and I held him so tight. We were all aware of the risks. I held him close and he promised to come home to me and the children.

We had been told it would be at least an 8-hour operation, and after only 3.5 hours the surgeon asked to see us. We tried to prepare for the worst news. Amazingly, the surgeon told us that the operation had been a complete success!

Mike recovered extremely well, and only eight days after the operation, he was home. It has been a long healing process, and he still has other medical issues which are being worked through with the medical professionals. One of the main concerns is that his heart has only slightly improved since the operation. Despite this, Mike is hoping that he will be able to return to work one day, however, doctors are still unsure whether this will be possible.

Overall, it has been a difficult journey, and has affected the whole family. Our 7-year old son Thomas has asked lots of questions, which we have always answered honestly, and we have explained to him what it means to be a carrier of Alpha-1. But to watch him with a great big smile on his face, kicking a ball around the garden with his daddy, fills my heart with so much joy! Mike is still not able to do loads but he is alive, and we are raising our children together.

Our children have their father again. Within the space of a few months, we went from planning Mike's funeral to planning our future. It is very true that you do not how much you love someone until you think you are going to lose them. Every day we are thankful to the donor who saved Mike's life.

In Memory of John Mugford

by the Board of Trustees

Sheila, John, Darryl, Paul and Ryan

5th October 1951 – 30th January 2017

Our dear friend John passed away peacefully at home, aged 65, and is survived by his wife Sheila and theirs sons, Paul, Darryl, and Ryan.

John was diagnosed with Alpha-1 Antitrypsin Deficiency in 1999 after contracting double pneumonia. Two years later he joined the Alpha-1 UK Support Group, became a member of the Committee in 2008 and was elected as Chair in 2010. He was instrumental in establishing the Group as a registered Charity, initially in England and Wales, and subsequently in Scotland.

John was always passionate about utilising the internet to improve his knowledge about all aspects of Alpha-1. He derived a great deal of pleasure and satisfaction from passing on that knowledge and from helping and supporting others in any way he could. Improving the lives of Alphas became John's mission. He was a committed and tenacious patient advocate - supporting Alphas and their friends and family became a full-time role for him. He established the group's social media presence, regularly shared relevant information and news and was the "communications centre" of the charity.

John was a very private man and didn't want anyone outside of his personal circle to know how ill he had become. His selfless dedication is further demonstrated by the fact that, still hours before he passed away, he was messaging group members and supporting them to the very end.

As many of you may know, John had a passion for music - he was a part-time musician, playing in a band up and down the country for 30 years. His "Song of the Day", which he posted on the group's Facebook page, was eagerly awaited by our members. John's last Song of the Day, posted on 29th January 2017, was aptly "I want to know what love is" by Foreigner – aptly, because the tributes received in John's memory have demonstrated how loved he was within the Alpha-1 community, not just in the UK but internationally, due to the kindness, support and professionalism he showed to all, even at times of great personal suffering. The "Song of the Day" tradition is continued in his memory by his son Ryan and our members.

John was a true gentleman and a devoted Alpha-1 advocate, and it was always a great pleasure to work alongside him. His efforts have made a significant difference to the development and growth of the charity, and he was delighted to see the establishment of specialist NHS Alpha-1 Clinics in several hospitals leading to better care for Alphas. With one augmentation therapy product now licensed in Europe, John remained hopeful that treatment was on the horizon for Alpha-1 patients in the UK.

John has left enormous shoes to fill, and the Trustees and Committee remain dedicated to continuing his good work. We would like to thank his wife Sheila, and his sons, for sharing him with us.

Breathing easy now John, our Oracle

Personal reflections about John from our members

"He united people across continents."

"A truly remarkable man."

"His generosity of spirit was boundless and his wisdom and kindness incalculable."

"I didn't actually know John, and he didn't actually know me, but he helped me just the same."

"He helped me more than he could ever know."

"Such a kind and helpful person. I will forever be grateful to him."

