Alpha-1 UK Support Group Newsletter

Issue 18
Autumn 2019

Welcome....

...to our Autumn 2019 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

"Another year over, and what have we done..."

By the Trustees (with a nod to John Lennon)

Having reached 'maturity' last year, what has happened in our 22nd year?

In order to capitalise on unique opportunities that arose from last year's annual meeting which gave Alpha-1 a national and public platform, we postponed our plan to give the charity a new look to this coming year.

The focal point of our annual meeting in Coventry last year, which was very well attended, was a stimulating, informative debate and 'Question Time' with a distinguished panel: two local Parliamentarians, Mark Pawsey MP (Rugby), a longstanding supporter of our charity and the Alpha-1 cause, and Matt Western (Warwick & Leamington) who has an expressed interest in healthcare issues; the Chief Executive Officer of University Hospitals Coventry and Warwickshire NHS Trust where one of the few Alpha-1 Specialist NHS Services are provided; Alpha-1 clinicians and industry representatives.

Both MPs pledged their continuing support for our efforts to obtain better access to care and effective treatments. Our meeting was 'live-streamed' and received much attention and positive feedback from both patients and MPs with constituents who are Alpha-1 patients.

The momentum generated by our meeting, together with Mark Pawsey's persistent and tenacious political pressure, resulted in securing the first ever cross-party Parliamentary Debate on Alpha-1 treatment, which was held in Westminster on 31st October 2019. During the debate, Mark, Matt and other MPs pressed the government on the following key issues: to ensure that the nationally commissioned NHS Highly Specialised Service for Alpha-1, which was approved in 2017 and has been budgeted for, becomes operational in a timely manner; that patients should be involved at all stages in the development and implementation of this service; and that the ongoing NICE evaluation of Respreeza®, the only Alpha-1 augmentation therapy currently

"Another year over, and what have we done..."
(continued)

licensed in the UK, is finally concluded. Alpha-1 patients, representatives of our charity and expert clinicians attended the debate and met with MPs and Nigel Adams, the Minister representing the Department of Health. The complete official report of the debate, which we recommend you read, can be easily found via this link https://hansard.parliament.uk/commons/2018-10-31/debates/D79EB4FC-284C-4B5F-8B68-0BFAAB302325/Alpha-1AntitrypsinDeficiency .

To our great disappointment, no discussions with NHS England and the Alpha-1 UK Support Group have yet taken place, despite the Department of Health's assurance during the Parliamentary Debate that there would be patient involvement in the development of the NHS Highly Specialised Service for Alpha-1. We will be pursuing this matter with NHS England.

In addition, final NICE guidance on the routine use of Respreeza® in the NHS was due in July this year but has still not been published. NICE has not been able to share with us the reasons for this delay or information on when we can expect the final recommendation to be issued. Whilst this is frustrating, it is also of concern to us that discussions with the manufacturer of Respreeza® may be underway with the aim of limiting the size of the eligible population for treatment with this therapy. You can rest assured that we will be seeking clarification and involvement in any such decisions.

We are very pleased that last year has seen significant clinical trial activity to study the safety and effectiveness of potential new treatments for Alpha-1, sponsored by Alnylam Pharmaceuticals, Arrowhead Pharmaceuticals and Mereo BioPharma. The UK remains one of the most active countries in Alpha-1 clinical research. It remains important for patients to continue to support the development of potential new therapies, and we encourage every patient to participate in clinical research, after being fully informed about the objectives of the study and carefully considering the potential benefits and disadvantages. Of course, not all research is aimed at identifying and testing potential novel treatments or may be of direct individual patient benefit, and anyone contemplating participation in research can contact the Alpha-1 UK Support Group for information and guidance.

Unfortunately, in the UK, there remains a significant imbalance between patient participation in clinical research activity and tangible therapeutic benefits arising for UK patients from this research. We have been working hard to shift this imbalance but a lot more needs to be done to achieve this, and we need your support! We will be conducting a national survey of Alpha-1 patients in the next year that will be designed to understand what patients in the UK want and value, in terms of clinical care in the NHS and clinical research. The results from this survey will help us to continue representing your interests as the landscape for Alpha-1 and healthcare in general changes. Keep an eye on our website for more information.

Sadly, this year we have lost more treasured members whose lives were shortened by Alpha-1, in many cases without them ever having smoked and none of whom had access to augmentation therapy that might have otherwise have changed the course of their disease, their lives and those of their families. They will all be deeply missed but not forgotten. We celebrate their lives and remember them and their loved ones in the obituaries.

We are lucky to have had keen fundraisers amongst our members and supporters, thanks to whom we have again managed to sustain a good income. Some of our members undertook amazing feats in order to raise funds for the charity and we celebrate their successes in this Newsletter.

We have again received generous donations from members and supporters and, occasionally, grants from pharmaceutical companies. One of the most valuable gifts we receive is the time that members and supporters commit to the charity, without which we would not be able to operate and work on behalf of the UK's Alpha-1 patient community. We would like to thank all of our members, donors, fundraisers and sponsors for their incredible and continuing support.

We welcome new members and new readers – all of our previous years' Newsletters are available on our website and provide valuable information for 'newcomers' about Alpha-1, the disease, the people, latest developments and the work of the charity.

Please contact us if you wish to become a trustee or want to support the charity in other ways – many hands make light work, and we need your help. Remember, the Alpha-1 UK Support Group is your charity!

Board of Trustees, Alpha-1 UK Support Group

Red & White Phoenix, The Adventures of a Hessle Road Lad

By the Trustees

It was our pleasure to welcome Keith Pollard to our annual meeting in September 2019. Keith kindly spoke to us about his book *Red and White Phoenix, The adventures of a Hessle Road Lad*. Keith was inspired to write this book after the sad loss of his son, Jason, to Alpha-1 in 2012. Keith wants his book to increase awareness of alpha-1 antitrypsin deficiency (AATD) as he had found how little was known about the condition at the time when his son was diagnosed.

Keith also feels that his son's journey is typical for many Alphas with liver problems that are incorrectly attributed to alcohol abuse rather than to Alpha-1. Being stigmatised as a heavy drinker whilst the real cause of the increasing health problems is not being identified is a deeply traumatising experience. Looking back at his family history, Keith found that a number of his relatives had respiratory problems and passed away at a fairly young age. Alpha-1 being a genetic condition makes clustering of affected people within one family much more likely which adds to the burden that affected patients suffer.

Keith wants to draw attention to these issues common to many Alphas and one of the key motivations for Keith to write this book is that he didn't want other people to endure the same experiences as he and his family.

Red and White Phoenix will be of interest to many Alphas, and the book's sales have already raised in excess of £3,000. Keith has kindly donated all of his book's proceeds to the Alpha-1 UK Support Group for which we are very grateful.

How to get involved and stay informed?

Join the Alpha-1 UK Support Group. Joining the support group is easy. Just visit our website: www.alpha1.org.uk, click the "Join Here" button and follow the "Email Forum" directions. There is also a link to our closed Facebook group, which we invite you to join.

- By joining our group, you will be able to connect with a large community of Alpha-1 patients, their families and carers.
- You can exchange and discuss your experiences with AATD and get advice and support from other members.
- We will keep you informed of the latest news and all relevant information about Alpha-1, including new clinical trials and how you could get considered to participate in them.
- We will regularly offer lifestyle tips to help you to keep fit and active.

