

Amyloidosis News

CARING FOR PATIENTS AND THEIR FAMILIES LIVING WITH AMYLOIDOSIS
ISSUE 2 2012

Correct diagnosis is vital

by Dick Wynveen (Victoria)

A few days after Christmas 2010, I was taking my grandchildren to a swimming pool when I suddenly experienced a very frightening sensation. I couldn't move and felt as though I was nailed to the ground. At the same time I had cramps in my stomach.

These feelings quickly passed and I felt fine for a week or so. Then it all happened again. Until that time I had been a fit, active 69 year old with my only real medical problem being high cholesterol for which I was on medication.

Over the Christmas period I had thoroughly enjoyed my food and put on four kilograms. I put my strange feelings down to my weight gain and set about watching my diet closely. Fairly quickly my weight dropped back and I felt well. However this did not last and I started to experience more days of feeling unwell, which gradually increased until there seemed to be no good days. I was constantly exhausted and everything became an effort.

My GP ordered an ECG (electrocardiogram) and cardiac ultrasound, and I was referred to a cardiologist who found that my heart muscle was considerably thickened. Following further tests, which still left my cardiologist questioning what was wrong, I was referred to the Alfred Hospital in Melbourne for a cardiac MRI but unfortunately had to wait two long months for this to be done.

The MRI confirmed that something was really wrong and I then underwent a biopsy of the heart. Amyloidosis was diagnosed in mid December. Following a bone marrow biopsy, the final diagnosis of AL amyloidosis was made. This was a great shock but I felt I just had to accept this and get on with the suggested treatment.

Early in 2012 I was referred back to Cabrini Hospital in Victoria where I underwent an array



Dick Wynveen

of tests to check whether my other organs had been affected by the amyloid deposition. No other organs appeared to be affected. While this seemed like good news to me, apparently it set off alarm bells for my haematologist. He then began to question the diagnosis. It was agreed that my biopsy slides should be sent to the Brisbane Amyloidosis Unit and my blood to the National Amyloidosis Centre in London for confirmation of the amyloidosis subtype.

Around the same time I started the stem cell mobilisation and was in the second day of collection when the procedure was stopped. My haematologist had received news from the two centres that I in fact had senile amyloidosis and not AL amyloidosis.

Although this was a big shock I was aware of how fortunate I was in having a haematologist who knew enough about amyloidosis to query the diagnosis when he found that I only had heart involvement. Senile amyloidosis is quite different from AL amyloidosis and if I had been treated with a stem cell transplant it would not have done me any good. In fact it may have caused me considerable problems.

continued on page 2

In this issue

Page 3
Free light chains

Page 5
Finding support

Page 6
The importance of
happiness

Page 11
News from around
Australia

From the editor



Welcome to our last edition of Amyloidosis News for the year. I hope you enjoy the two inspiring patient stories on pages one and four which illustrate how complicated this group of diseases is to diagnose, type and treat. The good news is that this is improving all the time but raising awareness is still as important as ever so that patients can be diagnosed as early as possible.

The articles also highlight how emotionally difficult the amyloidosis journey can be for patients and families, regardless of how strong they are. The positive side is that life with amyloidosis can be lived well and you are not alone. The Leukaemia Foundation exists to offer support.

On a personal note, this will be the last Amyloidosis News I will edit. Sheila Deuchars, Leukaemia Foundation Support Services Coordinator, has a strong interest in amyloidosis and will very competently take on this role with the ongoing help of the national amyloidosis patient editorial group. The Amyloidosis News will remain a national newsletter featuring information on all the types of amyloidosis.

My time with the Leukaemia Foundation as a volunteer working in the area of amyloidosis care has also come to an end. Since joining the Foundation in 2003 I have had the privilege of working with staff and patients to build education and support services across the country. In that time we have achieved a great deal, although I realise there is still a long way to go.

I personally will always be appreciative of the support I have received from the Foundation, particularly in

continued from page 1

It was obviously difficult for my haematologist to tell me that my original diagnosis was wrong and that I actually had a form of amyloidosis for which there was no active treatment. However the good news he gave me was that the prognosis for senile amyloidosis is often much better than for AL amyloidosis where there is heart involvement.

Because senile amyloidosis is usually not treated by a haematologist I was handed back to my cardiologist for him to concentrate on preserving my heart function. At first I was disappointed to think that there was no active treatment to stop the production of the amyloid protein depositing in my heart. But now I have come to accept it and am grateful I don't have to have chemotherapy or other treatments which may upset me and stop me from getting on with my life.

