

Amyloidosis News

CARING FOR PATIENTS AND THEIR FAMILIES LIVING WITH AMYLOIDOSIS
ISSUE 2 2011

The difficult road to diagnosis by David Birchenough

I am a 54-year-old automotive spray painter from Perth who has worked in and around a solvent-saturated environment most of my life.

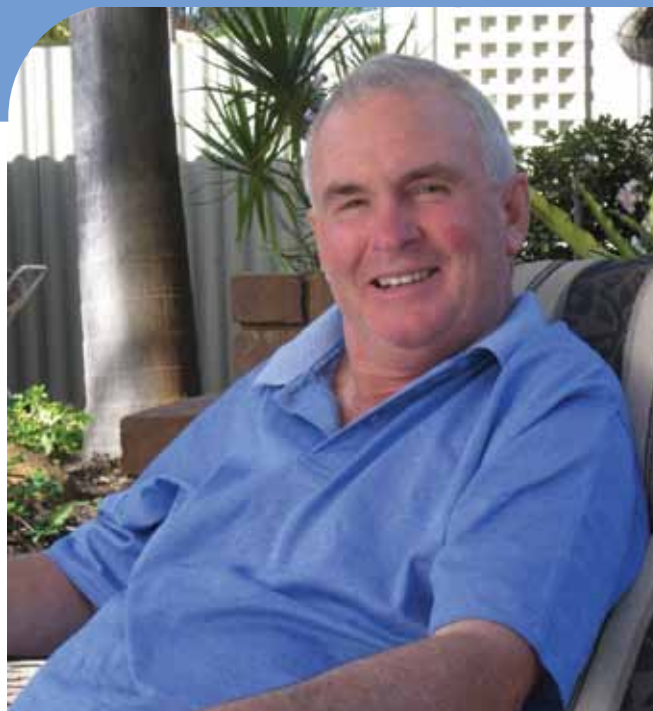
In November 2009, after two years of intermittent breathlessness, pains in my chest on exertion and many tests including an echocardiogram and cardiac angiogram, which appeared normal, I was diagnosed with AL amyloidosis. In the months before diagnosis my symptoms worsened with bloating, night sweats, clamminess, nausea and episodes of feeling very faint. On two occasions I was found to have protein in my urine that perhaps should have rung alarm bells but didn't.

After collapsing on the golf course, a further echocardiogram showed nothing. I then collapsed at home and a heart monitor in the accident and emergency department identified a period of excessive heartbeat and the ECG recorded an episode of tachycardia. I was told I wasn't going home until a reason was found.

Further tests and a renal biopsy finally identified AL amyloidosis affecting my heart and kidneys, which I quickly learnt was a rare and serious condition with dire consequences if left untreated and I would need a cardioverter-defibrillator implanted followed by chemotherapy and a stem cell transplant.

As we knew little about the disease we felt we needed a second opinion, but from where? This was not because I didn't trust my doctors but given that it was such a rare condition, I wondered how many people with this disease these doctors had actually treated.

My wife found the National Amyloidosis Centre in London on the internet. We emailed them



David Birchenough

with the proposed treatment and received an immediate answer assuring us that if my diagnosis was definitely AL amyloidosis, the suggested treatment was the best treatment for me, emphasising that time was of the essence. In November 2009 I received my first lot of chemotherapy followed by stem cell stimulating injections.

In December I received high-dose Melphalan before being given an infusion of my own stem cells. I quickly learnt that nothing goes entirely to plan and instead of the three weeks I thought I would spend in hospital I was actually there for seven weeks, three of them in intensive care on dialysis after developing sepsis and going into kidney failure.

There were periods when I wondered whether I would die and I remember feeling at one stage that it would be easier to just slip away. I might have done so if it wasn't for the support and love of my wife and children. I was eventually released to the outside world in January.

continued on page 10

In this issue

Page 3
News from London

Page 4-5
How amyloidosis affects your kidneys

Page 6-7
Questions & answers

Page 8
Research news

From the editor



For some 2011 has been a worrying year, facing a new diagnosis of amyloidosis or relapse of your disease. For others the happy news of remission has been received. Whatever your situation, I hope the Leukaemia Foundation or other amyloidosis support groups were able to give you emotional and practical support when you needed it.

I have always believed that good working relationships with other support agencies in Australia and around the world are very important. This close co-operation was illustrated recently when Australian patients were invited to submit their questions to a very experienced panel of doctors speaking at the American Amyloidosis Foundation's patient and family meeting in Detroit.

I was also delighted to be a guest at the Myeloma

Travelling with amyloidosis

If you have a pre-existing medical condition (known as an EMC within the travel insurance community) such as amyloidosis, what are your options when travelling?

Your first consideration is your destination, then the level of medical support that can be provided in relation to your condition.

Does the country you are planning to visit have a level of health care that is appropriate for your condition, and what costs are involved if you travel to a country that isn't covered by your travel insurer? When travelling with an EMC, regions such as South America, North America, Canada and Africa are considered 'high-risk' areas in terms of appropriate care.

