Update on new amyloidosis treatment developments and international meetings

GlaxoSmithKline have committed to development of the new treatment for clearance of systemic amyloid deposits invented by Professor Sir Mark Pepys. In November 2015 he gave invited keynote lectures about the treatment at the First European Congress on Hereditary TTR Amyloidosis in Paris, the Network Conference of the German Consortium on Systemic AL Amyloidosis (GERAMY) in Heidelberg and the German Society of Renal Physicians annual meeting in Mannheim. He also presented the latest results of CPHPC plus anti-SAP antibody treatment at the American Society of Hematology 2015 annual meeting in Orlando, Florida. GlaxoSmithKline held an Advisory Board Meeting there for world leaders in clinical amyloidosis to discuss plans for the phase II study. Sir Mark explained his invention and exchanged views about how best to proceed.

In Orlando Dr Ashutosh Wechalekar also presented NAC study results showing that doxycycline treatment combined with chemotherapy significantly improves survival in advanced cardiac AL amyloidosis patients.

At the Paris TTR Amyloidosis meeting Professor Philip Hawkins talked on “Imaging amyloid” and Dr Julian Gilmore presented an “Overview on clinical trials for ATTR amyloidosis”, an “Analysis of disease progression in patients with transthyretin cardiac amyloidosis” and showed the results of the Phase II trial of revusiran, one of the new genetic therapies being developed for ATTR amyloidosis. David Hutt, NAC Lead Nuclear Medicine Technician, presented the NAC experience of diagnostic DPD scans and Thirusha Lane, the NAC Lead Clinical Research Nurse, reviewed quality of life in ATTR amyloidosis. Dr Marianna Fontana and Janet Gilbertson also presented their work.
About half of the UK population use complementary or alternative medicine (CAM) at some point in their lives. It’s likely that CAM use is similarly widespread amongst patients with amyloidosis although exact figures aren’t available.

Conventional medicine strives to use treatments that have been tested in as thorough a scientific manner as possible and have been shown to be safe and effective. This approach is called “evidence based medicine” and is the cornerstone of modern Western medicine. There are clear and universally accepted rules governing the steps required and the proofs of efficacy and safety needed before a treatment is approved by the regulatory authorities. The National Institute for Health and Care Excellence (NICE) also bases NHS guidance on a scientific approach. Treatment of AL amyloidosis with various chemotherapy drugs is an example of evidence based conventional medical treatment. The side effects have been well studied and understood and the benefits have been shown to outweigh the risks.

In contrast, the terms “complementary” and “alternative” medicines or CAMs, are used to cover a wide variety of treatments that are not evidence based. These treatments include herbal and vitamin supplements, traditional Chinese and Indian (Aryurvedic) medicines, acupuncture, homeopathy, naturopathy, meditation, hypnotherapy, aromatherapy, osteopathy, chiropractic and colonic irrigation. There isn’t a clearly agreed definition of CAMs but NHS Choices distinguishes between complementary and alternative treatments as follows:

- “Complementary” medicine: treatments are used alongside conventional evidence based treatments with the goal of helping patients to cope with illness and with the side effects of treatment.
- “Alternative” medicine: treatments are used instead of conventional treatments with the goal of actually treating the condition.

In a very few cases there may be some evidence that some of these therapies may be effective in treating certain specific illnesses. However there is no evidence that any CAMs have efficacy in treating amyloidosis.

Natural medicines can’t do any harm ... or can they?

Common misconceptions about CAMs include the belief that these treatments are “natural” and therefore harmless. However, the reality is more complicated.

Firstly, use of CAMs may be harmful if it causes patients to delay conventional medical diagnosis and/or treatment. Outcomes in amyloidosis are often better the earlier that the diagnosis is made and conventional treatment started. Anything that delays this can be detrimental. It would also be harmful if a patient with amyloidosis were to use alternative treatments instead of conventional treatments.

Secondly, CAM medicines may contain substances that harm the body directly, or substances that interact with conventional medicines in a way that may decrease their efficacy or cause harmful, toxic effects.

Conventional medicines are required by law to comply with very strict formulation and labelling requirements. There are no such regulations for alternative medicines. Different formulations of the same alternative medicine product may contain widely differing concentrations of active ingredients, or the label may not list all the active (and sometimes harmful) ingredients that they contain. For some herbal remedies (eg. Echinacea), several different species or different preparations from different
parts of the plant may be all sold under the same name although they contain widely varying active components.