My cardiologist keeps a good eye on me and treats any fluid build-up I may experience with medication. In the course of all my investigations it was discovered that the drugs I had been taking to lower my cholesterol were actually causing the cramps in my stomach. An angiogram performed by my cardiologist also showed that my arteries were 100% clear. The tablets were immediately stopped with very favourable results.

Queensland, and from many medical professionals in Australia and overseas. I am also grateful to the hundreds of patients and families I have been privileged to know since I became involved in amyloidosis care in 1998 following the death of my husband. I have learnt so much from every one of you.

My sincere thanks go to everyone who has contributed to the Amyloidosis News during my time as editor.

I send you all my best wishes for a happy Christmas and a very peaceful 2013.

Pat Neely

Thank you Pat Neely

The Leukaemia Foundation would like to extend heartfelt thanks to Pat Neely for her years of volunteer service to the Foundation in the area of amyloidosis support. Since 2003 Pat has done a fantastic job in raising awareness of this little known group of diseases and has supported and cared for amyloidosis patients with a great deal of warmth and empathy. Her extensive knowledge of amyloidosis has been an asset, not only to our the organisation, but to the patients, families and medical professionals she has worked so closely with. Having lost her husband to amyloidosis in 1998, we understand that this is a cause close to Pat's heart and no doubt she will continue to be involved in spreading awareness about the disease and supporting patients and families. We wish Pat well.

I have learnt to live with the deficits caused by my damaged heart. Walking up a slope certainly slows me considerably. I know that I must get out of a chair or bed slowly to allow my heart to pump the blood around my body. I can do most things I used to do but in a slower fashion. My wife Connie and I walk together and we go away for a few days when we want to.

I am very positive in my outlook but realistic. Connie and I have been honest with each other and I have taught her all about the banking and how to program the video etc - jobs only I used to do! We now do them together.

Life remains very busy and I am as passionate as ever about the things I liked doing before my diagnosis such as building computers. Although I am very interested in any new treatments for senile amyloidosis, and would probably be the first to put myself forward if there was ever a trial in Australia, I often forget about my diagnosis and certainly don't live just in an amyloidosis world. My wife and I just enjoy each day and go away on holiday when we can.

New assay to measure free light chains

by Dr Peter Mollee

Measurement of serum free light chains is one of the cornerstones of the diagnosis and monitoring of AL amyloidosis. Until recently there was only one laboratory test that could measure serum free light chains and that was the Freelite® assay made by the Binding Site.

The Freelite® assay has undergone extensive testing in multiple countries and in thousands of patients with AL amyloidosis. This extensive testing led to the approval of the assay in most countries including Australia and to the incorporation of the important role of measuring serum free light chains in all current guidelines about AL amyloidosis. It also demonstrated that the Freelite® assay was not perfect and could occasionally give unusual results or “bounce around” even though a patient’s amyloid was quite stable.

A new test is now available called the N-Latex FLC assay® which is made by Siemens. Some laboratories in Australia have now introduced this new N-Latex FLC test and patients may notice that their free light chain results may vary with the introduction of this new test.

What do we know about this new N-Latex FLC assay?

To date not much has been published about this new assay in AL amyloidosis, but the amyloidosis centres in Italy, London and Brisbane have been examining it. The Italian centre presented their initial findings at the International Symposium on Amyloidosis in Groningen earlier this year. They concluded that the Binding Site and Siemens’ FLC assays have similar ability to diagnose AL amyloidosis and, when either

of these assays is used together with serum and urine protein immunofixation electrophoresis (other standard tests used in diagnosing all plasma cell diseases), 99% of AL amyloid cases were picked up. They also cautioned that the results of the two tests cannot be used interchangeably and that patients should have their AL amyloidosis monitored by the one assay all the time. The Amyloidosis Clinic at Brisbane’s Princess Alexandra Hospital, together with Pathology Queensland have also looked at the new N-Latex FLC® assay in 62 patients with AL amyloidosis and compared the results to the Freelite® assay. The results in diagnosis of AL amyloidosis were very similar to those findings by the Italian group. The use of the N-Latex FLC® assay to monitor patients’ response to treatment was also examined. While there were some differences between the two assays, both performed well in that response to treatment as measured by both assays predicted long-term outcome. These results remain preliminary and future work from amyloid centres will help to clarify the strengths and limitations of the new N-Latex FLC® assay.

What does this mean for patients with AL amyloidosis? Laboratories introducing the new assay are having a change over period where the results of both tests will be available. This will allow patients and their physicians to be able to see both results at once. Patients should not be alarmed when their free light chain result varies between the two tests. It remains very important to always have your blood and urine tests to monitor AL amyloidosis by the same assay in the same laboratory as results between different laboratories and between the Freelite® and N-Latex FLC® assays are not interchangeable.

