AL amyloidosis Matters

Issue 14 **Summer 2017**

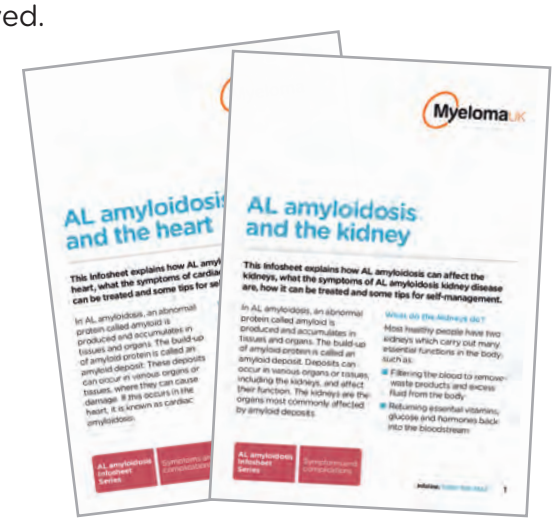
New patient information available from Myeloma UK

Myeloma UK has added to its range of AL amyloidosis information with two new Infosheets on the heart and the kidney.

The Infosheets aim to help patients understand more about how AL amyloidosis affects the heart and kidney, the symptoms that can be caused and how they can be treated and managed. Both Infosheets also contain tips for self-management when the kidney or heart is involved.

You can get copies of the new Infosheets by:

- Calling the **Myeloma UK Infoline** on **0800 980 3332** or emailing **askthenurse@myeloma.org.uk**
- Downloading from our website **www.myeloma.org.uk**



For a full list of all Myeloma UK AL amyloidosis publications, visit **www.myeloma.org.uk/publications**

IN THIS ISSUE

Welcome

Welcome to the 2017 summer issue of *AL amyloidosis Matters*.

In this edition you can read the second instalment of patient Richard Thompson's book – Meeting Amy – and articles on planning a holiday and complementary therapies.

Our patient experience, Gary Mines, shares his experience of being on the PRONTO clinical trial, and what it's like not knowing whether he's on a placebo or the active drug.

I hope you enjoy.

Jude Leitch
Editor

AL amyloidosis Matters

For feedback, comments and questions about the newsletter contact Jude Leitch on 0131 557 3332 or email jude.leitch@myeloma.org.uk

News&Notes

Active compound in green tea could be effective against AL amyloidosis

Researchers at Washington University in St. Louis, Missouri have discovered that an active compound found in green tea leaves affects the structure of the amyloid protein that builds up in AL amyloidosis. The team found that, in laboratory experiments, EGCG (epigallocatechine-3-gallte) affected how the amyloid proteins folded, preventing the formation of the misshapen amyloid deposits that can build up in the tissues and organs of patients. The authors had previously examined EGCG's effect on the proteins associated with Parkinson's and Alzheimer's diseases and found similar results. Further lab tests are planned and the research team hope to bring the compound into clinical trials involving AL amyloidosis patients in the future.

Six-minute walk test a potential outcome measure for cardiac AL amyloidosis

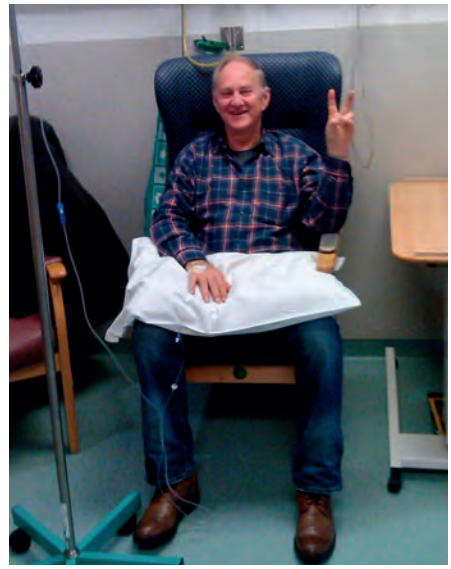
The six-minute walk test (6MWT) is a well-established outcome measure in a variety of diseases including heart failure. A study has been carried out to determine the impact of treatment on changes in the 6MWT in 22 cardiac AL amyloidosis patients. The patients performed the test at diagnosis and then again after bortezomib (Velcade®)-based treatment. 12 patients also went on to receive high-dose therapy and stem cell transplantation following their initial treatment. 6MWT distance improved in 18 of the 22 patients (81%), by an average increased distance walked of 90 metres. The study shows for the first time that the use of the 6MWT could be used as an objective marker of heart function improvement in cardiac AL amyloidosis patients.