Although CAM products may be marketed and promoted to consumers as “natural” or derived from a natural product, there are no legal requirements for this to be the case. Seemingly innocuous vitamin supplements and traditional Chinese and Indian (Aryurvedic) medicines have been found to be contaminated with dangerous substances such as mercury, iron and lead. So unless you have access to an analytical chemistry laboratory it’s hard to know with any certainty what you are really buying. You can’t rely on the label to be accurate or informative.

CAM products are not required by law to list possible harmful side effects on the label, which may lead people to perceive them as harmless. Some examples of severe and sometimes fatal side effects of alternative therapies include liver failure, bladder cancer, arsenic and lead poisoning from herbal supplements, hepatitis C transmission and pneumothoraces (collapsed lungs) from acupuncture, artery damage leading to stroke and paralysis or death from chiropractic manipulations and ruptured colon from colonic irrigation.

Of course conventional medicines also have side effects, which are sometimes severe. But pharmaceutical companies are required by law to research, document and list all possible side effects in the product information. Conventional medicines may only be prescribed by a qualified doctor who judges that the likely benefits of taking the medicine outweigh the risks. In contrast, CAMs are available over the counter, and as there are no legal requirements for the packaging to list the active ingredients or for the manufacturers to investigate and document the possible side effects, consumers have no idea of the dangers they may be incurring.

Here are some examples of specific pitfalls that patients with amyloidosis should avoid:

- Green tea and associated products have become popular in recent years as an alternative treatment for patients with cancer. It has been suggested that EGCG, an active component of green tea may have anti-amyloid effects. At present there is no evidence for such effects. But studies have suggested that green tea may block the activity of Velcade, an effective chemotherapy drug often prescribed for AL amyloidosis. Patients taking Velcade are therefore advised not to consume green tea.

- Patients taking Velcade are also advised to avoid taking vitamin C supplements because there is evidence that vitamin C may also block Velcade activity.

- St John’s Wort (Hypericum perforatum) should also be avoided with chemotherapy drugs such as Velcade, as it may alter exposure to these drugs unpredictably.

- Most herbal medicines and dietary supplements have not been studied in patients treated with chemotherapy so we don’t know what possible interactions may occur. It is therefore generally safer to avoid taking these products during chemotherapy.

- Garlic and cod liver oil may have anticoagulant effects which increases the risk of bleeding and may be harmful in large quantity.

- Patients taking warfarin should avoid garlic, ginger and turmeric supplements as these may increase bleeding risk.

- Beta carotene supplements may increase the risk of lung cancer.

- Kelp may interact with contrast agents containing iodine.

- Vitamin B6 can cause neuropathy, skin rashes and nausea if large amounts are taken for a long time.

- Therapy with combined high dose vitamin B6, B9 and B12 in patients with advanced diabetic kidney disease has been linked to worsening of kidney function. Patients with kidney disease due to amyloidosis should consult their doctors before taking such vitamins.

This is not a full list of all potentially problematic interactions so you should always discuss any alternative medicines you are thinking of taking with your doctors.

**Complementary therapies and quality of life**

Patients with amyloidosis may find that complementary therapies, used alongside conventional treatments make them feel better and help them to cope with their illness and with the treatment. Relaxation based therapies such as meditation, hypnotherapy, mindfulness based therapies, music therapy and massage are unlikely to cause harm as long as patients use them alongside, not instead of conventional therapies, and not in order to delay starting conventional treatment. These types of complementary therapies have not been studied in patients with amyloidosis but in patients with cancer they have been shown to help to improve mood and concentration, to reduce anxiety, enhance positive thinking and emotions and increase general feelings of wellbeing. All of these can contribute to improved quality of life.

It is important for patients with amyloidosis to be aware of the potential pitfalls of CAMs, and to consult with their doctors about all over the counter or alternative remedies they may be taking or considering taking.
Local Amyloidosis Patient Support Groups

Scotland Group (By George Turner, Scotland Group Organiser)

Following the first meeting of the Scotland Support Group in Glasgow in April with three members, the number of people now interested has grown. At our second meeting, again in Glasgow, in July, the group had grown to five. This was followed by our third and latest meeting at the Falkirk Wheel in October (see photo) when our numbers increased to 12. As well as amyloidosis patients, family members also attended.