Light the Night

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

Light the Night 2012 walks were held across the country recently to raise awareness of how diseases like amyloidosis affect our community as well as raise funds to invest in research to find the best possible treatments and cures.

Pictured right: Matt Teffer remembers his wife, Caryll, who passed away with amyloidosis last year. He was joined by daughter, Natasha, grandson Dylan; and son, Stuart.



Another chapter of my life began

by Allan Andrews

We all face problems in our lives and I am no different. It is perhaps how we cope with these problems that matters. None of us cope well all the time but my life philosophy has always been to try and live each day without hankering for the past and worrying too much about the future. I try to accept the bad things that happen and look for the good things. I believe this has helped me in my journey with amyloidosis.

In 2003 I received the news from my cardiologist that my heart was so damaged that without a heart transplant I would die. This stage of my life was very difficult. I had always been very fit, working and running marathons. I had always looked after my health so this seemed so unfair. My doctors were unable to fully explain why I had severe heart failure. I just had to get through each day hoping a heart would become available and in time it did! My phone rang at 1.32am, I phoned a taxi and another chapter of my life began.

As soon as I woke up from the surgery I felt the benefit of my new heart. My elation was dampened the following day when my transplant cardiologist gently told me that while the transplant had gone well, my heart failure was caused by an abnormal protein, amyloid, damaging my heart muscle and that I had a disease called amyloidosis – probably AL amyloidosis. I was also told that to try and ensure that this protein did not damage my new heart I would have to undergo a stem cell transplant when I was well enough.

I knew nothing about amyloidosis or stem cell transplants but I viewed this next procedure in a positive manner knowing that it had to be done to try and save my new heart and my life by stopping the production of the amyloid protein.

I left hospital five days after the heart transplant. As I was not to have the stem cell procedure for seven months I worked hard at becoming as fit as possible and quickly started to return to activities I enjoyed before I became ill. I was back playing bowls within four weeks and walking each day, feeling fitter all the time.

When I entered the hospital for my stem cell collection I felt I was as fit as I could be and ready for whatever was ahead. However this did prove to be quite a difficult time.

I was in hospital for four weeks and experienced many of the problems that can be associated with receiving the huge dose of chemotherapy before my stem cells were infused. An ulcerated mouth, loss of



Allan Andrews

weight and complete exhaustion made me feel very ill. But I tried to stick to my philosophy, taking one day at a time hoping that each day would bring me closer to leaving hospital with hopefully the amyloid protein under control.

Although I slowly began to feel better and my blood counts rose I began to lose my voice and needed surgery on my vocal cords. It was at this time that a haematologist filling in for my own doctor told me in front of my visitors that my prognosis was very poor. I was very angry at the rude way he delivered this news but instead of getting depressed I was determined to prove him wrong.

Again I was just getting my life back together again when I developed a very painful swelling around my left eye, which eventually burst. A diagnosis of nocardia was made, a dangerous infection which occurs mainly in people with weakened immune systems like me. I was rushed to intensive care where three lesions were found in my brain. Massive intravenous doses of antibiotics were given for two hours every day for three weeks. The treatment worked but I was left with my balance affected. Again this was another setback to overcome.

In early 2005 my light chains began to rise and I developed bruising on my face and around my eyes. The amyloid was on the move in spite of the stem cell transplant. My haematologist started me on Thalidomide which I remained on for 48 months. Although I went back into remission, the Thalidomide affected the nerves in my feet and hands (peripheral neuropathy) and consequently my balance got worse. I am now being supervised by an equilibrium specialist with good results.

My heart is no longer new and is functioning well. My AL amyloidosis seems to be in complete remission. I have recently undergone a successful knee replacement and after sticking diligently to my exercise program, I am now walking further each day and life is good.

Reflecting on the last nine years I realise how fortunate I was not to be diagnosed with amyloidosis

Finding support in your journey with amyloidosis

A diagnosis of amyloidosis may leave you feeling lonely, frightened and angry. At this time it may seem that no one has ever heard of this group of diseases and certainly will have little understanding of what you are going through. Sometimes it feels that even your doctors know little about amyloidosis.

Talking with others and learning more about your disease and treatment can ease the loneliness for some people. Ask your doctor or clinic nurse about the amyloidosis support services available in your area.

What does the Leukaemia Foundation offer?

The Leukaemia Foundation offers a range of practical support services which are offered free of charge to patients and their families. These include education, counselling, financial support, transportation and accommodation for rural and regional patients who need to move to a major city for treatment. In some areas the Foundation also runs specific amyloidosis support groups.