If you don't have access to the Internet, then you can contact us directly at the address on the back page of this Newsletter. We can then arrange to send you an information pack and our annual Newsletter.

Perhaps you could help raise funds to enable us to continue our work? Whether small or large, all donations will be put to good use, providing information, equipment and support to the UK Alpha-1 patient community. In addition, we aim to promote awareness and understanding of AATD and related diseases in the medical community.

Details of how to donate, or how to find us on *JustGiving*, *Virgin Money Giving,* or *Facebook* can be found on page 24 of this Newsletter, or visit our website www.alpha1.org.uk.

It is also possible to raise funds without any cost to yourself, by using the *Easyfundraising* or *AmazonSmile* websites to do your online shopping, or the *Easysearch* website for your internet searches. Again, further information can be found on page 24 of this Newsletter, or please visit our website and follow the directions on the homepage to sign up to either of these free services.

The members of our charity face many issues that impact on the quality of their lives and the lives of their families. Alpha-1 UK Support Group is committed to addressing these issues, improving access and equality in access to adequate clinical expertise and effective treatments and to bringing about a well informed and responsive community. We have been supporting Alpha-1 patients for over twenty years and we are backed by the UK's leading experts and researchers in the field of AATD.

For more information about the Alpha-1 UK Support Group, or to help us with our work then please visit our website: www.alpha1.org.uk

Could you be an Alpha?

A guide to alpha1-antitrypsin deficiency for patients and their families

Could your lung or liver problems be caused by a hereditary condition called alpha1-antitrypsin deficiency?

Alpha-1 antitrypsin deficiency is a hereditary condition, which may lead to lung or liver problems that can significantly affect your health. For the sake of convenience, the condition is often called 'Alpha-1' or 'AATD' and patients sometimes refer to themselves as 'Alphas'.

In the earliest stages of AATD, people are commonly diagnosed as having asthma or COPD and may be treated for these conditions for many years before the correct diagnosis is made. However, it is important to know you are an 'Alpha' as early as possible so that you can take steps to protect yourself and to get treatment from an expert in AATD.

Alpha-1 antitrypsin (AAT) is a protein that is produced in the liver and, in healthy people, it is released into the blood circulation so that it can protect the body, from the damaging effects of inflammation. The protein that is produced by Alphas does not function properly and gets trapped in the liver. This can cause damage to the liver and, because the protein can't reach the circulation, the lungs lack the protection they need from the damaging effects of pollutants (particularly cigarette smoke) and infections.

Although the lungs and, to a lesser extent, the liver are the most commonly affected organs in AATD, there are some rarer complications that can lead to problems with the skin, kidneys and pancreas.

Being an Alpha means that you have inherited a tendency to develop these problems, so it is important to know what this means for you and for your family.

How does AATD affect the lungs?

It is important for the body to have a defence system against attack from pollutants and infections but these defence systems can also cause damage to bodily tissues through inflammation. Under normal circumstances, this 'collateral damage' is kept in check by AAT. However, in Alphas, the function and the amount of available AAT is reduced, with the result that the unchecked inflammation can cause serious structural damage to tissues. The lungs are especially prone to this process, so that regular exposure to pollutants such as cigarette smoke, may lead to a type of emphysema that is more severe in Alphas who lack the level of protection from AAT than is seen in people with normal AAT.

What are the symptoms of AATD-related lung disease?

The earliest symptoms of lung disease are often quite minor, even when the lungs may already have been significantly damaged. Cough, wheeze or breathlessness when undertaking physical activity are the commonest symptoms but may be mistakenly put down to things like lack of fitness or incorrect diagnoses such as asthma or COPD.

What is emphysema?

Emphysema is a lung disease caused by loss of tissue at the ends of the airway branches at the site where the lungs take up oxygen into the bloodstream. The loss of tissue causes the lung structure to change in a way that makes the airways floppy and collapsible, and the lung's microscopic blood vessels disappear. Breathing tests show that lung function is adversely affected by these changes and deteriorates over time. Over the course of many years, cumulative tissue damage can lead to the lungs being unable to maintain their function even under circumstances of maximum supportive treatment.

How does AATD affect the liver?

Although the AAT protein that Alphas produce gets stuck and tends to accumulate in the liver, this does not usually cause many health problems.

In some patients, however, this causes damage to the liver which can lead to liver disease.

At birth, the build-up of AAT in the liver of Alphas can lead to yellow jaundice and, rarely, severe inflammation in the liver. It is not well understood why only some babies with AATD experience these problems. In later life, a minority of Alphas may experience symptoms of liver disease that arise from liver damage, such as the 'scarring' process known as cirrhosis.

Could you be an Alpha?
(continued)

When should AATD be suspected?

- When there is a family history of lung or liver disease
- When symptoms of asthma are present that do not fully respond to treatment (e.g. breathlessness, wheeze, cough and phlegm)
- When a diagnosis of COPD was made in early adulthood
- When unexplained lung or liver problems occur
- When breathing tests show that lung function deteriorates rapidly
- When liver function tests are abnormal

Who should be tested?

Organisations, such as the World Health Organisation (WHO), have made recommendations on testing for AATD. In general, testing is recommended in:

- Everyone with emphysema, chronic obstructive pulmonary disease (COPD), or chronic bronchitis
- People with bronchiectasis
- Newborns, children and adults with unexplained liver disease
- People with a family history of liver disease
- Blood relatives of a person diagnosed with Alpha-1
- Anyone with panniculitis, a skin disease

How is AATD testing done?

Testing for AATD is fairly simple and quick. It is usually done with a blood test. However, because AATD is an inherited condition, patients should always speak to their GP or their hospital doctor about the possibility of genetic testing for AATD if they think they may need it.

Genetic testing should not be undertaken lightly, as it is 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 test outcome.

Best practice therefore includes obtaining a referral from a GP, or specialist doctor if applicable, for genetic testing to be carried out. If your doctor thinks genetic testing for AATD may be appropriate, they should usually refer you for genetic counselling as well. It takes time and specialist expertise to be able to provide patients with an understandable and accurate explanation of the reasons for testing and the potential meaning of abnormal results, and the time to discuss 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.

For more information about testing for AATD, please refer to the relevant article in our 2017 Newsletter, also available at www.alpha1.org.uk.

What do the test results mean?

Genes contain instructions that tell our cells to make molecules called proteins, and we all have two copies for each gene – one from each parent. The AAT gene contains the code for the AAT protein. 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 AAT gene, with the Z or S versions being most common abnormal variants. It is generally the case that both inherited copies of the AAT gene have to be an abnormal variant in order to cause a significant risk to health.

AATD can be inherited from Alphas (people with two abnormal AAT genes) or 'carriers' (people with one normal and one abnormal AAT gene). The figure below shows what could happen for the children of parents who are both carriers (genotype MZ); when both inherited genes are M, this results in normal AAT protein levels but when both genes are Z, the level of AAT is low to undetectable.

There are many combinations of different AAT genes, but the commonest 'genotypes' are as follows:

Normal (MM)

- Does not have the disorder; does not carry any abnormal AAT genes.

Could you be an Alpha?
(continued)

Carrier (MZ)

- Mild to moderate AAT Deficiency — may get disease symptoms and does carry an abnormal AAT gene.