Members came from Ayrshire, Argyll, Lanarkshire, Fife and Edinburgh. We have also had interest from more people from Edinburgh and Aberdeen. As the group is spreading out across Scotland, we try to arrange our meetings at a place and time to suit as many as possible.

As well as our support group, some of us go to the Myeloma UK meetings at the Maggies Centre at Gartnavel Hospital in Glasgow. They have been very welcoming and supportive to all of us, not just to AL amyloidosis patients.

From being totally unaware of amyloidosis before being diagnosed in May 2014, and never having met anyone else with amyloidosis except at the NAC, it’s been really helpful and a great support to share with other members of the group our experiences of coping and living with amyloidosis.

We are not alone!

Mark McConway, another founder member of the group, wrote on the patient forum after the meeting: “It’s been nice to have a meet-up in a non-medical environment for a coffee or bite of lunch. Although amyloidosis has brought us together, it’s not the only thing to talk about and I feel that’s important to remember for anyone else thinking of joining us – or for starting their own group. Support can mean just being in the company of others who’ve experienced what you’ve been through”.

For information about the Scotland Group, please contact George: geordie_turner@hotmail.com.

London Group

The London group had their first meeting December 2015 at the NAC. The meeting was small (intimate!), but the members found it very worthwhile and would like to encourage other patients and family members in the London area to join.

The next meeting will be on Tuesday, 22 March between 5 and 6.30 pm. The provisional venue will be the Garden Gate Pub, 14 South End Road, London NW3 2QE (outside if fine weather!). The pub is a couple of minutes walk down the hill from the Royal Free and only a minute away from Hampstead Heath overground station. There is street pay-parking nearby.

Please contact David if you are interested in attending the London Group meetings or have any questions: davidgoldstone@btinternet.com.

Myeloma UK Groups - for patients with AL amyloidosis

The charity Myeloma UK also welcomes patients with AL amyloidosis at its support groups, which meet in around 60 locations around the UK – for more information see the Myeloma UK website: www.myeloma.org.uk.
From the patient online forum

The patient online support forum at http://amyloidosis.org.uk/forum has grown and expanded steadily since it went online a year ago. There are now over 200 members and some threads have been viewed more than 3000 times. The forum is most useful in providing information, support and a feeling of community for people who feel isolated by having a rare disease that most people they meet haven’t even heard of.

Some members recently showed how adversity can also spark creativity and humour can be a powerful coping tool. Here are examples:

Amyloidosis limericks

John Campbell wrote: “A few weeks ago it was National Poetry Day and I was kept awake in the dark hours of the night with the odd stabs, burns and tings in my legs and feet, so to keep myself occupied I dreamt up some limericks (it is too serious a subject to be sombre). They may not be very good but they are my own and I hope you enjoy them”.

I suffer from amyloidosis,
there is no other diagnosis,
because proteins corrupted
cause neurons disrupted,
which gives me a pain in my toeses.

Amyloidosis gives me the creeps,
it is interfering with my sleeps.
Let’s hope for a cure,
it’s coming I’m sure,
developed by Sir Mark Pepys

My senses are getting hazier
caused by plasma cell dyscrasia.
My paraesthesia gets no easier
and no time off for good behaviour.

These posts inspired Pat Pinchin to share a rather more serious poem that her daughter, Chloe, wrote after she was diagnosed:

An unusual diagnosis.
A sobering prognosis.
A rare debilitating illness.
A ferocious pathosis.
Clinically precocious.
The answers are ambiguous.
Other people are incredulous.
All so emotionally divisive.
Longing for something decisive,
instead of just derisive
to help me out of this crisis.
Don’t have room to list
all the things in life I miss.
Has it really come to this?
Got to focus on the positive,
despite the suffering and the sickness.
A CURE for AMYLOIDOSIS
is my single biggest wish.

Another patient, Kathy Koontz, added her own limerick written after a bone marrow biopsy:

I've got a hole in my pelvis,
can't shake my hips like Elvis.
It hurts in my marrow,
I'm stiff like a scarecrow.
I'll take it so I can get past this.