Travel and accommodation funding available for this year's Infoday

Our annual AL amyloidosis Patient and Family Infoday will take place on Friday 17 November 2017 at Friends House, Euston Road, London. For patients and family members based a little further afield, Myeloma UK has limited funding available to contribute towards the costs of travel and accommodation in order to attend the event. For those who have not attended before, the Infoday features experts talking about the latest in the treatment and management of AL amyloidosis and provides the chance to meet others affected by the condition. If you would like to request a funding application form, or have any queries regarding the event, please contact Hollie Nichol, Events Co-ordinator, on 0131 557 3332 or email infodays@myeloma.org.uk

Meeting Amy
– a cautionary tale (PART TWO)

BY RICHARD THOMPSON



Richard Thompson noticed swelling in his left ankle on Boxing Day 2014. Thinking it related to his prostate troubles, he thought not much of it and underwent a prostate operation in April 2015. In every issue of *AL amyloidosis Matters* in 2017, we will publish extracts from his book *Meeting Amy – a cautionary tale* about what followed.

August 2015

I arrive at a concrete and glass edifice where a receptionist directs us to the second floor along monochrome passages, through double doors onto some kind of ward. To my left are rows of people sitting quietly in numbered armchairs, attached to bottles dripping solutions of various colours into their arms.

I gape, my imagination running wild.

"Can I help you?" someone asks from a desk to my left. I want to say 'no thanks, I must have made a mistake, I should be somewhere else...', but nothing comes out of my mouth.

"You're here for your first Velcade?" I nod weakly, wondering if I'm going to be hooked up to a bag.

"I'm afraid it's going to be a bit of a wait while we prepare the medication. We do have sandwiches though... in the fridge. Help yourself."

Sandwiches eaten, blood pressure and temperature taken, I get the call, but not for a drip from a colourful hanging bag. My medication comes via injection in the stomach, so quick you hardly have time to wince.

Lovely nurses. Make you feel special even if you're not. Then they fill a carrier with more boxes of pills than I can count, while attempting to explain what they're all for. There are drugs to counteract other drugs, plus a daily anti-coagulant injection that sounds like Fragment. By the time I've taken this lot each day I'm going to rattle, or melt.

September-October 2015

I'm half way through my second five-week chemo cycle (Velcade, cyclophosphamide, dexamethasone).

It's been pretty gruesome, but I'm informed by The Boss that my 'kappa light chains' are diminishing and the little b******s messing with my kidneys are taking a hammering. I suppose that's good, but for three days a week it doesn't feel like it.

The only way to describe it is like when you had those adolescent hangovers after drinking cheap port all night and you wake up feeling like you've been filled with cement and your head's gone AWOL.

Still, mustn't grumble. Some people feel like hell all the time on this stuff.

28 November 2015

Now I'm on the mend. Official.

My second visit to the National Amyloidosis Centre in London, last week, concluded with a consultation in which a senior kidney specialist told me my offending 'kappa light chains' had been reduced to normal and I might not have to continue with chemotherapy.....! She's recommended a 'suspension' of chemo, which is medico speak for being given a get-out-of-jail-free card.

This means I'll continue to be monitored to ensure those nasty little proteins are under control. I'm not out of the woods yet and will have to carry on taking a cocktail of non-chemo stuff, but at least I won't be spending three days a week in a purple haze.

Is that a result? You're damned right it is.

Read the final instalment from Richard's book in the next issue of *AL amyloidosis Matters*.