"My favourite reason to check FB -- John's posts!"

"John was a truly inspirational gentleman."

"John was one of the most generous of all.

"His work will be an admirable remembrance and he will be missed by us all."

"...he instilled great knowledge and drive to keep the fight going…"

In Memory of Norah Elizabeth Oliver

by her husband Bob, Daughter Lisa Marie and her sisters Dolly & Ivy

21st July 1947 – 24th December 2016

It would have been Norah's 71st birthday on the 21st of July this year, and we all spent the day reminiscing of happy times in the past and of Norah's positive outlook on life, despite the challenges that she faced daily. She enjoyed a wonderful 70th last year and felt blessed at achieving this milestone following her diagnosis of Alpha-1 in her 40's.

Bob and Norah

Lisa Marie & Norah

Norah passed away peacefully at home on 23rd December 2016, surrounded by her family. It is now nine months since we lost her, but not a single day passes without thinking of her and missing her from the bottom of our hearts. Despite her ongoing pain and breathlessness over the years, Norah would always "put her face on" and face the world with a smile and always a kind word for anyone that she met. Norah really enjoyed going shopping with her sisters and, if the occasion was buying clothes, she would come home with quite a few bags and the comment "don't worry Bob, most of it is going back". This was because she simply did not have the energy to try on anything in the store.

Norah was very family-orientated, and her grandchildren Joseph and Erin were her world. She insisted that she wanted to see them, and for them to come to tea following school once a week 'kept her going'. Both Joseph and Erin have fond memories of holidays in Norfolk with Norah and loved to 'hitch a ride' on her scooter.

Despite Alpha-1 affecting her life so significantly, Norah never gave up and wanted to support others recently diagnosed or in need of advice. Lisa recalls her often talking about the people she met at the annual meetings, and knows how Norah would always strike up a conversation. She certainly was not shy and would know a persons potted history, normally within ten minutes of meeting them!

Following are memories from her sister Dolly and her sister Ivy.

Dolly: Norah was my youngest sister, with Ivy in between us. We grew up being very close. As teenagers, we all went dancing together which was an important part of our lives at that time. As time passed, we all got married and had families, but we all returned to Stamford to retire. Norah never let life get her down and she was always there for anyone who needed help or advice. As her illness progressed she was very brave. I miss her so much and won't ever forget her. I love you Norah.

Ivy: Norah had five sisters in total, all of whom grew up very close, but Dolly and myself were always extremely close to Norah and spent most of our time together, especially as teenagers.

Dolly, Lisa and myself phoned each other daily, and these chats are sorely missed by us all. Norah always had, or made, time to support her family and other friends - Bob is now having to develop his telephone communication skills!

Norah made some wonderful friends through the Alpha-1 community and, as is the way with this illness, quite a number of those friends are unfortunately no longer with us. Norah is now at rest and breathing easily.

Ivy, Dolly and Norah

In Memory of John W Walsh

by The Board of Trustees

4th February 1949 – 7th March 2017

Alpha-1 Patient, Co-founder Alpha-1 Foundation and COPD Foundation, USA

John W. Walsh, Alpha-1 patient and passionate, dedicated and visionary patient advocate, passed away on Tuesday, 7th March 2017 in Miami, USA, surrounded by his loving family.

John is survived by his devoted wife Diane, daughter Linda and granddaughter Lily. John also leaves his twin brother Fred Chase Walsh, and sisters Susan Walsh Ferro and Judith "Juba" Morton Walsh. They, along with an extended family of cousins, nieces, a nephew, their families, and many others, will miss John's warmth, compassion and spirit of life. John leaves behind an extraordinary legacy of civic service, advocacy and philanthropy.

In 1989, John was diagnosed with Alpha-1. Lung disease resulting from Alpha-1 had taken John's mother at the age of 48, and Alpha-1 also affects his sister Susan and his twin brother Fred, who received a double lung transplant.