Patient story: Pat Pinchin

“Thirteen years of amyloid mouth misery”

I had my dream job as the Modern Languages’ Consultant for Dorset Education Authority, when amyloidosis made me unable to continue. I live in Poole, with Patrick my wonderful husband who cares for me. I have a married son and daughter and four young grandchildren.

It all began in March 2003 when aged 55, I noticed a lumpy feeling around my upper right back teeth. My dentist checked for abnormalities but found nothing. Referral to a maxillo-facial consultant also led nowhere. I told him that in addition to the lumpiness, I had a powerful salty taste in the saliva flowing from my upper palate. “Stress”, he said, my jaw being rather stiff, and advised physiotherapy. My physiotherapist did observe abnormal hard lumps in my right cheek, which I now know was a marker. After an evening cinema visit, my husband exclaimed, “whatever have you done to your eyes?” In the mirror, I was astonished to see two enormous racoon-like deep purple bruises encircling my eyes. I had rubbed them! More unprovoked “eye episodes” ensued at intervals. Why?

In December 2004, my GP referred me to a haematologist “just in case” and said I should take along a photo showing my bruised eyes. Armed with my racoon eyed photo, I saw Dr Bell in 2005. From my GP’s referral letter and the photo, he concluded: “Your symptoms are a classic presentation of amyloidosis, a very rare, complex and serious disease. You need to have a blood test but in the meantime don’t search the internet because you will scare yourself”. Ignoring his “don’ts” I Googled amyloidosis. Aghast at the severity and incurability of the disease, I did not understand much of the terminology, and put it at the back of my mind. My blood test result showed that my free light chains were indeed elevated which together with a bone marrow biopsy, clinched a diagnosis of amyloidosis. Haematologist Dr Jack re-explained everything to me and Patrick. He said that I would be referred to the National Amyloidosis Centre (NAC) in London where I would have another blood test, echocardiogram, electrocardiogram and special SAP scan. The latter was the only scan in the world which could detect where masses of amyloid in the major organs had deposited. Dr Jack explained what amyloid is, what is meant by free light chains and the damage that amyloid could do to organ structure and function. I understood little, was dazed, and devastated. One positive was that I “was very lucky. Until about five years previously no effective treatment had existed for amyloidosis whereas now there was more hope”.

Patrick broke the news to our family who found it hard to take in, grappling with trying to understand the disease and what was happening to strong and healthy Mum. In an effort to turn negatives into positives, they all participated in challenging sporting fundraising activities for amyloidosis research.

At the NAC, to our great relief, my SAP scan proved that there was no amyloid involvement in my organs. Dr Offer diagnosed MGUS (monoclonal gammopathy of undetermined significance). So far not serious but as my abnormal protein was associated with systemic amyloidosis there could be a potentially serious outcome. His illustrated explanation was the best ever. I explained my unpleasant mouth symptoms and he recommended a head/neck CT scan and mouth biopsy. My maxillo-facial consultant agreed to the scan but as no abnormality was found he refused the biopsy. I re-emphasised the debilitating continuing salty taste in my palate; again to no avail. In 2007 a large lump on my upper arm was investigated by ultra-sound and diagnosed as an unusual lipoma. By 2008, the salty taste had become so overbearing that I had to retire. The arm lump had also increased in size. During another CT scan, a conscientious radiologist read my file and noted my amyloidosis diagnosis. She informed the consultant that the rather large hard lump could be amyloid. A biopsy confirmed this, as did biopsies of other lumps in my
breasts. I had further multiple lumps all over my upper body but none were troublesome. However the ones in the sub-mandibular glands “could be associated with the salty taste”. Although the last two investigations at the NAC still showed no organ involvement, blood test results were continuing to identify systemic AL amyloidosis.