The Leukaemia Foundation has also produced a booklet, *Understanding Amyloidosis* and distributes a twice-yearly newsletter, *Amyloidosis News*. It also runs Amyloidosis telephone forums (see page 11). Please phone **1800 620 420** or go to www.lfq.org.au and find out how the the Leukaemia Foundation can support you through your journey with amyloidosis.



New website
www.lfq.org.au

The Leukaemia Foundation of Queensland has a new website which contains a great deal of information about amyloidosis and the range of support services available to patients and their families.

Talk Blood Cancer
www.talkbloodcancer.com

Although amyloidosis is not a blood cancer it does come under the umbrella of the Leukaemia Foundation and Talk Blood Cancer discussion forums. This is a safe place to discuss with others living with amyloidosis and post any questions you may have.

Other useful websites

The websites below may also be useful in helping you find information about your disease and linking you with others.

Amyloidosis Australia
www.amyloidosis.com.au

The Amyloidosis Foundation
www.amyloidosis.org

Amyloid Support Networks
www.amyloidosissupport.com

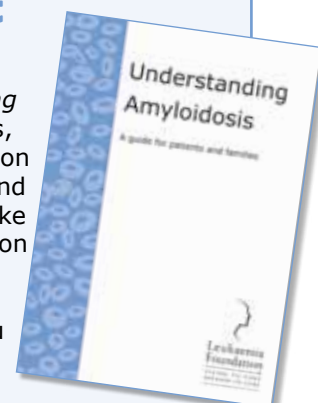
continued from page 4

before I underwent my heart transplant, as I probably would never have been approved for one. Perhaps for this reason I see the glass half full and not half empty. I am realistic about the fact that I have a chronic, maybe incurable disease. It is essential for me that I try to have good rapport with my doctors, ask questions and ensure that I fully understand my condition and treatments. I gratefully accept support from the Leukaemia Foundation staff when needed and I value highly the friendship and support I share with other patients I have met along the way.

I try not to live an "amyloidosis life" and rarely worry about tomorrow. I trust my doctors and do my part in keeping myself as fit as possible for whatever may come along. I enjoy my many friends and family and rejoice in each day.

Understanding Amyloidosis booklet

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 1800 620 420. You can download a copy from www.lfq.org.au.



The importance of happiness

by Neville Jones (Queensland)

A succession of medical incidents over a period of about five years involving three less-than-successful back operations, abdominal surgery and finally, a diagnosis of AL amyloidosis treated with chemotherapy and a stem cell transplant, left me feeling flat and less able to enjoy life than I previously had.

When diagnosed with a serious illness such as AL amyloidosis, it is understandable that a person's sense of well-being is seriously reduced. The treatments, including chemotherapy and stem cell transplant, are both physically demanding and psychologically stressful. Many people will have experienced feelings of anxiety and loss of control especially at diagnosis.

Meeting others through the Leukaemia Foundation of Queensland amyloidosis support meetings, many of whom have endured as much as I have and more, made me begin to realise that how I approached the ups and downs of my illness and treatment helped determine how well I would manage my life.

It then seemed to me that recent research and writing on the question of happiness was worthy of investigation. Below are some of the suggestions to be considered when you are trying to find some level of happiness again and take back control over your life.

- Social engagement which includes continuing your contacts and bonding with friends and acquaintances and developing new friendships;
- Develop a sense of community through activities such as volunteering;
- Humour can be a means of bonding and sharing. Laughter can often be the best medicine;
- Do random acts of kindness which not only have benefits for the recipient but also bring a feeling of wellbeing to the giver;
- Undertaking a hobby, sport or involvement in creative activities can have clear positive outcomes;
- Realise that despite the difficulties being experienced, life still has meaning and purpose;
- Frame issues in a positive manner – the glass is half full not half empty;
- Be less self-focused by looking out towards other people and being receptive to new ideas and experiences;



- Be grateful for kindnesses received and appreciate others;
- Count your blessings - you could be living in a war torn country or working in a factory in a polluted mega-city;
- Talk with your GP, haematologist, counsellor or psychologist if you are experiencing lasting feelings of sadness, depression or anxiety;
- Share your feelings with those nearest to you or talk with a friend.

To enhance wellbeing it is necessary to utilise your personal strengths and apply them in your activities. Where health permits, taking on even small challenges and enhancing your skills can help promote strength and cheerfulness.

Many people obtain great encouragement through religion, philosophy and spiritualism. These can provide a source of meaning and also contribute to happiness. Undertaking voluntary work can also be a source of encouragement.