After discovering how little was known about Alpha-1, and that only minimal research was being conducted in this area, John co-founded the Alpha-1 Foundation in 1995. The Alpha-1 Foundation is a not-for-profit organisation whose mission is to develop a cure for Alpha-1 and to improve the lives of those affected by the disease. Later that year, John co-founded AlphaNet, a not-for-profit organisation providing health management services to individuals with Alpha-1 in the US. AlphaNet utilises a unique business model and has, to date, contributed $50 million dollars to Alpha-1 research and related health programmes.

John led the Alpha-1 Foundation as President and CEO for more than 20 years. Under his leadership the organisation became internationally recognised for its commitment to people with Alpha-1 and for driving research for a cure and improving health services. To date, the Alpha-1 Foundation has invested over $65 million in research and health service programs worldwide.

When John started to reach out to other patients, physicians and researchers, he soon realised that Alpha-1 was often undiagnosed or misdiagnosed as common chronic obstructive pulmonary disease (COPD). This led John to commit to improving diagnostic rates of Alpha-1 in the COPD community. He quickly recognised that, like Alpha-1, COPD was also misunderstood, misdiagnosed, mistreated, underfunded and lacked adequate research. In response to the critical unmet need by the millions of diagnosed and undiagnosed COPD patients in the US, John formed the COPD Foundation in 2004. The COPD Foundation is a not-for-profit organisation, whose mission is to prevent and cure COPD and to improve the lives of people affected by it.

John's call to action was to educate, engage and empower patients. His contributions to patient empowerment and capacity building are unprecedented in the Alpha-1 community. John's voice impacted the direction of clinical research, strengthened the patient voice and changed the way patients are integrated into medical education. John forged a patient-centred movement in Alpha-1 bringing together patients, physicians, researchers, government agencies and pharmaceutical companies. The result of this longstanding collaboration has increased the longevity and the quality of life of those affected by Alpha-1 in the U.S., and a positive impact has started to be visible in many other countries globally.

John's entrepreneurial spirit, combined with his commitment to community service, set the stage for a patient-centred and visionary approach in the voluntary health sector that will likely remain unmatched.

John received numerous awards and accolades for his work. He often referred to himself as an "impatient patient" who felt compelled to take action, and he consistently cited patients, healthcare professionals, researchers, and his loving family as his source of strength and motivation.

John was a family man and a hero to many, and he has left us all with his common wish, "Keep the faith".

In Memory of Clare Dolan

by her sons Tom & Mike and daughter Emily

13th January 1970 – 9th July 2017

Tom's Memory

What can I say about my Mum or Clare? Many of you know her, although not necessarily by her name… My Mum was born 46 years ago in the hospital. As a child, she showed great spirit and loved the outdoor life. My Nana remembers chasing after her when she ventured out of the garden to try see the rest of the world. She was only two at the time! My Mum attended St Anne's School in Crumpsall until she moved to Levenshulme. She then attended St Bernard's School, where she became a vital member of the school's sports team and was involved in all activities.

As my Mum finished primary school and started to grow, she attended The Hollys High School where she made many friends who she retained throughout her life. My Mum and her friends often got up to all sorts of mischief, much to the nun's despair. My Grandad, Mum's father, was called in to take her home on occasion. Knowing my Nana was at work, my Mum loved this as she knew that, when Dad came, unlike her Mum, he would take her for drive and a shandy.

After leaving school my Mum made many new friends through work, due to her outgoing nature. She married at a young age and had two beautiful kids (if I may say so myself). When I was little, Mum often took us out and we would end up walking for hours. I also remember one time, when Mum told the school on a Friday at lunch time that myself and my brother Michael were going to the dentist, she would sneak us away on a train to Blackpool for the weekend, leaving us with fantastic memories. Nine years after having me, she had a much -loved daughter, Emily, who she loved to argue with and wind up.