Dr Jack decided that treatment was necessary to lower the excess of amyloid producing free light chains. He said that usually amyloid regression eventually follows. I began in 2008 with a three month course of dexamethasone. With no significant response, in 2009, I proceeded to four cycles of CDT but stopped the thalidomide after a rash during the first cycle. Again, no clonal response. On my next visit to the NAC, Dr Wechalekar, noting that my abnormal protein was IgM, the same as for lymphoma, prescribed the lymphoma chemotherapy R-CVP (Rituximab, Cyclophosphamide, Vincristine, Prednisolone). His diagnosis was nodular cutaneous amyloidosis (amyloid in the skin/soft tissue). Back at Poole, Dr Jack explained that he would have to make an application to the NHS Trust for me to have R-CVP. Unbelievably, it was that he would have to make an application to the NHS for the treatment was not weighty letter from Dr Wechalekar, the treatment was finally granted. I had eight cycles with a very good clonal response. Until now my FLCs have remained just about within the top end of normal.

Chemotherapy worsened the already powerful sore salty taste in my mouth. Nothing could quench my permanent thirst. Multiple amyloid lumps lined my cheeks and lips and I often had painful ulcers on my tongue. Decidedly I needed to find someone who could help me and I scoured the UK, consulting nine oral-maxillo specialists, including three London professors. All in vain. Diffuse amyloid deposits were impossible to remove surgically. A procedure would also be risky because amyloid is very vascular. I “was lucky not to have macroglossia” (amyloid swelling in the tongue). On reading a short article about amyloid involvement in the soft tissue, I discovered the reason for lack of improvement in my symptoms. The NAC confirmed that unlike in the internal organs, “amyloid in the soft tissue rarely changes over time”. I was in despair because I now knew it unlikely that I would get better. Others whose chemotherapy had resulted in some amyloid regression from organs were able to enjoy a better quality of life, but mine was deteriorating. My mouth symptoms were so overwhelming. Activity and conversation worsened them, so normal relationships with friends and family, social activities, days/evenings out, travel, cycling, gardening, holidays, everything I loved, became things of the past. My hard to endure symptoms are not understood so no-one can empathise. My doctors are sympathetic, but with my light chains stable and my disease non-life threatening I am of little concern.

A PET scan in 2013 requested by Dr Jack to check for a lurking lymphoma fortunately proved negative, but unknown to him, it did show very good imaging of “extensive deposits compatible with amyloid throughout the upper body and especially in the salivary glands”. At my 2014 NAC appointment Dr Wechalekar explained that amyloid had infiltrated my palatal nerves. At last, after 11 years with this distressing mouth misery I had an explanation. I read in a paper written by well respected amyloidosis specialists at The Mayo clinic in the USA, that patients (approximately 6%) with IgM related amyloidosis “have a higher prevalence of neuropathy”. What a revelation that was to me! My chronic nerve damage now made perfect sense, because “this suggests that the M protein may have a higher affinity for binding to nerve myelin”.

In 2015 the advice from the NAC was to pursue treatments for symptom control with a chronic pain consultant as there was nothing the Centre could offer me. Thanks to the efforts of Professor Sir Mark Pepys, I was referred to Dr Nandi, a leading UCL neurologist who unsurprisingly had never met a condition such as mine before. Like Dr Wechalekar he described my case as “unprecedented” in his experience. I had already tried a raft of drugs, including opioids, for neuropathic disorders but none provided effective relief. At Dr Nandi’s suggestion, and supervised by my local chronic pain consultant, Dr Hitchcock, I tried two infusions of Lidocaine, a powerful anaesthetic drug given for pain relief, but like many other drugs, it made my symptoms even worse. So many drugs, including those to reduce high blood pressure, further aggravate my chronically damaged palate nerves. I am now left with a combination regime of drugs used to treat neuropathic pain which I do not have! There is nothing for my specific disorder. The drugs provide minimal relief which is better than nothing. Even at the very low dose which I can just about tolerate, their side effects leave me fazed, lethargic, exhausted and affect my vision. I take a cocaine mouthwash which provides some benefit for 40 minutes three times per day. Worst of all, is that my very poor quality of unproductive life continues because my symptoms engulf me. I remain housebound except for gentle walks and medical appointments. My teeth decay repeatedly because of amyloid blocking the
normal flow of protective saliva. Chatting with family and friends is of necessity reduced, with a heart breaking emotional effect. I have to prevent myself from crying otherwise purple bruises encircle my eyes. I cannot be the wife and mother of my former self or the fun, active and avid story telling grandmother to my grandchildren I had always hoped to be.