Most travellers are unaware that in the following countries — New Zealand, United Kingdom, Republic of Ireland, Sweden, The Netherlands, Finland, Italy, Belgium, Malta, Norway — Australian residents are entitled to assistance with the cost of medical treatment under the Reciprocal Health Care Agreement (excluding elective or cosmetic surgery).

UK information day for AL amyloidosis patients and families in London. (Page 3).

The American Amyloidosis Support Group has recently run a successful patient and family meeting for those with familial amyloidosis. A panel of distinguished doctors spoke and these excellent talks can be accessed at www.amyloidosisupport.com.

I have become increasingly aware that we offer AL amyloidosis patients far more support and education than those living with AA and familial amyloidosis. I do hope this can be addressed in 2012. If you have these conditions I would love to speak with you.

I am, like many of you, very grateful to the Leukaemia Foundation for the support offered to patients and families around the country. I would particularly like to thank the Leukaemia Foundation of Queensland for its continued support in funding the publishing and distribution of approximately 500 copies of *Amyloidosis News* around Australia and overseas twice a year.

I also thank the patients and families who have had input into the articles for this publication, particularly David Birchenough for his personal story. I am also very grateful to the doctors who have taken time to check the medical articles. Please be encouraged to send your personal stories and article suggestions.

On behalf of all Leukaemia Foundation staff I'd like to wish our readers a happy and peaceful Christmas season.

Pat Neely
Amyloidosis Patient and Family Advocate
Leukaemia Foundation

These countries each provide care in different ways, so it is best to review the relevant services when you are planning your trip.

To gain access to care, you need your passport and a current Australian Medicare card. For more information visit www.medicareaustralia.gov.au. (Use the search facility by entering Reciprocal Health Care.)

For destinations not covered by the agreement, look into the state of medical care in each country because under the terms of your travel insurance, you may have to personally cover the cost of any treatment you require.

Communication is another consideration. Is not having an English-speaking doctor/surgeon a cause for concern?

Your chosen travel insurance provider will always ask you to submit a pre-existing medical assessment. Based on that assessment, they will either turn you down or accept coverage and require an additional premium payment.

For more information and guidance, contact Leisa Burdette, Travel Managers Australia on 0405 100 095 or email leisab@travelmanagers.com.au.

Amyloidosis news from London

by Pat Neely, Amyloidosis patient and family advocate, Leukaemia Foundation

I was delighted to be a guest at the Myeloma UK's AL Amyloidosis Patient and Family Information Day in London in September. The event was chaired by the Medical Director of the National Amyloidosis Centre (NAC), in London, Professor Philip Hawkins, and included a range of guest speakers (all NAC consultants).

Dr Julian Gilmore, senior lecturer and honorary consultant gave an overview of amyloidosis illustrating how complicated this group of diseases is both to diagnose and treat.

Dr Ashutosh Wechalekar, honorary consultant haematologist, spoke on current and future strategies for the treatment and management of AL amyloidosis. He gave an overview on how treatments have been borrowed from myeloma but emphasised the differences in treating amyloidosis where patients are often sick and frail due to the tissue and organ damage. Emphasis was on early diagnosis followed by correct typing of the amyloidosis. An explanation was given on how treatment decisions are made through thorough patient assessment taking into account age, general health, number of organs involved and severity of damage, especially if the heart is involved. The NAC has tended to use CTD (Cyclophosphamide, Thalidomide and Dexamethasone) more than Melphalan and Dexamethazone as first-line treatment. High-dose chemotherapy followed by stem cell transplant is usually only recommended for the younger, fitter patient with little organ damage.

Dr Wechalekar said that the prognosis for patients had improved greatly due to earlier diagnosis, thorough assessments and a greater understanding of treatments but there was still a need for new treatments to cure the disease. However great hope lies in changing amyloidosis treatment largely through the exciting research now being carried out by Professor Pepys and his team that is due to go to human trials next year. (See page 8)

Dr Wechalekar himself has been the chief investigator in the ALCHEMY study (AL amyloidosis chemotherapy). The results to date were presented at the International Myeloma Workshop in Paris earlier this year and have already led to change in clinical practice in the UK. (See page 8 for more information).

Dr Helen Lachmann, honorary consultant nephrologist, spoke on supportive care for damaged organs and how patients could help themselves.

The day finished with two excellent presentations by patients on their experiences.



I was invited to give a brief overview of amyloidosis support in Australia, which covered everything offered by the Leukaemia Foundation. *Amyloidosis News* raised particular interest.

Although Myeloma UK offers excellent support to AL amyloidosis patients and families through online educational publications, telephone support, information days and a support group, they do not offer services for other types of amyloidosis. Nor do they have a newsletter specifically for amyloidosis. At the end of the event it was announced that a questionnaire would be circulated to patients and families to review amyloidosis support services in the UK.

I had the opportunity to talk with patients and families and as always learnt a great deal from their stories. The general questions they asked the doctors mirrored those asked by Australian patients such as, "Why is amyloidosis often only diagnosed after many visits to different doctors?" and "Why don't doctors communicate better with one another, especially when there are a number of different specialist involved in treatment?"