Carrier (MS)

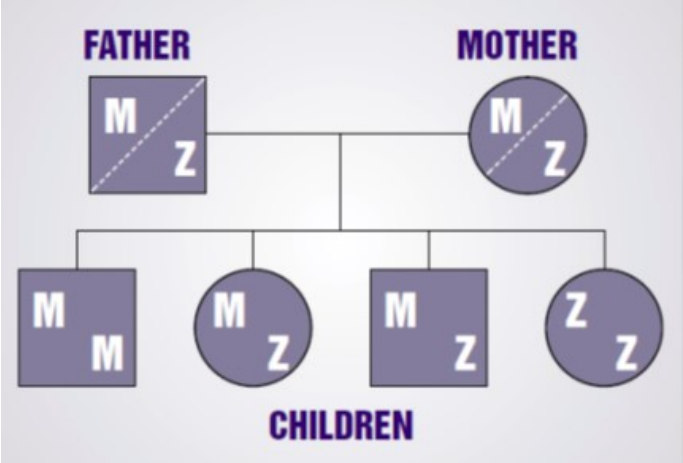
- It is unclear whether there is a risk for getting disease symptoms but does carry an abnormal AAT gene (most studies do not show an increased risk for disease).

Alpha-1 (SZ) or (ZZ)

- Moderate (SZ) to severe (ZZ) deficiency — could get disease and does carry two abnormal AAT genes.

Alpha-1 (SS)

- It is unclear whether there is a risk for getting disease symptoms but does carry two abnormal AAT genes (most studies do not show an increased risk for disease).



What does it mean to be a 'carrier'?

Carriers of AATD have one abnormal AAT gene and one normal gene, so that they have less functional AAT in the blood circulation than 'normal' people, but usually enough to prevent serious problems.

Being a carrier for AATD is not uncommon, and the vast majority of carriers are completely unaware that they have an abnormal gene and do not experience any AATD-related illness. However, carriers who have smoking-related COPD may be slightly more affected than patients with COPD who have two 'normal' AAT genes. It is therefore important for carriers to look after themselves and to avoid smoking or inhaling other pollutants. It is also sensible to avoid alcohol in order to minimise damage to the liver.

If I have AATD, how might this affect my children?

If you have AATD (two abnormal gene variants), it means that both of your AAT genes are faulty and that your children will inherit one of these. Your children will then be 'carriers' of this faulty gene. If your partner also has a faulty gene, there is then a chance of your children inheriting two faulty genes, i.e. one from you and one from your partner, that *could* lead to severe AATD. This is the reason why many partners of Alphas choose to have a genetic blood test themselves, to find out if they may also have a faulty gene.

If your child has inherited two abnormal AAT genes, this might lead to various lung and liver problems (as well as other rarer conditions), although it is impossible to predict whether, when and how your child's health might be affected by AATD. There are many Alphas with two faulty AATD genes who never experience serious health problems throughout their lives. In addition, 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 might experience ill-health, if at all. It is therefore important to receive genetic counselling and correct guidance and clinical advice in order to fully understand the implications of getting your child tested for AATD and the potential risks that AATD poses for their health.

How is AATD treated?

Symptoms are treatable using inhaled medications developed for use in asthma and COPD but these are not believed to significantly affect the underlying damage to the lungs. There are now many different types of inhalers and many different drugs for inhalation. The choice of what is best for individual patients depends on many factors, including personal preference for ease of use. Chest infections are thought to worsen lung damage and so it is always important to get these treated promptly, usually with antibiotics and oral steroids. Also, vaccinations against 'flu' (every year) and pneumonia (every five years) should not be forgotten because they reduce the chances and the severity of

Could you be an Alpha?
(continued)

chest infections. Your GP's surgery should arrange for these to be performed.

Replacement of the missing AAT protein is the logical treatment for AATD and is currently available as an intravenous preparation ('augmentation therapy') in some countries, including the US, Germany and Spain. Although an intravenous augmentation therapy product (Respreeza®) was licensed for use in the UK in 2015, it is currently not available on the NHS. NICE, the National Institute for Health and Care Excellence, is currently evaluating the effectiveness and the cost-effectiveness of Respreeza® and we are eagerly awaiting NICE's recommendation for or against the use of Respreeza® in the NHS. The Alpha-1 UK Support Group has been involved and has been representing the Alpha-1 patient voice in this NICE evaluation process.

A number of new treatment approaches to treatment are currently under investigation but, unfortunately, there are still no specific treatments for AATD available in the UK. For more information about ongoing research into AATD, please refer to our annual Newsletters and information on our website, available online at www.alpha1.org.uk.

What does a diagnosis of AATD mean for my future health?

A diagnosis of AATD can be unexpected and feel like a shock. For some people, the discovery of AATD may be by chance and not combined with any health-related problems. For other Alphas, it may come as a relief at the end of a frustrating journey of unexplained symptoms, worsening health and consultations with many different doctors, to finally obtain a clear diagnosis. The variability of the condition across patients means that your own experience is likely to be individual to you. However, what all Alphas have in common is the opportunity to take steps to avoid lifestyles that are known to pose a risk.

Even if you have developed symptoms, appropriate medication, treatments, exercise, and lifestyle choices can go a long way toward ensuring you live a long and happy life. Seeking care from an AATD expert will provide you with accurate information about AATD, about your own health, the best treatments and how to take good care of yourself.

What options are available to me?

It will help you if you develop an effective relationship with your family doctor and your consultants. However, it is important to recognise that AATD is just one amongst thousands of rare diseases, and most GPs and many specialists may not have much first-hand knowledge of the condition.

Expert knowledge and experience in the optimal management of AATD is therefore limited to a handful of clinical experts in the UK. You could ask your GP to refer you to one of the expert centres for AATD in the UK: Cambridge, Coventry, Birmingham and London. In the last year, a satellite clinic has been established in Exeter and we are hoping to see further satellites in the North in the future.

Contact information about existing centres and regular updates from existing and new centres are available on our website.

In addition, you can take a number of steps to help yourself:

- ✓ Create an exercise programme (under medical supervision)
- ✓ Create a nutrition programme (under medical supervision)
- ✓ Think about your health behaviour and life style (smoking, alcohol use and excess weight)
- ✓ Avoid risk factors:
  - Stop smoking and avoid second-hand smoke as much as possible
  - Avoid being around dust and fumes
- ✓ Discuss with your doctor who in your family should be informed and encourage anyone who might be affected to speak to their doctor about getting tested for AATD

Researchers around the world are studying Alpha-1 and learning more all the time. Visit the Alpha-1 UK Support Group website regularly for the latest news and advice on managing your Alpha-1.

You can find us at: www.alpha1.org.uk

An Athlete's Journey

By Lindsay Jarrett

For those of you who don't know me, my name is Lindsay, I am forty-eight years old and I live in Scotland. I have never smoked and have always been athletic.

When I was about thirty-seven, I was diagnosed with alpha-1 antitrypsin deficiency with a PiZZ phenotype, and I was on oxygen up until this year. I was a sponsored disabled athlete, with my sports being rock and ice climbing. Despite latterly being reduced to around 15-17% lung capacity, I was still able to continue with my climbing, whilst on supplementary oxygen, right up until January 2019.

I was referred to a hospital in England for lung transplantation assessment, as Scotland has no lung transplantation centres. I regularly attended this hospital for about eight years and, in all this time, I was never placed on the transplant register in the past. The reasons varied from year to year. Initially I was too well, which is the best reason of all, of course. Latterly, the reasoning changed somewhat, and I was told that I would never be suitable for lung transplantation.