I remain optimistic that the anti-SAP drug undergoing NAC/GSK clinical trials may become a reality and help me. Dr Hitchcock and Dr Nandi have told me that with the amyloid removed there are so many tiny nerves in the palate, that some of them may repair and symptom control may improve. In the meantime, my occupations have become uncharacteristically sedentary and solitary. Battling the relentlessly ferocious mouth symptoms is an isolating daily struggle. I try to stay cheerful and positive for the sake of my cherished family and live my life as best I can. Supporting the wonderful work of the NAC by helping with publicity for an inspirational fund-raiser has been therapeutic as is “meeting” other sufferers in the virtual world. The NAC forum has been of particular therapeutic support. Awareness of the disease is gathering momentum. My hope is that one day I may be well enough to be more actively involved in this. I urge all affected by amyloidosis to donate to fundraising appeals for the UCL Amyloidosis Research Fund at the NAC. Such funds free up other monies to be used for clinical work and research. We must support their earnest endeavours to bring us better treatments to improve the quality of life for all sufferers.

Fundraising News

For Don Prideaux (Father Extraordinaire)

By Ruth Prideaux

It never mattered what sport Dad was watching, he would always say “but look at the skill that must take”. He loved all sport, played badminton and tennis when he was a bit younger, moving onto golf when running around a court got a bit much and a sedate walk with a golf club a bit easier. He would watch any sport on the TV though and always admired the effort people put into honing their skills.

When Dad started to suffer from fatigue and dizziness he just assumed he was getting older, but after collapsing whilst we were on a family holiday, Mum insisted he go to the doctors and get checked out. It took about a year of tests and other procedures, and Dad getting weaker and weaker, until he was diagnosed with amyloidosis in June 2013. Despite his bravery and his determination, “I’m going to beat this thing kid, I’m not going anywhere”, he sadly passed away aged 69 on 28 August 2013 with all his family around him.

Dad was never one to dwell on sad times, he was such a kind soul, would talk to anyone (he could talk the hind leg off a donkey!!), and he always had a funny joke to tell that made us groan before he even started telling it. So, as his family, we of course mourned his passing, but then in his honour we got up, dusted ourselves off, and try to live life in his memory, laughing loudly and living every minute.

Losing Dad was not the only tragedy I have endured in quite a small space of time. I very nearly lost my five year old daughter to pneumonia in 2009 and then, four months later in 2010, I was diagnosed with breast cancer. They told me it had also spread to my lymph nodes which were removed and I went through a mastectomy, chemotherapy and radiotherapy and am still undergoing hormone treatment. In 2012 I had reconstructive surgery to rebuild my chest. To do this they stripped some of the muscle and skin from my back and wrapped it around my chest over an implant. This was successful but because they removed the muscle from my back I was told that I would always struggle to pull myself up things like ladders as I didn’t have my back muscle to brace me. Added to this, I also developed lymphoedema in my right arm due to having no lymph nodes and this means that I cannot lift heavy weights as my arm fills with lymph fluid and swells up quite alarmingly.

As part of my recover I started running so I could complete the ‘Race for Life’ to raise money, which I did in
I have continued to run ever since, up to half marathon distance, raising money for cancer research. Since I started running I had always toyed with the idea of running the WOLF Run (this stands for Woods, Obstacles, Lakes and Fields). It’s all set on a farm in the Warwickshire countryside and there are plenty of hills to run up and down, woods to run through, obstacles to get under, over and round, and lakes to swim, but I knew that I could never get round the course on my own as I would really struggle to pull myself over the obstacles.

My Brother Neil agreed to run it with me. We did some practice runs together to build up our distance and Neil lifted some weights in an effort to prepare himself to get me over the obstacles (there would never be enough time to train for this!!). The course is over six miles and, now I have completed it, I can confirm that there are a lot (and I mean a lot!!) of obstacles.

The day dawned cloudy and windy. Mum drove my brother and I to a remote farm in the middle of Warwickshire. We registered, had our numbers scrawled on our faces, and prepared to be called for our run time. Once we had been called into our group we were then helpfully warmed up by some army chaps, who proceeded to make us jump up and down for 10 minutes completing various exercises. I don’t mind telling you that at the end of that I could have done with a sit down, but then the 10 second countdown began, the horn blew, and we were off on the 6 mile trek.