To access the full power point presentations visit www.myeloma.org.uk. Go to patient services / patient and family info days / past info days.

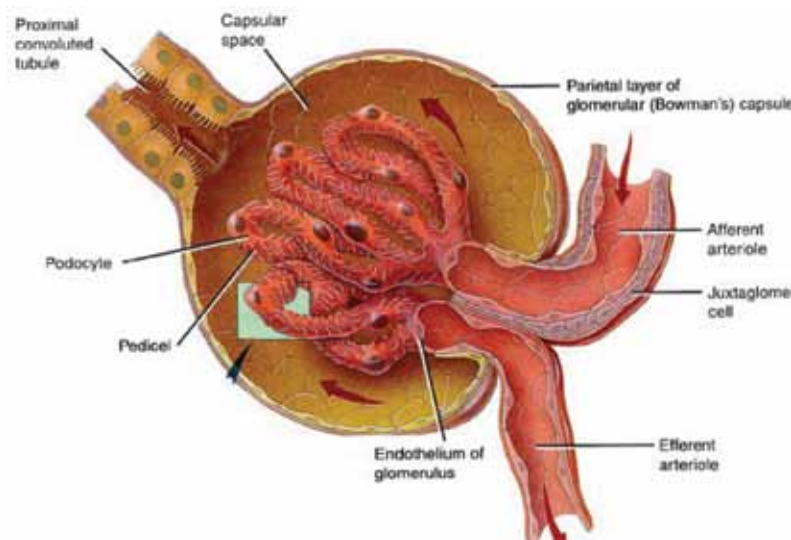
How amyloidosis affects your kidneys

The following is an overview of a talk given by Brisbane-based nephrologist, Dr Andrew Bofinger, to the Leukaemia Foundation's amyloidosis luncheon in Brisbane and to participants of the amyloidosis telephone forum.

The normal kidney

Most people are born with two kidneys. Each kidney has one million filters, called glomeruli. The kidneys receive about a fifth of the body's circulating blood supply. The role of the kidneys is to filter off fluid from the blood, leaving behind cells and big proteins to produce a pre-urine.

Once the pre-urine is made it travels along tubes, attached to each filter where cells lining each tube suck out everything that the body needs and returns it to the bloodstream.



The illustration shows the kidney filter known as the glomerulus which consists of very fine capillaries (fine blood vessels) known as tufts all within a capsule.

The blood vessels enter and leave at one end of the capsule. The blood enters, travels through the capillary loops, which are a little like a ball of string all wound up. As it travels through the thin-walled capillary, fluid leaks through.

This fluid leaks through a barrier which is made up of the cells lining the capillary (the endothelial cell), the basement membrane (a supporting scaffolding to which the endothelial cells are attached), and the podocytes which are outside the capillaries and have long fingers which wrap around the capillaries. Between these long fingers is a lattice of proteins

which act as a sieve. The fluid travels through or between the lining cells on the inside, across the basement membrane, through the lattice of proteins between the podocyte fingers and out into the space between the tuft and the capsule. This space is called Bowman's capsule.

What problems may arise?

The filters may become damaged and stop filtering as much urine or start leaking out things that should be retained in the body. When a significant number of the filters are damaged, the remaining filters have to work harder.

Eventually the remaining filters can be damaged and reduced further in numbers. Patients may get down to less than 10% of the normal numbers of filters and still be able to pass urine as the kidneys can adjust.

Amyloidosis and the kidney

Amyloidosis is an umbrella term for more than 25 different diseases in which an abnormal protein known as amyloid is produced. These amyloid fibrils deposit and accumulate in any of the organs and tissues of the body leading to organ dysfunction.

The kidney is the most commonly involved organ in AL amyloidosis. The kidneys can also be affected in AA amyloidosis and in the hereditary type of amyloidosis Afib.

The way the kidney is affected will depend on the extent of the amyloid infiltration and where the infiltration occurs.

Amyloid in the kidney most often presents within the glomeruli.

The most common presentation is through protein spilling into the urine. When this increases it becomes known as nephrotic syndrome.

Amyloid can also deposit just within the blood vessels, narrowing them and slowly reducing the blood flow resulting in declining kidney function in a way that is no different from someone with severe blood pressure. Amyloid patients with this presentation may not have protein in their urine.

In rare cases amyloid deposits in the tubes, damaging the mechanism for getting rid of the body's waste products. This may result in acid retention and build up in the blood. The body is then unable to concentrate the urine with the result that people urinate too often, a problem called poly ura.



What is nephrotic syndrome?

Normally we urinate 150 mgs or less of normal protein a day but in nephrotic syndrome it is 20-plus times that level. Albumin in the blood drops below the normal range and patients develop high cholesterol. Salt and water are retained which causes bloating.

Amyloid is one of the least common of the diseases that cause this syndrome but if a patient presents with nephrotic syndrome many renal physicians would suggest a kidney biopsy and other blood tests to check for autoimmune disease or other light chains or monoclonal proteins in the blood.

Definitive diagnosis of amyloid

Diagnosis usually results from a kidney biopsy in which a few cores of tissue are taken under a local anaesthetic. The pathologist stains this tissue with Congo red to diagnose amyloidosis and uses further stains to distinguish the type of amyloid.