Fortunately for me, the professor who is my consultant doctor in Scotland is also a professor at the Royal Brompton Hospital in London. He contacted a colleague of his at Harefield Hospital and told him all about me. Consequently, I received an assessment appointment within six weeks.

The night before the appointment I was in pieces. I did not want to go for fear that it was the start of another long, hard, frustrating and fruitless wait, as I had experienced before. Needless to say, that I did go to have the assessment; the staff were incredible, and I was so relieved. That was in December 2017. After the usual tests I was invited back in March 2018 and was placed on the register for lung transplantation.

Although this was excellent news on the one hand, it came as a shock on the other. I was a nervous wreck! You think you know how you will react when you are told that you have made it to the transplant waiting list, but really, I had not expected to feel so scared! I knew I would die without a transplantation, but I had gotten used to sitting on death row as it were. Now I was waiting to potentially put myself in the position of dying sooner, should lungs become available and the operation not be successful! Aaaaargh!

I maintained my fitness and health as best as I could so that if or when the time came, I would be fit enough for transplantation. My consultants in Scotland were extremely proud of how I was maintaining my health and often told me how incredible they found my physical capability, despite the obvious deterioration of my lungs. I truly believe that my mindset was the key to this survival and ability.

Nowadays, people love sharing positive messages, pictures and symbols on Facebook which, in my opinion are shared too frequently. These communications fail to inspire me anymore as many people who share them live anything but positive lives. I believe that actions are "where it is at". It is very easy to become swamped in ill health, in frustrations and inadequacies. In a blink of an eye we can feel worthless, useless, not to mention breathless.

I chose to be positive. I chose the harder route I believe. Being a mother to five children, survival was the only option for me. Each day since diagnosis and subsequent listing on the transplantation register, I reprogrammed my brain into positive thinking. Sure, there were days when I really couldn't be bothered. It would have been less hassle just to wallow, but somehow, I dug deep and pushed through, day after day and month after month. The more exercise I did, the better I felt, both physically and mentally.

At 9 am on Sunday, January 27th, 2019 I got THE CALL. This was my first call for lung transplantation, and I was taken aback! I ran around the house shouting "I've had my call for lungs." The response was muted and a bit

An Athlete's Journey
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bizarre... I don't think anyone thought the day would ever come!

That was the signal to trigger the transport plan to take me from the Highlands of Scotland to London as soon as possible. I phoned my local consultant and my MP. Both had met with me previously to put in place the transport plan and funding for the trip, should this day ever arise. Both acted professionally, quickly and with compassion and support. A helicopter picked me up, and we flew to Glasgow where a medically equipped aeroplane then took me to Luton. There I was picked up by an ambulance and taken to Harefield Hospital. I had to go alone as my partner could not accompany me in the helicopter as there was not enough fuel for an additional passenger! He got in the car and drove for ten hours to Harefield. My best friend made her way to the airport in Inverness, flew to Gatwick and then travelled across to Harefield.

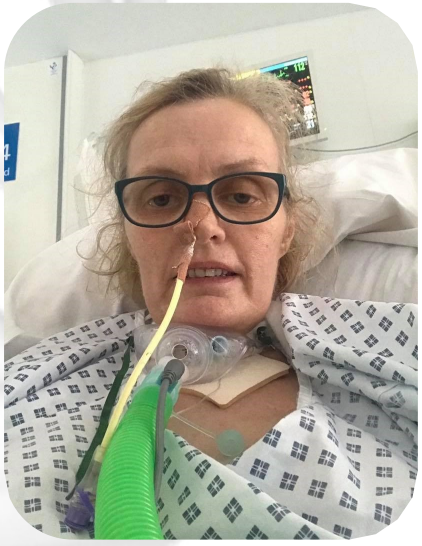
My partner made it to the hospital with fifteen minutes to spare before I was taken to theatre. The operation was much longer than expected, taking around fifteen hours. During the surgery, the surgeon had the task of scraping my lungs away from the chest wall as they had become affixed a couple of years previously. Apparently, I bled a lot during the operation and much of the surgeon's time was taken trying to deal with this. It took longer to remove my old lungs than to put the new ones in. I had what is called a bilateral single lung transplant where the two lungs are put in separately. The new left lung was put in first, but it did not work, and I had to be put on an Extra Corporeal Membrane Oxygenation (ECMO) machine to oxygenate my body. Eventually, both lungs were placed inside me successfully.

I was taken to the Intensive Care Unit where I was to stay for the next six weeks or more. Two days after transplantation I was taken back to theatre as there was a striation in my pulmonary artery. The connection between the donor organ and my heart was quite different in size and the left side of my heart had started to swell. I had no idea about any of this when it happened, as I was still under heavy sedation. In time, doctors tried to bring me round, but to no avail, as I had fallen into a coma. During this period, I fought off serious infections and kidney failure which resulted in receiving dialysis.

Eventually, a month after transplant, I sort of woke up. I was not coping with the intubation tube, so was given an emergency tracheotomy. That was the point at which it became obvious that I had lost my voice. My vocal cords had been damaged by the intubation tube. I am a chatterbox and I found this torture!! Hardly anyone was able to lip read and the alphabet board provided was a nightmare as I couldn't lift my hands to point at the board. It was like some bad joke! I could move my eyes, but that was about it!

I recalled lots of horrible dreams from my time in the coma too. Apparently, this is normal with all the drugs we are given. It took a psychologist to calm me down on a couple of occasions as the dreams and nightmares were so vivid once I had awoken. It was hard to tell what was real and what was not. I couldn't swallow either, so could not eat food. I was craving crazy things! Things that I had never even liked before!

My recovery was slow, very slow. However, once I got going, there was no stopping me.



I was transferred to the High Dependency Ward where I continued to work daily with the physiotherapists. Inability to keep food down after the tracheostomy tube was removed was a problem. I was so very thin and still struggle, to this day, to put on weight. Physically I was doing well, despite feeling very weak and noticing that I woke daily feeling very tight chested, almost as I had before the transplantation. Eventually, in April, three months after transplantation, I was allowed home.

The team keep a very close eye on me, with weekly, then fortnightly appointments.

An Athlete's Journey
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I had a stricture in the right bronchus, and I had to travel every two weeks to have a bronchoscopy and dilatation, during which the bronchus is stretched and gradually dilated. In total, I had six of these procedures. I last had one at the beginning of August but it's looking good and I don't have to return until September.

This summer I have canoed, rock climbed, camped and been away in my van. Where once there was a powered wheelchair in the rear, there is now a bed platform to convert it into a mini camper.

My journey has not been in the fast lane. Slow doesn't normally suit me but I had no choice as my pancreas, bowel and kidneys have all been under duress and I have ongoing issues with them. Maybe I was unlucky with some of the hurdles and complications I have been presented with. However, I can go about my daily life without needing supplementary oxygen for the first time in ten years.

I have been given the gift of life, but I had to fight tooth and nail for that life. My intention is to live it to the maximum and to do my donor proud.

The donor family have indicated that they are keen to find out how I am progressing. However, the transplant coordinator has recommended that I should wait until the first-year anniversary to contact them. I am conscious that this will be the first anniversary of their loved one's death and believe that this will be a particularly hard time for them.