When I grew up, Mum had many jobs, working in restaurants, bars and care homes where she made some fantastic friends. My Mum loved to travel and would often take us away as children. As she would not be able to afford to take all of us away at the same time, we would take turns for a break away. We children loved this, as we had her total attention for ourselves at these occasions.

Emily's Memory

My mum was an interesting person: talkative and very friendly, but she could also be short-tempered and very big-headed about her accomplishments. She would always want to 'one up me' about athletic achievements - clearly, she was very humble. I always thought that she was putting me down, when she was actually trying to push me to do better. Along with having her blue eyes and gobby attitude, I also seem to have her morals and the ability to stand up for what I believe in. Despite disagreeing on a lot of things, she taught me to be loyal and not let people walk all over me.

Alongside 17 years of great memories and fantastic trips together to places like Gambia and Florida, one of my fondest memories

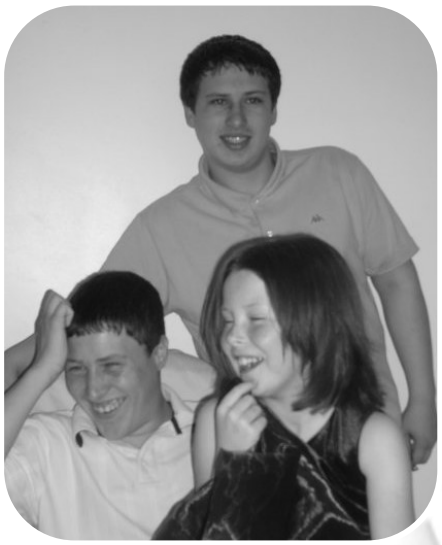
In Memory of Clare Dolan
(continued)

is a great example of how protective Mum was over me and how much she cared: I was around the age of 10 when my friends and I ventured through what we called “the brooks”. One day, a tall scary man shouted at us, and I was terrified. I scrambled my way home crying and my mother insisted on me taking her to this man. She followed me and my friends through the brooks to confront this man. The safer option would have been to tell me not to go near there again! I think deep down she knew I would not listen to her as she understood how much of a free spirit I was.

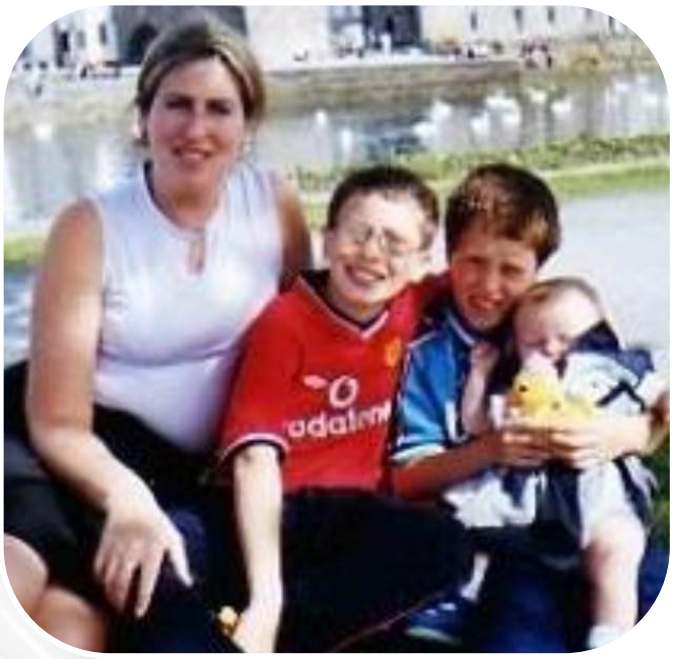
Mum was my best friend and my loving mother and will be deeply missed.

Mike’s memory

My Mum had a big heart with lots of room for others. She had three children as already mentioned - myself, Thomas and Emily. She also had people around her who she thought of as her children: two of them were her daughters-in-laws, Jenny and Shelly, but she also had Jordan often referred to as “her third (but often favourite) son, who helped after her motorcycle accident in Gambia. Also, Chelsea whom she thought of as another daughter, and the many people she helped in Gambia. Mum would help people on the street, or if someone needed an open ear, she was always there for them.