Meditation can be good for the soul and appreciation of the goodness in others. The Dalai Lama has written in *The Art of Happiness* that, "peace of mind or a state of calm is rooted in affection and compassion. If you possess this inner quality then, even if you lack various external facilities that you would normally consider necessary for happiness, it is still possible to live a happy and joyful life."

Many of these thoughts are applicable to life in general, and not just to those afflicted with a disease such as amyloidosis. It is important therefore to "smell the roses" and "close one's eyes and listen to the music." Notwithstanding the rigours of diagnosis and treatment it is possible to flourish and for life to become more meaningful.

A book that I have found very helpful was "The Discovery of Happiness" by Stuart McCready (editor), Source Books, 2001.

Q & A

What is senile amyloidosis and why have I been told that the prognosis is better than symptomatic cardiac AL amyloidosis?

Senile amyloidosis is a disease caused by extensive deposits of the normal (wild-type) protein transthyretin (TTR) in the heart. However this disease is not inherited like hereditary amyloidosis where the most common types are due to a transthyretin gene mutation.

AL amyloidosis is caused by an abnormal protein (the "light chain" of an immunoglobulin or antibody protein) made by abnormal plasma cells found in the bone marrow.

Senile amyloidosis occurs predominantly in men over the age of 80 and is exceptionally rare below 60, whereas in AL amyloidosis with heart involvement there is a 60-40 male to female ratio with patients usually being over the age of 50 but sometimes younger.

In senile amyloidosis only the heart is usually affected although some patients may also experience carpal tunnel syndrome often many years before the cardiac involvement.

In AL amyloidosis the amyloid protein can deposit in any organs or tissues of the body disrupting function. In both senile amyloidosis and (light chain) AL amyloidosis with heart involvement patients experience an infiltrating cardiomyopathy but on echocardiograms the ventricles are found to be thicker in senile amyloidosis.

It is essential that a correct diagnosis is made as treatment for these two diseases is different. AL amyloidosis is treated with chemotherapy and in some cases with a stem cell transplant but senile amyloidosis has no treatment available to stop the amyloid being produced and depositing in the heart.

However in spite of this lack of treatment the prognosis for patients with senile amyloidosis appears to often be better than for those with AL amyloidosis affecting the heart.

It seems that the two main reasons for this are:

- the slow deposition of amyloid in senile amyloidosis
- the absence of other major organ involvement.



Other factors that may also make a difference include:

- The different protein composition of the amyloid fibrils in AL amyloidosis and senile amyloidosis.
- The mechanism of infiltration of the amyloid within the heart which may cause worse heart failure in AL patients.
- The monoclonal light chains circulating in the blood may have a toxic property in AL amyloidosis.

Does it matter that when stem cells are collected some "baddies" are also then put back into you later?

The concern expressed in this question is that the contaminating plasma cells will be reinfused, engraft in the marrow and start producing amyloid again.

In short, we have no answer to this question in AL amyloidosis. What we know is learnt from myeloma where attempts have been made in the past to "purge" stem cell collections of myeloma cells. These efforts have failed for a variety of reasons but, perhaps most importantly, a clinical trial of "purged" (=CD34 selected) stem cells versus "non-purged" stem cells showed that the "purged" transplants had a worse outcome, in particular, with more infectious complications.

Also, in animal models, relapse in myeloma seems to come mostly from plasma cells left over in the body rather than from plasma cells reinfused in the stem cell collection. So, yes, stem cell collections in AL patients may contain small amounts of light chain producing plasma cells, but this does not seem to be a clinically concerning problem.

Patients from Australian amyloidosis clinics - a snapshot

There is no register to record the numbers and types of amyloidosis patients diagnosed each year in Australia nor the types and results from treatment. The information below is therefore useful in providing a snapshot of numbers, types and treatment results from the two designated amyloidosis centre in Australia.

Following is information from a retrospective cohort analysis of patients seen at two Australian amyloidosis clinics, the Westmead Hospital Amyloidosis clinic in Sydney and the Princess Alexandra Hospital Amyloidosis Clinic in Brisbane.

These two centres offer diagnostic sub-typing and management advice for patients with all types of amyloidosis.

This analysis of the characteristics, management and outcomes of 123 patients who were referred to these two clinics up to November 2011 was presented at the XIII International Symposium on Amyloidosis in Groningen, the Netherlands, in May this year.

The types of amyloidosis were broken down into:	
Systemic Light chain (AL) amyloidosis	68 patients
Localised AL amyloidosis	16 patients
Familial/hereditary amyloidosis	14 patients
Senile amyloidosis (SSA)	11 patients
AA amyloidosis	11 patients
Indeterminate	3 patients

SYSTEMIC AL AMYLOIDOSIS

Sixty patients had systemic AL amyloidosis. Six of the 60 also had multiple myeloma and three of the 60 had a lymphoproliferative neoplasm.