Going on holiday: things to think about

BY SELENA HALLAHAN
Patient Information Officer, Myeloma UK

Going on holiday offers a much needed break from daily routine as well as the chance to experience different countries and cultures. A diagnosis of AL amyloidosis does not need to mean an end to holidays, however there are a few things that patients should consider before travelling.

Planning the holiday

Planning the trip is an exciting part of a holiday. Be realistic when deciding where to go and what to do, and take into account the weather, travel distances, and anything else that might make you uncomfortable when travelling.

You should speak with your hospital doctor to confirm that you are ok to travel – this is particularly important if you are currently on treatment. It can sometimes be more difficult for AL amyloidosis patients to find travel insurance, so it is also important to look into this as early as possible when planning a holiday.

Travelling

If flying, speak to a doctor about your risk of blood clots. Clots are more likely on long haul flights and if you are on certain treatments, such as thalidomide and lenalidomide (Revlimid®). Doctors will usually advise getting up every so often, wearing compression socks and staying well-hydrated.

It is usually ok to carry medication for trips lasting less than one month, provided it is carried in hand luggage with a copy of the (repeat) prescription. For longer trips, an export licence from the Home Office may be needed.

When on holiday

Remember to use sunscreen and wear a sun hat and comfortable, loose cotton clothing to help to prevent sunburn. This is especially important if you are having, or have recently completed, chemotherapy treatment.

You may find it useful to carry a letter from your doctor outlining your treatment history and any medication you are currently taking. It is also a good idea to take extra medication when travelling, in case of unforeseen delays or complications such as stomach upsets or infections.

Holiday checklist:

Planning

- Discuss the trip with your doctor – they will advise you on whether or not you are fit enough to go away. If so, ask them for a 'fit to travel' letter
- Shop around insurance companies as soon as possible
- Be realistic in your travel and holiday plans

Travelling

- Pack medications in hand luggage along with a copy of the prescriptions or a letter from your doctor. If carrying syringes, check the airline's policy
- Notify the airline if you require additional assistance while travelling
- Long haul flights – be aware of the risk of blood clots and take preventative steps
- Keep comfortable – bring bottled water (purchase after security), a travel cushion, get up often and request extra leg room if it will help

Holidaying

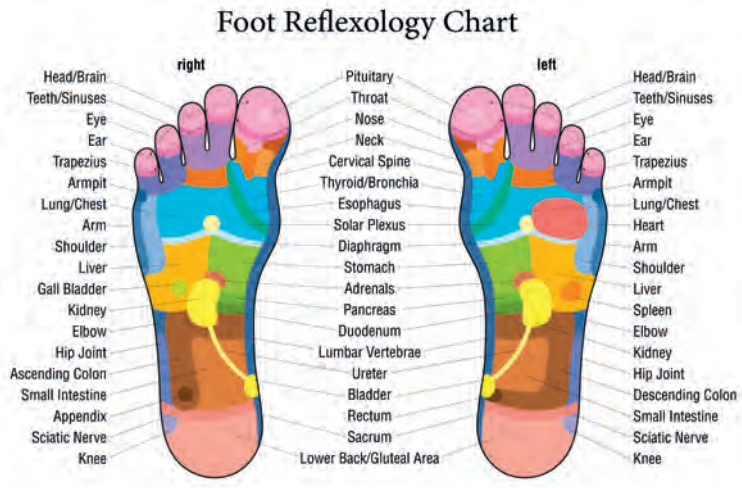
- Carry a letter outlining treatment history and details about your condition
- To prevent stomach upsets abroad, avoid tap water, ice, salads and under cooked meat or fish
- Use sun block, sun hats and wear light cotton clothing in hot countries

For more information see the *Travelling and travel insurance Infosheet* from Myeloma UK

Complementary therapies and AL amyloidosis

BY ALICE BARON
Patient Information Officer, Myeloma UK

Complementary therapies can be used alongside conventional treatment for AL amyloidosis to help improve your emotional, psychological and spiritual well-being. For example, complementary therapies may help to relieve stress and anxiety and promote relaxation and peacefulness.