It is essential to establish the type of amyloid before treatment to slow or stop the production of the amyloid begins.

Treatment for nephrotic syndrome

Treatments for nephrotic syndrome caused by amyloidosis are the same as those used for nephrotic syndrome caused by other problems.

- Restrict sodium in the diet to reduce swelling.
- Diuretic use to help pass extra salt and water in the urine and therefore hopefully reduce leg swelling.
- Drugs called ACE inhibitors may be used to help blood vessels enlarge or dilate and reduce blood pressure. These drugs also reduce pressure within the filters of the kidney and reduce protein leakage.
- Controlling the blood pressure with the goal of 120/70 or better which helps to reduce the protein loss.

- Preserving the blood vessels especially in patients with long-term kidney damage to reduce the risk of coronary heart disease and other vascular disease.
- Control high cholesterol
- People with bad nephrotic syndrome can lose clot-inhibiting proteins in their urine. Some patients may need to be treated with the drug Warfarin for a while.

Treatments to stop amyloid production

Treatments to stop or slow the production of the amyloid protein will depend on the type of amyloid, organ involvement, the degree of damage, and general health of the patient.

AL amyloidosis may be treated with chemotherapy, novel drugs and in certain patients a stem cell transplant.

AA amyloidosis is treated by bringing the chronic inflammatory disease under control.

Patients with certain types of hereditary amyloidosis may benefit from a liver transplant to remove the source of their amyloid.

If the amyloid continues to impair the kidney or kidney function deteriorates further because of treatment, calcium and phosphate balance then become important in the care of the patient.

End-stage disease

When the kidney reaches end stage, dialysis is a viable therapy. How well the patient does on dialysis will depend on their overall general health. If the amyloidosis patient is in remission without bad heart or nerve involvement, they will probably do better than someone who has considerable organ damage. In these patients kidney transplantation may be an option.

Q&A

Why is cardiac amyloidosis often not diagnosed until the heart is damaged?

Cardiac amyloidosis is very serious and its diagnosis and management is often complex. In cardiac amyloidosis (CA) the amyloidal proteins, which form insoluble fibrils, accumulate in the heart causing stiffening of the heart muscle resulting in impaired relaxation and contraction and sometimes damaging the electrical system. Cardiac amyloidosis is usually seen in the presence of other organ involvement.

The overall pace of the disease and the prognosis varies among the different types of amyloidosis and from patient to patient with the same condition. It does seem however that cardiac involvement in AL amyloidosis progresses faster than in senile amyloidosis or the hereditary type. It is rarely seen in patients with AA amyloidosis.

The medical literature states that AL patients sometimes deteriorate more rapidly than would seem likely from the degree of amyloid infiltration in their heart with the conclusion that perhaps the light chains themselves, independent of the amyloidal fibrils, may play a role in cardiac amyloidosis in AL patients.

Therapy for systemic amyloidosis is two fold-to stop or slow the production of the amyloid protein and preserve the damaged organ. It is therefore imperative that amyloidosis is diagnosed as early as possible so that patients can receive optimum treatment. But early diagnosis still remains a great challenge. Why?

It appears that patients with cardiac amyloidosis will undergo an asymptomatic preclinical stage. However, because amyloid proteins cause little or no local reaction it is unfortunately rarely diagnosed before there is some degree of organ failure.

Amyloidosis is relatively rare and there is no blood test available to diagnose it. A definitive diagnosis can only be made through biopsy. Patients often present with vague symptoms mimicking those seen in other diseases. These may include carpal tunnel syndrome, renal insufficiency, facial bruising, swelling of the legs, feeling of bloating and in the case of cardiac amyloidosis, exertional fatigue and breathlessness.

Diagnosis therefore often depends on the doctor having a high level of suspicion that these symptoms may be caused by infiltration of the amyloid protein. In the case of cardiac amyloidosis it appears that it is the subtle non-specific findings seen on testing,



such as the low voltage and the pseudoinfarction pattern on an ECG and the increased ventricular wall thickness on an electrocardiogram that should raise suspicion.

A test called a NT-pro BNP in conjunction with cardiac troponins (specific proteins found in the heart muscle) may reflect heart involvement. These tests and the free light chain assay are useful in diagnosis and measuring the results of treatment in AL amyloidosis.

To add to the difficulty of diagnosis cardiac amyloidosis in AL patients may present as a classic infiltration pattern resulting in restrictive cardiomyopathy or a distinct cardiovascular distribution, which may limit cardiac flow. These patients may not show the classic signs of left ventricular hypertrophy seen on the echocardiogram or low voltage on the ECG. This group of patients may also complain of an angina-type chest pain contrasting with the common presentation of exertional fatigue.

The authors of the paper, "How to diagnose cardiac amyloidosis early: impact of ECG, tissue Doppler echocardiography and myocardial biopsy" published in the March edition of the *Journal Amyloid* propose that there should be an accurate clinical ECG and echocardiographic evaluation of patients with heart failure of unknown origin.