It started off as really good fun. We were wading through water up to our chests and crawling through mud pits, and Neil helped me climb the walls and really big obstacles that I just wouldn’t have got over without him.

I have to admit that it was really tiring. I can road run six miles no problem, but it’s very different when it’s over rough terrain and you are clambering up, over and through things every 200 yards, but the worst bit was the swim. The organisers do advise that you have to be a strong swimmer to complete the swim. They tell you it’s cold and very draining so they give you an alternative route to run round the lake, so I guess Neil and I only had ourselves to blame when we plunged into the lake laughing. The laughter didn’t last long. I can honestly say I have never felt anything so cold before. I thought my heart was going to stop and it was difficult to breathe. There were people in canoes in the water in case anyone got into trouble so I just tried to focus on the opposite bank as I swam and concentrated on moving my arms and legs, which is difficult when you can’t actually feel them anymore. Eventually we got to the other side and we were told to keep running and not stop otherwise we would never get warm again.

The water slide was the other dramatic obstacle. Careering for a good 100 metres down a hill, you have to launch yourselves in pairs over the top of the slide and
then you slide at a really alarming rate down the hill, spinning round and round madly until you land in a pool of muddy water at the bottom. This would have been fine but as Neil launched himself a second before me, and I hurled myself over the top, I knew instantly that something had happened to my ribs as they collided with the top of the slide. I span wildly in circles before being deposited backwards into the water and couldn’t figure out for a second which way was up.

Having dragged myself out of the water at the end of the slide, muttering to Neil that I thought something had happened to my ribs, we carried on running. There were obstacles that I stood in front of and thought “how am I going to get over that?” and all the way around I could hear Dad’s voice in my head saying “You must be mad kid!!” But somehow we managed to get round and at the end we were on a complete high. It took me a couple of weeks to get over the run. My back muscle (wrapped round my chest) went into complete spasm which was so painful I can’t describe it. Once this had passed and the pain diminished, I discovered a new pain in my ribs as I couldn’t breathe or laugh without clutching my side and grimacing. My lymphoedemic arm was like a soggy noodle and swelled up, and my legs had cuts and bruises from the mud pits and colliding with branches that were hidden under the water.

As well as completing the WOLF Run, the fabulous people I work with helped me organise a charity day and we sold cakes and samosas to raise even more money. We managed to raise in total £1,187.14.

I still can’t work out if it was the worst thing I have ever done or the best fun ever, though I think I am leaning towards best fun ever. So many people sponsored us and supported us that it was quite overwhelming. I consider myself quite a lucky person.

I have had to fight some battles but because I have such amazing family and friends, I have fought through them. Despite my Dad passing he stays with me all the time and I hear his encouragement with every step I take.

A year of running for amyloidosis

By Ben Mason

In April 2014, I decided to start fundraising and also to raise awareness for the National Amyloidosis Centre, by taking part in a number of running events.

My Mum Margaret has AL amyloidosis. Following her diagnosis at the NAC, she underwent treatment with chemotherapy and this produced great results in slowing down the production of amyloid. Unfortunately, because diagnosis is so difficult and other more obvious diseases are investigated first, by the time she was diagnosed her kidney function had deteriorated so much that she now has to go to the hospital twice a week for dialysis. This process removes waste and excess water from the blood, a function normally performed by the kidneys.
Her light chain levels are monitored on a regular basis by the NAC, the only place in the UK dedicated to the needs of patients with amyloidosis. They perform a full body scan using a specialist machine called an SAP scanner. This enables them to monitor the amyloid deposits which helps plan her ongoing treatment.

My family and I are so grateful for everything the NAC has done for my Mum so to raise money and awareness I ran in a number of organised events over the course of a year and asked people to sponsor me.

I completed the following events:

- Pudsey (10K)
- Leeds (10K)
- Cheshire Arley Hall Half Marathon
- The Stoop Fell Race
- Chevin Chase Trail & Fell Race (on Boxing day!) – alongside, but very far behind, the famous Brownlee brothers (great British triathletes)
- Soreen Stanbury Splash Fell Race (in the snow!)
- Ilkley Moor Fell Race (on my birthday!)
- Bradford Parkrun
- Baildon Boundary Way Half Marathon
- Windermere Marathon
- Tough Mudder Midlands (11 mile obstacle course)

The day after the Windermere Marathon, I helped at Wilsden Trinity Church in Bradford, who kindly organised a coffee evening to raise further funds for the NAC.