I often think about what I might write to them. Of course, I want to thank them for their and their loved one's gift of life. However, I ask myself what thanks can I possibly give that would adequately convey my gratitude?

I think that to make the decision to share your loved one's organs, at a time when they are still alive, must be one of the most difficult things to do. For me, I think the best testimony I can give is to show my donor's family all the marvellous physical activities I have been able to do with my family and friends. I also want to thank them for enabling me to now look forward to the milestones in my children's lives, which I may never have done had they not gifted me their loved one's lungs.

My son Findlay, who is eleven, has asked me to write to them to let them know what it has meant to him. I imagine that will be an emotional read for me and for them, but it may also better and more fully convey our gratitude. Sometimes, children have the knack of keeping things simple, whereas adults can overthink or over complicate things.

I conclude my story by sharing with you an example of this clarity and simplicity. Lewis Capaldi's song, "Someone You Loved," is accompanied on YouTube by a very touching video, which I think portrays much of the reality surrounding organ donation. The haunting lyrics follow a grieving widower who meets a young woman who received organs from his much-loved wife. They share tender moments where they, and her young family, realise the momentous bonds they share. Her children present him with a drawing they have made. Written upon it are the simple words "Thank you for saving my Mummy."

Please watch this video. It is fitting and says everything I feel I want to say.

https://youtu.be/bCuhuePlP8o

Sarah Read, London Marathon, 28th April 2019

Written by Alpha-1 UK Support Group based on an account by Paul Carley

Paul Carley wrote telling us about his daughter Sarah who lives in Dubai, United Arab Emirates. Sarah obtained a non-charitable place in the Virgin Money London Marathon 2019 after completing the Dubai Marathon in 2018 and 2019. She readily agreed to her father's request that she run for our Alpha-1 UK Support Group, and, hopefully raise funds in support of our work, especially in the areas of research and potential treatments.

This is a cause close to her heart as Paul suffers from alpha-1 antitrypsin deficiency-related emphysema. He is at the stage where he needs ambulatory oxygen to walk when out and about. Unfortunately, Sarah's grandfather also died from this terrible disease, so she knows only too well what the future looks like for her father, without any specific treatment.

Following her decision to put her body through the torture of a marathon once again, Sarah carefully planned her training regime with a goal to complete the London Marathon in four hours. However, three weeks prior to flying over to the UK, she had a small accident resulting in a cut to her ankle. This required three stitches and meant that she was unable to complete some long distance training runs prior to running in London. As a result of the accident, Sarah's usual running style was affected and she was very nervous and had concerns about being able to complete an entire marathon. Despite that, she persevered, making the seven-hour flight from Dubai on the Thursday, arriving in London on Friday, just in time for the marathon on Sunday.

After a very early start on Sunday morning, Sarah ran her heart out and completed the hardest run of her life, the London Marathon, in four hours and twenty-one minutes, which is a remarkable time. On Monday, Sarah flew back to Dubai and on Tuesday she was back at work as a PE teacher – a 'flying visit' in many respects…

Paul described himself as an extremely proud Dad of his very own amazing Alpha-1 warrior. Donations were made via JustGiving and a large sum of money was raised by Sarah for this group, surpassing the fund-raising goal that she had set beforehand.

Sarah has since said that she would not run another full marathon, as the time required for training is considerable, especially while bringing up and supporting a family of three young children. However, Sarah has no regrets, especially as she knows how this rare disease has had a major effect on her family. Consequently, she is acutely aware of the need to develop effective treatments and, hopefully, a cure to save future generations from suffering from this debilitating and fatal genetic condition.

Paul has expressed thanks to all who supported Sarah and to the Alpha-1 UK Support Group.

We would like to echo his thanks to Sarah and congratulate her on this remarkable achievement and for having invested such a tremendous amount of time and effort – we are very grateful for Sarah's support of our charity.

Oliver's Fun Day

By Nicola Wright

Some kind friends of my late partner Simon and I decided to hold a Fun Day for our son Oliver, in memory of Simon and to raise awareness of Alpha-1.

Prior to Simon's diagnosis we had not heard of Alpha-1. Simon suffered from Crohn's Disease and had been poorly for some time. All his ailments were being blamed on the Crohn's. Then, after Simon collapsed unexplainably in October 2018, his Crohn's consultant sent him for further tests, which led to a diagnosis of liver disease. Crohn's Disease can result in liver damage, but the consultant pushed for more tests, which led to his very late Alpha-1 diagnosis. By this time, Simon was listed for liver transplantation, but one complication after another led to him being on and off the register.

He was very poorly at home and ended up in hospital with bowel problems in January 2019. Subsequently, he contracted pneumonia, and this affected his lungs and he ended up on a ventilator in the Intensive Care Unit. He did come off the ventilator and we prayed he would gain strength and get better. However, his body was just too weak, and his organs eventually failed. We lost our precious Simon on the 28th February 2019, the day after his 39th birthday. He left me and our 6-year-old son, Oliver, behind.

I felt completely lost and scared. With Simon being so poorly, me being back and forth to the hospital and taking our son to and from school, I hadn't had time, nor frankly even dared to begin to ask about or research Alpha-1. After Simon's passing though, realising it could now affect our young son, I decided I needed answers. Neither doctors nor even consultants had the information and knowledge I needed at the time, so that's when I reached out to the Alpha-1 UK Support Group. I found this organisation to be a fountain of knowledge, support and advice. I'm still learning and coming to terms with it all. Realising through the group that there was a lack of awareness for the condition, not only across the general public, but also within the clinical community, I asked if we could also use Oliver's Fun Day to help raise awareness of Alpha-1. Personally, I do not want my son to find it difficult to obtain information in the future, if needed, and I also felt the need to raise awareness generally.

On the 23rd June 2019, we held the Fun Day at the Lanchester Cricket Club in Durham. The weather was kind to us, and the day was a huge success all round. We had fabulous help from the Alpha-1 UK Support Group, who sent us posters, balloons, t-shirts, pens and magnets etc. for us to hand out at a stall where people could receive information about Alpha-1. Oliver had a great day celebrating his dad's memory. Additionally, we managed to raise awareness of Alpha-1 and raised a good sum of money to support the work of the charity. It was a fabulous day all round.

Top left: Nicola and Oliver. Top Right: Claire and Nicola. Middle: Andy and Nicola's brother-in-law Jonathan in the green. Bottom left: Oliver, Joseph Nicola's nephew, Nicola, Clodagh Nicola's sister and Hollie Oliver's best friend. Bottom right: Claire, Carol-Anne Nicola's sister, Danielle and Oliver

Oliver took pictures and a video into school and shared it with his class. Oliver's school also collected donations at a school event.

We will continue to do fundraising things for Alpha-1 in the future and thank everyone for their continuing and valuable support. We couldn't have done it without your help.

The Alpha-1 UK Support Group would like to sincerely thank Nicola and Oliver for their contribution to raising awareness of Alpha-1 in memory of Simon, particularly after their recent loss. The charity is very grateful for this support which allow us to continue our work for the Alpha-1 community and for fighting for better care and the development and access to effective medicines.

Clare Connolly, Chester Half Marathon, 19th May 2019

Written by Alpha-1 UK Support Group based on an account by Clare

Clare and Dave's third child James was born in January 2018. He was a beautiful, healthy baby and it was only after neonatal jaundice persisted for longer than two weeks, and he was referred for tests, that his parents began to realise that something may be wrong. After a stressful couple of months of countless tests and visits to the local hospital, James was diagnosed, at the age of three months, with alpha-1 antitrypsin deficiency (AATD), PiZZ phenotype.