I have heard of patients with amyloidosis and myeloma talking about receiving bone-strengthening drugs called bisphosphonates. I have AL amyloidosis but do not also have myeloma. Do I need these drugs?

In approximately 15 - 20% of patients, both AL amyloidosis and myeloma are present at the time of initial diagnosis. Less than 1% of patients with



isolated AL amyloidosis at diagnosis develop myeloma at a future time point.

Myeloma (also known as multiple myeloma) is a cancer of plasma cells where a large numbers of abnormal plasma cells called myeloma cells are made in the bone marrow. These cells multiply without any proper order, forming collections known as tumours that accumulate in different parts of the body, especially in the bone marrow and on the surfaces of different bones in the body. These tumours secrete chemicals that stimulate other bone marrow cells (osteoclasts) to remove calcium from the bone. As a result bones can become weaker, more brittle and break more easily. Bisphosphonates, such as zometa and aredia given through an IV drip are often used in myeloma treatment to strengthen bones.

In AL amyloidosis, the amyloid protein rarely causes problems in the bone structure. Therefore bisphosphonates are not usually recommended for patients with just AL amyloidosis. Doctors may recommend them for patients who have been on long-term steroid treatment or who already have problems with their bones. If you are concerned about not receiving these drugs you should discuss this with your haematologist.

There have been reports that a small percentage of patients taking bisphosphonates also develop osteonecrosis of the jaw (ONJ), the cause of which is not entirely clear.

Symptoms of ONJ include non-healing of a tooth socket after extraction; area of exposed bone in the mouth; swelling of gums; heavy or numb feeling in the jaw or pain; loosening of teeth and discharge of pus.

Antibiotics and painkillers are used to relieve symptoms. Aggressive surgery is usually avoided as this has not been reliably shown to help. An oral surgeon may, however, need to remove some of the dead tissue or bone from the area with a small operation (debridement).

The proven effectiveness of bisphosphonates in treating and preventing bone disease and changing

the outlook for myeloma patients has to be balanced against the relatively small risk of ONJ occurring.

The Leukaemia Foundation recommends that patients with both myeloma and amyloidosis who are prescribed bisphosphonate therapy should speak with their haematologist about the risk of developing osteonecrosis of the jaw.

Are you eligible for Medicare dental services?

Under the Medicare Chronic Disease Dental Scheme, benefits are available for most services provided by a dentist, dental specialist or dental prosthetist in private dental surgeries. Benefits are not available where services are provided to a person who has been admitted to a hospital.

To receive a Medicare benefit for dental services, you will first need to meet certain eligibility criteria and be referred by your GP to a dentist.

For more information go to www.health.gov.au.

What is the SAP (Serum amyloid P) scanner?

Information from the Centre for Amyloidosis and Acute Phase Proteins, London

In 1987 the National Amyloidosis Centre in London devised a completely new diagnostic test for systemic amyloidosis comprising a whole-body scanning procedure called SAP scintigraphy. This scan can show the distribution and amount of amyloid within the body's organs without the need for biopsies. SAP scans take about 45 minutes and are performed six to 24 hours after an intravenous injection of a small dose of radioactive tracer. The procedure delivers a very small radiation dose similar to a routine x-ray. The procedure is safe and painless and can be repeated every six to 12 months to monitor the course of the amyloid deposits and therefore help guide the need for on-going treatment. Over 5,000 scans have been performed and have greatly improved the understanding of amyloidosis and encouraged a much more vigorous approach to its treatment. In particular, it has been shown that amyloid deposits often disperse when the underlying disease is controlled, and this is usually accompanied by an improvement in general health.

NB: This test is unavailable in Australia.

Readers should not rely on information in this column without first seeking advice from their specialist.

If you have a question you would like answered contact pneely@leukaemia.org.au.

Research news

International meeting

The X111 International Symposium on Amyloidosis, will be held at the University Medical Centre, Groningen in the Netherlands from 6-10 May 2012. For more information go to the International Society of Amyloidosis at www.amyloidosis.nl.

Australian trial under way

An important trial currently under way in Australia is the randomised open-label multicenter phase III trial of Melphalan and Dexamethasone (MDex) versus Bortezomib, Melphalan and Dexamethasone (BMDex) for untreated patients with systemic light-chain (AL) amyloidosis. The international principal investigator is Dr Giampaolo Merlini, while the Australian principal investigator is Dr Peter Mollee.

This trial is an international collaboration with the European Myeloma Network. The international accrual target is 250 patients, and the Australasian Leukaemia and Lymphoma Group target is 30.

The anticipated study duration is four years. Tissue samples will be taken for the assessment of a new diagnostic method using laser capture microscopy and mass spectrometry. Serum samples will allow the creation of better assays to measure serum free light chain and also for infrared spectroscopy to try and determine which monoclonal free light chains fold abnormally and lead to amyloidosis. The trial is scheduled to open in the near future at the Princess Alexandra Hospital (Brisbane), Westmead Hospital (Sydney), Gosford Hospital, The Alfred Hospital (Melbourne), St Vincent's Hospital (Melbourne), Royal Adelaide Hospital and a site in Perth.