Although complementary therapies do not treat the underlying AL amyloidosis, some patients find them to be a helpful way of alleviating certain symptoms and side-effects such as pain or fatigue.

There are many complementary therapies available. Three examples are outlined below.

Reflexology

Reflexology is based on the theory that areas of your feet, legs, face and ears represent, and are connected with, the body's internal organs.

In a reflexology session the therapist will massage and apply pressure to different areas of your feet, for example, with the aim of helping to relieve symptoms in particular areas of your body, such as pain or nausea.

Reflexology is generally considered relaxing and may also be an effective way to alleviate stress, and enhance relaxation and sleep.

Aromatherapy

Aromatherapy uses essential oils with the aim of promoting physical and psychological well-being.

Aromatherapy can be carried out in a variety of ways. Most commonly, essential oils are added to a massage oil and massaged into your skin. The smell and touch should be soothing and calming.

If you are having chemotherapy, your skin may be more sensitive than usual, so the scent of the oils can also be enjoyed by adding them to your bath water, a diffuser or a handkerchief.

The scents of the oils are thought to elicit different effects, for example lavender can be calming while peppermint can be energising, so the aromatherapist will discuss how you are feeling to find the most suitable essential oils for you.

Tai chi

Tai chi is an exercise of slow continuous movements and deep breathing that may help to reduce stress and improve mobility. It can be enjoyed by all ages and it is very low impact form of exercise.

A teacher will guide you through a series of positions and movements that flow from one to the other. The movement can help strengthen your muscles and improve balance and posture.

Gentle exercise is considered beneficial for AL amyloidosis patients, however you should only do as much as you feel comfortable with. If a movement is too strenuous, your teacher can offer a suitable alternative.

There are many other complementary therapies available – check with your doctor or nurse which therapies are available in your local area.

To access a complementary therapy you can ask your doctor to refer you, put in a self-referral or find a private practice. It is always important to tell your doctor before starting a complementary therapy.

Summary

Using complementary therapies can give you a feeling of control over your AL amyloidosis and its treatment, although there is no medical evidence that they have any effect on the disease itself. Carers may also find therapies aimed at reducing tension and promoting relaxation useful.

PATIENT EXPERIENCE

Gary, 54, was diagnosed with AL amyloidosis ten years ago. The condition affects his kidneys and his heart. He was the first AL amyloidosis patient at the Royal Liverpool Hospital to have a pacemaker fitted to help counteract his cardiac symptoms.

Gary first featured in *AL amyloidosis Matters* in 2013. Here, we get an update from him on his general situation and he also tells us about being on the PRONTO clinical trial. PRONTO is a Phase II clinical trial studying the effects of the monoclonal antibody drug NEOD001 in AL amyloidosis patients with cardiac problems.

Since last writing for *AL amyloidosis Matters* in 2013, I have been on lenalidomide (Revlimid®) and dexamethasone after relapsing in July 2014. I had this for 5 cycles and fortunately had a complete response. This treatment combination was better for me than the previous combinations (involving bortezomib (Velcade®) and thalidomide) as I found it easier to cope with. As with all my previous treatments I had a course of cardiac rehabilitation after it.

In May / June of 2015, I was still having problems with atrial fibrillation (an irregular heartbeat) and shortness of breath so it was decided that I have an ablation (a procedure that aims to control or correct certain types of abnormally fast heart rhythms). I went into hospital on 1 September 2015 for the procedure in which they inserted a catheter into a vein in my groin, then used radiofrequency energy to 'burn' the walls of my heart to stop the irregular beats / flutters. Again I was the first person with cardiac AL amyloidosis to have this procedure done at the Liverpool Heart & Chest Hospital. I was in theatre around 5 hours. Unfortunately the procedure wasn't a success as they couldn't manage to correct all the irregular circuits. One of the main problems I had after the operation was that they couldn't stop the bleeding from the puncture hole in my groin (in which the catheter was inserted) which bled for around 28 hours. Eventually they stopped the bleeding by double stitching the wound.