Dave, James, Emily, Clare and Alice

Following this diagnosis, Clare joined the Alpha-1 UK Support Group in April 2018 and found the education and support available in this group invaluable. In addition to the kind and helpful advice and support which she received, Clare also shared her experiences and supported other members of the group with young children in a similar situation to James. Photographs of James which Clare posted to the group, show him to be a happy, bonny baby. James' parents and his big sisters, Emily and Alice, were tested for AATD and the results show that his siblings have PiMZ phenotypes.

Meanwhile, James, as a result of his prolonged jaundice, was immediately placed on medication to protect his liver from further damage caused by the excess bilirubin in his system. His parents were told that he may need a liver transplantation in the future (although this is only necessary in a small number of children with the condition.)

Over the next few months, James was regularly monitored, including attendance at the Leeds Specialist Children's Liver Unit. James was showing amazing signs of progress. The bilirubin was reducing, and ultrasounds showed his liver was normal. He was also putting on weight, something that James has always managed to do despite his condition. He was taken off the medication and is now monitored every six months to see if any further action needs to be taken. This came as a huge relief to James's family and they hope that he will continue with this healthy progress.

Through the support group, Clare was aware that AATD is a condition that can also affect the lungs. This is due to the lack of protective alpha-1 antitrypsin enzyme which is produced in the liver and transported through the body via blood circulation. In healthy individuals who have normal levels of alpha-1 antitrypsin, the enzyme protects the lungs against damage from inhaled pollutants and oxidative stress. However, many people with a deficiency in this enzyme, i.e. with AATD, suffer later in life with the lung diseases COPD and emphysema.

James' parents endeavour to protect him from environmental factors which may harm his lungs, keep him as healthy as possible and plan to educate him about his condition when he is old enough to understand.

Although 2018 was evidently a most challenging year for the Connolly family, Clare decided to do something positive to heighten awareness of AATD and made the decision to run in The Chester Half Marathon. She opened a fund-raising page with Virgin Money Giving and requested donations in support of the work of the Alpha-1 UK Support Group.

The half marathon was hard, but Clare completed it in a good time and raised a considerable sum of money for our charity.

In her message of thanks for all the support she received, Clare hopes that monies raised can further the work of the Alpha-1 UK Support Group in all aspects of its aims, including finding a cure for AATD.

The charity would like to congratulate Claire on her success of completing a Half Marathon, and express sincere gratitude for supporting the cause that we all feel very passionate about.

Evie's Story

By Karly Owens

I gave birth to my second beautiful daughter, Evie on the 16th September 2015. For my partner Simon and I, it was one of the happiest days of our lives. Evie suffered from jaundice at birth but, in due time, we went home to her elder sister Amelia and started our life as a family of four.

When Evie was six weeks old, I took her to the clinic where the Health Visitor advised me to consult my doctor as soon as possible as Evie was still jaundiced. The doctor sent us straight to the Royal Victoria Infirmary in Newcastle for bloods to be taken for testing. We were really scared as we didn't know what was going on.

When Evie's results came back, they showed that there was something wrong with her liver. We then spent a week in the Royal Victoria Infirmary. After numerous scans and investigations, one of them came back suggesting she could have biliary atresia. Our poor baby was very ill, and we were grief stricken, as well as scared. Simon still had to go to work and we had to leave Amelia with my Mam and Dad. It was all terribly stressful and upsetting. The doctors and nurses were great, but no amount of comfort could help us.

After being home a few days, we received a phone call advising us to go Leeds General Infirmary as Evie needed a small operation. It was there that we were told that Evie has alpha-1 antitrypsin deficiency. Simon and I were even more scared now, as we had never heard of it. Once again, the staff at the hospital were fantastic and explained everything to us. Also, through the Alpha-1 UK Support Group's website, I got in touch with Jemma Coad, the charity's Parent Support Coordinator.

We continued to travel between Leeds and Newcastle, seeing various doctors and having investigations, blood tests and scans. Then we received a telephone call asking us to attend the hospital. That is when we were told that Evie's liver was getting worse and that she would need a liver transplantation. It was one of the worst times of my life. Once again, I had to leave my eldest daughter with my Mam and Dad. Not only was I worrying about Evie, but I was also feeling guilty about leaving Amelia.

Every month, Evie continued to have blood tests. She wasn't gaining any weight, remained poorly and wasn't strong enough to have a transplantation. Consequently, she was given extra calories along with her milk to strengthen her. The doctors at Leeds decided eventually that Evie needed to be put on the transplant register. I had mixed feelings as I really wanted her to become fit and well but was worried about what she had to go through to get to that point.

We travelled to Leeds and spent a week there during which Evie had her transplant assessment. Once back in Newcastle, she had all her injections to prepare her for the transplantation. Then, we had to go back to Leeds to sign the consent forms. One week later, I received a phone call saying that they had a match for her. Simon was at work, so I had to ask my Mam to look after Amelia.

We were told the operation could last up to ten hours, so we went for a walk and had a cup of tea. Neither of us could concentrate as we were so worried. When I saw Evie after the operation, she had lots of tubes everywhere and I thought my heart would break.

She was at the Intensive Care Unit for three days, then she was transferred to a ward.

Evie had a remarkable recovery. The hospital staff were so pleased with her progress that we were allowed home after two weeks. Simon and our families were a great support throughout this very testing and stressful time.

We still had to go to Leeds every week for three months, then the visits became less frequent. The doctors were very happy with Evie and her new liver. We have had various health problems with Evie since, but nothing related to her liver.

Evie's Story
(Continued)

Evie is a real fighter. She attends nursery, plays with her sister and just enjoys life. We will be forever grateful to the liver donor and their family. Not only have they saved two lives, including my daughter's, but, they have also made so many people thankful and happy for Evie to live a happy and normal life. I understand that they had to lose someone they loved in order to make such a precious gift. Following Evie's transplantation, all our families are now on the donor register.

Thank you for reading Evie's story.

Jade Evans-Scott, Manchester Marathon, 7th April 2019
Written by Alpha-1 UK Support Group based on an account by Jade

For eighteen months, Jade's little boy Hugo was investigated for unexplained liver disease; many tests and examinations later, when Hugo was about two years of age, there was finally an answer. Hugo received a diagnosis of alpha-1 antitrypsin deficiency (AATD), with a PiZZ phenotype. At that stage, his family were very concerned about any irreversible liver damage that might already have occurred and the not knowing of what the future may hold for him. However, eight years later Hugo is doing very well; he is happy and healthy. Currently he has annual blood tests, scans and monitoring by his liver consultant and regular lung function tests.

Jade has stated that during those particularly stressful years, the Alpha-1 UK Support Group was amazing, with offers of reassurance, support and education. She appreciates the wealth of knowledge in the charity and support with signposting to specialist centres in and outside the UK. Jade has also said that she appreciates that all money donated to and raised for the charity goes directly to supporting affected patients and their families, towards campaigning to improve care and access to available treatments as well as the much-needed research that is needed to find a cure.