To read about three amyloid presentations at the HAA2011 meeting highlighting research within Australia go to www.haa-ap2011.org and search the site for "amyloid".

UK ALCHEMY study

Dr Ashutosh Wechalekar, honorary consultant haematologist at the National Amyloidosis Centre in London is the chief investigator in the ALCHEMY study (AL amyloidosis chemotherapy) This research study is funded by Myeloma UK and is designed to assess the outcomes of newly diagnosed AL patients attending the National Amyloidosis Centre (NAC). This study, now in its third and last year, uses a programme of intense monitoring focusing on the patient's life before, during and after chemotherapy and compares



these results with historical cases where patients did not undergo intensive monitoring. Patients recruited into the study have undergone initial treatment at the discretion of their treating doctor, as is UK practice. The majority of these patients have received the combination of cyclophosphamide, thalidomide and dexamethazone (CTD).

This study of over 200 patients has generated a mass of unique information, particularly relating to patients with advanced disease, which has already led to significant change in routine clinical practice at the NAC.

In light of the findings the NAC is offering all patients an additional clinical assessment after their three cycles of initial treatment giving the doctors the opportunity to switch to a Velcade-containing combination if the response to treatment has not been adequate. Future findings will also look at quality of life.

Exciting research moves into human trials

A team led by Mark Pepys, director of the Centre for Amyloidosis and Acute Phase Proteins and Professor of Medicine at the University College London Medical School, hypothesised that depleting SAP in serum and amyloid deposits could prevent new deposit formation and clear out established ones.

In 2002 the team designed a small molecule called CPHPC (R-1-[6-[R-2-carboxy-pyrrolidin-1-yl]-6-oxo-hexanoyl] pyrrolidine-2-carboxylic acid) that bound soluble SAP and cleared it from circulation, preventing the accumulation of SAP in the organs of both amyloidosis animal models and about 50 amyloidosis patients enrolled in a small pilot study.

However, CPHPC was unable to clear SAP in established amyloid deposits. For that task, the researchers decided to use antibodies that recognise

and bind multiple epitopes of SAP in both its soluble and fibril-bound forms.

Now, new findings show that an anti-SAP antibody plus CPHPC can lower amyloid deposits in the liver and spleen of transgenic mice with established amyloidosis compared with CPHPC only or no treatment. Additional studies showed that macrophages play a key role in the amyloid clearance.

Cardiac amyloidosis study

An interesting study, "The Return of the Normal Heart - Resolution of Cardiac Amyloidosis After Bone Marrow Transplant", was recently presented at the Australian Cardiology Conference and later at the ESC (European Society of Cardiology) in Paris.

The study was carried out by cardiologists Dr Ben Fitzgerald and Dr Greg Scalia and haematologist, Dr John Bashford, from the Wesley Hospital in Brisbane. To their knowledge this work has never been presented anywhere else in the world.

The Wesley Group observed unexpected resolution of cardiac amyloidosis in some AL patients after high-dose chemotherapy and bone marrow transplant.

AL amyloid is a blood cell dyscrasia in which the plasma cell produces the light chains which in turn produce the amyloid protein. This protein is different in each patient and its properties determine the tissue and clinical response. The amyloid protein deposits in organs and tissues of the body and without treatment the prognosis is often poor. With the development of cardiac amyloidosis the prognosis becomes worse.

The Wesley Group collected retrospective data from 30 AL amyloidosis patients with cardiac involvement who had been treated with high-dose chemotherapy and bone marrow transplant. This data included patient survival, time to normalisation of cardiac function following bone marrow transplant, ejection fraction of the heart, interventricular and posterior wall thickness, the degree of diastolic function, and the left atrial size.

This data showed that of the 30 patients identified with cardiac amyloidosis following ECG and echocardiographic evidence, 15 responded with normal heart function which took up to 25 months to be achieved.

The study concluded that the process that causes the laying down of the intercardiac amyloid protein can be reversed after chemotherapy and bone marrow transplant in some patients.

To view the full powerpoint presentation google "The Return of the Normal Heart - Resolution of Cardiac Amyloidosis."

Let's talk about practical matters

Have you made a will?

It is important that everyone over the age of 18 and of sound mind should consider making a will.

- A will leaves a clear guide of how you want any assets and belongings distributed.
- A will is cost efficient and may avoid lengthy court battles over who gets what.
- A will is particularly important if you have a family or other dependants, especially if you are a separated or unmarried parent.

A valid will is one that will be accepted by a court and is able to be put into effect. It must be in writing (handwritten, typed or printed) and signed in front of two witnesses.

A will can be made by buying and completing a will form from the post office or newsagent, through a solicitor, or through public trustees offices.

Some important questions:

- Do you have a power of attorney?
- Do you have a health directive?
- Have you discussed organ donation with your family?