The most common precursor light chain type was lambda and this was detected in 36% on blood testing with the 'serum free light chain assay' and in 28% in the urine as a lambda Bence Jones protein. One patient had no detectable precursor AL protein on blood, urine or bone marrow testing.

Organ involvement

On average two organs were involved with a range of one to seven organs involved. The most common organs involved were kidney 76%, heart 59% and nerves 37%.

Treatment

Cyclophosphamide, dexamethazone and thalidomide (CTD) was the most preferred treatment regimen after diagnosis to suppress the production of the amyloid protein. CTD was followed closely by Melphalan and dexamethasone. Nine of the 68 AL patients underwent a stem cell transplant.

Outcomes

56% of AL patients attained a haematological response after a median of 5 months. 30% achieved

an organ response after a median of 6 months. The median overall survival for AL patients using Kaplan Meier analysis was 66 months.

LOCALISED AMYLOIDOSIS

In the 16 patients analysed, the most commonly affected organs were the respiratory tract (nine patients), gastrointestinal tract (four patients), bladder (two patients) and skin (two patients).

FAMILIAL/HEREDITARY AMYLOIDOSIS

Fourteen patients belonged to 10 families. Three patients were the first to be identified as having inherited amyloidosis in their families.

The gene mutation type included TTR (seven patients), lysosome (two patients) and fibrinogen (one patient).

Gene mutations in the TTR (transthyretin) gene region were the most common type of inherited mutations. Inherited TTR patients had the nerves followed by the heart as the most common organs involved. For treatment, one patient with inherited TTR has been prescribed Diflunisal and one patient with inherited TTR amyloidosis is on the liver transplantation waiting list.

SENILE AMYLOIDOSIS

All 11 patients analysed were male and the median age at diagnosis was 67 years. The most common organ involvement was the heart (all 11 patients) with four patients having multi-organ disease. The drug Diflunisal was prescribed to one patient with SSA.

AA AMYLOIDOSIS

Of the 11 patients analysed the most common organ involvement was kidney (all 11 patients) with 45% requiring dialysis. Four patients had multi-organ involvement.

The majority had autoimmune diseases as the inflammatory disorder that created the amyloidogenic AA precursor protein. The most common causative disease was rheumatoid arthritis. In two patients no inflammatory disease could be found.

Conclusion

This cohort of Australian amyloidosis clinic patients reflects the recognised epidemiology and heterogeneity of the amyloidosis disorders.

Reference: A Cohort Analysis of Patients from Australian Amyloidosis Clinics by Fiona Kwok, Ming-Wei Lin, Graeme Stewart, David Booth, Nikky Isbel, Dariusz Korczyk, Sally Mapp, Patricia Renaut, John Taper, Amanda Johnston, Richard Phoon, Liza Thomas, Ragwha Sharma, Bartłomiej Getta, Steve Vucic, Peter Mollee David Gottlieb. Presented as a poster at the XIII International Symposium on Amyloidosis, Groningen, The Netherlands, May, 2012



Patients benefit from subsidy increase

The Leukaemia Foundation of Queensland has welcomed Queensland Health's announcement to increase the Patient Travel Subsidy Scheme (PTSS) for Queenslanders living in rural and remote areas.

From 1 January 2013 the PTSS will double from \$30 to \$60 per night per person and the mileage subsidy from 15 cents to 30 cents per kilometre.

It is the first time since 1987 that the accommodation subsidy has been increased.

The PTSS provides financial support for public and private patients to access specialist health services that are more than 50km away from their closest public hospital, with patients from regional, rural and remote locations the main users of the Scheme.

Chief Executive Officer of the Leukaemia Foundation of Queensland, Bill Petch, praised the initiative saying, "this much needed financial boost will allow the Foundation to alleviate accommodation costs and ultimately allow the Foundation to further invest funds into research, finding cures and better treatments for patients as well as providing a wider range of support services during and after treatment."

The patient transport system varies slightly in each state. Patients not living in Queensland should contact the Health Department in their state to find information on patient travel subsidies.

Research into anxiety and depression among AL amyloidosis patients: the role of cardiac symptoms

A research study has been conducted to gather data on the impact of diagnosis on AL patients. The study specifically looked at the role that symptoms, particularly cardiac symptoms, play in affecting anxiety and depression among patients.

The results of the study were published in the September 2012 edition of *Journal of Protein Folding Disorders "Amyloid"* in a paper entitled, "Anxiety and Depression among AL patients: the role of cardiac symptoms".