Consequently, Jade made the decision to run in The Manchester Marathon in order to raise funds for the vital work of the Alpha-1 UK Support Group. She set up a JustGiving page and asked for support with either a donation, sharing the message with friends and family to help raise awareness for the disease and our charity, or any words of encouragement and support to help see her over the finish line. Donations were also given by Jade's three children and their cousin, donating £5 each of their own money. Jade applied herself to a disciplined and arduous training programme, spurred on by much support and her belief in the worthwhile reasons for doing it.

During the marathon, just before mile twenty, Jade again started to think hard about her reasons for putting herself through this, namely for Hugo and the Alpha-1 charity. At that exact moment music from Hugo's favourite band, Stereophonics, came blasting out of a speaker; a sure sign to Jade to get her head down and run!

Jade succeeded in running the marathon in four hours and thirty-two minutes, describing the experience as incredible and amazing. Contributory to this was the fact that the crowds attending the event were wonderfully supportive and, also, she was spurred on by the generous donations which had been given. Moreover, she was greeted at the finish line by her three children, husband, mum, dad and sister! Jade was reminded, at this point, of how lucky she is to have had such unbelievable support throughout.

The support from her family and friends and her own determination helped Jade raise a large sum in donations to the Alpha-1 UK Support Group. She is extremely grateful to have received such generous donations and support and acknowledges that without it, she would not have been afforded the opportunity to be able to give something back.

The Alpha-1 UK Support Group would like to congratulate Jade for her amazing achievement and would like to sincerely thank her for generously supporting the charity and the cause we work for.

In Memory of Robert Dore

Memories of a life well lived, by Julie Dore

18th May 1946 – 11th May 2019

When Karen asked me if I would write about Robert (Rob to most people and Bob to his oldest childhood friend) I wasn't sure I could do it so soon after his passing. However, when would be a good time to do this? It's still too raw to write on a personal or detailed level, but a brief account of how Rob handled things since diagnosis may help others and give them strength.

I can hear Rob now, saying "nobody will want to hear about me". I disagree, as I shared and witnessed the way Rob lived his life, despite the progressive limitations Alpha-1 put on him. I can honestly say I never once heard him complain, ask "why me", or ever give in, right up to the end.

Rob was perhaps extremely lucky in that, for years he was totally oblivious to the fact he was alpha-1 antitrypsin deficient. He had no symptoms and was rarely ill, even with just a cold. He never smoked and, although he liked a drink it was never excessive, just the odd whisky or two socially.

Sport was a big part of his life. He played and watched cricket and football, but Aston Villa Football Club was his big passion from the time his uncle took him to Villa Park at the age of eight. This passion he passed on and shared with our daughter Becky, who, like her dad, became a very good player. He championed the game for girls. I remember a conversation he had with the head at Becky's junior school in which he said that girls should be allowed both to play football at school and to join the team representing the school. His actual meaning was that Becky should be allowed to do this; except he had such a way with people that he said things in a way that it would seem like it was their idea. She soon became a respected player in that school team. He was so proud of her and relished clocking up the miles taking her to matches and training as she progressed to playing for Aston Villa Girls. Subsequently, she played for the Aston Villa Ladies' Team, as well as attending the Birmingham centre of excellence, even though it was run by Birmingham City FC, the Villa's arch rivals!

As time passed, Rob added snooker and golf to the list of sports he played, and he started watching ice hockey at Coventry. The latter didn't last long as it wasn't played with a ball.

It was while playing golf that he started to notice he was out of breath after completing eighteen holes. He just thought he was getting old or unfit. However, it didn't seem too long after this that he really struggled to complete just nine holes and his recovery was taking far longer than it should.

Finally, after my nagging him, he went to see his doctor. After rounds of seeing heart doctors and all their tests coming back negative, it was suggested, almost as a lightbulb moment, that it was perhaps a lung problem. He had just turned fifty-eight when the results came back that he had the genetic condition alpha-1 antitrypsin deficiency. A few weeks later it was confirmed that he had the worst phenotype i.e. he was PiZZ. The doctors said that not smoking, not being much of a drinker and keeping fit contributed to the lateness in symptom onset. From being a baby, Rob was told often that he was underweight. He tried eating more, but never put a pound on. The past few years it has been a daily battle just to maintain the low weight he was.

What happened next you will perhaps all recognise - panic, fear, lack of knowledge or information from anyone really about what alpha-1 antitrypsin deficiency is. That was me of course. Rob seemed quite calm about it, something he always appeared to be.

Again, we were lucky as our GP knew of the condition, having already a twelve-year-old liver affected patient with AATD. Rob attended the ADAPT research programme in Birmingham, and a general respiratory consultant at our local hospital managed Rob's care in the NHS. Before they would test Becky, who was seventeen years old at the time, she and her dad were offered genetic counselling by our GP.

It was around this time that I found a brochure about the Alpha-1 UK Support Group and promptly joined the charity. I spoke to the lovely John Mugford who sent me

In Memory of Robert Dore
(continued)

so much information in a language we could understand. At last there were people who understood, who knew some of the answers and could give good advice built from experience. It was just what we needed. We made a whole new group of friends. Special friends, some we have been able to meet and many others around the world we haven't met and probably never will, but they are cherished friends, nonetheless. Sadly, so many of those first friends we made, like Rob, have since passed.

As things progressed, and Rob was still in good health, his respiratory consultant suggested looking at the possibility of a lung transplantation and wrote to the team at Cambridge. Most of the testing at this stage was done at the local hospital and the results were then sent to Cambridge. They kept coming back saying more tests were required. This happened a few times before the team concluded that there was a risk, in their considered opinion, that his quality of life would be better without a transplant than after a transplant. This was due to the slight problem they found. Rob was sixty-three by this time and past the usual age at which most transplant teams will accept a patient. At ADAPT, they disagreed with the view of the Cambridge team and, despite his age, Rob was referred to the Birmingham transplant team to look at the previous test results. Unfortunately, in the end they came to the same conclusion as the team at Cambridge.

Not long after came the news that there were to be four NHS funded Alpha-1 Specialist Centres to open in the UK: two of the four right here on our own patch. Rob chose Coventry and our GP requested an appointment.

From the day of the first appointment to the day of Rob's passing, he had the very best care available and help and advice was always available at the end of the telephone. Also, they were always there in person for both of us, even in Rob's last few hours. I will never be able to thank enough the Consultant at Coventry and his team or our GP practice and one very special doctor there. Both went far beyond the normal care expected for Rob, especially in the last two and a half years.

So, with that I come back to the beginning and that we consider we have been lucky. Lucky that when Rob's symptoms started to really affect his quality of life and prevent him from doing most things, care was still there and support for me too. I wish everyone had the privilege of the same care as Rob had as, I'm convinced that without this, we would have lost Rob much sooner. I will always admire the strength he had to fight for so long. Only in those last couple of days of his life did I fully understand the full extent of strength and willpower he had, after seeing how advanced the deterioration of his lungs were on a CT scan.

Rob was the kindest, gentlest and most selfless person I know. I don't think the thought of "why me" ever entered his head. He was so patient and aimed to make everyone around him feel good. Rob was one of those people who started to tell you a story and draw you in with all the detail, only for you to realise in the last line that he was telling a joke. He loved listening to recordings of Tony Hancock or Morecombe and Wise and he would roar with laughter.

Our daughter Rebecca wrote a poem to her father which I think sums up her dad, my husband, so well.

Poetry Please

In memory of Rob Dore

Dad—A poem by Rebecca Dore

All my memories of times spent together
They're here in my heart forever
The Early years we spent on the stands in B6
Me, stood on a milk crate – so I could see the pitch.