Useful suggestions

Place the following documents and information in an easy place for your family to find:

- Wills; birth and death certificates; adoption certificates; marriage certificates; prenuptial agreements; divorce decrees; immigration and citizenship documents; military service records; property deeds; mortgage papers; recent tax returns; all insurance policies; a list of all accounts-savings, stocks and shares, bonds, other investments, credit cards and frequent-flyer accounts.
- A list of the names of your financial institutions, type of accounts you have and the numbers and in what names the accounts are held.
- List the whereabouts of safety deposit boxes, valuables, jewellery etc
- All documents for any taxation lodgement.

If you are receiving medical treatment you should have the following information available:

- A short overview of any illness and treatment you are receiving
- Names, e-mail addresses and phone numbers of doctors
- List of medication taken regularly
- Names, phone numbers and email addresses of family and close friends

Treatment prompts questions

continued from front page

good news was that my free light chains were dropping which indicated the treatment was working and stopping the production of the amyloid proteins. I am pleased to say that I am doing well and my light chains are within the normal range.

I decided this year with the backing of my renal physician, to visit the National Amyloidosis Centre in London for a serum amyloid P (SAP) scan which is unavailable in Australia. (See page 7) There appears to be different opinions as to how helpful this test would be and that it would not alter the outcome. However, I decided to go ahead.

The SAP scan only showed a small loading of amyloid in my spleen that I had suspected due to tenderness in that area. The doctor said that it often does not show up in kidneys that have had chronic kidney disease for a prolonged time as in my case. He went through my story from the beginning and said that my symptoms were very typical including various symptoms I was unaware were associated with amyloidosis. He spent about an hour consulting with me and said I was in good shape considering I was at the top of the ladder as far as prognosis was concerned. He also said that if and when my clonal response to the treatment deteriorated, my body would be in a much better state to withstand a second round of treatment than it was when originally diagnosed. I couldn't have hoped for a better outcome and even though it doesn't change anything, I feel incredibly encouraged by the results and feel a lot better psychologically. For me it was worth the UK£ 3363 cost.

As I reflect on my journey over the past two-and-a-half years, I'd like to share what questions I still have and what I have learnt.

I now understand the importance of early diagnosis in the treatment of amyloidosis and I still wonder why my original echocardiograms didn't pick up the amyloid in my heart sooner. I also wonder whether there have recently been developments in the way cardiac amyloidosis is diagnosed.

I question, as I am sure many patients do, whether there is any relationship between a person's working environment and amyloidosis and whether any research has been done in this area.

It is so important to understand why your doctor



David Birchenough

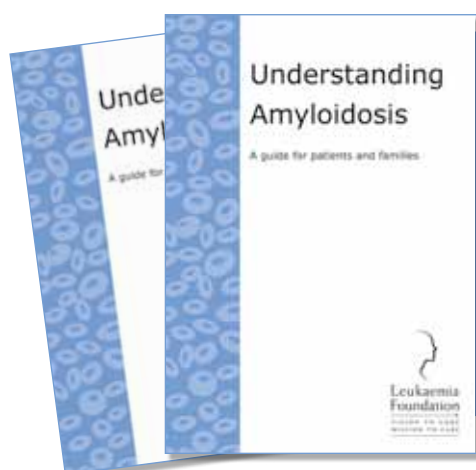
makes particular decisions about your treatment before agreeing to go ahead with it. Be fully informed. Asking appropriate questions at the appropriate time is vital to this understanding.

Once I had made the decision to accept the suggested treatment, my amyloidosis became almost secondary to surviving the ever-changing side effects of that treatment and my world became very narrow. My wife was left to worry about the wider picture.

It is not just the physical aspect of treatment that affects you but also your psychological state is vitally important. Although I did see a psychologist when I was in hospital it was only when I began to feel better physically that I found I was suddenly experiencing very emotional moods, often bursting into tears for no reason. Normally this left me feeling better afterwards. I am sure many of us experience anxiety before our follow-up appointments and the results of our free light chain assays.

Quite some time after my stem cell transplant my GP arranged a care plan, a government paid initiative which included a pharmacist visiting me at home to explain my medications, an exercise and nutrition program and sessions with a psychologist. Every aspect of this package has been extremely helpful and I would recommend it to everyone. I firmly believe that exercise is the best treatment for fatigue.

It was some months after my treatment finished that my hematologist began to talk about prognosis and how well I had done compared with others with the disease affecting their heart. He talked about guilt from surviving and encouraged me to join a support group. I am a member of the Australian Amyloidosis Society and also join the Leukaemia Foundation's amyloidosis telephone forums. This has given me the opportunity to communicate with other patients which I found very helpful.



The Leukaemia Foundation has produced a booklet, *Understanding Amyloidosis*, which offers patients, carers and medical staff information about the disease, its diagnosis and treatment options. If you would like a copy of the booklet or information about any of the Foundation's amyloidosis support services, please contact the Support Services Department on **1800 620 420**. You can also download a copy from www.leukaemia.org.au.

News from around Australia

Queensland

Three successful amyloidosis lunch seminars were held for patients and families in Brisbane this year covering topics including exercise for cancer survivors and how amyloidosis affects the kidneys and heart. Future lunch seminars will resume when the Leukaemia Foundation of Queensland new village at the Boggo Road Precinct at Dutton Park in Brisbane is completed. Patients will be kept informed of the dates. This year I have been working closely with Dr Peter Mollee and the social workers at Princess Alexandra Hospital's Amyloidosis Diagnosis and Treatment Centre to support newly diagnosed patients and their families.