I have recently had my pacemaker and medication adjusted and my periods of atrial fibrillation seem to have been reduced. My kidney function is stable at present but this can go down with every infection I pick up.

Overall my condition is stable as I am still in a period of remission after my last treatment with lenalidomide and dexamethasone 28 months ago.

Overview of PRONTO clinical trial – NEOD001 vs placebo

This trial is taking place in the UK, Europe and the USA. It is testing a drug called NEOD001 in AL amyloidosis patients with heart involvement, who have previously responded to treatment and have ongoing cardiac problems. The purpose of the trial is to evaluate whether the drug is safe and effective, and also whether it improves heart function.

NEOD001 is a monoclonal antibody drug that has been developed to target the abnormal proteins (amyloid) that causes AL amyloidosis.

In the trial patients either receive the NEOD001 drug or a placebo drug. The trial is 'double blinded' which means neither the patient nor the National Amyloidosis Centre (NAC) know whether the drug or the placebo is being given – only the drug company, Prothena, know.

There is an initial screening process to see if patients meet the criteria to be eligible for the trial. Some of the things they look at are kidney and heart function tests, along with other critical criteria.

Once assigned onto the trial patients are given either the drug or placebo, both coming in a saline pack. If patients receive the drug, it will have been put into the saline before being sent out to the participating hospitals.

Patients receive an infusion every 28 days for a period of 12 months.

They have various blood samples taken before each infusion, then some following it, and are also closely monitored during and after receiving the infusion. They also check patients' heart function through echo scans, ECGs (electrocardiograms) and a six minute walking test.

If, after the 12 month period, patients find out they have been receiving the placebo, they will be invited to participate in what's called an "open–label extension study" of NEOD001 for a further 12 months.

Whilst on the PRONTO trial patients cannot receive any other treatment for their AL amyloidosis, or participate in any other clinical trial. Patients can come off the trial at any point but would need to continue to be monitored for a period of time.

Gary receiving the drug or placebo as part of the PRONTO trial

My PRONTO trial experience

In September 2016 I was accepted onto the PRONTO trial. I travel to the Royal Free Hosptial in London to participate and my first drug infusion was on 4 October 2016. Just like every patient involved, I'm not sure whether I am receiving placebo or NEOD001.

I've had 9 infusions to date, with some symptoms. Obviously I'm not sure if these are side-effects of the drug or normal symptoms related to my AL amyloidosis.

Each infusion / visit to the Royal Free is done over two days. On the first day I have bloods taken, I'm weighed and have my blood pressure checked. Then I discuss any concerns, new symptoms etc with a Consultant from the NAC. If all is ok then I proceed to day two.

On day two I report to the Oncology ward in the Royal Free Hospital. Again, I'm checked over and if ok to proceed I wait for the drug / placebo to be delivered from the pharmacy. I have found this to be the main problem with the trial - the amount of time wasted waiting for the pharmacy to deliver. Another problem for me on the trial is getting down to London from Liverpool every 28 days for a two day period. This obviously affects my work but my employer has been very understanding.

Once the pharmacy deliver the drug / placebo, I am given any pre-medication needed, then a cannula (thin tube) is inserted into a vein and I'm given the drug or placebo over a given time period (usually 1 – 1.5 hours). Then if I'm feeling ok, I'm allowed to leave after a further 1.5 hours.

On certain visits I have other tests carried out, for example ECGs, urine tests, walking tests and heart echo scans. I also update a log with questions regarding my health and wellbeing on some visits.

After 9 infusions I still have no idea if I am on the drug or the placebo. My blood results are fluctuating up and down, so I cannot really get a good sense of what's going on. At least they're not getting steadily worse! My inkling would be that I am not on the drug, but who knows?!

I think being on the trial is very interesting. I would advise anyone that is asked to go on a trial to accept as the trial drugs may not only help you, but the results will also help future patients.

My hope is that we begin to see more drugs being developed for AL amyloidosis that, like NEOD001, aim to disperse amyloid deposits in the organs and give people like myself a better quality of life.