Even when my Mum became very ill, many things still brought a smile to her face. Having her grandkids Dexter and Duncan around always made her smile and cheered her up when she was feeling down. Being able to see the smile on her face at my wedding was priceless and will be a memory I will cherish.



In the later years, my favorite memories of my Mum were the small things, like having Dexter secretly walk into her room and how she would light up and be active with him. Or the way Duncan would always go upstairs and say good-bye to her, which is not something usually in his character. My Mum often talked about this with pure joy.

My Mum spent a lot of time on social media, as I am sure many of you know her from her countless posts each day. The majority of her time online was spent campaigning for worthy causes especially FGM, Alpha-1 Antitrypsin Deficiency and homelessness where any donations will be given on her behalf.

After many years of suffering and battling illness, my Mum sadly passed away due to Alpha-1 on the 9th July 2017. My mother’s wishes were to be cremated and her ashes to be scattered off the west coast of Ireland, a place she adored.

On behalf of my Mum I thank you all for attending the ceremony and showing your support in celebrating my mum’s life.

In Memoriam

All Alpha friends that we have lost have left their mark on our lives, and it was a privilege to have known them.

Reflections *by Joe Lyons*

For the people who've gone before us, your fight was not in vain
Our thoughts and prayers are with you, we tried to ease your pain
We know you were the bravest, the best that you could be
And even then you smoothed a path for someone just like me

In life we all need heroes who would fight and be strong
You are all classed amongst them, even though you've gone
In our thoughts you're always there, we'd think of what you'd do
Even when life is a struggle we strive to be as good as you

Life takes so many heroes before we can get it right
Our thoughts are always with them throughout the day and night
So pause just for a moment let your mind free to take stock
Be thankful in that moment remembering what you've got

Carol Thomas
8 Oct 2016

Sioned Lewis
28 October 2016

Sue Wynn-Jones
20 Nov 2016

Corinne Freeder
30 Nov 2016

Ray Overton
15 Dec 2016

Norah Oliver
23 Dec 2016

Tina Taylor
1 Jan 2017

John Mugford
30 Jan 2017

Clare Dolan
9 Jul 2017

Marian Wild
21 Aug 2017

Cindyanne Forster
28 Aug 2017

Poetry Please

by Joe Lyons

The Valentine

I've just received a card
I don't know who it's from
Because it had my name on it
I suppose that's why it's come
On reading someone cares for me
They really care a lot
They're sending all their kisses
And all the love they've got
I don't know if I believe it,
Or if it's just a game
For if they really cared so much
Should they have put their name?

Poetry Please

by Joe Lyons

The Extraction

I sit waiting patiently for my name to be called out
The pain won't last for very long, I must relax try not to shout
Now tiredness dulls my eyes and my head feels rather sore
Too many nights without sleep, with details I won't bore
At last my name is called I take a seat in the rising chair
The mask is on the light is bright I sit without a care
At last here comes the needle, you know I hardly felt it prick
Just relax take deep breaths, with any luck it will not stick
With a quick wrench it's gone I did not have time to shout
Apart from mouthwash and cotton wool no pain and yet it's out
Must remember, cut down on sweets try and eat fruit instead
No fizzy drinks just water or fruit juice I'll try instead
They're a pain when they come just the same when they go
I will try and slow down there departure
Do my best I know
The gaps are now bigger with the teeth no longer there
I must try to be more careful or it's dentures I will wear.

Fundraising and Awareness

A Big Thank You to everyone involved in fundraising activities and for donations to the group. This year supporters have been active as ever - running marathons, baking cakes, organising awareness stands, having a full body wax, donating in memory of loved ones and special occasions, selling sweets, making regular donations, and many other fundraising activities.