The results of the study concluded that although psychological support should be offered to all AL amyloidosis patients during the course of the disease, it seems to be crucial at cardiac symptom onset more than at diagnosis communication, in order to assist patients to accept AL disease when it becomes visible and present in their daily lives. Moreover, psychological support should be intensified with the worsening of cardiac symptoms.

Although AL patients have the worst prognosis, no data was available about the incidence of the disease-related psychological impact of this illness. Most studies have been conducted on hereditary forms of amyloidosis. These investigations which have focused on advanced disease stages have analysed the impact of liver transplantation on anxiety and depression among hereditary amyloidosis patients.

AL amyloidosis represents the most common type of systemic amyloidosis and cardiac involvement is seen in over half of patients at diagnosis. One of the main determinants of prognosis is cardiac involvement and among the different forms of cardiac amyloidosis (hereditary or acquired), AL amyloidosis has the worst prognosis. In addition to affecting the prognosis, cardiac symptoms also impact a person's daily activities.

The aim of the study was to evaluate the role of time that had passed since the diagnosis was communicated, the time that had passed since the onset of cardiac symptoms, and the effect that cardiac symptom severity has on levels of anxiety, depression and psychological stress among cardiology patients with AL.

Thirty-two AL amyloidosis patients (15 males) aged 45-83 (mean age 66) participated in the study. On average AL amyloidosis had been diagnosed 20 months prior to the patients' participation. The mean diagnosis for the onset of cardiac symptoms was 12 months prior to the patients' participation in the research. (Diagnosis was mainly made eight months prior to the onset of cardiac symptoms.)

The present study provides useful information for the organisation of psychological support programs for AL amyloidosis patients since our results suggest that the timing of the intervention is important.

The study was conducted by Martina Smorti, Faculty of Education, Free University of Bolzano, Bressanone, Italy; Francesco Cappelli, Heart and Vessel Department, Intensive Cardiac Care Unit, AOU Careggi, Florence, Italy; and Franco Bergesio and Federico Perfetto from the Tuscan Regional Amyloid Centre, AOU Careggi, Florence, Italy.

Although the authors acknowledge this study has a number of limitations and further longitudinal studies with an increased number of subjects are needed to better understand how psychological aspects change over time, they hope this study provides useful information for the organisation of psychological support programs for AL amyloidosis patients.

Making the most of your doctor's appointments

The diagnosis of a life-threatening disease such as amyloidosis can leave you in shock and unable to think properly. However good your doctors may be at communicating, many patients say they have difficulty retaining any information given, except perhaps how serious amyloidosis is.

If you or your family wishes to make informed decisions about treatment, you need to have the facts. Patients vary in how much they want to know and what they can understand. Some patients want to know as much as possible about their disease from the beginning and will ask many questions. Other people would rather not know very much at first or are overwhelmed when they meet their doctor and are unable to ask questions.

In the course of the diagnosis and assessment of your disease you may see a number of specialists. They will try to be sensitive to your needs and give the information they perceive you want. It is often a good idea to take your partner or a friend to your appointments. Some people like to tape the consultation. It is wise to ask the doctor whether he/she is happy with this. Always carry a pen and paper to make notes.

Think about and write down your questions before your doctor's appointment. The mind tends to go blank when we enter the doctor's consulting room.

A diagnosis of amyloidosis means that you will be quickly learning a new vocabulary. Asking the questions that are relevant to you can help you understand these new terms and build a better understanding between you and your doctor.

Below are some suggested questions that you may want to ask your doctor. Obviously you may not want to ask all these questions at one time and the relevant questions will change as you move through your treatment.

The disease

- What is amyloidosis?
- What type of amyloidosis do I have?
- How serious is this disease?
- How can I learn more about my disease? Do you have any written material I can read?
- Are there many other people you know with this disease?
- Are there any support groups or people I could talk with?

Treatment program

- What exactly is the treatment?
- What are the objectives of treatment?



- Over what period would treatment be given?
- How will the treatment be given?
- How often would I have to visit hospital?
- Will I have to stay in hospital?
- Will I be able to work or look after my children during treatment?
- How do people usually feel during treatment?
- How long would the treatment last?
- How long would I take to get over it?
- What will happen after the treatment is finished?
- Why have you chosen this treatment for me?
- Are there any costs attached to the drugs recommended for my treatment?
- What happens if this treatment does not work?

Past experience

- How many patients have you treated with this treatment regime?
- How much experience is there with this treatment in Australia and around the world?
- What is the likelihood of achieving a complete or partial remission?
- How long have other people remained in remission?
- In the event of the disease coming back, would there be other treatments I could have?
- What factors influence outcomes?
- If I should develop any pain, nausea or other problems through the treatment would there be medicines to help me?
- How will you know whether the treatment is working?