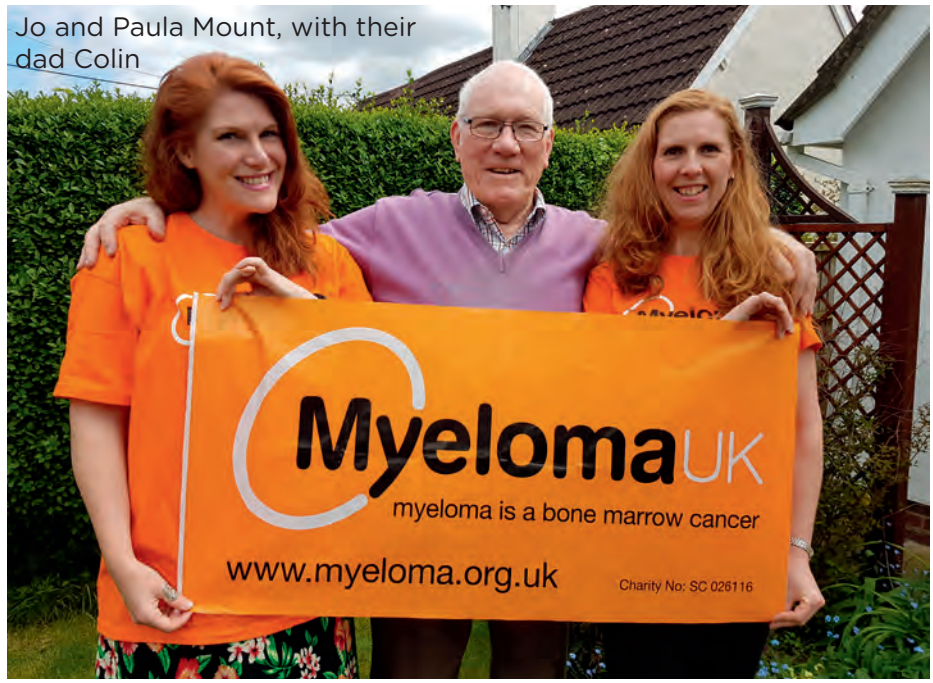
Even better, a drug may be found that can totally cure AL amyloidosis.

Raising funds and awareness for AL amyloidosis

Jo Mount and family decided to fundraise for Myeloma UK in memory of her mum, Margaret, after she passed away in November 2016, having been diagnosed with both cardiac AL amyloidosis and myeloma just 4 weeks earlier.

Jo explained they planned lots of different types of fundraising because "we recognise there are many charities out there and lots of fundraising events every day, so we are mixing it up by choosing fundraising ideas that either push us to our limits or aim to bring people together." So far the family have raised over £4,400 from the events they've held, and through their Virgin Money Giving page set up for people to donate in memory of Margaret.

They chose to hold a tea party as their first fundraising event as a way to bring together everyone in the local community who knew their mum. Since then, Myeloma UK has been chosen as a local charity supported by the Waitrose token scheme and a friend of the family, Leanne Barrett, ran the Devizes Half Marathon to raise money for Myeloma UK. Jo's sister, Paula, will be doing a Wing Walk (walking on the wings of an aeroplane during flight), organising a cabaret night and singing at a fundraising dinner being arranged by the Dorset Myeloma Support Group. Jo is currently training to do a half marathon later in the year.



Jo and Paula Mount, with their dad Colin

> **"Fundraising has been a way of channelling our energy into something positive. If we can help educate everyone we come into contact with, by raising awareness and encouraging them to question and talk about this disease, this will go some way towards evoking change."**

After seeing it take over a year for the doctors to find out what was wrong with their mum, the Mount family also wanted to help raise awareness of AL amyloidosis by talking about the rare condition widely within their communities. "It has been devastating to lose our mother so young. We had a choice and decided that although we could not change what had happened to our beautiful mum, we could stand up and fight for better awareness so GPs, key medical professionals and cardiologists understand more about this condition."

Virgin Money Giving page
http://uk.virginmoneygiving.com/SomeoneSpecial/margaretmount

Thank you for your generous support.

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