Through your activities we are able to continue funding our programmes of providing support and education for patients, families, carers and friends who are affected directly or indirectly by Alpha-1 Antitrypsin Deficiency. We aim to do this by:

- growing a social network for patients,
- providing discussion groups focusing on how better to cope with their condition aiming towards improving quality of life,
- advancing education, understanding and awareness of the condition, in particular among medical professionals, including information relating to genetic implications, treatment, and lifestyle choices, and
- supporting research and campaigning for better access to treatment for Alpha-1 patients.

We know there is so much more we can and need to do to promote better knowledge and understanding of Alpha-1 Antitrypsin Deficiency but we are limited by the funds we receive, so your support is valued and very much appreciated.

Jimmy Campbell, Captain 2016, chose our charity to donate the proceeds of the clubs annual fundraising events in memory of his wife Gillian

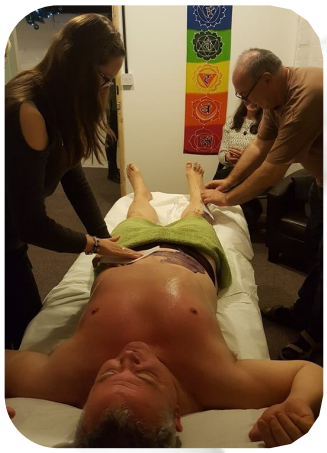
16/10/2016 - Hannah Jones & Kieran Endicott completed the Great West Run in Exeter

25/09/2016 - Emma and Jim Hunt completed the Windsor Half Marathon

The grounds of Windsor Great Park are beautiful and the marathon route went through private areas of the Queen's Estate which are not generally open to the public. Great weather and stunning scenery to keep you going when it was tough.

It was a lonely run for Emma being very near the back due to having to walk a lot due to injuring her hip around 8 miles into the race.

Jim completed the marathon and achieved his target time of 2hrs 30mins. Emma was an hour behind and her children met her on the final mile and they ran over the line together.



Hannah and her dad Mike raising awareness and donations in memory of Wendy Kilby, Hannah's nanny and Mike's mother-in-law.

Mike bravely agreed to a full body wax and their family and friends were delighted to pay extra for the privilege of ripping a wax strip off themselves.

They also held a Christmas Fayre.

Fundraising and Awareness

07/01/2017 - Callum Overton, Ridgway Cycle
Callum cycled with his friend Pete Wood in memory of his Dad, Ray Overton

Anne Cole and Jones Dorling, Racing
Raising awareness and collecting donations in the teams British Superbike hospitality area at venues around the UK.

02/04/2017 - Philip Bowden, Greater Manchester Marathon
Phil ran the marathon with his friends Lee and Matt

25/06/2017 - Liam Harris completed the Swansea Half Marathon

03/11/2016 - Jane Purves
Held a Cakes for Charity event

Tina Walker donated her BBC iPlayer Fees from "Lizzie's Last Stand", an episode she wrote, featuring a patient with Alpha-1

Fay Whittaker and Ina Smith at Kirkcaldy Victoria Hospital raising awareness and collecting donations

Sarah Willacy raising awareness and funds as she was diagnosed with Alpha-1 shortly after she got married

01/11/2016 - Imogen Charlotte Goode's niece, held a School Sweet Sale

Donations

Diane and Mick Stobart	Monthly donations
Elaine Bradshaw	Donation
Jenny and Mike Clarke	Golden Wedding Anniversary
Berenice White	Monthly Donations
Monica and Peter Ward	Donation
Emma Hunt	Donation