Running in the back yard
Such energy you had
You always had time
And I'm so very glad

We listened to the Beatles
Elvis too
How I loved growing up
Sharing all this with you

And as I've grown older, I want you to know
Your values I've grown up with, will always show
I took it all in, I heard every word
You showed me how to laugh, be kind
To have a light heart

You were always there with a Listening ear
Your great advice would allay any fear

All of this I will miss dearly, but I shall take heart
In many ways we will never be apart
Be it hearing an old song, or at Villa Park
Wherever I am - you won't be far

Poetry Please

A Poem by Ronnie Owen

At times when Barb was feeling blue
We looked to friends we thought were true
But those sunshine pals were never there
In stormy weather they just don't care

Then in the local news on day
We saw an item to our dismay
It spoke of Mardi who had Alpha-1
And her bucket list she had begun

We finally got to speak I'm glad
As she told us of the friends she had
On a Facebook forum for alpha ones
Where others care and do not run

Well we joined these groups and I have to say
The love and support was there each day
We were never alone like we were before
They loved each other when their health was poor

Sad to say my Barbies gone
Now she's with Mardi and not with Ron
I'm really not alone at all
With Alpha-1 friends I still stand tall

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

Paul Lowe
November 2018

Michael Caudle
December 2018

Peter Moore
December 2018

Jim Quill
January 2019

Simon Wright
February 2019

Barbara Warner
April 2019

Rob Dore
May 2019

Janey Leary
June 2019

Mary Doyle
June 2019

Linda Parking
July 2019

Keith Evans
August 2019

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, partaking in tough mudders and lake swims, baking cakes and hosting coffee mornings and gala days, organising awareness stands, donating in memory of loved ones and special occasions, making regular donations and even donating their hair.

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.

Alpha-1 Warriors Land's End to John O'Groats Saturday 11th August 2018

Paul & Becca Seager's vision for the alpha-1 community to set off from Land's End and reach John O'Groats, covering 874 miles, in under 24 hours was smashed achieving a staggering 1,102 miles.

Donations continued to roll in for this amazing team effort. Every penny raised for this event, and more, will be donated for research with the aim to improve the lives of those affected by alpha-1. Thank you to everyone who helped to make this event a huge success.

Broadway United Reformed Church, Christmas Fair Saturday 17th November 2018

Every year the church holds their annual Christmas Fair supporting two charities.

Jane Ratcliffe asked the church elders to support the Alpha-1 UK Support Group in memory of her beloved husband Michael Ratcliffe's 10th anniversary (Sunrise 15th April 1967, Sunset 5th January 2009).

Karen with Jane holding a photograph of Michael

Karen O'Hara, our Chair, had the great honour of speaking to the church congregation about alpha-1 before officially opening the fair.

The cake stall was incredibly popular and they had sold out by the time we reached the stall.

Jane's mum and her sewing group also held a coffee morning to raise additional funds for the two nominated charities.

Fundraising and Awareness

Paula Brown & NatWest held a bake off and coffee morning. This amazing cake was individually raffled.

Chris Thompson completed the ‘Iron’ 3.8km Ullswater Epic Lakes Swim in support of Oliver

Jim Hunt made a donation in honour of his being elected Master of the Lodge for the 2nd time

Liam Harris ran the Endurancelife Gower Ultra, 35 miles of downland, woodland, cliff tops, sand dunes and world famous beaches.

Graham Dalton & Mark Worswick created Facebook Fundraisers to raise awareness and Alpha-1 and raise money for our Charity

Connor & Brooke completed a 5km Tough Mudder with 13 obstacles, anyone that knows them knows this was quite a challenge

Fay Whittaker and Ina Smith raise awareness, collect donations and sell merchandise at Kirkcaldy Victoria Hospital throughout the year

Jenny Walker and her 11 year old daughter Meg donated their hair to the Little Princess Trust and raised money for our Charity in memory of James Murphy

Tanya & Simon Jones celebrated 10 years of marriage and instead of presents donated to our Charity

Fundraising and Awareness

Stephanie Baillie
Elizabeth Bulley
Emma Churchman
Erin Coad
Alison Fitzherbert-Stewart
Sandra Nestler-Parr
Emma Norman
Helen Shaw
Elizabeth Siayn
Jade Taylor
Libby Ware

Donations

Anne	In lieu of Christmas Cards
Anonymous	Monthly donations
Anonymous	Spring Charity Conference donation
Anonymous	Donations
Derek Arnold	Donation
Diane and Mick Stobart	Monthly donations
Susan & Doddie Turner	Donation
Berenice White	Monthly donations
Andrew Willis	Donation

Donations in Memory

Anonymous	In memory of Stephen Hundtofte
Anonymous	In memory of Vera
Amanda Barnett	In memory of Stephen Hundtofte
Andria Ellis	In memory of Beryl Saunders
Friends & Family	In Memory of Robert Dore
Brian Saunders	In memory of Beryl Saunders

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**

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You can raise money for the Alpha-1 UK Support Group via JustGiving, Virgin Money Giving and Facebook. It's easy (and completely free) to set up a fundraiser 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.

easyfundraising.org.uk

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

1. Start at easyfundraising

Go to easyfundraising.org.uk

2. Search for a Cause

Enter ***alpha-1 uk support group***

3. Choose Support This Cause

Create your account with Facebook or an email address

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!

amazonsmile
You shop. Amazon gives.

Join Amazon Smile and raise funds for us every time you shop on Amazon.

https://smile.amazon.co.uk/ch/1146330-0

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.

Expedia.co.uk · TESCO direct · The National Lottery · GAP · Argos · Boots · ebay · LOVEFILM · DELL · amazon.co.uk · LAKELAND · GROUPON · John Lewis · Toys"R"Us · Viking · Boden · DEBENHAMS

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 2019

Here is our 2019 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
Co-ordinator,
Parent
Support

Dr Sandra Nestler-Parr
Trustee,
Strategy and
External
Relations

Tanya Jones
Trustee,
Social Media &
Data Protection
Officer

Julie Dore
Fundraising
Co-ordinator

Mel Brolly
Fundraising
Co-ordinator

Lindsay Jarrett
Lead Scotland

Fay Whittaker
Fundraising
Co-ordinator
Scotland

Patrons

Professor Robert A Stockley

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

Professor William MacNee

Consultant Physician, Emeritus Professor of Respiratory Medicine University of Edinburgh, Royal Infirmary of Edinburgh

Supporters

ADAPT

Alpha 1 Alliance

The InterNet Factory

CSL Behring
Biotherapies for Life™

British Lung Foundation

α1 ALPHA-1 CANADA

alpha-1 GLOBAL

Boehringer Ingelheim

Almirall

1 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:

a) Providing advice, support and equipment with a view to improve the quality of life of those suffering with AATD, their families, carers and friends;

b) Advancing awareness and knowledge of AATD, in particular among affected patients and medical professions and healthcare provider organisations;

c) Fostering improvements in access and equality of access to clinical expertise and optimal disease management and treatments for AATD;

d) Supporting initiatives aimed at development, introduction and widespread adoption of novel therapies for AATD, including research.

e) 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.

Find us on Facebook
alpha1uksupportgroup

YAHOO! Groups
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Alpha1UKSupport

www.alpha1.org.uk

info@alpha1.org.uk

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