I kept a blog about the above events and my site also contains information and useful links about amyloidosis. With over 2000 visitors reading my blog I feel that more awareness has been raised about this rare disease. If you would like to read my blog, please go to: [http://benmasonamyloidosis.weebly.com](http://benmasonamyloidosis.weebly.com). So far I have raised over £3,000 including gift aid.

**Dobie brothers’ fundraising swim**

By Tim Dobie

My brother Laurence and I signed up for the Boulters to Bray Swim near Maidenhead on 4 July 2015 to raise money for our Mum Anna, who was diagnosed with AL Amyloidosis in 2014. The swim is 2.8 km down the Thames, unsurprisingly from Boulters to Bray. We set our alarm for 4 am for the drive from London to Boulters. We assembled with about 150 other sleepy looking wetsuit clad folk at 6 am for the ‘safety briefing’ which consisted of a few anecdotes about how ill we were going to get from the water and that a can of diet coke will kill pretty much all harmful bacteria. We hoped that the 2.8 km swim would be aided by a healthy downstream flow but, unfortunately, the river was virtually at a standstill between locks so it was going to be down to us to swim the lot.
Neither of us had swum this distance before and certainly not in the Thames, so we were somewhat nervous, but once we were in the murk of the water the nerves settled as we waited for the starting gun. We set off at 6.15 am. Starting a race like this is always slightly eerie. There is pandemonium of thrashing limbs, kicking, splashing, etc but not a word uttered by anyone, just everyone jostling for position and trying to settle into a rhythm. Before long the pack stretches out somewhat and you find your rhythm and you just grind it out whilst trying to keep tabs on how far you have swum. Towards the end it seemed to drag out forever but eventually you find yourself dragging yourself up the river bank at the finish line, somewhat relieved.

Looking back on it, it was a great experience. They close the river to boats for the swimmers so it’s quite a privilege. It’s certainly an invigorating way to start the day! We were surprised by our times; Tim 46 minutes, 34 seconds and Laurence 54 minutes flat. Maybe the river did help us a bit after all! ... and yes we did hastily consume plenty of diet coke straight afterwards, just in case.

Donations

Research and development of new treatments for amyloidosis in the Centre have made enormous contributions to understanding of the disease. Availability of resources in the UCL Amyloidosis Research Fund is of immense benefit for this research. The resources provided by the UCL Amyloidosis Research Fund, through the generosity of donors, undoubtedly makes major contributions towards alleviating the suffering and saving the lives of many thousands of people afflicted by this terrible disease. Every penny is received with sincere gratitude and is used specifically and in its entirety for amyloidosis research.

The UCL Amyloidosis Research Fund is a charitable fund within University College London (UCL) which qualifies for Gift Aid. This increases the amount donated by 25% without any extra cost to the donor. It does so by allowing UCL to claim back the basic rate tax paid by the donor. To qualify for Gift Aid the donor must be a UK taxpayer and his/her combined income and capital gains tax bill must equal or exceed the amount UCL claims back on the gift. The gift can be for any amount and applies to one-off gifts and regular gifts made over a number of years. For UCL to claim the tax benefits the donor must complete a Gift Aid form. To claim back the Gift Aid cheques should be made payable to “UCL Development Fund”. This money is then transferred to the Amyloidosis Research Fund, together with the reclaimed tax. If a donor does not qualify for Gift Aid, or does not wish to contribute in this way, cheques should be made payable to: “UCL Amyloidosis Research Fund”.

Donations may also be made online at:
www.ucl.ac.uk/amyloidosis/support-us
or by post. Gift Aid forms are also available online or from:

Beth Jones
National Amyloidosis Centre
Division of Medicine, Royal Free Campus
University College London
Rowland Hill Street
London NW3 2PF UK
Tel: 020 7433 2802
Email: beth.jones@ucl.ac.uk

Newsletter funded by a bequest from Laura Lock

Newsletters are available at the NAC, by post and email, and online at www.ucl.ac.uk/amyloidosis/nac/newsletters
If you would like to receive copies by post or email please contact Beth Jones (contact details above)