Donations in Memory

Arran Lewis	In memory of Linda Lee
Anonymous	In memory of Gerald Whybrow
Anonymous	In memory of Frances Barnsley
CJ and JF Derose	In memory of Corinne Freeder
Dolly Walters	In memory of John Mugford
Dolly Walters	In memory of Norah Oliver
Elaine Bradshaw	In memory of Norah Oliver
Friends and Family	In memory of Jane Hill
Friends and Family	In memory of John Mugford
Friends and Family	In memory of Linda Cooke
Friends and Family	In memory of Mark Bradford
Friends and Family	In memory of Norah Oliver
Friends and Family	In memory of Phil Orchard
Hannah Cowley	In memory of Wendy Kilby
Heather Barrett	In memory of John Mugford
Ivy Carter	In memory of Norah Oliver
Ivy Carter	In memory of John Mugford
Jenny and Mike Clarke	In memory of John Mugford

Donations in Memory

John and Joy Hehir	In memory of Norah Oliver
Karen Stocks	In memory of Norah Oliver
Keith Weller	In memory of Lillian Weller
Lyn Holmes	In memory of Ann Goddard
Mary Doyle	In memory of Norah Oliver
MF and LM Durke	In memory of Corinne Freeder
Mick and Mel	In memory of John Mugford
Mr and Mrs Clarke	In memory of John Mugford
Mr and Mrs Fortnum	In memory of Norah Oliver
Mr and Mrs Keys	In memory of Ray Overton
Mr and Mrs Pearce	In memory of John Mugford
Pat and Keith Evans	In memory of John Mugford
Paul Harrison	In memory of John Mugford
P J Smith	In memory of Norah Oliver
Rebekah Lilley	In memory of Ray Overton
RE and SE Parker	In memory of Norah Oliver
Rob & Julie Dore	In memory of Edna Dore
Sharon Thomas	In memory of Corinne Freeder

Raising Funds - How You Can Help

Perhaps you could help raise funds to enable us to continue our work? Whether £5 or £500, all donations will be put to good use, providing information, equipment and support for all Alpha-1 patients.

In addition we aim to promote better awareness and understanding of the condition throughout the medical profession, support research and campaign for better services and treatment for Alpha-1 patients in the UK. **Please visit our Website for more information: www.alpha1.org.uk**

JustGiving™

You will raise more for Alpha-1 UK Support Group on **JustGiving**. It's easy (and completely free) to set up a fundraising page for your favourite charity. It only takes 60 seconds to get up and running.

You can write out your personal fundraising story, add photos and even video and colour to your page. Best of all, it's all incredibly simple to do giving you the best tools to make it easy to ask friends to sponsor you.

If you are a UK tax payer our charity can also claim back via Gift Aid the basic rate tax already paid on donations by the donor. This means we can claim back from the government on your behalf 25p for every £1 donated, boosting the value of the donation by a quarter.

You can also use your mobile to send a donation. Text "ALPH10 £amount to donate" to 70070 to donate to Alpha-1 UK Support Group.

JustTextGiving is powered by Vodafone.

justtextgiving.com

easyfundraising.org.uk

easyfundraising.org.uk is a great way to raise money for our charity just by shopping online.

1. Start at easyfundraising

Let's say you want to buy a pair of shoes from John Lewis.

Instead of going to johnlewis.com directly, you first go to easyfundraising.org.uk.

2. Make a purchase

From the easyfundraising website, click through to John Lewis to make your purchase. This tells John Lewis you came from easyfundraising. The price of the shoes is exactly the same.

3. Get a donation

After you buy your shoes, John Lewis will give you a cash

reward that you can turn into a donation for your good cause. easyfundraising collect this and send it on at no extra cost.

4. Get the easyfundraising Donation Reminder

You can skip steps 1 and 2 with the easyfundraising Donation Reminder. Just click the Reminder when you shop to receive any eligible donations. You'll never forget a free donation again!

easysearch.org.uk

easysearch.org.uk is a free search engine that enables you to raise funds for the good cause of your choice whenever you search the Web. It costs nothing - **easysearch** is completely free.

How does it work?

If **easysearch** is used as the search engine for a web search, **easysearch** will donate half a penny to the Alpha-1 UK Support Group for every search you make. This is an easy way to raise money, so please use easysearch.