I would like to take this opportunity to wish all readers a safe and happy Christmas and look forward to seeing you in 2012.

Sheila Deuchars

Support Services Coordinator
Leukaemia Foundation of Queensland

Victoria

Almost 400 patients and carers attended Victoria's annual patient conference in Melbourne in September. Twelve of Melbourne's top haematologists presented on a variety of topics including chemotherapy, radiotherapy, bone marrow transplantation and the late effects of cancer treatments.

The myeloma and amyloidosis session, presented by Professor Miles Prince, was well received and patients and carers said they appreciated the opportunity to meet others living with amyloidosis.

The support services team in Victoria and Tasmania are interested in running regular support forums for amyloidosis patients and carers. For more information and to express your interest please contact me on (03) 9863 6957.

Amyloidosis patients often see a number of specialist doctors in the management of their disease. I have recently commenced work on reviewing the pathways of referral for patients in Victoria. Interviews are taking place with haematologists from each treating centre to identify the key health professionals in each health service. The aim is to develop a local directory to assist newly diagnosed amyloidosis patients and their carers to navigate the system in their chosen venue of care. Once the methodology is finalised, the same process will be conducted in Tasmania. Contact me if you would like more information.

Sara Andrews

Support Services Manager

South Australia

The Leukaemia Foundation of South Australia is holding an education session on 30 May 2012 at the BioSA Incubator Conference Centre. Haematologist, Dr Noemi Horvath, will be discussing the latest information and findings from the International Amyloidosis Conference she is attending in Groningen next May. The Leukaemia Foundation's Amyloidosis Patient and Family Advocate, Pat Neely, will also facilitate an educational session and open up the discussion to those in attendance.

We would welcome feedback from our amyloidosis patients and their families on suggestions for topics for future support group events. Please contact Louise Bastian on 08 8273 3515 or Debbie Newton on 08 8273 3510.

Louise Bastian

Support Services Manager

Amyloidosis telephone forums

The next amyloidosis telephone forum will be held on Wednesday, 14 December. The Leukaemia Foundation's Amyloidosis Patient and Family Advocate Pat Neely will discuss the highlights from the Myeloma UK's AL amyloidosis patient and family information day.

Kaye Hose

National Myeloma Coordinator
Email: myeloma@leukaemia.org.au
Ph: 03 9863 6951/0412 681 646

Light the Night

Amyloidosis patients were among thousands of Australians who shone their lanterns for the *Light the Night* sunset walks in support of those who have been affected by blood cancers and disorders.

Light the Night walks were held across the country in September and October creating incredible seas of lights as participants carried coloured lanterns which contained a tiny light and had a special meaning:

- white for blood cancer patients and survivors
- blue for supporters
- gold for those remembering a loved one lost.

Special thanks go to our *Light the Night* sponsors Bridgestone Australia and Seeley International.



Building progress on track

Take a look at the latest construction progress for the new ESA Village in Brisbane and the expansion of Queensland Freemasons Village in Townsville.

It is expected that patients and families will move into these new state-of-the-art accommodation facilities in mid-2012.

If you would like to donate to our fundraising appeal for these projects, please phone 3318 4418.



Brisbane construction



Brisbane construction



Townsville construction



Townsville construction



Townsville construction

Support services

Queensland – Barbara Hartigan

Amyloidosis patient and family advocate – Pat Neely

National Support Services Manager – Anthony Steele

Victoria/Tasmania – Sara Andrews

New South Wales/ACT – Kath Skinner

South Australia/Northern Territory – Louise Bastian

Western Australia – Sarah Langmead

Useful websites

www.amyloidosis.com.au.

www.ucl.ac.uk/medicine/amyloidosis.

www.amyloidosis.org.

www.amyloidosisupport.com.

www.heartfoundation.org.au.

www.kidney.org.au.

**For help all patient enquiries call 1800 620 420
or visit www.leukaemia.org.au.**



Leukaemia
Foundation

VISION TO CURE
MISSION TO CARE

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Brisbane QLD 4001
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Leukaemia Foundation
of Queensland ©

Our Vision to Cure and Mission to Care.

The Leukaemia Foundation of Queensland is a not-for-profit organisation focused on the care and support of patients and their families living with leukaemias, lymphomas, myeloma and related blood disorders.

The Foundation does this by providing emotional support, accommodation, transportation and practical assistance for patients and their families. The Leukaemia Foundation also funds research into cures and better treatments for blood cancers.

The Leukaemia Foundation receives no direct ongoing government funding and relies on the continuous support of individuals and corporate partners to expand its services.

To find out more about the work of the Leukaemia Foundation of Queensland and how you can help, phone 1800 620 420 or visit the Foundation's website at www.leukaemia.org.au.

Disclaimer: No person should rely on the contents of this publication without first obtaining advice from their treating specialist.

If you do not wish to receive future editions of this publication please contact the Leukaemia Foundation Support Services Division on 07 3840 3844.