Side effects

- What side effects do people usually get on the treatment you have suggested?
- When would I begin to experience any side effects?
- Could any side effects be life threatening or cause pain and permanent damage?
- Will I be offered treatment for any side effects?

Alternatives

- What are the alternatives to the treatment you are recommending?
- What would be the good and bad things about the alternative treatment?
- How affective might the alternative treatment be for me?

News from around Australia

Queensland

Our new ESA Village at Dutton Park is fully occupied and operational and was officially opened on 27 October by Queensland Governor Penelope Wensley AC. We held two amyloidosis support luncheons in July and November at the new village which were well attended.

A satisfaction questionnaire was completed at the July meeting and the results were mailed out to our Queensland residents. The Leukaemia Foundation of Queensland anticipates that the results will guide our support meetings for the future, ensuring that we are continuing to meet the needs of our patients and families.

Thank you to the amyloidosis patients and families who attended *Light the Night* walks around Queensland. I think you will all agree it was a spectacular night and was a great opportunity to raise awareness of amyloidosis and show support for those affected by amyloidosis and related blood disorders.

The Leukaemia Foundation of Queensland has recently launched its new website, www.lfq.org.au, which contains a great deal of disease information, including amyloidosis. It also provides comprehensive information on available support services, fundraising events and research information.

In 2013 I will take over from Pat Neely as editor of *Amyloidosis News*, so if you would like to share your story or have any ideas for articles please let me know. I will also continue to work closely with patients and their families. Please call me on 07 3252 2277 if you would like to chat or ask any questions.

Sheila Deuchars
Support Services Coordinator

South Australia

The Leukaemia Foundation in South Australia and Northern Territory will again be hosting its major patient education day on 22 June, 2013. While the program and speakers are yet to be finalised, the theme of the day will be "Emergence of personalised medications - targeted therapies." The amyloidosis session will be facilitated by Amyloidosis Patient and Family Advocate, Pat Neely.

Louise Bastian
Support Services Manager

Victoria and Tasmania

The Annual Victorian and Tasmanian patient conference was held in Melbourne on Saturday 6 October. With an ever increasing interest in this event, the decision was made to offer a number of new disease specific streams including a morning dedicated to amyloidosis. The invitation to the event was extended across a number of states and we were pleased to welcome 12 attendees to the program.

The organising committee was thrilled to invite Dr Peter Mollee from the Amyloidosis Centre at the Princess Alexandra Hospital in Brisbane. Dr Mollee provided an extensive overview of the different types of amyloidosis along with a review of treatment options, novel therapies and the importance of good supportive care for patients and their loved ones. The response from attendees was very positive with benefits including a greater understanding of what they were dealing with and also a great sense of peer support from others in the room. As a result there are ongoing plans to extend our support options for this group of patients.

A total of 447 patients and carers attended this year's event and plans will commence very early in 2013 to ensure that the next event is just as well received.

Sara Andrews
Support Services Manager

Amyloidosis telephone forums

The Leukaemia Foundation's amyloidosis telephone forums are designed to bring together people with amyloidosis in the comfort of their own home. If you live in a regional area or are unable to travel to amyloidosis luncheons, you might not meet others with amyloidosis. The forums can offer you that opportunity and ability to discuss topics related to your disease as well as share experiences and tips. A very informative and interactive forum was held on 7 November with guest speaker, Professor Miles Prince sharing with patients and carers how amyloidosis may affect different organs, the way the disease is being currently treated and possible emerging treatments in the future. The challenges and future areas of research were also explored. Professor Prince is Professor of Medicine at both Melbourne and Monash Universities and Director of the Centre for Blood Cell Therapies at the Peter MacCallum Cancer Centre, Melbourne and consultant haematologist at Cabrini Hospital, Melbourne. Forum participants were also invited to share their stories and experiences. If you would like information on forums for 2013 please contact Kaye Hose on 03 98636951 or email myeloma@leukaemia.org.au

2013 Amyloidosis Queensland support luncheons

Tuesday 19 February, 21 May,
27 August and 26 November

Amyloidosis T-Shirts

On sale for only \$5

Why not buy an amyloidosis t-shirt to help raise awareness. We now only have smaller sizes (extra small, small) available. Contact Sheila Deuchars on 3252 2277.



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**Leukaemia
Foundation's**
*Christmas
Appeal*

Donate online at
www.lfqappeals.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

**GPO Box 9954
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 1800 620 420