Alpha-1 UK Support Group Merchandise

On our website we have a selection of Alpha-1 merchandise available for purchase, including T-Shirts, wristbands, trolley key rings, badges and Christmas cards.

Alpha-1 information packs, booklets and posters are also available at no cost.
Please e-mail us with your full name and address at:

info@alpha1.org.uk

We are happy to supply our materials to healthcare professionals.

CHRISTMAS CARDS for 2017

Here is our 2017 selection of Christmas Cards. Please order either in our online Shop: alpha1.org.uk or by email: info@alpha1.org.uk The cost is £3.75 for a pack of 10 (140mm/5" square).

Snow Santa

Gold Tree

Twelve Days of Christmas

Santa's Reindeer

Star Christmas Wishes

Trustees & Committee

Karen O'Hara
Chairman
Trustee,
Treasurer

Bev Burroughs
Trustee,
Admin Support

Jemma Coad
Trustee,
Fundraising
Awareness
Co-ordinator
Parent
Support

Dr Sandra Nestler-Parr
Trustee,
Strategy and
External
Relations

Mel Brolly
Fundraising
Awareness
Co-ordinator

Graham Dalton
Scottish
Representative

Fay Whittaker
Scottish
Representative

Patrons

Professor Robert A Stockley

Professor of Medicine at University Hospital Birmingham, Director of Lung Immunobiochemical Research Laboratory

Professor William MacNee

Professor of Respiratory and Environmental Medicine at University of Edinburgh, Honorary Consultant Physician at Lothian Health

Supporters

ADAPT

Alpha 1 Alliance

The InterNet Factory

CSL Behring
Biotherapies for Life™

British Lung Foundation

ALPHA-1 CANADA

alpha-1 GLOBAL

Boehringer Ingelheim

Almirall

ALPHA-1 FOUNDATION
Research for a Cure

BRITISH LIVER TRUST

Chiesi

KAMADA
High Quality Pharmaceuticals

Children's Liver Disease Foundation
fighting childhood liver disease

Supporting alphas, their families, carers and friends since 1997

Alpha-1 UK Support Group
24 Pelham Road
Droitwich
Worcestershire
WR9 8NT

We are a registered Charity

England and Wales (1146330)

Scotland (SC043177)

www.alpha1.org.uk

info@alpha1.org.uk

Who are we?

The Alpha-1 UK Support Group is a not for profit organisation and registered charity founded in 1997 by those diagnosed with the genetic condition Alpha-1 Antitrypsin Deficiency who are dedicated to help, advise and support fellow sufferers, their families, carers and friends.

What are our objectives?

To relieve the needs of individuals suffering from the genetic condition Alpha-1 Antitrypsin Deficiency (AATD), their families, carers and friends, in particular, but not exclusively by:

- Providing advice, support and equipment with a view to improve the quality of life of those suffering with AATD, their families, carers and friends;
- Advancing awareness and knowledge of AATD, in particular among affected patients and medical professions and healthcare provider organisations;
- Fostering improvements in access and equality of access to clinical expertise and optimal disease management and treatments for AATD;
- Supporting initiatives aimed at development, introduction and widespread adoption of novel therapies for AATD, including research.
- Working collaboratively with our members and relevant external individuals, groups of individuals, organisations and institutions in order to achieve a) - d).

What is Alpha-1 Antitrypsin Deficiency?

Alpha-1 Antitrypsin Deficiency also known as Alpha-1, A1AD or AATD is an inherited, genetic condition that is passed on from generation to generation. As the name suggests it is a deficiency of Alpha-1 antitrypsin (AAT) in the bloodstream. AAT is an enzyme produced in the liver to help protect the tissues of the body during infections. The low level of AAT in the blood occurs because the AAT is abnormal and cannot be released from the liver at the normal rate. This leads to a build up of abnormal AAT in the liver that can cause liver disease and a decrease of AAT in the blood can lead to lung disease.

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